Synopsis

International digest of children’s palliative care research abstracts

Editors: Dr Satbir Singh Jassal, Dr Susie Lapwood, Dr Linda Maynard, Sue Langley and Lizzie Chambers
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Together for Short Lives is a UK wide charity that, together with our members, speaks out for all children and young people who are expected to have short lives. Together with everyone who provides care and support to these children and families we are here to help them have as fulfilling lives as possible and the very best care at the end of life. We can’t change the diagnosis, but we can help children and families make the most of their time together.

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East Anglia’s Children’s Hospices (EACH) – New library and information service for children’s hospices

For staff working in children’s hospices it’s not always easy to access the most up-to-date resources to support professional and evidence based practice. However, the good news is that a new online library service is now available to all children’s hospices via the EACH library service.

The library has more than 3,000 resources, specialising in all aspects of caring for children and their families with life-limiting conditions. Library services and information can be provided over the telephone, by post or via email.

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For more details about the long distance service visit www.each.org.uk/library

For information on membership, please contact: Sue Langley, Library and Information Services Manager sue.langley@each.org.uk; 01223 815103
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Selected abstracts: July 2017 – February 2018

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Selected abstracts from 1st July 2017 to 28th February 2018
Clinical and Ethical Decision Making


Whether children should be offered genetic testing for cancer risk is much debated but young voices are rarely heard in these conversations. The current study explored perspectives of genetic testing held by adolescents and emerging adults in families with Li-Fraumeni syndrome (LFS). Twelve 12-to 25-year-olds in families with LFS completed qualitative interviews for this study. All believed that testing should be offered for children but many qualified this statement saying parental approval would be needed and testing should be optional. Genetic testing was seen as way to learn of risk status, allow for disease prevention efforts, and reduce uncertainty and anxiety. Perceived disadvantages included negative emotions associated with the testing result. Participants generally felt that children should be involved in the testing decision, but that parents could unilaterally decide to have a child tested in certain circumstances (e.g., young age, high risk). All who were aware of having been tested and of their test result (n = 7; 4 positive) said testing had no negative impact on their outlook and they agreed with the decision to undergo testing. Implications of these findings for clinical practice and future research are discussed.


We will examine several ethical considerations in the resuscitation of infants born at the margin of gestational viability in analyzing a case of preterm labor. More specifically, we will discuss the obligations of physicians in characterizing expected outcomes, both mortality and long-term morbidity, for extremely premature infants and how potential adverse outcomes should be framed-as complications of prematurity itself or as iatrogenic complications of care. We will also explore how the concept of a “trial of therapy” can support parents and neonatologists in decision making concerning withholding or withdrawing care for periviable infants.


This document is the result of previous work carried out by different expert groups and submitted to multidisciplinary debate at a Conference about controversial, deficient, or new aspects in the field of neonatal palliative care, such as: 1) the deliberative decision-making process, 2) hospital and domiciliary palliative care, 3) donation after controlled cardiac death, and 4) moral stress in professionals. The most relevant conclusions were: the need to instruct professionals in bioethics and in the deliberative method to facilitate thorough and reasonable decision-making; the lack of development in the field of perinatal palliative care and domiciliary palliative care in hospitals that attend newborns; the need to provide neonatal units with resources that help train professionals in communication skills and in the management of moral distress, as well as delineate operational procedure and guidelines for neonatal organ donation.


The purpose of this study was to explore the experience of decision making in the care of children with palliative care needs in Jordan, from the perspective of their mothers. This study employed a collective qualitative case study approach. Data were collected in 3 pediatric wards in a Jordanian hospital. The study used 2 data collection methods: participant observation (197 observational hours) and 56 semi-structured interviews with 24 mothers,
12 physicians and 20 nurses. The findings show how Jordanian mothers seek to transfer the role of decision making to physicians, as they perceive themselves to be unable to make decisions about critical issues related to the treatment of their children. Mothers had a widespread apprehension of “future guilt,” especially when they feared that any decisions they might make could have an adverse impact on their children. Contrary to the predominant pattern, some mothers took a proactive approach towards decision making about their children’s treatment. These mothers requested detailed information from primary physicians and sought different sources of knowledge such as second opinions, reading online resources, or talking to other parents who had a child with similar circumstances. The study concludes that mothers prefer to involve physicians in decisions about their children’s healthcare and treatment to eliminate their fear of probable future guilt; this modifies any tendency to autonomously decide for their children. These findings are underpinned by the Jordanian culture in which doctors’ opinions are highly regarded.


As more countries adopt laws and regulations concerning euthanasia, pediatric euthanasia has become an important topic of discussion. Conceptions of what constitutes harm to patients are fluid and highly dependent on a myriad of factors including, but not limited to, health care ethics, family values, and cultural context. Euthanasia could be viewed as iatrogenic insofar as it results in an outcome (death) that some might consider inherently negative. However, this perspective fails to acknowledge that death, the outcome of euthanasia, is not an inadvertent or preventable complication but rather the goal of the medical intervention. Conversely, the refusal to engage in the practice of euthanasia might be conceived as iatrogenic as it might inadvertently prolong patient suffering. This article will explore cultural and social factors informing families’, health care professionals’, and society’s views on pediatric euthanasia in selected countries.


BACKGROUND: Early engagement in advance care planning (ACP) is seen as fundamental for ensuring the highest standard of care for children and young people with a life-limiting condition (LLC). However, most families have little knowledge or experience of ACP. OBJECTIVE: To investigate how parents of children and young people with LLCs approach and experience ACP. METHODS: Open-ended, semi-structured interviews were conducted with parents of 18 children; nine children who were currently receiving palliative care services, and nine children who had received palliative care and died. Verbatim transcripts of audiotaped interviews were analysed following principles of grounded theory while acknowledging the use of deductive strategies, taking account of both the child’s condition, and the timing and nature of decisions made. RESULTS: Parents reported having discussions and making decisions about the place of care, place of death and the limitation of treatment. Most decisions were made relatively late in the illness and by parents who wished to keep their options open. Parents reported different levels of involvement in a range of decisions; many wished to be involved in decision making but did not always feel able to do so. DISCUSSION: This study highlights that parents’ approaches to decision making vary by the type of decision required. Their views may change over time, and it is important to allow them to keep their options open. We recommend that clinicians have regular discussions over the course of the illness in an effort to understand parents’ approaches to particular decisions rather than to drive to closure prematurely.


OBJECTIVE: In English paediatric practice, English law requires that parents and clinicians agree the ‘best interests’ of children and, if this is not possible, that the courts decide. Court intervention is rare and the concept of best interests is ambiguous. We report qualitative research exploring how the best interests standard operates in practice, particularly with decisions related to planned non-treatment. We discuss results in the light of accounts
of best interests in the medical ethics literature. DESIGN: We conducted 39 qualitative interviews, exploring decision making in the paediatric intensive care unit, with doctors, nurses, clinical ethics committee members and parents whose children had a range of health outcomes. Interviews were audio-recorded and analysed thematically. RESULTS: Parents and clinicians indicated differences in their approaches to deciding the child’s best interests. These were reconciled when parents responded positively to clinicians’ efforts to help parents agree with the clinicians’ view of the child’s best interests. Notably, protracted disagreements about a child’s best interests in non-treatment decisions were resolved when parents’ views were affected by witnessing their child’s physical deterioration. Negotiation was the norm and clinicians believed avoiding the courts was desirable. CONCLUSIONS: Sensitivity to the long-term interests of parents of children with life-limiting conditions is defensible but must be exercised proportionately. Current approaches emphasise negotiation but offer few alternatives when decisions are at an impasse. In such situations, the instrumental role played by a child’s deterioration and avoidance of the courts risks giving insufficient weight to the child’s interests. New approaches to decision making are needed.


AIDS has devastated communities across southern Africa, leaving many children orphaned. Grandmothers are considered ideal caregivers because of cultural expectations of intergenerational care, and because they have not been decimated by AIDS to the same extent as younger adults. However, these grandmothers, who currently carry the majority of the burden of care for AIDS orphans, are themselves aging and dying. I argue here that in Lesotho, the caregiving demanded of grandmothers late into their lives not only alters kin relations for the living but has increasingly made a “good” death unachievable for elderly caregivers.


OBJECTIVE: In the paediatric intensive care unit (PICU), medication administration is challenging. Empirical studies demonstrate that interruptions occur frequently and that nurses are fundamental in the delivery of medication. However, little is known about nurse’s decision making when interrupted during medication administration. Therefore, the objective is to understand decision making when interrupted during medication administration within the PICU. RESEARCH DESIGN: A qualitative study incorporating non-participant observation and audio recorded semi-structured interviews. A convenience sample of ten PICU nurses were interviewed. Each interview schedule was informed by two hours of observation which involved a further 29 PICU nurses. Data was analysed using Framework Analysis. SETTING: A regional PICU located in a university teaching hospital in the United Kingdom. FINDINGS: Analysis resulted in four overarching themes: (1) Guiding the medication process, (2) Concentration, focus and awareness, (3) Influences on interruptions (4) Impact and recovery CONCLUSION: Medication administration within the PICU is an essential but complex activity. Interruptions can impact on focus and concentration which can contribute to patient harm. Decision making by PICU nurses is influenced by interruption awareness, fluctuating levels of concentration, and responding to critically ill patient and families’ needs.


BACKGROUND: In the last 20 years, the chances for intact survival for extremely preterm infants have increased in high income countries. Decisions about withholding or withdrawing intensive care remain a major challenge in infants born at the limits of viability. Shared decision-making regarding these fragile infants between health care professionals and parents has become the preferred model today. However, there is an ongoing ethical debate on how decisions regarding life-sustaining treatment should be reached and who should have the final word
when health care professionals and parents do not agree. We designed a survey among neonatologists and neonatal nurses to analyze practices, difficulties and parental involvement in end-of-life decisions for extremely preterm infants. METHODS: All 552 physicians and nurses with at least 12 months work experience in level III neonatal intensive care units (NICU) in Switzerland were invited to participate in an online survey with 50 questions. Differences between neonatologists and NICU nurses and between language regions were explored. RESULTS: Ninety six of 121 (79%) physicians and 302 of 431(70%) nurses completed the online questionnaire. The following difficulties with end-of-life decision-making were reported more frequently by nurses than physicians: insufficient time for decision-making, legal constraints and lack of consistent unit policies. Nurses also mentioned a lack of solidarity in our society and shortage of services for disabled more often than physicians. In the context of limiting intensive care in selected circumstances, nurses considered withholding tube feedings and respiratory support less acceptable than physicians. Nurses were more reluctant to give parents full authority to decide on the course of action for their infant. In contrast to professional category (nurse or physician), language region, professional experience and religion had little influence if any on the answers given. CONCLUSIONS: Physicians and nurses differ in many aspects of how and by whom end-of-life decisions should be made in extremely preterm infants. The divergences between nurses and physicians may be due to differences in ethics education, varying focus in patient care and direct exposure to the patients. Acknowledging these differences is important to avoid potential conflicts within the neonatal team but also with parents in the process of end-of-life decision-making in preterm infants born at the limits of viability.


In most children's hospitals, there are very few ethics consultations, even though there are many ethically complex cases. We hypothesize that the reason for this may be that hospitals develop different mechanisms to address ethical issues and that many of these mechanisms are closer in spirit to the goals of the pioneers of clinical ethics than is the mechanism of a formal ethics consultation. To show how this is true, we first review the history of collaboration between philosophers and physicians about clinical dilemmas. Then, as a case-study, we describe the different venues that have developed at one children's hospital to address ethical issues. At our hospital, there are nine different venues in which ethical issues are regularly and explicitly addressed. They are (1) ethics committee meetings, (2) Nursing Ethics Forum, (3) ethics Brown Bag workshops, (4) PICU ethics rounds, (5) Grand Rounds, (6) NICU Comprehensive Care Rounds, (7) Palliative Care Team (PaCT) case conferences, (8) multidisciplinary consults in Fetal Health Center, and (9) ethics consultations. In our hospital, ethics consultants account for only a tiny percentage of ethics discussions. We suspect that most hospitals have multiple and varied venues for ethics discussions. We hope this case study will stimulate research in other hospitals analyzing the various ways in which ethicists and ethics committees can build an ethical environment in hospitals. Such research might suggest that ethicists need to develop a different set of "core competencies" than the ones that are needed to do ethics consultations. Instead, they should focus on their skills in creating multiple "moral spaces" in which regular and ongoing discussion of ethical issues would take place. A successful ethicist would empower everyone in the hospital to speak up about the values that they believe are central to respectful, collaborative practice and patient care. Such a role is closer to what the first hospital philosophers set out to do than in the role of the typical hospital ethics consultant today.


When baby Charlie Gard was diagnosed with a rare mitochondrial disease, his parents located a Professor of Neurology in the USA willing to provide nucleoside therapy which offered a theoretical chance of improvement and successfully raised pound1.3 million through crowd funding. The decision that unproven therapy was contrary to Charlie Gard’s best interests and that life-sustaining treatment should be withdrawn was devastating for his parents and difficult for their supporters to comprehend. The decision was upheld at three levels of appeal and Charlie died in July 2017 aged 11 months. This commentary provides a critical analysis of the legal principles surrounding unproven treatment and application of the best interests test in the different contexts of hospital and court. It draws attention to conflicting guidance and explores differences in approach in relation to unproven treatment for adults lacking capacity and children.

BACKGROUND: The world literature shows that empirical research regarding the process of decision-making when cancer in adolescents is no longer curable has been conducted in High-income, English speaking countries. The objective of the current study was to explore in-depth and to explain the decision-making process from the perspective of Mexican oncologists, parents, and affected adolescents and to identify the ethical principles that guide such decision-making. METHODS: Purposive, qualitative design based on individual, fact-to-face, semi-structured, in-depth interviews. The participants were thirteen paediatric oncologists, 13 parents or primary carers, and six adolescents with incurable cancer. The participants were recruited from the paediatric oncology services of three national tertiary-care medical centres in Mexico City. RESULTS: The oncologists stated that they broach the subject of palliative management when they have determined that curative treatment has failed. Respect for autonomy was understood as the assent of the parent/adolescent to what the oncologist determined to be in the best interest of the adolescent. The oncologists thought that the adolescent should be involved in the decision-making. They also identified the ability to count on a palliative care clinic or service as an urgent need. For the parents, it was essential that the oncologist be truly interested in their adolescent child. The parents did not consider it necessary to inform the child about impending death. The adolescents stated that the honesty of their oncologists was important; however, several of them opted for a passive role in the decision-making process. CONCLUSION: The findings of this study evidence that to achieve good medical practice in low-middle income countries, like Mexico, it is urgent to begin effective implementation of palliative care, together with appropriate training and continuing education in the ethics of clinical practice.


There are more migrants, refugees, and immigrants adrift in the world today than at any time in the recent past. Doctors and hospitals must care for people from many different cultures, countries, and religious backgrounds. We sometimes find our own deeply held beliefs and values challenged. In this “Ethics Rounds,” we present a case in which a Pakistani immigrant family faces a tragic medical situation and wants to deal with it in ways that might be normative in their own culture but are aberrant in ours. We asked the American doctors and 2 Pakistani health professionals to think through the issues. We also invited the father to talk about his own experience and preferences. We conclude that strict adherence to Western ethical norms may not always be the best choice. Instead, an approach based on cultural humility may often allow people on both sides of a cultural divide to learn from one another.


Euthanasia was first legalised in the Netherlands in 2002, followed by similar legislation in Belgium the same year. Since the beginning, however, only the Netherlands included the possibility for minors older than 12 years to request euthanasia. In 2014, the Belgian Act legalising euthanasia was amended to include requests by minors who possess the capacity of discernment. This amendment sparked great debate, and raised difficult ethical questions about when and how a minor can be deemed competent. We conducted a systematic review of argument-based literature on euthanasia in minors. The search process followed PRISMA guidelines. Thirteen publications were included. The four-principle approach of medical ethics was used to organise the ethical arguments underlying this debate. The justification for allowing euthanasia in minors is buttressed mostly by the principles of beneficence and respect for autonomy. Somewhat paradoxically, both principles are also used in the literature to argue against the extension of legislation to minors. Opponents of euthanasia generally rely on the principle of non-maleficence. CONCLUSION: The present analysis reveals that the debate surrounding euthanasia in minors is at an early stage. In order to allow a more in-depth ethical discussion, we suggest enriching the four-principle approach by including a care-ethics approach. What is Known: * The Netherlands and Belgium are the

https://academic.oup.com/medlaw/advance-article-abstract/doi/10.../medlaw/.../4748889
only two countries in the world with euthanasia legislation making it possible for minors to receive euthanasia. * This legislation provoked great debate globally, with ethical arguments for and against this legislation. What is New: * A systematic description of the ethical concepts and arguments grounding the debate on euthanasia in minors, as reported in the argument-based ethics literature. * A need has been identified to enrich the debate with a care-ethics approach to avoid oversimplifying the ethical decision-making process.

https://www.ncbi.nlm.nih.gov/pubmed/28573404


OBJECTIVE: We reviewed our decisions about continuation/withdrawal of life-sustaining treatments in a group of critically ill newborns who were discussed in structured medical ethical decision-making meetings, and provide the surviving children’s outcomes at 2-year follow-up. STUDY DESIGN: In an explorative observational study, 61 cases were evaluated. The children involved had been discussed in such a structured way from 2009 to 2012 in a level III-D neonatal intensive care unit. RESULTS: Decisions made were: full treatment (n=6), earlier restriction cancelled (n=3), treatment restriction (n=30) and palliative care (n=22). Parents of six children disagreed with the decision proposed. Thirteen (54%) of the 24 children who survived (39%) had moderate to severe neurological problems; 8 (33%) had additional sequelae; only one 2-year-old child was healthy. CONCLUSIONS: Decisions made varied to a large extent. The poor outcomes should be disseminated among decision makers. Future studies must explore new ways to improve outcome prediction, extend follow-up periods and consider what living with severe handicaps really means for both child and family.


Illness narratives have become very popular. The stories of children, however, are rarely ever studied. This paper aims to provide insight into how children, parents and physicians make sense of progressive childhood cancer. It also explores how this meaning-giving process interacts with cultural dominant stories on cancer and dying. The presented data come from 16 open-ended face-to-face interviews with palliative paediatric patients, their parents and physicians. The interviews were carried out in eight paediatric oncology centres in Switzerland. Data analysis followed Arthur Frank’s dialogical narrative analysis. Quest narratives were relatively rare compared to both chaos and restitution stories. All participants welcomed chaos stories as a liminal haven between quest and restitution. The possibility that the child could die was either ignored or briefly contemplated, but then immediately pushed away. Except for one patient, children never directly addressed the topic of death. The way in which death was presented raises important questions about how the social discourse on dying is framed in terms of choice, autonomy and individuality. This discourse not only determines the way in which children and adults relate to the minor’s death, it also constitutes an obstacle to children’s participation in decision-making.


Requests for life-prolonging treatments can cause irresolvable conflicts between health-care providers and surrogates. The Multiorganization Policy Statement (Bosset et al. 2015) with recommendations to prevent and manage these conflicts creates a good opportunity to examine how end-of-life decisions are made in Dutch neonatal intensive care units and how medical futility is defined. The Dutch equivalent of medical futility in the context of NICU care has grown and developed rather independently, within the typical legal, ethical, and cultural framework of Dutch society. Moreover, intractable conflicts and legal claims about life-prolonging treatments of newborns are very rare. This makes the Multiorganization Statement recommendations only partly relevant to Dutch practice. Critics are right in their comments that a substitution for the term futility, as recommended in the statement, is unlikely to prevent or solve irresolvable conflicts. But the recommended transparency about limit setting may work well, provided physicians make sure that comfort and palliative care are always provided.

https://www.ncbi.nlm.nih.gov/m/pubmed/29375078

CONTEXT: Human rights standards to address palliative care have developed over the last decade. OBJECTIVES: This article aims to examine key milestones in the evolution of human rights standards to address palliative care, relevant advocacy efforts, and areas for further growth. METHODS: The article provides an analysis of human rights standards in the context of palliative care through the lens of the right to health, freedom from torture and ill treatment, and the rights of older persons and children. RESULTS: Significant developments include the following: 1) the first human rights treaty to explicitly recognize the right to palliative care, the Inter-American Convention on the Rights of Older Persons; 2) the first World Health Assembly resolution on palliative care; 3) a report by the UN Special Rapporteur on Torture with a focus on denial of pain treatment; 4) addressing the availability of controlled medicines at the UN General Assembly Special Session on the World Drug Problem. CONCLUSION: Development of human rights standards in relation to palliative care has been most notable in the context of the right to health, freedom from torture and ill treatment, and the rights of older persons. More work is needed in the context of the rights of children, and human rights treaty bodies are still not consistently addressing state obligations with regards to palliative care.


Withdrawing Artificial Nutrition and Hydration (WANH) in the neonatal intensive care units (NICUs) has long been controversial. In France, the practice has become a legal option since 2005. But even though, the question remains as to what the stakeholders’ experience is, and whether they consider it ethically appropriate. In order to contribute to the debate, we initiated a study in 2009 to evaluate parental and health care professionals (HCP) perspectives, after they experienced WAHN for a newborn. The study included 25 cases from 5 different clinical neonatology departments. We interviewed both the parents and some of the HCP who cared for the baby, at least 6 months after this latter deceased. We proceeded through in-depth individual qualitative interviews. Content was analyzed for themes and patterns that emerged from the data. Some parents expressed that WANH offered an opportunity to the family to spend a few demedicalized days with the baby before she dies, without any tubes and machines, and to be well supported by the HCP during this palliative stage. But others evaluated the practice in retrospect as a terrible ordeal. All said that, at least, the time of waiting for death to ensue should not last too long. After a few days, it becomes unbearable because of the transformation of the baby’s appearance and because they, as parents, began to wonder if she was not dying from starvation rather than from her initial disease. An important proportion of HCP also expressed some kind of ethical unease. This was due to the psychological violence involved in the decision for a human being to stop feeding a little one, and also to the difficulty to deal with the fundamental intention behind the decision of WANH: indeed, could it be claim that it does not presuppose the intention of provoking the infant’s death? The discussion focuses on the point to know if WANH can be considered as a source of progress from an ethical point of view, particularly in comparison with earlier practices—that in France could involve active euthanasia by lethal injection. We argue that when HCP are merely focused on avoiding that WAHN could be construed as a way of intending to hasten the baby’s death, the practice is at risk to be implemented in a way that becomes ethically counterproductive. Focusing on this intention easily distract the clinical teams from what should be their ultimate concern, namely the baby’s comfort during the dying process, as well as the support owned to her parents. To conclude, we suggest that the ethical priorities, when WANH is decided, should be the support due to the patient and her family on the one hand, and, on the other hand to implement it in such a way that at least the baby seems to have died of her initial disease and never of starvation. This means that HCP have a duty to control the timing of death, even though this might be incompatible with the worry to avoid the intention of hastening the baby’s death.


Recent research has focused attention on the role of patients’ and clinicians’ cultural skills and values in generating inequalities in health care experiences. Yet, examination of how social structural factors shape people’s abilities to build, refine, and leverage strategies for navigating the health care system have received less attention. In this paper I place focus on one such social structural factor, social support, and examine how social support operates as a flexible resource that helps people navigate the health care system. Using the case of families navigating pediatric cancer care this study combines in-depth interviews with parents of pediatric cancer patients (N = 80), direct observation of clinical interactions between families and physicians (N = 73), and in-depth interviews with pediatric oncologists (N = 8). Findings show that physicians assess parental visibility in the hospital, medical vigilance, and adherence to their child’s treatment and use these judgments to shape clinical decision-making. Parents who had help from their personal networks had more agility in balancing competing demands, and this allowed parents to more effectively meet institutional expectations for appropriate parental involvement in the child’s health care. In this way, social support served as a flexible resource for some families that allowed parents to more quickly adapt to the demands of caring for a child with cancer, foster productive interpersonal relationships with health care providers, and play a more active role in their child’s health care.


Emeritus Professor Edward Alan Glasper from the University of Southampton discusses the complexities of care delivery to children in hospital who have life limiting medical conditions.


BACKGROUND: The ability to save the life of an extremely premature baby has increased substantially over the last decade. This survival, however, can be associated with unfavourable outcomes for both baby and family. Questions are now being asked about quality of life for survivors of extreme prematurity. Quality of life is rightly deemed to be an important consideration in high technology neonatal care; yet, it is notoriously difficult to determine or predict. How does one define and operationalise what is considered to be in the best interest of a surviving extremely premature baby, especially when the full extent of the outcomes might not be known for several years? RESEARCH QUESTION: The research investigates the caregiving dilemmas often faced by neonatal nurses when caring for extremely premature babies. This article explores the issues arising for neonatal nurses when they considered the philosophical and ethical questions about quality of life in babies </24 weeks gestation. PARTICIPANTS: Data were collected via a questionnaire to Australian neonatal nurses and semi-structured interviews with 24 neonatal nurses in New South Wales, Australia. Ethical considerations: Ethical processes and procedures have been adhered to by the researchers. FINDINGS: A qualitative approach was used to analyse the data. The theme ‘difficult choices’ was generated which comprised three sub-themes: ‘damaged through survival’, ‘the importance of the brain’ and ‘families are important’. The results show that neonatal nurses believed that quality of life was an important consideration; yet they experienced significant inner conflict and uncertainty when asked to define or suggest specific elements of quality of life, or to suggest how it might be determined. It was even more difficult for the nurses to say when an extremely premature baby’s life possessed quality. Their previous clinical and personal experiences led the nurses to believe that the quality of the family’s life was important, and possibly more so than the quality of life of the surviving baby. This finding contrasts markedly with much of the existing literature in this field. CONCLUSION: Quality of life for extremely premature babies was an important consideration for neonatal nurses; however, they experienced difficulty deciding how to operationalise such considerations in their everyday clinical practice.

Richard Griffith, Senior Lecturer in Health Law at Swansea University, reviews how the courts assist in settling disputes over the care of seriously ill babies and describes the test used to inform decisions about their treatment.


This discussion follows a series of high profile cases involving a terminally ill child, Charlie Gard. These cases are significant as they trace the complexities that arise when parents and medical teams do not agree as well as addressing the question of whether there is a right to access experimental treatment.
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5715039/


There is an inconsistency in the ways that doctors make clinical decisions regarding the treatment of babies born extremely prematurely. Many experts now recommend that clinical decisions about the treatment of such babies be individualized and consider many different factors. Nevertheless, many policies and practices throughout Europe and North America still appear to base decisions on gestational age alone or on gestational age as the primary factor that determines whether doctors recommend or even offer life-sustaining neonatal intensive care treatment. These policies are well intentioned. They aim to guide doctors and parents to make decisions that are best for the baby. That is an ethically appropriate goal. But in relying so heavily on gestational age, such policies may actually do the babies a disservice by denying some babies treatment that might be beneficial and lead to intact survival. In this paper, we argue that such policies are unjust to premature babies and ought to be abolished. In their place, we propose individualized treatment decisions for premature babies. This would treat premature babies as we treat all other patients, with clinical decisions based on an individualized estimation of likelihood that treatment would be beneficial.


BACKGROUND: Digital communication between a patient and their clinician offers the potential for improved patient care, particularly for young people with long term conditions who are at risk of service disengagement. However, its use raises a number of ethical questions which have not been explored in empirical studies. The objective of this study was to examine, from the patient and clinician perspective, the ethical implications of the use of digital clinical communication in the context of young people living with long-term conditions. METHODS: A total of 129 semi-structured interviews, 59 with young people and 70 with healthcare professionals, from 20 United Kingdom (UK)-based specialist clinics were conducted as part of the LYNC study. Transcripts from five sites (cancer, liver, renal, cystic fibrosis and mental health) were read by a core team to identify explicit and implicit ethical issues and develop descriptive ethical codes. Our subsequent thematic analysis was developed iteratively with reference to professional and ethical norms. RESULTS: Clinician participants saw digital clinical communication as potentially increasing patient empowerment and autonomy; improving trust between patient and healthcare professional; and reducing harm because of rapid access to clinical advice. However, they also described ethical challenges, including: difficulty with defining and maintaining boundaries of confidentiality; uncertainty regarding the level of consent required; and blurring of the limits of a clinician’s duty of care when unlimited access is possible. Paradoxically, the use of digital clinical communication can create dependence rather than promote autonomy in some patients. Patient participants varied in their understanding of, and concern about, confidentiality in the context of digital communication. An overarching theme emerging from the data was a shifting of the boundaries of the patient-clinician relationship and the professional duty of care in the context of use of clinical digital communication. CONCLUSIONS: The ethical implications of clinical digital communication are complex and go beyond concerns about confidentiality and consent. Any development of this form of
communication should consider its impact on the patient-clinician relationship, and include appropriate safeguards to ensure that professional ethical obligations are adhered to.


Approval of Spinraza (nusinersen) for treatment of spinal muscular atrophy prompts consideration of a number of ethical issues that arise whenever a new treatment is proposed for a serious condition, especially one that is rare and can devastatingly affect children. Patients, families, clinicians, researchers, institutions and policymakers all must take account of the ways that newly available treatments affect informed and shared decision-making about therapeutic and research options. The issues to consider include: addressing what is still uncertain and unknown; the possibility that potential benefits will be exaggerated and potential harms underemphasized in the media, by advocacy organizations, and in consent forms and processes; the high cost of many novel drugs and biologics; the effects of including conditions of variable phenotype in state-mandated newborn screening panels; and how new treatments can change the standard of care, altering what is and is not known about a disorder and posing challenges for decision-making at both individual and policy levels. The good news that Spinraza brings thus requires additional attention to its ethical and policy implications, to improve counseling and shared decision-making about treatment and research options for patients and all involved in their care.


ECMO has proven to be a life-saving intervention for a variety of disease entities with a high rate of survival in the neonatal population. However, ECMO requires clinical teams to engage in many ethical considerations. Even with ongoing improvements in technology and expertise, some patients will not survive a course of ECMO. An unsuccessful course of ECMO can be difficult to accept and cause a great deal of angst. These questions can result in real conflict both within the care team, and between the care team and the family. Herein we explore a range of ethical considerations that may be encountered when caring for a patient on ECMO, with a particular focus on those courses where it appears likely that the patient will not survive. We then consider how a palliative care approach may provide a tool set to help engage the team and family in confronting the difficult decision to discontinue ECMO.

https://www.ncbi.nlm.nih.gov/m/pubmed/29331209


OBJECTIVES: In the context of serious or life-limiting illness, pediatric patients and their families are faced with difficult decisions surrounding appropriate resuscitation efforts in the event of a cardiopulmonary arrest. Code status orders are one way to inform end-of-life medical decision making. The objectives of this study are to evaluate the extent to which pediatric providers have knowledge of code status options and explore the association of provider role with (1) knowledge of code status options, (2) perception of timing of code status discussions, (3) perception of family receptivity to code status discussions, and (4) comfort carrying out code status discussions. DESIGN: Nurses, trainees (residents and fellows), and attending physicians from pediatric units where code status discussions typically occur completed a short survey questionnaire regarding their knowledge of code status options and perceptions surrounding code status discussions. SETTING: Single center, quaternary care children's hospital. MEASUREMENTS AND MAIN RESULTS: 203 nurses, 31 trainees, and 29 attending physicians in 4 high-acuity pediatric units responded to the survey (N = 263, 90% response rate). Based on an
objective knowledge measure, providers demonstrate poor understanding of available code status options, with only 22% of providers able to enumerate more than two of four available code status options. In contrast, provider groups self-report high levels of familiarity with available code status options, with attending physicians reporting significantly higher levels than nurses and trainees (p = 0.0125). Nurses and attending physicians show significantly different perception of code status discussion timing, with majority of nurses (63.4%) perceiving discussions as occurring “too late” or “much too late” and majority of attending physicians (55.6%) perceiving the timing as “about right” (p<0.0001). Attending physicians report significantly higher comfort having code status discussions with families than do nurses or trainees (p<0.0001). Attending physicians and trainees perceive families as more receptive to code status discussions than nurses (p<0.0001 and p = 0.0018, respectively). CONCLUSIONS: Providers have poor understanding of code status options and differ significantly in their comfort having code status discussions and their perceptions of these discussions. These findings may reflect inherent differences among providers, but may also reflect discordant visions of appropriate care and function as a potential source of moral distress. Lack of knowledge of code status options and differences in provider perceptions are likely barriers to quality communication surrounding end-of-life options. 


Many times, we make decisions in our lives that have unanticipated impacts. Decisions made can change our careers and outlook toward life. As a younger doctor, I considered illness and disease abstractions, and good management with optimal outcomes for the patient was our focus, as we strive toward our professional goals. But having experienced the real pain and helplessness that patients and their parents undergo during critical illnesses, the importance of empathy together with the knowledge and skills needed to treat a child should never be undervalued.


Care for children as they near the end of life is difficult and very complex. More difficult still are the decisions regarding what interventions are and are not indicated during these trying times. Occasionally, families of children who are nearing the end of life disagree with the assessment of the medical team regarding these interventions. In rare cases, the medical team can be moved to enact a do not attempt resuscitation order against the wishes of the patient’s parents. This article presents one such illustrative case and discusses the ethical issues relevant to such challenging clinical scenarios. The authors posit that such a unilateral do not attempt resuscitation order is only appropriate in very limited circumstances in pediatric care. Instead, focus should be placed on open discussion between parents and members of the clinical team, shared decision making, and maintenance of the clinician-parent relationship while simultaneously supporting members of the clinical team who express discomfort with parental decisions. The authors propose an alternative framework for approaching such a conflict based on clinician-parent collaboration and open communication.


Palliative care for children who can expect only a short life has expanded over the last decade. Greater understanding of the measures required to ensure comfort and acceptable quality of life within the critical care environment has grown in tandem. Some more invasive interventions may be considered a “step too far” by some practitioners, including feeding gastrostomy, contracture release, and tracheostomy. Tracheostomy can facilitate a number of measures, which may enhance the brief life of the child and their family. However, tracheostomy is associated with some challenges, which may make it less suitable for some families. We discuss 3 cases where this intervention was carried out.


The perinatal world is unique in its dutiful consideration of two patients along the lines of decision-making and clinical management - the fetus and the pregnant woman. The potentiality of the fetus-newborn is intertwined with the absolute considerations for the woman as autonomous patient. From prenatal diagnostics, which may be quite extensive, to potential interventions prenatally, postnatal resuscitation, and neonatal management, the fetus and newborn may be anticipated to survive with or without special needs and technology, to have a questionable or guarded prognosis, or to live only minutes to hours. This review will address the ethical ramifications for prenatal diagnostics, parental values and goals clarification, birth plans, the fluidity of decision-making over time, and the potential role of prenatal and postnatal palliative care support.


Patient and public involvement (PPI) is important both in research and in quality improvement activities related to healthcare services. While PPI activities do not require formal ethical approval, they can raise a number of ethical concerns, through the introduction of complex technical medical concepts, challenging language or sensitive subject areas. There is very little published literature to guide ethical practice in this area. We have been conducting PPI with children and young people throughout a research study in paediatric palliative care. PPI started during the application process and continued to guide and shape the research as it progressed. Ethical issues can arise at any time in PPI work. Although many can be predicted and planned for, the nature of PPI means that researchers can be presented with ideas and concepts they had not previously considered, requiring reflexivity and a reactive approach. This paper describes how we considered and addressed the potential ethical issues of PPI within our research. The approach that emerged provides a framework that can be adapted to a range of contexts and will be of immediate relevance to researchers and clinicians who are conducting PPI to inform their work.


Many babies with trisomy 13 and 18 die in the first year of life. Survivors all have severe cognitive impairment. There has been a debate among both professionals and parents about whether it is appropriate to provide life-sustaining interventions to babies with these serious conditions. On one side of the debate are those who argue that there is no point in providing invasive, painful, and expensive procedures when the only outcomes are either early death or survival with severe cognitive impairment. Others suggest that, although mortality is high and cognitive impairment universal, babies with these conditions have an acceptable quality of life. In this paper, we will discuss both points of view. We will review the ways in which these conditions are portrayed in pediatrics textbooks and on social media sites that offer support to parents. We will then suggest an appropriate way to deal with clinical decisions for babies with these trisomies.


BACKGROUND: In the last decade, the number of children with life-limiting and life-threatening conditions in England has almost doubled, and it is estimated that worldwide, there are 1.2 million children with palliative care needs. Families and professionals caring for children with life-limiting conditions are likely to face a number of difficult treatment decisions and develop plans for future care over the course of the child's life, but little is known about the process by which these decisions and plans are made. METHODS: The purpose of this review is to synthesize findings from qualitative research that has investigated decision-making and future planning for children with life-limiting conditions. A systematic search of six online databases was conducted and identified
887 papers for review; five papers were selected for inclusion, using predefined criteria. Reference list searching and contacting authors identified a further four papers for inclusion. RESULTS: Results sections of the papers were coded and synthesized into themes. Nineteen descriptive themes were identified, and these were further synthesized into four analytical themes. Analytical themes were ‘decision factors’, ‘family factors’, ‘relational factors’ and ‘system factors’. CONCLUSIONS: Review findings indicate that decision-making and future planning is difficult and needs to be individualized for each family. However, deficits in understanding the dynamic, relational and contextual aspects of decision-making remain and require further research.


We report about an infant who was diagnosed with spinal muscular atrophy type 1 (Werdnig-Hoffmann) at the age of 7 weeks. In a detailed discussion with the physicians the parents decided against mechanical ventilation in the event of either acute or chronic respiratory failure. During care at home all palliative actions were taken to optimize the quality of life for the child. The boy died at the age of 7 months at home. Whether continuous mechanical ventilation in a child with spinal muscular atrophy type 1 should be the individual decision of the parents or whether it should never be offered by the physicians is discussed controversially in the present medical literature.


The discontinuation of life sustaining medical treatment (LSMT) in severely and permanently impaired neonates, especially artificial nutrition and hydration (ANH) is subject to uncertainty and controversy. Definitive clinical guidelines are lacking, clinical research is limited, ethical disagreement is commonplace, and while case and statutory law provide legal underpinning for the practice in defined circumstances, uncertainty in this realm likely influences clinical practice. We use the case of a neurologically devastated neonate to highlight and review these arenas, and show how, using available legal, ethical, and clinical standards and practice, the case of Baby O was resolved, and to underline the need for further research in neonatal palliative care.


BACKGROUND: Pediatric oncologists are often faced with situations in which parents or guardians refuse recommended treatment for curable childhood cancer. Deciding how to proceed in such situations is an ethical dilemma. The aim of this article is to consider optimal approaches when parents are strongly against oncological treatment, potentially compromising their childrens rights for health care and to the chance for cure. CASES: In this paper, we report two cases of treatment refusal from our department and the impact of such decisions on the children themselves. Case no. 1 describes a child with retinoblastoma whose parents refused standard treatment in order to seek alternative treatment abroad. Case no. 2 describes a patient with a primary lymphoma of bone who received treatment by a court order after parental refusal. CONCLUSION: When parents refuse a treatment for potentially curable cancer, the medical team often focuses on the certainty of death without treatment. In the background, there is a smaller but still significant risk that - even if the treatment is eventually accepted or compelled - the child will still die from treatment-related complications or refractory disease, possibly with considerable suffering. The reasons for refusing a treatment vary. The entire medical team is tasked with trying to
respectfully understand the reasoning behind the parents unwillingness to accept the treatment, in order to address all possible misunderstandings and to propose solutions that could be acceptable for the parents. In some situations however, it is necessary to resolve the dilemma by legal means in order to protect the life of the child. Key words: oncology - ethics - decision making - treatment refusal - legal guardians. The authors declare they have no potential conflicts of interest concerning drugs, products, or services used in the study. The Editorial Board declares that the manuscript met the ICMJE recommendation for biomedical papers. Submitted: 7. 8.2017 Accepted: 7. 9. 2017.


OBJECTIVES: Typically pediatric end-of-life decision-making studies have examined the decision-making process, factors, and doctors’ and parents’ roles. Less attention has focussed on what happens after an end-of-life decision is made; that is, decision enactment and its outcome. This study explored the views and experiences of bereaved parents in end-of-life decision-making for their child. Findings reported relate to parents’ experiences of acting on their decision. It is argued that this is one significant stage of the decision-making process. METHODS: A qualitative methodology was used. Semi-structured interviews were conducted with bereaved parents, who had discussed end-of-life decisions for their child who had a life-limiting condition and who had died. Data were thematically analysed. RESULTS: Twenty-five bereaved parents participated. Findings indicate that, despite differences in context, including the child’s condition and age, end-of-life decision-making did not end when an end-of-life decision was made. Enacting the decision was the next stage in a process. Time intervals between stages and enactment pathways varied, but the enactment was always distinguishable as a separate stage. Decision enactment involved making further decisions - parents needed to discern the appropriate time to implement their decision to withdraw or withhold life-sustaining medical treatment. Unexpected events, including other people’s actions, impacted on parents enacting their decision in the way they had planned. Several parents had to re-implement decisions when their child recovered from serious health issues without medical intervention. Significance of results A novel, critical finding was that parents experienced end-of-life decision-making as a sequence of interconnected stages, the final stage being enactment. The enactment stage involved further decision-making. End-of-life decision-making is better understood as a process rather than a discrete once-off event. The enactment stage has particular emotional and practical implications for parents. Greater understanding of this stage can improve clinician’s support for parents as they care for their child.


Most childhood deaths in the United States occur in hospitals. Pediatric intensive care clinicians must anticipate and effectively treat dying children’s pain and suffering and support the psychosocial and spiritual needs of families. These actions may help family members adjust to their loss, particularly bereaved parents who often experience reduced mental and physical health. Candid and compassionate communication is paramount to successful end-of-life (EOL) care as is creating an environment that fosters meaningful family interaction. EOL care in the pediatric intensive care unit is associated with challenging ethical issues, of which clinicians must maintain a sound and working understanding.


Many families of patients hold the view that it is their right to be present during a loved one’s resuscitation, while the majority of patients also express the comfort and support they would feel by having them there. Currently, family presence is more commonly accepted in paediatric cardiopulmonary resuscitation (CPR) than adult CPR. Even though many guidelines are in favour of this practice and recognise potential benefits, healthcare professionals are hesitant to support adult family presence to the extent that paediatric family presence is supported. However, in this paper, we suggest that the ethical case to justify family presence during paediatric resuscitation (P-FPDR) is weaker than the justification of family presence during adult resuscitation (A-FPDR). We go on to support this claim using three main arguments that people use in clinical ethics to justify FPDR. These include scarcity of evidence documenting disruption, psychological benefits to family members following the incident and respect for patient autonomy. We demonstrate that these arguments actually apply more strongly to A-FPDR compared with P-FPDR, thereby questioning the common attitude of healthcare professionals of allowing the latter while mostly opposing A-FPDR. Importantly, we do not wish to suggest that P-FPDR should not be allowed. Rather, we suggest that since P-FPDR is commonly (and should be) allowed, so should A-FPDR. This is because the aforementioned arguments that are used to justify FPDR in general actually make a stronger case for A-FPDR.


Pediatric health care is practiced with the goal of promoting the best interests of the child. Treatment generally is rendered under a presumption in favor of sustaining life. However, in some circumstances, the balance of benefits and burdens to the child leads to an assessment that forgoing life-sustaining medical treatment (LSMT) is ethically supportable or advisable. Parents are given wide latitude in decision-making concerning end-of-life care for their children in most situations. Collaborative decision-making around LSMT is improved by thorough communication among all stakeholders, including medical staff, the family, and the patient, when possible, throughout the evolving course of the patient’s illness. Clear communication of overall goals of care is advised to promote agreed-on plans, including resuscitation status. Perceived disagreement among the team of professionals may be stressful to families. At the same time, understanding the range of professional opinions behind treatment recommendations is critical to informing family decision-making. Input from specialists in palliative care, ethics, pastoral care, and other disciplines enhances support for families and medical staff when decisions to forgo LSMT are being considered. Understanding specific applicability of institutional, regional, state, and national regulations related to forgoing LSMT is important to practice ethically within existing legal frameworks. This guidance represents an update of the 1994 statement from the American Academy of Pediatrics on forgoing LSMT.

Education, Research and Professional Issues


BACKGROUND: Although medical marijuana (MM) may have utility in the supportive care of children with serious illness, it remains controversial. We investigated interdisciplinary provider perspectives on legal MM use in children with cancer. **METHODS**: We sent a 32-item, cross-sectional survey to 654 pediatric oncology providers in Illinois, Massachusetts, and Washington characterizing MM practices, knowledge, attitudes, and barriers. Forty-eight percent responded; 44% (n = 288) were included in analyses. Providers were stratified by status as legally eligible to certify (ETC) for MM. We used Fisher's exact and Wilcoxon rank tests and univariate and multivariate logistic regression models for group comparisons. **RESULTS**: The provider median age was 35 years (range 22-70 years); 33% were ETC (83 physicians; 13 Washington state advance practice providers). Thirty percent of providers received >/=1 request for MM in the previous month. Notably, only 5% of all providers knew state-specific regulations. ETC providers were more likely to know that MM is against federal laws (P < .0001). Whereas most providers (92%) reported willingness to help children with cancer access MM, in adjusted models, ETC providers were less likely to indicate approval of patient MM use by smoking, oral formulations, as cancer-directed therapy, or to manage symptoms (P < .005 for all). Forty-six percent of all providers cited the absence of standards around formulations, potency, or dosing to be the greatest barrier to recommending MM. **CONCLUSIONS**: Most pediatric oncology providers are willing to consider MM use in children with cancer and receive frequent inquiries. However, ETC providers endorse less favorable attitudes overall. The absence of standards is an important barrier to recommending MM.


AIMS: This study explored how paediatric healthcare professionals experienced and coped with end-of-life conflicts and identified how to improve coping strategies. **METHODS**: A questionnaire was distributed to all 2300 professionals at a paediatric university hospital, covering the frequency of end-of-life conflicts, participants, contributing factors, resolution strategies, outcomes and the usefulness of specific institutional coping strategies. **RESULTS**: Of the 946 professionals (41%) who responded, 466 had witnessed or participated in paediatric end-of-life discussions: 73% said these had led to conflict, more frequently between professionals (58%) than between professionals and parents (33%). Frequent factors included professionals' rotations, unprepared parents, emotional load, unrealistic parental expectations, differences in values and beliefs, parents' fear of hastening death, precipitated situations and uncertain prognosis. Discussions with patients and parents and between professionals were the most frequently used coping strategies. Conflicts were frequently resolved by the time of death. Professionals mainly supported designating one principal physician and nurse for each patient, two-step interdisciplinary meetings - between professionals then with parents - postdeath ethics meetings, bereavement follow-up protocols and early consultations with paediatric palliative care and clinical ethics services. **CONCLUSION**: End-of-life conflicts were frequent and predominantly occurred between healthcare professionals. Specific interventions could target most of the contributing factors.


OBJECTIVE: Parents of seriously ill children require attention to their spiritual needs, especially during end-of-life care. The objective of this study was to characterize parental attitudes regarding physician inquiry into their belief system. **Materials and Main Results**: A total of 162 surveys from parents of children hospitalized for >48 hours in pediatric intensive care unit in a tertiary academic medical center were analyzed. Forty-nine percent of all respondents and 62% of those who identified themselves as moderate to very spiritual or religious stated that
their beliefs influenced the decisions they made about their child's medical care. Although 34% of all respondents would like their physician to ask about their spiritual or religious beliefs, 48% would desire such enquiry if their child was seriously ill. Those who identified themselves as moderate to very spiritual or religious were most likely to welcome the discussion (P < .001). Two-thirds of the respondents would feel comforted to know that their child's physician prayed for their child. One-third of all respondents would feel very comfortable discussing their beliefs with a physician, whereas 62% would feel very comfortable having such discussions with a chaplain.

CONCLUSION: The study findings suggest parental ambivalence when it comes to discussing their spiritual or religious beliefs with their child's physicians. Given that improved understanding of parental spiritual and religious beliefs may be important in the decision-making process, incorporation of the expertise of professional spiritual care providers may provide the optimal context for enhanced parent-physician collaboration in the care of the critically ill child.


AIM: This study determined whether there was a difference in the conclusions reached by neonatologists in morbidity and mortality conferences based on their level of involvement in a case. METHODS: All neonatal deaths occurring between August 2014 and September 2015 at the neonatal intensive care unit of Sainte-Justine Hospital, Montreal, Quebec, Canada, were reviewed by internal physicians involved in the case and external physicians who were not. The reviewers were asked to identify positive and negative clinical practice items and provide written recommendations. These were classified into eight categories and compared for each case.

RESULTS: During the study, 55 patients died leading to 110 reviews and a total of 590 positive and negative items. Most items were in the communication (25.2%), ethical decision-making (16.7%) and clinical management (14.8%) categories. Both the internal and external reviewers were in agreement 48.5% of the time for positive items and 44.8% for negative items. There were 242 written recommendations, which differed significantly among the internal and external reviewers. CONCLUSION: Reviews of neonatal deaths by two independent reviewers, internal physicians and external physicians, led to different positive and negative practice items and recommendations. This could allow for a richer discussion and improve recommendations for patient care.


BACKGROUND: Following a perinatal death, a formal standardised multi-disciplinary review should take place, to learn from the death of a baby and facilitate improvements in future care. It has been recommended that bereaved parents should be offered the opportunity to give feedback on the care they have received and integrate this feedback into the perinatal mortality review process. However, the MBRRACE-UK Perinatal Confidential Enquiry (2015) found that only one in 20 cases parental concerns were included in the review. Although guidance suggests parental opinion should be sought, little evidence exists on how this may be incorporated into the perinatal mortality review process. The purpose of the PARENTS study was to investigate bereaved parents' views on involvement in the perinatal mortality review process. METHODS: A semi-structured focus group of 11 bereaved parents was conducted in South West England. A purposive sampling technique was utilised to recruit a diverse sample of women and their partners who had experienced a perinatal death more than 6 months prior to the study. A six-stage thematic analysis was followed to explore parental perceptions and expectations of the perinatal mortality review process. RESULTS: Four over-arching themes emerged from the analysis: transparency; flexibility combined with specificity; inclusivity; and a positive approach. It was evident that the majority of parents were supportive of their involvement in the perinatal mortality review process and they wanted to know the outcome of the meeting. It emerged that an individualised approach should be taken to allow flexibility on when and how they could contribute to the process. The emotional aspects of care should be considered as well as the clinical care. Parents identified that the whole care pathway should be examined during the review including antenatal, postnatal, and neonatal and community based care. They agreed that there should be an opportunity for parents to give feedback on both good and poor aspects of their care. CONCLUSION: Parents were unaware that a review of their baby's death took place in the hospital. Parental involvement in the perinatal mortality review process would promote an open culture in the healthcare system and learning from adverse events including deaths. Further research should focus on designing and evaluating a perinatal mortality
BACKGROUND: The perinatal mortality review meeting that takes place within the hospital following a stillbirth or neonatal death enables clinicians to learn vital lessons to improve care for women and their families for the future. Recent evidence suggests that parents are unaware that a formal review following the death of their baby takes place. Many would welcome the opportunity to feedback into the meeting itself. Parental involvement in the perinatal mortality review meeting has the potential to improve patient satisfaction, drive improvements in patient safety and promote an open culture within healthcare. Yet evidence on the feasibility of involving bereaved parents in the review process is lacking. This paper describes the protocol for the Parents’ Active Role and Engagement in the review of their Stillbirth/perinatal death study (PARENTS 2), whereby healthcare professionals’ and stakeholders’ perceptions of parental involvement will be investigated, and parental involvement in the perinatal mortality review will be piloted and evaluated at two hospitals.

METHODS AND ANALYSIS: We will investigate perceptions of parental involvement in the perinatal mortality review process by conducting four focus groups. A three-round modified Delphi technique will be employed to gain a consensus on principles of parental involvement in the perinatal mortality review process. We will use three sequential rounds, including a national consensus meeting workshop with experts in stillbirth, neonatal death and bereavement care, and a two-stage anonymous online questionnaire. We will pilot a new perinatal mortality review process with parental involvement over a 6-month study period. The impact of the new process will be evaluated by assessing parents’ experiences of their care and parents’ and staff perceptions of their involvement in the process by conducting further focus groups and using a Parent Generated Index questionnaire.

ETHICS AND DISSEMINATION: This study has ethical approval from the UK Health Research Authority. We will disseminate the findings through national and international conferences and international peer-reviewed journals.


While availability of palliative care consultation for children with advanced heart disease increases, little is known about cardiologist attitudes towards palliative care. We sought to describe perspectives of cardiologists regarding palliative care and to characterize their perceived competence in palliative care concepts. A cross-sectional survey of pediatric cardiologists and cardiac surgeons from 19 pediatric medical centers was performed. Overall response rate was 31% (183/589). Respondents had a median of 18 years of experience since medical school (range 2-49) and most practiced at academic centers (91%). Sixty-percent of respondents felt that palliative care consultations occur “too late” and the majority (85%) agreed that palliative care consultations are helpful. Barriers to requesting palliative care consultation were most frequently described as “referring to palliative care services too early will undermine parents' hope” (45%) and “concern that parents will think I am giving up on their child” (56%). Only 33% of cardiologists reported feeling “very” or “moderately” competent in prognosticating life expectancy while over 60% felt competent caring for children with heart disease around end of life, and nearly 80% felt competent discussing goals of care and code status. Greater perceived competence was associated with subspecialty (heart failure/intensivist vs. other) (OR 3.6, 95% CI 1.6-8.1, p = 0.003) and didactic training (OR 6.27, 95% CI 1.8-21.8, p = 0.004). These results underscore the need for further training in palliative care skills for pediatric cardiologists. Enhancing palliative care skills among cardiologists and facilitating partnership with subspecialty palliative care teams may improve overall care of children with advanced heart disease.


BACKGROUND: While the importance of pediatric palliative care (PPC) for children with life-threatening illness is increasingly recognized, little is known about physicians’ attitudes toward palliative care for children with heart disease. OBJECTIVE: To compare the perspectives of PPC physicians and pediatric cardiologists regarding...
palliative care in pediatric heart disease. DESIGN: Cross-sectional web-based surveys. RESULTS: Responses from 183 pediatric cardiologists were compared to those of 49 PPC physicians (response rates 31% [183/589] and 28% [49/175], respectively). Forty-eight percent of PPC physicians and 63% of pediatric cardiologists agreed that availability of PPC is adequate (p = 0.028). The majority of both groups indicated that PPC consultation occurs “too late.” Compared with pediatric cardiologists, PPC physicians reported greater competence in all areas of advance care planning, communication, and symptom management. PPC physicians more often described obstacles to PPC consultation as “many” or “numerous” (42% vs. 7%, p < 0.001). PPC physicians overestimated how much pediatric cardiologists worry about PPC introducing inconsistency in approach (60% vs. 11%, p < 0.001), perceive lack of added value from PPC (30% vs. 7%, p < 0.001), believe that PPC involvement will undermine parental hope (65% vs. 44%, p = 0.003), and perceive that PPC is poorly accepted by parents (53% vs. 27%, p < 0.001). CONCLUSIONS: There are significant differences between pediatric cardiologists and PPC physicians in perception of palliative care involvement and perceived barriers to PPC consultation. An intervention that targets communication and exchange of expertise between PPC and pediatric cardiology could improve care for children with heart disease.


AIM: Our study aimed to assess physicians’ experiences and education regarding advance care planning (ACP) in paediatrics. We aimed to assess barriers to ACP initiation, including the adequacy of exposure and education regarding ACP and whether practitioners would deem improved education and resource provision useful. METHODS: A 25-question survey was designed following literature review. Paediatricians, intensivists and advanced trainees at Sydney Children’s Hospital were invited to complete the online survey. Ninety-two responses were obtained over a 10-week period. RESULTS: Patients with life-limiting conditions are encountered frequently, with 57% of respondents caring for at least 10 such patients during the last 2 years. In total, 64% of respondents felt that ACP discussions should occur early around the time of diagnosis or during a period of stability; however, 57% observed discussions occurring late in illness after multiple acute, severe deteriorations. In total, 46% felt that multidisciplinary teams were the most appropriate to initiate ACP discussions. Prognostic uncertainty was the most common barrier to ACP initiation. Lack of experience and education were identified as barriers by 43 and 32%, respectively. The majority of respondents regarded exposure to ACP and education during training as inadequate. CONCLUSIONS: ACP discussions are being initiated later than physicians deem optimal. Of concern, clinicians prefer ACP discussions to be initiated by multidisciplinary teams, which may create a barrier to timely initiation. Barriers due to lack of education and experience could be overcome with improvements in training. Provision of education and resources would be welcomed and improve clinician skills in this area.


In this study we utilized the framework of patient-centered communication to explore the influence of physician gender and physician parental status on (1) physician-parent communication and (2) care of pediatric patients at the end of life (EOL). The findings presented here emerged from a larger qualitative study that explored physician narratives surrounding pediatric EOL communication. The current study includes 17 pediatric critical care and pediatric emergency medicine physician participants who completed narrative interviews between March and October 2012 to discuss how their backgrounds influenced their approaches to pediatric EOL communication. Between April and June of 2013, participants completed a second round of narrative interviews to discuss topics generated out of the first round of interviews. We used grounded theory to inform the design and analysis of the study. Findings indicated that physician gender is related to pediatric EOL communication and care in two primary ways: (1) the level of physician emotional distress and (2) the way physicians perceive the influence of gender on communication. Additionally, parental status emerged as an important theme as it related to EOL decision-making and communication, emotional distress, and empathy. Although physicians reported experiencing more emotional distress related to interacting with patients at the EOL after they became parents, they also felt that they were better able to show empathy to parents of their patients.


Pediatric end-of-life care (EOL care) entails challenging tasks for health care professionals (HCPs). Little is known about HCPs’ experiences and needs when providing pediatric EOL care in Switzerland. This study aimed to describe the experiences and needs of HCPs in pediatric EOL care in Switzerland and to develop recommendations for the health ministry. The key aspect in EOL care provision was identified as the capacity to establish a relationship with the dying child and the family. Barriers to this interaction were ethical dilemmas, problems in collaboration with the interprofessional team, and structural problems on the level of organizations. A major need was the expansion of vocational training and support by specialized palliative care teams. We recommend the development of a national concept for the provision of EOL care in children, accompanied by training programs and supported by specialized pediatric palliative care teams located in tertiary children’s hospitals.


BACKGROUND AND OBJECTIVE: Chronic pain is associated with significant functional and social impairment. The objective of this review was to assess the characteristics and quality of randomized controlled trials (RCTs) evaluating pain management interventions in children and adolescents with chronic pain. METHODS: We performed a systematic search of PubMed, Embase and the Cochrane Library up to July 2017. We included RCTs that involved children and adolescents (3 months-18 years) and evaluated the use of pharmacological or non-pharmacological intervention(s) in the context of pain persisting or re-occurring for more than 3 months. Methodological quality was evaluated using the Cochrane Risk of Bias (ROB) Tool. RESULTS: A total of 58 RCTs were identified and numbers steadily increased over time. The majority were conducted in single hospital institutions, with no information on study funding. Median sample size was 47.5 participants (Q1,Q3: 32, 70). Forty-five percent of RCTs included both adults and children and the median of the mean ages at inclusion was 12.9 years (Q1,Q3: 11, 15). Testing of non-pharmacological interventions was predominant and only 5 RCTs evaluated analgesics or co-analgesics. Abdominal pain, headache/migraine and musculoskeletal pain were the most common types of chronic pain among participants. Methodological quality was poor with 90% of RCTs presenting a high or unclear ROB. CONCLUSIONS: Evaluation of analgesics targeting chronic pain relief in children and adolescents through RCTs is marginal. Infants and children with long-lasting painful conditions are insufficiently represented in RCTs. We discuss possible research constraints and challenges as well as methodologies to circumvent them. SIGNIFICANCE: There is a substantial research gap regarding analgesic interventions for children and adolescents with chronic pain. Most clinical trials in the field focus on the evaluation of non-pharmacological interventions and are of low methodological quality. There is also a specific lack of trials involving infants and children and adolescents with long-lasting diseases.


BACKGROUND: Young people with cancer have distinct clinical and psychosocial needs during and after cancer treatment. However, as adolescent and young adult (AYA) cancer is rare, and only recently recognized as specialty, health professionals may not have the skills, competence, and confidence to meet the needs of the young patient with cancer. The aim of this study was to identify the learning needs of health professionals providing cancer care to adolescents and young adults before and following the introduction of a state-wide AYA cancer education program. METHODS: A survey of educational needs of health professionals was undertaken in 2013 at the commencement of the Queensland Youth Cancer Service. The survey was used to develop the education program of the service. The education program was delivered across the state in a variety of formats, covering a range of topics throughout 2013-2016. The second survey was completed in 2017. Results were compared to identify if educational needs or the self-rated confidence of health professionals in regard to AYA cancer care had changed over time. RESULTS: One hundred twenty-two participants completed the first survey and 73 completed the second. The most prominent educational needs in 2013 were palliative care and biomedical topics such as understanding AYA growth and development as well as specific AYA cancers and
treatment. The second survey identified that palliative care education remained important; however, there was a shift toward health professionals request for more psychosocial and practical education on topics including fertility, sexuality, and managing late effects. CONCLUSION: To provide high-quality healthcare to AYAs with cancer, health professionals require ongoing opportunities for education and training.


BACKGROUND: Pediatric fellows receive little palliative care (PC) education and have few opportunities to practice communication skills. OBJECTIVE: In this pilot study, we assessed (1) the relative effectiveness of simulation-based versus didactic education, (2) communication skill retention, and (3) effect on PC consultation rates. DESIGN: Thirty-five pediatric fellows in cardiology, critical care, hematology/oncology, and neonatology at two institutions enrolled: 17 in the intervention (simulation-based) group (single institution) and 18 in the control (didactic education) group (second institution). Intervention group participants participated in a two-day program over three months (three simulations and videotaped PC panel). Control group participants received written education designed to be similar in content and time. MEASUREMENTS: (1) Self-assessment questionnaires were completed at baseline, post-intervention and three months; mean between-group differences for each outcome measure were assessed. (2) External reviewers rated simulation-group encounters on nine communication domains. Within-group changes over time were assessed. (3) The simulation-based site's PC consultations were compared in the six months pre- and post-intervention. RESULTS: Compared to the control group, participants in the intervention group improved in self-efficacy (p = 0.003) and perceived adequacy of medical education (p < 0.001), but not knowledge (p = 0.20). Reviewers noted nonsustained improvement in four domains: relationship building (p = 0.01), opening discussion (p = 0.03), gathering information (p = 0.01), and communicating accurate information (p = 0.04). PC consultation rate increased 64%, an improvement when normalized to average daily census (p = 0.04). CONCLUSIONS: This simulation-based curriculum is an effective method for improving PC communication, education, and consults. More frequent practice is likely needed to lead to sustained improvements in communication competence.


BACKGROUND: Research exploring nurses' experiences working with families in paediatric intensive care unit (PICU) is limited. No studies have been undertaken in a mixed adult-paediatric ICU. OBJECTIVES: To explore nurses' perceptions of working with families of critically ill children in a mixed adult-paediatric intensive care unit (ICU). DESIGN: Descriptive qualitative design. METHODOLOGY: Five PICU nurses participated in semi-structured interviews. Data were analysed using thematic analysis. Trustworthiness was enhanced using an audit trail, member checks and peer review of all data. RESULTS: Three main themes and one overarching theme emerged. Role confusion and delineation examined the roles which nurses ascribed to themselves and the families; and demonstrated the conflict which could arise if roles were challenged. Information sharing demonstrated the positive and negative ways in which nurses utilized information with families in their daily practice. The contextual environment of the PICU scrutinized the physical, cultural and institutional factors which impacted on the nurses' ability to work with families in the PICU. Finally, the overarching theme Competing values explores the interplay between the nurses' personal values and those of the PICU and the institution. CONCLUSIONS: Working with families in a mixed adult-paediatric ICU is influenced by multiple personal and institutional factors. The value placed on families and on the time nurses spent with them often competed for priority with nurses' other values and the wider culture of the PICU. The potential for role confusion, the management of information and the physical environment of the PICU further contributed to variability in nurses' working with families. RELEVANCE TO CLINICAL PRACTICE: The results highlighted a need for education for both nurses and medical staff who work with families of critically ill children. Additionally, the need for each PICU to have a written policy on family
The presence and participation is crucial to guide practice and maintain continuity of care. 


OBJECTIVES: This study explores the influences of the paediatric intensive care environment on relationships between parents and healthcare providers when children are dying. It forms part of a larger study, investigating parental experiences of the death of their child in intensive care. RESEARCH METHODOLOGY: Constructivist grounded theory. SETTING: Four Australian paediatric intensive care units. MAIN OUTCOME MEASURES: Audio-recorded, semi-structured interviews were conducted with twenty-six bereaved parents. Data were analysed using the constant comparison and memoing techniques common to grounded theory. FINDINGS: The physical and social environment of the intensive care unit influenced the quality of the parent-healthcare provider relationship. When a welcoming, open environment existed, parents tended to feel respected as equal and included members of their child's care team. In contrast, environments that restricted parental presence or lacked resources for parental self-care could leave parents feeling like 'watchers', excluded from their child's care. CONCLUSIONS: The paediatric intensive care unit environment either welcomes and includes parents of dying children into the care team, or demotes them to the status of 'watcher'. Such environments significantly influence the relationships parents form with healthcare staff, their ability to engage in elements of their parental role, and their experiences as a whole. 


Recruitment of participants into bereavement research may present many challenges for the research team. At present, there is little consensus for researchers and ethics committees on the most appropriate method of recruitment. There is some evidence that participants prefer to be contacted about research studies via letters. However, recruitment involving the use of a letter can occur in a number of ways, each with ethical and practical benefits and limitations. In a study of the experiences of bereaved parents, we used letters in three ways: direct mailing from the research team with an opt-out option; permission to mail letters obtained by social workers from a hospital-based follow-up program during routine contact; and letters mailed from the hospital's PICU research nurse at the hospital with instruction on how to opt in. In this paper, the practical and ethical realities of each method are highlighted, using examples from our own experiences. Nineteen parents also provided reflections in follow-up phone calls. While direct researcher contact is perhaps the most feasible for researchers, ethical concerns may render it unacceptable. While contact via a known member of a follow-up program is more ethically appropriate for participants, it also presents significant practical issues. We suggest that contact via a representative of the healthcare institution provides the best balance of ethical and practical acceptability for both participants and the research team, but responsiveness to the ethical and practical requirements of the study is crucial in ensuring it can be successfully undertaken. (c) 2017 Wiley Periodicals, Inc.


AIM: Pain management is correlated with pain assessment in the newborn infant. The aim of this study was to assess the impact of a 2-week training program composed of short (20min), repeated training sessions conducted in the unit. METHODS: Pain assessment was studied by means of audits. Each audit included data recorded from the newborn infant's medical charts on the day the infant was admitted to the unit and 3 days before the audit. An audit was performed before the training program and then repeated every month for 12 months. RESULTS: Eighty-eight (53.7%) members of the neonatology staff were trained during the 2-week training program. After the training program, pain assessment "at least once a day" increased by 39.0% and pain assessment "at least once a shift" increased by 21.5% compared to baseline (P<0.05). The effects of the training program were maintained after 12 months (P<0.05). CONCLUSION: A training program with short, repeated sessions conducted in the unit trained 53.7% of the neonatology staff and increased the frequency of pain assessment.

CONTEXT: Clinicians draw upon experiential knowledge to manoeuvre difficult conversations, using tacit knowledge that is difficult to explicitly teach. Instead, learners are taught to communicate through role-play and checklists, both of which are approaches that may fail during moments of clinical complexity. We know that difficult conversations may provoke anxiety in learners, but we know little about how they learn to navigate them. Without a deeper understanding, we may fail to equip learners with the skills to manoeuvre these conversations in practice. METHODS: Using constructivist grounded theory, we applied the sensitising concepts of self-monitoring and reflection-in-action both to explore the process in which trainees engage to navigate difficult conversations and to expand understanding about these theories. We situated our research in the neonatal intensive care unit (NICU), in which difficult conversations are ubiquitous. Fifteen resident and fellow trainees drew rich pictures about difficult conversations, and shared their drawings and experiences during semi-structured interviews. Interview transcripts were analysed using constant comparative analysis. RESULTS: Participants described how they responded when checklist approaches became ineffective during moments of unexpected uncertainty and complexity. For participants, these indeterminate zones of practice triggered a process of seeing families differently and then pausing to understand problems that arose with the checklist-based approach. Throughout this process, learners actively observed others’ communication approaches, negotiated their roles within difficult conversations, and abandoned the checklist to engage differently with families. CONCLUSIONS: Our findings suggest links between the theories of self-monitoring and reflection-in-action, and describe the engagement of both processes in the context of NICU conversations. Self-monitoring may lead to the realisation of an indeterminate zone of practice, after which trainees may respond through reflection-in-action. We recognise that training programmes may need to teach a checklist-based approach as a starting point. We suggest that trainees also be given purposeful opportunities and support to depart from checklists in order to compassionately and flexibly navigate difficult conversations with families.


OBJECTIVES: The goal of this study was to assess pediatric oncology providers’ perceptions of palliative care in order to validate previously identified barriers and facilitators to early integration of a pediatric palliative care team (PCT) in the care of children with cancer. METHODS: A 36-question survey based on preliminary, single-institution data was electronically distributed to pediatric oncology physicians, nurse practitioners, nurses, and social workers nationally. The principal outcomes measured included perceived barriers and facilitators to early integration of pediatric palliative care. Data were analyzed using Rv3.1.2 statistical software. RESULTS: Most respondents agreed that the PCT does not negatively impact the role of the oncologist; however, there were concerns that optimal patient care may be limited by pediatric oncologists’ need to control all aspects of patient care (P < 0.001). Furthermore, oncologists, more than any provider group, identified that the emotional relationship they form with the patients and families they care for, influences what treatment options are offered and how these options are conveyed (P < 0.01). Education and evidence-based research remain important to all providers. Respondents reached consensus that early integration of a PCT would provide more potential benefits than risks and most would not limit access to palliative care based on prognosis. CONCLUSIONS: Overall, providers endorse early integration of the PCT for children with cancer. There remains a continued emphasis on provider and patient education. Palliative care is generally accepted as providing a benefit to children with cancer, though barriers persist and vary among provider groups.


PURPOSE: We report on an in-depth interview and participant observation study that uses data from multiple sources to determine how the involvement of teenagers with leukaemia is understood and enacted in healthcare. In this article, we investigate healthcare professionals’ (HCP) views of teenagers’ involvement in decisions about their care and treatment for leukaemia. METHODS: We conducted participant observation at 98 multi-disciplinary meetings and 95 open-ended, semi-structured interviews and informal conversations with clinical teenage cancer teams at one UK tertiary referral centre. Data were collected over a 9-month period, audio-recorded, transcribed verbatim and analysed using principles of grounded theory. RESULTS: HCP revealed principles relating to the involvement of teenagers with leukaemia in decision making: (1) do the ‘right thing’, (2) act on the care and treatment preferences of the teenager and (3) openly disclose information about the teenagers’ condition. These principles were prioritised and utilised uniquely in each situation, reliant on three mediating factors: (1) family communication styles, (2) stage of illness and (3) nature of the disease. CONCLUSIONS: Specialist haematology teams are aware of the individual, and shifting and situational preferences of teenagers. They follow the lead which teenagers give them with regard to these preferences. If actual practice with regard to the involvement of teenagers is found to be wanting, this study refutes that this should be ascribed to insensitivity on the part of HCP about teenagers informational and decisional role preferences. https://www.ncbi.nlm.nih.gov/pubmed/29218835


OBJECTIVE: Palliative care for children is becoming an important subspecialty of healthcare. Although concurrent administration of curative and palliative care is recommended, timely referral to pediatric palliative care (PPC) services remains problematic. This literature review aims to identify barriers and recommendations for proper implementation of palliative care for children through the looking glass of PPC guidelines. METHOD: To identify studies on PPC guidelines, five databases were searched systematically between 1960 and 2015: Scopus, PubMed, PsycINFO, the Web of Science, and CINAHL. No restrictions were placed on the type of methodology employed in the studies. RESULTS: Concerning barriers, most of the papers focused on gaps within medical practice and the lack of evidence-based research. Common recommendations therefore included: training and education of healthcare staff, formation of a multidisciplinary PPC team, research on the benefits of PPC, and raising awareness about PPC. A small number of publications reported on the absence of clear guidance in PPC documents regarding bereavement care, as well as on the difficulties and challenges involved in multidisciplinary care teams. SIGNIFICANCE OF RESULTS: Our results indicate that a critical assessment of both the research guidelines and medical practice is required in order to promote timely implementation of PPC for pediatric patients. https://www.ncbi.nlm.nih.gov/pubmed/28065197


PURPOSE: Describe practice patterns among obstetrician/gynecologists (OB/GYNs) when caring for women with pregnancy complicated by fetal trisomy 13 (T13) or 18 (T18) and compare these between maternal-fetal medicine (MFM) and non-MFM providers. MATERIALS AND METHODS: We conducted an electronic survey using the American College of Obstetricians and Gynecologists database. Using simple statistics, we describe demographics and practice patterns among respondents and compare those of MFM practitioners with non-MFM providers. RESULTS: The survey was sent to 300 individuals, 161 individuals verified email receipt, and 105 had complete response and were included. The median age was 58 (IQR 53.62). Sixty percent were female, 69% were private practice, and 38% were MFM. All providers were more likely to offer than to recommend antenatal and intrapartum interventions. MFM were more likely to offer growth ultrasounds and neonatal hospice consults (53% vs. 29%, p = .02; 88% vs. 60%, p < .01). During labor, MFM were more likely to offer no fetal heart rate monitoring, (90% vs. 52%, p < .01), 60% of all providers offer breech vaginal delivery; 32% offer cesarean delivery for fetal distress. CONCLUSION: Many providers offer antepartum and intrapartum interventions for pregnancies complicated by T13/18. We recommend that providers elicit each woman’s goals for pregnancies complicated by T13/18 and tailor management options to meet these goals. https://www.ncbi.nlm.nih.gov/pubmed/28629247

Child death review teams (CDRTs) focus on the prevention of child deaths, but a comprehensive understanding of their activities is lacking. This exploratory study addressed this gap through a qualitative analysis of reported CDRT activities using the “spectrum of prevention” framework. We collected state-level CDRT reports published 2006-2015, recorded their activities (n = 193), and coded them using the “spectrum of prevention” framework. The highest percentage (64.2%) of activities was categorized under “fostering coalitions and networks.” We recommend that CDRTs increase their reporting of activities so others can better understand their potential impact on preventing child deaths.


BACKGROUND: Paediatric palliative care is a nuanced area of practice with additional complexities in the context of intellectual disability. There is currently minimal research to guide clinicians working in this challenging area of care. METHOD: This study describes the complex care of children with life-limiting conditions and intellectual disability by means of a literature synthesis and commentary with "best-practice" guide. RESULTS: As few articles concerning children with intellectual disability and palliative care needs were identified by formal systematic review, our expert consensus group has drawn from the paediatric palliative, oncology and adult intellectual disability literature to highlight common clinical challenges encountered in the day-to-day care of children with intellectual disability and life-limiting conditions. CONCLUSION: A longitudinal child- and family-centred approach is key to ensuring best-practice care for families of children with life-limiting conditions and intellectual disability. As highlighted by the great absence of literature addressing this important patient population, further research in this area is urgently required.


PURPOSE: Children with cancer are faced with many challenges related to their disease that disturbs their comfort. The aim of this study was to apply Kolcaba's comfort theory for a child with cancer. DESIGN: A case study design was used. METHODS: We applied Kolcaba's comfort theory for a young boy with cancer who was sad and in discomfort because of intravenous access procedures. Following Kolcaba’s taxonomy of needs for comfort in the spiritual and mental level, we designed a new intervention. FINDINGS: Kolcaba's comfort theory is an appropriate approach which not only helps to assess and evaluate comfort holistically but also assists in performing innovative interventions to provide comfort for children with cancer. CONCLUSIONS: Kolcaba's comfort theory is a practical theory for oncology nurses. CLINICAL RELEVANCE: Nursing theories can improve the quality of clinical care.


PURPOSE: There is an increasing demand for quality palliative care teaching within undergraduate medical education. Studies suggest that many junior doctors feel underprepared to perform end-of-life care. Previous systematic reviews on palliative care teaching within medical schools have identified significant variability and lack of consistency in teaching. This review aims to update the literature on the current status of palliative care teaching to undergraduates within medical schools. METHOD: A systematic review was undertaken on articles published from December 2001 to November 2015 on palliative care teaching for undergraduate medical students. In all, 650 abstract citations were obtained, of which 126 were relevant to the research questions. Thematic analysis was performed on remaining articles according to whether they discussed content and/or methodology of palliative care education, and data collated. RESULTS: There is greater consistency in the content being delivered as part of end-of-life care education within medical schools. The most frequently taught topics include attitudes to death and dying, communication skills, and pain management. Pediatric care and religious/cultural issues are less frequently addressed. Teaching institutions are also utilising a broader range of teaching modalities. CONCLUSION: There is significant progress in palliative care education within medical schools. Ongoing challenges relate to correlating our current practice in medical education to professional recommendations and the expressed needs of junior doctors to practice competent end-of-life care.

Sudden unexpected death in epilepsy (SUDEP) is an important cause of mortality in epilepsy. To date, there is only one published UK study evaluating information provision of SUDEP among parents of children with epilepsy (CWE), and there are no studies published from Asia. Although SUDEP information provision is recommended among parents of CWE, it is uncertain if these recommendations are applicable to Asian countries due to the different cultural attitude towards epilepsy. Our prospective cohort study consisted of multiethnic parents of children with epilepsy (CWE) seen in a tertiary hospital in Malaysia. Information on SUDEP was delivered to parents using an epilepsy educational software program. Participants completed a set of standardized questionnaire and Depression Anxiety Stress Scales-Short Form (DASS-21) immediately after and retested 3-6 months after the SUDEP information provision. A total of 127 parents (84 mothers) participated in the study. The CWE consisted of 3 ethnic groups (38% Malay, 30% Chinese, 32% Indian) with a mean age of 9.6 years. Majority (70.9%) felt positive after SUDEP information provision, 90.6% wanted SUDEP discussion for themselves with 70.1% wanted SUDEP discussion with their child, and a lower proportion (58.3%) would discuss SUDEP with their child. None of the participants reported increased symptoms of depression, stress or anxiety attributed to SUDEP information provision. Most parents took steps to reduce SUDEP risk, and most parents did not report an impact on their own functioning. However, there was an increase in parental report over time of impact on their child’s functioning following SUDEP information provision (P < 0.05). In conclusion, most Malaysian parents of CWE wanted SUDEP information. Following SUDEP information disclosure, majority did not report negative emotions; however, an increase in parents over time reported an impact on their child. Our findings reiterate that provision of SUDEP information should form part of care of CWE and parents should receive ongoing support as they undergo a period of parenting adjustment when dealing with the information provided.


AIM: To characterise neonatal intensive care unit (NICU) staff perceptions regarding factors which may lead to more challenging staff-parent interactions, and beneficial strategies for working with families with whom such interactions occur. METHODS: A survey of 168 physician and nursing staff at two NICUs in American teaching hospitals inquired about their perceptions of challenging parent-staff interactions and situations in which such interactions were likely to occur. RESULTS: From a medical perspective, staff perceptions of challenging interactions were noted when infants had recent decompensation, high medical complexity, malformations or long duration of stay in the NICU. From a psychological/social perspective, a high likelihood of challenging interactions was noted with parents who were suspicious, interfere with equipment, or parents who hover in the NICU, express paranoid or delusional thoughts, repeat questions, perceive the staff as inaccessible, are managing addictions, or who require child protective services involvement. Frequent family meetings, grieving opportunities, education of parents, social work referrals, clearly defined rules, partnering in daily care and support groups were perceived as the most beneficial strategies for improving difficult interactions. CONCLUSION: This study delineates what staff perceive as challenging interactions and provides support for an educational and interventional role that incorporates mental health professionals.


https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5722497/


BACKGROUND The decision to utilize antimicrobials in end-of-life situations is complex. Understanding the reasons why physicians prescribe antimicrobials in this patient population is important for informing the design of antimicrobial stewardship interventions. METHODS A 51-item survey containing both closed and open-ended
questions on end-of-life antimicrobial use was administered to physicians affiliated with the University of Pennsylvania and Children's Hospital of Philadelphia from January through April 2017. A mixed-methods approach was used to analyze responses. RESULTS Of 637 physicians surveyed, 283 responses (44.4%) were received. Most (86.2%) physicians believed that respecting a patient’s wish to continue antimicrobials was important. Approximately half of physicians (49.8%) believed that antimicrobial use at the end of life contributes to resistance. A higher proportion of pediatricians would often or always continue antimicrobial treatment for active infections and for hospice patients whose death was imminent compared to adult physicians (P < 0.001). Analysis of free-text responses revealed additional reasons why physicians may continue antimicrobials at end of life, including meeting family expectations, wanting to avoid the perception of “giving up,” uncertainty about prognosis, and reducing patient pain or discomfort. CONCLUSIONS Physician decision making concerning antimicrobial use in patients at the end of life is multifactorial. Clinicians may overweight the benefits of antimicrobial therapy in end-of-life situations and view the importance of adhering to stewardship policies differently. Pediatric and adult clinicians have different approaches to this patient population. Better understanding of the complex decision making that occurs in the end-of-life patient population can help guide antimicrobial stewardship policies and improve patient care. Infect Control Hosp Epidemiol 2018;39:383-390. https://www.ncbi.nlm.nih.gov/pubmed/29428002


OBJECTIVE: Perinatal loss (stillbirth or early infant death) is often a sudden, unexpected event for families. We evaluated who communicates the loss to the parents and who is there for support at the delivery or death. STUDY DESIGN: We conducted a mail survey of 900 bereaved and 500 live-birth mothers to assess emotional, physical and reproductive health outcomes. RESULTS: We had a 44% response rate at 9 months after birth or loss from 377 bereaved mothers and 232 with surviving infants. Bereaved women were less likely to have hospital staff or family members present at delivery. African-American (versus Caucasian) mothers were half as likely to have first heard about their stillbirth from a physician or midwife. CONCLUSION: This is the first large study documenting who communicates perinatal death to families and who is present for support. Hospitals should be aware that many bereaved families may lack support at critical times. https://www.ncbi.nlm.nih.gov/pubmed/29192693


INTRODUCTION: Pediatric surgeons are often involved in the management of severely or terminally ill patients. However, articles addressing their specific roles in the context of palliative care are almost inexistential. We sought to characterize the involvement of pediatric surgeons caring for children near end of life. METHODS: Chart review of children who had a procedure under general anesthesia within 6 months of their death over a five-year period at a tertiary children's hospital (excluding traumas and neonatology cases). In addition to demographic and clinical data, we recorded the aim of the procedures performed, the involvement of the palliative care service, and presence of DNAR orders. RESULTS: The analysis included 83 patients (mean age: 8 years). Forty-four children had more than one procedure (range 2-10). Pediatric palliative care service was involved in 66 cases (80%). A majority of patients had cancer (50%), and the most frequent cause of death was oncologic progression (46%). Ten patients died of a complication following their intervention. The aim of the procedure was palliative in 48 cases (29 for symptoms control and 19 to facilitate care), diagnostic in 16, and curative in 19. Forty-five procedures were performed urgently and 14 despite DNAR orders. CONCLUSION: Surgeon involvement with children near end of life is not infrequent. The procedures performed are varied and can be categorized according to their aim. Lack of formal palliative care training by surgeons highlights the need for increased collaboration with palliative care services to provide children optimal care when they need it most. LEVEL OF EVIDENCE: IV. https://www.ncbi.nlm.nih.gov/pubmed/29526348


BACKGROUND: Parental presence in the neonatal intensive care unit (NICU) is essential for families to participate in infant care and prepare them to transition from hospital to home. Nurses are the principal caregivers in the
NICU. The nurse work environment may influence whether parents spend time with their hospitalized infants. PURPOSE: To examine the relationship between the NICU work environment and parental presence in the NICU using a national data set. METHODS: We conducted a cross-sectional, observational study of a national sample of 104 NICUs, where 6060 nurses reported on 15,233 infants cared for. Secondary analysis was used to examine associations between the Practice Environment Scale of the Nursing Work Index (PES-NWI) (subscale items and with a composite measure) and the proportion of parents who were present during the nurses’ shift. RESULTS: Parents of 60% (SD = 9.7%) of infants were present during the nurses’ shift. The PES-NWI composite score and 2 domains—Nurse Participation in Hospital Affairs and Manager Leadership and Support—were significant predictors of parental presence. A 1 SD higher score in the composite or either subscale was associated with 2.5% more parents being present. IMPLICATIONS FOR PRACTICE: Parental presence in the NICU is significantly associated with better nurse work environments. NICU practices may be enhanced through enhanced leadership and professional opportunities for nurse managers and staff. IMPLICATIONS FOR RESEARCH: Future work may benefit from qualitative work with parents to illuminate their experiences with nursing leaders and nurse-led interventions in the NICU and design and testing of interventions to improve the NICU work environment.


CONTEXT: Infants of age less than one year have the highest mortality rate in pediatrics. The American Academy of Pediatrics published guidelines for palliative care in 2013; however, significant variation persists among local protocols addressing neonatal comfort care at the end-of-life (EOL). OBJECTIVES: The purpose of this study was to evaluate current neonatal EOL comfort care practices and clinician satisfaction across America. METHODS: After institutional review board approval (516005), an anonymous, electronic survey was sent to members of the American Academy of Pediatrics Section on Neonatal-Perinatal Medicine. Members of the listserv include neonatologists, neonatal fellow physicians, neonatal nurses, and neonatal nurse practitioners from across America (U.S. and Canada). RESULTS: There were 346/3000 (11.5%) responses with wide geographic distribution and high levels of intensive care responding (46.1% Level IV, 50.9% Level III, 3.0% Level II). Nearly half (45.2%) reported that their primary institution did not have neonatal comfort care guidelines. Of those reporting institutional neonatal comfort care guidelines, 19.1% do not address pain symptom management. Most guidelines also do not address gastrointestinal distress, anxiety, or secretions. Thirty-nine percent of respondents stated that their institution did not address physician compassion fatigue. Overall, 91.8% of respondents felt that their institution would benefit from further education/training in neonatal EOL care. CONCLUSION: Across America, respondents confirmed significant variation and verified many institutions do not formally address neonatal EOL comfort care. Institutions with guidelines commonly appear to lack crucial areas of palliative care including patient symptom management and provider compassion fatigue. The overwhelming majority of respondents felt that their institutions would benefit from further neonatal EOL care training.


OBJECTIVES: When contemplating tracheostomy placement in a pediatric patient, a family-physician conference is often the setting for the disclosure of risks and benefits of the procedure. Our objective was to compare benefits and risks of tracheostomy presented during family-physician conferences to an expert panel’s recommendations for what should be presented. DESIGN: We conducted a retrospective review of 19 transcripts of audio-recorded family-physician conferences regarding tracheostomy placement in children. A multicenter, multidisciplinary expert panel of clinicians was surveyed to generate a list of recommended benefits and risks for comparison. Primary analysis of statements by clinicians was qualitative. SETTING: Single-center PICU of a tertiary medical center. SUBJECTS: Family members who participated in family-physician conferences regarding tracheostomy placement for a critically ill child from April 2012 to August 2014. MEASUREMENTS AND MAIN RESULTS: We identified 300 physician statements describing benefits and risks of tracheostomy. Physicians were more likely to discuss benefits than risks (72% vs 28%). Three broad categories of benefits were identified: 1) tracheostomy would limit the impact of being in the PICU (46%); 2) perceived obstacles of tracheostomy can be overcome (34%); and 3) tracheostomy optimizes respiratory health (20%). Risks fell into two categories: tracheostomy involves a big commitment (71%), and it has complications (29%). The expert panel’s recommendations were similar to risks and benefits discussed during family conferences; however, they suggested physicians present an equal balance of discussion of risks and benefits. CONCLUSIONS: When discussing tracheostomy placement,
physicians emphasized benefits that are shared by physicians and families while minimizing the risks. The expert panel recommended a balanced approach by equally weighing risks and benefits. To facilitate educated decision making, physicians should present a more extensive range of risks and benefits to families making this critical decision.


Nursing education needs to prepare students for care of dying patients. The aim of this study was to describe the development of nursing students’ attitudes toward caring for dying patients and their perceived preparedness to perform end-of-life care. A longitudinal study was performed with 117 nursing students at six universities in Sweden. The students completed the Frommelt Attitude Toward Care of the Dying Scale (FATCOD) questionnaire at the beginning of first and second year, and at the end of third year of education. After education, the students completed questions about how prepared they felt by to perform end-of-life care. The total FATCOD increased from 126 to 132 during education. Five weeks’ theoretical palliative care education significantly predicted positive changes in attitudes toward caring for dying patients. Students with five weeks’ theoretical palliative care training felt more prepared and supported by the education to care for a dying patient than students with shorter education. A minority felt prepared to take care of a dead body or meet relatives.


On the day of his birth in 1971, David Vetter was “temporarily” placed in a sterile isolator to wait for a bone marrow donor who would cure his Severe Combined Immunodeficiency Syndrome. After enduring 12 years in isolation, David, now known to the world as “The Boy in the Bubble”, received a bone marrow transplant from his unmatched sister and died 4 months later. Like Severe Combined Immunodeficiency Syndrome, pediatric heart failure is a rare and life-threatening condition for which organ transplantation is often the only option for survival. Nearly 15 years ago, the Berlin Heart EXCOR ventricular assist device was developed, as the isolator was for David, to be a bridge to transplantation for infants and children with unstable heart failure. Yet, when symptoms progress faster than the patient’s suitability for transplant can be evaluated, this device may be implanted as a “bridge-to-decision” while a transplant evaluation is completed. Therein lies the potential for history to repeat itself. As biomedical science carries us ever forward in our ability to sustain life, we must always bear in mind that the miraculous EXCOR must not be a machine that simply sustains the circulation. It must be, and always remain, a bridge to somewhere.


OBJECTIVE: To assess the prevalence of symptoms of acute stress reactions (ASR) and post-traumatic stress disorder (PTSD) in paediatric trainees following their involvement in child death. DESIGN: A survey designed to identify trainees’ previous experiences of child death combined with questions to identify features of PTSD. Quantitative interpretation was used alongside a chi(2) test. A p value of <0.05 was considered significant. SETTING: 604 surveys were distributed across 13 UK health education deaneries. PARTICIPANTS: 303/604 (50%) of trainees completed the surveys. RESULTS: 251/280 (90%) of trainees had been involved with the death of a child, although 190/284 (67%) had no training in child death. 118/248 (48%) of trainees were given a formal debrief session following their most recent experience. 203/251 (81%) of trainees reported one or more symptoms or behaviours that could contribute to a diagnosis of ASR/PTSD. 23/251 (9%) of trainees met the complete criteria for ASR and 13/251 (5%) for PTSD. Attending a formal debrief and reporting feelings of guilt were associated with an increase in diagnostic criteria for ASR/PTSD (p=0.036 and p<0.001, respectively). CONCLUSIONS: Paediatric trainees are at risk of developing ASR and PTSD following the death of a child. The feeling of guilt should be identified and acknowledged to allow prompt signposting to further support, including psychological assessment or intervention if required. Clear recommendations need to be made about the safety of debriefing sessions as, in keeping with existing evidence, our data suggest that debrief after the death of a child may be associated with

AIMS: To determine if there are brain activity differences between paediatric intensive care nurses and allied health professionals during pain intensity rating tasks and test whether these differences are related to the population observed (infant or adult) and professional experience. BACKGROUND: The underestimation of patients' pain by healthcare professionals has generally been associated with patterns of change in neural response to vicarious pain, notably reduced activation in regions associated with affective sharing and increased activation in regions associated with regulation, compared with controls. Paediatric nurses, however, have recently been found to provide higher estimates of infants' pain in comparison to allied health controls, suggesting that changes in neural response of this population might be different than other health professionals. DESIGN: Cross-sectional study. METHODS: Functional MRI data were acquired from September 2014-June 2015 and used to compare changes in brain activity in 27 female paediatric care nurses and 24 allied health professionals while rating the pain of infants and adults in a series of video clips. RESULTS: Paediatric nurses rated infant and adult pain higher than allied health professionals, but the two groups' neural response only differed during observation of infant pain; paediatric nurses mainly showed significantly less activation in the medial prefrontal cortex (linked to cognitive empathy) and in the left anterior insula and inferior frontal cortex (linked to affective sharing). CONCLUSIONS: Patterns of neural activity to vicarious pain may vary across healthcare professions and patient populations and the amount of professional experience might explain part of these differences. 


OBJECTIVE: Compassion fatigue (CF) is secondary traumatic distress experienced by providers from contact with patients' suffering. Burnout (BO) is job-related distress resulting from uncontrollable workplace factors that manifest in career dissatisfaction. Compassion satisfaction (CS) is emotional fulfillment derived from caring for others. The literature on BO in healthcare providers is extensive, whereas CF and CS have not been comprehensively studied. Because of ongoing exposure to patient and family distress, pediatric palliative care (PPC) providers may be at particular risk for CF. We conducted a cross-sectional pilot study of CF, BO, and CS among PPC providers across the United States. METHOD: The Compassion Fatigue and Satisfaction Self-Test for Helpers and a questionnaire of professional and personal characteristics were distributed electronically and anonymously to PPC physicians and nurses. Logistic and linear regression models for CF, BO, and CS as a function of potential risk factors were constructed. RESULTS: The survey response rate was 39%, primarily consisting of female, Caucasian providers. The prevalence of CF, BO, and CS was 18%, 12%, and 25%, respectively. Distress about a "clinical situation," physical exhaustion, and personal loss were identified as significant determinants of CF. Distress about "coworkers," emotional depletion, social isolation, and "recent involvement in a clinical situation in which life-prolonging activities were not introduced" were significant determinants of BO. Physical exhaustion, personal history of trauma, "recent involvement in a clinical situation in which life-prolonging activities were not introduced," and not discussing distressing issues were significant predictors of lower CS scores. Significance of results CF and BO directly influence the well-being and professional performance of PPC providers. To provide effective compassionate care to patients, PPC providers must be attentive to predictors of these phenomena. Further work is needed to explore additional causes of CF, BO, and CS in PPC providers as well as potential interventions.


OBJECTIVES: In the context of serious or life-limiting illness, pediatric patients and their families are faced with difficult decisions surrounding appropriate resuscitation efforts in the event of a cardiopulmonary arrest. Code status orders are one way to inform end-of-life medical decision making. The objectives of this study are to evaluate the extent to which pediatric providers have knowledge of code status options and explore the
association of provider role with (1) knowledge of code status options, (2) perception of timing of code status discussions, (3) perception of family receptivity to code status discussions, and (4) comfort carrying out code status discussions. DESIGN: Nurses, trainees (residents and fellows), and attending physicians from pediatric units where code status discussions typically occur completed a short survey questionnaire regarding their knowledge of code status options and perceptions surrounding code status discussions. SETTING: Single center, quaternary care children's hospital. MEASUREMENTS AND MAIN RESULTS: 203 nurses, 31 trainees, and 29 attending physicians in 4 high-acuity pediatric units responded to the survey (N = 263, 90% response rate). Based on an objective knowledge measure, providers demonstrate poor understanding of available code status options, with only 22% of providers able to enumerate more than two of four available code status options. In contrast, provider groups self-report high levels of familiarity with available code status options, with attending physicians reporting significantly higher levels than nurses and trainees (p = 0.0125). Nurses and attending physicians show significantly different perception of code status discussion timing, with majority of nurses (63.4%) perceiving discussions as occurring “too late” or “much too late” and majority of attending physicians (55.6%) perceiving the timing as “about right” (p<0.0001). Attending physicians report significantly higher comfort having code status discussions with families than do nurses or trainees (p<0.0001). Attending physicians and trainees perceive families as more receptive to code status discussions than nurses (p<0.001 and p = 0.0018, respectively). CONCLUSIONS: Providers have poor understanding of code status options and differ significantly in their comfort having code status discussions and their perceptions of these discussions. These findings may reflect inherent differences among providers, but may also reflect discordant visions of appropriate care and function as a potential source of moral distress. Lack of knowledge of code status options and differences in provider perceptions are likely barriers to quality communication surrounding end-of-life options.


OBJECTIVES: To measure the level of moral distress in PICU and neonatal ICU health practitioners, and to describe the relationship of moral distress with demographic factors, burnout, and uncertainty. DESIGN: Cross-sectional survey. SETTING: A large pediatric tertiary care center. SUBJECTS: Neonatal ICU and PICU health practitioners with at least 3 months of ICU experience. INTERVENTIONS: A 41-item questionnaire examining moral distress, burnout, and uncertainty. MEASUREMENTS AND MAIN RESULTS: The main outcome was moral distress measured with the Revised Moral Distress Scale. Secondary outcomes were frequency and intensity Revised Moral Distress Scale subscores, burnout measured with the Maslach Burnout Inventory depersonalization subscale, and uncertainty measured with questions adapted from Mishel's Parent Perception of Uncertainty Scale. Linear regression models were used to examine associations between participant characteristics and the measures of moral distress, burnout, and uncertainty. Two-hundred six analyzable surveys were returned. The median Revised Moral Distress Scale score was 96.5 (interquartile range, 69-133), and 58% of respondents reported significant work-related moral distress. Revised Moral Distress Scale items involving end-of-life care and communication scored highest. Moral distress was positively associated with burnout (r = 0.27; p < 0.001) and uncertainty (r = 0.04; p = 0.008) and inversely associated with perceived hospital supportiveness (r = 0.18; p < 0.001). Nurses reported higher moral distress intensity than physicians (Revised Moral Distress Scale intensity subscores: 57.3 vs 44.7; p = 0.002). In nurses only, moral distress was positively associated with increasing years of ICU experience (p = 0.02) and uncertainty about whether their care was of benefit (r = 0.11; p < 0.001) and inversely associated with uncertainty about a child’s prognosis (r = 0.03; p = 0.03). CONCLUSIONS: In this single-center, cross-sectional study, we found that moral distress is present in PICU and neonatal ICU health practitioners and is correlated with burnout, uncertainty, and feeling unsupported.


BACKGROUND: Postmortem examination is the single most useful investigation in providing information to parents about why their baby or child died. Despite this, uptake remains well below the recommended 75%.

OBJECTIVE: To address the question ‘what are the barriers and motivators to perinatal, prenatal and paediatric PM examination?’ SEARCH STRATEGY: Key databases including Pubmed and CINAHL; Cochrane library, websites of relevant patient organisations, hand search of key journals, first and last authors and references. SELECTION CRITERIA: Peer-reviewed qualitative, quantitative or mixed methods research examining factors affecting uptake or decline of perinatal or paediatric postmortem examination. DATA COLLECTION AND ANALYSIS: Narrative
synthesis; findings were compared across studies to examine interrelations. MAIN RESULTS: Seven major themes describing barriers to postmortem uptake were identified: dislike of invasiveness, practicalities of the procedure, organ retention issues, protective parenting, communication and understanding, religion and culture and professional or organisational barriers. Six major themes related to factors which facilitated parental consent were identified: desire for information, contributing to research, coping and well-being, respectful care, minimally invasive options, and policy and practice. There were a number of themes in the literature that reflected best practice. CONCLUSION: Findings highlight the need for better health professional education and the fact some concerns may be mitigated if less invasive methods of postmortem were routinely available. New consent packages and codes of practice may have a positive impact on perception of examination after death. The landscape is changing; further research is necessary to assess the impact on postmortem uptake rates.
TWEETABLE ABSTRACT: Systematic review to explore the barriers and motivators to perinatal, prenatal and paediatric postmortem examination.


OBJECTIVE: To assess health professionals' and coroners' attitudes towards non-minimally and minimally invasive autopsy in the perinatal and paediatric setting. METHODS: A qualitative study using semi-structured interviews. Data were analysed thematically. RESULTS: Twenty-five health professionals (including perinatal/paediatric pathologists and anatomical pathology technologists, obstetricians, fetal medicine consultants and bereavement midwives, intensive care consultants and family liaison nurses, a consultant neonatologist and a paediatric radiologist) and four coroners participated. Participants viewed less invasive methods of autopsy as a positive development in perinatal and paediatric care that could increase autopsy rates. Several procedural and psychological benefits were highlighted including improved diagnostic accuracy in some circumstances, potential for faster turnaround times, parental familiarity with imaging and laparoscopic approaches, and benefits to parents and faith groups who object to invasive approaches. Concerns around the limitations of the technology such not reaching the same levels of certainty as full autopsy, unsuitability of imaging in certain circumstances, the potential for missing a diagnosis (or misdiagnosis) and de-skilling the workforce were identified. Finally, a number of implementation issues were raised including skills and training requirements for pathologists and radiologists, access to scanning equipment, required computational infrastructure, need for a multidisciplinary approach to interpret results, cost implications, equity of access and acceptance from health professionals and hospital managers. CONCLUSION: Health professionals and coroners viewed less invasive autopsy as a positive development in perinatal and paediatric care. However, to inform implementation a detailed health economic analysis and further exploration of parental views, particularly in different religious groups, are required.


OBJECTIVE: Little is known about the current views and practices of healthcare professionals (HCPs) in Sub-Saharan Africa (SSA) regarding delivery of hospital palliative care. The present qualitative study explored the views of nursing staff and medical professionals on providing palliative and end-of-life care (EoLC) to hospital inpatients in Tanzania. METHOD: Focus group discussions were conducted with a purposive sample of HCPs working on the medical and paediatric wards of the Kilimanjaro Christian Medical Centre, a tertiary referral hospital in northern Tanzania. Transcriptions were coded using a thematic approach. RESULTS: In total, 32 healthcare workers were interviewed via 7 focus group discussions and 1 semistructured interview. Four major themes were identified. First, HCPs held strong views on what factors were important to enable individuals with a life-limiting diagnosis to live and die well. Arriving at a state of "acceptance" was the ultimate goal; however, they acknowledged that they often fell short of achieving this for inpatients. Thus, the second theme involved identifying the "barriers" to delivering palliative care in hospital. Another important factor identified was difficulty with complex communications, particularly "breaking bad news," the third theme. Fourth, participants were divided about their personal preferences for "place of EoLC." but all emphasized the benefits of the hospital setting so as to enable better symptom control. SIGNIFICANCE OF RESULTS: Despite the fact that all the HCPs interviewed were regularly involved in providing palliative and EoLC, they had received limited formal training in its provision, although they identified such training as a universal requirement. This training gap is likely to be present across much of SSA. Palliative care training, particularly in terms of communication skills, should be
comprehensively integrated within undergraduate and postgraduate education. Research is needed to develop culturally appropriate curricula to equip HCPs to manage the complex communication challenges that occur in caring for a diverse inpatient group with palliative care needs.


INTRODUCTION: In 2013, 55,000 infants and children, aged 0 to 14, died in the United States. Nearly 7,000 of those deaths were attributed to traumatic causes. A child's death significantly affects emergency service personnel (ESP) caring for children and families. This study explores the lived experience of ESP involved in unsuccessful pediatric resuscitation efforts and how this experience affects them professionally and personally.

METHODS: A phenomenologic approach guided this study. Using an open-ended format, an interview was conducted with a purposive sample of ESP who experienced unexpected pediatric death. Eight ESP participated in semistructured, face-to-face interviews, ranging in length from 35 to 75 minutes. The research question asked: "What is it like for you when a child dies after an unsuccessful resuscitation attempt?" Data were analyzed using thematic analysis. RESULTS: Van Manen's 4 existentials guided this study, and 10 subthemes emerged that included: "what if," "dying before my eyes," "team," "what if it was were my child?/being a parent," "the environment," "being trapped," "wounded healer," "education," "anger," and "coping." DISCUSSION: This study explores the experience of ESP involved in unsuccessful pediatric resuscitation that resulted in unexpected pediatric death and ESP's perceptions of this experience: thoughts of loss, a sense of anger, and a lack of preparation to cope with unexpected pediatric death and the unknowns of life.


Midwifery students with perinatal palliative care education develop a skillset to provide holistic midwifery care to women and families who are experiencing stillbirth or life-limiting fetal diagnoses. This paper presents a model of perinatal palliative care in a United States midwifery education program. By utilizing evidence based practices and national programs, perinatal palliative care can be threaded through midwifery curricula to achieve international standards of practice and competencies. Most importantly, enhancing perinatal palliative care education will better prepare future midwives for when a birth outcome is not what was expected at the outset of a pregnancy.


BACKGROUND: The majority of patients desire all available prognostic information, but some physicians hesitate to discuss prognosis. The objective of the current study was to examine outcomes of prognostic disclosure among the parents of children with cancer. METHODS: The authors surveyed 353 parents of children with newly diagnosed cancer at 2 tertiary cancer centers, and each child's oncologist. Using multivariable logistic regression, the authors assessed associations between parental report of elements of prognosis discussions with the oncologist (quality of information/communication and prognostic disclosure) and potential consequences of these discussions (trust, hope, peace of mind, prognostic understanding, depression, and anxiety). Analyses were stratified by oncologist-reported prognosis. RESULTS: Prognostic disclosure was not found to be associated with increased parental anxiety, depression, or decreased hope. Among the parents of children with less favorable prognoses (<75% chance of cure), the receipt of high-quality information from the oncologist was associated with greater peace of mind (odds ratio [OR], 5.23; 95% confidence interval [95% CI], 1.81-15.16) and communication-related hope (OR, 2.54; 95% CI, 1.00-6.40). High-quality oncologist communication style was associated with greater trust in the physician (OR, 2.45; 95% CI, 1.09-5.48) and hope (OR, 3.01; 95% CI, 1.26-7.19). Accurate prognostic understanding was less common among the parents of children with less favorable prognoses (OR, 0.39; 95% CI, 0.17-0.88). Receipt of high-quality information, high-quality communication, and
Inadequate resuscitation leads to death or brain injury. Recent recommendations for resuscitation team training to complement knowledge and skills training highlighted the need for development of an effective team resuscitation training session. This study aimed to evaluate and revise an interprofessional team training session which addressed roles and performance during provision of paediatric resuscitation, through incorporation of real-time, real team simulated training episodes. This study was conducted applying the principles of action research. Two cycles of data collection, evaluation and refinement of a 30-40 minute resuscitation training session for doctors and nurses occurred. Doctors and nurses made up 4 groups of training session participants. Their responses to the training were evaluated through thematic analysis of rich qualitative data gathered in focus groups held immediately after each training session. Major themes included the importance of realism, teamwork, and reflective learning. Findings informed important training session changes. These included: committed in-situ
training; team diversity; realistic resources; role flexibility, definition and leadership; increased debriefing time and the addition of a team goal. In conclusion, incorporation of interprofessional resuscitation training which addresses team roles and responsibilities into standard medical and nursing training will enhance preparedness for participation in paediatric resuscitation.


OBJECTIVE: To analyse the referral patterns of perinatal patients referred to a specialist palliative care service (SPCS), their demographics, diagnoses, duration of illness, place of death and symptom profile. DESIGN: A retrospective chart review of all perinatal referrals over a 4-year period to the end of 2015. SETTING: A consultant-led paediatric SPCS at Our Lady's Children's Hospital, Crumlin, Dublin, and the Coombe Women & Infants University Hospital, Dublin. RESULTS: 83 perinatal referrals were received in a 4-year period. Chromosomal abnormalities accounted for 35% of diagnoses, congenital heart disease 25%, complex neurological abnormalities 11% and renal agenesis 4%. 22 referrals (26.5%) were made antenatally, with 61 (73.5%) postnatally. Of the postnatal referrals, 27 (44%) were asymptomatic on referral. An opioid medication was recommended (regularly or as required) in 46 cases. Symptom control was achieved without dose titration in 43 of these cases (93%). Of 47 deaths in this group referred postnatally, 22 of these (47%) died at home with support from community teams. Discharge home for best supportive care required complex interagency communication and cooperation. CONCLUSIONS: Perinatal palliative care requires effective multidisciplinary work, whether delivered in the inpatient setting or in the community. With appropriate support, end-of-life care can be delivered in the community.


BACKGROUND: Multidisciplinary teamwork is considered central to pediatric palliative care. Although different studies state that volunteers play an essential role in palliative care, little is known about the collaboration between volunteers and staff. AIM: This study aims to explore and compare the perspectives of volunteers and staff regarding collaboration in a pediatric palliative care unit. DESIGN: A mixed-methods approach was chosen to appropriately reflect the complex aspects of collaboration. SETTING/PARTICIPANTS: Both face-to-face interviews with staff who work together with volunteers and a group discussion with all volunteers were conducted. These were supplemented by 2 questionnaires designed for this study that examined participants' characteristics and their estimation of what information volunteers need before they meet a patient. RESULTS: Nine staff members and 7 volunteers participated in this study. Their ideas of collaboration could be grouped into 3 categories: (i) factual level of collaboration, (ii) relationship level of collaboration, and (iii) overall appraisal of collaboration (suggestions for improvement). CONCLUSION: Communication can be considered a key factor in successful collaboration between volunteers and staff. Because many patients in pediatric palliative care units are not able to communicate verbally, good information flow between volunteers and staff is crucial for ensuring quality patient care. Moreover, communication is the key to establishing a team philosophy by clarifying roles and building relationships between volunteers and staff.


The clinical practicum is one of the most anticipated components of the nursing program for nursing students. However, the practicum can be anxiety producing for students, especially when it is their first placement in an emotional demanding setting like pediatric oncology unit. Taking care of children with cancer and who are facing the death trajectory is complex and demanding not only for students but also for the experienced nurse. In this qualitative research, the purpose was to explore senior student perceptions and self-reflective accounts of what it was like to care for children with cancer and their family throughout the course of their first practicum on a pediatric oncology unit that also provided children palliative care as needed. Data from the self-reflective journals and interviews were analyzed together using conventional content analysis. The three resultant categories that emerged: state of shock and getting lost, walking in to a mind shaking world and finding the way provided in-
RATIONALE: The Burkitt lymphoma (BL) is a very aggressive B-cell non-Hodgkin’s lymphoma. It accounts for 34% of lymphoma cases in children. PATIENT CONCERNS: We present the case of a 6-year-old boy diagnosed with BL, who presented multiple contrasting elements of the disease: silent symptomatology, without involvement of the bone marrow at first, but with multiorgan infiltration and a fast evolution, despite starting the treatment shortly after the symptoms appeared. DIAGNOSES: He was diagnosed with BL after immunophenotyping from the pleural fluid. INTERVENTIONS: After a week from admission, chemotherapy was initiated according to protocol NH-BFM therapeutic group III- cytoreductive phase in the acute care ward and subsequently the AA 24 treatment. OUTCOMES: Following the treatment, the patient developed medullary aplasia and cutaneous toxicity. The patient’s general state remained severe during the hospitalization. LESSONS: Even though the prognosis of BL has improved over time (up to 90% survival rate), in this case the evolution was unfavorable. In our patient, the symptoms appeared abruptly. They appeared late in the phase of multiple-organ dissemination, which generated the pessimistic prognosis.

BACKGROUND: The Institute of Medicine and the American Academy of Pediatrics has called for improvement in education and training of pediatricians in pediatric palliative care (PPC). Given the shortage of PPC physicians and the immediate need for PPC medical education, this study reports the outcomes of a problem-based learning (PBL) module facilitated by academic general and subspecialty pediatric faculty (non-PPC specialists) to third year medical students. Objectives/Setting: To test the effectiveness of a PPC-PBL module on third year medical students’ and pediatric faculty’s declarative knowledge, attitudes toward, perceived exposure, and self-assessed competency in PPC objectives. DESIGN: A PBL module was developed using three PPC learning objectives as a framework: define core concepts in palliative care; list the components of a total pain assessment; and describe key principles in establishing therapeutic relationships with patients. A PPC physician and nurse practitioner guided pediatric faculty on facilitating the PPC-PBL. In Part 1, students identified domains of palliative care for a child with refractory leukemia and self-assigned questions to research and present at the follow-up session. In Part 2, students were expected to develop a care plan demonstrating the three PPC objectives. MEASUREMENTS: Measures included a knowledge exam and a survey instrument to assess secondary outcomes. RESULTS: Students’ declarative knowledge, perceived exposure, and self-assessed competency in all three PPC learning objectives improved significantly after the PPC-PBL, p = 0.002, p < 0.001, and p < 0.001, respectively. There were no significant differences in faculty knowledge test scores from baseline to follow-up, but scores were generally high (median >80%). Students and faculty rated palliative care education as “important or very important” at baseline and follow-up. CONCLUSIONS: This study suggests that key concepts in PPC can be taught to medical students utilizing a PBL format and pediatric faculty resulting in improved knowledge and self-assessed competency in PPC.

OBJECTIVE: Prognostication of survival is difficult in children with life-limiting illnesses because of the rarity of these conditions and technological advances improving survival. The objective of this article is to describe the characteristics of children with life-limiting illnesses who survived longer than the expectations of health-care providers. STUDY DESIGN: This is a retrospective cohort study conducted in a tertiary-care children’s hospital in North Carolina. “Unexpected survivors,” defined as children who survived despite a prognosis of imminent death or significantly longer than prognosticated by health-care providers, were identified from among 349 children enrolled in a pediatric palliative care program between March 2008 and October 2012. Children’s clinical courses


were followed until September 2015 or their death. RESULTS: Eighteen (5%) children were identified as unexpected survivors; 17 (10 girls and 7 boys) were included. Congenital anomalies were the most common diagnoses. Neonatal intensive care unit was the most frequent setting of prognostication. Thirteen children used some form of medical technology at the time of prognostication. Eleven children received hospice services. Eight died during the observation period but survived significantly longer than expected (median survival time 1.5 years), and 9 survived beyond the observation period (median survival time 5.9 years). CONCLUSIONS: Unexpected survivors are a small group of children with life-limiting conditions. Clinicians should be aware of the possibility of prognostic inaccuracy, able to communicate prognostic uncertainty to parents, and engage supportive services when prognosticating poor survival. Prospective studies are needed to understand outcomes of children with life-limiting illnesses of uncertain prognosis.


Given the broad focus of pediatric palliative care (PPC) on the physical, emotional, and spiritual needs of children with potentially life-limiting illnesses and their families, PPC research requires creative methodological approaches. This manuscript, written by experienced PPC researchers, describes issues encountered in our own areas of research and the novel methods we have identified to target them. Specifically, we discuss potential approaches to: assessing symptoms among nonverbal children, evaluating medical interventions, identifying and treating problems related to polypharmacy, addressing missing data in longitudinal studies, evaluating longer-term efficacy of PPC interventions, and monitoring for inequities in PPC service delivery.


INTRODUCTION: Adequate communication by medical personnel is especially important at certain points during the treatment of childhood cancer patients. AIM: To investigate the timing and manner of communication with parents concerning the introduction of palliative care in pediatric oncology. METHOD: Structured interviews, containing 14 questions, were carried out with physicians working in pediatric oncology (n = 22). Codes were generated inductively with the aid of Atlas.ti 6.0 software. RESULTS: Interviews show a tendency of a one-step transition to palliative care following curative therapy. Another expert is usually involved in communication, most likely a psychologist. Regarding communication, there are expressions utilized or avoided, such as expressing clarity, self-defense and empathy. The communication of death and dying was the most contradictory. CONCLUSION: This was the first investigation regarding communication in pediatric palliative care in Hungary. Our results show that a modern perspective of palliative communication is present, but necessitates more time to become entrenched. Orv Hetil. 2017; 158(30): 1175-1181.

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Since the sixteenth century, competition between midwives and surgeons has created a culture of blame around the difficult delivery. In the late seventeenth century, 100 years before oxygen was discovered, researchers associated “apparent death of the newborn” with impaired respiratory function of the placenta. The diagnosis “birth asphyxia” replaced the term “apparent death of the newborn” during the mass phobia of being buried alive in the eighteenth century. This shifted the interpretation from unavoidable fate to a preventable condition. Although the semantic inaccuracy (“pulselessness”) was debated, “asphyxia” was not scientifically defined until 1992. From 1792 the diagnosis was based on a lack of oxygen. “Blue” and “white” asphyxia were perceived as different disorders in the eighteenth, and as different grades of the same disorder in the nineteenth century. In 1862, William Little linked birth asphyxia with cerebral palsy, and although never confirmed, his hypothesis was accepted by scientists and the public. Fetal well-being was assessed by auscultating heart beats since 1822, and continuous electronic fetal monitoring was introduced in the 1960s without scientific assessment. It neither diminished the incidence of birth asphyxia nor of cerebral palsy, but rather raised the rate of cesarean sections and litigation against obstetricians and midwives.


CONTEXT: The families of oncology patients requiring intensive care often face increasing complexity in communication with their providers, particularly when patients are cared for by providers from different disciplines. OBJECTIVE: The objective of this study was to describe experiences and challenges faced by pediatric oncologists and intensivists and how the oncologist-intensivist relationship impacts communication and initiation of goals of care discussions (GCDs). METHODS: We conducted semi-structured interviews with a convenience sample of 10 physicians, including pediatric oncology and intensive care attendings and fellows. RESULTS: We identified key themes (three barriers and four facilitators) to having GCDs with families of oncology patients who have received intensive care. Barriers included challenges to communication within teams because of hierarchy and between teams due to incomplete sharing of information and confusion about who should initiate GCDs; provider experiences of internal conflict about how to engage parents in decision-making and about the "right thing to do" for patients; and lack of education and training in communication. Facilitators included team preparation for family meetings; skills for partnering with families; the presence of palliative care specialists; and informal education in communication and willingness for further training in communication. Notably, the education theme was identified as both a barrier and resource. CONCLUSION: We identified barriers to communication with families both within and between teams and for individual physicians. Formal communication training and processes that standardize communication to ensure completeness and role delineation between clinical teams may improve oncologists' and intensivists' ability to initiate GCDs, thereby fulfilling their ethical obligations of decision support.


AIM: Medical providers may face unique emotional challenges when confronted with the suffering of chronically ill, dying, and bereaved children. This study assessed the preliminary outcomes of participation in a group-based multimodal mindfulness training pilot designed to reduce symptoms of burnout and mental health symptoms in providers who interact with children in the context of end-of-life care. METHODS: A total of 13 medical providers who care for children facing life-threatening illness or bereaved children participated in a 9-session multimodal mindfulness session. Mental health symptoms and burnout were assessed prior to the program, at the program midpoint, and at the conclusion of the program. RESULTS: Participation in the pilot was associated with significant reductions in depressive and posttraumatic stress disorder (PTSD) symptoms among providers (P < .05). CONCLUSION: Mindfulness-based programs may help providers recognize and address symptoms of depression and PTSD. Additional research is needed to enhance access and uptake of programming among larger groups of participants.


Communicating diagnostic news in health contexts is a potentially difficult event for all parties involved. However, despite this task's presence in the physician-patient context, it is rarely addressed during clinical training. The current study thus aimed to describe and evaluate how difficult news can be toned down during genetic counseling sessions involving cases of fetal syndromes and/or malformations. The study analyzed 33 naturalistic interactions (i.e. real situations), taped and transcribed, according to the theoretical and methodological perspective of Conversation Analysis, with an ethnomet hodological basis. These interactions consisted of sessions in clinical genetics with pregnant women seen at the fetal medicine service of a reference hospital for maternal and child health in the Brazilian Unified National Health System (SUS). The analysis showed that communicating difficult news can be accompanied by optimistic perspectives that are scaled-up according to each situation's severity. In the absence of a positive diagnosis, the appointments can conclude with positive aspects such as recommendations for palliative care, so that the patient always leaves the appointment with some kind of recommendation. This study proposes to innovate and expand the scope of studies on communicating difficult news in the physician-patient relationship in Brazil, precisely by developing an analysis of real interactions in
genetic counseling and thus providing interactional backing for training health professionals that deal with this challenge in their routine work.


PURPOSE: This study aimed to identify and compare hospice care research topics between Korean and international nursing studies using text network analysis. METHODS: The study was conducted in four steps: 1) collecting abstracts of relevant journal articles, 2) extracting and cleaning keywords (semantic morphemes) from the abstracts, 3) developing co-occurrence matrices and text-networks of keywords, and 4) analyzing network-related measures including degree centrality, closeness centrality, betweenness centrality, and clustering using the NetMiner program. Abstracts from 347 Korean and 1,926 international studies for the period of 1998-2016 were analyzed. RESULTS: Between Korean and international studies, six of the most important core keywords -“hospice,” “patient,” “death,” “RNs,” “care,” and “family”-were common, whereas “cancer” from Korean studies and “palliative care” from international studies ranked more highly. Keywords such as “attitude,” “spirituality,” “life,” “effect,” and “meaning” for Korean studies and “communication,” “treatment,” “USA,” and “doctor” for international studies uniquely emerged as core keywords in recent studies (2011~2016). Five subtopic groups each were identified from Korean and international studies. Two common subtopics were “hospice palliative care and volunteers” and “cancer patients.” CONCLUSION: For a better quality of hospice care in Korea, it is recommended that nursing researchers focus on study topics of patients with non-cancer disease, children and family, communication, and pain and symptom management.


A lack of knowledge and skills in pediatric palliative care may create hesitation in caring for children with serious life-threatening conditions and their families. Our research examined the effectiveness of pediatric palliative care training for pediatric clinicians. A pretest-posttest study provided educational training in pediatric palliative care to pediatric clinicians and used a pretest and a posttest to assess outcomes. Fifty pediatric clinicians attended this research with 83.3% response rate. After training, participants reported significantly increased confidence in a variety of areas, including providing emotional support to clinicians, personal knowledge, skills, and communication; ethical and legal concerns; and providing emotional support to dying children and their families. Results showed a significant main effect of training on confidence levels (p < .000). This suggests that education can effectively boost pediatric clinicians' confidence regarding providing pediatric palliative care and therefore should regularly be provided to clinicians.


BACKGROUND: Clinical observation is a key component of medical ability, enabling immediate evaluation of the patient's emotional state and contributing to a clinical clue that leads to final decision making. In medical schools, the art of learning to look can be taught using medical humanities and especially visual arts. By presenting a Ramsay sedation score (RSS) integrated with Caravaggio’s paintings during a procedural sedation conference for pediatric residents, we want to test the effectiveness of this approach to improve the quality of learning. METHODS: In this preliminary study, we presented videos showing sedated pediatric patients in the setting of a procedural sedation lesson to two randomized groups of residents, one attending a lesson on RSS explained through the masterpieces of Caravaggio, the other without artistic support. A week later we tested their learning with ten multi-choice questions focused on theoretical questions about sedation monitoring and ten more questions focused on recognizing the appropriate RSS viewing the videos. The primary outcome was the comparison of the total number of RSS layers properly recognized in both groups. We also evaluated the appreciation of the residents of the use of works of art integrated with the lesson. RESULTS: Eleven students were randomized to each group. Two residents in the standard lesson did not attend the test. The percentage of correct answers on the theoretical part was similar, 82% in the art group and 89% in the other (p > 0.05). No difference was found in the video recognition part of the RSS recognition test. Residents exposed to paintings showed great appreciation for the integration of the lesson with the Caravaggio’s masterpieces. CONCLUSIONS:
BACKGROUND: Current conceptualisations of moral distress largely portray a negative phenomenon that leads to burnout, reduced job satisfaction and poor patient care. OBJECTIVE: To explore clinical experiences, perspectives and perceptions of moral distress in neonatology. DESIGN: An anonymous questionnaire was distributed to medical and nursing providers within two tertiary level neonatal intensive care units (NICUs)-one surgical and one perinatal-seeking their understanding of the term and their experience of it. Open-ended questions were analysed using qualitative methodology. RESULTS: A total of 345 healthcare providers from two NICUs participated (80% response rate): 286 nurses and 59 medical providers. Moral distress was correctly identified as constrained moral judgement resulting in distress by 93% of participants. However, in practice the term moral distress was also used as an umbrella term to articulate different forms of distress. Moral distress was experienced by 72% of providers at least once a month. Yet despite the negative sequelae of moral distress, few (8% medical, 21% nursing providers) thought that moral distress should be eliminated from the NICU. Open-ended responses revealed that while interventions were desired to decrease the negative impacts of moral distress, moral distress was also viewed as an essential component of the caring profession that prompts robust discussion and acts as an impetus for medical decision-making. CONCLUSIONS: Moral distress remains prevalent within NICUs. While the harmful aspects of moral distress need to be mitigated, moral distress may have a positive role in advocating for and promoting the interests of the neonatal population.

OBJECTIVES: The benefits of non-invasive ventilation (NIV) have been clearly demonstrated in pediatrics. In palliative care, NIV can improve the level of comfort and quality of life and can decrease dyspnea. The objective was to survey pediatricians’ opinions and practices regarding NIV in palliative care in France. DESIGN: A mail survey was conducted among pediatric pneumologists, intensivists and palliative medicine consultants from February 2015 to March 2015. RESULTS: In case of acute respiratory failure, 84% of the responding practitioners found NIV appropriate in do-not-intubate (DNI) children, while only 35% of them found it appropriate in comfort-measures-only (CMO) children (P<0.0001). In case of progressive respiratory failure, 68% of the responders found NIV appropriate in DNI children, while only 30% in CMO children (P<0.05). The major criterion for initiating NIV in pediatric palliative care was the presence of dyspnea. In pediatric palliative care, the efficacy of NIV was evaluated primarily clinically in terms of the improvement of the child’s comfort level, as well as the child’s and family’s satisfaction. Hypercapnia and desaturation were rarely measured to initiate NIV or to assess its efficacy. Sixty percent of the responding practitioners indicated that referral to NIV was anticipated with children and family before acute events or end-of-life occurred. CONCLUSION: French pediatricians habitually use NIV for management of acute or progressive respiratory symptoms in DNI children. In CMO children, a majority of responding practitioners find NIV inappropriate. In palliative care, the indications for and efficacy of NIV are evaluated based on clinical criteria and rarely on gasometric criteria.

BACKGROUND: Despite the number of interprofessional team members caring for children at the end of life, little evidence exists on how institutions can support their staff in providing care in these situations. OBJECTIVE: We sought to evaluate which aspects of the hospital work environment were most helpful for multidisciplinary team members who care for patients at the end of life and identify areas for improvement to better address staff needs. DESIGN: Qualitative thematic analysis was completed of free-text comments from a survey distributed to interprofessional staff members involved in the care of a recently deceased pediatric patient. A total of 2701 surveys were sent; 890 completed. Free-text responses were provided by 306 interprofessional team members. SETTING/SUBJECTS: Interprofessional team members involved in the care of a child who died at a 348 bed...
academic children’s hospital in the Midwestern United States. MEASUREMENTS: Realist thematic analysis of free-text responses was completed in Dedoose using a deductive and inductive approach with line-by-line coding. Descriptive statistics of demographic information was completed using Excel. RESULTS: Thematic analysis of the 306 free-text responses identified three main support-related themes. Interprofessional team members desire to have (1) support through educational efforts such as workshops, (2) support from colleagues, and (3) support through institutional practices. CONCLUSIONS: Providers who participate in end-of-life work benefit from ongoing support through education, interpersonal relationships, and institutional practices. Addressing these areas from an interprofessional perspective enables staff to provide the optimal care for patients, patients’ families, and themselves. 


Paediatric palliative care (PPC) endeavours to alleviate the suffering and improve the quality of life of children with serious illnesses and their families. In the past two decades since WHO defined PPC and called for its inclusion in paediatric oncology care, rigorous investigation has provided important insights. For example, the first decade of research focused on end-of-life experiences of the child and the family, underscoring the high prevalence of symptom burden, the barriers to parent-provider concordance with regards to prognosis, as well as the need for bereavement supports. The second decade expanded PPC oncology investigation to include the entire cancer continuum and the voices of patients. Other studies identified the need for support of parents, siblings, and racial and ethnic minority groups. Promising interventions designed to improve outcomes were tested in randomised clinical trials. Future research will build on these findings and pose novel questions about how to continue to reduce the burdens of paediatric cancer. 


OBJECTIVE: To describe our institutional experience with a four-week pediatric HPM elective rotation and its impact on residents’ self-rated competencies. BACKGROUND: In the spirit of bolstering primary hospice and palliative medicine (HPM) skills of all pediatricians, it is unclear how best to teach pediatric HPM. An elective rotation during residency may serve this need. METHODS: An anonymous online survey was distributed to pediatric and internal medicine/pediatrics residents at a single, tertiary academic children's hospital. Respondents were asked to rate education, experience, and comfort with five aspects of communication with families of children with terminal illnesses and six domains of managing the symptoms of terminal illnesses. Self-ratings were recorded on a 1-5 scale: none, minimal, moderate, good, or excellent. Demographic data, including details of training and prior HPM training, were collected. Respondents completed a set of six questions gauging their attitude toward palliative care in general and at the study institution specifically. RESULTS: All respondents desire more HPM training. Those residents who self-selected to complete a pediatric HPM elective rotation had significantly higher self-ratings in 10 of 11 competency/skill domains. Free-text comments expressed concern about reliance on the specialty HPM team. DISCUSSION: A pediatric HPM elective can significantly increase residents’ self-rated competency. Such rotations are an under-realized opportunity in developing the primary HPM skills of pediatricians, but wider adoption is restricted by the limited availability of pediatric HPM rotations and limited elective time during training. 


BACKGROUND: Fifteen evidence-based Standards for Psychosocial Care for Children with Cancer and Their Families (Standards) were published in 2015. The Standards cover a broad range of topics and circumstances and require qualified multidisciplinary staff to be implemented. This paper presents data on the availability of psychosocial staff and existing practices at pediatric oncology programs in the United States, providing data that can be used to advocate for expanded services and prepare for implementation of the Standards. PROCEDURE: Up to three healthcare professionals from 144 programs (72% response rate) participated in an online survey
Pulmonary arteriovenous malformations (PAVMs) are structurally abnormal vascular communications that provide a continuous right-to-left shunt between pulmonary arteries and veins. Their importance stems from the risks they pose (>1 in 4 patients will have a paradoxical embolic stroke, abscess or myocardial infarction while life-threatening haemorrhage affects 1 in 100 women in pregnancy), opportunities for risk prevention, surprisingly high prevalence and under-appreciation, thus representing a challenging condition for practising healthcare professionals. The driver for the current Clinical Statement was the plethora of new data since previous hereditary haemorrhagic telangiectasia (HHT) guidelines generated in 2006 and a systematic Cochrane Review for PAVM embolisation in 2011. The British Thoracic Society (BTS) identified key areas in which there is now evidence to drive a change in practice. Due to the paucity of data in children, this Statement focused on adults over 16 years. The Statement spans the management of PAVMs already known to be present (interventional and medical), and identifies challenges in the delivery of psychosocial care. RESULTS: Over 90% of programs have social workers and child life specialists who provide care to children with cancer and their families. Fewer programs have psychologists (60%), neuropsychologists (31%), or psychiatrists (19%). Challenges in psychosocial care are primarily based on pragmatic issues related to funding and reimbursement.

CONCLUSION: Most participating pediatric oncology programs appear to have at least the basic level of staffing necessary to implement some of the Standards. However, the lack of a more comprehensive multidisciplinary team is a likely barrier in the implementation of the full set of Standards.


OBJECTIVE: To describe neonatologist and pediatric intensivist attitudes and practices relevant to high-stakes decisions for children with chronic critical illness, with particular attention to physician perception of professional duty to seek treatment team consensus and to disclose team conflict. DESIGN: Self-administered online survey. SETTING: U.S. neonatal ICUs and PICUs. SUBJECTS: Neonatologists and pediatric intensivists. INTERVENTIONS: None. MEASUREMENTS AND MAIN RESULTS: We received 852 responses (333 neonatologists, denominator unknown; 319 of 1,290 pediatric intensivists). When asked about guiding a decision for tracheostomy in a chronically critically ill infant, only 41.7% of physicians indicated professional responsibility to seek a consensus decision, but 73.3% reported, in practice, that they would seek consensus and make a consensus-based recommendation; the second most common practice (15.5%) was to defer to families without making recommendations. When presented with conflict among the treatment team, 63% of physicians indicated a responsibility to be transparent about the decision-making process and reported matching practices. Neonatologists more frequently reported a responsibility to give decision making fully over to families; intensivists were more likely to seek out consensus among the treatment team. CONCLUSIONS: ICU physicians do not agree about their responsibilities when approaching difficult decisions for chronically critically ill children. Although most physicians feel a professional responsibility to provide personal recommendations or defer to families, most physicians report offering consensus recommendations. Nearly all physicians embrace a sense of responsibility to disclose disagreement to families. More research is needed to understand physician responsibilities for making recommendations in the care of chronically critically ill children.

BACKGROUND: Hospice and Palliative Medicine (HPM) competencies are of growing importance in training general pediatricians and pediatric sub-specialists. The Accreditation Council for Graduate Medical Education (ACGME) emphasized pediatric trainees should understand the “impact of chronic disease, terminal conditions and death on patients and their families.” Currently, very little is known regarding pediatric trainee education in HPM. METHODS: We surveyed all 486 ACGME-accredited pediatric training program directors (PDs) - 200 in general pediatrics (GP), 57 in cardiology (CARD), 64 in critical care medicine (CCM), 69 in hematology-oncology (ONC) and 96 in neonatology (NICU). We collected training program’s demographics, PD’s attitudes and educational practices regarding HPM. RESULTS: The complete response rate was 30% (148/486). Overall, 45% offer formal HPM curriculum and 39% offer a rotation in HPM for trainees. HPM teaching modalities commonly reported included conferences, consultations and bedside teaching. Eighty-one percent of all respondents felt that HPM curriculum would improve trainees’ ability to care for patients. While most groups felt that a HPM rotation would enhance trainees’ education [GP (96%), CARD (77%), CCM (82%) and ONC (95%)], NICU PDs were more divided (55%; p < 0.05 for all comparisons vs. NICU). CONCLUSION: While most programs report perceived benefit from HPM training, there remains a paucity of opportunities for pediatric trainees. Passive teaching methods are frequently utilized in HPM curricula with minimal diversity in methods utilized to teach HPM. Opportunities to further emphasize HPM in general pediatric and pediatric sub-specialty training remains.


OBJECTIVES: To determine which prognostic information sources parents find informative and which are associated with better parental understanding of prognosis. METHODS: Prospective, questionnaire-based cohort study of parents and physicians of children with cancer at 2 academic pediatric hospitals. We asked parents how they learned about prognoses and evaluated relationships between information sources and prognostic understanding, defined as accuracy versus optimism. We excluded parents with pessimistic estimates and whose children had such good prognoses that optimism relative to the physician was impossible. Analytic cohort of 256 parent-physician pairs. RESULTS: Most parents considered explicit sources (conversations with oncologists at diagnosis, day-to-day conversations with oncologists, and conversations with nurses) “very” or “extremely” informative (73%-85%). Implicit sources (parent’s sense of how child was doing or how oncologist seemed to feel child was doing) were similarly informative (84%-87%). Twenty-seven percent (70/253) of parents reported prognostic estimates matching physicians’ estimates. Parents who valued implicit information had lower prognostic accuracy (odds ratio [OR] 0.50; 95% confidence interval 0.29-0.88), especially those who relied on a “general sense of how my child’s oncologist seems to feel my child is doing” (OR 0.47; 0.22-0.99). Parents were more likely to use implicit sources if they reported receiving high-quality prognostic information (OR 3.02; 1.41-6.43), trusted the physician (OR 2.01; 1.01-3.98), and reported high-quality physician communication (OR 1.81; 1.00-3.27). CONCLUSIONS: Reliance on implicit sources was associated with overly-optimistic prognostic estimates. Parents who endorsed strong, trusting relationships with physicians were not protected against misinformation.


From the time of diagnosis through either survivorship or end of life, communication between healthcare providers and patients or parents can serve several core functions, including fostering healing relationships, exchanging information, responding to emotions, managing uncertainty, making decisions, and enabling patient/family self-management. We systematically reviewed all studies that focused on communication between clinicians and patients or parents in pediatric oncology, categorizing studies based on which core functions of communication they addressed. After identifying gaps in the literature, we propose a research agenda to further the field.


Sepsis is the leading cause of child's death, yet it is well known that the rapid initiation of simple, timely interventions reduces morbidity and mortality. This paper shares our findings on the barriers doctors in training have identified to delivering such care. We also share the results of a pilot paediatric intensive care unit outreach teaching programme designed to directly address these highlighted concerns.


Due to the lack of evidence-based guidelines, management strategies for neonatal MI should be individualized and administered largely at the discretion of responsible treating teams. Supportive care with a focus on preserving adequate circulation and antithrombotic therapy with a view to restoring vascular patency are the mainstays of treatment. Thrombolytic therapy of neonatal MI includes a chance to completely restore myocardial function. Understanding the resilience of the neonatal heart and mechanism of cardiac cell repair in neonates may spark novel treatment strategies for severe MI in the large number of affected individuals in an aging population.


OBJECTIVES: Our aim in this study was to understand usage patterns of pediatric palliative care (PPC) consultation and associations with end-of-life preparation among pediatric patients who are deceased. METHODS: We reviewed 233 pediatric mortalities. Data extraction from the electronic health record included determination of PPC consultation by using Current Procedural Terminology codes. Diagnoses were identified by International Classification of Disease codes and were classified into categories of life-threatening complex chronic conditions (LT-CCCs). Data analysis included Student's t test, Wilcoxon rank test, Fisher's exact test, chi(2) test, and multivariable logistic regression. RESULTS: The overall PPC consultation rate for pediatric patients who subsequently died was 24%. A PPC consultation for patients admitted to the pediatric ward and PICU was more likely than for patients cared for in the NICU (31% vs 12%, P < .01) and was more likely for those with an LT-CCC (40% vs 10%, P < .01), particularly malignancy (65% vs 35%, P < .01). Also noted were increased completion of Physician Orders for Life-Sustaining Treatment forms (8 vs 0, P < .01) and increased documentation of mental health disorders (60% vs 40%, P = .02). CONCLUSIONS: Our findings suggest that PPC consultation for patients in the pediatric ward and PICU is more likely among patients with a greater number of LT-CCCs, and is associated with increased Physician Orders for Life-Sustaining Treatment preparation and documentation of mental health disorders. Patients at risk to not receive PPC consultation are those with acute illness and patients in the NICU.


CONTEXT: Pediatric palliative care consults for children with cancer often occur late in the course of disease and close to death, when earlier involvement would reduce suffering. The perceptions that pediatric oncology providers hold about the pediatric palliative care service (PPCS) may shape referral patterns. OBJECTIVES: To explore how pediatric oncology providers at one institution perceived the hospital's PPCS and the way these perceptions may influence the timing of consultation. METHODS: We conducted semistructured qualitative interviews with pediatric oncology providers at a large children’s hospital. Interviews were audio-recorded, transcribed, and analyzed by two coders using a modified grounded theory approach. RESULTS: We interviewed 16 providers (10 physicians, one nurse practitioner, two social workers, two psychologists, and one child life specialist). Three core perceptions emerged: 1) the PPCS offers a diverse range of valuable contributions to the care of children with advancing cancer; 2) providers held favorable opinions about the PPCS owing to positive interactions with individual palliative care specialists deemed extraordinarily emotionally skilled; and 3) there is
considerable emotional labor involved in calling a PPCS consult that serves as a barrier to early initiation. CONCLUSION: The pediatric oncology providers in our study held a highly favorable opinion about their institution’s PPCS and agreed that early consultation is ideal. However, they also described that formally consulting PPCS is extremely difficult because of what the PPCS symbolizes to families and the emotional labor that the provider must manage in introducing them. Interventions to encourage the early initiation of palliative care in this population may benefit from a focus on the emotional experiences of providers.


BACKGROUND: Non-medical prescribing is well established within the British health service, with increasing numbers of nurses practicing within children’s hospices. AIM: To identify the context of non-medical prescribing in children’s hospices in the UK, focusing on the perceived benefits and challenges. METHOD: Internet-based questionnaires were sent to 55 UK children’s hospices, exploring the practice and context of prescribing. RESULTS: Of the 55 invited, 20 children’s hospices responded to the questionnaire, 14 of which employed a total of 39 non-medical prescribers (NMPs). Sixteen individual NMPs responded, of which half (50%) prescribed to enable the continuation of existing medicines, 37.5% prescribed independently surrounding symptom management and control and 31.3% in end-of-life care. Perceived benefits of prescribing included timely access to medicines, increased efficiency and accuracy in the admissions process and medicine reconciliation and the increased ability to offer choice in the place of palliative and hospice care. Perceived barriers to prescribing surrounded opportunities to develop confidence, defining the scope of practice and the time required to assess, diagnose and treat. CONCLUSION: NMPs are making a significant contribution to the prescribing workforce within hospices; however, a number of challenges need to be addressed to enable hospices to realise the benefits.


BACKGROUND: Children’s hospices are a key provider of palliative care for children and young people with life-limiting and life-threatening conditions. However, despite recent policy attention to the provision of paediatric palliative care, little is known about the role of children’s hospice staff and the factors that may impact on their wellbeing at work. This study explored the rewards and challenges of working in a children’s hospice with an aim to identify staff support and development needs. METHODS: We conducted an exploratory, qualitative study involving thematic analysis of semi-structured interviews with 34 staff and three focus groups with 17 staff working in a multi-disciplinary care team in a UK children’s hospice. RESULTS: Participants identified rewards and challenges related to the direct work of caring for children and their families; team dynamics and organisational structures; and individual resilience and job motivation. Participants described the work as emotionally intensive and multi-faceted; ‘getting it right’ for children was identified as a strong motivator and reward, but also a potential stressor as staff strived to maintain high standards of personalised and emotional care. Other factors were identified as both a reward and stressor, including team functioning, the allocation of work, meeting parent expectations, and the hospice environment. Many participants identified training needs for different aspects of the role to help them feel more confident and competent. Participants also expressed concerns about work-related stress, both for themselves and for colleagues, but felt unable to discuss this at work. Involuntary support from colleagues and group clinical reflection were identified as primary resources to reflect on and learn from and for emotional support. However, opportunities for this were limited. CONCLUSIONS: Providing regular, structured, and dedicated clinical reflection provides a mechanism through which children’s hospice staff can come together for support and learning, and demonstrates an organisational commitment to staff wellbeing and development. Being aware of children’s hospice specific rewards and challenges can help to ensure that staff feel supported and competent in their role. Breaking down barriers to discussing work-related stress and enhancing awareness about early signs of burnout is also important.


Background Treating children with cancer requires multiple different skills. For the healthcare personnel (HCP) in Germany the practice of ongoing training to improve professional skills is almost non-existent. Therefore, we
developed a programme called ‘SICKO’ to support HCPs skills and attitudes by means of a multidisciplinary workshop. Methods Following a qualitative analysis, we then designed a modular (3 day) workshop. During day one (8 h) participants learn practical skills, the fundamentals of chemotherapy, and effective communication skills. Workshop day 2 (8 h) includes education regarding the complications of cancer therapy (e.g. tumour-lysis syndrome, delayed methotrexate excretion), and their management. Topics during day 3 (8 h) include ‘breaking bad news’, conflict management in the team, infusion-related complications and ‘crew resource management’ (CRM). Results 43 nurses and 33 physicians participated between 2013 and 2015. All participants highly recommend the workshop. Participants felt that knowledge increased significantly after workshops and were more confident regarding challenging communications. Discussion/Conclusions Although long-term effects have not yet been evaluated, ‘SICKO’ offers the opportunity for HCP to train and experience simulated day-to-day challenges in the field of paediatric oncology.


There are currently no evidence-based oxygen saturation targets for treating children with life-threatening conditions. We reviewed evidence of SpO2 targets for oxygen therapy in children with emergency signs as per WHO Emergency Triage Assessment and Treatment guidelines. We systematically searched for physiological data and international guidelines that would inform a safe approach. Our findings suggest that in children with acute lung disease who do not require resuscitation, a threshold SpO2 for commencing oxygen of 90% will provide adequate oxygen delivery. Although there is no empirical evidence regarding oxygen saturation to target in children with emergency signs from developing countries, a SpO2 of >/= 94% during resuscitation may help compensate for common situations of reduced oxygen delivery. In children who do not require resuscitation or are stable post resuscitation with only lung disease, a lower limit of SpO2 for commencing oxygen of 90% will provide adequate oxygen delivery and save resources.


INTRODUCTION: Short bowel syndrome (SBS) is the main cause of intestinal failure (IF) in the pediatric population. To promote the standardization of care of these patients, the registry of Pediatric Intestinal Rehabilitation and Transplantation (PIRAT) has been established. The aim of this study is to describe patients with IF using PIRAT database. MATERIALS AND METHODS: Data from two tertiary care European referral Centers registered in PIRAT (https://www.studeon.eu/pirat) were analyzed (1994-2015). Neonatal SBS-related IF was defined as need for parenteral nutrition (PN) to sustain life and growth for more than 75 days, after extensive bowel resection during neonatal period. Data included patient demographics, disease at birth, residual small intestine, and intestinal autonomy (PN on/off). RESULTS: In this study, 114 children with SBS-related IF were identified (male 60%). Median gestational age was 35.3 weeks (interquartile range [IQR]: 33.0-38.0); median birth weight was 2,440 g (IQR: 1,700-2,990). The main causes of SBS were intestinal atresia in 31 (27%), midgut volvulus in 29 (25%), necrotizing enterocolitis in 23 (20%), and gastroschisis in 12 (11%). Nine (7.9%) patients died on PN (six sepsis, two IF-associated liver disease, and one multiorgan failure). Median residual small bowel length was 46 cm (IQR: 13.0-92.5). Ileocecal valve was resected in 48 patients (42%). Intestinal autonomy was achieved in 68% patients. CONCLUSION: We present the web-based registry PIRAT and the first results of patients with IF registered from two European Centers. PIRAT could give the opportunity to create a dedicated international network (IF-net) to standardize, improve, and spread the therapeutic paths for the rare and heterogeneous condition of SBS-related IF.


BACKGROUND: Over the last decade, paediatric palliative care teams (PPCTs) have been introduced to support children with life-limiting diseases and their families and to ensure continuity, coordination and quality of paediatric palliative care (PPC). However, implementing a PPCT into an organisation is a challenge. The objective of this study was to identify barriers and facilitators reported by healthcare professionals (HCPs) in primary, secondary or tertiary care for implementing a newly initiated multidisciplinary PPCT to bridge the gap between hospital and home. METHODS: The Measurement Instrument for Determinants of Innovations (MIDI) was used to assess responses of 71 HCPs providing PPC to one or more of the 129 childr

OBJECTIVES: To describe enteral feeding practices in pre and postoperative infants with congenital heart disease in European PICUs. DESIGN: Cross-sectional electronic survey. SETTING: European PICUs that admit infants with congenital heart disease pre- and postoperatively. PARTICIPANTS: One senior PICU physician or designated person per unit. INTERVENTIONS: None. MEASUREMENTS AND MAIN RESULTS: Fifty-nine PICUs from 18 European countries responded to the survey. PICU physicians were involved in the nutritional care of children with congenital heart disease in most (76%) PICUs, but less than 60% of units had a dedicated dietician. Infants with congenital heart disease were routinely fed preoperatively in only 63% of the PICUs, due to ongoing concerns around prostaglandin E1 infusion, the presence of umbilical venous and/or arterial catheters, and the use of vasoactive drugs. In three quarters of the PICUs (76%), infants were routinely fed during the first 24 hours postoperatively. Units cited, the most common feeding method, both pre and postoperatively, was intermittent bolus feeds via the gastric route. Importantly, 69% of European PICUs still did not have written guidelines for feeding, but this varied for pre and postoperative patients. CONCLUSIONS: Wide variations in practices exist in the nutritional care between European PICUs, which reflects the absence of local protocols and scientific society-endorsed guidelines. This is likely to contribute to suboptimal energy delivery in this particularly vulnerable group.

Central venous access devices (CVADs) form an important component of modern paediatric healthcare, especially for children with chronic health conditions such as cancer or gastrointestinal disorders. However device failure and complications rates are high. Over 2(1/2) years, a child requiring parenteral nutrition and associated vascular access dependency due to ‘short gut syndrome’ (intestinal failure secondary to gastroschisis and resultant significant bowel resection) had ten CVADs inserted, with nine subsequently failing. This resulted in multiple anaesthetics, invasive procedures, injuries, vascular depletion, interrupted nutrition, delayed treatment and substantial healthcare costs. A conservative estimate of the institutional costs for each insertion, or rewiring, of her tunnelled CVAD was $A10 253 (2016 Australian dollars). These complications and device failures had significant negative impact on the child and her family. Considering the commonality of conditions requiring prolonged vascular access, these failures also have a significant impact on international health service costs.

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major facilitator for implementing a PPCT. Tailored organisational strategies such as working arrangements by management, concrete information about the PPCT itself and the type of support provided by the PPCT should be clearly communicated to involved HCPs to increase awareness about benefits of the PPCT and ensure a successful implementation. New PPCTs need protection and resources in their initial year to develop into experienced and qualified PPCTs.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5810030/


BACKGROUND/AIM: Somatosensation is the ability to detect and recognise body sensations such as touch, vibration, pressure, pain, temperature and proprioception. Cerebral palsy is a neurological disorder that is often accompanied by impairments in somatosensation. Current somatosensory assessments have limited psychometrics established for use with these children. The aim of this study was to identify therapists' current practice and perspectives related to the assessment of somatosensation in children with neurological disorders. METHODS: A cross-sectional questionnaire was used to identify the somatosensory assessments currently used in clinical practice, time allocated to assessment, and therapists' satisfaction and confidence using the available assessments of somatosensation. The questionnaire was adapted from a previously utilised questionnaire that identified therapists' use of somatosensory assessments with adults post-stroke. RESULTS: A total of 135 therapists responded to the questionnaire. Seventy-nine (92%) occupational therapists and 44 (89.7%) physiotherapists indicated that they currently assessed or treated children with somatosensory deficits. Sixty-four (82.1%) occupational therapists and 38 (86.3%) physiotherapists regarded assessment of somatosensation in children with neurological disorders as important to very important. However, only seven (8.8%) occupational therapists and seven (15.9%) physiotherapists reported confidence in their ability to do so. The methods with which therapists detect and measure somatosensory impairment in children with neurological disorders are variable, with non-standardised and/or informal assessments most frequently used. CONCLUSION: Despite there being recommendations of best practice for the assessment of specific domains of somatosensation in children with cerebral palsy, current practice does not yet mirror these recommendations. Additionally, therapists have low satisfaction and confidence with what they are currently using, highlighting the need for a comprehensive and standardised assessment of somatosensation for use in children with neurological disorders.


OBJECTIVES: Create trustworthy, rigorous, national clinical practice guidelines for the practice of pediatric donation after circulatory determination of death in Canada. METHODS: We followed a process of clinical practice guideline development based on World Health Organization and Canadian Medical Association methods. This included application of Grading of Recommendations Assessment, Development, and Evaluation methodology. Questions requiring recommendations were generated based on 1) 2006 Canadian donation after circulatory determination of death guidelines (not pediatric specific), 2) a multidisciplinary symposium of national and international pediatric donation after circulatory determination of death leaders, and 3) a scoping review of the pediatric donation after circulatory determination of death literature. Input from these sources drove drafting of actionable questions and Good Practice Statements, as defined by the Grading of Recommendations Assessment, Development, and Evaluation group. We performed additional literature reviews for all actionable questions. Evidence was assessed for quality using Grading of Recommendations Assessment, Development, and Evaluation and then formulated into evidence profiles that informed recommendations through the evidence-to-decision framework. Recommendations were revised through consensus among members of seven topic-specific working
groups and finalized during meetings of working group leads and the planning committee. External review was provided by pediatric, critical care, and critical care nursing professional societies and patient partners. RESULTS: We generated 63 Good Practice Statements and seven Grading of Recommendations Assessment, Development, and Evaluation recommendations covering 1) ethics, consent, and withdrawal of life-sustaining therapy, 2) eligibility, 3) withdrawal of life-sustaining therapy practices, 4) ante and postmortem interventions, 5) death determination, 6) neonatal pediatric donation after circulatory determination of death, 7) cardiac and innovative pediatric donation after circulatory determination of death, and 8) implementation. For brevity, 48 Good Practice Statement and truncated justification are included in this summary report. The remaining recommendations, detailed methodology, full Grading of Recommendations Assessment, Development, and Evaluation tables, and expanded justifications are available in the full text report. CONCLUSIONS: This process showed that rigorous, transparent clinical practice guideline development is possible in the domain of pediatric deceased donation. Application of these recommendations will increase access to pediatric donation after circulatory determination of death across Canada and may serve as a model for future clinical practice guideline development in deceased donation.


The concept of 'beginner's mind' invites the expert medical professional to be present to their patients, remaining curious and responsive in the face of the individuality of illness. Each patient is a universe of unknowns, presenting with suffering which cannot always be classified with a diagnosis. Improvisation and openness may not just benefit our patients enduring their patient journeys but may also revive and reconnect us with our own humanity. Why this matters to me: My experience as cancer patient brought home to me the value of encountering a doctor who was present and able to listen and respond to me as an individual. It did not necessarily take longer, but was about an attitude of heart. Like any professional, as GPs we are at risk of presumption and habitual thinking. Beginner's mind, that is recapturing the openness and curiosity modelled to us by children, can powerfully transform the medical encounter, allowing space for emergence of patient perspective and doctor response. Key message: Embrace curiosity and learning into our expert clinical practice.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5694793/
Epidemiology and Pathology


We estimated mortality rate and predictors of death in children and adolescents who acquired HIV through mother-to-child transmission in Paraguay. In 2000-2014, we conducted a cohort study among children and adolescents aged < 15 years. We abstracted data from medical records and death certificates. We used the Cox proportional hazards model for the multivariable analysis of mortality predictors. A total of 302 subjects were included in the survey; 216 (71.5%) were younger than 5 years, 148 (51.0%) were male, and 214 (70.9%) resided in the Asuncion metropolitan area. There were 52 (17.2%) deaths, resulting in an overall mortality rate of 2.06 deaths per 100 person-years. The children and adolescents with hemoglobin levels \( < / = 9 \) g/dL at baseline had a 2-times higher hazard of death compared with those who had levels \( > 9 \) g/dL (HR 2.27, 95% CI 1.01-5.10). The mortality of HIV-infected children and adolescents in Paraguay is high, and anemia is associated with mortality. Improving prenatal screening to find cases earlier and improving pediatric follow-up are needed.


BACKGROUND: In 2015, the second cycle of the CONCORD programme established global surveillance of cancer survival as a metric of the effectiveness of health systems and to inform global policy on cancer control. CONCORD-3 updates the worldwide surveillance of cancer survival to 2014. METHODS: CONCORD-3 includes individual records for 37.5 million patients diagnosed with cancer during the 15-year period 2000-14. Data were provided by 322 population-based cancer registries in 71 countries and territories, 47 of which provided data with 100% population coverage. The study includes 18 cancers or groups of cancers: oesophagus, stomach, colon, rectum, liver, pancreas, lung, breast (women), cervix, ovary, prostate, and melanoma of the skin in adults, and brain tumours, leukemias, and lymphomas in both adults and children. Standardised quality control procedures were applied; errors were rectified by the registry concerned. We estimated 5-year net survival. Estimates were age-standardised with the International Cancer Survival Standard weights. FINDINGS: For most cancers, 5-year net survival remains among the highest in the world in the USA and Canada, in Australia and New Zealand, and in Finland, Iceland, Norway, and Sweden. For many cancers, Denmark is closing the survival gap with the other Nordic countries. Survival trends are generally increasing, even for some of the more lethal cancers: in some countries, survival has increased by up to 5% for cancers of the liver, pancreas, and lung. For women diagnosed during 2010-14, 5-year survival for breast cancer is now 89.5% in Australia and 90.2% in the USA, but international differences remain very wide, with levels as low as 66.1% in India. For gastrointestinal cancers, the highest levels of 5-year survival are seen in southeast Asia: in South Korea for cancers of the stomach (68.9%), colon (71.8%), and rectum (71.1%); in Japan for oesophageal cancer (36.0%); and in Taiwan for liver cancer (27.9%). By contrast, in the same world region, survival is generally lower than elsewhere for melanoma of the skin (59.9% in South Korea, 52.1% in Taiwan, and 49.6% in China), and for both lymphoid malignancies (52.5%, 50.5%, and 38.3%) and myeloid malignancies (45.9%, 33.4%, and 24.8%). For children diagnosed during 2010-14, 5-year survival for acute lymphoblastic leukaemia ranged from 49.8% in Ecuador to 95.2% in Finland. 5-year survival from brain tumours in children is higher than for adults but the global range is very wide (from 28.9% in Brazil to nearly 80% in Sweden and Denmark). INTERPRETATION: The CONCORD programme enables timely comparisons of the overall effectiveness of health systems in providing care for 18 cancers that collectively represent 75% of all cancers diagnosed worldwide every year. It contributes to the evidence base for global policy on cancer control. Since 2017, the Organisation for Economic Co-operation and Development has used findings from the CONCORD programme as the official benchmark of cancer survival, among their indicators of the quality of health care in 48 countries worldwide. Governments must recognise population-based cancer registries as key policy tools that can be used to evaluate both the impact of cancer prevention strategies and the effectiveness of health systems for all patients diagnosed with cancer. FUNDING: American Cancer Society; Centers for Disease Control and

Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) are life-limiting and progressive neuromuscular conditions with significant comorbidities, many of which manifest during adolescence. BMD is a milder presentation of the condition and much less prevalent than DMD, making it less represented in the literature, or more severely affected individuals with BMD may be subsumed into the DMD population using clinical cutoffs. Numerous consensus documents have been published on the clinical management of DMD, the most recent of which was released in 2010. The advent of these clinical management consensus papers, particularly respiratory care, has significantly increased the life span for these individuals, and the adolescent years are now a point of transition into adult lives, rather than a period of end of life. This review outlines the literature on DMD and BMD during adolescence, focusing on clinical presentation during adolescence, impact of living with a chronic illness on adolescents, and the effect that adolescents have on their chronic illness. In addition, we describe the role that palliative-care specialists could have in improving outcomes for these individuals. The increasing proportion of individuals with DMD and BMD living into adulthood underscores the need for more research into interventions and intricacies of adolescence that can improve the social aspects of their lives.


PURPOSE: The gut microbiome plays a critical role in maintaining children's health and in preventing and treating children's disease. Current application of the gut microbiome in childhood cancer is still lacking. This study aimed to systematically review the following: (1) alternations in the gut microbiome throughout cancer treatment trajectories in children, (2) the associations between the gut microbiome and gastrointestinal (GI) symptoms and psychoneurological symptoms (PNS), and (3) the efficacy of therapeutic interventions in the gut microbiome in children with cancer. METHODS: PubMed, EMBASE, the Cochrane Library, and the American Society of Clinical Oncology abstract were searched. Eligible studies included all study types in which the gut microbiome was primarily reported in children with cancer. The Mixed Methods Assessment Tool was used to evaluate the methodology quality of included studies. Seven studies met our eligibility criteria, including two cohort studies, two case-control studies, and three randomized controlled trials. RESULTS: The findings showed that the diversity estimates of the gut microbiome in children with cancer were lower than those of healthy controls both pre- and post-treatment. Children with cancer showed a significantly lower relative abundance of healthy gut microbiome (e.g., Clostridium XIVa and Bifidobacterium) during and after cancer treatment. No adequate literature was identified to support the associations between dysbiosis of the gut microbiome and GI symptoms/PNS. The use of prebiotics (fructooligosaccharides) and probiotics (Bifidobacterium or Lactobacilli) appears to improve the microenvironment of the gut around 1 month (4-5 weeks) during chemotherapy rather than at the beginning of treatment. Data also suggest that both prebiotic and probiotic interventions decrease clinical side effects (e.g., infection and morbidity risk) in children with cancer. CONCLUSIONS: This study adds to the evidence that dysbiosis of the gut microbiome can be improved using prebiotic and probiotic supplementations in children with cancer. More well-designed experimental studies are needed to confirm this conclusion. Further studies are needed to examine the associations between the gut microbiome and GI symptoms/PNS in childhood cancer.

AIMS: New Zealand currently defines the adolescent and young adult (AYA) group for cancer services as young people 12-24 years of age, while other countries favour a designation of 15-29 years. This study was undertaken to compare cancer incidence and survival among 20-29 year olds to New Zealand’s younger AYA population and to assess survival for our 15-29 year population against international benchmarks. METHODS: Diagnostic and demographic information for cancer registrations between 2000 and 2009 for 25-29 year olds was obtained from the New Zealand Cancer Registry. Incidence rates (IR) and five-year relative survival estimates were calculated according to AYA diagnostic group/sub-group, sex and prioritised ethnicity. RESULTS: 1,541 new primary malignant cancers were diagnosed (IR: 588 per million). Five-year relative survival was 85%, but was significantly lower for Maori and Pacific peoples (both 77%) compared to non-Maori/non-Pacific peoples (88%). In the overall 15-29 year AYA cohort, disease-specific outcomes for bone tumours (46%) and breast cancer (64%) were inferior to international standards. CONCLUSION: New Zealand 25 to 29 year olds are at twice the risk of developing cancer as those 15-24 years. Given that the survival disparities identified were remarkably consistent with those for younger AYA, consideration should be given widening New Zealand's AYA age range.


CONTEXT: Specialized pediatric palliative home care (SPPHC) is the main pediatric palliative care structure in Germany. Detailed data on patient characteristics and care are sparse. Describing this population in terms of diagnoses and care needs is essential for further development of palliative care services for these patients.

OBJECTIVES: We asked whether the population at our center 1) was representative compared with national mortality statistics; 2) showed differences in the clinical course among the four diagnostic categories established by the Association for Children with Terminal Conditions/Royal College of Paediatrics and Child Health; and 3) was different to published populations in pediatric palliative care regarding diagnoses, care, and place of death.


RESULTS: Main International Statistical Classification of Diseases and Related Health Problems, 10th Revision groups were nervous system, congenital abnormalities, neoplasia, and metabolic disease, reflecting the mortality statistics for patients one to 20 years. Thirty-six percent of patients were assigned to ACT-3, 34% to ACT-4, 26% to ACT-1, and 4% to ACT-2. ACT-1 patients mostly needed high-intensity care for short durations, ACT-4 patients showed long survival times with mostly intermittent care. Seventy-five percent of patients showed nervous system involvement. Eighty-four percent died at home, 12% in hospital, and 4% in a hospce, with 96% dying at their preferred place. CONCLUSION: Our data on SPPHC show 1) significant differences between Association for Children with Terminal Conditions/Royal College of Paediatrics and Child Health groups in terms of care needs and survival; 2) a high prevalence of children with neurological problems; and 3) a large majority of children dying at home.


OBJECTIVES: To investigate survival up to early adulthood for children with intellectual disability and compare their risk of mortality with that of children without intellectual disability. STUDY DESIGN: This was a retrospective cohort study of all live births in Western Australia between January 1, 1983 and December 31, 2010. Children with an intellectual disability (n = 10,593) were identified from the Western Australian Intellectual Disability Exploring Answers Database. Vital status was determined from linkage to the Western Australian Mortality database. Kaplan-Meier product limit estimates and 95% CIs were computed by level of intellectual disability. Hazard ratios (HRs) and 95% CIs were calculated from Cox proportional hazard regression models adjusting for potential confounders. RESULTS: After adjusting for potential confounders, compared with those without intellectual disability, children with intellectual disability had a 6-fold increased risk of mortality at 1-5 years of age (adjusted HR [aHR] = 6.0, 95%CI: 4.8, 7.6), a 12-fold increased risk at 6-10 years of age (aHR = 12.6, 95% CI: 9.0, 17.7) and a 5-fold increased risk at 11-25 years of age (aHR = 4.9, 95% CI: 3.9, 6.1). Children with severe intellectual disability were at even greater risk. No difference in survival was observed for Aboriginal children with intellectual disability compared with non-Aboriginal children with intellectual disability. CONCLUSIONS: Although children with intellectual disability experience higher mortality at all ages compared with those without intellectual disability, the greatest burden is for those with severe intellectual disability. However, even children with mild to moderate intellectual disability have increased risk of death compared with unaffected children.
Congenital Zika syndrome (CZS) causes early brain development impairment by affecting neural progenitor cells (NPCs). Here, we analyze NPCs from three pairs of dizygotic twins discordant for CZS. We compare by RNA-Seq the NPCs derived from CZS-affected and CZS-unaffected twins. Prior to Zika virus (ZIKV) infection the NPCs from CZS babies show a significantly different gene expression signature of mTOR and Wnt pathway regulators, key to a neurodevelopmental program. Following ZIKV in vitro infection, cells from affected individuals have significantly higher ZIKV replication and reduced cell growth. Whole-exome analysis in 18 affected CZS babies as compared to 5 unaffected twins and 609 controls excludes a monogenic model to explain resistance or increased susceptibility to CZS development. Overall, our results indicate that CZS is not a stochastic event and depends on NPC intrinsic susceptibility, possibly related to oligogenic and/or epigenetic mechanisms.


Sarcomas arise from primitive mesenchymal cells, which are classified, into two main groups: Bone and soft tissue sarcomas. We have searched all-important electronic databases including Google scholar and PubMed for the collection of latest literature pertaining to pediatric sarcomas. Latest literature confirmed that these tumors are relatively rare and represent only 1% of all malignancies but they have higher incidence in children. Pediatric sarcomas comprise about 13% of all pediatric malignancies and are ranked third in childhood cancers. The highest incidence rates are reported among rhabdomyosarcoma, osteosarcoma and Ewing’s sarcomas in children. All of these neoplasms often display highly aggressive behavior with tendency to form metastases. Important globally used management avenues include surgery with systemic chemotherapy and have success rate of 70% at 5-years. Furthermore, in the cases of advanced stages, the prognosis is poor, chances of treatment failure and recurrence are quite high. Utilization of cancer stem cells is the latest approach with great potential in management of above pathological state. The present review article discuss all-important aspects of commonly found pediatric sarcomas throughout the world.


Mitochondrial disorders are amongst the most common groups of inborn errors of metabolism. They are caused by deficiencies in the final pathway of the cellular energy production, the mitochondrial respiratory chain. The disorders are clinically and genetically heterogeneous and the aetiology could be found in the mitochondrial, or in the nuclear genome. We searched important e-databases for the collection of latest literature on the mitochondrial disease especially in pediatric population. Most of the studies in the recent past have focused on the understanding of the clinical phenotypes and pathophysiological mechanisms. Leigh syndrome is a common severe, neurodegenerative disease of early childhood. A defect in the POLG gene is another common observation in most of the cases leading to Alpers syndrome. The review concludes that pediatric mitochondrial disorders are severe, progressive and usually multi-systemic. Further, whole genome sequencing is an excellent diagnostic option.

Central nervous system (CNS) tumors are a leading cause of death in pediatric oncology. New drugs are desperately needed to improve survival. We evaluated the outcome of children and adolescents with CNS tumors participating in phase I trials within the Innovative Therapies for Children with Cancer (ITCC) consortium. Patients with solid tumors aged < 18 years at enrollment in their first dose-finding trial between 2000 and 2014 at eight ITCC centers were included retrospectively. Survival was evaluated using univariate/multivariate analyses. Overall, 114 patients were included (109 evaluable for efficacy). Median age was 10.2 years (range 1.0–17.9). Main diagnoses included: medulloblastoma/primitive neuroectodermal tumors (32.5%) and high-grade gliomas (23.7%). Complete/full responses (CR/PR) were reported in 7.3% patients and stable disease (SD) in 23.9%. Performance status at 90–100%, school/work attendance, normal ALT/AST and CR/PR/SD correlated with better overall survival (OS) in the univariate analysis. No variables assessable at screening/enrollment were associated with OS in the multivariate analysis. Five patients (4.5%) were discontinued from study due to toxicity. No toxic deaths occurred. Median OS was 11.9 months with CR/PR, 14.5 months with SD and 3.7 months with progressive disease (p < 0.001). The enrollment of children and adolescents with CNS tumors in phase I trials is feasible, safe and offers potential benefit for the patients. Sustained disease stabilization has a promising role as a marker of anti-tumor activity in children with CNS tumors participating in phase I trials.


OBJECTIVE: To describe the clinical course of paediatric patients undergoing surgery for congenital heart disease in UMAE of Yucatan. METHODS: Descriptive review was performed on the records of paediatric patients undergoing surgery for congenital heart disease from 1 November 2011 to 30 November 2013. RESULTS: The most frequent heart diseases were persistent ductus arteriosus (37.6%) and transposition of the great vessels. The median intensive care stay was 3 days. Mortality was 11.76%, with septic shock (44.4%) in most cases. The most frequent complications were sepsis (5.9%), low cardiac output syndrome (4.7%), cardiac arrest, and AV block and ventricular tachycardia (2.4% each). There was a moderate positive correlation between surgical complications and survival or death. CONCLUSIONS: The number of surgical patients is lower compared to reference centres for cardiovascular surgery. There is a marked tendency to perform corrective and palliative surgeries in specific disease in patients with added risk or ‘bad’ cardiac anatomy that prevent full correction at the first attempt. Prospective epidemiological and clinical studies should be conducted to understand the behaviour of congenital heart diseases treated in the region.


Importance: Neurologic disorders with isolated symptoms or complex syndromes are relatively frequent among mitochondrial inherited diseases. Recessive RTN4IP1 gene mutations have been shown to cause isolated and syndromic optic neuropathies. Objective: To define the spectrum of clinical phenotypes associated with mutations in RTN4IP1 encoding a mitochondrial quinone oxidoreductase. Design, Setting, and Participants: This study involved 12 individuals from 11 families with severe central nervous system diseases and optic atrophy. Targeted and whole-exome sequencing were performed at Hospital Angers (France), Institute of Neurology Milan (Italy), Imagine Institute Paris (France), Helmholt Zentrum of Munich (Germany), and Beijing Genomics Institute (China) to clarify the molecular diagnosis of patients. Each patient’s neurologic, ophthalmologic, magnetic resonance imaging, and biochemical features were investigated. This study was conducted from May 1, 2014, to June 30, 2016. Main Outcomes and Measures: Recessive mutations in RTN4IP1 were identified. Clinical presentations ranged from isolated optic atrophy to severe encephalopathies. Results: Of the 12 individuals in the study, 6 (50%) were male and 6 (50%) were female. They ranged in age from 5 months to 32 years. Of the 11 families, 6 (5 of whom were consanguineous) had a member or members who presented isolated optic atrophy with the already reported p.Arg103His or the novel p.Ile362Phe, p.Met43Ile, and p.Tyr51Cys amino acid changes. The 5 other families had a member or members who presented severe neurologic syndromes with a common core of
symptoms, including optic atrophy, seizure, intellectual disability, growth retardation, and elevated lactate levels. Additional clinical features of those affected were deafness, abnormalities on magnetic resonance images of the brain, stridor, and abnormal electroencephalographic patterns, all of which eventually led to death before age 3 years. In these patients, novel and very rare homozygous and compound heterozygous mutations were identified that led to the absence of the protein and complex I disassembly as well as mild mitochondrial network fragmentation. Conclusions and Relevance: A broad clinical spectrum of neurologic features, ranging from isolated optic atrophy to severe early-onset encephalopathies, is associated with RTN4IP1 biallelic mutations and should prompt RTN4IP1 screening in both syndromic neurologic presentations and nonsyndromic recessive optic neuropathies.


Mechanical birth-related injuries to the neonate are declining in incidence with advances in prenatal diagnosis and care. These injuries, however, continue to represent an important source of morbidity and mortality in the affected patient population. In the United States, these injuries are estimated to occur among 2.6% of births. Although more usual in context of existing feto-maternal risk factors, their occurrence can be unpredictable. While often superficial and temporary, functional and cosmetic sequelae, disability or even death can result as a consequence of birth-related injuries. The Agency for Healthcare research and quality (AHRQ) in the USA has developed, through expert consensus, patient safety indicators which include seven types of birth-related injuries including subdural and intracerebral hemorrhage, epiphragmatic subaponeurotic hemorrhage, skeletal injuries, injuries to spine and spinal cord, peripheral and cranial nerve injuries and other types of specified and non-specified birth trauma. Understandably, birth-related injuries are a source of great concern for the parents and clinician. Many of these injuries have imaging manifestations. This article seeks to familiarize the reader with the clinical spectrum, significance and multimodality imaging appearances of neonatal multi-organ birth-related trauma and its sequelae, where applicable. Teaching points * Mechanical trauma related to birth usually occurs with pre-existing feto-maternal risk factors. * Several organ systems can be affected; neurologic, musculoskeletal or visceral injuries can occur.* Injuries can be mild and transient or disabling, even life-threatening.* Imaging plays an important role in injury identification and triage of affected neonates.


Although whole-exome sequencing (WES) is the gold standard for the diagnosis of neurodevelopmental disorders (NDDs), it remains expensive for some genetic centers. Commercialized panels comprising all OMIM-referenced genes called "medical exome" (ME) constitute an alternative strategy to WES, but its efficiency is poorly known. In this study, we report the experience of 2 clinical genetic centers using ME for diagnosis of NDDs. We recruited 216 consecutive index patients with NDDs in 2 French genetic centers, corresponded to the daily practice of the units and included non-syndromic intellectual disability (NSID, n = 33), syndromic ID (NSID = 122), pediatric neurodegenerative disorders (n = 7) and autism spectrum disorder (ASD, n = 54). We sequenced samples from probands and their parents (when available) with the Illumina TruSight One sequencing kit. We found pathogenic or likely pathogenic variants in 56 index patients, for a global diagnostic yield of 25.9%. The diagnosis yield was higher in patients with ID as the main diagnosis (32%) than in patients with ASD (3.7%). Our results suggest that the use of ME is a valuable strategy for patients with ID when WES cannot be used as a routine diagnosis tool.


BACKGROUND: Cystic fibrosis (CF) is a chronic life-threatening disease. In patients who suffer from chronic disease, Attention Deficit Hyperactivity Disorder (ADHD) is associated with functional impairment that can affect
adherence to treatment and consequently influence prognosis. METHODS: CF patients filled in the ADHD Rating Scale (ADHD-RS) adapted to the DSM5 and were assessed on a continuous performance task (MOXO-CPT), a standardized-computerized test designed to evaluate several domains of attention. RESULTS: Of the 175 patients (99 males), 18% presented ADHD symptoms, according to ADHD-RS, 16% in the younger group (<18 years), and 18.9% in the adult group. The male to female ratio was 3:1 in children and 1:1 in adults. CONCLUSIONS: The occurrence of ADHD symptoms in patients with CF is substantially higher than in the general population and should be recognized as a co-morbidity of CF. As ADHD can impair adherence to therapy, further research is needed to investigate the effect of ADHD therapy on adherence. 


The objective of this study was to investigate racial/ethnic differences in survival for pediatric high-grade glioma (HGG) and medulloblastoma in the state of California. We obtained data from the California Cancer Registry on 552 high-grade glioma patients (110 brainstem, 442 non-brainstem) and 648 medulloblastoma patients ages 0-19 years from 1988 to 2012. Using multivariate Cox proportional hazards regression, we examined the impact of individual and neighborhood characteristics on survival. Socioeconomic quintile and insurance status differed significantly by race for both diagnoses. Hispanic children with non-brainstem HGG had worse survival than non-Hispanic white children: hazard ratio (HR) 1.62; 95% confidence interval (CI) 1.24-2.11, but the difference was mitigated some by accounting for socioeconomic status (HR 1.48, CI 1.10-1.99). Racial/ethnic differences in survival exist for children with high-grade glioma, particularly Hispanic children with non-brainstem high-grade glioma, and are likely related to sociologic factors. 


Evidence has demonstrated iron accumulation in specific brain regions of patients suffering from neurodegenerative disorders, and this metal has been recognized as a contributing factor for neurodegeneration. Using an experimental model of brain iron accumulation, we have shown that iron induces severe memory deficits that are accompanied by oxidative stress, increased apoptotic markers, and decreased synaptophysin in the hippocampus of rats. The present study aims to characterize iron loading effects as well as to determine the molecular targets of iron accumulation (brainstem), the main non-psychomimetic compound of Cannabis sativa, on mitochondria. Rats received iron in the neonatal period and CBD for 14 days in adulthood. Iron induced mitochondrial DNA (mtDNA) deletions, decreased epigenetic modulation of mtDNA, mitochondrial ferritin levels, and succinate dehydrogenase activity. CBD rescued mitochondrial ferritin and epigenetic modulation of mtDNA, and restored succinate dehydrogenase activity in iron-treated rats. These findings provide new insights into molecular targets of iron neurotoxicity and give support for the use of CBD as a disease modifying agent in the treatment of neurodegenerative diseases.


BACKGROUND: Acquired neutropenia in immunocompetent children is common and its differential diagnosis ranges from benign causes to life-threatening diseases. We described the etiology, clinical picture and outcome of new onset neutropenia in immunocompetent children assessed in the emergency department and hospitalized at our medical center. PATIENTS AND METHODS: Previously healthy children admitted with neutropenia (absolute neutrophil count [ANC] < 1.5 x 10/L) were included. Serious bacterial infections (SBI) were defined as culture-positive blood, urine, CSF, articular fluid or stool infections, pneumonia, Brucellosis and Rickettsiosis. RESULTS: 601 patients (aged 5 days-202 months) were enrolled; 3 (0.5%), 48 (8%), 165 (27.5%) and 385 (64%) had ANCs <0.2, 0.2-0.5, 0.5-1.0 and 1.0-1.5 x 10/L, respectively. Associated leukopenia and thrombocytopenia were diagnosed in 186 (39%) and 71 (11.8%) patients. 316/601 (52.6%) and 519/601 (86.4%) were <2 or 36 months of age, respectively. Fever at admission was present in 27.6% patients. SBIs were diagnosed in 106 (17.6) patients. Brucellosis and rickettsiosis were diagnosed in 8/52 (15.4%) and 9/39 (23.1%) tests obtained. RSV was diagnosed
in 17/33 (51.5%) nasal washes. An infectious etiology was determined in 171 (28.5%) patients. Acute leukemia was diagnosed in 6 patients. A significant correlation was found between resolution of neutropenia and patient age, infectious etiology and severity of neutropenia. CONCLUSIONS: 1. Severe neutropenia was rare; 2. More than half of patients were <2 months of age; 3. An infectious etiology was diagnosed in a high number of patients and SBIs were frequent; 4. Brucella spp. and rickettsial infections were frequent etiologies associated with neutropenia in our setting. https://www.ncbi.nlm.nih.gov/pubmed/29319583


Germline mutations in the SAMD9 and SAMD9L genes, located in tandem on chromosome 7, are associated with a clinical spectrum of disorders including the MIRAGE syndrome, ataxia-pancytopenia syndrome and myelodysplasia and leukemia syndrome with monosomy 7 syndrome. Germline gain-of-function mutations increase SAMD9 or SAMD9L’s normal antiproliferative effect. This causes pancytopenia and generally restricted growth and/or specific organ hypoplasia in non-hematopoietic tissues. In blood cells, additional somatic aberrations that reverse the germline mutation’s effect, and give rise to the clonal expansion of cells with reduced or no antiproliferative effect of SAMD9 or SAMD9L include complete or partial chromosome 7 loss or loss-of-function mutations in SAMD9 or SAMD9L. Furthermore, the complete or partial loss of chromosome 7q may cause myelodysplastic syndrome in these patients. SAMD9 mutations appear to associate with a more severe disease phenotype, including intratuerine growth restriction, developmental delay and hypoplasia of adrenal glands, testes, ovaries or thymus, and most reported patients died in infancy or early childhood due to infections, anemia and/or hemorrhages. SAMD9L mutations have been reported in a few families with balance problems and nystagmus due to cerebellar atrophy, and may lead to similar hematological disease as seen in SAMD9 mutation carriers, from early childhood to adult years. We review the clinical features of these syndromes, discuss the underlying biology, and interpret the genetic findings in some of the affected family members. We provide expert-based recommendations regarding diagnosis, follow-up, and treatment of mutation carriers. https://www.ncbi.nlm.nih.gov/pubmed/29535429


Duchenne Muscular Dystrophy (DMD; MIM 310200) is one of the most common and severe type of hereditary muscular dystrophies. The disease is caused by mutations in the dystrophin gene. The dystrophin gene is associated with X-linked recessive Duchenne and Becker muscular dystrophy. This disease occurs almost exclusively in males. The clinical symptoms of muscle weakness usually begin at childhood. The main symptoms of this disorder are gradually muscular weakness. The affected patients have inability to standing up and walking. Death is usually due to respiratory infection or cardiomyopathy. In this article, we have reported the discovery of a new nonsense mutation that creates abnormal stop codon in the dystrophin gene. This mutation was detected using Next Generation Sequencing (NGS) technique. The subject was a 17-year-old male with muscular dystrophy that who was suspected of having DMD. He was referred to Hakim medical genetics center of Neyshabur, IRAN. https://www.ncbi.nlm.nih.gov/pubmed/29246534


AIM: The aim of the study was to assess the incidence, mortality and morbidity of dilated cardiomyopathy (DCM) and noncompaction of the left ventricle (LVNC) in Swedish children. METHODS: We reviewed hospital records of all children with dilated cardiomyopathy (DCM) or left ventricular noncompaction cardiomyopathy (LVNC) up to the age of 18 in the healthcare region of western Sweden from 1991 to 2015. RESULTS: In total, 69 cases (61% males) were identified. The combined incidence of DCM and LVNC was 0.77 (95% CI 0.59-0.96) per 100 000 person years. Children were divided into six groups, and their outcomes were analysed depending on their aetiology. Idiopathic DCM was reported in 43%, and familial dilated and left ventricular noncompaction aetiology was present in 32%. DCM due to various diseases occurred in 8%. DCM associated with neuromuscular diseases was present in 16%. The overall risk of death or receiving transplants in children with idiopathic and familial DCM was 30% over the study period, and 21% died in the first year after diagnosis. CONCLUSION: The combined
incidence of DCM and LVNC was similar to previous reports. Most children with idiopathic DCM presented during infancy, and mortality was highest during the first year after diagnosis. https://www.ncbi.nlm.nih.gov/pubmed/29224255


Little is known about the specific causes of neonatal and under-five childhood death in high-mortality geographic regions due to a lack of primary data and dependence on inaccurate tools, such as verbal autopsy. To meet the ambitious new Sustainable Development Goal 3.2 to eliminate preventable child mortality in every country, better approaches are needed to precisely determine specific causes of death so that prevention and treatment interventions can be strengthened and focused. Minimally invasive tissue sampling (MITS) is a technique that uses needle-based postmortem sampling, followed by advanced histopathology and microbiology to definitely determine cause of death. The Bill & Melinda Gates Foundation is supporting a new surveillance system called the Child Health and Mortality Prevention Surveillance network, which will determine cause of death using MITS in combination with other information, and yield cause-specific population-based mortality rates, eventually in up to 12-15 sites in sub-Saharan Africa and south Asia. However, the Gates Foundation funding alone is not enough. We call on governments, other funders, and international stakeholders to expand the use of pathology-based cause of death determination to provide the information needed to end preventable childhood mortality. https://www.ncbi.nlm.nih.gov/pubmed/28719334


For many neuromuscular diseases (NMDs), cardiac disease represents a major cause of morbidity and mortality. The management of cardiac disease in NMDs is made challenging by the broad clinical heterogeneity that exists among many NMDs and by limited knowledge about disease-specific cardiovascular pathogenesis and course-modifying interventions. The overlay of compromise in peripheral muscle function and other organ systems, such as the lungs, also makes the simple application of endorsed adult or pediatric heart failure guidelines to the NMD population problematic. In this statement, we provide background on several NMDs in which there is cardiac involvement, highlighting unique features of NMD-associated myocardial disease that require clinicians to tailor their approach to prevention and treatment of heart failure. Undoubtedly, further investigations are required to best inform future guidelines on NMD-specific cardiovascular health risks, treatments, and outcomes. https://www.ncbi.nlm.nih.gov/pubmed/28838934


OBJECTIVE: We aimed to assess Child Death Overview Panel (CDOP) data validity, and cause of death classification, by comparison with information from a local birth cohort study (Born in Bradford, BiB), and another cause of death coding system (causes of death and associated conditions-CODAC). We then aimed to use CDOP data to calculate ethnic-specific infant mortality rates (IMRs), and compare characteristics of infants who died of congenital anomalies (CA) with those who died from other causes (non-CA). DESIGN: Retrospective cohort study. SETTING: Bradford Metropolitan District. PATIENTS: All infant deaths, 2008 to 2013. MAIN OUTCOME MEASURES: Infant mortality rates from CA and non-CA causes. RESULTS: 315 infant deaths were included, 56 of whom were BiB recruits. Agreement between CDOP and BiB was moderate to perfect for all characteristics except ethnicity, which showed weak agreement (kappa=0.58). The same deaths (27/56) were classified as CA by CDOP and CODAC. IMRs (per 1000 live births, 2009-2013) were highest in Pakistani infants (all causes 9.8, CA cause 5.5) compared with white British (all causes 4.3, CA cause 1.3) and other infants (all causes 5.1, CA cause 1.4). In multivariate analysis, infants who died of CA cause were more likely to have been born at term (OR 3.18) and to consanguineous parents (OR 3.28) than infants who died of non-CA cause. CONCLUSIONS: Excess Pakistani
mortality appears to be partly explained by an excess of deaths from CA, which in this population appears associated with a greater prevalence of consanguinity. 


BACKGROUND: Children and adolescents dying from complex chronic conditions require paediatric palliative care. One aim of palliative care is to enable a home death if desired and well supported. However, there is little data to inform care, particularly from countries without paediatric palliative care, which constitute the majority worldwide. METHODS: This is an epidemiological study analysing death certificate data of decedents aged between 0 and 17 years in Portugal, a developed Western European country without recognised provision of paediatric palliative care, from 1987 to 2011. We analysed death certificate data on cause and place of death; the main outcome measure was home death. Complex chronic conditions included cancer, cardiovascular, neuromuscular, congenital/genetic, respiratory, metabolic, gastro-intestinal, renal, and haematology/immunodeficiency conditions. Multivariate analysis determined factors associated with home death in these conditions. RESULTS: Annual deaths decreased from 3268 to 572. Of 38,870 deaths, 10,571 were caused by complex chronic conditions, their overall proportion increasing from 23.7% to 33.4% (22.4% to 45.4% above age 1-year). For these children, median age of death increased from 0.5 to 4.32-years; 19.4% of deaths occurred at home, declining from 35.6% to 11.5%; factors associated with home death were year of death (adjusted odds ratio 0.89, 95% confidence interval 0.89-0.90), age of death (6-10 year-olds 21.46, 16.42-28.04, reference neonates), semester of death (October-March 1.18, 1.05-1.32, reference April-September), and cause of death (neuromuscular diseases 1.59, 1.37-1.84, reference cancer), with wide regional variation. CONCLUSIONS: This first trend analysis of paediatric deaths in Portugal (an European country without paediatric palliative care) shows that palliative care needs are increasing. Children are surviving longer and, in contrast with countries where paediatric palliative care is thriving, there is a long-term trend of dying in hospital instead of at home. Age, diagnosis, season and region are associated with home death, and should be considered when planning services to support families choosing this option. Priorities should address needs of the youngest children, those with cancer, neuromuscular and cardiovascular conditions, as well as inequities related to place of residence. 


Occipital lobe epilepsy of childhood includes two entities: Panayiotopoulos syndrome in pre-school children, and idiopathic childhood occipital epilepsy of Gastaut (ICOEG) in school-age children. The typical initial manifestation of the former is vomiting, and that of the latter is visual hallucinations. Ictal cardiopulmonary arrest at initial presentation has been reported for Panayiotopoulos syndrome, but not for ICOEG. We document a 7-year-old previously healthy girl who experienced an acute elemental visual hallucination of seeing insects, followed by a new-onset generalized seizure. Upon arrival at the local hospital, she was unconscious and soon thereafter, developed respiratory arrest. She was resuscitated and initiated on mechanical ventilation. An electroencephalogram taken three days after seizure cessation showed frequent occipital spikes, consistent with the diagnosis of ICOEG. The sequence of acute elemental visual hallucination followed by a motor seizure, and then witnessed respiratory arrest illustrated occurrence of life-threatening autonomic involvement at initial onset in ICOEG. We speculate that the epileptic propagation from the occipital lobes eventually compromised the respiratory center in the brainstem. The possibility of occipital lobe epilepsy should be considered in school-age children presenting with acute visual hallucination followed by respiratory arrest. Such a presentation should prompt an urgent electroencephalogram and initiation of antiepileptic treatment if indicated. 


Survival for childhood central nervous system (CNS) tumours varies across Europe, partly because of the difficulty of distinguishing malignant from non-malignant disease. This study examines bias in CNS tumours survival analysis to obtain the reliable and comparable survival figures. We analysed survival data for about 15,000
children (age <15) diagnosed with CNS between 2000 and 2007, from 71 population-based cancer registries in 27 countries. We selected high-quality data based on registry-specific data quality indicators and recorded observed 1-year and 5-year survival by countries and CNS entity. We provided age-adjusted survival and used a Cox model to calculate the hazard ratios (HRs) of death, adjusting by age, site and grading by country. Recording of non-malignant lesions, use of appropriate morphology codes and completeness of life status follow-up differed among registries. Five-year survival by countries varied less when non-malignant tumours were included, with rates between 79.5% and 42.8%. The HRs of dying, for registries with good data, adjusting by age and grading, were between 0.7 and 1.2; differences were similar when site (supra- and infra-tentorial) was included. Several sources of bias affect the correct definition of CNS tumours, the completeness of incidence series and the goodness of follow-up. The European Network of Cancer Registries needs to improve childhood cancer registration and stress the need to update the International Classification for Cancer. Since survival differences persisted even when restricting the analysis to registries with satisfactory data, and since diagnosis of CNS tumours is difficult and treatment complex, national plans must aim for the revision of the diagnosis and the coordination of care, with adequate national and international networks.


**BACKGROUND:** Unique features and worse outcomes have been reported for cancers among adolescents and young adults (AYAs; 15-39 years old). The aim of this study was to explore the mortality and survival patterns of malignant central nervous system (CNS) tumors among AYAs in Southern-Eastern Europe (SEE) in comparison with the US Surveillance, Epidemiology, and End Results (SEER) program. METHODS: Malignant CNS tumors diagnosed in AYAs during the period spanning 1990-2014 were retrieved from 14 population-based cancer registries in the SEE region (n = 11,438). Age-adjusted mortality rates were calculated and survival patterns were evaluated via Kaplan-Meier curves and Cox regression analyses, and they were compared with respective 1990-2012 figures from SEER (n = 13,573). RESULTS: Mortality rates in SEE (range, 11.9-18.5 deaths per million) were higher overall than the SEER rate (9.4 deaths per million), with decreasing trends in both regions. Survival rates increased during a comparable period (2001-2009) in SEE and SEER. The 5-year survival rate was considerably lower in the SEE registries (46%) versus SEER (67%), mainly because of the extremely low rates in Ukraine; this finding was consistent across age groups and diagnostic subtypes. The highest 5-year survival rates were recorded for ependymomas (76% in SEE and 92% in SEER), and the worst were recorded for glioblastomas and anaplastic astrocytomas (28% in SEE and 37% in SEER). Advancing age, male sex, and rural residency at diagnosis adversely affected outcomes in both regions. CONCLUSIONS: Despite definite survival gains over the last years, the considerable outcome disparities between the less affluent SEE region and the United States for AYAs with malignant CNS tumors point to health care delivery inequalities. No considerable prognostic deficits for CNS tumors are evident for AYAs versus children. Cancer 2017;123:4458-71. (c) 2017 American Cancer Society.


Epileptic encephalopathies encompass a heterogeneous group of epilepsy syndromes that manifest with cognitive, behavioral, and neurologic deficits, seizures that are often intractable and multiform, aggressive electroencephalographic paroxysmal activity, and sometimes early death. As more is learned about the etiologies and manifestations of epileptic encephalopathies, progress has been made toward better treatment options. However, there is still a great need for further randomized controlled trials and research to help create clinically effective therapies. The 2015 Neurobiology of Disease in Children symposium, held in conjunction with the 44th annual meeting of the Child Neurology Society, aimed to (1) describe the clinical concerns involving diagnosis and treatment, (2) review the current status of understanding in the pathogenesis of epileptic encephalopathy, (3) discuss clinical management and therapies for epileptic encephalopathy, and (4) define future directions of
Pulmonary complications and liver failure. What is known: Neonatal presentation is a rare form of NPC with exclusively visceral involvement in the newborn period. A 1-month-old patient without any neurological deficit. CONCLUSIONS: Neonatal presentation is a rare form of NPC with an additional accompanying feature. All but one died due to pulmonary complications (n = 6) and liver insufficiency. Confirmation was done by molecular analysis, indicating NPC1 (n = 8) and NPC2 (n = 2) mutations. A history of parental consanguinity (n = 8) and first or second degree relative with NPC (n = 3). Patients were characterized by alterations in mental status, hypotonia, seizures, and abnormalities in feeding and respiration. The most common cause of neonatal encephalopathy is hypoxic-ischemic encephalopathy, for which treatment with 72 hours of therapeutic hypothermia is associated with reduced death or disability.


Purpose To assess the spectrum of associated anomalies, the intrauterine course, the outcome and possible prognostic markers in prenatally diagnosed Ebstein's anomaly (EA). Materials and Methods All cases of EA diagnosed over a period of 13 years with a minimum follow-up of 1 year were retrospectively collected in 4 tertiary referral centers in Germany. Results In the study period 76 cases of EA were prenatally diagnosed. The mean gestational age at diagnosis was 25.0 weeks (range: 13-35). 41 (53.9 %) cases were isolated and 35 (46.1 %) had other cardiac and/or extracardiac anomalies. 19 (25.0 %) pregnant women opted for termination of pregnancy. Intrauterine fetal death occurred in 7 cases (9.2 %), neonatal death in 14 cases (18.4 %), death in infancy or childhood in 9 cases (11.8 %) and 27 children (35.5 %) were alive at the last follow-up. After exclusion of terminations, the only parameter inversely correlated with intrauterine survival was hydrops fetalis. Prognostic parameters significantly associated with postnatal non-survival were an abnormal Celermajer index (right atrium/heart ratio > 0.7), cardiomegaly (cardiothoracic circumference ratio > 0.5), absence of antegrade flow over the pulmonary valve and earlier diagnosis in pregnancy. Conclusion Prenatally diagnosed EA has a high morbidity and mortality with the highest loss rate in the intrauterine and neonatal period. In our study, hydrops fetalis was the only parameter significantly associated with intrauterine demise, while other prenatal markers were only significantly associated with postnatal mortality.


Niemann-Pick disease type C (NPC) is a neurovisceral lysosomal storage disorder with a great variation in clinical spectrum and age at presentation. Clinical features of 10 NPC patients who presented in the newborn period between 1993 and 2015 at our center were retrospectively analyzed. Males and females were equally distributed; there was a history of parental consanguinity (n = 8) and first-degree relative with NPC (n = 3). Patients were symptomatic between 1 and 10 days (mean 3.6 +/- 2.6 days). Age at diagnosis was between 1 and 30 days (mean 14.6 +/- 13.3 days). Laboratory work-up included bone marrow aspiration (n = 8) and/or filipin staining (n = 4). Confirmation was done by molecular analysis, indicating NPC1 (n = 8) and NPC2 (n = 2) mutations. All patients had neonatal cholestasis and hepatosplenomegaly. Pulmonary involvement (n = 9) and fetal ascites (n = 2) were additional accompanying features. All but one died due to pulmonary complications (n = 6) and liver insufficiency (n = 3) between 1.5 and 36 months of age (mean 8.1 +/- 10.8 months). Currently, one patient is alive at the age of 11 months without any neurological deficit. CONCLUSIONS: Neonatal presentation is a rare form of NPC with exclusively visceral involvement in the newborn period and poor prognosis leading to premature death due to pulmonary complications and liver failure. What is known: * Neonatal presentation is a rare form of NPC with
Progressive liver disease is the most common cause of death among neonatal-onset NPC patients. What is new: * Natural course of neonatal-onset NPC may show variations. * Pulmonary involvement should be considered as an important cause of death in neonatal-onset cases, and appropriate precautions should be taken to prevent complications of respiratory insufficiency and airway infections.


BACKGROUND: Measurement of changes in health across locations is useful to compare and contrast changing epidemiological patterns against health system performance and identify specific needs for resource allocation in research, policy development, and programme decision making. Using the Global Burden of Diseases, Injuries, and Risk Factors Study 2016, we drew from two widely used summary measures to monitor such changes in population health: disability-adjusted life-years (DALYs) and healthy life expectancy (HALE). We used these measures to track trends and benchmark progress compared with expected trends on the basis of the Socio-demographic Index (SDI). METHODS: We used results from the Global Burden of Diseases, Injuries, and Risk Factors Study 2016 for all-cause mortality, cause-specific mortality, and non-fatal disease burden to derive HALE and DALYs by sex for 195 countries and territories from 1990 to 2016. We calculated DALYs by summing years of life lost and years of life lived with disability for each location, age group, sex, and year. We estimated HALE using age-specific death rates and years of life lived with disability per capita. We explored how DALYs and HALE differed from expected trends when compared with the SDI: the geometric mean of income per person, educational attainment in the population older than age 15 years, and total fertility rate. FINDINGS: The highest globally observed HALE at birth for both women and men was in Singapore, at 75.2 years (95% uncertainty interval 71.9–78.6) for females and 72.0 years (68.8–75.1) for males. The lowest for females was in the Central African Republic (45.6 years [42.0–49.5]) and for males was in Lesotho (41.5 years [39.0–44.0]). From 1990 to 2016, global HALE increased by an average of 6.24 years (5.97–6.48) for both sexes combined. Global HALE increased by 6.04 years (5.74–6.27) for males and 6.49 years (6.08–6.77) for females, whereas HALE at age 65 years increased by 1.78 years (1.61–1.93) for males and 1.96 years (1.69–2.13) for females. Total global DALYs remained largely unchanged from 1990 to 2016 (-2.3% [-5.9 to 0.9]), with decreases in communicable, maternal, neonatal, and nutritional (CMNN) disease DALYs offset by increased DALYs due to non-communicable diseases (NCDs). The exemplars, calculated as the five lowest ratios of observed to expected age-standardised DALY rates in 2016, were Nicaragua, Costa Rica, the Maldives, Peru, and Israel. The leading three causes of DALYs globally were ischaemic heart disease, cerebrovascular disease, and lower respiratory infections, comprising 16.1% of all DALYs. Total DALYs and age-standardised DALY rates due to most CMNN causes decreased from 1990 to 2016. Conversely, the total D Aly burden rose for most NCDs; however, age-standardised DALY rates due to NCDs declined globally. INTERPRETATION: At a global level, DALYs and HALE continue to show improvements. At the same time, we observe that many populations are facing growing functional health loss. Rising SDI was associated with increases in cumulative years of life lived with disability and decreases in CMNN DALYs offset by increased NCD DALYs. Relative compression of morbidity highlights the importance of continued health interventions, which has changed in most locations in pace with the gross domestic product per person, education, and family planning. The analysis of DALYs and HALE and their relationship to SDI represents a robust framework with which to benchmark location-specific health performance. Country-specific drivers of disease burden, particularly for causes with higher-than-expected DALYs, should inform health policies, health system improvement initiatives, targeted prevention efforts, and development assistance for health, including financial and research investments for all countries, regardless of their level of sociodemographic development. The presence of countries that substantially outperform others suggests the need for increased scrutiny for proven examples of best practices, which can help to extend gains, whereas the presence of underperforming countries suggests the need for devotion of extra attention to health systems that need more robust support. FUNDING: Bill & Melinda Gates Foundation.
The number of children without a diagnosis in pediatric palliative care and the process of decision-making in these children are widely unknown. The study was conducted as single-center retrospective cohort study. Between January 2013 and September 2016, 198 children and young adults were cared for; 27 (13.6%) of these were without a clear diagnosis at the start of pediatric palliative care. A definite diagnosis was ultimately achieved in three children. Median age was 7 years (0-25), duration of care 569 days (2-2638), and number of home visits 7.5 (2-46). Most patients are still alive (19; 70.4%). Median number of drugs administered was eight (range 2-19); antiepileptics were given most frequently. Despite the lack of a clear diagnosis (and thus prognosis), 13 (48.1%) parents faced with their critically ill and clinically deteriorating children decided in favor of a DNAR order. Comparing this with 15 brain-injured children, signs, symptoms, and supportive needs were similar in both groups. CONCLUSION: Children without a clear diagnosis are relatively common in pediatric palliative care and have-like all other patients-the right to receive optimized and symptom-adapted palliative care. Parents are less likely to choose treatment limitation for children who lack a definitive diagnosis. What is Known: * A clear diagnosis is usually considered important for best-practice pediatric palliative care (PPC) including advanced care planning (ACP). * Timely initiation of pediatric palliative care (PPC) is highly recommended in children with life-limiting conditions. What is New: * SWAN (syndrome without a name) children show similar signs and symptoms (mostly neurological) and have similar supportive needs as brain-injured children. * Defining treatment limitations in advance care planning is more difficult for parents of SWAN compared to brain-injured children.


The tragedy of epilepsy emerges from the combination of its high prevalence, impact upon sufferers and their families, and unpredictability. Childhood epilepsies are frequently severe, presenting in infancy with pharmaco-resistant seizures; are often accompanied by debilitating neuropsychiatric and systemic comorbidities; and carry a grave risk of mortality. Here, we review the most current basic science and translational research findings on several of the most catastrophic forms of pediatric epilepsy. We focus largely on genetic epilepsies and the research that is discovering the mechanisms linking disease genes to epilepsy syndromes. We also describe the strides made toward developing novel pharmacological and interventional treatment strategies to treat these disorders. The research reviewed provides hope for a complete understanding of, and eventual cure for, these childhood epilepsy syndromes.


Osteogenesis imperfecta (OI) is a rare hereditary skeletal disease leading to recurrent fractures, short stature and impaired mobility. The phenotype varies from mildly affected patients to perinatal lethal forms. In most cases an impaired collagen production due to mutations in COL1A1 or COL1A2 cause this hereditary bone fragility syndrome with an autosomal dominant inheritance. Currently an interdisciplinary therapeutic approach with antiresorptive drugs, physiotherapy and surgical procedures is the state of the art therapy. The effect of such a therapy is evaluated by measuring different surrogate parameters like areal bone mineral density or by using different mobility tests or questionnaires. Up till now the impact of these parameters on quality of life of the patients is not evaluated. Currently pharmacological strategies are based on antiresorptive treatment with bisphosphonates. In this trial we investigated the effect of an antiresorptive therapy with the monoclonal antibody denosumab decreasing the activity of osteoclasts. Denosumab was administered subcutaneously in a dose of 1mg/kg body weight in 10 children with OI (5-10 years of age) every 12 weeks for 48 weeks. Areal bone mineral density, mobility, pain scores and quality of life were measured. The results showed a good effect of the treatment on bone mineral density but this improvement showed no correlation to pain and quality of life. In conclusion further trials have to define parameters to assess interventions which influence activities of daily life of the patients. An interdisciplinary approach including physicians, basic researchers and patient organisations is needed to focus research on topics improving quality of life of patients with severe skeletal diseases.

Due to advances in caring for critically ill children and those with chronic diseases, rates of deep vein thrombosis (DVT) are increasing in children. Risk factors consist of central venous catheters, chronic medical conditions, thrombophilia, and various medications. Compression Doppler ultrasonography is the method most commonly used to diagnose DVT, and patients will usually present with pain and swelling of the affected limb. Anticoagulation via subcutaneous injection is the most common treatment regime for children with DVT, and the new, direct oral anticoagulants are currently under investigation. Prevention techniques are not established, but clinical studies are addressing this need. 


BACKGROUND: Recent estimates of the number of children and young people with life-limiting conditions derived from routine inpatient data are higher than earlier estimates using death record data. AIM: To compare routine inpatient data and death records as means of identifying life-limiting conditions in children and young people. DESIGN: Two national cohorts of children and young people with a life-limiting condition (primary cohort from England with a comparator cohort from Scotland) were identified using linked routinely collected healthcare and administrative data. PARTICIPANTS: A total of 37,563 children and young people with a life-limiting condition in England who died between 1 April 2001 and 30 March 2015 and 2249 children and young people with a life-limiting condition in Scotland who died between 1 April 2003 and 30 March 2014. RESULTS: In England, 16,642 (57%) non-neonatal cohort members had a life-limiting condition recorded as the underlying cause of death; 3364 (12%) had a life-limiting condition-related condition recorded as the underlying cause and 3435 (12%) had life-limiting conditions recorded only among contributing causes. In all, 5651 (19%) non-neonates and 3443 (41%) neonates had no indication of a life-limiting condition recorded in their death records. Similar results were seen in Scotland (overall, 16% had no indication of life-limiting conditions). In both cohorts, the recording of life-limiting condition was highest among those with haematology or oncology diagnoses and lowest for genitourinary and gastrointestinal diagnoses. CONCLUSION: Using death record data alone to identify children and young people with life-limiting condition - and therefore those who would require palliative care services - would underestimate the numbers. This underestimation varies by age, deprivation, ethnicity and diagnostic group.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5788076/


Pyruvate dehydrogenase complex (PDHC) deficiency is a rare metabolic disorder that affects tissues with high energy demand such as the central nervous system. The clinico-radiological phenotype of Leigh's disease is one of its common presentations. We present a 9-month-old boy with rapidly progressive infantile Leigh's disease. PDHA1 gene sequencing revealed a pathological homozygous missense mutation c.131A>G or p.H44R in exon 3 consistent with PDHC deficiency. H44R is among the five mutations (H44R, R88S, G89S, R263G, and V389fs) in E1 alpha subunit that is thiamine responsive. The child was initiated on thiamine, riboflavin, carnitine, coenzyme Q, and sodium benzoate supplementation. Mild recovery was noted at 6 months follow up as no further episodes of encephalopathy occurred. Thereafter, the child was treated with Ketonogenic diet which resulted in increased levels of activity and alertness. Despite an improving course, the child had a sudden unexpected death at the age of 21 months.


BACKGROUND: A substantial proportion of infants born with tetralogy of Fallot (TOF) die in infancy. A better understanding of the heterogeneity associated with TOF, including extracardiac malformations and chromosomal anomalies is vital to stratifying risk and optimizing outcomes during infancy. METHODS: Using the North Carolina Birth Defects Monitoring Program, infants diagnosed with TOF and born between 2003 and 2012 were included. Kaplan-Meier survival curves were used to estimate cumulative 1-year mortality, stratified by the presence of concomitant birth defects (BDs) and chromosomal anomalies. Multivariable logistic regression was used to estimate the direct effect of each concomitant BD, after adjusting for all others. RESULTS: A total of 496 infants

65
with TOF were included, and 15% (n = 76) died. The number of concomitant BD systems was significantly associated with the risk of death at 1-year, p < 0.0001. Specifically, the risk of mortality was 8% among infants with TOF with or without additional cardiac defects, 16% among infants with TOF and 1 extracardiac BD system, 19% among infants with 2 extracardiac BD systems, and 39% among infants with >/= 3 extracardiac BD systems. After adjustment, concomitant eye and gastrointestinal defects were significantly associated increased with 1-year mortality, odds ratio 2.83 (95% confidence interval, 1.08-7.32) and odds ratio 4.43 (95% confidence interval, 1.57, 12.45), respectively. Infants with trisomy 13 or trisomy 18 were also significantly more likely to die, p < 0.0001.

CONCLUSION: Both concomitant BDs and genetic anomalies increase the risk of mortality among infants with TOF. Future studies are needed to identify the underlying genetic and socioeconomic risk factors for high-risk TOF infants. Birth Defects Research 109:1154-1165, 2017. (c) 2017 Wiley Periodicals, Inc.


AIMS: Out-of-hospital sudden cardiac arrest (SCA) is a rare but devastating event in children and adolescents. The risk is assumed to be higher in children with congenital heart defects (CHDs) than in healthy individuals. The aim of the present study was to investigate the rate of and survival after out-of-hospital cardiac arrest in children 2-18 years old with CHDs. METHODS AND RESULTS: Data concerning all live births in Norway between 1994 and 2009 were retrieved from the Medical Birth Registry of Norway, the patient administrative systems at all hospitals in Norway, the Oslo University Hospital's Clinical Registry for Congenital Heart Defects and the Norwegian Cause of Death Registry. Survivors were followed through 2012, and supplementary information for the deceased children was retrieved from medical records at Norwegian hospitals. Among the 943 871 live births in Norway from 1994 to 2009, 11 272 (1.2%) children had a CHD. We identified 11 (0.1%) children 2-18 years old with CHDs who experienced out-of-hospital SCA. The estimated rate of out-of-hospital SCA in children 2-18 years old with CHD was 10 per 100 000 person-years. Early cardiopulmonary resuscitation was initiated in all patients. Three children survived. CONCLUSIONS: The incidence of and survival after out-of-hospital SCA in children with CHDs were comparable to the reported rates in the general child population.


BACKGROUND: Over half of under-five deaths occur in sub-Saharan Africa and appropriate, timely, quality care is critical for saving children's lives. This study describes the context surrounding children's deaths from the time the illness was first noticed, through the care-seeking patterns leading up to the child's death, and identifies factors associated with care-seeking for these children in rural Rwanda. METHODS: Secondary analysis of a verbal and social autopsy study of caregivers who reported the death of a child between March 2013 to February 2014 that occurred after discharge from the child's birth facility in southern Kayonza and Kirehe districts in Rwanda. Bivariate analyses using Fisher's exact tests were conducted to identify child, caregiver, and household factors associated with care-seeking from the formal health system (i.e., community health worker or health facility). Factors significant at alpha = 0.10 significance level were considered for backwards stepwise multivariate logistic regression, stopping when remaining factors were significantly associated with care-seeking at alpha = 0.05 significance level. RESULTS: Among the 516 eligible deaths among children under-five, 22.7% (n = 117) did not seek care from the health system. For those who did, the most common first point of contact was community health workers (45.8%). In multivariate logistic regression, higher maternal education (OR = 3.36, 95% CI: 1.89, 5.98), having diarrhea (OR = 4.21, 95% CI: 1.95, 9.07) or fever (OR = 2.03, 95% CI: 1.11, 3.72), full household insurance coverage (3.48, 95% CI: 1.79, 6.76), and longer duration of illness (OR = 22.19, 95% CI: 8.88, 55.48) were significantly associated with formal care-seeking. CONCLUSION: Interventions such as community health workers and insurance promote access to care, however a gap remains as many children had no contact with the health system prior to death and those who sought formal care still died. Further efforts are needed to respond to urgent cases in communities and further understand remaining barriers to accessing appropriate, quality care.


BACKGROUND: Leukodystrophies consist of degenerative neurogenetic diseases often associated with comorbidities that extend beyond the neurological system. Despite their impacts on patients' quality of life and risks of complications, head and neck symptomology is poorly reported in the literature. The objective of this study was to identify and quantify the main head and neck complaints among a cohort of patients diagnosed with leukodystrophies and define the role of the otolaryngologist as part of a multidisciplinary team for treating these patients.

METHODS: During the First Canadian National Conference on Leukodystrophies held at the Montreal's Children Hospital, a cohort of 12 patients diagnosed with leukodystrophies were recruited and evaluated by a multidisciplinary team. An otolaryngology-focused assessment was done through history and physical examination, and included a screening questionnaire for 23 common otolaryngology issues. If families reported a history of salivary, a validated questionnaire (Drool Quality of Life Assessment Questionnaire (DroolQoL)) was subsequently distributed. Results from the questionnaires were then compiled and analyzed. RESULTS: Of the 12 recruited patients, 83% (10/12) were known to an otolaryngologist. Drooling affected 67% (8/12) of patients although only 37.5% (3/8) of patients had undergone medical or surgical therapies for this issue. Four patients experienced at least one aspiration pneumonia. 58% (7/12) of the patients had dysphagia, of whom 43% (3/12) were fed exclusively via gastrostomy tube and 28% (2/7) required thickening of feeds. Two patients, despite suspicion of dysphagia and aspiration, had never undergone evaluation. As for otologic issues, it was noted that 25% (3/12) of patients had a history of pressure equalizing tubes (PETs) and one patient had a history of hearing loss. CONCLUSION: Head and neck comorbidities affect children with leukodystrophies. Therefore, the otolaryngologist should be part of the multidisciplinary team, specifically for the management of dysphagia and sialorrhea.


Trisomy 13 (T13) is accompanied by severe complications, and it can be challenging to achieve long-term survival without aggressive treatment. However, recently, some patients with T13 have been receiving home care. We conducted this study to investigate factors related to home health-care transition for patients with T13. We studied 28 patients with T13 born between January 2000 and December 2014. We retrospectively compared nine home care transition patients (the home care group) and 19 patients that died during hospitalization (the discharge at death group). The median gestational age of the patients was 36.6 weeks, with a median birth weight of 2,047 g. Currently, three patients (11%) have survived, and 25 (89%) have died. The home care group exhibited a significantly longer gestational age (38.9 vs. 36.3 weeks, p = 0.039) and significantly larger occipitofrontal circumference Z score (-0.04 vs. -0.09, p = 0.019). Congenital heart defects (CHD) was more frequent in the discharge at death group, with six patients in the home care group and 18 patients in the discharge at death group (67% vs. 95%, p = 0.047), respectively. Survival time was significantly longer in the home care group than in the discharge at death group (171 vs. 19 days, p = 0.012). This study has shown that gestational age, occipitofrontal circumference Z score at birth, and the presence of CHD are helpful prognostic factors for determining treatment strategy in patients with T13.


OBJECTIVE: Infantile-onset spinal muscular atrophy (SMA) is the most common genetic cause of infant mortality, typically resulting in death preceding age 2. Clinical trials in this population require an understanding of disease progression and identification of meaningful biomarkers to hasten therapeutic development and predict outcomes. METHODS: A longitudinal, multicenter, prospective natural history study enrolled 26 SMA infants and 27 control infants aged <6 months. Recruitment occurred at 14 centers over 21 months within the NINDS-sponsored NeuroNEXT (National Network for Excellence in Neuroscience Clinical Trials) Network. Infant motor function scales (Test of Infant Motor Performance Screening Items [TIMPSI], The Children’s Hospital of Philadelphia Infant Test for Neuromuscular Disorders, and Alberta Infant Motor Score) and putative physiological and molecular biomarkers were assessed preceding age 6 months and at 6, 9, 12, 18, and 24 months with
progression, correlations between motor function and biomarkers, and hazard ratios analyzed. RESULTS: Motor function scores (MFS) and compound muscle action potential (CMAP) decreased rapidly in SMA infants, whereas MFS in all healthy infants rapidly increased. Correlations were identified between TIMPSI and CMAP in SMA infants. TIMPSI at first study visit was associated with risk of combined endpoint of death or permanent invasive ventilation in SMA infants. Post-hoc analysis of survival to combined endpoint in SMA infants with 2 copies of SMN2 indicated a median age of 8 months at death (95% confidence interval, 6, 17). INTERPRETATION: These data of SMA and control outcome measures delineates meaningful change in clinical trials in infantile-onset SMA. The power and utility of NeuroNEXT to provide “real-world,” prospective natural history data sets to accelerate public and private drug development programs for rare disease is demonstrated. Ann Neurol 2017;82:883-891. https://www.ncbi.nlm.nih.gov/pubmed/29149772


AIM: To investigate if the serum biomarkers of cerebral injury, neuron-specific enolase and S100b protein, may classify unfavourable neurological outcome after paediatric cardiac arrest. METHODS: We performed a retrospective study of neuron-specific enolase and S100b measurements from 95 children treated in our paediatric cardiac intensive care unit after cardiac arrest. Neurological outcome at discharge was evaluated using the paediatric cerebral performance category scale, with unfavourable outcome defined as a change of >1 compared to pre-arrest status or death. RESULTS: Fifty-eight patients (61.1%) survived to discharge with 48 (50.5%) having a favourable neurological outcome. We observed significantly higher levels of both biomarkers in the unfavourable outcome group at designated time points (neuron-specific enolase at 24, 48, and 72h and S100b at 12, 24, and 48h after cardiac arrest, p<0.05). Receiver operating characteristic areas under the curve for neuron-specific enolase were 0.83, 0.80, and 0.73 at time points 24, 48, and 72h and 0.87, 0.81, and 0.82 for S100b at 12, 24, and 48h after cardiac arrest, respectively. Neuron-specific enolase measurement at 24h after cardiac arrest was an independent predictor of unfavourable outcome in a multivariable analysis. CONCLUSIONS: Neuron-specific enolase and S100b classify unfavourable neurological outcome in this large paediatric cardiac arrest cohort. Further multi-institutional prospective studies to comprehensively evaluate the diagnostic accuracy of these biomarkers under various clinical conditions and to determine reliable cut-off values in children are warranted. https://www.ncbi.nlm.nih.gov/pubmed/28939504


BACKGROUND: Classic Rett Syndrome (RS) is a disabling condition mainly caused by MECP2 mutations. Girls with RS are at risk of developing bone fragility and fractures at a young age which results in pain and may seriously impair quality of life. OBJECTIVE: To retrospectively assess the safety and efficacy of IV bisphosphonates on fracture, bone mineral density (BMD) and bone markers in RS girls with bone fragility. METHODS: RS girls received either IV pamidronate (n = 19) or IV zoledronate (n = 1) for 2 years. RESULTS: Of 20 patients studied (age: 12.5 years [6; 39]), 14 were non-ambulatory. The incidence of fracture decreased from 37 fractures in 20 patients, to 1 fracture during or after treatment (follow-up: 3.1 years [1.5; 5]). The spine BMD Z-score improved from -3.2 [-5.6; -0.1] to -2.2 [-3.8; 0.0], p = 0.0006. Most parents reported decreases in chronic pain and 2 patients started to walk. Urinary calcium excretion decreased from 0.7 [0.18; 1.5] to 0.2 [0.03; 0.67] mM/MV of creatinine (p = 0.0001). Pamidronate was well tolerated. CONCLUSION: RS girls should be screened for impaired bone mineralization and preventive measures should be taken. In girls experiencing fractures, IV bisphosphonates constitute a beneficial adjuvant treatment to diminish the risk of fracture and restore bone density. https://www.ncbi.nlm.nih.gov/pubmed/29073271


BACKGROUND: GM1 gangliosidosis is a rare lysosomal storage disorder caused by GLB1 mutations. Because of its extreme rarity and symptoms that overlap with other neurodegenerative diseases, its diagnosis is sometimes challenging, especially in the late infantile form with less severe phenotype. We aim to expand the clinical and genetic spectrum of late infantile GM1 gangliosidosis. METHODS: We confirmed a diagnosis of GM1...
CONCLUSION: There were distinctive geographic patterns of potential and realized need with high density of the Los Angeles area. Over 30 pediatric hospice providers supplied hospice care need. Sensitivity analysis revealed neighborhood surrounding the metropolitan areas of Los Angeles and San Diego had the highest density of realized need. The highest density of potential need was found in the areas surrounding Los Angeles. Almost 90% of the children and adolescents had a potential need for hospice care, whereas more than 10% had a realized need. Geographic information systems were used to create heat maps for analysis.

OBJECTIVE: To map and describe the geographic distribution of pediatric hospice care need versus supply in California over a 4-year time period (2007-2010). METHODS: Multiple databases were used for this descriptive longitudinal study. The sample consisted of 2036 children and adolescent decedents and 136 pediatric hospice providers. Geocoded data were used to create the primary variables of interest for this study-need and supply of pediatric hospice care. Geographic information systems were used to create heat maps for analysis. RESULTS: Almost 90% of the children and adolescents had a potential need for hospice care, whereas more than 10% had a realized need. The highest density of potential need was found in the areas surrounding Los Angeles. The areas surrounding the metropolitan communities of Los Angeles and San Diego had the highest density of realized hospice care need. Sensitivity analysis revealed neighborhood-level differences in potential and realized need in the Los Angeles area. Over 30 pediatric hospice providers supplied care to the Los Angeles and San Diego areas. CONCLUSION: There were distinctive geographic patterns of potential and realized need with high density of.
potential and realized need in Los Angeles and high density of realized need in the San Diego area. The supply of pediatric hospice care generally matched the needs of children and adolescents. Future research should continue to explore the needs of children and adolescents at end of life at the neighborhood level, especially in large metropolitan areas.


In the past 25 years, major advances were achieved in the nososcopy of cardiomyopathies, influencing the definition and taxonomy of this important chapter of cardiovascular disease. Nearly, 50% of patients dying suddenly in childhood or adolescence or undergoing cardiac transplantation are affected by cardiomyopathies. Novel cardiomyopathies have been discovered (arrhythmogenic, restrictive, and noncompacted) and added to update the World Health Organization classification. Myocarditis has also been named inflammatory cardiomyopathy. Extraordinary progress accomplished in molecular genetics of inherited cardiomyopathies allowed establishment of dilated cardiomyopathy as mostly cytoskeleton, force transmission disease; hypertrophic-restrictive cardiomyopathies as sarcomeric, force generation disease; and arrhythmogenic cardiomyopathy as desmosome, cell junction disease. Channelopathies (short and long QT, Brugada, and catecholaminergic polymorphic ventricular tachycardia syndromes) should also be considered cardiomyopathies because of electric myocyte dysfunction. Cardiomyopathies are easily diagnosed but treated only with palliative pharmacological or invasive therapy. Curative therapy, thanks to insights into the molecular pathogenesis, has to target the fundamental mechanisms involved in the onset and progression of these conditions.


Ovarian Sertoli-Leydig cell tumours (OSLCT) are rare and typically present with androgenic manifestations in women of the 2nd-3rd decade. Out of 228 diagnoses of ovarian sex cord-stromal tumours recorded at an academic institution during a 14-year period, eight women were surgically treated for OSLCT. Patient mean age was 54.8 years (range 19-81), five women being in the postmenopausal stage (62.5%). Only one woman presented with androgenic manifestations (12.5%), four with abnormal/postmenopausal uterine bleeding (50%), and three with abdominal pain (37.5%). Fertility sparing or radical surgery was performed depending on patient age and stage of disease. The only patient with an advanced disease (FIGO stage IV) was referred to palliative care postoperatively. The other seven were at FIGO stage I. Five of them were free from disease at a mean follow-up of 67 months, while the remaining two were lost at follow-up. The youngest woman of the series, treated with fertility-preserving unilateral salpingo-oophorectomy at the age of 19, had two spontaneous pregnancies and deliveries of healthy babies during a 10-year follow-up period. In conclusion, our single institution 14-year experience demonstrates that the diagnosis of OSLCT is particularly challenging since many patients are older than expected and lack androgenic manifestations. Impact statement * What is already known on this subjectOvarian Sertoli-Leydig cell tumours (OSLCT) are rare and are thought to typically present with androgenic manifestations in women of the 2nd-3rd decade. * What the results of this study addOur single institution 14-year experience shows that a high proportion of women with ovarian Sertoli-Leydig cell tumours may not present with androgenic manifestations, and many of them also are in the postmenopausal stage. Most patients have a good prognosis and fertility-preserving surgery in younger women can lead to spontaneous pregnancies and deliveries of healthy children after treatment. * What are the implications of these findings for clinical practice and/or further researchThe diagnosis of OSLCT is particularly challenging and therefore not reached before surgery in most of the cases. However, while hysterectomy with bilateral salpingo-oophorectomy and surgical staging are recommended for women with higher stage or no fertility wish, fertility-sparing surgery should be considered in younger women with early disease. Therefore, further research should focus on non-invasive diagnosis possibly by means of laboratory or imaging techniques.

Background: Monitoring levels and trends in premature mortality is crucial to understanding how societies can address prominent sources of early death. The Global Burden of Disease 2016 Study (GBD 2016) provides a comprehensive assessment of cause-specific mortality for 264 causes in 195 locations from 1980 to 2016. This assessment includes evaluation of the expected epidemiological transition with changes in development and where local patterns deviate from these trends. METHODS: We estimated cause-specific deaths and years of life lost (YLLs) by age, sex, geography, and year. YLLs were calculated from the sum of each death multiplied by the standard life expectancy at each age. We used the GBD cause of death database composed of: vital registration (VR) data corrected for under-registration and garbage coding; national and subnational verbal autopsy (VA) studies corrected for garbage coding; and other sources including surveys and surveillance systems for specific causes such as maternal mortality. To facilitate assessment of quality, we reported on the fraction of deaths assigned to GBD Level 1 or Level 2 causes that cannot be underlying causes of death (major garbage codes) by location and year. Based on completeness, garbage coding, cause list detail, and time periods covered, we provided an overall data quality rating for each location with scores ranging from 0 stars (worst) to 5 stars (best). We used robust statistical methods including the Cause of Death Ensemble model (CODEm) to generate estimates for each location, year, age, sex, and cause. 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birth complications, HIV/AIDS, chronic obstructive pulmonary disease, and neonatal encephalopathy due to birth asphyxia and trauma. Ischaemic heart disease was the leading cause of total YLLs in 113 countries for men and 97 countries for women. Comparisons of observed levels of YLLs by countries, relative to the level of YLLs expected on the basis of SDI alone, highlighted distinct regional patterns including the greater than expected level of YLLs from malaria and from HIV/AIDS across sub-Saharan Africa; diabetes mellitus, especially in Oceania; interpersonal violence, notably within Latin America and the Caribbean; and cardiomyopathy and myocarditis, particularly in eastern and central Europe. The level of YLLs from ischaemic heart disease was less than expected in 117 of 195 locations. Other leading causes of YLLs for which YLLs were notably lower than expected included neonatal preterm birth complications in many locations in both south Asia and southeast Asia, and cerebrovascular disease in western Europe. INTERPRETATION: The past 37 years have featured declining rates of communicable, maternal, neonatal, and nutritional diseases across all quintiles of SDI, with faster than expected gains for many locations relative to their SDI. A global shift towards deaths at older ages suggests success in reducing many causes of early death. YLLs have increased globally for causes such as diabetes mellitus or some neoplasms, and in some locations for causes such as drug use disorders, and conflict and terrorism. Increasing levels of YLLs might reflect outcomes from conditions that required high levels of care but for which effective treatments remain elusive, potentially increasing costs to health systems. FUNDING: Bill & Melinda Gates Foundation. https://www.ncbi.nlm.nih.gov/pubmed/28919116


Rapid whole-exome sequencing (rWES) is used in critically ill newborn infants to inform about diagnosis, clinical management, and prognosis. Here we report a male newborn infant with hydrops, pancytopenia, and acute liver failure who was listed for liver transplantation. Given the acuteness of the presentation, the procedure-related morbidity and mortality, and lack of diagnosis, we used rWES in the proband and both parents with a turnaround time of 10 business days. rWES returned one maternally inherited, likely pathogenic and one paternally inherited, likely pathogenic variant in NPC1, suggestive of a diagnosis of Niemann-Pick disease type C (NPC). Interestingly, a diagnosis of NPC was entertained prior to rWES, but deemed unlikely in light of absent cholesterol storage on liver biopsy and near-normal oxysterol levels in dried blood. The diagnosis of NPC was confirmed on filipin stain in fibroblasts demonstrating defective cholesterol trafficking. NPC is a slowly progressive neurodegenerative disorder that may also affect the liver with overall poor prognosis. It was decided to take the infant off the transplant list and transfer to palliative care, where he died after 4 wk. This case highlights the utility of rWES in an acute clinical setting for several domains of precision medicine including (1) diagnosis, (2) prognosis and outcome, (3) management and therapy, and (4) utilization of resources. https://www.ncbi.nlm.nih.gov/pubmed/28802248


Hereditary angioedema (HAE) with C1-inhibitor (C1-Inh) deficiency (C1-Inh-HAE) is a rare, life-threatening, and disabling genetic disorder characterized by self-limited tissue swelling caused by deficiency or dysfunction of C1-Inh. Our aim in this update is to discuss new advances in HAE therapy, focusing mainly on the various treatment options that have been available recently and also drugs that are under trial for prophylaxis to prevent attacks. There is a paradigm shift to where the treatment of HAE is headed, focusing now on prophylactic treatment rather than abortive management. https://www.ncbi.nlm.nih.gov/pubmed/28781749


OBJECTIVE: To define a distinct SCN1A developmental and epileptic encephalopathy with early onset, profound impairment, and movement disorder. METHODS: A case series of 9 children were identified with a profound developmental and epileptic encephalopathy and SCN1A mutation. RESULTS: We identified 9 children 3 to 12 years of age; 7 were male. Seizure onset was at 6 to 12 weeks with hemiconic seizures, bilateral tonic-clonic seizures, or spasms. All children had profound developmental impairment and were nonverbal and nonambulatory, and 7 of 9 required a gastrostomy. A hyperkinetic movement disorder occurred in all and was
OPINION STATEMENT: Leukemia is the most common pediatric cancer and accounts for approximately one third of childhood malignancies. There are germline genetic alterations that significantly increase the risk of developing hematopoietic malignancies in childhood. In this review, we describe a number of these hereditary disorders and their clinical features. These predispositions to cancer syndromes can be attributed to DNA repair/genetic instability, RAS pathway dysfunction, bone marrow failure, telomeropathies, immunodeficiencies, transcription factor abnormalities, pure familial leukemia, and aneuploidy. We focus especially on acute myeloid leukemia associated with Down syndrome, but also include other hereditary syndromes in this review. Recent advances in high-throughput genotyping technology have identified new genetic variants prone to human leukemia. Understanding of the mechanism of leukemia development in these hereditary syndromes allows us to gain insight into leukemogenesis in general and suggests therapeutic strategies based on these findings.


OBJECTIVE: To assess the spectrum of genetic anomalies in a cohort of children presenting at least one cerebral or spinal pial arteriovenous fistula (AVF), and to describe their clinical characteristics. METHODS: From 1988 to 2016, all consecutive patients with at least one cerebral or spinal pial AVF were screened for genetic disease. All patients aged <18 years were included. Symptoms associated with AVF were recorded: heart failure, neurological deficit/seizure, and hemorrhage. The outcome was assessed using the modified Rankin Scale and school performance in children with cerebral AVF and the American Spinal Injury Association impairment scale in children with spinal AVF. RESULTS: Forty-three children were included. Twenty-five children were male and 18 were female. A germline mutation was identified in 23 probands (53.5 +/- 14.9%): 8 in ENG (34.8 +/- 14.2%), 1 in ACVR1L (4.3 +/- 6%) leading to a diagnosis of HHT, and 14 in RASA1 (60.9 +/- 14.4%) leading to a diagnosis of capillary malformation/arteriovenous malformation type 1. No EphB4 gene mutation was identified. HHT patients presented a significantly lower rate of heart failure at diagnosis (p = 0.047). A trend toward an increased bleeding rate at presentation was observed in HHT (p = 0.069) and an increased rate of giant venous pouch in children in whom no mutation was identified (p = 0.097). Finally, an association with RASA1 mutation was observed in children with associated skin capillary hemangioma (p < 0001). INTERPRETATION: These results highlight the importance of genetic testing in this setting in view of the high frequency of gene mutations in pediatric cerebrospinal AVFs, and show the predominance of RASA1 over HHT mutations. Ann Neurol 2017;82:972-980.


INTRODUCTION: During recent decades, the prognosis of childhood acute lymphoblastic leukemia (ALL) has improved dramatically, nowadays, reaching a cure rate of almost 90%. These results are due to a better management and combination of old therapies, refined risk-group stratification and emergence of minimal residual disease (MRD) combined with treatment’s intensification for high-risk subgroups. However, the subgroup of patients with refractory/relapsed ALL still presents a dismal prognosis indicating necessity for innovative therapeutic approaches. Areas covered: We performed an exhaustive review of current first-line therapies for childhood ALL in the worldwide main consortia, summarized the major advances for front-line and relapse
treatment and highlighted recent and promising innovative therapies with an overview of the most promising ongoing clinical trials. Expert opinion: Two major avenues marked the beginning of 21(st) century. First, is the introduction of tyrosine-kinase inhibitor coupled to chemotherapy for treatment of Philadelphia positive ALL opening new treatment possibilities for the recently identified subgroup of Ph-like ALL. Second, is the breakthrough of immunotherapy, notably CAR T-cell and specific antibody-based therapy, with remarkable success observed in initial studies. This review gives an insight on current knowledge in these innovative therapeutic directions, summarizes currently ongoing clinical trials and addresses challenges these approaches are faced with.


PurposeProteus syndrome is a rare mosaic overgrowth disorder that is associated with severe complications. While anecdotal data have suggested that the life span of affected patients is reduced, this has not been measured. Mortality data on rare diseases is critical for assessing treatments and other interventions.MethodsTo address this we used the clinical research records of 64 patients in a longitudinal natural history cohort at the National Institutes of Health to ascertain the data in an organized manner and estimate survival using a Kaplan-Meier approach.

ResultsThe median age of diagnosis was 19 months. Based on this analysis, there was 25% probability of death by 22 years of age. Ten of the 11 patients who died were younger than 22 years of age, and there was only a single death after this age.

ConclusionThese data quantify the risk of premature death in Proteus syndrome, which can be used to support interventions and trials. Although the risk of death is substantial, the fact that only one patient died after 22 years of age supports anecdotal evidence that the disease process moderates after the end of adolescence. Interventions to reduce mortality should be targeted to the pediatric age range.


Spastic paraplegia type 5 (SPG5) is a rare subtype of hereditary spastic paraplegia, a highly heterogeneous group of neurodegenerative disorders defined by progressive neurodegeneration of the corticospinal tract motor neurons. SPG5 is caused by recessive mutations in the gene CYP7B1 encoding oxysterol-7alpha-hydroxylase. This enzyme is involved in the degradation of cholesterol into primary bile acids. CYP7B1 deficiency has been shown to lead to accumulation of neurotoxic oxysterols. In this multicentre study, we have performed detailed clinical and biochemical analysis in 34 genetically confirmed SPG5 cases from 28 families, studied dose-dependent neurotoxicity of oxysterols in human cortical neurons and performed a randomized placebo-controlled double blind interventional trial targeting oxysterol accumulation in serum of SPG5 patients. Clinically, SPG5 manifested in childhood or adolescence (median 13 years). Gait ataxia was a common feature. SPG5 patients lost the ability to walk independently after a median disease duration of 23 years and became wheelchair dependent after a median 33 years. The overall cross-sectional progression rate of 0.56 points on the Spastic Paraplegia Rating Scale per year was slightly lower than the longitudinal progression rate of 0.80 points per year. Biochemically, marked accumulation of CYP7B1 substrates including 27-hydroxycholesterol was confirmed in serum (n = 19) and cerebrospinal fluid (n = 17) of SPG5 patients. Moreover, 27-hydroxycholesterol levels in serum correlated with disease severity and disease duration. Oxysterols were found to impair metabolic activity and viability of human cortical neurons at concentrations found in SPG5 patients, indicating that elevated levels of oxysterols might be key pathogenic factors in SPG5. We thus performed a randomized placebo-controlled trial (EudraCT 2015-000978-35) with atorvastatin 40 mg/day for 9 weeks in 14 SPG5 patients with 27-hydroxycholesterol levels in serum as the primary outcome measure. Atorvastatin, but not placebo, reduced serum 27-hydroxycholesterol from 853 ng/ml [interquartile range (IQR) 683-1113] to 641 (IQR 507-694) (31.5%, P = 0.001, Mann-Whitney U-test). Similarly, 25-hydroxycholesterol levels in serum were reduced. In cerebrospinal fluid 27-hydroxycholesterol was reduced by 8.4% but this did not significantly differ from placebo. As expected, no effects were seen on clinical outcome parameters in this short-term trial. In this study, we define the mutational and phenotypic spectrum of SPG5, examine the correlation of disease severity and progression with oxysterol concentrations, and demonstrate in a randomized controlled trial that atorvastatin treatment can effectively lower 27-
hydroxycholesterol levels in serum of SPG5 patients. We thus demonstrate the first causal treatment strategy in hereditary spastic paraplegia.


OBJECTIVE: To describe the presentation and identify the cause of a new clinical phenotype, characterized by early severe neurodegeneration with myopathic and myasthenic features. METHODS: This case study of 5 patients from 3 families includes clinical phenotype, serial MRI, electrophysiologic testing, muscle biopsy, and full autopsy. Genetic workup included whole exome sequencing and segregation analysis of the likely causal mutation. RESULTS: All 5 patients showed severe muscular hypotonia, progressive cerebral atrophy, and therapy-refractory epilepsy. Three patients had congenital contractures. All patients died during their first year of life. In 2 of our patients, electrophysiologic testing showed abnormal decrement, but treatment with pyridostigmine led only to temporary improvement. Causative mutations in ALG14 were identified in all patients. The mutation c.220 G>A (p.Asp74Asn) was homozygous in 2 patients and heterozygous in the other 3 patients. Additional heterozygous mutations were c.422T>G (p.Val141Gly) and c.326G>A (p.Arg109Gln). In all cases, parents were found to be heterozygous carriers. None of the identified variants has been described previously. CONCLUSIONS: We report a genetic syndrome combining myasthenic features and severe neurodegeneration with therapy-refractory epilepsy. The underlying cause is a glycosylation defect due to mutations in ALG14. These cases broaden the phenotypic spectrum associated with ALG14 congenital disorders of glycosylation as previously only isolated myasthenia has been described. https://www.ncbi.nlm.nih.gov/pubmed/29126212


The outcome of treatment-refractory and/or relapsed pediatric T cell acute lymphoblastic leukemia (T-ALL) is extremely poor, and the genetic basis for this is not well understood. Here we report comprehensive profiling of 121 cases of pediatric T-ALL using transcriptome and/or targeted capture sequencing, through which we identified new recurrent gene fusions involving SPI1 (STMN1-SPI1 and TCF7-SPI1). Cases positive for fusions involving SPI1 (encoding PU.1), accounting for 3.9% (7/181) of the examined pediatric T-ALL cases, showed a double-negative (DN; CD4(-)CD8(-)) or CD8(+) single-positive (SP) phenotype and had uniformly poor overall survival. These cases represent a subset of pediatric T-ALL distinguishable from the known T-ALL subsets in terms of expression of genes involved in T cell precommitment, establishment of T cell identity, and post-beta-selection maturation and with respect to mutational profile. PU.1 fusion proteins retained transcriptional activity and, when constitutively expressed in mouse stem/progenitor cells, induced cell proliferation and resulted in a maturation block. Our findings highlight a unique role of SPI1 fusions in high-risk pediatric T-ALL. https://www.ncbi.nlm.nih.gov/pubmed/28733338


Pediatric brain tumors are the leading cause of childhood cancer mortality. Recurring genetic abnormalities play an essential role in the diagnosis and prognosis of pediatric brain tumors. However, clinical workup has not routinely included whole genome assessment. Here, we present high resolution whole genome array results in 11 pediatric brain tumors. Array identified clinically relevant abnormalities in all samples. Copy number aberrations with targeted therapy implication, GOPC-ROS1 fusion, CDK4 amplification, and NF1 deletion, were detected in 3 cases. In addition, array detected recurring genetic abnormalities, including KIAA1549-BRAF fusion, 19q13.42 amplification, i(17q), and monosomy 6, which assisted accurate histological diagnosis in pediatric brain tumors. In conclusion, our results show that whole genome high-resolution array detects diagnostic and treatment-relevant copy number abnormalities in pediatric brain tumors. https://www.ncbi.nlm.nih.gov/pubmed/28844173
This follow-up study of a subgroup of the patients seen in a natural history study of mucopolysaccharidosis type IIIA (Sanfilippo syndrome type A) addressed the adaptive and medical characteristics of their advanced disease manifestations. Of the original 24 patients, specific data was collected on only 58% primarily due to difficulty in locating families and coordinating time for interviews two to four years after the original study. At the last contact with the patient, age range was 8 to 24 years of age. Data were collected from telephone interviews from the Vineland Adaptive Behavior Scales II and medical and treatment history. We report the case data from rapid progressing and slow progressing patients separately. By the end of our data collection, 5 patients had died; 4 rapid progressing patients between 8 and 12 years of age and 1 slow progressing patient at age 21. Two patients were in out-of-home placements in the year before they died. We found that the incidence of surgeries and epilepsy was relatively low and that behavior problems largely subsided. Adaptive levels were very low with children functioning at below a two-year age equivalent level in all adaptive functions, but motor skills were slightly more intact. Only one slow progressing patient was functioning above a three-year level. Parent burden had shifted from behavioral control to physical management. Although their quality of life was clearly negatively impacted by physical management and palliative care, parents were more able to cope and adapt to such demands than in the initial stages of the disease.


BACKGROUND: Little information exists on health of children with developmental disabilities (DDs) in the Canadian province of Manitoba. METHOD: The present authors linked 12 years of administrative data and compared health status, changes in health and access to health and social services between children with (n = 1877) and without (n = 5661) DDs living in the province, matched by age, sex and region of residence. RESULTS: Children with DDs were significantly more likely than children in the matched comparison group to die before the age of 17 and have a history of respiratory illness, diabetes and injury-related hospitalizations. Children with DD also had significantly higher average number of ambulatory physician visits and higher rate of continuity of care. CONCLUSIONS: Children with DDs had poorer health status than the matched comparison group. The health disparities experienced by children with DDs persisted over time. Further population-based longitudinal research is needed in this area.


BACKGROUND: Duchenne muscular dystrophy (DMD) is the most common muscular dystrophy. There are no large studies describing its natural course from India. MATERIALS AND METHODS: Immunohistochemically/genetically confirmed DMD patients diagnosed between 1998 and 2014 were ambispectively included. The main aim was to study the natural course of motor milestones, i.e., age at onset of wheelchair status, bedbound state, and age at death, which were considered as primary outcome measures. We also correlated the DMD genotype with the motor milestones and other phenotypic features. RESULTS: A total of 500 DMD patients were included and 275 participated in the study. The mean age at symptom onset was 3.7 +/- 1.9 years, mean age at presentation was 8.1 +/- 2.5 years, and mean duration of illness was 4.4 +/- 2.6 years. On following them over 15 years, 155 (56.4%) had attained at least one of the primary outcome measures. Wheelchair status was attained in 124 (45.1%) [mean age: 10.4 +/- 1.6 years] and bedbound state in 24 (8.7%; mean age: 11.8 +/- 2.2 years) patients. Seven patients (2.6%) died during the follow-up period (mean age: 15.2 +/- 2.4 years). There was no significant impact of the genotypic or phenotypic features on the primary outcome. CONCLUSION: The pattern of major motor milestones (primary outcome measures) in this large cohort is comparable with that of the Western population despite variability in medical care. The genotypic pattern was
also similar to other large studies, which suggests that DMD is a more homogeneous disorder with limited ethnic variability in its geno-phenotypic expression.


Muscular dystrophies are a clinically and heterogeneous group of disorders that all share clinical characteristics of progressive muscular weakness. Duchenne muscular dystrophy (DMD) is the most common X-linked disorder muscular dystrophy in children, presenting in early childhood and characterized by proximal muscle weakness and calf hypertrophy in affected boys. There is usually delay in motor development and eventually wheelchair confinement followed by premature death from cardiac or respiratory complications. Treatment modalities such as corticosteroid therapy and use of intermittent positive pressure ventilation have provided improvements in function, ambulation, quality of life, and life expectancy, although novel therapies still aim to provide a cure for this devastating disorder. Here, we present a case of DMD in a 12-year-old male with remarkable clinical and oral manifestations.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5787973/


BACKGROUND/OBJECTIVES: This study aimed to investigate the association of dietary patterns with overweight risk and all-cause mortality in pediatric cancer patients. SUBJECTS/METHODS: Prospective cohort study was undertaken; 83 cancer patients admitted to the pediatric cancer ward at a university hospital in Seoul were included and followed for obesity and death over 24 months. Food consumption data were collected from patients using validated meal order sheets for breakfast, lunch, and dinner at the pediatric cancer ward over 3 days. Using principal component analysis, three dietary patterns were derived from 29 food groups. RESULTS: Eighteen deaths occurred among the patient cohort during the follow-up period. The “spicy & fried meat and fish” dietary pattern was positively associated with overweight risk at both baseline [odds ratio (OR) = 4.396, 95% confidence interval (CI) = 1.111-17.385, P for trend = 0.023] and after 6 months (OR = 4.088, 95% CI = 1.122-14.896, P for trend = 0.025) as well as all-cause mortality (hazard ratios = 5.124, 95% CI = 1.080-24.320, P for trend = 0.042), when comparing the highest and lowest tertiles after adjusting for covariates. The “fish, egg, meat, and fruits & vegetables” dietary pattern was associated with lower overweight risk after 24 months (OR = 0.157, 95% CI = 0.046-0.982, P for trend = 0.084). CONCLUSION: The results imply that dietary patterns might be associated with weight gain and premature death among pediatric cancer patients.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5712500/


Attention deficit hyperactivity disorder (ADHD), affecting 11% of children and adolescents, increases risk for injury and may predispose children to illness. However, the prevalence of ADHD and other developmental disorders in the pediatric intensive care unit (PICU) has not been previously studied. We performed a single-center, prospective cohort study of children aged 6 to 12 years who were hospitalized in the PICU from May through August 2016. Parents described their child’s educational and neurodevelopmental history, and completed ADHD and emotional/behavioral disorder screening on enrollment and 1 month after discharge. Twenty-four children were enrolled. Ten patients (42%) had a prior neurodevelopmental diagnosis, and 7 (29%) met study criteria for ADHD. Children hospitalized for critical illness have a high prevalence of neurodevelopmental disabilities and are more susceptible to the impact of critical illness on development and behavior. More research is needed to better understand how to support this vulnerable population after critical illness.


The field of intestinal transplantation has experienced dramatic growth since the first reported cases 3 decades ago. Improvements in operative technique, donor assessment and immunosuppressive protocols have afforded children who suffer from life-threatening complications of intestinal failure a chance at long-term survival. As
experience has grown, newer diseases, with more systemic manifestations have arisen as potential indications for transplant. After discussing the historical developments of intestinal transplant as a backdrop, this review focuses on the specific pre-operative indications for transplant as well as the great success that intestinal rehabilitation has witnessed over the past decade. A detailed discussion of evolution of immunosuppressive strategies is followed a general review of the common infectious complications experienced by children after intestinal transplant as well as the current long- and short-term results, including a section on new research on the quality of life in this challenging population of patients.


Fetal death is an important indicator of national health care. In Korea, the fetal mortality rate is likely to increase due to advanced maternal age and multiple births, but there is limited research in this field. The authors investigated the characteristics of fetal deaths, the annual changes in the fetal mortality rate and the perinatal mortality rate in Korea, and compared them with those in Japan and the United States. Fetal deaths were restricted to those that occurred at 20 weeks of gestation or more. From 2009 to 2014, the overall mean fetal mortality rate was 8.5 per 1,000 live births and fetal deaths in Korea, 7.1 in Japan and 6.0 in the United States. While the birth rate in Korea declined by 2.1% between 2009 and 2014, the decrease in the number of fetal deaths was 34.5%. The fetal mortality rate in Korea declined by 32.9%, from 11.0 in 2009 to 7.4 in 2014, the largest decline among the 3 countries. In addition, rates for receiving prenatal care increased from 53.9% in 2009 to 75.0% in 2014. Perinatal mortality rate I and II were the lowest in Japan, followed by Korea and the United States, and Korea showed the greatest decrease in rate of perinatal mortality rate II. In this study, we identified that the indices of fetal deaths in Korea are improving rapidly. In order to maintain this trend, improvement of perinatal care level and stronger national medical support policies should be maintained continuously.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5494332/


Postmortem examination of 7 neonates with congenital Zika virus infection in Brazil revealed microcephaly, ventriculomegaly, dystrophic calcifications, and severe cortical neuronal depletion in all and arthrogryposis in 6. Other findings were leptomeningeal and brain parenchymal inflammation and pulmonary hypoplasia and lymphocytic infiltration in liver and lungs. Findings confirmed virus neurotropism and multiple organ infection.


Developmental and epileptic encephalopathies (DEE) are a heterogeneous group of neurodevelopmental disorders with poor prognosis. Recent discoveries have greatly expanded the repertoire of genes that are mutated in epileptic encephalopathies and DEE, often in a de novo fashion, but in many patients, the disease remains molecularly uncharacterized. Here, we describe a new form of DEE in patients with likely deleterious biallelic variants in PTPN23. The phenotype is characterized by early onset drug-resistant epilepsy, severe and global developmental delay, microcephaly, and sometimes premature death. PTPN23 encodes a tyrosine phosphatase with strong brain expression, and its knockout in mouse is embryonically lethal. Structural modeling supports a deleterious effect of the identified alleles. Our data suggest that PTPN23 mutations cause a rare severe form of autosomal-recessive DEE in humans, a finding that requires confirmation.


Drugs and biologics developed to treat children with cancer have been historically developed in adults for adult indications. Although leading to many useful drugs and biologics to treat pediatric cancer, future development of molecularly targeted therapies (MTTs) should be directed toward pediatric tumors more specifically to maximize antitumor efficacy while minimizing acute morbidity and long-term disability. This will put pediatric clinicians closer to the goal of cure for all children diagnosed with cancer.


Gliomas are the most common CNS tumors in children and adolescents, and they show an extremely broad range of clinical behavior. The majority of pediatric gliomas present as benign, slow-growing lesions classified as grade I or II by the WHO classification of CNS tumors. These pediatric low-grade gliomas (LGGs) are fundamentally different from IDH-mutant LGGs occurring in adults, because they rarely undergo malignant transformation and show excellent overall survival under current treatment strategies. However, a significant fraction of gliomas develop over a short period of time and progress rapidly and are therefore classified as WHO grade III or IV high-grade gliomas (HGGs). Despite all therapeutic efforts, they remain largely incurable, with the most aggressive forms being lethal within months. Thus, the intentions of neurosurgeons, pediatric oncologists, and radiotherapists to improve care for pediatric patients with glioma range from increasing quality of life and preventing long-term sequelae in what is often a chronic, but rarely life-threatening disease (LGG), to uncovering effective treatment options to prolong patient survival in an almost universally fatal setting (HGG). The last decade has seen unprecedented progress in understanding the molecular biology underlying pediatric gliomas, fueling hopes to achieve both goals. Large-scale collaborative studies around the globe have cataloged genomic and epigenomic alterations in gliomas across ages, grades, and histologies. These studies have revealed biologic subgroups characterized by distinct molecular, pathologic, and clinical features, with clear relevance for patient management. In this review, we summarize hallmark discoveries that have expanded our knowledge in pediatric LGGs and HGGs, explain their role in tumor biology, and convey our current concepts on how these findings may be translated into novel therapeutic approaches.


Approximately 5000 to 10,000 children suffer an in-hospital cardiac arrest requiring cardiopulmonary resuscitation (CPR) each year in the United States. Importantly, 2% to 6% of all children admitted to pediatric intensive care units (ICUs) receive CPR, as do 4% to 6% of children admitted to pediatric cardiac ICUs. Survival from pediatric ICU cardiac arrest has improved substantially during the past 20 years presumably due to improved training methods, CPR quality, and post-resuscitation care. Extracorporeal life support CPR remains an important treatment option for both cardiac and noncardiac ICU patients.


PURPOSE: Adolescents with cancer have had less improvement in survival than other populations in the United States. This may be due, in part, to adolescents not receiving treatment at Children’s Oncology Group (COG) institutions, which have been shown to increase survival for some cancers. The objective of this ecologic study was to examine geographic distance to COG institutions and adolescent cancer mortality. METHODS: We calculated cancer mortality among adolescents and sociodemographic and healthcare access factors in four geographic zones at selected distances surrounding COG facilities: Zone A (area within 10 miles of any COG institution), Zones B and C (concentric rings with distances from a COG institution of >10-25 miles and >25-50 miles, respectively), and Zone D (area outside of 50 miles). RESULTS: The adolescent cancer death rate was highest in Zone A at 3.21 deaths/100,000, followed by Zone B at 3.05 deaths/100,000, Zone C at 2.94 deaths/100,000, and Zone D at 2.88 deaths/100,000. The United States-wide death rate for whites without Hispanic ethnicity, blacks without Hispanic ethnicity, and persons with Hispanic ethnicity was 2.96 deaths/100,000, 3.10 deaths/100,000, and 3.26 deaths/100,000, respectively. Zone A had high levels of poverty (15%), no health insurance coverage (16%), and no vehicle access (16%). CONCLUSIONS: Geographic access to COG institutions, as measured by distance alone, played no evident role in death rate differences across zones. Among adolescents,
socioeconomic factors, such as poverty and health insurance coverage, may have a greater impact on cancer mortality than geographic distance to COG institution.


BACKGROUND: Acute lymphoblastic leukemia (ALL) is the most common childhood malignancy. This report describes the survival of children with ALL in the United States using the most comprehensive and up-to-date cancer registry data. METHODS: Data from 37 state cancer registries that cover approximately 80% of the US population were used. Age-standardized survival up to 5 years was estimated for children aged 0-14 years who were diagnosed with ALL during 2 periods (2001-2003 and 2004-2009). RESULTS: In total, 17,500 children with ALL were included. The pooled age-standardized net survival estimates for all US registries combined were 95% at 1 year, 90% at 3 years, and 86% at 5 years for children diagnosed during 2001-2003, and 96%, 91%, and 88%, respectively, for those diagnosed during 2004-2009. Black children who were diagnosed during 2001-2003 had lower 5-year survival (84%) than white children (87%) and had less improvement in survival by 2004-2009. For those diagnosed during 2004-2009, the 1-year and 5-year survival estimates were 96% and 89%, respectively, for white children and 96% and 84%, respectively, for black children. During 2004-2009, survival was highest among children aged 1 to 4 years (95%) and lowest among children aged <1 year (60%). CONCLUSIONS: The current results indicate that overall net survival from childhood ALL in the United States is high, but disparities by race still exist, especially beyond the first year after diagnosis. Clinical and public health strategies are needed to improve health care access, clinical trial enrollment, treatment, and survivorship care for children with ALL. Cancer 2017;123:5178-89. Published 2017. This article is a U.S. Government work and is in the public domain in the USA. https://www.ncbi.nlm.nih.gov/pubmed/29205314


Despite significant advances in basic research, the treatment of degenerative diseases of the nervous system remains one of the greatest challenges for translational medicine. The childhood onset motor neuron disorder spinal muscular atrophy (SMA) has been viewed as one of the more tractable targets for molecular therapy due to a detailed understanding of the molecular genetic basis of the disease. In SMA, inactivating mutations in the SMN1 gene can be partially compensated for by limited expression of SMN protein from a variable number of copies of the SMN2 gene, which provides both a molecular explanation for phenotypic severity and a target for therapy. The advent of the first tailored molecular therapy for SMA, based on modulating the splicing behaviour of the SMN2 gene provides, for the first time, a treatment which alters the natural history of motor neuron degeneration. Here we consider how this will change the landscape for diagnosis, clinical management and future therapeutic trials in SMA, as well as the implications for the molecular therapy of other neurological diseases.


Philadelphia chromosome-like acute lymphoblastic leukemia (Ph-like ALL) is a subtype of B-lineage ALL (B-ALL) that displays a gene expression profile (GEP) similar to Philadelphia chromosome-positive ALL (Ph(+) ALL). It has a diverse range of genetic alterations that activate cytokine receptor genes and kinase signaling pathways, frequently accompanied by abnormal transcription factors related to lymphatic development. Children with Ph-like ALL account for 15% of children with high-risk B-ALL. It has adverse clinical features and a poor prognosis. Tyrosine kinase inhibitors combined with chemotherapy can significantly improve the prognosis of children with Ph(+) ALL, suggesting that targeted therapy based on the molecular cytogenetic abnormalities of Ph-like ALL has good research prospects. This paper expounds the genetic alterations, pathogenesis, clinical features, diagnostic measures, and potential therapeutic approaches of Ph-like childhood ALL based on recent research progress in Ph-like ALL.


Congenital and infantile malignant melanomas are rare and typically carry poor prognosis. The purpose of this article was to review the data on congenital and infantile malignant melanomas of the scalp in order to understand its presentation, diagnosis, management, and outcomes of congenital melanoma of scalp. We searched PubMed, CINAHL and Cochrane databases. Ten cases of congenital and 3 cases of infantile malignant melanoma of scalp were identified. The diagnosis was confirmed by biopsy and histological analysis for confirmation. The prognosis depends on the origin of disease (congenital melanocytic nevus, transplacental metastasis, or de-novo), tumor thickness, the presence of ulceration and/or necrosis, and anatomic site (scalp lesions having poor prognosis). The most commonly used treatment of the reported cases of congenital and infantile melanoma was surgical excision of the primary lesion. Further modes of treatment may be extrapolated from the treatment of childhood and adult melanomas.


Acute lymphoblastic leukemia is the most common (77%) childhood leukemia and also the most common neoplastic disease in children. Acute lymphoblastic leukemia initially present with hyperleukocytosis (WBC count more than 50,000/mm(3)) in twenty percent cases. These children are particularly at risk of development of tumor lysis syndrome (TLS). Tumor lysis syndrome is a metabolic disorder consists of hyperuricaemia, hyperkalemia, hyperphosphataemia, hypocalcaemia with or without renal insufficiency. It is the most common disease related emergency encountered by physicians caring for cancer of children and adult. This cross sectional observational study was conducted in Department of Paediatric Haematology and Oncology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh from November 2011 to April 2012 and was designed to assess early biochemical changes associated with hyperleukocytosis which may follow tumor lysis syndrome. Children with Acute lymphoblastic leukemia with hyperleukocytosis who are potential to develop tumor lysis syndrome were selected for the study in their initial presentation. Thirty patients with newly diagnosed acute lymphoblastic leukemia with initial high WBC count; more than 50,000/mm(3) were selected for the study. The objectives of the study were to observe the development of tumor lysis syndrome with hyperleukocytosis, detection of early signs of electrolyte changes and early detection of tumor lysis syndrome which can help further in the management of such patients. Majority of the cases were in age group 7 years to 15 years with male predominance (83%). Hyperphosphataemia (80%) followed by hyperuricaemia (53%) were the most common biochemical findings. Hyperkalemia and hypocalcaemia were present in 33% and 26% patients. Laboratory tumor lysis syndrome was developed in 40% of patients and clinical tumor lysis syndrome was developed in 20% patients with hyperleukocytosis meeting the criteria for definition of tumor lysis syndrome by Cairo Bishop in 2004. It was observed that TLS increased with higher WBC. Tumor lysis syndrome developed in 26% patients with WBC count below one lac, 50% with WBC count 1 lac to 2 lacs and increasing upto 100% with WBC count more than 3 lacs. Developing tumor lysis syndrome in high LDH (44% when LDH is more than 1000 unit/l) was also observed.


The United States has poorer child health outcomes than other wealthy nations despite greater per capita spending on health care for children. To better understand this phenomenon, we examined mortality trends for the US and nineteen comparator nations in the Organization for Economic Cooperation and Development for children ages 0-19 from 1961 to 2010 using publicly available data. While child mortality progressively declined across all countries, mortality in the US has been higher than in peer nations since the 1980s. From 2001 to 2010 the risk of death in the US was 76 percent greater for infants and 57 percent greater for children ages 1-19. During this decade, children ages 15-19 were eighty-two times more likely to die from gun homicide in the US. Over the fifty-year study period, the lagging US performance amounted to over 600,000 excess deaths. Policy interventions should focus on infants and on children ages 15-19, the two age groups with the greatest disparities, by addressing perinatal causes of death, automobile accidents, and assaults by firearm.

Rett syndrome (RTT) is a severe neurodevelopmental disorder typically affecting females. It is mainly caused by 


Colorectal carcinoma is a well-known malignancy in adults. However, it is rare in children. Besides, it also has different behaviour in paediatric age-group and usually presents with non-specific symptoms like abdominal pain, weight loss, and anaemia. This usually leads to delay in diagnosis. Adenocarcinoma in children has unfavourable tumour histology (mucinous subtype) and advanced disease stage at presentation which lead to poorer prognosis in children. Family history, genetic typing and sibling screening are essential components of management as this malignancy is frequently seen associated with hereditary syndromes. We describe a case of unusual presentation of rectal carcinoma in a 12-year-old girl.


large volumes of data are generated in hospital settings, including clinical and physiological data generated during the course of patient care. our goal, as proof of concept, was to identify early clinical factors or traits useful for predicting the outcome, of death, intubation, or transfer to ICU, for children with pediatric respiratory failure. we implemented both supervised and unsupervised methods to extend our understanding on statistical relationships in clinical and physiological data. as a supervised learning method, we use binary logistic regression to predict the risk of developing DIT outcome. next, we implemented unsupervised k-means algorithm on principal components of clinical and physiological data to further explore the contribution of clinical and physiological data on developing DIT outcome. our results show that early signals of DIT can be detected in physiological data, and two risk factors, blood pressure and oxygen level, are the most important determinant of developing DIT.

Diseases, Injuries, and Risk Factors Study 2016." The Global Burden of Disease, Injuries, and Risk Factors Study 2016 (GBD 2016) provides a comprehensive assessment of prevalence, incidence, and years lived with disability (YLDs) for 328 causes in 195 countries and territories from 1990 to 2016. METHODS: We estimated prevalence and incidence for 328 diseases and injuries and 2982 sequelae, their non-fatal consequences. We used DisMod-MR 2.1, a Bayesian meta-regression tool, as the main method of estimation, ensuring consistency between incidence, prevalence, remission, and cause of death rates for each condition. For some causes, we used alternative modelling strategies if incidence or prevalence needed to be derived from other data. YLDs were estimated as the product of prevalence and a disability weight for all mutually exclusive sequelae, correcting for comorbidity and aggregated to cause level. We updated the Socio-demographic Index (SDI), a summary indicator of income per capita, years of schooling, and total fertility rate. GBD 2016 complies with the Guidelines for Accurate and Transparent Health Estimates Reporting (GATHER). FINDINGS: Globally, low back pain, migraine, age-related and other hearing loss, iron-deficiency anaemia, and major depressive disorder were the five leading causes of YLDs in 2016, contributing 57.6 million (95% uncertainty interval [UI] 40.8-75.9 million [7.2%, 6.0-8.3]), 45.1 million (29.0-62.8 million [5.6%, 4.0-7.2]), 36.3 million (25.3-50.9 million [4.5%, 3.8-5.3]), 34.7 million (23.0-49.6 million [4.3%, 3.5-5.2]), and 34.1 million (23.5-46.0 million [4.2%, 3.2-5.3]) of total YLDs, respectively. Age-standardised rates of YLDs for all causes combined decreased between 1990 and 2016 by 2.7% (95% UI 2.3-3.1). Despite mostly stagnant age-standardised rates, the absolute number of YLDs from non-communicable diseases has been growing rapidly across all SDI quintiles, partly because of population growth, but also the ageing of populations. The largest absolute increases in total numbers of YLDs globally were between the ages of 40 and 69 years. Age-standardised YLD rates for all conditions combined were 10.4% (95% UI 9.0-11.8) higher in women than in men. Iron-deficiency anaemia, migraine, Alzheimer's disease and other dementias, major depressive disorder, anxiety, and all musculoskeletal disorders apart from gout were the main conditions contributing to higher YLD rates in women. Men had higher age-standardised rates of substance use disorders, diabetes, cardiovascular diseases, cancers, and all injuries apart from sexual violence. Globally, we


The most common neurodegenerative disease in childhood is spinal muscular atrophy (SMA). The severe infantile type 1 (Werdnig-Hoffman disease) makes 60% of SMA in total. These children usually die within 18 months without ventilation. New therapeutic approaches have led from the theoretical concept to randomized controlled clinical trials in patients. For the first time, a pharmacological treatment of SMA has been approved. The early detection of the disease is decisive for the success of therapy. All previous data suggest starting treatment early and when possible prior to the onset of symptoms considerably improves the outcome in comparison to a delayed start. The goal must be the presymptomatic diagnosis in order to initiate treatment before motor neuron degeneration. Technical and ethical prerequisites for a molecular genetic newborn screening are given.


Over the past several decades, neurofibromatosis type 1 (NF1) has become increasingly recognized as a neurodevelopmental disorder conferring increased risk for several important neurodevelopmental problems. In this review, we summarize the specific neurodevelopmental problems encountered in the context of NF1. These include impairments in general cognitive function, deficits in specific cognitive domains such as executive function and visuospatial processing and risk for specific learning disorders, impairments in attention and social skills and the overlap with attention-deficit-hyperactivity disorder and autism spectrum disorder, and the risk of developing other psychiatric conditions including anxiety and depression. Early recognition of these developmental impairments is important for the effective treatment of children with NF1, and further characterization is essential to improve our understanding of how mutations in the NF1 gene create the diversity of clinical neuropsychiatric symptomatology observed in this at-risk population.


BACKGROUND: As mortality rates decline, life expectancy increases, and populations age, non-fatal outcomes of diseases and injuries are becoming a larger component of the global burden of disease. The Global Burden of Diseases, Injuries, and Risk Factors Study 2016 (GBD 2016) provides a comprehensive assessment of prevalence, incidence, and years lived with disability (YLDs) for 328 causes in 195 countries and territories from 1990 to 2016. METHODS: We estimated prevalence and incidence for 328 diseases and injuries and 2982 sequelae, their non-fatal consequences. We used DisMod-MR 2.1, a Bayesian meta-regression tool, as the main method of estimation, ensuring consistency between incidence, prevalence, remission, and cause of death rates for each condition. For some causes, we used alternative modelling strategies if incidence or prevalence needed to be derived from other data. YLDs were estimated as the product of prevalence and a disability weight for all mutually exclusive sequelae, corrected for comorbidity and aggregated to cause level. We updated the Socio-demographic Index (SDI), a summary indicator of income per capita, years of schooling, and total fertility rate. GBD 2016 complies with the Guidelines for Accurate and Transparent Health Estimates Reporting (GATHER). FINDINGS: Globally, low back pain, migraine, age-related and other hearing loss, iron-deficiency anaemia, and major depressive disorder were the five leading causes of YLDs in 2016, contributing 57.6 million (95% uncertainty interval [UI] 40.8-75.9 million [7.2%, 6.0-8.3]), 45.1 million (29.0-62.8 million [5.6%, 4.0-7.2]), 36.3 million (25.3-50.9 million [4.5%, 3.8-5.3]), 34.7 million (23.0-49.6 million [4.3%, 3.5-5.2]), and 34.1 million (23.5-46.0 million [4.2%, 3.2-5.3]) of total YLDs, respectively. Age-standardised rates of YLDs for all causes combined decreased between 1990 and 2016 by 2.7% (95% UI 2.3-3.1). Despite mostly stagnant age-standardised rates, the absolute number of YLDs from non-communicable diseases has been growing rapidly across all SDI quintiles, partly because of population growth, but also the ageing of populations. The largest absolute increases in total numbers of YLDs globally were between the ages of 40 and 69 years. Age-standardised YLD rates for all conditions combined were 10.4% (95% UI 9.0-11.8) higher in women than in men. Iron-deficiency anaemia, migraine, Alzheimer's disease and other dementias, major depressive disorder, anxiety, and all musculoskeletal disorders apart from gout were the main conditions contributing to higher YLD rates in women. Men had higher age-standardised rates of substance use disorders, diabetes, cardiovascular diseases, cancers, and all injuries apart from sexual violence. Globally, we
noted much less geographical variation in disability than has been documented for premature mortality. In 2016, there was a less than two times difference in age-standardised YLD rates for all causes between the location with the lowest rate (China, 9201 YLDs per 100 000, 95% UI 6862-11943) and highest rate (Yemen, 14 774 YLDs per 100 000, 11 018-19 228). INTERPRETATION: The decrease in death rates since 1990 for most causes has not been matched by a similar decline in age-standardised YLD rates. For many large causes, YLD rates have either been stagnant or have increased for some causes, such as diabetes. As populations are ageing, and the prevalence of disabling disease generally increases steeply with age, health systems will face increasing demand for services that are generally costlier than the interventions that have led to declines in mortality in childhood or for the major causes of mortality in adults. Up-to-date information about the trends of disease and how this varies between countries is essential to plan for an adequate health-system response. FUNDING: Bill & Melinda Gates Foundation, and the National Institute on Aging and the National Institute of Mental Health of the National Institutes of Health.


**BACKGROUND:** Detailed assessments of mortality patterns, particularly age-specific mortality, represent a crucial input that enables health systems to target interventions to specific populations. Understanding how all-cause mortality has changed with respect to development status can identify exemplars for best practice. To accomplish this, the Global Burden of Diseases, Injuries, and Risk Factors Study 2016 (GBD 2016) estimated age-specific and sex-specific all-cause mortality between 1970 and 2016 for 195 countries and territories and at the subnational level for the five countries with a population greater than 200 million in 2016. METHODS: We have evaluated how well civil registration systems captured deaths using a set of demographic methods called death distribution methods for adults and from consideration of survey and census data for children younger than 5 years. We generated an overall assessment of completeness of registration of deaths by dividing registered deaths in each location-year by our estimate of all-age deaths generated from our overall estimation process. For 163 locations, including subnational units in countries with a population greater than 200 million with complete vital registration (VR) systems, our estimates were largely driven by the observed data, with corrections for small fluctuations in numbers and estimation for recent years where there were lags in data reporting (lags were variable by location, generally between 1 year and 6 years). For other locations, we took advantage of different data sources available to measure under-5 mortality rates (USMR) using complete birth histories, summary birth histories, and incomplete VR with adjustments; we measured adult mortality rate (the probability of death in individuals aged 15-60 years) using adjusted incomplete VR, sibling histories, and household death recall. We used the USMR and adult mortality rate, together with crude death rate due to HIV in the GBD model life table system, to estimate age-specific and sex-specific death rates for each location-year. Using various international databases, we identified fatal discontinuities, which we defined as increases in the death rate of more than one death per million, resulting from conflict and terrorism, natural disasters, major transport or technological accidents, and a subset of epidemic infectious diseases; these were added to estimates in the relevant years. In 47 countries with an identified peak adult prevalence for HIV/AIDS of more than 0.5% and where VR systems were less than 65% complete, we informed our estimates of age-sex-specific mortality using the Estimation and Projection Package (EPP)-Spectrum model fitted to national HIV/AIDS prevalence surveys and antenatal clinic serosurveillance systems. We estimated stillbirths, early neonatal, late neonatal, and childhood mortality using both survey and VR data in spatiotemporal Gaussian process regression models. We estimated abridged life tables for all location-years using age-specific death rates. We grouped locations into development quintiles based on the Socio-demographic Index (SDI) and analysed mortality trends by quintile. Using spline regression, we estimated the expected mortality rate for each age-sex group as a function of SDI. We identified countries with higher life expectancy than expected by comparing observed life expectancy to anticipated life expectancy on the basis of development status alone. FINDINGS: Completeness in the registration of deaths increased from 28% in 1970 to a peak of 45% in 2013; completeness was lower after 2013 because of lags in reporting. Total deaths in children younger than 5 years decreased from 1970 to 2016, and slower decreases occurred at ages 5-24 years. By contrast, numbers of adult deaths increased in each 5-year age bracket above the age of 25 years. The distribution of annualised rates of change in age-specific mortality rate differed over the period 2000 to 2016 compared with earlier decades: increasing annualised rates of change were less frequent, although rising annualised rates of change still occurred in some locations, particularly for adolescent and younger adult age groups. Rates of stillbirths and under-5 mortality both decreased globally from 1970. Evidence for global convergence of death rates was mixed; although the absolute difference between age-standardised death rates
narrowed between countries at the lowest and highest levels of SDI, the ratio of these death rates—a measure of relative inequality—increased slightly. There was a strong shift between 1970 and 2016 toward higher life expectancy, most noticeably at higher levels of SDI. Among countries with populations greater than 1 million in 2016, life expectancy at birth was highest for women in Japan, at 86.9 years (95% UI 86.7–87.2), and for men in Singapore, at 81.3 years (78.8–83.7) in 2016. Male life expectancy was generally lower than female life expectancy between 1970 and 2016, and the gap between male and female life expectancy increased with progression to higher levels of SDI. Some countries with exceptional health performance in 1990 in terms of the difference in observed to expected life expectancy at birth had slower progress on the same measure in 2016.

**INTERPRETATION:** Globally, mortality rates have decreased across all age groups over the past five decades, with the largest improvements occurring among children younger than 5 years. However, at the national level, considerable heterogeneity remains in terms of both level and rate of changes in age-specific mortality; increases in mortality for certain age groups occurred in some locations. We found evidence that the absolute gap between countries in age-specific death rates has declined, although the relative gap for some age-sex groups increased. Countries that now lead in terms of having higher observed life expectancy than that expected on the basis of development alone, or locations that have either increased this advantage or rapidly decreased the deficit from expected levels, could provide insight into the means to accelerate progress in nations where progress has stalled. **FUNDING:** Bill & Melinda Gates Foundation, and the National Institute on Aging and the National Institute of Mental Health of the National Institutes of Health.


Osteosarcoma is a common malignant tumor in childhood and adolescence (nearly 5% of all cases of cancer in children), as well as a type of tumor with poor prognosis. However, the pathogenesis and molecular mechanisms of osteosarcoma remains to be elucidated. The aim of the current study was to determine the association between methylation and gene expression changes in osteosarcoma cell line. Microarray data were obtained from the Gene Expression Omnibus database (GSE36004). Genomewide methylation status was determined in 19 different osteosarcoma cell lines and 6 normal controls. Differentially expressed genes (DEGs) were identified from cancer cells with genefilter package in R and differentially methylated sites were screened with CpGassoc package in R. Integrated gene expression with methylation profiles, genes differentially expressed and methylated, were obtained, and transcriptional regulatory network construction was performed. Functional annotation was performed for genes in the network using the DAVID online tool. Following integrated analysis, a total of 75 methylated sites were demonstrated to be localized at a transcription factor binding region. These sites may be bound by 83 transcription factors which will then alter the expression of 75 downstream DEGs. In the regulatory network, seizure related 6 homolog like 2 (SEZ6L2) had the highest degree of upregulation and was demonstrated to be regulated by 12 transcription factors. Furthermore, kin of IRRE like (KIRREL), centrosomal protein 72 (CEP72) and cyclindependent kinase 4 (CDK4) were also regulated by more than three transcription factors. Functional annotation revealed that the upregulated genes were primarily involved in the cell cycle pathway. Several differentially methylated sites were associated with upregulation of SEZ6L2, KIRREL, CEP72 and CDK4 and may have an important role in the pathogenesis of osteosarcomas through promotion of cell proliferation and metastasis.


DiGeorge/22q11.2 Deletion Syndrome (22q11DS) is a common genetic microdeletion syndrome that underlies several neurodevelopmental disorders including autism, attention deficit/hyperactivity disorder, and schizophrenia. In addition to cognitive impairments, those with 22q11DS have disrupted feeding and swallowing from birth onward. This perinatal dysphagia significantly compromises nutritional status, impairs appropriate weight gain, and can lead to life threatening aspiration-based infections. Appropriately timed excitation and inhibition of brainstem hypoglossal motor neurons, which innervate tongue muscles, is essential for proper feeding and swallowing. In this study we have examined changes in hypoglossal motor neuron function in the LgDel mouse model of 22q11DS. Hypoglossal motor neurons from LgDel mouse pups have action potentials with afterhyperpolarizations, mediated by a large conductance charybdotoxin-sensitive Ca-activated K current, that are significantly shorter in duration and greater in magnitude than those in wild-type pups. In addition, the amplitude, but not frequency, of glutamatergic excitatory glutamatergic postsynaptic currents (EPSCs) is
diminished, and GABAergic, but not glycineric, neurotransmission to hypoglossal motor neurons was reduced in LgDel animals. These observations provide a foundation for understanding the neurological changes in hypoglossal motor neuron function and their contribution to swallowing abnormalities that occur in DiGeorge/22q11.2 Deletion Syndrome.


Mortality surveillance and vital registration are limited in Sierra Leone, a country with one of the highest mortality rates among children aged <5 years worldwide, approximately 120 deaths per 1,000 live births (1,2). To inform efforts to strengthen surveillance, stillbirths and deaths in children aged <5 years from multiple surveillance streams in Bombali Sebora chiefdom were retrospectively reviewed. In total, during January 2015-November 2016, 930 deaths in children aged <5 years were identified, representing 73.3% of the 1,269 deaths that were expected based on modeled estimates. The “117” telephone alert system established during the Ebola virus disease (Ebola) epidemic captured 683 (73.4%) of all reported deaths in children aged <5 years, and was the predominant reporting source for stillbirths (n = 172). In the absence of complete vital events registration, 117 call alerts markedly improved the completeness of reporting of stillbirths and deaths in children aged <5 years.


The Regional Infant and Child Mortality Review Committee serves 10 counties in southeastern South Dakota and aims to use its reviews to prevent future loss of life during childhood. In 2016, the Committee reviewed 25 deaths (compared to 32 cases in 2013, 25 in 2014, and 24 in 2015). In 2016, three deaths in the region were attributable to maltreatment. This is an outlier from previous years when typically one such tragedy occurs and reveals the fragility of young life in stressed and unstable home environments. In 2016, there was also an increase from recent years in accidental deaths that included three children who were not properly restrained as passengers or while driving. In 2016, five infants died during sleep compared to seven the previous year and four of these deaths occurred with risks present in the sleep environment. The Committee has not seen progress towards decreasing infant deaths during sleep in our region. In fact, their number may even be increasing. The report provides the Committee's recommendations for community action that could prevent future deaths of infants and children.


Black patients have a twofold increased risk of induction mortality compared to White patients with acute myeloid leukemia (AML). We reviewed diagnosis and billing data from Pediatric Health Information System for 28 AML Induction I deaths to investigate conditions preceding death in White and Black patients. Half of deaths occurred within 10 days of initial diagnostic admission. Respiratory, cardiac, renal, and infectious complications were common prior to both White and Black deaths. Deaths in White patients were more commonly preceded by intracranial hemorrhage compared to deaths in Black patients. Future studies should assess management approaches of complications by race to identify modifiable processes that reduce mortality.


AIM: To describe the cases of Niemann-Pick type C (NP-C) disease in a United Kingdom epidemiological study of progressive intellectual and neurological deterioration in childhood. METHOD: Paediatricians notified cases via the British Paediatric Surveillance Unit between 1997 and 2015. RESULTS: Fifty-three NP-C patients were identified: 29 females, 24 males. Fifteen cases had a systemic presentation (neonatal jaundice and/or
hepatosplenomegaly). Thirty-eight had a neurological onset, the commonest presenting symptom being gait disturbance/ataxia (29 cases, 76%). Forty-nine cases eventually had neurological problems, the commonest were school/cognitive difficulties (40, 82%), seizures (33, 67%), dysphagia (20, 41%), dysarthria (18, 37%), cataplexy (17, 35%), and visual deterioration (8, 16%); their commonest abnormal physical signs were vertical supranuclear gaze palsy (35, 71%), hypotonia (19, 39%) and hepatosplenomegaly (19, 39%). The median diagnostic delay in the 38 neurological onset cases was 3 years (range 0.3-12.8). Confirmatory investigations included filipin staining of skin fibroblasts (42 cases), bone marrow examination in 30 (the last in 2011), and increasingly DNA studies, mutations in NP-C1 being found in 20 cases. INTERPRETATION: NP-C should be considered in children with progressive neurological deterioration. Subtle neurological problems combined with a history of prolonged neonatal jaundice and/or hepatosplenomegaly may provide early evidence of the disease.


Congenital anomalies that are diagnosed in at least 120,000 US infants every year are the leading cause of infant death and contribute to disability and pediatric hospitalizations. Several large-scale epidemiologic studies have provided substantial evidence of an association between congenital anomalies and cancer risk in children, suggesting potential underlying cancer-predisposing conditions and the involvement of developmental genetic pathways. Electronic medical records from 1,107 pediatric, adolescent, and young adult oncology patients were reviewed. The observed number (O) of congenital anomalies among children with a specific pediatric cancer subtype was compared to the expected number (E) of anomalies based on the frequency of congenital anomalies in the entire study population. The O/E ratios were tested for significance using Fisher's exact test. The Kaplan-Meier method was used to compare overall and neurological malignancy survival rates following tumor diagnosis. Thirteen percent of patients had a congenital anomaly diagnosis prior to their cancer diagnosis. When stratified by congenital anomaly subtype, there was an excess of neurological anomalies among children with central nervous system tumors (O/E = 1.56, 95%CI 1.13-2.09). Male pediatric cancer patients were more likely than females to have a congenital anomaly, particularly those <5 years of age (O/E 1.35, 95%CI 0.97-1.82). Our study provides additional insight into the association between specific congenital anomaly types and pediatric cancer development. Moreover, it may help to inform the development of new screening policies and support hypothesis-driven research investigating mechanisms underlying tumor predisposition in children with congenital anomalies.


Background: Pneumonia is now the second leading cause of death for children aged <5 years worldwide. However, analyses of the long-term evolution of under-5 mortality from pneumonia are still scarce in the literature. We aimed to explore long-term trends of under-5 mortality from pneumonia in 56 countries from 1960 to 2012. Methods: Data on under-5 mortality from pneumonia were extracted from the World Health Organization mortality database. Long-term trends were assessed for 56 countries and for 4 national income transition groups. We also used joinpoint regression analysis to detect distinct period segments of long-term trends and estimate the annual percent of changes of each period segment. Results: The average mortality rate from pneumonia for children aged 0-4 years in 56 countries declined from 163.0 per 100000 children (95% confidence interval [CI], 119.4 to 212.8) in 1960 to 9.9 per 100000 children (95% CI, 6.4 to 13.4) in 2012, with an average annual percent of change of -5.6% (95% CI, -7.2% to -3.9%). The temporal trends of childhood mortality were different between national income transition groups. Conclusions: Our findings suggest a striking overall downward trend in under-5 mortality from pneumonia between 1960 and 2012. However, the rate and absolute terms of decline differ by national income transition groups. These variable patterns between national income transition groups may inform further intervention setting and priority setting.


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Purpose: Pediatric sarcomas provide a unique diagnostic challenge. There is considerable morphologic overlap between entities, increasing the importance of molecular studies in the diagnosis, treatment, and identification of therapeutic targets. We developed and validated a genome-wide DNA methylation based classifier to differentiate between osteosarcoma, Ewing's sarcoma, and synovial sarcoma. Materials and Methods: DNA methylation status of 482,421 CpG sites in 10 Ewing’s sarcoma, 11 synovial sarcoma, and 15 osteosarcoma samples were determined using the Illumina Infinium HumanMethylation450 array. We developed a random forest classifier trained from the 400 most differentially methylated CpG sites within the training set of 36 sarcoma samples. This classifier was validated on data drawn from The Cancer Genome Atlas (TCGA) synovial sarcoma, TARGET Osteosarcoma, and a recently published series of Ewing’s sarcoma. Results: Methylation profiling revealed three distinct patterns, each enriched with a single sarcoma subtype. Within the validation cohorts, all samples from TCGA were accurately classified as synovial sarcoma (10/10, sensitivity and specificity 100%), all but one sample from TARGET-OS were classified as osteosarcoma (85/86, sensitivity 98%, specificity 100%) and 14/15 Ewing’s sarcoma samples classified correctly (sensitivity 93%, specificity 100%). The single misclassified osteosarcoma sample demonstrated high EWSR1 and ETV1 expression on RNA-seq although no fusion was found on manual curation of the transcript sequence. Two additional clinical samples, that were difficult to classify by morphology and molecular methods, were classified as osteosarcoma when previously suspected to be a synovial sarcoma and Ewing's sarcoma on initial diagnosis, respectively. Conclusion: Osteosarcoma, synovial sarcoma, and Ewing's sarcoma have distinct epigenetic profiles. Our validated methylation-based classifier can be used to provide diagnostic assistance when histological and standard techniques are inconclusive.

Outcomes and Instruments


OBJECTIVE: Trisomies 13 and 18 are among the most common autosomal aneuploidies associated with high mortality rates. Conventional management strategies offer to limit interventional support; however, some of the recent studies suggest that intervention does make a difference in terms of survival. STUDY DESIGN: A retrospective cohort study was performed between January 1996 and January 2016, covering all cases with such trisomies. A total of 69 cases were reviewed for clinical aspects, outcome, and management strategies. RESULTS: In almost all pregnancies with follow-up, at least one indication present for invasive testing (54/55). Invasive testing was not performed in 18.5% of such cases. All parents opted for termination in cases with prenatal diagnosis. None of the liveborns had prenatal diagnoses, thus, neonatal resuscitation and intensive care unit admission were not withheld in such infants. Major intervention was done in only one patient with full trisomy 13. Median survival for infants with full trisomies 13 and 18 was 36 and 60 days, respectively. Almost half the patients died within 1 month. CONCLUSION: To which extent the major interventions should be withheld is an issue of debate in managing such infants; however, current approaches are subject to change, given the technological advances.


OBJECTIVE: To measure the cross-informant variant of pediatric quality of life (QoL) based on self-reports and parent proxy measures. METHODS: A secondary analysis of baseline data obtained from two independent studies measuring the QoL based on the pediatric PROMIS-25 self-report and the PROMIS parent-proxy items banks. A scoring manual associated raw scores to a T score metric (mean = 50; SD = 10). Reliability of QoL ratings utilized the ICC while comparison of mean T Scores utilized the unpaired t-test. RESULTS: A total of 289 parent-child dyads comprised our study responders. Average age for parents and children was 41.27 years and 12.52 years, respectively. The mean T score (child self-report: parent proxy) for each QoL domains were: mobility (50.82:52.58), anxiety (46.73:44.21), depression (45.18:43.60), fatigue (45.59:43.92), peer-relationships (52.15:52.88) and pain interference (47.47:44.80). CONCLUSION: Parents tend to over-estimate their child’s QoL based on measures of anxiety, depression, fatigue, peer-relationships and pain interference.


BACKGROUND: Parents of very or extremely low birth weight infants have fewer subsequent children after preterm birth. Whether this applies to parents of less preterm infants is unknown. METHODS: In this nationwide cohort study, we identified all 230 308 traceable (>99%) singletons (9983 preterm, 4.3%) live born in Finland between January 1, 1987, and September 30, 1990, and their parents. Quantitative contribution of gestational age of child to the birth of parental subsequent children was assessed by multivariate Cox regression models, stratifying by the number of previous children. The impact of gestational age on sibling count was estimated at individual and population level. RESULTS: Mothers of extremely preterm (23-27 completed weeks) infants were, compared with mothers of term infants (39-41 weeks), less likely to have a subsequent live-born child (adjusted hazard ratio [HR]: 0.74; 95% confidence interval: 0.63-0.86). Corresponding HRs and confidence intervals were as follows: 28 to 31 weeks: 0.72 (0.65-0.80), 32 to 33 weeks: 0.82 (0.74-0.90), and 34 to 36 weeks: 0.90 (0.87-0.93). These HRs were consistent with those of fathers and couples. The cohort included 8002 firstborn preterm children, of whom 356 (4.4%) died in infancy. The 8002 children had a total of 13 826 subsequent siblings (1138 less than expected); per 1000 preterm births, this translates to the death of 44 preterm infants and 142 missing subsequent siblings. CONCLUSIONS: Families with a preterm singleton child have fewer subsequent children. In a high-income country, the main population effect of preterm birth is caused by these “missing siblings,” whose number exceeds the number of those preterm infants who die.


BACKGROUND: Oral mucositis can be a frequent and severe complication of chemotherapy in children. It can result in pain, infection, depression, prolonged admission, treatment delays, increase in patient morbidity, and increased costs. AIM: To record the prevalence and severity of oral mucositis among inpatients and explore the relationship of risks factors and the development of oral mucositis. DESIGN: During an 18-month period 643 clinical inpatient assessments were completed on 73 children who were admitted and had received chemotherapy in the last 14 days. RESULTS: There were 43 episodes of oral mucositis in 31 children; 42.5% of the inpatient population. World Health Organization assessment identified 32.6% of episodes were grade 1, 34.9% grade 2, 14.0% grade 3, and 18.6% grade 4. Analysis revealed significant associations between patient diagnosis (P<0.0001), chemotherapy cycles (P<0.0001), day 8 and 9 of the chemotherapy cycle (P<0.05), and neutropenia (P<0.0001) and oral mucositis. Children had increased length of admission with increasing severity of oral mucositis (P=0.0005). CONCLUSIONS: The prevalence of oral mucositis was 42.5% among inpatients and admission length was increased with increasing severity. Patient diagnosis, chemotherapy treatment block, day of chemotherapy cycle, and neutropenic status were shown to influence the risk of developing oral mucositis. https://www.ncbi.nlm.nih.gov/pubmed/29045267


OBJECTIVE: The objective was to describe outcomes and investigate factors affecting prognosis at 1 year post intervention for infants with surgical necrotising enterocolitis (NEC). DESIGN: Using the British Association of Paediatric Surgeons Congenital Anomalies Surveillance System, we conducted a prospective, multicentre cohort study of every infant reported to require surgical intervention for NEC in the UK and Ireland between 1 March 2013 and 28 February 2014. Association of independent variables with 1-year mortality was investigated using multivariable logistic regression analysis. SETTING: All 28 paediatric surgical centres in the UK and Ireland. PATIENTS: Infants were eligible for inclusion if they were diagnosed with NEC and deemed to require surgical intervention, regardless of whether that intervention was delivered. OUTCOMES: Primary outcome was mortality within 1 year of the decision to intervene surgically. RESULTS: 236 infants were included in the study. 208 (88%) infants had 1-year follow-up. 59 of the 203 infants with known survival status (29%, 95% CI 23% to 36%) died within 1 year of the decision to intervene surgically. Following adjustment, key factors associated with reduced 1-year mortality included older gestational age at birth (adjusted OR (aOR) 0.87, 95% CI 0.78 to 0.96). Being small for gestational age (SGA) (aOR 3.6, 95% CI 1.4 to 9.5) and requiring parenteral nutrition at 28 days post-decision to intervene surgically (aOR 3.5, 95% CI 1.1 to 11.03) were associated with increased 1-year mortality. CONCLUSIONS: Parents of infants undergoing surgery for NEC should be counselled that there is approximately a 1:3 risk of death in the first post-operative year but that the risk is lower for infants who are of greater gestational age at birth, who are not SGA and who do not require parenteral nutrition at 28 days post-intervention.


INTRODUCTION AND OBJECTIVES: Baclofen is a drug used mainly to treat muscle spasticity. Its overdose can lead to life-threatening clinical symptoms, including acute respiratory failure requiring mechanical ventilation. The aim of this study was to assess the prevalence of selected clinical symptoms associated with baclofen poisoning comparing to an ingested dose. MATERIAL AND METHODS: 60 cases of oral baclofen poisoning were analyzed. Gender, age distribution, and correlation between the dose of ingested baclofen were studied, as well as and following clinical parameters: degree of altered consciousness, heart rate, blood pressure, presence of acute respiratory failure, duration of mechanical ventilation, and presence of psychotic symptoms. RESULTS: The study found statistically significant correlations between dosage of ingested baclofen and presence of acute respiratory failure, as well as duration of mechanical ventilation. No statistically significant correlations were found between the dose of ingested baclofen and presence of hypertension, bradycardia, acute psychotic symptoms, or level of consciousness disturbance. However, it was found that patients who suffered from hypertension, bradycardia, and altered mental status ingested a larger dose of baclofen. CONCLUSIONS: There is a statistically significant
correlation between the dose of ingested baclofen and the presence of acute respiratory failure, and duration of mechanical ventilation. Patients who have taken a single dose of baclofen of 200 mg, or higher, should be managed in centres able to provide continuous monitoring of life functions. Those with a higher level of a single dose of baclofen ingestion (>500 mg), should be hospitalized in a Toxicology Unit or Intensive Care Unit able to provide airway support and mechanical ventilation.


OBJECTIVE AND BACKGROUND: Few previous studies have explored how pediatric palliative care (PPC) influences hospital utilization. We evaluated this among PPC recipients in a single center. METHODS: This is a retrospective cohort study of 109 patients >2 years of age who received PPC consultation at a large quaternary children’s hospital from April 2009 to September 2010. We assessed frequencies of hospital admissions and emergency department (ED) visits, use of intensive interventions, and hospital costs. Generalized estimating equations were used to compare outcomes in the two years before and after PPC consultation, stratifying by whether a patient survived two or more years following PPC enrollment. RESULTS: Median age at PPC consultation was 13 years (interquartile range 6-18); 56.0% were male (n = 61), 69.7% white non-Hispanic (n = 76). Fifty-nine percent (n = 64) of patients died during the study period. Overall, annual hospital admission rates decreased from 4.6 (95% confidence interval [CI] 4.0-5.4) before PPC consultation to 3.7 (95% CI 3.4-4.4) after (p = 0.025). Annual ED visits decreased from 0.9 (95% CI 0.7-1.2) to 0.6 (95% CI 0.4-0.8) (p = 0.030). Survivors had significantly decreased hospital admissions [rate ratio (RR) 0.57 (95% CI 0.45-0.73), p < 0.001] and ED visits [RR 0.33 (95% CI 0.20-0.54), p < 0.001]. Decedents had increased intensive care unit use (p = 0.029) but decreased operations (p = 0.002); survivors experienced no change in these outcomes after PPC consultation. Hospital costs remained stable for all (p = 0.929). DISCUSSION: PPC involvement may contribute to decreased hospital and ED use, without escalating costs. These outcomes are most evident in survivors. Hence, PPC may have a measurable long-term impact on hospital use in seriously ill children.


We conducted a retrospective cohort study of 125 pediatric oncology patients who died in 2010-2014 to explore how healthcare utilization, pediatric palliative care (PPC) receipt, and end-of-life care (EOLC) differed between patients enrolled in early phase clinical trials (EP) and those not enrolled (NEP). Baseline characteristics and healthcare utilization did not significantly differ between groups. EP patients received PPC consultation closer to death than NEP patients (median days before death = 58 [interquartile range = 16-84] vs. 85 [32-173]; P = 0.04). Our findings suggest that early phase trial enrollment does not substantially alter EOLC for children with advanced cancer but may contribute to later PPC engagement. Future studies should definitively assess the relationship between trial enrollment and PPC timing.


BACKGROUND: Valid observational pain scales are needed to assess pain and ensure sufficient treatment of pain in children that lack the verbal ability to self-report pain. Published reviews attempt to synthesize results from primary studies validating these scales and based on the findings recommendations may be given, for example which pain scales are the most appropriate for use in different pediatric populations. OBJECTIVES: The aims of this review were to describe how systematic reviews have evaluated and recommended observational pain scales for use in children aged 0-18 years and appraise the evidence underlying these recommendations. DESIGN: Systematic review of reviews. DATA SOURCES: The Cochrane Library, PubMed/MEDLINE, CINAHL, Web of Science, and PsychINFO were searched from inception to September 2016. Reference lists and gray literature were searched for additional studies. REVIEW METHODS: Study selection and data extraction were performed by two reviewers independently with a disagreement procedure in place. Methodological quality or study validity was measured using the Assessment of Multiple Systematic Reviews checklist and risk of bias or internal validity was measured using the Risk of Bias in Systematic Reviews tool. The review protocol was registered with PROSPERO:
OBJECTIVE: The goal of pediatric palliative care (PPC) is to maintain the quality of life (QoL) of children whose lives are threatened. However, there are sparse scientific data on the domains of QoL in this particular context, and no measurement strategies are available. The present study aims to describe the domains of QoL in the context of PPC in oncology, according to the perceptions of professional caregivers. METHOD: Semistructured interviews were conducted with a random sample of 20 professional caregivers from the Division of Hematology/Oncology at Le Centre Hospitalier Universitaire Sainte-Justine (Montreal, Canada). The caregivers were asked about their perceptions about the QoL of the children they have cared for in this context. The data were analyzed using inductive thematic content analysis. RESULTS: The analysis allowed us to identify seven domains of QoL: “physical comfort,” “alleviation of psychological suffering,” “fun and the present moment,” “sense of control,” “feeling valued and appreciated,” “feeling that life goes on,” and “meaningful social relationships.” SIGNIFICANCE OF RESULTS: Caregivers recount the regard that should be accorded to maintaining well-being and a sense of fun, as well as fostering the child’s abilities, taking account of the progression of the disease, and to fulfilling his or her needs, especially social ones. Our results also demonstrate that all domains were positively referred to by professional caregivers. The data from our study will lead to better assessment of QoL according to the trajectory of a child with advanced cancer while undergoing PPC.


RATIONALE: Multiple observational coding systems have been developed and validated to assess parent-child interactions during painful procedures. Most of these coding systems are neither theory-based nor do they well represent parent nonverbal behaviours. AIMS: Develop the Parent Caring Response Scoring System (P-CaReSS) based on Swanson’s Theory of Caring and test its psychometric properties in children in cancer port starts.

METHODS: A hybrid approach of inductive and deductive coding was used to formulate the preliminary observational codes for the P-CaReSS. Twenty-nine children, each with one video-recording of port start available, were selected from the parent study (R01CA138981) to refine the P-CaReSS, train coders and test inter-rater reliability. Videos of another 43 children were used to evaluate the construct validity of P-CaReSS. Per cent agreement and Cohen’s kappa were used to present the inter-rater reliability. Spearman rank-order correlations were used to report the construct validity. RESULTS: The 18-item P-CaReSS includes three types of parent behaviours: verbal, nonverbal and emotional behaviours. These parent interaction behaviours comprise five caring domains - knowing, being with, doing for, enabling, and maintaining belief - and one noncaring domain. On average the per cent agreement was 0.82 for the P-CaReSS overall, with average per cent agreements above 0.80 for both verbal and nonverbal behaviours. Kappa coefficient was 0.81 for the emotional behaviour. The behavioural codes in the P-CaReSS showed significant correlations with independent ratings of parent distress, child distress and child cooperation. CONCLUSIONS: The P-CaReSS is a promising tool that can be used to evaluate parent verbal, nonverbal and emotional behaviours during cancer-related port starts. This observational tool can be used to guide the development of nursing interventions to help parents caring for their child during cancer procedures.


Pediatric lung transplantation has been undertaken since the 1980s, and it is today considered an accepted therapy option in carefully selected children with end-stage pulmonary diseases, providing carefully selected children a net survival benefit and improved health-related quality of life. Nowadays, >100 pediatric lung transplants are done worldwide every year. Here, specific pediatric aspects of lung transplantation are reviewed such as the surgical challenge, effects of immunosuppression on the developing pediatric immune system, and typical infections of childhood, as it is vital to comprehend that children undergoing lung transplants present a real challenge as children are not ‘just small adults’. Further, an update on the management of the pediatric lung transplant patient is provided in this review, and future challenges outlined. Indications for lung transplantation in children are different compared to adults, the most common being cystic fibrosis (CF). However, the primary diagnoses leading to pediatric lung transplantation vary considerably by age group. Furthermore, there are regional differences regarding the primary indication for lung transplantation in children. Overall, early referral, careful patient selection and appropriate timing of listing are crucial to achieve real survival benefit. Although allograft function is to be preserved, immunosuppressant-related side effects are common in children post-transplantation. Strategies need to be put into practice to reduce drug-related side effects through careful therapeutic drug monitoring and lowering of target levels of immunosuppression, to avoid acute-reversible and chronic-irreversible renal damage. Instead of a ‘one fits all approach’, tailored immunosuppression and a personalized therapy is to be advocated, particularly in children. Further, infectious complications are a common in children of all ages, accounting for almost 50% of death in the first year post-transplantation. However, chronic lung allograft dysfunction (CLAD) remains the major obstacle for improved long-term survival.


INTRODUCTION: In this study we examined the feasibility of assessing motor milestone performance of infants with spinal muscular atrophy (SMA) using the Hammersmith Infant Neurological Exam-Part 2 (HINE-2) in a phase 2 study of nusinersen. METHODS: Nineteen SMA infants were assessed using the HINE-2 at baseline (<=7 months of age), and periodically up to 39 months of age. We evaluated whether the HINE-2 was feasible, reliable, and sensitive to change. RESULTS: Motor milestone assessments in SMA infants were feasible using the HINE-2. Baseline test-retest reliability was excellent (R = 0.987; P < 0.0001). SMA infants were extremely low functioning at baseline and the HINE-2 was able to detect changes over time in 16 of 19 infants within all 8 domains. HINE-2 improvements were correlated with changes in other neuromuscular outcome measures. CONCLUSION: Results support the use of the HINE-2 motor milestone assessment in clinical trials of SMA infants. Muscle Nerve 57: 143-146, 2017.


Breast milk is the recommended nutrition for infants. For preterm infants, when mother’s milk is not available, pasteurized donor milk is recommended (1). Non-Hispanic black mothers are at increased risk for having a preterm birth and for not breastfeeding (2,3); however, it is not known whether demographic disparities exist in the use of breast milk in neonatal intensive care units (NICUs). Data from CDC’s 2015 Maternity Practices in Infant Nutrition and Care (mPINC) survey, which does not collect patient-level demographics, were linked to the 2011-2015 U.S. Census Bureau’s American Community Survey (ACS)* to examine use of breast milk in NICUs based on demographic makeup of the hospital's postal code area. Among U.S. hospitals with a NICU, the use of mother’s own milk and donor milk were examined by the percentage of non-Hispanic black (black) residents in the hospital postal code area, categorized as being above or below the national average (12.3%). In postal codes with >12.3% black residents, 48.9% of hospitals reported using mothers’ own milk in >75% of infants in the NICU, and 38.0% reported not using donor milk, compared with 63.8% and 29.6% of hospitals, respectively, in postal codes with <=12.3% black residents. Further investigation is needed to understand variations in breast milk use in NICUs. Targeted efforts to increase breast milk use in hospitals located in postal codes where the percentage of black mothers is above the national average might help ensure more equitable access to breast milk for preterm and other high-risk infants.
Interventions for bereaved children and families range from supportive counseling, designed to promote social connectedness and expression of feelings and thoughts about the deceased, to intensive trauma/grief-specific therapy, designed to ameliorate symptoms of posttraumatic stress disorder (PTSD) and depression. That said, professionals have few brief assessment instruments to match response and functioning to appropriate interventions. To expedite the screening and referral process for bereaved families, Brown, Goodman, and Swiecicki (2008) developed the PTSD and Depression Screener for Bereaved Youth, a 19-item measure of bereavement-related history and symptoms of PTSD and depression. The current study is a psychometric evaluation of the Screener for Bereaved Youth. Data were collected from 284 bereaved children, 6-17 years of age (M = 12.4; SD = 2.9). A factor analysis revealed distinct subscales for PTSD (8 items) and Depression (4 items). The PTSD and Depression subscales showed both concurrent and discriminant validity. Endorsement of four items on either subscale was associated with meeting full criteria on more extensive measures of PTSD and depression. These findings are discussed with specific consideration to the multiple systems in which the measure could be used and applications to clinical services.


BACKGROUND: The question 'would you be surprised if this patient died in the next 12-months' is widely used for identifying adult patients in the last year of life. However, this has not yet been studied in children. AIM: To assess the prognostic accuracy of the surprise question when used by a multidisciplinary team to predict survival outcomes of children with life-limiting conditions over a 3 and 12 month period. DESIGN: A prospective cohort study. SETTING/PARTICIPANTS: Six multidisciplinary team members working in a children's hospice answered a 3 and 12 month surprise question about 327 children who were either newly referred or receiving care at the hospice between 2011 and 2013. RESULTS: The prognostic accuracy of the multidisciplinary team for the 3 (and 12)month surprise question were: sensitivity 83.3% (83.3%), specificity 93.2% (70.7%), positive predictive value 41.7% (23.6%), negative predictive value 99% (97.5%) and accuracy 92.6% (71.9%). Patients with a 'no' response had an increased risk of death at 3 (hazard ratio, 22.94, p 0.001) and 12 months (hazard ratio, 6.53, p 0.001). CONCLUSION: The surprise question is a highly sensitive prognostic tool for identifying children receiving palliative care who are in the last 3 and 12 months of life. The tool is accurate at recognising children during stable periods demonstrated through a high negative predictive value. In practice, this tool could help identify children who would benefit from specialist end of life care, act as a marker to facilitate communications on advance care planning and assist in resource allocation.


Respiratory syncytial virus (RSV), responsible for more than three million yearly hospitalizations and up to 118 000 deaths in children under 5 years, is the leading pulmonary cause of death for this age group that lacks a licensed vaccine. Ninety-nine percent of deaths due to the virus occur in developing countries. In-hospital RSV fatalities affect previously healthy term infants in association with bacterial sepsis, clinically significant pneumothoraces and, to a lesser extent, comorbid conditions. Community deaths affect low-income children from socially vulnerable families and appear to be as frequent as inpatient fatalities. In industrialized countries, RSV deaths occur almost exclusively in children with premorbid conditions. In a sense, RSV is an "opportunistic" killer. It needs a synergistic premorbid, medical practice-related, infectious, or social co-factor to cause a fatal outcome. But while the complex problems associated with these co-factors await solutions, candidate vaccines, long-lived monoclonal antibodies and antivirals against RSV are under clinical evaluation. It seems reasonable to predict that the landscape of RSV infections will look different in the next decade.

RATIONALE: Cerebellar liponeurocytoma is a rare tumor of the central nervous system (CNS) characterized by low proliferation but high likelihood of recurrence. Because of its rarity and the paucity of systematic follow-up, the biological behaviors and clinical features of this tumor are still poorly understood. We herein reported a case of cerebellar liponeurocytoma originating in the cerebral hemisphere. PATIENT CONCERNS: A 11-year-old male with intermittent headache, nausea, and vomiting. The first computed tomography revealed a large mass in the right cerebral hemisphere. He was transferred to our institution for neurosurgical treatment. DIAGNOSIS: Magnetic resonance imaging showed a large cystic-solid mass in the right fronto lobe with obvious contrast enhancement. Histopathological examinations showed sheets of isomorphic small neoplastic cells with clear cytoplasm and focal lipomatous differentiation. On immunohistochemistry, tumor cells were positive for synaptophysin, microtubule-associated protein 2, and neuronal nuclei antigen. INTERVENTIONS: The patient was performed a right frontoparietal craniotomy, and gross total resection of the tumor was achieved without adjuvant therapy. OUTCOMES: No clinical or neuroradiological evidence of recurrence or residual of the tumor was found 6 years and 2 months after initial surgery. LESSONS: Cerebellar liponeurocytoma developing in supratentorial cerebral hemisphere was first reported in the present study. The radiological and histopathological features may be useful in differentiating this rare tumor from other tumors at similar locations. A change in the nomenclature of cerebellar liponeurocytomas should be considered in future World Health Organization (WHO) classifications.  


BACKGROUND: Invasive mechanical ventilation (IMV) is a common practice in pediatric intensive care unit (PICU). However, the role of oxygenation (OI) and ventilation (VI) indices regarding the time on IMV has not been fully understood. BASIC PROCEDURES: The study was conducted with infants up to 24 months of age, hospitalized in PICU for two consecutive years. The values of ventilatory parameters, OI, VI, and blood gas of infants, collected in the first seven days in IMV, were associated with the time on IMV. IMV was classified into: short (< seven days) and long time (> seven days). The comparison was made from the first to the seventh day. Alpha=0.05. MAIN FINDINGS: Of 142 infants [mean age=7.51+/−6.33 months], 59 (41.5%) remained on IMV for a short time and 83 (58.5%) for a long time. Differences in PaO2 values were found on the second day, and PaO2/FiO2 ratio on the second, third and fourth days, with higher values in the short-term IMV. For FiO2 from the second to the fifth day; PInsp from the first to the seventh day; PEEP from the second to the sixth day; mechanical respiratory frequency from the second to the seventh day, PaCO2 on the second day; Paw from the first to the seventh day, OI from the second to the sixth day, and VI from the first to the seventh day, the values were higher in the long-term IMV. CONCLUSIONS: The OI and VI can be considered as potential predictors of long-term IMV, along with other markers obtained during the IMV.  


BACKGROUND: Perinatal stroke causes lifelong motor disability, affecting independence and quality of life. Non-invasive neuromodulation interventions such as transcranial direct current stimulation (tDCS) combined with intensive therapy may improve motor function in adult stroke hemiparesis but is under-explored in children. Measuring cortical metabolites with proton magnetic resonance spectroscopy (MRS) can inform cortical neurobiology in perinatal stroke but how these change with neuromodulation is yet to be explored. METHODS: A double-blind, sham-controlled, randomized clinical trial tested whether tDCS could enhance intensive motor learning therapy in hemiparetic children. Ten days of customized, goal-directed therapy was paired with cathodal tDCS over contralesional primary motor cortex (M1, 20 min, 1.0 mA, 0.04 mA/cm(2)) or sham. Motor outcomes were assessed using validated measures. Neuronal metabolites in both M1s were measured before and after intervention using fMRI-guided short-echo 3T MRS. RESULTS: Fifteen children [age(range) = 12.1(6.6-18.3) years] were studied. Motor performance improved in both groups and tDCS was associated with greater goal achievement. After cathodal tDCS, the non-lesioned M1 showed decreases in glutamate/glutamine and creatine while no metabolite changes occurred with sham tDCS. Lesioned M1 metabolite concentrations did not change post-intervention. Baseline function was highly correlated with lesioned M1 metabolite concentrations (N-acetyl-aspartate, choline, creatine, glutamate/glutamine). These correlations consistently increased in strength following
intervention. Metabolite changes were not correlated with motor function change. Baseline lesioned M1 creatine and choline levels were associated with clinical response. CONCLUSIONS: MRS metabolite levels and changes may reflect mechanisms of tDCS-related M1 plasticity and response biomarkers in hemiparetic children with perinatal stroke undergoing intensive neurorehabilitation. https://www.ncbi.nlm.nih.gov/pubmed/28958737


Atypical teratoid/rhabdoid tumors (AT/RT) are highly aggressive, malignant tumors and are the most common malignant brain tumor in children under 6 months of age. Currently, there is no standard treatment for AT/RT. Recent studies have reported potential anti-tumoral properties of ribavirin, a guanosine analog and anti-viral molecule approved by the Food and Drug Administration for treatment of hepatitis C. We previously demonstrated that ribavirin inhibited glioma cell growth in vitro and in vivo. Based on these results and the fact that no pre-clinical model of ribavirin in AT/RT exists, we decided to investigate the effect of ribavirin on several human AT/RT cell lines (BT12, BT16, and BT37) both in vitro and in vivo. We provide evidence that ribavirin has a significant impact on AT/RT cell growth and increases cell cycle arrest and cell death, potentially through modulation of the elf4E and/or EZH2 pathways. Interestingly, using scratch wound and transwell Boyden chamber assays, we observed that ribavirin also impairs AT/RT cell migration, invasion, and adhesion. Finally, we demonstrate that ribavirin significantly improves the survival of mice orthotopically implanted with BT12 cells. Our work establishes that ribavirin is effective against AT/RT by decreasing tumoral cell growth and dissemination and could represent a new therapeutic option for children with this deadly disease. https://www.ncbi.nlm.nih.gov/pubmed/29487714


OBJECTIVES: This study aimed to examine the role of nutrition in pediatric dilated cardiomyopathy (DCM). BACKGROUND: In adults with DCM, malnutrition is associated with mortality, whereas obesity is associated with survival. METHODS: The National Heart, Lung, and Blood Institute-funded Pediatric Cardiomyopathy Registry was used to identify patients with DCM and categorized by anthropometric measurements: malnourished (MN) (body mass index [BMI] <5% for age >/=2 years or weight-for-length <5% for <2 years), obesity (BMI >95% for age >/=2 years or weight-for-length >95% for <2 years), or normal bodyweight (NB). Of 904 patients with DCM, 23.7% (n = 214) were MN, 13.3% (n=120) were obese, and 63.1% (n=570) were NB. RESULTS: Obese patients were older (9.0 vs. 5.7 years for NB; p < 0.001) and more likely to have a family history of DCM (36.1% vs. 23.5% for NB; p = 0.023). MN patients were younger (2.7 years vs. 5.7 years for NB; p < 0.001) and more likely to have heart failure (79.9% vs. 69.7% for NB; p = 0.012), cardiac dimension z-scores >2, and higher ventricular mass compared with NB. In multivariable analysis, MN was associated with increased risk of death (hazard ratio [HR]: 2.06; 95% confidence interval [CI]: 1.66 to 3.65; p < 0.001); whereas obesity was not (HR: 1.49; 95% CI: 0.72 to 3.08). Competing outcomes analysis demonstrated increased risk of mortality for MN compared with NB (p = 0.03), but no difference in transplant rate (p = 0.159). CONCLUSIONS: Malnutrition is associated with increased mortality and other unfavorable echocardiographic and clinical outcomes compared with those of NB. The same effect of obesity on survival was not observed. Further studies are needed investigating the long-term impact of abnormal anthropometric measurements on outcomes in pediatric DCM. (Pediatric Cardiomyopathy Registry; NCT00005391). https://www.ncbi.nlm.nih.gov/pubmed/29428438


OBJECTIVES: To evaluate the effects of closed endotracheal tube suctioning on systemic oxygen saturation, cerebral regional oxygen saturation, and somatic regional (renal) oxygen saturation and hemodynamic variables in children. DESIGN: Prospective observational. SETTING: A tertiary care PICU. SUBJECTS: Children aged 0-18
years, requiring invasive mechanical ventilation and with an arterial line. INTERVENTIONS: Closed endotracheal suction. MEASUREMENTS AND MAIN RESULTS: The study included 19 sedated and intubated children, 0-18 years old. They were enrolled in an ongoing prospective observational study. We used near-infrared spectroscopy for cerebral regional oxygen saturation and somatic regional (renal) oxygen saturation. The timing of each closed endotracheal tube suctioning event was accurately identified from video recordings. We extracted systemic oxygen saturation, cerebral regional oxygen saturation, somatic regional (renal) oxygen saturation, heart rate, and systolic blood pressure and diastolic blood pressure for 5 minutes before and 5 minutes after each event and used these data for analysis. One-minute average values of these variables were used for repeated-measures analysis. We analyzed 287 endotracheal tube suctioning episodes in 19 children. Saline was instilled into the endotracheal tube during 61 episodes. The mean heart rate (107.0 +/- 18.7 vs 110.2 +/- 10.4; p < 0.05), mean arterial blood pressure (81.5 +/- 16.1 vs 83.0 +/- 15.6 mm Hg; p < 0.05), and the mean cerebral regional oxygen saturation (64.8 +/- 8.3 vs 65.8 +/- 8.3; p < 0.05) were increased after suctioning. The mean systemic oxygen saturation (96.9 +/- 2.7 vs 96.7 +/- 2.7; p = 0.013) was decreased, whereas the mean somatic regional (renal) oxygen saturation was not significantly different after endotracheal tube suctioning. Repeated-measures analysis revealed transient increases in heart rate, respiratory rate, systolic blood pressure, and diastolic blood pressure; a sustained increase in cerebral regional oxygen saturation; and transient decreases in systemic oxygen saturation and somatic regional (renal) oxygen saturation. Saline instillation did not affect oxygenation or hemodynamic variables. CONCLUSIONS: Closed endotracheal tube suctioning in sedated children is associated with transient but clinically insignificant changes in heart rate, blood pressure, cerebral regional oxygen saturation, systemic oxygen saturation, and somatic regional (renal) oxygen saturation. Saline instillation during endotracheal tube suctioning had no adverse effects on systemic or cerebral oxygenation.


BACKGROUND: Central venous pressure (CVP) is an important factor affecting capillary blood flow, and it is associated with poor outcomes in adult septic shock patients. However, whether a similar association exists in pediatric patients remains unclear. METHODS: We retrospectively analyzed data from patients admitted to our pediatric intensive care unit (PICU) between February 2009 and July 2015. Patients were divided into two groups-survivors and nonsurvivors-according to 28-day mortality. The associations between (a) mortality and CVP at 6, 24, 48, and 72 h after initiating treatment for established septic shock was analyzed and (b) initial serum lactic acid levels and 6-h CVP. RESULTS: Two hundred twenty-six patients were included in this study, and the mortality rate was 29.6% (67 deaths, nonsurvivor group). Initial serum lactic acid levels, Pediatric Risk of Mortality (PRISM) III score, and Vasoactive-Inotropic Score (VIS) within 24 h after PICU admission were significantly higher in the nonsurvivors than in survivors (1.3 [0.9, 2.4] vs. 3.9 [1.6, 8.0] mmol/l, 11.0 [7.0, 15.0] vs. 17.0 [10.0, 21.5], 12.0 [7.0, 25.0] vs. 22.5 [8.0, 55.0], respectively with p-values < 0.001, < 0.001, and 0.009, respectively). In addition, compared to survivors, a greater percentage of nonsurvivors required mechanical ventilation (92.5% vs. 51.6%, p < 0.001) and showed a greater extent of fluid overload at 48 h after admission (3.9% vs. 1.9%, p = 0.006), along with higher 6-h CVP (10.0 [7.0, 16.0] vs. 8.0 [5.0, 11.0] mmHg, p < 0.001). Patient survival according to levels of CVP (CVP < 8 mmHg, CVP 8-12 mmHg, or CVP > 12 mmHg) showed that the CVP > 12-mmHg group had significantly greater mortality rates (50.0%, p = 0.002) than the other groups (21.3% and 27.5%). Furthermore, multivariate analysis identified significant associations of CVP > 12 mmHg, serum lactic acid levels, and the need for mechanical ventilation with mortality (OR: 2.74, 1.30, and 12.51, respectively; 95% CI: 1.11-6.72, 1.12-1.50, and 4.12-37.96, respectively). CONCLUSIONS: Elevated CVP is an independent risk factor for mortality in pediatric septic shock patients.


OBJECTIVE: To evaluate the in-hospital consequences of prolonged respiratory support with invasive mechanical ventilation in very low birth weight infants. STUDY DESIGN: A cohort study was performed using prospectively collected data from 69 neonatal intensive care units participating in the Korean national registry. In total, 3508 very low birth weight infants born between January 1, 2013 and December 31, 2014 were reviewed. RESULTS: The adjusted hazard ratio for death increased significantly for infants who received mechanical ventilation for more than 2 weeks compared with those who were mechanically ventilated for 7 days or less. The individual mortality rate increased after 8 weeks, reaching 50% and 60% at 14 and 16 weeks of cumulative mechanical ventilation,
respectively. After adjusting for potential confounders, the cumulative duration of mechanical ventilation was associated with a clinically significant increase in the odds of bronchopulmonary dysplasia and pulmonary hypertension. Mechanical ventilation exposure for longer than 2 weeks, compared with 7 days or less, was associated with retinopathy of prematurity requiring laser coagulation and periventricular leukomalacia. The odds of abnormal auditory screening test results were significantly increased in infants who needed mechanical ventilation for more than 4 weeks. A longer cumulative duration of mechanical ventilation was associated with increased lengths of hospitalization and parenteral nutrition and a higher probability of discharge with poor achievement of physical growth. CONCLUSIONS: Although mechanical ventilation is a life-saving intervention for premature infants, these results indicate that it is associated with negative consequences when applied for prolonged periods.


BackgroundNon-accidental head injury (NAI) is an inflicted injury usually on a child, often resulting in long-term neurological impairment and occasionally death. This study aimed to investigate the predictive values of acute findings, especially ocular, for long-term neurological outcomes.MethodsMedical records including retinal images of all children who attended the local Children's hospital with a diagnosis of NAI from over a period of 5 years were reviewed and data collected via the electronic patient record system. Patient demographics, injuries sustained, wide-field digital retinal images, visual acuity and sequelae, neurological function, and global function was noted. IBM SPSS software program was used for statistical analysis. ResultsOf the 38 patients (24 males, 14 females), 12 children died acutely from the head injury with the remaining 26 children available for long-term follow-up. A younger age of injury (P=0.004) was the only statistically significant predictor of good neurological outcome as compared with absence of macular retinoschisis, unilateral retinal haemorrhage, and unilateral subdural haemorrhage. Of the 38 children, 17 children had retinoschisis; 9 children with macular retinoschisis died acutely while 4 suffered a degree of developmental delay and only 4 were developmentally normal at the last follow-up. Long-term visual acuity data was available for 18 of the 26 survivors (range: NPL to Snellen 6/5). A statistical significance was noted between retinoschisis and worsened visual acuity (P<0.05). ConclusionsBilateral macular retinoschisis on acute presentation of NAI is associated with a seven-fold and unilateral with a four-fold increase in the development of a poor neurological outcome and eventual death. Conflicting to other studies, younger children presented better neurological outcomes.


BACKGROUND: Around the world, different models of paediatric palliative care have responded to the unique needs of children with life shortening conditions. However, research confirming their utility and impact is still lacking. This study compared patient-related outcomes and healthcare expenditures between those who received home-based paediatric palliative care and standard care. The quality of life and caregiver burden for patients receiving home-based paediatric palliative care were also tracked over the first year of enrolment to evaluate the service's longitudinal impact. METHOD: A structured impact and cost evaluation of Singapore-based HCA Hospice Care's Star PALS (Paediatric Advance Life Support) programme was conducted over a three-year period, employing both retrospective and prospective designs with two patient groups. RESULTS: Compared to the control group (n = 67), patients receiving home-based paediatric palliative care (n = 71) spent more time at home than in hospital in the last year of life by 52 days (OR = 5.230, 95% CI: 25.44-79.17) with at least two fewer hospital admissions (OR = 2.46, 95% CI: 0.43-4.48); and were five times more likely to have an advance care plan formulated (OR = 5.51, 95% CI: 1.55-19.67). Medical costs incurred by this group were also considerably lower (by up to 87%). Moreover, both patients' quality of life (in terms of pain and emotion), and caregiver burden showed improvement within the first year of enrolment into the programme. DISCUSSION: Our findings suggest that home-based paediatric palliative care brings improved resource utilization and cost-savings for both patients and healthcare providers. More importantly, the lives of patients and their caregivers have improved, with terminally ill children and their caregivers being able to spend more quality time at home at the final stretch of the disease. CONCLUSIONS: The benefits of a community paediatric palliative care programme have been validated. Study findings can become key drivers when engaging service commissioners or even policy makers in appropriate settings.
OBJECTIVES: To evaluate functional outcomes and evaluate predictors of an unfavorable functional outcome in children following a critical illness. DESIGN: Prospective observational longitudinal cohort study. SETTING: Two tertiary care, Canadian PICUs: McMaster Children's Hospital and London Health Sciences. PATIENTS: Children 12 months to 17 years old, admitted to PICU for at least 48 hours with one or more organ dysfunction, were eligible. Patients not expected to survive, direct transfers from neonatal ICU and patients in whom long-term follow-up would not be able to be conducted, were excluded. INTERVENTIONS: None. MEASUREMENTS AND MAIN RESULTS: The primary endpoint was functional outcome up to 6 months post PICU discharge, measured using the Pediatric Evaluation of Disabilities Inventory Computer Adaptive Test. Secondary outcomes included predictors of unfavorable functional outcome, caregiver stress, health-related quality-of-life, and clinical outcomes such as mortality, length of stay, and PICU-acquired complications. One hundred eighty-two patients were enrolled; 78 children (43.6%) had functional limitations at baseline and 143 (81.5%) experienced functional deterioration following critical illness. Ninety-two (67.1%) demonstrated some functional recovery by 6 months. Higher baseline function and a neurologic insult at PICU admission were the most significant predictors of functional deterioration. Higher baseline function and increasing age were associated with slower functional recovery. Different factors affect the domains of functioning differently. Preexisting comorbidities and iatrogenic PICU-acquired morbidities were associated with persistent requirement for caregiver support (responsibility function) at 6 months. The degree of functional deterioration after critical illness was a significant predictor of increased hospital length of stay. CONCLUSIONS: This study provides new information regarding functional outcomes and the factors that influence meaningful aspects of functioning in critically ill children. Identifying patients at greatest risk and modifiable targets for improvement in PICU care guides us in developing strategies to improve functional outcomes and tailor to the rehabilitation needs of these patients and their families.


OBJECTIVE In the past, the outcome of surgical treatment for thalamic tumor was poor. These lesions were often considered inoperable. However, contemporary microsurgical techniques, together with improvements in neuroimaging that enable accurate presurgical planning, allow resection to be accomplished in a safer way. METHODS The medical records, imaging studies, and operative and pathology reports obtained for pediatric patients who were treated for thalamic tumors at the authors’ department were reviewed. Neuronavigation and intraoperative monitoring of motor and somatosensory evoked potentials were used. Preoperative tractography, which helped to identify internal capsule fibers, was very important in selecting the surgical strategy. Postoperatively, an MRI study performed within 24 hours was used to assess the extent of tumor resection as partial (<\(\leq\) 90%), subtotal (> 90%), or gross total (no residual tumor). RESULTS Since 2002, 27 children with thalamic tumors have been treated at the authors' department. There were 9 patients with unilateral thalamic tumors, 16 with thalamopeduncular tumors, and 2 with a bilateral tumor. These last 2 patients underwent nine tumor debulking procedures in the remaining 25 patients. Different surgical approaches were chosen according to tumor location and displacement of the posterior limb of the internal capsule (as studied on axial T2-weighted MRI) and corticospinal tract (as studied on diffusion tensor imaging with tractography, after it became available). In 12 cases, multiple procedures were performed; in 7 cases, these were done as part of a planned multistage resection. In the remaining 5 cases, the second procedure was necessary because of late recurrence or regrowth of residual tumor. At the end of the surgical phase, of 25 patients, 15 (60%) achieved a gross-total resection, 4 (16%) achieved a subtotal resection, and 6 (24%) achieved a partial resection. Eighteen patients harbored low-grade tumors in our series. In this group, the mean follow-up was 45 months (range 4-132 months). At the end of follow-up, 1 patient was dead, 12 patients were alive with no evidence of disease, 4 patients were alive with stable disease, and 1 was lost to follow-up. All patients were independent in their daily lives. The outcome of high-grade tumors in 9 patients was very poor: 2 patients died immediately after surgery, 6 died of progressive disease, and 1

was alive with residual disease at the time of this report. CONCLUSIONS This institutional review seems to offer further evidence in favor of attempts at radical resection in pediatric patients harboring unilateral thalamic or thalamopeduncular tumors. In low-grade gliomas, radical resection in a single or staged procedure can be curative without complementary treatment. Recurrences or residual regrowth can be safely managed surgically. In high-grade tumors, the role of and opportunity for radical or partial resection remains a matter of debate. https://www.ncbi.nlm.nih.gov/pubmed/29271729


OBJECTIVE: Psychometric properties of the Fear of Progression Questionnaire - Short Form (FoP-Q-SF) were shown to be good in samples of adult cancer patients and their partners but have so far not been investigated in parents of children with cancer. This study therefore aimed to examine psychometric properties of the previously adapted parent version of the Fear of Progression Questionnaire (FoP-Q-SF/PR) in pediatric oncology. METHODS: N=181 parents (119 mothers, 62 fathers) of n=128 children with diverse cancer entities, up to ten years after diagnosis were recruited at six hospitals and six registered parent associations in Germany and Austria between 06/2015 and 05/2016 (cross-sectional design). Parents provided medical information about their child and completed standardized questionnaires (Hospital Anxiety and Depression Scale, HADS; State-Trait Anxiety Inventory, STAI; Impact of Event Scale-Revised, IES-R; Ulm Quality of Life Inventory for Parents, ULQIE; Giessen Physical Complaints Inventory for children and adolescents, GBB-KJ). RESULTS: Exploratory factor analysis yielded two factors (50.2% explained variance) and internal consistency was good (Cronbach’s alpha=0.89). Significant medium to large correlations of the FoP-Q-SF/PR were observed with anxiety (HADS: r=0.68; STAI: r=0.60-0.61), depression (HADS: r=0.58), posttraumatic stress (IES-R: r=0.42-0.64) and quality of life (ULQIE: r=-0.59). The FoP-Q-SF/PR discriminated between sub-groups, e.g. parents with and without clinical anxiety levels (Cohen’s d=1.26). CONCLUSION: The FoP-Q-SF/PR demonstrated good reliability and validity for parents of children with cancer. The FoP-Q-SF/PR is a feasible screening instrument, which is suitable for the assessment of parental FoP in pediatric oncology. https://www.ncbi.nlm.nih.gov/pubmed/29502766


BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization (WHO) guidelines for pharmacological treatments for children’s persisting pain acknowledge that pain in children is a major public health concern of high significance in most parts of the world. While in the past, pain was largely dismissed and was frequently left untreated, views on children’s pain have changed over time, and relief of pain is now seen as important. We designed a suite of seven reviews on chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol) in order to review the evidence for children’s pain utilising pharmacological interventions in children and adolescents. As the leading cause of morbidity in the world today, chronic disease (and its associated pain) is a major health concern. Chronic pain (that is pain lasting three months or longer) can occur in the paediatric population in a variety of pathophysiological classifications (nociceptive, neuropathic, or idiopathic) relating to genetic conditions, nerve damage pain, chronic musculoskeletal pain, and chronic abdominal pain, and for other unknown reasons. Antiepileptic (anticonvulsant) drugs, which were originally developed to treat convulsions in people with epilepsy, have in recent years been used to provide pain relief in adults for many chronic painful conditions and are now recommended for the treatment of chronic pain in the WHO list of essential medicines. Known side effects of antiepileptic drugs range from sweating, headache, elevated temperature, nausea, and abdominal pain to more serious effects including mental or motor function impairment. OBJECTIVES: To assess the analgesic efficacy and adverse events of antiepileptic drugs used to treat chronic non-cancer pain in children and adolescents aged between birth and 17 years, in any setting. SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online, MEDLINE via Ovid, and Embase via Ovid from inception to 6 September 2016. We also searched the reference lists of retrieved studies and reviews as well as online clinical trial registries. SELECTION CRITERIA: Randomised controlled trials, with or without blinding, by any route, treating chronic non-cancer pain in children and adolescents, comparing any antiepileptic drug with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for
On NRSTS consisting of doxorubicin and surgical removal of the tumor performed within 3 months after diagnosis. Two of them were mistaken for nonsecreting hepatoblastomas at diagnosis and had recurrence shortly after initial diagnosis, followed by surgical resection. Three patients (50%) died of disease. Two of them were mistaken for nonsecreting hepatoblastomas at diagnosis and had recurrence shortly after completion of treatment. The third one presented a cardiac right atrium thrombus. Three patients (50%) are long-term survivors; they received multimodal therapy including chemotherapy according to protocol EppSG NRSTS consisting of doxorubicin and surgical removal of the tumor performed within 3 months after diagnosis. One patient had adjuvant radiotherapy. CONCLUSION: According to our results, search of SMARCB1 mutation or


BACKGROUND: Rhabdoid tumors (RTs) of the liver are rare, aggressive and nonsecreting malignancies occurring mainly during the first year of life. Definition of RT relies on characteristic morphology and on the inactivation of the SMARCB1 tumor suppressor gene. The aim of this study was to analyze clinical data, treatments and outcomes in our patients. PATIENTS AND METHODS: 6 cases of patients treated in our institution for RT of the liver between January 2007 and January 2015 are reported. Variables examined included age at diagnosis, tumor stage, treatment and long-term survival. RESULTS: Median age at diagnosis was 5 months (range: 4-23). Normal for age serum AFP levels was observed in all patients. No patient presented with metastasis at diagnosis. The diagnosis of RT based on the loss of SMARCB1 was made early in 4 patients. The other two patients were initially diagnosed as nonsecreting hepatoblastomas. Median follow-up was 6 years (range: 2-9). All patients received chemotherapy, with variable regimens depending on initial diagnosis, followed by surgical resection. Three patients (50%) died of disease. Two of them were mistaken for nonsecreting hepatoblastomas at diagnosis and had recurrence shortly after completion of treatment. The third one presented a cardiac right atrium thrombus. Three patients (50%) are long-term survivors; they received multimodal therapy including chemotherapy according to protocol EppSG NRSTS consisting of doxorubicin and surgical removal of the tumor performed within 3 months after diagnosis. One patient had adjuvant radiotherapy. CONCLUSION: According to our results, search of SMARCB1 mutation or

alternative immunohistochemical assay for SMARCB1 in nonsecreting hepatoblastomas is mandatory to exclude RT. Chemotherapy according to EpSSG NRSTS protocol together with a surgical treatment seems justified to improve long-term survival. TYPE OF STUDY: Retrospective study. LEVEL OF EVIDENCE: Level IV. https://www.ncbi.nlm.nih.gov/pubmed/28966010


CONTEXT: This review summarizes the current randomized controlled trials literature on psychological and physical outcomes of psychosocial interventions in pediatric oncology. OBJECTIVES: The objective of this study was to evaluate the effectiveness and impact of psychosocial interventions in children with cancer. METHODS: A search of the literature resulted in a total of 12 randomized clinical trials and these have evaluated psychosocial interventions in children younger than 18 years with current and previous diagnoses of cancer. Outcome measures were both psychological (e.g., symptoms of anxiety, depression, quality of life, and self-esteem) and physical (e.g., cancer symptoms, treatment adherence, and pain). Interventions identified included cognitive behavioral therapy (CBT; n = 4), joint CBT and physical exercise therapy (n = 1), family therapy (n = 2), therapeutic music video (n = 2), self-coping strategies (n = 1), a wish fulfillment intervention (n = 1), and joint family therapy and CBT (n = 1). RESULTS: Nine studies reported statistically significant improvements on psychological outcomes. These findings suggest that psychosocial interventions are effective at reducing anxiety and depressive symptoms as well as improving quality of life. Additionally, six studies found psychosocial interventions to have a positive impact on physical symptoms and well-being, including a reduction in procedural pain and symptom distress. CONCLUSION: These findings suggest that mental health needs in pediatric oncology patients can and should be addressed, potentially which will lead to better mental and physical health outcomes. https://www.ncbi.nlm.nih.gov/pubmed/28962919


OBJECTIVE: Allogeneic stem-cell transplant (allo-SCT) is the standard of care for pediatric patients with acute lymphoblastic leukemia (ALL) who relapse after frontline chemotherapy; however, for patients who relapse after allo-SCT, outcomes are very poor. Few studies have examined overall survival in this patient population, particularly in patients who received a second allo-SCT. METHODS: This was a retrospective analysis using data from the Center for International Blood and Marrow Transplant Research (CIBMTR) registry. The study population included patients aged 3 to 21 years who were diagnosed with B-ALL and underwent their first allo-SCT between 2009 and 2013. The primary endpoint was the time from the date of posttransplant relapse to the date of death due to any reason. RESULTS: Outcomes in 1349 pediatric and young-adult patients were included in this analysis. The Kaplan-Meier estimated probability of survival at 3 years after first allo-SCT was 63.1% (95% CI, 60.2%-65.8%). Overall, 29.2% of patients relapsed after first allo-SCT and had a median survival of 7.4 months (95% CI, 6.0-9.6 months). Twenty-five patients in the analysis developed secondary malignancies, most of which were lymphoproliferative disorders. CONCLUSIONS: Survival rates are low in pediatric and young-adult patients who relapse after first and second allo-SCT, and new therapies are needed to improve outcomes in this population. This data can be used as a historical comparison for single-arm trials of novel therapies for this patient population, including chimeric antigen receptor T-cell therapy. https://www.ncbi.nlm.nih.gov/pubmed/28945102


OBJECTIVE: We designed a prospective, randomized, controlled, double-blind study to evaluate the efficacy of hippocampal deep brain stimulation (Hip-DBS) in patients with refractory temporary lobe epilepsy (TLE). METHODS: Sixteen adult patients with refractory TLE were studied. Patient's workup included medical history, interictal and ictal electroencephalography (EEG), and high-resolution 1.5T magnetic resonance imaging (MRI). Patients were randomized on a 1:1 proportion to an active (stimulation on) or to a control (no stimulation) arm. After implantation, patients were allowed to recover for 1 month, which was followed by a 1-month titration (or sham) period. The 6-month blinded phase started immediately afterward. A postoperative MRI confirmed the
electrode’s position in all patients. All patients received bipolar continuous stimulation. Stimulus duration was 300 ms and frequency was 130 Hz; final intensity was 2 V. Patients were considered responders when they had at least 50% seizure frequency reduction. RESULTS: All patients had focal impaired awareness seizures (FIAS, complex partial seizures), and 87% had focal aware seizures (FAS, simple partial seizures). Mean preoperative seizure frequency was 12.5 +/- 9.4 (mean +/- standard deviation) per month. MRI findings were normal in two patients, disclosed bilateral mesial temporal sclerosis (MTS) in three, left MTS in five, and right MTS in six patients. An insertional effect could be noted in both control and active patients. In the active group (n = 8), four patients became seizure-free; seven of eight were considered responders and one was a nonresponder. There was a significant difference regarding FIAS frequency between the two groups from the first month of full stimulation (p < 0.001) until the end of the blinded phase (p < 0.001). This was also true for FAS, except for the third month of the blinded phase. SIGNIFICANCE: Hip-DBS was effective in significantly reducing seizure frequency in patients with refractory TLE in the active group, as compared to the control group. Fifty-percent of the patients in the active group became seizure-free. The present study is the larger prospective, controlled, double-blind study to evaluate the effects of Hip-DBS published to date.


Pain behaviors are important indicators of functioning in chronic pain; however, no self-reported pain behavior instrument has been developed for pediatric populations. The purpose of this study was to create a brief pediatric measure of patient-reported pain behaviors as part of the Patient-Reported Outcome Measurement Information System (PROMIS). A pool of 47 candidate items for this measure had been previously developed through qualitative research. In this study, youth with chronic pain associated with juvenile fibromyalgia, juvenile idiopathic arthritis, or sickle cell disease (ages 8-18 years) from 3 pediatric centers completed all 47 candidate items for development of the pain behavior item bank along with established measures of pain interference, depressive symptoms, fatigue, average pain intensity, and pain catastrophizing. Caregivers reported on sociodemographic information and health history. Psychometric properties of the pain behavior items were examined using an item response theory framework with confirmatory factor analysis and examination of differential item functioning, internal consistency, and test information curves. Results were used along with expert consensus and alignment with the adult PROMIS pain behavior items to arrive at an 8-item pediatric pain behavior short form, and all 47 items were retained in a calibrated item bank. Confirmatory factor analysis and correlations with validated measures of pain, pain interference, and psychosocial functioning provided support for the short form’s reliability and validity. The new PROMIS pediatric pain behavior scale provides a reliable, precise, and valid measure for future research on pain behavior in school-aged children with chronic pain.


PURPOSE: The purposes of this study are to describe sleep quality and sleep disturbance among caregivers of children in the maintenance phase of acute lymphoblastic leukemia (ALL) and to examine the relationship between sleep quality, child sleep disturbance, and caregiver guilt and worry. METHODS: Caregivers of 68 children with ALL aged 3 to 12 years old, completed measures of caregiver guilt and worry, caregiver sleep quality, and child’s developmental history and sleep habits. Demographic and treatment correlates of poor caregiver sleep were examined, and caregiver guilt and worry was tested as a moderator between child and caregiver sleep. RESULTS: More than half of caregivers (55.9%) reported clinically significant poor sleep and less than 40% were obtaining adequate sleep durations. Caregiver sleep was significantly related to child age at diagnosis, child sleep, and caregiver guilt and worry. Caregiver guilt and worry did not moderate the relationship between child sleep and caregiver sleep. CONCLUSIONS: Poor sleep is common in caregivers of children with cancer. Further research on the timing of sleep interventions and the most effective intervention targets are needed to maximize caregiver functioning during a child’s cancer treatment. Targeted interventions seeking to improve caregiver sleep should be directed towards caregivers of children diagnosed in early childhood, caregivers of children with poor sleep, and caregivers with high guilt and worry.
https://www.ncbi.nlm.nih.gov/m/pubmed/29046955
Juvenile dermatomyositis (JDM) is a rare autoimmune disease that may lead to serious complications, even to death. We develop a 2-state Markov regression model in a Bayesian framework to characterise disease progression in JDM over time and gain a better understanding of the factors influencing disease risk. The transition probabilities between disease and remission state (and vice versa) are a function of time-homogeneous and time-varying covariates. These latter types of covariates are introduced in the model through a latent health state function, which describes patient-specific health over time and accounts for variability among patients. We assume a nonparametric prior based on the Dirichlet process to model the health state function and the baseline transition intensities between disease and remission state and vice versa. The Dirichlet process induces a clustering of the patients in homogeneous risk groups. To highlight clinical variables that most affect the transition probabilities, we perform variable selection using spike and slab prior distributions. Posterior inference is performed through Markov chain Monte Carlo methods. Data were made available from the UK JDM Cohort and Biomarker Study and Repository, hosted at the UCL Institute of Child Health. [https://www.ncbi.nlm.nih.gov/pubmed/29462840](https://www.ncbi.nlm.nih.gov/pubmed/29462840)

**OBJECTIVE:** To characterize patterns of care at the end of life for children and young adults with life-threatening complex chronic conditions (LT-CCCs) and to compare them by LT-CCC type. **STUDY DESIGN:** Cross-sectional survey of bereaved parents (n = 114; response rate of 54%) of children with noncancer, noncardiac LT-CCCs who received care at a quaternary care children’s hospital and medical record abstraction. **RESULTS:** The majority of children with LT-CCCs died in the hospital (62.7%) with more than one-half (53.3%) dying in the intensive care unit. Those with static encephalopathy (AOR, 0.19; 95% CI, 0.04-0.98), congenital and chromosomal disorders (AOR, 0.28; 95% CI, 0.09-0.91), and pulmonary disorders (AOR, 0.08; 95% CI, 0.01-0.77) were significantly less likely to die at home compared with those with progressive central nervous system (CNS) disorders. Almost 50% of patients died after withdrawal or withholding of life-sustaining therapies, 17.5% died during active resuscitation, and 36% died while receiving comfort care only. The mode of death varied widely across LT-CCCs, with no patients with pulmonary disorders dying receiving comfort care only compared with 66.7% of those with CNS progressive disorders. A majority of patients had palliative care involvement (79.3%); however, in multivariable analyses, there was distinct variation in receipt of palliative care across LT-CCCs, with patients having CNS static encephalopathy (AOR, 0.07; 95% CI, 0.01-0.68) and pulmonary disorders (AOR, 0.07; 95% CI, 0.01-0.09) significantly less likely to have palliative care involvement than those with CNS progressive disorders. **CONCLUSIONS:** Significant differences in patterns of care at the end of life exist depending on LT-CCC type. Attention to these patterns is important to ensure equal access to palliative care and targeted improvements in end-of-life care for these populations. [https://www.ncbi.nlm.nih.gov/pubmed/29174080](https://www.ncbi.nlm.nih.gov/pubmed/29174080)

Mild Zellweger spectrum disorder, also described as Infantile Refsum disease, is attributable to mutations in PEX genes. Its clinical course is characterized by progressive hearing and vision loss, and neurodevelopmental regression. Supportive management is currently considered the standard of care, as no treatment has shown clinical benefits. LT was shown to correct levels of circulating toxic metabolites, partly responsible for chronic neurological impairment. Of three patients having undergone LT for mild ZSD, one died after LT, while the other two displayed significant neurodevelopmental improvement on both the long-term (17 years post-LT) and short-term (9 months post-LT) follow-up. We documented a sustained improvement of biochemical functions, with a complete normalization of plasma phytanic, pristanic, and pipecolic acid levels. This was associated with stabilization of hearing and visual functions, and improved neurodevelopmental status, which has enabled the older patient to lead a relatively autonomous lifestyle on the long term. The psychomotor acquisitions have been markedly improved as compared to their affected siblings, who did not undergo LT and exhibited a poor neurological outcome with severe disabilities. We speculate that LT performed before the onset of severe sensorineural defects in mild ZSD enables partial metabolic remission and improved long-term clinical outcomes.
INTRODUCTION: Pediatric acute liver failure (ALF) due to inherited metabolic diseases (IMD) is a rare life-threatening condition with a poor prognosis. Early intervention may be lifesaving. OBJECTIVE: To describe clinical presentation, investigation and outcomes of ALF related to IMD in young children. MATERIAL AND METHODS: Retrospective review of the medical records of children aged up to 24 months, admitted to a tertiary pediatric and neonatal Intensive Care Unit during a 27-year period, fulfilling the ALF criteria, with documented metabolic etiology. RESULTS: From 34 ALF cases, 18 were related to IMD: galactosemia (4), mitochondrial DNA depletion syndrome (MDS) (3), ornithine transcarbamilase deficiency (3), congenital defects of glycosylation (2), tyrosinemia type 1 (2), long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency (1), hereditary fructose intolerance (1), classic methylmalonic aciduria (1) and citrulinaemia type 1 (1). The median age was 1.3 months. At least one previous suggestive sign/symptom of IMD (vomiting, failure to thrive, hypotonia or developmental delay) was observed in 67% of the cases. The most common physical signs at admission included: hepatomegaly (72%), jaundice (67%) and encephalopathy (44%). The peak laboratorial findings were: mean international normalized ratio 4.5, median lactate 5mmol/L, mean bilirubin 201umol/L, median alanine aminotransferase (ALT) 137 UI/L and median ammonia 177umol/L. One patient was submitted to liver transplant in ALF context (MSD). The mortality rate was 44%. DISCUSSION: The identification of IMD as a frequent cause of ALF allowed specific therapeutic measures and adequate family counselling. Particular clinical features and moderated ALT and bilirubin levels can lead to its suspicion.


OBJECTIVE: To evaluate the reliability and validity of the Brazilian version of the Echelle Douleur Inconfort Nouveau-Ne, which measures prolonged pain in neonates. METHOD: A methodological study carried out with 96 neonates. The Brazilian versions of the Echelle Douleur Inconfort Nouveau-Ne and the Children's and Infants' Postoperative Pain Scale were used for data collection. For reliability, equivalence measured by intraobserver agreement and homogeneity were considered. To evaluate the validity, the convergent construct approach was considered correlating the Brazilian versions of the Echelle Douleur Inconfort Nouveau-Ne and the Children's and Infants' Postoperative Pain Scale. RESULTS: In assessing reliability, the coefficient of agreement between observers varied between 0.64 and 0.85 for the items that make up the instrument, and 0.96 for the total score. Cronbach's alpha was 0.82. Regarding the convergent validity evaluation, Spearman correlation coefficient between the values found for both scales was 0.79 (p< 0.0001). CONCLUSION: The Brazilian version of the Echelle Douleur Inconfort Nouveau-Ne is a reliable and valid instrument for assessing prolonged pain in neonates.

allograft is important with regards to impact on outcomes. Retransplantation is rare in pediatric heart transplant recipients. Pediatric heart transplantation continues to evolve in order to address the challenges of the diverse group of patients that reach end-stage heart failure during childhood. [https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5827130/]


CONTEXT: Pediatric palliative care has no evidence-based needs assessment measure. The Parent and Child Needs Survey (PCNeeds) is a new instrument designed to assess the needs of children in palliative care, including children receiving end-of-life care, and their families. OBJECTIVES: This study examines the psychometrics of and respondents’ perceptions about the PCNeeds. METHODS: Parents of children in four outpatient pediatric palliative care programs completed the PCNeeds and the World Health Organization Quality of Life-Brief tool (WHOQOL-BREF). Parents answered questions about demographics and the experience of completing the PCNeeds. Internal scale reliability was measured with Cronbach’s alpha. Validity was assessed by correlating the PCNeeds total and subscale scores with the WHOQOL-BREF subscales. Additional respondent perceptions were obtained via written comments and analyzed using content analysis. RESULTS: The 93 respondents were predominantly female (n = 69, 74%); white (n = 79, 85%); college graduates (n = 71, 76%); and married or partnered (n = 75, 81%). Internal reliability was acceptable (Cronbach’s alpha = 0.83), and validity correlations with the WHOQOL-BREF subscales were consistent with theoretical expectations (moderate negative correlations ranging from −0.36 to −0.51). The most frequently cited need not addressed by our survey was sibling impact (n = 17, 18%). Twelve parents (13%) indicated that no content was missing. The least met needs were financial impact, family impact, and the child’s physical problems besides pain. Sixty-eight percent of parents (n = 63) rated completion of the survey as “easy” or “very easy.” CONCLUSION: Initial psychometric analysis of the PCNeeds is encouraging, but further study of reliability and validity with more diverse respondents is needed. [https://www.ncbi.nlm.nih.gov/pubmed/29288879]


Sleep problems are a common and serious issue in children with life-limiting conditions (LLCs) and severe psychomotor impairment (SPMI). The "Sleep Questionnaire for Children with Severe Psychomotor Impairment" (Schlaffragebogen fur Kinder mit Neurologischen und Anderen Komplexen Erkrankungen, SNAKE) was developed for this unique patient group. In a proxy rating, the SNAKE assesses five different dimensions of sleep (associated) problems (disturbances going to sleep, disturbances remaining asleep, arousal and breathing disorders, daytime sleepiness, and daytime behavior disorders). It has been tested with respect to construct validity and some aspects of criterion validity. The present study examined whether the five SNAKE scales are consistent with parents’ or other caregivers’ global ratings of a child’s sleep quality. Data from a comprehensive dataset of children and adolescents with LLCs and SPMI were analyzed through correlation coefficients and Mann-Whitney U testing. The results confirmed the consistency of both sources of information. The highest levels of agreements with the global rating were achieved for disturbances in terms of going to sleep and disturbances with respect to remaining asleep. The results demonstrate that the scales and therefore the SNAKE itself is well-suited for gathering information on different sleep (associated) problems in this vulnerable population. [https://www.ncbi.nlm.nih.gov/pubmed/29389907]


BACKGROUND: In palliative care contexts, support programs for families with a severely ill parent and minor children are few, and even fewer have been evaluated scientifically. The aims of this study are to examine feasibility and potential effects of a modified version of the Family Talk Intervention (FTI) in palliative care. METHODS: This ongoing family-centered intervention has a quasi-experimental design comparing one intervention and one comparison group. The intervention includes severely ill parents who have minor children (aged 6-19 yrs) and are receiving advanced homecare in Stockholm, Sweden between March 2017 and March 2018. The main goal of the FTI is to support family communication through psycho-education and narrative therapy. The modified FTI consists of six meetings with family members, and is held by two interventionists. Each
family sets up needs-based goals for the intervention. For evaluation purposes, data are collected by questionnaire before the intervention, within two months after baseline, and one year after baseline. Interviews will be conducted within two months after FTI is completed. Notes taken by one of the interventionists during the family meetings will also be used. Questionnaire data analysis will focus on patterns over time using descriptive statistics. For interview data and notes, content analysis will be used. DISCUSSION: This study will add knowledge about palliative care for parents who have minor children. It will contribute by testing use of FTI in palliative care, and point out directions for future evaluations of FTI in palliative care settings. TRIAL REGISTRATION: ClinicalTrials.gov Identifier NCT03119545, retrospectively registered in April 18, 2017. https://www.ncbi.nlm.nih.gov/pubmed/29154891


AIMS: The risk of ischaemic heart disease (IHD) death in early type 1 diabetes onset was assessed using death certification data. METHODS: The Yorkshire Register of type 1 Diabetes in Children and Young People was linked to clinically validated death certification data for those diagnosed under 15 years. Standardised mortality ratios (SMRs) were calculated using the England and Wales population and IHD death rates between 1978 and 2014 by 5-year age group and sex. RESULTS: The cohort included 4382 individuals (83 euro per thousand 097 person years). Of 156 deaths, nine were classed as IHD deaths before clinical validation. After clinical validation, 14 IHD deaths were classified, with an SMR of 13.8 (95% CI 8.2 to 23.3) and median age at death of 35.1 years (range 21.9 euro 47.9 years). CONCLUSIONS: There is an early emergence of death from IHD in early onset type 1 diabetes. Underascertainment of IHD deaths was present without clinical validation of death certification. https://www.ncbi.nlm.nih.gov/pubmed/29367262


CONTEXT: Reliable identification of lower respiratory tract pathogens is crucial in the management of cystic fibrosis (CF). The multitude of treatments and clinical procedures are a considerable burden and are potentially provoking pain. OBJECTIVES: As part of another study (NCT02363764), investigating the bacterial yield of three sampling methods, nasal swabs (NSs), cough swabs (CSs), and (induced) sputum samples ([S]Ss), in both expectorating patients (EPs) and non-expectorating patients (NEPs) with CF, the present study aimed to explore the prevalence of respiratory culture sampling-related pain as assessed by self-report within a cohort of children and adults. METHODS: Literate patients with CF (aged six years or older) completed a questionnaire on pain perception related to the three aforementioned sampling methods (No/Yes; visual analogue scale for pain [VAS-Pain] [0-10 cm]). In addition, patients were asked to rank these methods by their own preference without taking into account the presumed bacterial yield. RESULTS: In total, 119 questionnaires were returned. In the EPs-group, CS was most frequently (n%; mean VAS-Pain if pain [range]) reported as painful method: overall (n = 101; 12.9%; 1.8 [0.2-4.8]), children (n = 41; 22.0%; 1.4 [0.2-2.7]), and adults (n = 60; 6.7%; 2.5 [0.5-4.8]). Highest pain intensity scores were observed with NS overall (3.0%; 2.4 [0.3-6.2]) and in children (4.9%; 3.3 [0.3-6.2]), but not in adults (1.7%; 0.6 [-]). NEPs-children (n = 17) reported ISS most frequently and as most painful sampling method (17.6%; 2.0 [1.0-4.0]). The only NEP-adult did not perceive pain. NEPs preferred NS > CS > ISS (61.1%, 33.3%, 5.6%, respectively [P = 0.001]) as primary sampling method, whereas EPs preferred SS > NS > CS (65.7%, 26.3%, 8.1%, respectively [P < 0.0001]). Patients' preference for a specific method inversely correlated to pain perception and intensity in EPs (phi = -0.155 [P = 0.007] and rho = -0.626 [P = 0.001]) and in children (phi = -0.226 [P = 0.097] and rho = -0.135 [P = 0.798], respectively). CONCLUSION: A relatively large range of pain experiences was observed in patients with CF during respiratory culture sampling, which underlines the importance of individual pain assessment. Nevertheless, clinicians can confidently choose the sampling method based on validity over patients' preference. https://www.ncbi.nlm.nih.gov/pubmed/29154891


OBJECTIVE: To describe the patient demographics and outcome analysis in paediatric non-Hodgkin lymphoma (NHL) patients. STUDY DESIGN: An observational study. PLACE AND DURATION OF STUDY: The Hematology/Oncology Unit of The Children's Hospital and Institute of Child Health, Lahore, from January 2012 till
the nusinersen group than in the control group had a motor result prompting early termination of the trial. In the final analysis, a significantly higher percentage of infants in the nusinersen group than in the control group had a motor milestone response (37 of 73 infants [51%] vs. 0 of 1). This result prompted early termination of the trial. In the final analysis, a significantly higher percentage of infants in the nusinersen group than in the control group had a motor-milestone response (37 of 73 infants [51%] vs. 0 of 1).
37 [0%]), and the likelihood of event-free survival was higher in the nusinersen group than in the control group (hazard ratio for death or the use of permanent assisted ventilation, 0.53; P=0.005). The likelihood of overall survival was higher in the nusinersen group than in the control group (hazard ratio for death, 0.37; P=0.004), and infants with a shorter disease duration at screening were more likely than those with a longer disease duration to benefit from nusinersen. The incidence and severity of adverse events were similar in the two groups.

CONCLUSIONS: Among infants with spinal muscular atrophy, those who received nusinersen were more likely to be alive and have improvements in motor function than those in the control group. Early treatment may be necessary to maximize the benefit of the drug. (Funded by Biogen and Ionis Pharmaceuticals; ENDEAR ClinicalTrials.gov number, NCT02193074 ).


BACKGROUND: Critically ill pediatric patients with endotracheal tubes routinely receive endotracheal tube suctioning to clear secretions and ensure tube patency. This common practice can result in adverse events. OBJECTIVES: The aim of this study was to evaluate the research literature on the stressors of endotracheal suctioning and consequent effect on the pediatric patient. METHODS: An integrative review was conducted using the Whittomore and Knaff modified framework for integrative reviews, and article selection was guided by the Preferred Reporting Items for Systematic Reviews and Meta-analyses flow diagram. A literature search was conducted via PubMed, the Cumulative Index to Nursing and Allied Health Literature, and Scopus. Selected articles were evaluated to present the current evidence on the stressors of endotracheal suctioning in the pediatric population. RESULTS: This review includes 14 articles, with a total of 849 patients, ranging in age from premature neonates to 17 years of age. The available literature aligned into 3 categories: neurovascular effects, respiratory systems effects, and pain related to endotracheal tube suctioning. Pain was the most prevalent category, with half of the studies using endotracheal suctioning as a painful procedure to validate pain assessment tools rather than examining the effect of suctioning. A majority of the studies (67%) were conducted in the premature neonate population. Children with congenital cardiac or pulmonary defects, genetic syndromes, or neurological injuries were frequently excluded. CONCLUSIONS: Literature regarding the effects of endotracheal suctioning in children is limited. There are many extrapersonal, interpersonal, and intrapersonal stressors associated with endotracheal suctioning that merit future research.


Effective deployment of limited spiritual care resources requires valid and reliable methods of screening that can be used by nonchaplain health care professionals to identify and refer patients with potential religious/spiritual (R/S) need. Research regarding the validity of existing approaches to R/S screening is limited. In a sample of 1,399 hematopoietic stem cell transplant survivors, we tested the validity of the Rush Protocol and two alternative versions of it. The negative religious coping subscale of the Brief RCOPE provided the reference standard. Based on the Protocol, 21.9% of the survivors were identified as having potential R/S struggle. The sensitivity of the Protocol was low (42.1%) and the specificity was marginally acceptable (81.3%). The sensitivity and specificity of the two alternative versions were similar to those for the unmodified Protocol. Further research with the Rush Protocol, and other models, should be pursued to develop the best evidence-based approaches to R/S screening.


BACKGROUND: Death audits have been used to describe pediatric mortality in under-resourced settings, where record keeping is often a challenge. This information provides the cornerstone for the foundation of quality improvement initiatives. Malawi, located in sub-Saharan Africa, currently has an Under-5 mortality rate of 64/1000. Kamuzu Central Hospital, in the capital city Lilongwe, is a busy government referral hospital, which admits up to 3000 children per month. A study published in 2013 reported mortality rates as high as 9%. This is the first known audit of pediatric death files conducted at this hospital. METHODS: A retrospective chart review on all pediatric deaths that occurred at Kamuzu Central Hospital (excluding deaths in the neonatal nursery) during a 13-month period was done using a standardized death audit form. A descriptive analysis was completed,
including patient demographics, HIV and nutritional status, and cause of death. Modifiable factors were identified that may have contributed to mortality, including a lack of vital sign collection, poor documentation, and delays in the procurement or results of tests, studies, and specialist review. RESULTS: Seven hundred forty three total pediatric deaths were recorded and 700 deceased patient files were reviewed. The mortality rate by month ranged from a low of 2.2% to a high of 4.4%. Forty-four percent of deaths occurred within the first 24 h of admission, and 59% occurred within the first 48 h. The most common causes of death were malaria, malnutrition, HIV-related illnesses, and sepsis. CONCLUSIONS: The mortality rate for this pediatric referral center has dramatically decreased in the 6 years since the last published mortality data, but remains high. Areas identified for continued development include improved record keeping, improved patient assessment and monitoring, and more timely and reliable provision of testing and treatment. This study demonstrates that in low-resource settings, where reliable record keeping is often difficult, death audits are useful tools to describe the sickest patient population and determine factors possibly contributing to mortality that may be amenable to quality improvement interventions.


PURPOSE OF REVIEW: Immunotherapy for the treatment of cancer has advanced at a tremendous pace over the last decade. In this review, we provide an overview of recent progress in immunotherapy for the treatment of leukemia, focusing on antibody-drug conjugates (ADC), bi-specific T-cell engagers (BiTE), and chimeric antigen receptor (CAR) T cells. RECENT FINDINGS: Ongoing clinical trials of CAR T cells directed against CD19 have produced complete remission rates as high as 93%, prompting global multicenter phase 2 trials and the first FDA approval of a CAR T-cell therapy. Insights into cytokine release syndrome, a toxicity of CAR T-cell therapy, and the cause for relapse after CAR T-cell therapy are evolving. The bispecific antibody blinatumomab and the ADCs inotuzumab and gemtuzumab have also recently received FDA approval for ALL and AML, respectively, moving these agents into a more prominent role in the relapse setting. SUMMARY: The use of immunotherapy for leukemia has been successful in creating durable remissions for multiply relapsed and refractory patients who previously had little chance of cure. The ongoing clinical and preclinical work continues to advance our understanding of these immune-based therapies, and will shape the next generation of clinical trials.


Mucopolysaccharidoses (MPS) types I, II and VI are associated with deficiencies in alpha-L-iduronidase, iduronate-2-sulfatase and N-acetylglactosamine-4-sulfatase, respectively, and generally involve progressive and multisystemic clinical manifestations. Enzyme replacement therapy (ERT) appears to be reasonably well tolerated. The aim of this study was to examine clinical and diagnostic findings of a series of pediatric and adult MPS patients, and assess the safety and efficacy of ERT in children and adults with MPS type I, II and VI. Pediatric and adult patients were treated weekly with 1 mg/kg recombinant human N-acetylglactosamine-4-sulfatase (rhASB), 0.45 mg/kg alpha-L-iduronidase, or 0.5 mg/kg iduronate-2-sulfatase. Clinical and biochemical parameters with ERT were evaluated for a mean duration of 5 years. Mantel-Haenszel risk ratios and associated 95% confidence intervals (CIs) were calculated for rates of death among different types of enzyme replacement therapies (ERTs). Twenty-seven patients (mean ages pediatric: 6.8 years; adult: 29 years) were included. ERT was found to be consistently well tolerated and effective in attenuating symptoms, but did not prevent the progression of the disease or reduce mortality rates. Our findings demonstrated that early diagnosis and initiation of ERT are critical for improvements in patient-important outcomes and quality of life, although disease progression and mortality rates remain high.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5608928/


This article aims to describe the clinical features of electric powered indoor/outdoor wheelchair (EPIOC) users with cerebral palsy (CP) that are problematic to optimal prescription and to explore comorbidities, features of CP, and
conditions secondary to disability impacting on equipment provision for children and adults. The method is a cross-sectional study of EPIOC users (n = 102) with a primary diagnosis of CP. This is a retrospective review of electronic and case note records of EPIOC recipients attending a specialist wheelchair service in 2007-2008. Records were reviewed by a rehabilitation consultant. Data were extracted under three themes; demographic, diagnostic/clinical and wheelchair factors. There were 48 males mean age 27.5 (range 8-70, SD 13.9) years and 54 females, mean age 29.5 (range 7-68, SD 14.6) years with CP. Sixteen comorbidities, nine features of CP, and five features of disability influenced wheelchair prescription. Sixty-four users were provided with specialized seating (SS) and 47 with tilt-in-space (TIS) seats. Complex controls were provided to 16 users, 12 tray-mounted. The majority of users had both SS and TIS. Powered wheelchair prescription has important therapeutic roles in clinical management in addition to enhancing mobility, independence and participation. Clinical features such as spasticity and problematic pain appeared less well managed in adults than in children.


OBJECTIVE: To determine how many children are admitted to paediatric intensive care unit (PICU) with life-limiting conditions (LLCs) and their outcomes. DESIGN: National cohort, data-linkage study. SETTING: PICUs in England. PATIENTS: Children admitted to a UK PICU (1 January 2004 and 31 March 2015) were identified in the Paediatric Intensive Care Audit Network dataset. Linkage to hospital episodes statistics enabled identification of children with a LLC using an International Classification of Diseases (ICD10) code list. MAIN OUTCOME MEASURES: Random-effects logistic regression was undertaken to assess risk of death in PICU. Flexible parametric survival modelling was used to assess survival in the year after discharge. RESULTS: Overall, 57.6% (n=89 127) of PICU admissions and 72.90% (n=4821) of deaths in PICU were for an individual with a LLC. The crude mortality rate in PICU was 5.4% for those with a LLC and 2.7% of those without a LLC. In the fully adjusted model, children with a LLC were 75% more likely than those without a LLC to die in PICU (OR 1.75 (95% CI 1.64 to 1.87)). Although overall survival to 1 year postdischarge was 96%, children with a LLC were 2.5 times more likely to die in that year than children without a LLC (OR 2.59 (95% CI 1.64 to 2.71)). CONCLUSIONS: Children with a LLC accounted for a large proportion of the PICU population. There is an opportunity to integrate specialist paediatric palliative care services with paediatric critical care to enable choice around place of care for these children and families.


BACKGROUND: Current palliative care tools do not address distressing chronic symptoms that are most relevant to cystic fibrosis. METHODS: A CF-specific structured assessment based on a primary palliative care framework was administered to 41 adolescents and adults with CF. Descriptive and correlational analyses were conducted. RESULTS: Patients reported numerous physical and psychological symptoms (mean of 10 per patient), with psychological symptoms rated as more distressing. Anxiety (34%) and depression (44%) were prevalent and correlated with distress attributable to physical symptoms and difficulty with CF self-management, but did not correlate with disease severity. CONCLUSIONS: Individuals with CF, regardless of disease severity, face challenges managing symptom burden. Frequently reported symptoms are not consistently associated with distress, suggesting the importance of individualized evaluation. The CF-CARES (Coping, goal Assessment, and Relief from Evolving CF Symptoms) primary palliative care assessment model provides a framework for patients experiencing chronic symptoms to explore interventional options with their clinicians.


BACKGROUND: The rate of childhood cancer survival has recently reached >80%. Various adverse events among childhood cancer survivors (CCS) have been reported. Proton beams are able to avoid unnecessary irradiation to normal/vital organs. We conducted a quality of life (QOL) study for CCS who were treated with proton beam
therapy (PBT). METHODS: We included those patients treated with PBT to the brain, head, or neck and who were <15 years old at the University of Tsukuba Hospital between 1983 and 2011. Clinical information was collected from medical records. Questionnaires including the Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core Scales (which assess health-related quality of life) were sent to the families/patients. RESULTS: Sixty patients were included. Median age at treatment was 6.2 years. The number of patients with status alive/dead/unknown was 32/24/4. Median follow-up period was 63.0 months (range, 48-340 months) for survivors. Questionnaires were sent to 25 families/patients and 19 were returned. PedsQL was assessed for 17 patients. Eleven of 32 living patients had at least one comorbidity grade 3/4. Average QOL score was above that for Japanese schoolchildren and adolescents. There was no correlation with comorbidity, and only longer time from treatment was correlated with a higher PedsQL score (P = 0.006). CONCLUSION: CCS who were treated with multimodal treatment using PBT had a higher QOL score. Higher score was related to longer time since treatment, regardless of comorbidity.


OBJECTIVES/HYPOTHESIS: To assess the longitudinal risk of death following tracheostomy in the pediatric age group. STUDY DESIGN: Retrospective cohort study. METHODS: Hospital records of 513 children (<18 years) at a tertiary care children's hospital who underwent tracheostomy between 1984 and 2015 were reviewed. The primary outcome measure was time from tracheostomy to death. Secondary patient demographic and clinical characteristics were assessed, with likelihood of death using chi(2) tests and the Cox proportional hazards model. RESULTS: Median age at time of tracheostomy was 0.8 years (interquartile range, 0.3-5.2 years). The highest mortality rate (27.8%) was observed in patients in the 13- to 18-year-old age category; their mortality rate was significantly higher when compared to the lowest mortality risk group patients (age 1-4 years, P = .031). Timing of death was evenly distributed: <90 days (37.6%), 90 days to 1 year (27.1%), and >1 year after tracheostomy (35.3%). Patients who underwent tracheostomy for cardiopulmonary disease had an increased risk of mortality compared with airway obstruction (adjusted hazard ratio: 3.53, 95% confidence interval: 1.72-7.24, P < .001) and other indications. Adjusted hazard ratios for bronchopulmonary dysplasia (BPD) and congenital heart disease (CHD) were 2.63 and a 2.61, respectively (P < .001). CONCLUSIONS: Pediatric patients with tracheostomy have a high mortality rate, with an increased risk of death associated with a cardiopulmonary indication for undergoing tracheostomy. The majority of deaths occur after the index hospitalization during which the tracheostomy was performed. BPD and CHD are independent predictors of mortality in pediatric tracheostomy patients. LEVEL OF EVIDENCE: 4 Laryngoscope, 127:1701-1706, 2017.


OBJECTIVE: Congenital heart disease is commonly a manifestation of genetic conditions. Surgery and/or extracorporeal membrane oxygenation were withheld in the past from some patients with genetic conditions. We hypothesized that surgical care of children with genetic conditions has increased over the last decade, but their cardiac extracorporeal membrane oxygenation use remains lower and mortality greater. DESIGN: Retrospective cohort study. SETTING: Patients admitted to the Pediatric Health Information System database 18 years old or younger with cardiac surgery during 2003-2014. Genetic conditions identified by International Classification of Diseases, 9th Edition codes were grouped as follows: trisomy 21, trisomy 13 or 18, 22q11 deletion, and all "other" genetic conditions and compared with patients without genetic condition. PATIENTS: A total of 95,253 patients met study criteria, with no genetic conditions (85%), trisomy 21 (10%), trisomy 13 or 18 (0.2%), 22q11 deletion (1%), and others (5%). INTERVENTIONS: None. MEASUREMENTS AND MAIN RESULTS: Annual surgical cases did not vary over time. Compared to patients without genetic conditions, trisomy 21 patients, extracorporeal membrane oxygenation use was just over half (odds ratio, 0.54), but mortality with and without extracorporeal membrane oxygenation were similar. In trisomy 13 or 18 patients, extracorporeal membrane oxygenation use was similar to those without genetic condition, but all five treated with extracorporeal membrane oxygenation died. 22q11 patients compared with those without genetic condition had similar extracorporeal membrane oxygenation use, but greater odds of extracorporeal membrane oxygenation mortality (odds ratio, 3.44). Other genetic conditions had significantly greater extracorporeal membrane oxygenation use (odds ratio, 1.22), mortality with extracorporeal membrane oxygenation (odds ratio, 1.42), and even greater mortality odds without (odds ratio, 2.62). CONCLUSIONS: The proportion of children undergoing cardiac surgery who have genetic conditions did
not increase during the study. Excluding trisomy 13 or 18, all groups of genetic conditions received and benefited from extracorporeal membrane oxygenation, although extracorporeal membrane oxygenation mortality was greater for those with 22q11 deletion and other genetic conditions.


BACKGROUND: Malignant rhabdoid tumor of the kidney (MRTK) is the most aggressive childhood renal tumor with overall survival (OS) rates ranging from 22% to 42%. Whether high-dose chemotherapy with autologous stem-cell transplantation (HD SCT) in an intensive first-line treatment offers additional benefit is an ongoing discussion. METHODS: A retrospective analysis of all 58 patients with MRTK from Austria, Switzerland, and Germany treated in the framework of consecutive, prospective renal/rhabdoid tumor studies SIOP9/GPO, SIOP93-01/GPOH (where SIOP is International Society of Pediatric Oncology and GPOH is German Society of Pediatric Oncology and Hematology), SIOP2001/GPOH, and European Rhabdoid Tumor Registry from 1991 to 2014. RESULTS: Median age at diagnosis was 11 months. Fifty percent of patients had metastases or multifocal disease at diagnosis (Stage IV). Local stage distribution was as follows: not done/I/II/III-1/6/11/40. Fifteen (26%) patients underwent upfront surgery. Thirty-seven (64%) patients achieved a complete remission, 17 (29%) relapsed, 34 (59%) died of disease progression, and two (3%) died of treatment-related complication. Mean time to the first event was 3.5 months. Two-year EFS/OS (where EFS is event-free survival) for the whole group was 37 +/- 6%/38 +/- 6%. Metastases/multifocal disease, younger age, and local stage III were associated with significantly inferior survival. Eleven (19%) patients underwent HD SCT (carboplatin + thiotepa, n = 6; carboplatin + etoposide + melphalan, n = 4; others, n = 1); 2-year OS in this group was 60 +/- 15% compared to 34 +/- 8% in the non-HD SCT group (P = 0.064). However, the time needed from radiologic to histologic diagnosis, stem-cell harvest, and HD SCT must also be taken into account to avoid selection bias by excluding the highest risk group with early progression (<90 days). Thus, 2-year EFS only for patients without progression until day 90 was 60 +/- 16% consolidated by HD SCT compared to 62 +/- 11% without (P = 0.8). CONCLUSION: Our retrospective analysis suggests comparable outcomes for patients with and without HD SCT, if adjusted for early disease progression.


BACKGROUND: Opsoclonus-myoclonus syndrome is an autoimmune neurological disorder characterized by opsonoclonus, myoclonus, ataxia, and behavioral changes. Although long-term outcomes have historically been poor, including motor and cognitive disabilities, the advent of new and more aggressive immunotherapy regimens may be improving prognosis in opsonoclonus-myoclonus syndrome. METHODS: We retrospectively reviewed the records of all children diagnosed with opsonoclonus-myoclonus syndrome at BC Children’s Hospital from 2000 to 2010. Neurological outcomes were compared with those previously reported in the literature. RESULTS: Twelve children with opsonoclonus-myoclonus syndrome were identified, four of whom had an associated neuroblastoma. Two thirds of patients received initial treatment with a combination of corticosteroids, intravenous immunoglobulin (IVIG), and an additional immunosuppressant agent. After a median follow-up of three years from diagnosis, ten patients had no or minimal neurological abnormalities. Two patients had poor outcome with significant cognitive impairment. CONCLUSIONS: Most patients in this series were treated with early multimodal immunotherapy, and neurological outcomes were better than those in most historical reports. This finding is consistent with recent studies that suggest multimodal immunotherapy regimens may be improving the prognosis in this challenging disease. However, some individuals did well with less aggressive treatment, and further studies are required to determine optimal treatment approach.


BACKGROUND: The Questionnaire on Pain caused by Spasticity (QPS) is a modular patient- and observer-reported outcome measure of spasticity-related pain (SRP) in children with cerebral palsy (CP). Originally
developed for an English-speaking population, we conducted a psychometric validation of a recently developed Chinese language version of the QPS. METHODS: This was a prospective, observational study involving 137 children/adolescents with CP and upper and/or lower limb spasticity and their parents at three sites in China. Six QPS modules were used, three each for upper and lower limb SRP assessment: a patient self-report module; an interviewer-administered module used by site staff based on the cognitive, communicative, and motor abilities of a patient; and a parent/caregiver module administered for all children as an observer-reported outcome to complement the patient-reported outcome. If no assessment by the patient was possible because of age or cognitive impairments, only the parent/caregiver module was completed. Two visits with a 3-week interval provided data to evaluate and establish administrative ease of use, scoring of the QPS (factor analyses, Rasch analyses), reliability (Cronbach’s alpha, intraclass correlation coefficient), validity (correlations with quality of life [PedsQL], motor impairment [Gross Motor Function Classification System, Gross Motor Function Measure-66, Manual Ability Classification System], and spasticity [Ashworth Scale, Modified Tardieu Scale]). RESULTS: For most children, clinic staff reported no difficulties associated with general QPS use or deciding which module to use. Children (and parents) who reported more demanding activities also reported higher levels of associated SRP (or observed SRP behavior). Activity-related SRP items were combined for a total QPS score. Cronbach’s alpha was low for child self-report, but was acceptable for interviewer-administered and parent reports on SRP. Test-retest reliability was high for all modules. Moderate-strong associations were frequently seen between QPS and quality of life, and were particularly strong in the child self-report group. Relatively weak associations were observed between QPS and motor impairment and spasticity. CONCLUSIONS: This first study was successful in providing initial evidence for the psychometric properties. Clinic staff were able to administer the QPS modules easily, and both children and parents were able to complete the designated QPS appropriately.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5704623/


BACKGROUND AND PURPOSE: The diagnostic and prognostic potential of brain MR imaging before term-equivalent age is limited until valid MR imaging scoring systems are available. This study aimed to validate an MR imaging scoring system of brain injury and impaired growth for use at 29 to 35 weeks postmenstrual age in infants born at <31 weeks gestational age. MATERIALS AND METHODS: Eighty-three infants in a prospective cohort study underwent early 3T MR imaging between 29 and 35 weeks’ postmenstrual age (mean, 32(+2) +/- 1(+3) weeks); 49 males, born at median gestation of 28(+4) weeks; range, 23(+6)-30(+6) weeks; mean birthweight, 1068 +/- 312 g). Seventy-seven infants had a second MR scan at term-equivalent age (mean, 40(+6) +/- 1(+3) weeks). Structural images were scored using a modified scoring system which generated WM, cortical gray matter, deep gray matter, cerebellar, and global scores. Outcome at 12–months corrected age (mean, 12 months 4 days +/- 1(+2) weeks) consisted of the Bayley Scales of Infant and Toddler Development, 3rd ed. (Bayley III), and the Neuro-Sensory Motor Developmental Assessment. RESULTS: Early MR imaging global, WM, and deep gray matter scores were negatively associated with Bayley III motor (regression coefficient for global score beta = -1.31; 95% CI, -2.39 to -0.23; P = .02), cognitive (beta = -1.52; 95% CI, -2.39 to -0.65; P < .01) and the Neuro-Sensory Motor Developmental Assessment outcomes (beta = -1.73; 95% CI, -3.19 to -0.28; P = .02). Early MR imaging cerebellar scores were negatively associated with the Neuro-Sensory Motor Developmental Assessment (beta = -5.99; 95% CI, -11.82 to -0.16; P = .04). Results were reconfirmed at term-equivalent-age MR imaging. CONCLUSIONS: This clinically accessible MR imaging scoring system is valid for use at 29 to 35 weeks postmenstrual age in infants born very preterm. It enables identification of infants at risk of adverse outcomes before the current standard of term-equivalent age.


BACKGROUND: Treacher Collins syndrome (TCS) mainly presents with severe craniofacial developmental abnormalities characterized by a combination of bilateral downward-slanting palpebral fissures, colobomas of the lower eyelids, hypoplasia of the facial bones, cleft palate, malformation of the external ears, atresia of the external auditory canals, and bilateral conductive hearing loss. It is due to mutations in Treacher Collins syndrome 1 (TCOF1) (5q32-q33.1) and Polymerase RNA 1 polypeptides D and C (POLR1D [13q12.2], and POLR1C [6p21.1]) genes, which are responsible for increased neuroepithelial apoptosis during embryogenesis resulting in the lack
of neural crest cells involved in facial bone and cartilage formation. Altered function of the upper digestive tract has been reported, whereas severe dysmotility disorders have never been reported. We describe here the first case of TCS associated with histologically proven chronic intestinal pseudo-obstruction (CIPO) in humans. Case presentation: A 12-year-old boy with TCS due to TCOF1 gene deletion experienced nutritional difficulties and digestive intolerance from birth. CIPO was suspected during childhood because of severe intestinal dysmotility leading to enteral-jejunal nutrition intolerance and dependence on total parenteral nutrition. Diagnosis of CIPO with nervous abnormalities was histologically confirmed on a surgical rectal biopsy that showed enlarged ganglionic myenteric plexus. At the age of 9 years, an isolated colonic stenosis without dilatation responsible for severe abdominal pain and altered quality of life led to digestive derivation contributing to rapid disappearance of chronic abdominal pain. At the age of 12 years, the patient was still dependent on total home parenteral nutrition 7 days a week to maintain regular growth velocity. CONCLUSION: Recently, mice studies have pointed out the role played by TCOF1 in ganglionic cell migration in the foregut, suggesting that the synergistic haploinsufficiency of Tcof1 and Pax3, a transcription factor regulating the RET gene involved in disorders of neural crest cell development, probably results in colonic aganglionosis and may explain the association described here between TCS and CIPO. This case may correspond to this possible mechanism in humans. These findings and our clinical report suggest that CIPO may be assessed as unusual digestive manifestations in TCS with TCOF1 deletion.


PURPOSE: Medical clowning has proven effective for reducing pain, anxiety, and stress, however, its differential effects on children from different cultures have not yet been researched. This study evaluated the effects of medical-clowning intervention on anxiety and pain among Jewish and Bedouin children, and anxiety among their parents, in southern Israel. PATIENTS AND METHODS: The study was conducted in hospital pediatric departments and employed a pre-post design involving quantitative and qualitative methods. The study included 89 children whose ages ranged from 7.5 to 12 years (39 Jewish and 50 Bedouin) and 69 parents (19 Jewish and 50 Bedouin). Questionnaires assessing pain, anxiety, and demographics were used at the pre-intervention stage and pain, anxiety, and enjoyment of different aspects of the intervention were evaluated following the intervention. The intervention stage lasted for 8-10 minutes and included the use of word play, body language, and making faces, as well as the use of props brought by the clown. Semi-structured interviews were also conducted at the post-intervention stage. RESULTS: The intervention reduced pain and anxiety among both groups of children and reduced anxiety among both groups of parents. However, anxiety levels were reduced more significantly among Bedouin children. The nonverbal components of the clowns’ humor were most central, but it was the verbal components that mediated the reduction in anxiety among the Bedouin children. CONCLUSION: This study underscored the effectiveness and importance of medical clowning in reducing pain and anxiety among children in different cultural contexts. Moreover, the issue of culturally appropriate humor was underscored and implications for intercultural clown training are discussed.


Spinal muscular atrophy (SMA) is one of the most common juvenile neurodegenerative diseases, which can be associated with child mortality. SMA is caused by a mutation of ubiquitously expressed gene, Survival Motor Neuron1 (SMN1), leading to reduced SMN protein and the motor neuron death. The disease is incurable and the only therapeutic strategy to follow is to improve the expression of SMN protein levels in motor neurons. Significant numbers of motor neurons in SMA mice and SMA cultures are caspase positive with condensed nuclei, suggesting that these cells are prone to a process of cell death called apoptosis. Searching for other potential molecules or signaling pathways that are neuroprotective for central nervous system (CNS) insults is essential for widening the scope of developmental medicine. PTEN, a Phosphatase and Tensin homologue, is a tumor suppressor, which is widely expressed in CNS. PTEN depletion activates anti-apoptotic factors and it is evident that the pathway plays an important protective role in many neurodegenerative disorders. It functions as a negative regulator of PIP3/AKT pathway and thereby modulates its downstream cellular functions through lipid phosphatase activity. Moreover, previous reports from our group demonstrated that, PTEN depletion using viral vector delivery system in SMN delta7 mice reduces disease pathology, with significant rescue on survival rate and
the body weight of the SMA mice. Thus knockdown/depletion/mutation of PTEN and manipulation of PTEN medicated Akt/PKB signaling pathway may represent an important therapeutic strategy to promote motor neuron survival in SMA.


Risk stratification in Brugada syndrome in young patients remains challenging. We investigated the clinical characteristics, prognosis, and risk in young patients with the Brugada syndrome. We studied 95 patients with the Brugada syndrome aged <\=19 years. The median age at diagnosis was 12.9 years. The clinical presentation was sudden cardiac death in 7% and syncope in 21%. The remaining 72% were asymptomatic at diagnosis. Electrical abnormalities were present in 36%, including spontaneous type I electrocardiogram (12%), sinus node dysfunction (9%), atrioventricular block (17%), intraventricular conduction delay (16%), and atrial arrhythmias (8%). An electrophysiologic study was performed in 75%; ventricular arrhythmias were induced in 3%. An implantable cardioverter-defibrillator was placed in 25%. During a mean follow-up of 59 months, 9 patients presented with arrhythmic events (event rate: 1.9% per year). Variables significantly associated with events were: presentation with sudden cardiac death or syncope, spontaneous type I electrocardiogram, sinus node dysfunction and/or atrial tachycardia, conduction abnormality, and induction of ventricular arrhythmias during programmed ventricular stimulation. A model including the previous 4 main clinical variables (1, sudden cardiac death or syncope; 2, spontaneous type I electrocardiogram; 3, sinus node dysfunction and/or atrial tachycardia; and 4, conduction abnormality) had a high predictive power (C: 0.93) for the risk of lethal events. A score of >\=4 conferred a 5-year event probability of 30% that increased to 53% if the score was >\=6. In conclusion, our study validated a model to predict risk in young patients with the Brugada syndrome, which takes into account 4 clinical measures.


BACKGROUND: Standing frames are used for children with cerebral palsy (CP). They may improve body structure and function (e.g., reducing risk of hip subluxation, and improving bladder and bowel function), improving activity (e.g., motor abilities) and participation (e.g., interaction with peers), but there is little evidence that they do. We aimed to identify current UK standing frame practice for children with CP and to understand stakeholder views regarding their clinical benefits and challenges to use. METHOD: Three populations were sampled: clinicians prescribing standing frames for children with CP (n = 305), professionals (health and education) working with children with CP who use standing frames (n = 155), and parents of children with CP who have used standing frames (n = 91). Questionnaires were developed by the co-applicant group and piloted with other professionals and parents of children with CP. They were distributed online via clinical and parent networks across the UK. RESULTS: Prescribing practice was consistent, but achieving the prescribed use was not always possible. Respondents in all groups reported the perceived benefits of frames, which include many domains of the International Classification of Functioning Disability and Health for Children and Youth. Challenges of use are related to physical space and child-reported pain. CONCLUSIONS: These survey findings provide information from key stakeholders regarding current UK standing frame practice.


BACKGROUND: Consensus opinion supports standing frame use as part of postural management for nonambulant young people with cerebral palsy. Although the rationale for standing frame use and the associated challenges have been described, little attention has been given to the users’ experiences. The aim of the current study was to explore young people’s positive and negative experiences, and attitudes regarding standing frame
use. METHODS: Framework analysis informed an open exploration of young people's opinions of standing frames. Using semistructured interviews, 12 young people with cerebral palsy (6 female) were interviewed, providing the data set for transcription and thematic analysis. FINDINGS: The first theme "attitudes to standing frames" describes the young people's understanding of why they use standing frames. Although standing frames can be painful, some young people believe they should be endured to improve their body structure and function. There were mixed views about the impact standing frames have socially, with some young people feeling excluded from their peers, and others feeling as though standing frames helped them "fit in." Some young people are not offered a choice about how and when they use their standing frame. The second theme "challenges of standing frame use" highlights the issues with standing frame use such as manual handling, interference from siblings, and the lack of aesthetically pleasing standing frame designs. CONCLUSIONS: Young people report benefits related to choice, pain relief, and participation but can also cause pain, discomfort, and reduced independence and participation. Healthcare professionals should have open, informative conversations about potential benefits and challenges of standing frames on all aspects of the young people's lives, including participation and activity.


BACKGROUND AND OBJECTIVES: Despite significant morbidity and mortality associated with ESRD, these patients receive palliative care services much less often than patients with other serious illnesses, perhaps because they are perceived as having less need for such services. We compared characteristics and outcomes of hospitalized patients in the United States who had a palliative care consultation for renal disease versus other serious illnesses. DESIGN, SETTING, PARTICIPANTS, & MEASUREMENTS: In this observational study, we used data collected by the Palliative Care Quality Network, a national palliative care quality improvement collaborative. The 23-item Palliative Care Quality Network core dataset includes demographics, processes of care, and clinical outcomes of all hospitalized patients who received a palliative care consultation between December of 2012 and March of 2016. RESULTS: The cohort included 33,183 patients, of whom 1057 (3.2%) had renal disease as the primary reason for palliative care consultation. Mean age was 71.9 (SD=16.8) or 72.8 (SD=15.2) years old for those with renal disease or other illnesses, respectively. At the time of consultation, patients with renal disease or other illnesses had similarly low mean Palliative Performance Scale scores (36.0% versus 34.9%, respectively; P=0.08) and reported similar moderate to severe anxiety (14.9% versus 15.3%, respectively; P=0.90) and nausea (5.9% versus 5.9%, respectively; P>0.99). Symptoms improved similarly after consultation regardless of diagnosis (P>/=0.50), except anxiety, which improved more often among those with renal disease (92.0% versus 66.0%, respectively; P=0.002). Although change in code status was similar among patients with renal disease versus other illnesses, from over 60% full code initially to 30% full code after palliative care consultation, fewer patients with renal disease were referred to hospice than those with other illnesses (30.7% versus 37.6%, respectively; P<0.001). CONCLUSIONS: Hospitalized patients with renal disease referred for palliative care consultation had similar palliative care needs, improved symptom management, and clarification of goals of care as those with other serious illnesses.


BACKGROUND: Empirically derived and tested models are necessary to develop effective, holistic interventions to improve positive health outcomes in adolescents and young adults (AYA) with cancer, yet few exist. This article is the second of 2 articles reporting on evaluation of the Resilience in Illness Model (RIM) as a predictive model to guide positive health research and practice. OBJECTIVE: The aim of this study was to report the confirmatory model evaluation of the RIM. METHODS: A confirmatory evaluation of RIM was done using baseline data from a sample of 113 AYA aged 11 to 24 years who were undergoing hematopoietic stem cell transplant and enrolled in a randomized controlled trial of a behavioral intervention to enhance resilience. Data were analyzed using latent variable structural equation modeling. RESULTS: Goodness-of-fit indices supported RIM as a confirmed model that accounted for large amounts of variance in the outcomes of self-transcendence (62%) and resilience (72%), and in 3 of 5 mediators, specifically social integration (74%), courageous coping (80%), and hope-derived meaning (87%), as well as small to moderate amounts of variance in the remaining mediators of defensive coping (1%) and family environment (35%). CONCLUSIONS: Findings establish the RIM as a plausible predictive model.
framework for explaining ways AYA with cancer transcend their illness and achieve resilience resolution and for guiding intervention studies in this population. Additional research is needed to explore RIM’s transferability based on stage of illness, other chronic diseases, and cultural diversity. IMPLICATIONS FOR PRACTICE: Results support the RIM as an appropriate guide for developing and evaluating interventions to foster positive adjustment in AYA with cancer.


The purpose of our study was to evaluate surgical enteric access in pediatric cancer patients to determine factors associated with postoperative complications. We performed a single-institution retrospective review of all patients below 21 years old with a primary cancer diagnosis who underwent surgical procedures for enteral access between 2004 and 2014. Multivariate logistic regression was performed to determine independent predictors of postoperative complications. During the study period, 122 patients had surgically placed feeding tubes, of whom 58% developed >/=1 complication(s) and 16% experienced a major complication. No single factor was significantly associated with developing any complication or major complication. Several trends were noted including increased complications associated with jejunostomy tubes, percutaneous endoscopic gastrostomy tubes, and abdominal radiation. Surgically placed enteric access in pediatric and adolescent cancer patients is associated with an extremely high complication rate emphasizing the importance of careful evaluation of these patients before embarking on surgical feeding access. Future work should evaluate mechanisms to decrease complications and/or explore alternative methods to provide supplemental nutrition in children and adolescents with cancer.


Children with single-ventricle disease experience high mortality and complex care. In other life-limiting childhood illnesses, paediatric palliative care may mitigate maternal stress. We hypothesised that early palliative care in the single-ventricle population may have the same benefit for mothers. In this pilot randomised trial of early palliative care, mothers of infants with prenatal single-ventricle diagnoses completed surveys measuring depression, anxiety, coping, and quality of life at a prenatal visit and neonatal discharge. Infants were randomised to receive early palliative care - structured evaluation, psychosocial/spiritual, and communication support before surgery - or standard care. Among 56 eligible mothers, 40 enrolled and completed baseline surveys; 38 neonates were randomised, 18 early palliative care and 20 standard care; and 34 postnatal surveys were completed. Baseline Beck Depression Inventory-II and State-Trait Anxiety Index scores exceeded normal pregnant sample scores (mean 13.76+/-8.46 versus 7.0+/-5.0 and 46.34+/-12.59 versus 29.8+/-6.35, respectively; p=0.0001); there were no significant differences between study groups. The early palliative care group had a decrease in prenatal to postnatal State-Trait Anxiety Index scores (-7.6 versus 0.3 in standard care, p=0.02), higher postnatal Brief Cope Inventory positive reframing scores (p=0.03), and a positive change in PedsQL Family Impact Module communication and family relationships scores (effect size 0.46 and 0.41, respectively). In conclusion, these data show that mothers of infants with single-ventricle disease experience significant depression and anxiety prenatally. Early palliative care resulted in decreased maternal anxiety, improved maternal positive reframing, and improved communication and family relationships.


Probiotics are used in the prophylaxis and treatment of several conditions, including irritable bowel syndrome, diarrhoea, necrotising enterocolitis (NEC) and colic in infants. Despite the long history of probiotic use in humans, there is still significant debate about their efficacy and safety, particularly in HIV-infected and immunocompromised individuals. Here, we reviewed the safety and adverse event (AE) reporting from clinical trials that have tested probiotics in at risk populations, including HIV-infected individuals, the terminally ill and elderly, and neonates. Our analysis suggests that the benefits of probiotic therapy outweigh their potential risks.
in HIV-infected populations, and in the treatment of colic and NEC in low birth weight or premature neonates. Most case reports of severe AEs were in the elderly and terminally ill, or in those with additional severe medical conditions. We conclude that probiotic use, as adjunctive treatment, is effective and safe in the majority of patients including HIV-infected individuals, although special care should be taken in individuals with extreme immunosuppression and severe medical conditions in all ages.


OBJECTIVE: Despite growing interest in children and young people’s (CYP) perspectives on healthcare, they continue to be excluded from many patient experience surveys. This study investigated the feasibility of, and additional information gained by, measuring CYP experiences of a recent hospital admission. DESIGN: Cross-sectional analysis of national survey data. SETTING: Inpatients aged 8-15 years in eligible National Health Service hospitals, July-September 2014. PARTICIPANTS: 6204 parents/carers completed the parent section of the survey. The CYP section of the survey was completed by CYP themselves (n=3592), parents (n=849) or jointly (n=1763). MAIN OUTCOME MEASURES: Pain relief, involvement, quality of staff communication, perceived safety, ward environment, overall experience. ANALYSES: Single-measures intraclass correlations (ICCs) were used to assess the concordance between CYP and parent responses about the same inpatient episode. Multilevel logistic regression models, adjusted for individual characteristics, were used to compare the odds of positive responses when the CYP section of the survey was completed by parents, by CYP themselves or jointly. RESULTS: The CYP section of the survey was completed independently by 57.8% of CYP. Agreement between CYP and parent responses was reasonably good for pain relief (ICC=0.61 (95% CI 0.58 to 0.63)) and overall experience (ICC=0.70 (95% CI 0.68 to 0.72)), but much lower for questions comparing professionals’ communication with CYP and with their parents (ICC range=0.28 (95% CI 0.24 to 0.32) to 0.51 (95% CI 0.47 to 0.54)). In the regression models, CYP were significantly less likely than parents to report feeling safe (adjusted OR (AOR)=0.54 (95% CI 0.38 to 0.76)), involvement in decisions (AOR=0.66 (95% CI 0.46 to 0.94)) or adequate privacy (AOR=0.68 (95% CI 0.52 to 0.89)). CONCLUSIONS: Including CYP (8-15 years) in patient experience surveys is feasible and enhances what is known from parents’ responses.

https://adc.bmj.com/content/early/2018/02/06/archdischild-2017-313801


Diffuse intrinsic pontine glioma (DIPG) is a rare but uniformly fatal cancer of the brain, with peak incidence in children of 5-7 years of age. In contrast to most types of human cancer, there has been no significant improvement in treatment outcomes for patients with DIPG. Since DIPG occurs in the brainstem, a vital region of the brain, there are no surgical options for providing relief to patients, and chemotherapy as well as radiation therapy provide palliative relief at best. To date, more than 250 clinical trials evaluating radiotherapy along with conventional cytotoxic chemotherapy, as well as newer biologic agents, have failed to improve the dismal outcome when compared with palliative radiation alone. The recent discovery of somatic oncogenic histone gene mutations affecting chromatin regulation in DIPG has dramatically improved our understanding of the disease pathogenesis in DIPG, and these findings have stimulated the development of novel therapeutic approaches targeting epigenetic regulators for disease treatment. This review will discuss about the role of histone modification in chromatin machinery and epigenetic therapeutic strategies for the treatment of DIPG.


Children with severe impairment of the central nervous system (CNS) experience gastrointestinal (GI) symptoms at a high rate and severity, including retching, vomiting, GI tract pain, and feeding intolerance. Commonly recognized sources of symptoms include constipation and gastroesophageal reflux disease. There is growing awareness of sources due to the impaired nervous system, including visceral hyperalgesia due to sensitization of sensory neurons in the enteric nervous system and central neuropathic pain due to alterations in the thalamus. Challenging the management of these symptoms is the lack of tests to confirm alterations in the nervous system as a cause of symptom generation, requiring empirical trials directed at such sources. It is also common to have
multiple reasons for the observed symptoms, further challenging management. Recurrent emesis and GI tract pain can often be improved, though in some not completely eliminated. In some, this can progress to intractable feeding intolerance. This comprehensive review provides an evidence-based approach to care, a framework for recurrent symptoms, and language strategies when symptoms remain intractable to available interventions. This summary is intended to balance optimal management with a sensitive palliative care approach to persistent GI symptoms in children with severe impairment of the CNS.


The focus of critical care has evolved from saving lives to preservation of function. Morbidity rates in pediatric critical care are approximately double mortality rates. Morbidity includes complications of disease and medical care. In pediatric critical care, functional status morbidity is an intermediate outcome in the progression toward death and is the result of the same factors associated with mortality, including physiologic profiles and case-mix factors. The Functional Status Scale developed by Collaborative Pediatric Critical Care Research Network is a validated, granular, age-independent measure of functional status that has proved valuable and practical even in large outcome studies.


Pain-related functional limitations represent an important outcome domain to assess in children and adolescents with chronic pain. The aim of this study was to extend the empirical support of the 21-item Child Activity Limitations Interview (CALI-21), a well-validated measure of activity limitations, using a large, multisite sample and to develop a brief form of the measure with more interpretable scoring. A sample of 1616 youth and 1614 parents completed the CALI-21 at an initial appointment in 1 of 3 pain specialty clinics in the Midwest or Northwest United States, or as part of a research study after this initial visit. All youth also reported on usual pain intensity. The CALI-21 data from 1236 youth and parents were used in analyses. Results of the exploratory and confirmatory factor analyses supported a common 2-factor structure (Active and Routine factors) for both child- and parent-report versions. Using item reduction, the 9-item measure (CALI-9) was developed with both child and parent versions showing good internal consistency and high cross-informant reliability. Initial validity was shown by the ability of the CALI-9 to distinguish by level of pain intensity. Findings suggest that the CALI-9 is a promising brief tool for the evaluation of pain-related activity limitations in youth with chronic pain and for proxy report by parents. Advantages of the shortened scale include the revised 0 to 100-point scale, which increases interpretability, and further validation of the subscale scoring to assess specific limitations in Active and Routine physical functioning domains.


BACKGROUND: Withdrawal of mechanical ventilation is an important, but rarely explored issue in Asia during end-of-life care. This study aimed to describe the clinical characteristics and survival outcomes of terminally ill patients undergoing withdrawal of mechanical ventilation in Taiwan. METHODS: One-hundred-thirty-five terminally ill patients who had mechanical ventilation withdrawn between 2013 and 2016, from a medical center in Taiwan, were enrolled. Patients’ clinical characteristics and survival outcomes after withdrawal of mechanical ventilation were analyzed. RESULTS: The three most common diagnoses were organic brain lesion, advanced cancer, and newborn sequelae. The initiator of the withdrawal process was family, medical personnel, and patient him/herself. The median survival time was 45 min (95% confidence interval, 33-57 min) after the withdrawal of mechanical ventilation, and 102 patients (75.6%) died within one day after extubation. The median time from diagnosis of disease to receiving life-sustaining treatment and artificial ventilation support, receiving life-sustaining treatment and artificial ventilation support to "Withdrawal meeting," "Withdrawal meeting" to ventilator withdrawn, and ventilator withdrawn to death was 12.1 months, 19 days, 1 day, and 0 days, respectively.

Patients with a diagnosis of advanced cancer and withdrawal initiation by the patients themselves had a significantly shorter time interval between receiving life-sustaining treatment and artificial ventilation support to
tumors were treated with proton therapy at a single academic medical center in the United States. Median age

METHODS: From 2008 to 2011, 166 U.K. children with approved CNS tumors were treated with proton therapy to a North American facility. The aim of this study is to report patient outcomes of U.K. children referred for proton therapy early in the course of their disease. The data were obtained from Oct 1, 2008 to June 30, 2011. Conclusions: The PGIS is comparable to the PGS, has a lower response burden, and can reliably and validly predict women who may experience future intense grief associated with perinatal loss.

OBJECTIVES: Head trauma is one of the main causes of death in childhood and often leaves severe disability with serious neurological damage. Appropriate treatment must be provided immediately to improve outcomes. This study was performed to identify factors associated with a poor prognosis at an early stage of severe head injury in children. METHODS: The subjects were registered in the Japan Neurotrauma Data Bank. They were 119 children (mean age, 8 years; male, 67.2%) with severe head injury registered during a period of 4 years (from July 1, 2004 to June 30, 2006 and from July 1, 2009 to June 30, 2011). Univariate and multivariate analyses were performed to examine relationships among factors and outcome 6 months after discharge. Logistic regression analysis was performed to develop models for poor prognosis and death. RESULTS: Outcome was evaluated based on the Glasgow Outcome Scale. Seventy-three children (61.3%) had good recovery, 11 (9.2%) had moderate disability, 8 (6.7%) had severe disability, 4 (3.3%) were in a vegetative state, and 23 (19.3%) had died. Four factors were identified as predictors of a poor prognosis: serum glucose level greater than or equal to 200 mg/dL, Glasgow Coma Scale score on admission less than or equal to 5, presence of mydriasis, and presence of traumatic subarachnoid hemorrhage. Three factors were identified as predictors of death: serum glucose level greater than or equal to 200 mg/dL, Glasgow Coma Scale score on admission less than or equal to 5, and presence of mydriasis. CONCLUSIONS: Using these predictors, subsequent exacerbation may be predicted just after arrival at the hospital and appropriate treatment can be provided immediately.

OBJECTIVE: The Perinatal Grief Intensity Scale (PGIS) was developed for clinical use to identify and predict intense grief and need for follow-up after perinatal loss. This study evaluates the validity of the PGIS via its ability to predict future intense grief based on a PGIS score obtained early after a loss. METHODS: A prospective observational study was conducted with 103 international, English-speaking women recruited at hospital discharge or via the internet who experienced a miscarriage, stillbirth, or neonatal death within the previous 8 weeks. Survey data were collected at baseline using the PGIS and the Perinatal Grief Scale (PGS). Follow-up data on the PGS were obtained 3 months later. Data analysis included descriptive statistics, Cronbach's alpha, receiver operating characteristic curve analysis, and confirmatory factor analysis. RESULTS: Cronbach's alphas were >0.70 for both instruments. PGIS factor analysis yielded three factors as predicted, explaining 57.7% of the variance. The optimal cutoff identified for the PGIS was 3.535. No difference was found when the ability of the PGIS to identify intense grief was compared to the PGS (p=0.754). The PGIS was not inferior to the PGS (AUC=0.78, 95% CI 0.68-0.88, p<0.001) in predicting intense grief at the follow-up. A PGIS score >/=3.53 at baseline was associated with increased grief intensity at Time 2 (PGS: OR=1.97, 95% CI 1.59-2.34, p<0.001). CONCLUSIONS: The PGIS is comparable to the PGS, has a lower response burden, and can reliably and validly predict women who may experience future intense grief associated with perinatal loss.

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BACKGROUND: International, multidisciplinary care of children with central nervous system (CNS) tumors presents unique challenges. The aim of this study is to report patient outcomes of U.K. children referred for proton therapy early in the course of their disease. METHODS: The subjects were registered in the Japan Neurotrauma Data Bank. They were 119 children (mean age, 8 years; male, 67.2%) with severe head injury registered during a period of 4 years (from July 1, 2004 to June 30, 2006 and from July 1, 2009 to June 30, 2011). Univariate and multivariate analyses were performed to examine relationships among factors and outcome 6 months after discharge. Logistic regression analysis was performed to develop models for poor prognosis and death. RESULTS: Outcome was evaluated based on the Glasgow Outcome Scale. Seventy-three children (61.3%) had good recovery, 11 (9.2%) had moderate disability, 8 (6.7%) had severe disability, 4 (3.3%) were in a vegetative state, and 23 (19.3%) had died. Four factors were identified as predictors of a poor prognosis: serum glucose level greater than or equal to 200 mg/dL, Glasgow Coma Scale score on admission less than or equal to 5, presence of mydriasis, and presence of traumatic subarachnoid hemorrhage. Three factors were identified as predictors of death: serum glucose level greater than or equal to 200 mg/dL, Glasgow Coma Scale score on admission less than or equal to 5, and presence of mydriasis. CONCLUSIONS: Using these predictors, subsequent exacerbation may be predicted just after arrival at the hospital and appropriate treatment can be provided immediately.

was 7 years (range, 1-19). Median follow-up was 2.6 years. RESULTS: The 3-year actuarial overall survival (OS) and local control (LC) rates were 96% and 91%, respectively, for the overall group, 92% and 85% for the ependymoma subgroup (n = 57), 95% and 88% for the low-grade glioma subgroup (n = 54), and 100% and 100%, respectively, for the craniopharyngioma subgroup (n = 45). Cyst expansion was observed in 13 patients, including one case resulting in visual impairment. Serious side effects included new-onset seizures in three patients (1.8%), symptomatic vasculopathy in three patients (1.8%), and symptomatic brainstem necrosis in one patient (0.6%). CONCLUSIONS: In this cohort of British children referred overseas for proton therapy, disease control does not appear compromised, toxicity is acceptable, and improvement in long-term function is anticipated in survivors owing to the reduced brain exposure afforded by proton therapy. 


OBJECTIVES: To describe the frequency of postnatal discussions about withdrawal or withholding of life-sustaining therapy (WWLST), ensuing WWLST, and outcomes of infants surviving such discussions. We hypothesized that such survivors have poor outcomes. STUDY DESIGN: This retrospective review included registry data from 18 centers of the National Institute of Child Health and Human Development Neonatal Research Network. Infants born at 22-28 weeks of gestation who survived >12 hours during 2011-2013 were included. Regression analysis identified maternal and infant factors associated with WWLST discussions and factors predicting ensuing WWLST. In-hospital and 18- to 26-month outcomes were evaluated. RESULTS: WWLST discussions occurred in 529 (15.4%) of 3434 infants. These were more frequent at 22-24 weeks (27.0%) compared with 27-28 weeks of gestation (5.6%). Factors associated with WWLST discussion were male sex, gestational age (GA) of <24 weeks, birth weight small for GA, congenital malformations or syndromes, early onset sepsis, severe brain injury, and necrotizing enterocolitis. Rates of WWLST discussion varied by center (6.4%-29.9%) as did WWLST (5.2%-20.7%). Ensuing WWLST occurred in 406 patients; of these, 5 survived to discharge. Of the 123 infants for whom intensive care was continued, 58 (47%) survived to discharge. Survival after WWLST discussion was associated with higher rates of neonatal morbidities and neurodevelopmental impairment compared with babies for whom WWLST discussions did not occur. Significant predictors of ensuing WWLST were maternal age >25 years, necrotizing enterocolitis, and days on a ventilator. CONCLUSIONS: Wide center variations in WWLST discussions occur, especially at <24 weeks GA. Outcomes of infants surviving after WWLST discussions are poor. TRIAL REGISTRATION: ClinicalTrials.gov: NCT00063063. 


BACKGROUND: We aimed to test a novel method of delivery of chloral hydrate (CH) sedation in ventilated critically ill young children. METHODS: Children < 12 years old, within 72 hours of admission, who were ventilated, receiving enteral tube feeds, with intermittent CH ordered were enrolled after signed consent. Patients received a CH loading-dose of 10 mg/kg enterally, then a syringe-pump enteral infusion at 5 mg/kg/hour, increasing to a maximum of 9 mg/kg/hour. Cases were compared to historical controls matched for age group and Pediatric Risk of Mortality score (PRISM) category, using Fisher’s exact test and the t test. The primary outcome was feasibility, defined as the use of an enteral CH continuous infusion without discontinuation attributable to a pre-specified potential harm. RESULTS: There were 21 patients enrolled, at age 11.4 (12.1) months, with bronchiolitis in 10 (48%), a mean Pediatric Logistic Organ Dysfunction (PELOD) score of 6.2 (5.2), and having received enteral CH continuous infusion for 4.5 (2.2) days. Infusion of CH was feasible in 20/21 (95%); 95% CI 76-99% patients, with one (5%) adverse event of duodenal ulcer perforation on day 3 in a patient with croup receiving regular ibuprofen and dexamethasone. The CH infusion dose (mg/kg/h) on day 2 (n = 20) was 8.9 (IQR 5.9, 9), and on day 4 (n = 11) was 8.8 (IQR 7, 9). Days to titration of adequate sedation (defined as <3 PRN doses/shift) was 1 (IQR 0.5, 2.5), and hours to awakening for extubation was 5 (IQR 2.9). Cases (versus controls) had less positive fluid balance at 48 h (-2 (45) vs. 26 (46) ml/kg, p = 0.051), and a decrease in number of PRN sedation doses from 12 h pre to 12 hours post starting CH (4.7 (3.3) to 2.6 (2.8), p = 0.009 versus 2.9 (3.9) to 3.4 (5), p = 0.74). There were no statistically significant differences between cases and controls in inotrope scores, signs or treatment of withdrawal, or PICU days. CONCLUSIONS: Delivering CH by continuous enteral infusion is feasible, effective, and
BACKGROUND: Many adult patients with cancer who know they are dying choose less intense care; additionally, high-intensity care is associated with worse caregiver outcomes. Little is known about intensity of end-of-life care in children with cancer. METHODS: By using the California Office of Statewide Health Planning and Development administrative database, we performed a population-based analysis of patients with cancer aged 0 to 21 who died between 2000 and 2011. Rates of and sociodemographic and clinical factors associated with previously-defined end-of-life intensity indicators were determined. The intensity indicators included an intense medical intervention (cardiopulmonary resuscitation, intubation, ICU admission, or hemodialysis) within 30 days of death, intravenous chemotherapy within 14 days of death, and hospital death. RESULTS: The 3732 patients were 34% non-Hispanic white, and 41% had hematologic malignancies. The most prevalent intensity indicators were hospital death (63%) and ICU admission (20%). Sixty-five percent had > =1 intensity indicator, 23% > =2, and 22% > =1 intense medical intervention. There was a bimodal association between age and intensity: ages < 5 years and 15 to 21 years was associated with intense care. Patients with hematologic malignancies were more likely to have high-intensity end-of-life care, as were patients from underrepresented minorities, those who lived closer to the hospital, those who received care at a nonspecialty center (neither Children's Oncology Group nor National Cancer Institute Designated Cancer Center), and those receiving care after 2008. CONCLUSIONS: Nearly two-thirds of children who died of cancer experienced intense end-of-life care. Further research needs to determine if these rates and disparities are consistent with patient and/or family goals.

BACKGROUND: Studies of adolescent and young adult (AYA) oncology end-of-life care utilization are critical because cancer is the leading cause of nonaccidental AYA death and end-of-life care contributes significantly to health care expenditures. This study was designed to determine the quantity of and disparities in inpatient utilization in the last year of life of AYAs with cancer. METHODS: The California Office of Statewide Health Planning and Development administrative discharge database, linked to death certificates, was used to perform a population-based analysis of cancer patients aged 15 to 39 years who died in 2000-2011. The number of hospital days and the inpatient costs were determined for each patient in the last year of his or her life, as were clinical and sociodemographic factors associated with high inpatient utilization. Admission patterns as death approached were also evaluated. RESULTS: The 12,883 patients were admitted for 40 days on average in the last year of life, and this cost $151,072 per patient in inpatient costs. As death approached, the admission rates and the percentage of all admissions occurring at nonspecialty centers increased. Five percent of patients used 20% of bed days in the last year (high utilizers). Factors associated with high utilization included younger age (15-30 years), Hispanic ethnicity, non-health maintenance organization insurance, and hematologic malignancies. CONCLUSIONS: AYA oncology decedents were admitted for 40 days in their last year of life. Subgroups with high utilization had distinct sociodemographic and clinical characteristics, and nonspecialty center admissions increased as death approached. This demonstrates the need for palliative care at nonspecialty centers. Future studies need to determine whether these patterns are goal-concurrent, include high utilizers, and monitor the effects of health care reform.


BACKGROUND: Objective parameters predicting futility of care in severely injured pediatric patients are lacking. Although futility of care has been investigated in a limited number of studies in trauma patients, none of these studies achieves a 100% success rate in a large cohort of pediatric patients. The purpose of the current study was to identify extreme laboratory values that could be used to predict 100% mortality in severely injured children.


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STUDY DESIGN: We evaluated a registry-based, historical cohort of all severely injured children (Level I trauma, younger than 16 years old) who were not dead on arrival between January 2010 and December 2016 from a single Level I trauma center. Extreme arrival laboratory data were evaluated both alone and in conjunction with traumatic brain injury. RESULTS: There were 1,292 patients who met inclusion criteria, of which 1,169 (90.5%) survived and 123 (9.5%) died. Those who died were significantly younger, with higher head Abbreviated Injury Scale scores and overall Injury Severity Scores. Single extreme laboratory values were identified that predicted mortality perfectly (100% positive predictive value): international normalized ratio > 3.0, pH < 6.95, base excess < -22, platelet count < 30,000, hemoglobin < 5.0 g/dL, rapid thromboelastography < 30 mm, and rapid thromboelastography lysis at 30 minutes > = 50%. When 2 laboratory values or the presence of traumatic brain injury were added, lower thresholds for futility were noted. CONCLUSIONS: Extreme admission laboratory values are capable of predicting 100% mortality and futility of additional care in severely injured children with a high level of accuracy. Validation of these single-center findings is warranted and, if supported, should initiate a discussion within the pediatric trauma community about application and cessation of resuscitation efforts to optimize resource use.


Objectives: To examine parent-child communication (i.e., openness, problems) and child adjustment among youth with advanced or non-advanced cancer and comparison children. Methods: Families (n = 125) were recruited after a child’s diagnosis/relapse and stratified by advanced (n = 55) or non-advanced (n = 70) disease. Comparison children (n = 60) were recruited from local schools. Children (ages 10-17) reported on communication (Parent-Adolescent Communication Scale) with both parents, while mothers reported on child adjustment (Child Behavior Checklist) at enrollment (T1) and one year (T2). Results: Openness/problems in communication did not differ across groups at T1, but problems with fathers were higher among children with non-advanced cancer versus comparisons at T2. Openness declined for all fathers, while changes in problems varied by group for both parents. T1 communication predicted later adjustment only for children with advanced cancer. Conclusions: Communication plays an important role, particularly for children with advanced cancer. Additional research with families affected by life-limiting conditions is needed.


BACKGROUND: The aim of this study was to determine the tube-related complications and feeding outcomes of infants discharged home from the neonatal intensive care unit (NICU) with nasogastric (NG) tube feeding or gastrostomy (G-tube) feeding. MATERIALS AND METHODS: We performed a chart review of 335 infants discharged from our NICU with home NG tube or G-tube feeding between January 2009 and December 2013. The primary outcome was the incidence of feeding tube-related complications requiring emergency department (ED) visits, hospitalizations, or deaths. Secondary outcome was feeding status at 6 months postdischarge. Univariate and multivariate analyses were conducted. RESULTS: There were 322 infants discharged with home enteral tube feeding (NG tube, n = 84; G-tube, n = 238), with available outpatient data for the 6-month postdischarge period. A total of 115 ED visits, 28 hospitalizations, and 2 deaths were due to a tube-related complication. The incidence of tube-related complications requiring an ED visit was significantly higher in the G-tube group compared with the NG tube group (33.6% vs 9.5%, P < .001). Two patients died due to a G-tube-related complication. By 6 months postdischarge, full oral feeding was achieved in 71.4% of infants in the NG tube group compared with 19.3% in the G-tube group (P < .001). Type of feeding tube and percentage of oral feeding at discharge were significantly associated with continued tube feeding at 6 months postdischarge. CONCLUSION: Home NG tube feeding is associated with fewer ED visits for tube-related complications compared with home G-tube feeding. Some infants could benefit from a trial home NG tube feeding.

Recurrence of pediatric high-grade glioma is a leading cause of cancer-related death in children. We report results of a systematic review and meta-analysis investigating survival outcome in pediatric patients with recurrent high-grade glioma over the last 20 years. MEDLINE/PubMed, EMBASE, Web of Science and Cochrane Review databases were searched for relevant studies reporting on survival outcomes for pediatric patients with recurrent high-grade glioma treated between 1996 and 2016. Progression-free survival (PFS) and overall survival (OS) were calculated cumulatively over all studies, by therapy subgroup, and by decade of treatment. Random effects models were used to control for heterogeneity as measured by the I^2 statistic. A total of 17 studies across 4 treatment strategies were included. Eleven investigated traditional chemotherapy, 1 investigated targeted therapy, 3 investigated immunotherapy, and 2 investigated radiotherapy. A total of 129 patients were included with a median age of 10.0 years. Cumulative PFS was 3.5 months (95% CI 2.1-5.0). Cumulative OS was 5.6 months (95% CI 3.9-7.3). OS was 4.0 months (95% CI 1.9-6.1) using traditional chemotherapy, 9.3 months using targeted therapies (95% CI 5.4-13), 6.9 months using immunotherapy (95% CI 2.1-12), and 14 months using reirradiation (95% CI 2.8-25). OS between 1996 and 2006 was 4.2 months (95% CI 2.1-6.2) compared to 8.5 months (95% CI 5.6-11) after 2006. Pediatric patients with recurrent high-grade glioma suffer from poor PFS and OS, regardless of therapy. There may be a trend towards improved OS in the last decade. https://www.ncbi.nlm.nih.gov/pubmed/29204840


Reappraisal, a cognitive approach intended to alter an emotional response, is generally associated with prefrontal cortical recruitment and decreased limbic activity. However, the extent to which neurofunctional activity predicts successful reappraisal is unclear. During fMRI, 60 healthy participants completed a reappraisal paradigm, which included reappraising negative images to reduce emotional reactivity ('ReappNeg') and viewing negative images and experiencing the negative affect they evoke ('LookNeg'). After each trial, participants rated their emotional response on a Likert-type scale where higher values indicated more negative affect. Reappraisal ability was based on a difference value (DeltaReappNeg-LookNeg) such that negative values signified successful reappraisal ('SR'; n=38) and positive values, unsuccessful reappraisal ('USR'; n=22). Neural activity based on ReappNeg-LookNeg conditions from 37 regions of interest encompassing cortical and limbic areas was submitted to Principal Component Analysis (PCA). Resulting PCA factors were submitted to discriminant function analysis to evaluate which factor(s) predicted SR and USR groups. Results showed a factor with high loadings for certain frontal areas (e.g., left dorsomedial prefrontal cortex) and limbic regions (e.g., bilateral amygdala) predicted 71.1% of cases in the SR group and 68.2% of cases in the USR group. Additionally, successful reappraisal corresponded with more activation in the factor with high loadings for frontal areas and less activity in the factor associated with limbic regions. Results are consistent with studies of individual differences where more prefrontal engagement and less limbic activity is associated with effectual reappraisal, but for the first time, a neural 'signature' for successful reappraisal has been demonstrated. https://www.ncbi.nlm.nih.gov/pubmed/29061386


The responsive neurostimulation (RNS) system, an adjunctive treatment for pharmacoresistant partial-onset seizures with 1 or 2 foci, has been available to patients aged 18 years or older since the device’s FDA approval in 2013. Herein, the authors describe their off-label application of this technology in 2 pediatric patients and the consequent therapeutic benefit without surgical complications or treatment side effects. A 14-year-old nonambulatory, nonverbal male with severe developmental delay was considered for RNS therapy for medically and surgically refractory epilepsy with bilateral seizure onsets in the setting of a normal radiological examination and a known neuropathological diagnosis of type I cortical dysplasia. The RNS system was implanted with strip electrodes placed on the left lateral frontal and right lateral temporal neocortex. At 19 months’ follow-up, cortical stimulation resulted in sustained reduction in both seizure frequency-3 seizures per day down from 15 to 30 per day-and seizure severity. The patient subsequently underwent a trial of corticothalamic stimulation with a right temporal cortical strip and a left thalamic depth electrode, which resulted in a further 50% reduction in seizure

frequency. In a second case, a 9-year-old right-handed female with radiological evidence of a small watershed infarct on the left and medically refractory seizures was referred for presurgical evaluation. Invasive monitoring revealed an unresectable seizure focus in the eloquent cortex of the left posterior frontal and parietal lobes. The RNS device was implanted with cortical leads placed at the putative seizure focus. At 21 months after surgery, the patient had been seizure free for 4 months, following a 17-month period in which the seizure frequency had decreased from 12 per month to 2 per month, with associated functional and behavioral improvement. The authors’ results suggest that RNS may be a palliative option for children with intractable seizures whose condition warrants off-label use of the surgical device. The improved therapeutic effect noted with time and sustained RNS treatment points to a possible neuromodulatory effect.


BACKGROUND: The sense of taste is very much essential to the overall health of an individual. It is a necessary component to enjoy one’s food, which in turn provides nutrition to an individual. Any disturbance in taste perception can hamper quality of life in such patients by influencing their appetite, body weight and psychological well-being. Taste disorders have been treated using different modalities of treatment and there is no consensus for the best intervention. Hence this Cochrane Review was undertaken. This is an update of the Cochrane Review first published in November 2014. OBJECTIVES: To assess the effects of interventions for the management of patients with taste disturbances. SEARCH METHODS: Cochrane Oral Health’s Information Specialist searched the following databases: Cochrane Oral Health’s Trials Register (to 4 July 2017); the Cochrane Central Register of Controlled Trials (CENTRAL; 2017 Issue 6) in the Cochrane Library (searched 4 July 2017); MEDLINE Ovid (1946 to 4 July 2017); Embase Ovid (1980 to 4 July 2017); CINAHL EBSCO (1937 to 4 July 2017); and AMED Ovid (1985 to 4 July 2017). The US National Institutes of Health Ongoing Trials Register ClinicalTrials.gov (www.clinicaltrials.gov) and the World Health Organization International Clinical Trials Registry Platform were searched for trials. Abstracts from scientific meetings and conferences were searched on 25 September 2017. No restrictions were placed on the language or date of publication when searching the electronic databases. SELECTION CRITERIA: We included all randomised controlled trials (RCTs) comparing any pharmacological agent with a control intervention or any non-pharmacological agent with a control intervention. We also included cross-over trials in the review. DATA COLLECTION AND ANALYSIS: Two pairs of review authors independently, and in duplicate, assessed the quality of trials and extracted data. Wherever possible, we contacted trial authors for additional information. We collected adverse events information from the trials. MAIN RESULTS: We included 10 trials (581 participants), nine of which we were able to include in the quantitative analyses (566 participants). We assessed three trials (30%) as having a risk of bias, four trials (40%) at high risk of bias and three trials (30%) as having an unclear risk of bias. We only included studies on taste disorders in this review that were either idiopathic, or resulting from zinc deficiency or chronic renal failure. Of these, nine trials with 544 people compared zinc supplements to placebo for patients with taste disorders. The participants in two trials were children and adolescents with respective mean ages of 10 and 11.2 years and the other seven trials had adult participants. Out of these nine, two trials assessed the patient-reported outcome for improvement in taste acuity using zinc supplements (risk ratio (RR) 1.40, 95% confidence interval (CI) 0.94 to 2.09; 119 participants, very low-quality evidence). We meta-analysed for taste acuity improvement using objective outcome (continuous data) in idiopathic and zinc-deficient taste disorder patients (standardised mean difference (SMD) 0.44, 95% CI 0.23 to 0.65; 366 participants, three trials, very low-quality evidence). We also analysed one cross-over trial separately using the first half of the results for taste detection (mean difference (MD) 2.50, 95% CI 0.93 to 4.07; 14 participants, very low-quality evidence), and taste recognition (MD 3.00, 95% CI 0.66 to 5.34; 14 participants, very low-quality evidence). We meta-analysed taste acuity improvement using objective outcome (dichotomous data) in idiopathic and zinc-deficient taste disorder patients (RR 1.42, 95% CI 1.09 to 1.84; 292 participants, two trials, very low-quality evidence). Out of the nine trials using zinc supplementation, four reported adverse events like eczema, nausea, abdominal pain, diarrhoea, constipation, decrease in blood iron, increase in blood alkaline phosphatase, and minor increase in blood triglycerides. One trial tested taste discrimination using acupuncture (MD 2.80, 95% CI -1.18 to 6.78; 37 participants, very low-quality evidence). No adverse events were reported in the acupuncture trial. None of the included trials could be included in the meta-analysis for health-related quality of life in taste disorder patients. AUTHORS’ CONCLUSIONS: We found very low-quality evidence that was insufficient to conclude on the role of zinc supplements to improve taste acuity reported by patients and very low-quality evidence that zinc supplements improve taste acuity in patients with zinc deficiency/idiopathic taste disorders. We did not find any evidence to conclude the role of zinc supplements for improving taste discrimination, or any evidence addressing health-related quality of life due to taste disorders. We found very low-quality evidence that is not sufficient to conclude on the role of acupuncture for improving taste discrimination in
cases of idiopathic dysgeusia (distortion of taste) and hypogeusia (reduced ability to taste). We were unable to draw any conclusions regarding the superiority of zinc supplements or acupuncture as none of the trials compared these interventions. https://www.ncbi.nlm.nih.gov/pubmed/29260510


BACKGROUND & AIMS: Gastrostomies are widely used to provide long-term enteral nutrition to patients with neurologic conditions that affect swallowing (eg, following a cerebrovascular accident or for patients with motor neuron disease) or with oropharyngeal malignancies. The benefits derived from this intervention are uncertain for patients and caregivers. We conducted a prospective, multicenter cohort study to determine how gastrostomies affect health-related quality of life (HRQoL) in recipients and caregivers. METHODS: We performed a study of 100 patients who received gastrostomies (55% percutaneous endoscopic gastrostomy, 45% radiologically inserted) at 5 centers in the United Kingdom, 100 caregivers, and 200 population control subjects. We used the EuroQol-5D (comprising a questionnaire, index, visual analogue scale) to assess HRQoL for patients and caregivers before the gastrostomy insertion and then 3 months afterward; findings were compared with those from control subjects. Ten patients and 10 caregivers were also interviewed after the procedure to explore quantitative findings. Findings from the EuroQol-5D and semi-structured interviews were integrated using a mixed-methods matrix. RESULTS: Six patients died before the 3-month HRQoL reassessments. We observed no significant longitudinal changes in mean EuroQol-5D index scores for patients (0.70 before vs 0.710 after; P = .83) or caregivers (0.95 before vs 0.95 after; P = .32) following gastrostomy insertion. The semi-structured interviews revealed problems in managing gastrostomy tubes, social isolation, and psychological and emotional consequences that reduced HRQoL. CONCLUSIONS: We performed a mixed-methods prospective study of the effects of gastrostomy feeding on HRQoL. HRQoL did not significantly improve after gastrostomy insertion for patients or caregivers. The lack of significant decrease in HRQoL after the procedure indicates that gastrostomies may help maintain HRQoL. Findings have relevance to those involved in gastrostomy insertion decisions and indicate the importance of carefully selecting patients for this intervention, despite the relative ease of insertion. https://www.ncbi.nlm.nih.gov/pubmed/27840184


Denosumab is a fully human recombinant monoclonal antibody to the receptor activator of nuclear factor-kappaB ligand. Denosumab is used in the treatment of postmenopausal osteoporosis and cancer-related bone disorders. There are only very scarce data on denosumab treatment in children. 14-year-old boy with spinal muscular atrophy (SMA) and severe disuse osteoporosis (spinal bone mineral density L1-L4 BMD-6.2SD Z-score) and two prevalent fragility fractures was treated with denosumab. He received 60 mg subcutaneous injection at the baseline and seven months later. Six months after the initial injection there was a 19% increase in L1-L4 BMD. The injections were well tolerated without any adverse reactions. Calcemia remained stable (2.3-2.4 mmol/L). He was scheduled for the third denosumab injection six months later. Prior to this date, he acquired pneumonia and died due to respiratory failure, which is a frequent cause of death in patients with SMA. There was no relation to the denosumab treatment. In conclusion, one dose of denosumab significantly increased BMD in a child with severe osteoporosis. https://www.ncbi.nlm.nih.gov/pubmed/29228533


PURPOSE: The primary objective was to describe anxiety measurement instruments used in children and adolescents with cancer or undergoing hematopoietic stem cell transplantation (HSCT) and summarize their content and psychometric properties. METHODS: We conducted searches of MEDLINE, Embase, PsycINFO, HAPI, and CINAHL. We included studies that used at least one instrument to measure anxiety quantitatively in children or adolescents with cancer or undergoing HSCT. Two authors independently identified studies and abstracted study demographics and instrument characteristics. RESULTS: Twenty-seven instruments, 14 multi-item and 13 single-item, were used between 78 studies. The most commonly used instrument was the State-Trait Anxiety
BACKGROUND: Advances in pain assessment approaches now indicate which measures should be used to capture chronic pain experiences in children and adolescents. However, there is little guidance on how these tools should best be administered and reported, such as which time frames to use or how pain scores are categorised as mild, moderate, or severe. OBJECTIVE: To synthesise current evidence on unidimensional, single-item pain intensity scale selection, administration, interpretation, and reporting. METHODS: Databases were searched (inception: 18 January 2016) for studies in which unidimensional pain intensity assessments were used with children and adolescents with chronic pain. Ten quality criteria were developed by modifying existing recommendations to evaluate the quality of administration of pain scales most commonly used with children. RESULTS: Forty-six studies met the inclusion criteria. The highest score achieved was 7 out of a possible 10 (median: 5; IQR: 4–6). Usage of scales varied markedly in administrator/completer, highest anchors, number of successive assessments, and time referent periods used. CONCLUSIONS: Findings suggest these scales are selected, administered, and interpreted inconsistently, even in studies of the same type. Furthermore, methods of administration are rarely reported or justified making it impossible to compare findings across studies. This article concludes by recommending criteria for the future reporting of paediatric chronic pain assessments in studies.


INTRODUCTION: In 2013, the Pediatric Association of the Netherlands launched an evidence-based guideline 'Palliative care for children'. To promote implementation in daily practice and hereby improve quality of paediatric palliative care, we aimed to develop a functional individualised paediatric palliative care plan (IPPCP) that covers physical, psychological, spiritual and social functioning, with great emphasis on the guideline's recommendations, advance care planning and patients' and parents' preferences and desires. METHODS: A Dutch working group (28 individuals) with a strong multidisciplinary character developed a draft IPPCP, which was piloted retrospectively.
and prospectively. In the pilots we completed, the IPPCPs for patients who were recently diagnosed with a life-threatening or life-limiting condition and evaluated completeness, usability and user-friendliness. RESULTS: The final IPPCP comprised five domains: (1) IPPCP data, (2) basics, (3) social, (4) psychosocial and spiritual, and (5) physical care. Each domain covered various components. In both pilots, the IPPCP was considered a comprehensive document that covered all areas of paediatric palliative care and was experienced as an improvement to the present situation. However, the current form was regarded to lack user-friendliness.

CONCLUSION: We propose a set of essential components of a comprehensive IPPCP for paediatric palliative care with extra attention for advance care planning and anticipatory action. Patients’ and parents’ preferences and desires are included next to the recommendations of the evidence-based guideline ‘Palliative care for children’.


Children and adolescents who require limb amputation as part of cancer treatment face many physical and emotional challenges. Preparatory interventions may serve to facilitate positive coping and improve long-term adjustment during pediatric cancer treatment, including decreasing anxiety and postoperative distress. This review aimed to examine and identify the type and degree of psychosocial preparation provided to the child with cancer and family prior to amputation. Electronic databases including Embase, PubMed, and PsyCINFO were searched for relevant research articles. Five studies were identified that satisfied inclusion criteria and revealed common themes for preparatory interventions, but results were limited by a lack of empirical approaches and revealed little consensus on pre-operative support prior to amputation. These findings demonstrate that there is a lack of studies to date that have adequately addressed psychosocial preparation prior to amputation for pediatric oncology patients. Future research on preparatory interventions is needed using prospective and quantitative research to establish evidence-based recommendations for interventions to support this vulnerable population.


OBJECTIVES: To identify patient-reported paediatric advance care planning (pACP) needs of adolescents living with HIV and to examine the congruence with their family’s perception of their needs. METHODS: A cross-sectional survey among six paediatric hospital-based outpatient HIV specialty clinics. Participants included 48 adolescent/family dyads (n=96 participants) within a larger study facilitating pACP. The main outcome measure was the Lyon Advance Care Planning Survey - Adolescent and Surrogate Versions - Revised. RESULTS: Adolescents’ mean age was 18 years (range >/=14 -<21); 54% male; 92% African-American; 27% with prior AIDS diagnosis. If dying, 92% believed in completing an advance directive; 85% preferred to die at home; 88% knowing how to say good bye; 71% being off machines that extend life and 77% dying a natural death. Best timing for end-of-life (EOL) decisions was while healthy (38%), when first diagnosed (17%), when first sick from a life-threatening illness (4%), when first hospitalised (8%), if dying (4%) and all of the above (19%). Prevalence-adjusted bias-adjusted Kappa (PABAK) measured congruence in pACP needs within adolescent/family dyads. There was substantial congruence in that being free from pain (PABAK=0.83), and understanding your treatment choices (PABAK=0.92) were very important or important. There was discordance about being off machines that extend life (PABAK=0.08) and when is the best time to bring up EOL decisions (PABAK=0.32). CONCLUSIONS: Areas of discordance were associated with life-sustaining choices and when to have the EOL conversation. Targeted, adolescent/family-centred, evidence-based pACP interventions are needed to improve family understanding of youth’s EOL wishes.

TRIAL REGISTRATION NUMBER: NCT01289444; Results.


The objective of this study is to determine if pediatric advance care planning (pACP) increases adolescent/family congruence in end-of-life (EOL) treatment preferences longitudinally. Adolescents aged 14-21 years with HIV/AIDS and their families were randomized (N = 105 dyads) to three-60-minute sessions scheduled one week
for possible agitation) were found to be associated with acute laryngeal lesions. Adequate sedation and

1.070; P = 0.041). The amount of tube repositioning episodes and the need for extra d

of sedation per day of intubation, there was also an increase of 3.5% on the same baseline risk (RR 95% CI 1.001

with repositioning of the endotracheal tube, there was an increase of 7.3% (RR 95% CI 1.012

presented moderate to severe

children were enrolled between November 2005 and December 2015. At FFL examination, 102 children (44.15%)

8 h after extubation. A blinded researcher identified and classified laryngeal lesions based on recorded media. 231

patients considered on palliative care. All patients underwent flexible fiber

history of laryngeal disease, current or past tracheostomy, the presence of craniofacial malformations and

required endotracheal intubation for at least 24 h were enrolled. Exclusion

The objective of this study is to determine the inc


The objective of this study is to determine the incidence of post-extubation acute laryngeal lesions in a pediatric

intensive care unit (PICU) and potential risk factors. Children, aged 28 days to 5 years, admitted to the PICU who

quired endotracheal intubation for at least 24 h were enrolled. Exclusion criteria were a previous intubation,

history of laryngeal disease, current or past tracheostomy, the presence of craniofacial malformations and

patients considered on palliative care. All patients underwent flexible fiber-optic laryngoscopy (FFL) not later than

8 h after extubation. A blinded researcher identified and classified laryngeal lesions based on recorded media. 231

children were enrolled between November 2005 and December 2015. At FFL examination, 102 children (44.15%)

presented moderate to severe laryngeal lesions. On a multivariable analysis, we found that for each additional day

of sedation per day of intubation, there was also an increase of 3.5% on the same baseline risk (RR 95% CI 1.001-

1.070; P = 0.041). The amount of tube repositioning episodes and the need for extra doses of sedation (as a proxy

for possible agitation) were found to be associated with acute laryngeal lesions. Adequate sedation and


BACKGROUND: Few reports of palliative radiotherapy (RT) for pediatrician malignant diseases have been published. We described clinical indications, outcomes, and toxicities for children who received palliative RT. PROCEDURE: Pediatric patients (age </=18 years) treated with palliative RT for incurable cancer from January 1 2008 to February 26, 2014 were included. Diagnosis, details of RT, treatment response, toxicity, and survival were retrospectively reviewed. RESULTS: Forty-six patients received 76 RT courses. Fifteen patients (33%) had >/=2 courses. Median age at palliative RT was 10.3 years; 54% were male. The most common diagnoses were neuroblastoma (20%) and diffuse intrinsic pontine glioma (17%). The most common indications for RT were oligometastatic disease in asymptomatic patients (39%) and pain (25%). The most common treatment sites were brain (32%) and bone (29%). Median RT dose was 30 Gy. Median number of RT fractions was 12. Sixty-five treatment courses (86%) were delivered with fraction sizes >/=2.5 Gy. Twenty-seven treatment courses (36%) were given under general anesthesia. Median follow-up was 3.9 months. Grade 1-2 RT-related toxicity occurred in 21% of treatment courses and 4-8% up to 12 months after RT. Two patients had Grade 3 toxicity during RT (esophagitis). Of symptomatic patients, 91%, 73%, 58%, and 43% had improved or stable symptoms during RT and 0-3, 3-6, and 6-12 months afterwards, respectively. Median survival after palliative RT was 4.2 months. Four of 21 surviving patients (19%) had hospice care at last follow-up. CONCLUSIONS: Palliative RT was well tolerated in children with incurable malignancies, with most cases associated with acceptable toxicity, and improved or stable symptoms.


minimized tube repositioning should be pursued to possibly prevent the development of post-extubation airway compromise.


CONTEXT: Although access to subspecialty pediatric palliative care (PPC) is increasing, little is known about the role of PPC for children with advanced heart disease (AHD). OBJECTIVES: The objective of this study was to examine features of subspecialty PPC involvement for children with AHD. METHODS: This is a retrospective single-institution medical record review of patients with a primary diagnosis of AHD for whom the PPC team was initially consulted between 2011 and 2016. RESULTS: Among 201 patients, 87% had congenital/structural heart disease, the remainder having acquired/nonstructural heart disease. Median age at initial PPC consultation was 7.7 months (range 1 day-28.8 years). Of the 92 patients who were alive at data collection, 73% had received initial consultation over one year before. Most common indications for consultation were goals of care (80%) and psychosocial support (54%). At initial consultation, most families (67%) expressed that their primary goal was for their child to live as long and as comfortably as possible. Among deceased patients (n = 109), median time from initial consultation to death was 33 days (range 1 day-3.6 years), and children whose families expressed that their primary goal was for their child to live as comfortably as possible were less likely to die in the intensive care unit (P = 0.03) and more likely to die in the setting of comfort care or withdrawal of life-sustaining interventions (P = 0.008). CONCLUSION: PPC involvement for children with AHD focuses on goals of care and psychosocial support. Findings suggest that PPC involvement at end of life supports goal-concordant care. Further research is needed to clarify the impact of PPC on patient outcomes.


BACKGROUND: Pediatric patients, especially in the preverbal stage, cannot self-report intensity of pain therefore several validated observational tools, including the Face, Legs, Activity, Cry, Consolability (FLACC) Behavioral Scale, have been used as a benchmark to evaluate pediatric pain. Unfortunately, this scale is currently unavailable in Japanese, precluding its widespread use in Japanese hospitals. OBJECTIVES: To translate and verify the validity and reliability of the Japanese version of the FLACC Behavioral Scale. METHOD: Back-translation was first conducted by eight medical researchers, then an available sample of patients at the University of Tsukuba Pediatric Intensive Care Unit (from May 2017 to August 2017) was enrolled in a clinical study. Two researchers evaluated the validity of the translated FLACC Behavioral Scale by weighted kappa coefficient and intraclass correlation coefficients (ICC). Observational pain was simultaneously measured by the visual analog scale (VAS obs) and reliability was evaluated by correlation analysis. RESULT: The original author approved the translation. For the clinical study, a total of 121 observations were obtained from 24 pediatric patients. Agreement between observers was highly correlated for each of the FLACC categories (Face: kappa = 0.85, Leg: kappa = 0.74, Activity: kappa = 0.89, Cry: kappa = 0.93, Consolability: kappa = 0.93) as well as the total score (Total: kappa = 0.95). Correlation analysis demonstrated a good criterion validation between the FLACC scale and the VAS obs. (r = 0.96). CONCLUSION: Our Japanese version of the FLACC Behavioral Scale shows high validity and reliability.


OBJECTIVE: This multicenter, parallel-group, randomized trial examined the effects of an animal-assisted intervention on the stress, anxiety, and health-related quality of life for children diagnosed with cancer and their parents. METHOD: Newly diagnosed patients, aged 3 to 17 years (n = 106), were randomized to receive either standard care plus regular visits from a therapy dog (intervention group), or standard care only (control group). Data were collected at set points over 4 months of the child’s treatment. Measures included the State-Trait Anxiety Inventory, Pediatric Quality of Life Inventory, Pediatric Inventory for Parents, and child blood pressure and
heart rate. All instruments were completed by the child and/or his/her parent(s). RESULTS: Children in both groups experienced a significant reduction in state anxiety (P < .001). Parents in the intervention group showed significantly decreased parenting stress (P = .008), with no changes in stress among parents in the control group. However, no significant differences between groups over time on any measures were observed. CONCLUSIONS: Animal-assisted interventions may provide certain benefits for parents and families during the initial stages of pediatric cancer treatment.


Aim The aims of this study were to examine the prevalence and potential correlates of feeding difficulties in infants who underwent cardiac surgery in the neonatal period and to investigate resource utilisation by infants with feeding difficulties. METHODS: All neonates who underwent their first cardiac surgery at the Heart Centre for Children, The Children's Hospital at Westmead, between January and December, 2009 were included. Demographic, preoperative, intraoperative, and postoperative data were collected via electronic medical records. For the purpose of this study, feeding difficulty was defined as the requirement for ongoing tube feeding at the time of discharge home or transfer to another hospital. RESULTS: Out of a total of 79 neonates, 24 (30%) were discharged home or transferred to another hospital with a feeding tube. Feeding difficulties were associated with the presence of a genetic syndrome (p<0.0001), assisted feeding preoperatively (odds ratio (OR)=4.4, p=0.03), and having a palliative procedure before biventricular repair (OR=5.1, p=0.02). Infants with feeding difficulties had significantly more reviews by speech pathologists (M=5.9, SD=7.9), dieticians (M=5.9, SD=5.4), and cardiac clinical nurse consultants (M=1.2, SD=1.4) compared with those without feeding difficulties. CONCLUSIONS: This study identified factors that can be used in the early recognition of infant feeding difficulties, to help guide the direction of limited health resources, as well as being focal points for future research and clinical practice improvement.


CONTEXT: Brain injury during prenatal and preoperative postnatal life might play a major role in neurodevelopmental impairment in infants with congenital heart disease (CHD) who require corrective or palliative surgery during infancy. A systematic review of cerebral findings during this period in relation to neurodevelopmental outcome (NDO), however, is lacking. OBJECTIVE: To assess the association between prenatal and postnatal preoperative cerebral findings and NDO in infants with CHD who require corrective or palliative surgery during infancy. DATA SOURCES: PubMed, Embase, reference lists. STUDY SELECTION: We conducted 3 different searches for English literature between 2000 and 2016; 1 for prenatal cerebral findings, 1 for postnatal preoperative cerebral findings, and 1 for the association between brain injury and NDO. DATA EXTRACTION: Two reviewers independently screened sources and extracted data on cerebral findings and neurodevelopmental outcome. Quality of studies was assessed using the Newcastle-Ottawa Quality Assessment Scale. RESULTS: Abnormal cerebral findings are common during the prenatal and postnatal preoperative periods. Prenatally, a delay of cerebral development was most common; postnatally, white matter injury, periventricular leukomalacia, and stroke were frequently observed. Abnormal Doppler measurements, brain immaturity, cerebral oxygenation, and abnormal EEG or amplitude-integrated EEG were all associated with NDO. LIMITATIONS: Observational studies, different types of CHD with different pathophysiological effects, and different reference values. CONCLUSIONS: Prenatal and postnatal preoperative abnormal cerebral findings might play an important role in neurodevelopmental impairment in infants with CHD. Increased awareness of the vulnerability of the young developing brain of an infant with CHD among caregivers is essential.


PURPOSE: The aim is to study the clinicodemographic profile and treatment outcome of ocular surface squamous neoplasia (OSSN). METHODS: This was a retrospective observational study of 57 eyes (56 cases) with clinically diagnosed OSSN, presenting in our center over the past year. RESULTS: The median age of presentation was 55
years with male:female ratio being 4.5:1. Systemic predisposing conditions were xeroderma pigmentosa (1) postkidney transplant immunosuppression (1), and human immunodeficiency virus infection (1). Patients with predisposing conditions had a younger median age of onset (33 years). The majority of tumors were nodular (61.4%), gelatinous (61.4%), and had limbal involvement (96%). On ultrasound biomicroscopy (UBM), mean tumor height was 2.93 +/- 1.02 mm, and intraocular extension was evident in seven eyes. OSSN with intraocular extension had a mean tumor height of 4.3 +/- 1.32 mm. Nodal metastasis was seen in one case at presentation. As per American Joint Committee for Cancer Classification seventh edition staging-two cases were T1, one was T2, 46 were T3 and eight were T4. Treatment advised included conservative therapy for 39; wide local excision (4 mm margin clearance) with cryotherapy for seven; enucleation in four; and exenteration in four eyes. Overall, complete regression was achieved in 88% of cases during a mean follow-up of 13.5 +/- 4.6 months. Recurrence was seen in three cases, which were treated with exenteration, radical neck dissection, and palliative chemoradiotherapy, respectively. CONCLUSION: Although associated with old age, earlier onset of OSSN is seen in patients with systemic predisposing conditions. Thicker tumors in the setting of a previous surgery or immunocompromised status should be considered high-risk features for intraocular extension and should be evaluated on UBM.


BACKGROUND: Nusinersen is an antisense oligonucleotide drug that modulates pre-messenger RNA splicing of the survival motor neuron 2 (SMN2) gene. It has been developed for the treatment of spinal muscular atrophy (SMA). METHODS: We conducted a multicenter, double-blind, sham-controlled, phase 3 trial of nusinersen in 126 children with SMA who had symptom onset after 6 months of age. The children were randomly assigned, in a 2:1 ratio, to undergo intrathecal administration of nusinersen at a dose of 12 mg (nusinersen group) or a sham procedure (control group) on days 1, 29, 85, and 274. The primary end point was the least-squares mean change from baseline in the Hammersmith Functional Motor Scale-Expanded (HFMSE) score at 15 months of treatment; HFMSE scores range from 0 to 66, with higher scores indicating better motor function. Secondary end points included the percentage of children with a clinically meaningful increase from baseline in the HFMSE score (>/>=3 points), an outcome that indicates improvement in at least two motor skills. RESULTS: In the prespecified interim analysis, there was a least-squares mean increase from baseline to month 15 in the HFMSE score in the nusinersen group (by 4.0 points) and a least-squares mean decrease in the control group (by -1.9 points), with a significant between-group difference favoring nusinersen (least-squares mean difference in change, 5.9 points; 95% confidence interval, 3.7 to 8.1; P<0.001). This result prompted early termination of the trial. Results of the final analysis were consistent with results of the interim analysis. In the final analysis, 57% of the children in the nusinersen group as compared with 26% in the control group had an increase from baseline to month 15 in the HFMSE score of at least 3 points (P<0.001), and the overall incidence of adverse events was similar in the nusinersen group and the control group (93% and 100%, respectively). CONCLUSIONS: Among children with later-onset SMA, those who received nusinersen had significant and clinically meaningful improvement in motor function as compared with those in the control group. (Funded by Biogen and Ionis Pharmaceuticals; CHERISH ClinicalTrials.gov number, NCT02292537 ).


BACKGROUND: Neurological dysfunction may occur after corrective cardiac surgery using cardio-pulmonary bypass (CPB) with or without circulatory arrest. Different neurophysiological monitoring systems have been employed to detect neurological complications and possible brain injury in infants and children during and after cardiac surgery. The value of Electroencephalogram (EEG) in infants and children at risk for neurological sequelae has not been systematically studied. METHODS: Sequential performance of two EEGs before and after cardiac surgery at a tertiary University Hospital to screen for possible brain injury after cardiac surgery in neonates and children undergoing CPB surgery. In addition, a complete neurological examination and assessment by a physiotherapist was performed. RESULTS: Over a 4-year period, in 313 patients (age: 54.2 +/- 55.7 months; normal initial EEG) after cardiac surgery CPB (duration of surgery: 146.0 +/- 58.9 min; aortic cross clamp time: 34.1
+/- 19.1 min), a 19-channel EEG recording was performed 2.4 +/- 1.8 days prior to and 11.6 +/- 5.3 days after cardiac surgery. An abnormal EEG was detected in only 8 of 313 patients (2.5%; focal slowing: 1, generalised slowing: 5, epileptiform discharges: 2) after cardiac surgery, while the EEG was normal in the remaining 305 patients (97.5%). In 1 patient, an intra-cerebral pathology was seen on MRI (ischemic); in 5 patients, follow-up EEGs were performed, which revealed normalized findings. None of the 8 patients demonstrated new focal neurological deficits on physical examination, but 33 (9.7%) children demonstrated minor abnormalities (e.g., subtle motor asymmetry, increase in muscle tone, etc.), which were unrelated to abnormal EEG findings.

CONCLUSIONS: According to the used protocol, pathological EEG findings were very infrequent in our study cohort. The routine and indiscriminate recording of EEGs in children before and after corrective or palliative cardiac surgery for congenital heart disease using CPB is not recommended. Further intra-operative neuromonitoring methods with immediate intervention should be evaluated.


OBJECTIVE: Guidelines recommend individual decision making on resuscitating infants of 22-24 weeks' gestational age (GA) at birth. When the decision not to resuscitate is made, infants would likely die soon after delivery, and under some circumstances such neonatal deaths may be registered as stillbirths occurring during delivery (intrapartum stillbirth). Thus we assessed whether socioeconomic factors are associated with perinatal death (during or within 1 hour of delivery) of infants delivered at 22-24 weeks' gestation. METHODS: We analysed 14 726 singletons of 22-24 weeks’ GA using the 2003-2011 Japanese vital statistics, and assessed how maternal characteristics influence risk of perinatal death as well as intrauterine fetal death (IUFD) and death after 1 hour of age until 40 weeks postmenstrual age. RESULTS: Living in a municipality with low-average income (lowest tertile (risk ratio 1.32, 95% CI 1.20 to 1.44), middle tertile (risk ratio 1.08, 95% CI 0.98 to 1.19)), younger maternal age (age <20 (risk ratio 1.43, 95% CI 1.17 to 1.75), age 20-34 (risk ratio 1.14, 95% CI 1.03 to 1.27)) and having previous live births (risk ratio 1.08, 95% CI 1.01 to 1.17) increased risk of perinatal deaths, but did not increase risk of IUFD or deaths after 1 hour of age. Perinatal death was twice as likely to occur in births to multiparous teenage mothers in a low-income municipality, compared with those of older primiparous mothers in a wealthier municipality. CONCLUSIONS: Socioeconomic factors substantially influence whether births of 22-24 weeks’ GA survive delivery and the first hour of life. Such disparities may reflect the impact of socioeconomic situations on decision making for resuscitation.


We aimed to retrospectively assess treatments/outcomes, including the value of high-dose-chemotherapy and autologous-stem-cell-rescue (HDC + AuSCR) and re-irradiation, in a large, European patient-cohort with relapsed intracranial germ-tumors (GCTs) receiving uniform first-line therapy, including radiotherapy as standard-of-care. Fifty-eight UK/German patients (48 male/10 female) with relapsed intracranial-GCTs (13 germinoma/45 non-germinomatous GCT (NGGCT)) treated 1996-2010 as per the SIOP-CNS-GCT-96 protocol were evaluated. For germinoma, six patients relapsed with germinoma and five with NGGCT (one palliative, one teratoma patient excluded). Five-year overall survival (OS) for the whole-group (n = 11) was 55%. Four of six germinoma relapses and two of five relapsing with NGGCT were salvaged; patients were salvaged with either standard-dose-chemotherapy (SDC) and re-irradiation or HDC + AuSCR with/without re-irradiation. Of 45 relapsed NGGCT patients, 13 were excluded (three non-protocol adherence, five teratoma, five palliation). Five-year OS for the remaining 32 relapsed malignant NGGCT patients treated with curative intent was 9% (95%CI: 2-26%). By treatment received, 5-year OS for the 10 patients receiving SDC and 22 patients treated with intention for HDC + AuSCR was 0% (0-0%) and 14% (3-36%), respectively. The three relapsed NGGCT survivors had raised HCG markers alone; two received additional irradiation. Patients with relapsed germinoma had better 5-year OS than those with relapsed NGGCT (55 vs. 9%; p = 0.007). Patients with relapsed germinoma were salvaged both with SDC and re-irradiation or HDC + AuSCR with/without re-irradiation; both represent valid treatment options. Outcomes for malignant relapse following initial diagnosis of NGGCT were exceptionally poor; the few survivors received thiotepa-based HDC + AuSCR, which is a treatment option at first malignant relapse for such patients, with further surgery/irradiation where feasible.

OBJECTIVE: Glioblastoma multiforme (GBM) is an aggressive primary brain tumor with dismal survival. This study aims to examine the prognostic value of primary tumor sites and race on survival outcomes. METHODS: Patient data obtained from the Scott and White Hospital Brain Tumor Registry (1976-2013) were stratified according to sex, age, race, primary tumor site, vital status, and survival. RESULTS: Of the 645 patients, 580 (89.9%) were diagnosed with GBM not otherwise specified (GBM NOS), 57 (8.8%) with GBM, and 8 (1.2%) with giant–cell GBM. Most were male (53.5%), aged 50 years or older (78.7%). The white population had the highest GBM prevalence (87.1%) and the lowest overall survival versus all other race groups (6.6% vs. 30.1%; P < 0.01). The black population had a relatively low prevalence of GBM (5.9%) and the greatest overall survival versus all others (47.4% vs. 7.3%; P < 0.01). Primary tumor sites located in the temporal (25.8% vs. 20.2%; P = 0.03), occipital (8.1% vs. 2.9%; P = 0.05), and parietal lobes (24.2% vs. 20.8%; P = 0.05) had a greater occurrence in surviving individuals. The overall survival for men versus women was (62.9% vs. 37.1%; P = 0.12). CONCLUSIONS: Black racial background and temporal, occipital, or parietal primary tumor sites are suggestive of positive survival outcomes. Conversely, white racial background with primary tumor sites in the brain overlapping and NOS areas seem to be associated with negative outcomes and decreased survival. Thus, racial background and primary tumor site may be useful prognostic factors in patients with GBM.


OBJECTIVE: To describe the perceptions of patients, their caregivers, and their healthcare providers to the development of a new specific instrument for assessment of the quality of life (QoL) in patients with mucopolysaccharidoses (MPS) using a qualitative focus group (FG) design. FGs were held in two Brazilian states (Rio Grande do Sul and Rio de Janeiro). RESULTS: Three versions of the new instrument were developed, each for a different age group: children (age 8-12 years), adolescents (age 13-17), and adults (age >/= 18). The FGs mostly confirmed the relevance of items. All FGs unanimously agreed on the facets: School, Happiness, Life Prospects, Religiosity, Pain, Continuity of Treatment, Trust in Treatment, Relationship with Family, Relationship with Healthcare Providers, Acceptance, and Meaning of Life. The overall concept of QoL (as proposed by the WHO-World Health Organization) and its facets apply to this patient population. However, other specific facets—particularly concerning clinical manifestations and the reality of the disease-were suggested, confirming the need for the development of a specific QoL instrument for MPS.

inotropes/vasopressors, hemodialysis, and antibiotics. Almost none of the patients had advance directives in place. Palliative care team was available in only few cases. https://www.ncbi.nlm.nih.gov/pubmed/28206733


BACKGROUND: Preterm infant pain can be relieved by combining non-nutritive sucking (sucking), oral sucrose, and facilitated tucking (tucking), but the pain-relief effects of oral expressed breast milk (breast milk) are ambiguous. AIMS: We compared the effects of combined sucking + breast milk, sucking + breast milk + tucking, and routine care on preterm infant pain during and after heel-stick procedures. DESIGN: A prospective, randomized controlled trial. SETTINGS: Level III neonatal intensive care unit and a neonatal unit at a medical center in Taipei. PARTICIPANTS/SUBJECTS: Preterm infants (N=109, gestational age 29-37 weeks, stable disease condition) needing procedural heel sticks were recruited by convenience sampling and randomly assigned to three treatment conditions: routine care, sucking + breast milk, and sucking + breast milk + tucking. METHODS: Pain was measured by watching video recordings of infants undergoing heel-stick procedures and scoring pain at 1-min intervals with the Premature Infant Pain Profile. Data were collected over eight phases: baseline (phase 1, 10min without stimuli before heel stick), during heel stick (phases 2 and 3), and a 10-min recovery (phases 4-8). RESULTS: For infants receiving sucking + breast milk, pain-score changes from baseline across phases 2-8 were 2.634, 4.303, 2.812, 2.271, 1.465, 0.704, and 1.452 units lower than corresponding pain-score changes of infants receiving routine care (all p-values <0.05 except for phases 6 and 7). Similarly, for infants receiving sucking + breast milk + tucking, pain-score changes from baseline were 2.652, 3.644, 1.686, 1.770, 1.409, 1.165, and 2.210 units lower than corresponding pain-score changes in infants receiving routine care across phases 2-8 (all p-values <0.05 except for phase 4). After receiving sucking + breast milk + tucking and sucking + breast milk, infants' risk of mild pain (pain score >/=6) significantly decreased 67.0% and 70.1%, respectively, compared to infants receiving routine care. After receiving sucking + breast milk + tucking and sucking + breast milk, infants' risk of moderate-to-severe pain (pain score >/=12) decreased 87.4% and 95.7%, respectively, compared to infants receiving routine care. CONCLUSION: The combined use of sucking + breast milk + tucking and sucking + breast milk effectively reduced preterm infants' mild pain and moderate-to-severe pain during heel-stick procedures. Adding facilitated tucking helped infants recover from pain across eight phases of heel-stick procedures. Our findings advance knowledge on the effects of combining expressed breast milk, sucking, and tucking on preterm infants' procedural pain. https://www.ncbi.nlm.nih.gov/pubmed/29100198


OBJECTIVE: Seizure onset within the insula is increasingly recognized as a cause of intractable epilepsy. Surgery within the insula is difficult, with considerable risks, given the rich vascular supply and location near critical cortex. MRI-guided laser interstitial thermal therapy (LiTT) provides an attractive treatment option for insular epilepsy, allowing direct ablation of abnormal tissue while sparing nearby normal cortex. Herein, the authors describe their experience using this technique in a large cohort of children undergoing treatment of intractable localization-related epilepsy of insular onset. METHODS: The combined epilepsy surgery database of Cook Children's Medical Center and Dell Children's Hospital was queried for all cases of insular onset epilepsy treated with LiTT. Patients without at least 6 months of follow-up data and cases preoperatively designated as palliative were excluded. Patient demographics, presurgical evaluation, surgical plan, and outcome were collected from patient charts and described. RESULTS: Twenty patients (mean age 12.8 years, range 6.1-18.6 years) underwent a total of 24 LiTT procedures; 70% of these patients had normal findings on MRI. Patients underwent a mean follow-up of 20.4 months after their last surgery (range 7-39 months), with 10 (50%) in Engel Class I, 1 (5%) in Engel Class II, 5 (25%) in Engel Class III, and 4 (20%) in Engel Class IV at last follow-up. Patients were discharged within 24 hours of the procedure in 15 (63%) cases, in 48 hours in 6 (24%) cases, and in more than 48 hours in the remaining cases. Adverse functional effects were experienced following 7 (29%) of the procedures: mild hemiparesis after 6 procedures (all patients experienced complete resolution or had minimal residual dysfunction by 6 months), and expressive language dysfunction after 1 procedure (resolved by 3 months). CONCLUSIONS: To their knowledge, the authors present the largest cohort of pediatric patients undergoing insular surgery for treatment of intractable epilepsy. The patient outcomes suggest that LiTT can successfully treat intractable seizures originating
within the insula and offers an attractive alternative to open resection. This is the first description of LiTT applied to insular epilepsy and represents one of only a few series describing the use of LiTT in children. The results indicate that seizure reduction after LiTT compares favorably to that after conventional open surgical techniques. https://www.ncbi.nlm.nih.gov/pubmed/29027866


Half of children admitted after surgery experience intense pain in hospital, and many experience continued pain and delayed functional recovery at home. However, there is a gap in tools available to measure acute functional ability in pediatric postsurgical settings. We aimed to validate the Youth Acute Pain Functional Ability Questionnaire (YAPFAQ) in a large inpatient pediatric surgical population, evaluate its responsiveness to expected functional recovery, and develop a short form for broad clinical implementation. The YAPFAQ is a self-report measure assessing acute functional ability, developed in children admitted for acute sickle cell pain. We evaluated psychometric properties of the measure in 564 children ages 8 to 18 years admitted after surgery. A sample of 54 participants completed the YAPFAQ daily for 3 days after major surgery to assess responsiveness. The measure showed good reliability (Cronbach alpha = .96) and construct validity, with expected relationships with physical health-related quality of life ($r = -.53, P < .001$) and pain intensity ($r = .42, P < .001$). YAPFAQ scores decreased over time showing good responsiveness to expected recovery. A 3-item short form of the YAPFAQ showed promising psychometric properties. Early assessment of functioning after surgery may identify children at risk for poor functional outcomes and allow targeting of therapies to improve postsurgical recovery. PERSPECTIVE: The YAPFAQ showed promising psychometric properties in a pediatric postsurgical population. This study addresses a gap in tools available to monitor functional recovery during hospitalization after pediatric surgery. Early detection of problems with recovery may enable targeted therapies to improve postsurgical outcomes. https://www.ncbi.nlm.nih.gov/pubmed/28576670


BACKGROUND: Complete repair of pulmonary atresia (PA) ventricular septal defect (VSD) with hypoplastic or absent native pulmonary arteries, often with major aortopulmonary collateral arteries (MAPCAs), involves construction of an adequate sized pulmonary arterial tree. We report our results with a previously described staged strategy using initial right ventricle (RV)-to-reconstructed pulmonary arterial tree (RV-PA) connection to promote pulmonary arterial growth and facilitate later ventricular septation. METHODS: We retrospectively reviewed data for all patients (N = 10) with initial echocardiographic diagnosis of PA-VSD and hypoplastic pulmonary arteries operated in our center from October 2008 to August 2016. Pulmonary arterial vessel size measured on preoperative and postoperative angiography was used to calculate Nakata index. RESULTS: Seven patients had PA-VSD, three had virtual PA-VSD, and seven had MAPCAs. All underwent creation of RV-PA connection at a median age of 7.5 days and weight 3.6 kg. Eight patients had RV-PA conduits, two had a transannular patches, and seven had major pulmonary artery reconstruction simultaneously. There were no deaths or serious morbidity; one conduit required revision prior to complete repair. Complete repair with ventricular septation and RV pressure less than half systemic was achieved in all patients at a median age of 239 days. Nakata index in neonatal period was 54 mm²/m² (range 15-144 mm²/m²) and at time of septation 184 mm²/m² (range 56-510 mm²/m²); P = .004). Growth rates of right and left branch pulmonary arteries were similar. The 10 patients underwent 28 catheterizations with 13 interventions in 8 patients prior to full repair. CONCLUSION: Early palliative RV-PA connection promotes pulmonary arterial growth and facilitates eventual full repair with VSD closure with low RV pressure and operative risk. https://www.ncbi.nlm.nih.gov/pubmed/28901225


BACKGROUND/OBJECTIVES: The practice of palliative radiation therapy (RT) is based on extrapolation from adult literature. We evaluated patterns of pediatric palliative RT to describe regimens used to identify opportunity for
future pediatric-specific clinical trials. DESIGN/METHODS: Six international institutions with pediatric expertise completed a 122-item survey evaluating patterns of palliative RT for patients ≤21 years old from 2010 to 2015. Two institutions use proton RT. Palliative RT was defined as treatment with the goal of symptom control or prevention of immediate life-threatening progression. RESULTS: Of 3,225 pediatric patients, 365 (11%) were treated with palliative intent to a total of 427 disease sites. Anesthesia was required in 10% of patients. Treatment was delivered to metastatic disease in 54% of patients. Histologies included neuroblastoma (30%), osteosarcoma (18%), leukemia/lymphoma (12%), rhabdomyosarcoma (12%), medulloblastoma/ependymoma (12%), Ewing sarcoma (8%), and other (8%). Indications included pain (43%), intracranial symptoms (23%), respiratory compromise (14%), cord compression (8%), and abdominal distention (6%). Sites included nonspine bone (35%), brain (16% primary tumors, 6% metastases), abdomen/pelvis (15%), spine (12%), head/neck (9%), and lung/mediastinum (5%). Re-irradiation comprised 16% of cases. Techniques employed three-dimensional conformal RT (41%), intensity-modulated RT (23%), conventional RT (26%), stereotactic body RT (6%), protons (1%), electrons (1%), and other (2%). The most common physician-reported barrier to consideration of palliative RT was the concern about treatment toxicity (83%). CONCLUSION: There is significant diversity of practice in pediatric palliative RT. Combined with ongoing research characterizing treatment response and toxicity, these data will inform the design of forthcoming clinical trials to establish effective regimens and minimize treatment toxicity for this patient population.


OBJECTIVES: The aim of this study was to analyze changes in health care utilization and cost among a sample of highly impaired children and adolescents who sought a 3-week intensive interdisciplinary pain treatment (IIPT). MATERIALS AND METHODS: Claims data from 7 statutory health insurance companies were analyzed for 65 children and adolescents who sought IIPT at the German Paediatric Pain Centre. The annual health care utilization and cost were determined for the following 4 areas: outpatient care, inpatient care, medications, and remedies and aids. We analyzed the changes in resource utilization in the year before (pre_1 y) IIPT and in the subsequent year (post_1 y). RESULTS: Within the first year after IIPT, overall health care costs did not decrease significantly. However, the pattern of health care utilization changed. First, significantly more children and adolescents started outpatient psychotherapy (P=0.001). Second, the number of hospitalized children decreased significantly from 1-year pre to 1-year post (P=0.001). Accordingly, there were significantly fewer hospitalizations for primary chronic pain disorders at 1-year post (P<0.001). The prescription of nonopioids, co-analgesics and opioids was significantly reduced from 1-year pre to 1-year post (all P<0.013). DISCUSSION: The present results indicate that the health care costs of children and adolescents with severe chronic pain disorders do not significantly decrease 1 year after IIPT; however, the treatment becomes more goal-focused. Differential diagnosis measures and nonindicated therapeutic interventions decreased, and more indicated interventions, such as psychotherapy, were used. Future research is needed to investigate the economic long-term changes after IIPT.


BACKGROUND: Non-invasive ventilation (NIV) significantly changed the management of respiratory distress syndrome (RDS) in preterm infants. Further perspectives for neonatologists regard the assessment of different NIV strategies in terms of availability, effectiveness, and failure. OBJECTIVE: The aim of the present study is to evaluate the effectiveness of three different NIV strategies: nasal continuous positive airway pressure (N-CAPAP), nasal synchronized intermittent positive pressure ventilation (N-SIPPV), and nasal bilevel-CAPAP (BiPAP), as first intention treatment for RDS in very low birth-weight infants (VLBW). METHODS: A multicenter retrospective study was conducted in three neonatal intensive care unit (NICUs) that enrolled 191 VLBW infants complicated by RDS, who received, as first intention treatment for RDS, three different NIV approaches (N-CAPAP: n = 66; N-SIPPV: n = 62, BiPAP: n = 63). We evaluated the performance of different NIV strategies by primary (failure within the first 5 d of life) and some selected secondary end-points. RESULTS: The incidence of NIV failure was significantly higher in the N-CAPAP group (22/66) versus N-SIPPV/BiPAP groups (11/62; 11/63) (p < .05 for both), while no difference was observed between N-SIPPV and BiPAP groups. Moreover, no differences were found between the three groups regarding secondary outcomes. CONCLUSIONS: The present study shows that first intention N-
SIPPPV/BiPAP, as NIV support, augment the beneficial effects of N-CPAP contributing to a reduced risk of failure in VLBW infants complicated by RDS. Data open up to further RCTs on a wider population to evaluate NIV effectiveness on long-term outcomes.


The purpose of this study was to clarify ownership and usage of mobile phones among young patients with brain tumors in Japan. The subjects of this study were patients with brain tumors diagnosed between 2006 and 2010 who were between the ages of 6 and 18 years. The target population for the analysis was 82 patients. Patients were divided into two groups: 16 patients who were mobile phone owners 1 year before diagnosis, and 66 patients who did not own mobile phones (non-owners). Using data on the mobile phone ownership rate obtained from three general-population surveys, we calculated the expected number of mobile phone owners. The three age-adjusted standardized ownership ratios were 0.83 (95% confidence interval [CI]: 0.56-1.22), 0.51 (95% CI: 0.24-1.04), and 0.75 (95% CI: 0.42-1.32). The mobile phone ownership prevalence among the young Japanese patients with brain tumors in the current study does not differ from available estimates for the general population of corresponding age. However, since the use of mobile phones among children is increasing annually, investigations into the health effects of mobile phone use among children should continue. Bioelectromagnetics. 38:349-355, 2017. (c) 2017 Wiley Periodicals, Inc. https://www.ncbi.nlm.nih.gov/pubmed/28342194


Pediatric Risk of Mortality Score (PRISM III-12) is a physiology-based predictor for risk of mortality. We conducted prospective study from January 1, 2014 to 2015 in pediatric oncology intensive care unit (POICU) at South Egypt Cancer Institute, Egypt to explore the ability of 1st PRISM III-12 to predict the risk of mortality in critically ill cancer patients and the ability of serial PRISM III measured every 72 hours to follow-up the patients’ clinical condition during POICU stay. In total, 123 (78 males) children were included. Median age was 5 years (1 to 15 y). Death rate was 20%. 1st PRISM III-12 mean was 19 (0 to 61). The mean 1st PRISM III-12 for survivors was significantly higher compared with nonsurvivors (15 vs. 37 respectively; P<0.001). 1st PRISM III-12 mean was significantly correlated to the reasons for admission and organ failures' number (P<0.001 and <0.001). 1st PRISM III-12 correlated weakly positive with the length of stay (r=0.2; P=0.024). Receiver operator curve for 1st PRISM III-12 was 0.913 (95% confidence interval, 0.85-0.98; P<0.001). Decline in serial PRISM III was significantly correlated with favorable (survivor) outcome (P<0.001). We concluded that PRISM III-12 can be used effectively in predicting the risk of mortality and following the clinical condition of patients during POICU stay. https://www.ncbi.nlm.nih.gov/pubmed/29176465


Anthracycline-induced cardiotoxicity remains a significant contributor to late morbidity/mortality in children and young adults with acute myeloid leukemia (AML). The cardioprotectant dexrazoxane can be used as prophylaxis to diminish risk for cardiomyopathy but whether it affects risk of relapse in pediatric AML is unclear. Our institution adopted the use of dexrazoxane before anthracyclines administration for all oncology patients in 2011. We compared patients with AML (ages, 0 to 21 y) who received or did not receive dexrazoxane during the years 2008 to 2013. In total, 44 patients with AML (ages, 4.5 mo to 21.7 y) were included. We identified no statistical difference in 2-year event rate (62% vs. 50%, P=0.41) or 2-year overall survival (69% vs. 69%, P=0.53) between patients receiving (n=28) or not receiving (n=16) dexrazoxane. Ejection fraction (P=0.0262) and shortening fraction (P=0.0381) trended significantly higher in patients that received dexrazoxane compared with those that did not receive dexrazoxane. Utilization of the cardioprotectant dexrazoxane before anthracycline chemotherapy in pediatric patients with AML demonstrated no significant difference in either event rate or overall survival.
relative to institutional controls and seems to improve cardiac function indices. Further studies in this patient population are needed to confirm these findings.


BACKGROUND: Low case volume has been associated with lower survival after pediatric lung transplantation. Our aim was to analyze waitlist outcomes among pediatric lung transplant centers in the USA. METHODS: We studied a cohort of 1,139 pediatric candidates listed in the Organ Procurement and Transplantation Network for lung transplantation between 2002 and 2014. Of these candidates, 720 (63.2%) received a transplant. Candidates were divided into groups according to the clinical activity of the center of listing: high-volume pediatric (>4 transplants per year); low-volume pediatric (<4 transplants per year); and adult (transplant volume predominantly in adults). We used multivariate Cox regression analysis to identify independent risk factors for waitlist mortality. We also determined the transplant rate and likelihood of transplant after listing over the study period. RESULTS: Fifty-eight percent of the children and adolescents were listed in adult centers where the resultant transplant rate was low-only 42% received a transplant compared with 93% in pediatric programs. Listing in an adult program was also the most significant risk factor for death on the waiting list (hazard ratio 15.6, 95% confidence interval 5.8 to 42.1). CONCLUSIONS: Most children (58%) are listed for lung transplantation in adult centers and have a reduced rate of transplantation and a greater chance of waitlist mortality.


The objective of the study was to determine predictors of survival among HIV-positive children (<15 years) in Swaziland. A retrospective cohort analysis of medical records for 4 167 children living with HIV who were initiated on antiretroviral therapy (ART) between 2004 and 2008, and followed up until 2014 was conducted in clinical settings at 36 health facilities. The Kaplan Meier Estimator, signed-ranks test, and the Cox proportional hazards regression model were applied to determine survival probabilities, significant difference among stratified survival functions and adjusted hazard ratios respectively. The results reveal that the median survival time for children was 78 months (95% CI: 77-79). Children who were initiated early on ART had higher survival probability over time (HR: 0.35 [95% CI: 0.21-0.57], p < 0.001) compared to those whose ART initiation was delayed. Children within the age group of <1 years had higher hazard (HR = 1.55 [95% CI: 1.16-2.08], p < 0.001) of death than children within the age group of 1-14 years. Children who were nourished had 88% lower hazard of death (HR: 0.12 [95% CI: 0.07-0.19], p < 0.001) than severely malnourished children. The study demonstrates that ART paediatric services are effective in increasing survival among HIV infected children and early initiated children have high survival probability. Active tuberculosis (TB), malnutrition, and delayed ART initiation remain predictors of poor survival among children living with HIV.


Importance: Pneumonia is a leading cause of morbidity and mortality in children. It is important to identify the clinical symptoms and physical examination findings associated with pneumonia to improve timely diagnosis, prevent significant morbidity, and limit antibiotic overuse. Objective: To systematically review the accuracy of symptoms and physical examination findings in identifying children with radiographic pneumonia. Data Sources and Study Selection: MEDLINE and Embase (1956 to May 2017) were searched, along with reference lists from retrieved articles, to identify diagnostic studies of pediatric pneumonia across a broad age range that had to include children younger than age 5 years (although some studies enrolled children up to age 19 years); 3644 unique articles were identified, of which 23 met inclusion criteria. Data Extraction and Synthesis: Two authors independently abstracted raw data and assessed methodological quality. A third author resolved disputes. Main Outcomes and Measures: Likelihood ratios (LRs), sensitivity, and specificity were calculated for individual symptoms and physical examination findings for the diagnosis of pneumonia. An infiltrate on chest radiograph was considered the reference standard for the diagnosis of pneumonia. Results: Twenty-three prospective cohort studies of children (N = 13833) with possible pneumonia were included (8 from North America), with a range of
78 to 2829 patients per study. The prevalence of radiographic pneumonia in North American studies was 19% (95% CI, 11%-31%) and 37% (95% CI, 26%-50%) outside of North America. No single symptom was strongly associated with pneumonia; however, the presence of chest pain in 2 studies that included adolescents was associated with pneumonia (LR, 1.5-5.5; sensitivity, 8%-14%; specificity, 94%-97%). Vital sign abnormalities such as fever (temperature >37.5 degrees C [LR range, 1.7-1.8]; sensitivity, 80%-92%; specificity, 47%-54%) and tachypnea (respiratory rate >40 breaths/min; LR, 1.5 [95% CI, 1.3-1.7]; sensitivity, 79%; specificity, 51%) were not strongly associated with pneumonia diagnosis. Similarly, auscultatory findings were not associated with pneumonia diagnosis. The presence of moderate hypoxemia (oxygen saturation <96%); LR, 2.8 [95% CI, 2.1-3.6]; sensitivity, 64%; specificity, 77%) and increased work of breathing (grunting, flaring, and retractions; positive LR, 2.1 [95% CI, 1.6-2.7]) were signs most associated with pneumonia. The presence of normal oxygenation (oxygen saturation >96%) decreased the likelihood of pneumonia (LR, 0.47 [95% CI, 0.32-0.67]). Conclusions and Relevance: Although no single finding reliably differentiates pneumonia from other causes of childhood respiratory illness, hypoxia and increased work of breathing are more important than tachypnea and auscultatory findings.


Importance: Hypothermia for 72 hours at 33.5 degrees C for neonatal hypoxic-ischemic encephalopathy reduces death or disability, but rates continue to be high. Objective: To determine if cooling for 120 hours or to a temperature of 32.0 degrees C reduces death or disability at age 18 months in infants with hypoxic-ischemic encephalopathy. Design, Setting, and Participants: Randomized 2 x 2 factorial clinical trial in neonates (>=36 weeks’ gestation) with hypoxic-ischemic encephalopathy at 18 US centers in the Eunice Kennedy Shriver National Institute of Child Health and Human Development Neonatal Research Network between October 2010 and January 2016. Interventions: A total of 364 neonates were randomly assigned to 4 hypothermia groups: 33.5 degrees C for 72 hours (n = 95), 32.0 degrees C for 72 hours (n = 90), 33.5 degrees C for 120 hours (n = 96), or 32.0 degrees C for 120 hours (n = 83). Main Outcomes and Measures: The primary outcome was death or moderate or severe disability at 18 to 22 months of age adjusted for center and level of encephalopathy. Severe disability included any of Bayley Scales of Infant Development III cognitive score less than 70, Gross Motor Function Classification System (GMFCS) level of 3 to 5, or blindness or hearing loss despite amplification. Moderate disability was defined as a cognitive score of 70 to 84 and either GMFCS level 2, active seizures, or hearing with amplification. Results: The trial was stopped for safety and futility in November 2013 after 364 of the planned 726 infants were enrolled. Among 347 infants (95%) with primary outcome data (mean age at follow-up, 20.7 [SD, 3.5] months; 42% female), death or disability occurred in 56 of 176 (31.8%) cooled for 72 hours and 54 of 171 (31.6%) cooled for 120 hours (adjusted risk ratio, 0.92 [95% CI, 0.68-1.25]; adjusted absolute risk difference, -1.0% [95% CI, -10.2% to 8.1%]) and in 59 of 185 (31.9%) cooled to 33.5 degrees C and 51 of 162 (31.5%) cooled to 32.0 degrees C (adjusted risk ratio, 0.92 [95% CI, 0.68-1.26]; adjusted absolute risk difference, -3.1% [95% CI, -12.3% to 6.1%]). A significant interaction between longer and deeper cooling was observed (P = .048), with primary outcome rates of 29.3% at 33.5 degrees C for 72 hours, 34.5% at 32.0 degrees C for 72 hours, 34.4% at 33.5 degrees C for 120 hours, and 28.2% at 32.0 degrees C for 120 hours. Conclusions and Relevance: Among term neonates with moderate or severe hypoxic-ischemic encephalopathy, cooling for longer than 72 hours, cooling to lower than 33.5 degrees C, or both did not reduce death or moderate or severe disability at 18 months of age. However, the trial may be underpowered, and an interaction was found between longer and deeper cooling. These results support the current regimen of cooling for 72 hours at 33.5 degrees C. Trial Registration: clinicaltrials.gov Identifier: NCT01192776.


OBJECTIVES: Neurologic and functional morbidity occurs in ~30% of PICU survivors, and young children may be at particular risk. Bronchiolitis is a common indication for PICU admission among children less than 2 years old.
Two single-center studies suggest that greater than 10-25% of critical bronchiolitis survivors have neurologic and functional morbidity but those estimates are 20 years old. We aimed to estimate the burden of neurologic and functional morbidity among more recent bronchiolitis patients using two large, multicenter databases. DESIGN: Analysis of the Pediatric Health Information System and the Virtual Pediatric databases. SETTING: Forty-eight U.S. children’s hospitals (Pediatric Health Information System) and 40 international (mostly United States) children’s hospitals (Virtual Pediatric Systems). PATIENTS: Previously healthy PICU patients less than 2 years old admitted with bronchiolitis between 2009 and 2015 who survived and did not require extracorporeal membrane oxygenation or cardiopulmonary resuscitation. INTERVENTIONS: None. Neurologic and functional morbidity was defined as a Pediatric Overall Performance Category greater than 1 at PICU discharge (Virtual Pediatric Systems subjects), or a subsequent hospital encounter involving developmental delay, feeding tubes, MRI of the brain, neurologist evaluation, or rehabilitation services (Pediatric Health Information System subjects). MEASUREMENTS AND MAIN RESULTS: Among 3,751 Virtual Pediatric Systems subjects and 9,516 Pediatric Health Information System subjects, ~20% of patients received mechanical ventilation. Evidence of neurologic and functional morbidity was present at PICU discharge in 707 Virtual Pediatric Systems subjects (18.6%) and more chronically in 1,104 Pediatric Health Information System subjects (11.6%). In both cohorts, neurologic and functional morbidity was more common in subjects receiving mechanical ventilation (27.5% vs 16.5% in Virtual Pediatric Systems; 14.5% vs 11.1% in Pediatric Health Information System; both p < 0.001). In multivariate models also including demographics, use of mechanical ventilation was the only variable that was associated with increased neurologic and functional morbidity in both cohorts. CONCLUSIONS: In two large, multicenter databases, neurologic and functional morbidity was common among previously healthy children admitted to the PICU with bronchiolitis. Prospective studies are needed to measure neurologic and functional outcomes using more precise metrics. Identification of modifiable risk factors may subsequently lead to improved outcomes from this common PICU condition.


BACKGROUND: Although central venous catheters (CVCs) are essential to pediatric cancer care, complications are common (e.g., occlusion, central line-associated bloodstream infection [CLABSI]). Parenteral nutrition (PN) and external CVCs are associated with an increased complication risk, but their interaction is unknown. METHODS: A retrospective matched cohort study of pediatric oncology patients who received PN through subcutaneous ports or external CVCs. Complication rates were compared between CVC types during PN and non-PN periods (log-negative binomial model). RESULTS: Risk of CLABSI was higher during PN for children with ports (relative risk [RR] = 39.6; 95% confidence interval, 5.0-309) or external CVCs (RR = 2.9; 95% confidence interval, 1.1-7.4). This increased risk during PN was greater for ports than for external CVCs (ratio of relative risks = 13.6). Occlusion risk was higher during PN in both groups (RR = 10.0 for ports; RR = 2.0 for external CVCs), and the increase was significantly greater in ports (ratio of relative risks, 4.9). Overall, complication rates for ports were much lower than for external CVCs during the non-PN period but similar during the PN period. CONCLUSION: Children with cancer who receive PN have increased risk of CLABSI and occlusion. The risk increase is greatest in children with ports: a 40- and 10-fold increase in infection risk and occlusion, respectively, resulting in similar complication rates during PN regardless of CVC type and negating the usual benefits of ports. Children with cancer who will require PN should have primary insertion of external CVCs where possible.


BACKGROUND: There are few studies assessing the birth measures of patients with congenital heart disease (CHD). Our aim to evaluate their progression and impact over the outcome. METHODS: The cases consisted of patients with CHD during their first hospitalization in a reference cardiac and pediatric intensive care unit (ICU) from Southern Brazil. Controls were composed of patients with no clinical evidence of CHD hospitalized soon after cases. The cases underwent high-resolution karyotype and fluorescence in situ hybridization (FISH) for 22q11 microdeletion. We analyzed birth weight, length and head circumference of patients of both groups. For CHD patients, we evaluated their progression and impact until hospitalization at ICU. RESULTS: Our sample was
INTRODUCTION: Pediatric cardiac arrest is an uncommon but critical life-threatening event requiring effective cardiopulmonary resuscitation. High-quality cardio-pulmonary resuscitation (CPR) is essential, but is poorly performed, even by highly skilled healthcare providers. The recently described two-thumb chest compression technique (nTTT) consists of the two thumbs directed at the angle of 90 degrees to the chest while having the fingers fist-clenched. This technique might facilitate adequate chest-compression depth, chest-compression rate and rate of full chest-pressure relief. METHODS: 42 paramedics from the national Emergency Medical Service of Poland performed three single-rescuer CPR sessions for 10 minutes each. Each session was randomly assigned to the conventional two-thumb (TTHT), the conventional two-finger (TFT) or the nTTT. The manikin used for this study was connected with an arterial blood pressure measurement device and blood measurements were documented on a 10-seconds cycle. RESULTS: The nTTT provided significant higher systolic (82 vs. 30 vs. 41 mmHg). A statistically significant difference was noticed between nTTT and TFT (p<.001), nTTT and TTHT (p<.001), TFT and TTHT (p=.003). The median diastolic pressure using nTTT was 16 mmHg compared with 9 mmHg for TFT (p<.001), and 9.5 mmHg for TTHT (p=.001). Mean arterial pressure using distinct methods varied and amounted to 40 vs. 22. vs. 26 mmHg (nTTT vs. TFT vs. TTHT, respectively). A statistically significant difference was noticed between nTTT and TFT (p<.001), nTTT and TTHT (p<.001), and TFT and TTHT (p<.001). The highest median pulse pressure was obtained by the nTTT 67.5 mmHg. Pulse pressure was 31.5 mmHg in the TTHT and 24 mmHg in the TFT. The difference between TFT and TTHT (p=.025), TFT and nTTT (p<.001), as well as between TTHT and nTTT (p<.001) were statistically significant. CONCLUSIONS: The new nTTT technique generated higher arterial blood pressures compared to established chest compression techniques using an infant manikin model, suggesting a more effective chest compression. Our results have important clinical implications as nTTT was simple to perform and could be widely taught to both healthcare professionals and bystanders. Whether this technique translates to improved outcomes over existing techniques needs further animal studies and subsequent human trials.

OBJECTIVE: Genetic surfactant dysfunction causes respiratory failure in term and near-term newborn infants, but little is known of such condition in prematures. We evaluated genetic surfactant dysfunction in premature newborn infants with severe RDS. PATIENTS AND METHODS: A total of 68 preterm newborn infants with gestational age <32 weeks affected by unusually severe RDS were analysed for mutations in SFTPB, SFTPC and ABCA3. Therapies included oxygen supplementation, nasal CPAP, different modalities of ventilatory support, administration of exogenous surfactant, inhaled nitric oxide and steroids. Molecular analyses were performed on genomic DNA extracted from peripheral blood and Sanger sequencing of whole gene coding regions and intron junctions. In one case histology and electron microscopy on lung tissue was performed. RESULTS: Heterozygous previously described rare or novel variants in surfactant proteins genes ABCA3, SFTPB and SFTPC were identified in 24 newborn infants. In total, 11 infants died at age of 2 to 6 months. Ultrastructural analysis of lung tissue of one infant showed features suggesting ABCA3 dysfunction. DISCUSSION: Rare or novel genetic variants in genes encoding surfactant proteins were identified in a large proportion (35%) of premature newborn infants with particularly severe RDS. We speculate that interaction of developmental immaturity of surfactant production in association with abnormalities of surfactant metabolism of genetic origin may have a synergic worsening phenotypic effect.


OBJECTIVE: To compare the neurodevelopmental outcomes at 18 to 21 months corrected age (CA) of infants born at <29 weeks that received room air, an intermediate oxygen concentration or 100% oxygen at the initiation of resuscitation. STUDY DESIGN: In this retrospective cohort study, we compared neonatal and neurodevelopmental outcomes at 18 to 21 months CA among inborn infants born before 29 weeks' gestation that received room air, intermediate oxygen concentration or 100% oxygen at the initiation of resuscitation. RESULTS: Of 1509 infants, 445 received room air, 483 received intermediate oxygen concentrations and 581 received 100% oxygen. Compared to infants that received room air, the primary outcome of death or neurodevelopmental impairment (NDI) was not different in intermediate oxygen (adjusted odds ratio (aOR) 1.01; 95% confidence interval (CI) 0.77, 1.34) or 100% oxygen (aOR 1.03; 95% CI 0.78, 1.35). Compared to room air, there was no difference in odds of death or severe NDI in intermediate oxygen (aOR 1.14; 95% CI 0.82, 1.58) or 100% oxygen group (aOR 1.22; 95% CI 0.90, 1.67). The odds of severe NDI among survivors were significantly higher in infants that received 100% oxygen as compared to room air (aOR 1.57, 95% CI 1.05, 2.35). CONCLUSIONS: We observed no significant difference in the primary composite outcomes of death or NDI and death or severe NDI at 18 to 21 months CA between infants that received room air, intermediate oxygen concentration or 100% oxygen at the initiation of resuscitation. However, use of 100% oxygen was associated with increased odds of severe NDI among survivors as compared to room air.


BACKGROUND: Interdisciplinary pain treatment has been shown to be effective for children and adolescents with chronic pain, both in an outpatient and inpatient setting. Until now, the effectiveness has been analyzed with various outcome measures. Although it has only rarely been used for adolescents so far, Chronic Pain Grading (CPG) developed by Von Korff could be an appropriate general outcome measure. OBJECTIVE: The study aims at prospectively investigating and comparing the therapy outcome one year after initial presentation for both outpatients and inpatients using the CPG. MATERIALS AND METHODS: Data of 258 adolescents were gathered at initial presentation and one year later and analyzed using the CPG. Changes from pretreatment to follow-up and predictors of good therapy outcome were investigated for the whole sample and separately for outpatients and inpatients. RESULTS: Compared to inpatients, outpatients were characterized by a lower CPG both before and one year after initial presentation. Large effects were found both for outpatient and inpatient therapy regarding the
improvement of the CPG. In outpatient therapy, boys were two times more likely to display therapy success. CONCLUSION: The study shows that the CPG is an appropriate outcome measure to display the long-term effectiveness of an inpatient and outpatient interdisciplinary pain treatment. The interdisciplinary pain treatment needs to be better tailored to girls to improve its effectiveness.


Diseases causing hematochezia range from benign to potentially life-threatening. Systematic pediatric data on the causes of hematochezia are scarce. We studied the underlying causes and long-term outcome of hematochezia in children. We further investigated the relevance of antibiotic-associated hemorrhagic colitis in children, especially if caused by Klebsiella oxytoca. Infants, children, and adolescents with hematochezia were recruited prospectively. Patients were grouped according to age (<1 year, 1-5 years, 6-13 years, >14 years). In addition to routine diagnostics, K oxytoca stool culture and toxin analysis was performed. We collected data on history, laboratory findings, microbiological diagnostic, imaging, final diagnosis, and long-term outcome. We included 221 patients (female 46%; age 0-19 years). In 98 (44%), hematochezia was caused by infectious diseases. Endoscopy was performed in 30 patients (13.6%). No patient died due to the underlying cause of hematochezia. The most common diagnoses according to age were food protein-induced protocollitis in infants, bacterial colitis in young children, and inflammatory bowel disease in children and adolescents. Seventeen (7.7%) had a positive stool culture for K oxytoca. Antibiotic-associated colitis was diagnosed in 12 (5%) patients: 2 caused by K oxytoca and 2 by Clostridium difficile; in the remaining 8 patients, no known pathobiont was identified. Infections were the most common cause of hematochezia in this study. In most patients, invasive diagnostic procedures were not necessary. Antibiotic-associated hemorrhagic colitis caused by K oxytoca was an uncommon diagnosis in our cohort. Antibiotic-associated colitis with hematochezia might be caused by pathobionts other than C difficile or K oxytoca.


Context: Tumors of the central nervous system (CNS) constitute the second most common pediatric cancers. Unlike leukemia, management of CNS tumors requires a good multidisciplinary team. Higher rates of treatment abandonment are documented in view of complexity of the treatment with long duration, involving neurosurgery, radiation, chemotherapy, and high cost of treatment. Morbidity associated with CNS tumors may be significant in terms of physical deficits as well as neuropsychological and neuroendocrine sequelae. Pediatric neurooncology is still at a very nascent stage in the developing countries. There are only a few reports on the multidisciplinary approach and outcomes of pediatric brain tumors in developing countries. Aims: The aim of this study is to identify the clinicopathological profile of Pediatric CNS tumors in a tertiary care center located in South India in comparison with reports from other low- and middle-income Countries. Settings and Design: A retrospective analysis of medical records of all children diagnosed with brain tumors from January 2012 to November 2016 at our institute was done. Subjects and Methods: A retrospective study of clinical, pathological profile, and outcomes of children <18 years diagnosed with brain tumors at our institute from January 2012 to November 2016 was done. Histopathological categorization was done as per the WHO classification 2007. The multidisciplinary treatment with respect to surgery, radiation, and chemotherapy was noted and the outcomes were recorded. Statistical Analysis Used: R for Statistical Computing (Version 3.0.2; 2013-09-25). Results: A total of 52 children were diagnosed with male preponderance of 66.6%. Highest incidence was noted in the age group of 0-4 years (50%). Majority of them were supratentorial (59.6%). CNS embryonal tumors contributed to 48% of all our brain tumors. 73% of them underwent either resection or biopsy. Eight (15.3%) of them died due to the progression of disease, but 44% abandoned treatment due to the progression/recurrence of disease. Those lost to follow-up were mostly among the high-risk groups with poor prognosis such as pontine glioma, medulloblastoma (high risk), and primitive neuroectodermal tumor. Conclusions: Although brain tumors constituted 30% of all our solid tumors, only 56% of them received appropriate treatment and 25% abandoned treatment. High rates of abandonment were a consequence of late diagnosis, complex multidisciplinary treatment
involved, high treatment cost, lack of uniformity in management between different oncology centers and poor prognosis of the tumor subtype.


BACKGROUND: Substantial variability exists among countries regarding the modes of death in pediatric intensive care units (PICUs). However, there is limited information on end-of-life care in Japanese PICUs. Thus, this study aimed to elucidate the characteristics of end-of-life care practice for children in a Japanese PICU. METHODS: We examined life-sustaining treatment (LST) status at the time of death based on medical chart reviews from 2010 to 2014. All deaths were classified into 3 groups: limitation of LST (limitation group, death after withholding or withdrawal of LST or a do not attempt resuscitation order), no limitation of LST (no-limitation group, death following failed resuscitation attempts), or brain death (brain death group). RESULTS: Of the 62 patients who died, 44 (71%) had limitation of LST, 18 (29%) had no limitation of LST, and none had brain death. In the limitation group, the length of PICU stay was longer than that in the no-limitation group (13.5 vs 2.5 days; P = .01). The median time to death after the decision to limit LST was 2 days (interquartile range: 1-5.5 days), and 94% of the patients were on mechanical ventilation at the time of death in the limitation group. CONCLUSIONS: Although limiting LST was a common practice in end-of-life care in a Japanese PICU, a severe limitation of LST such as withdrawal from the ventilator was hardly practiced, and a considerable LST was still provided at the time of death.


BACKGROUND: Maintenance of a patent airway while the neonate is on nasal continuous positive airway pressure (nasal CPAP) requires vigilant monitoring and oral/nasopharyngeal suctioning. Currently, no evidence-based guidelines for safe suctioning in neonates while on bubble nasal CPAP have been published. PURPOSE: (1) To characterize the clinical and behavioral responses of neonates on bubble nasal CPAP in a level III neonatal intensive care unit following routine oral and nasopharyngeal suctioning. METHODS: This pilot study has a 1-sample within-subject repeated-measures design in which neonates (N = 16) served as their own control. Data on a neonate’s physiological and behavior measures (heart rate, respiratory rate, oxygen saturation [SaO2], and Premature Infant Pain Profile [PIPP] score) were collected before, during, and after the completion of suctioning sequence. FINDING/RESULTS: A total of 16 neonates with a mean gestational age of 29.76 weeks and an average day of life of 3.4 were enrolled. The infant's heart rates did not differ significantly (P = .51) across the suctioning sequence. There were no statistical significant changes in the average respiratory rate across the suctioning sequences (P = .79). SaO2 demonstrated a drop between baseline and after each suctioning (P < .001). The PIPP score demonstrated a precipitous increase throughout the procedure (P < .001). On average, it took 9.5 seconds (SD = 4.9) to complete the suctioning sequences. IMPLICATIONS FOR PRACTICE: Our results suggest that the guideline tested is safe and tolerated by infants. IMPLICATIONS FOR RESEARCH: This guideline should be tested in a larger sample and with neonates on other nasal CPAP systems.

compared with the non-VAP ones. The VAP rate was 13.3% or 10.1 per 1000 ventilator days. According to the multivariate analysis, a birthweight less than 750 g [adjusted odds ratio (aOR)=10.75, 95% confidence interval (CI)=2.35-49.16; P=0.021] and gestational age less than 32 weeks [adjusted odds ratio (aOR)=2.90, 95% confidence interval (CI)=1.23-6.73; P=0.002]. INTERPRETATION: Adverse events after BoNT-A injections are common but mostly mild and self-limiting. Children in GMFCS levels IV and V are at increased risk of systemic adverse events. The relationship between CP severity and BoNT-A adverse events is complex and further research is required to better understand this relationship. WHAT THIS PAPER ADDS: Adverse events reported at the time of botulinum toxin A injection occurred in 6% of injection episodes. Adverse events were reported at follow-up in 22% of injection episodes. Children in Gross Motor Function Classification System (GMFCS) levels IV and V have increased rates of systemic adverse events. Children in GMFCS levels IV and V report less local weakness and pain.


**BACKGROUND:** Mucopolysaccharidosis type IIIb syndrome (also known as Sanfilippo type B syndrome) is a lysosomal storage disease resulting in progressive deterioration of cognitive acquisition after age 2-4 years. No treatment is available for the neurological manifestations of the disease. We sought to assess the safety and efficacy of a novel intracerebral gene therapy. **METHODS:** Local regulatory authorities in France allowed inclusion of up to four children in this phase 1/2 study. Treatment was 16 intraparenchymal deposits (four in the cerebellum) of a recombinant adenovassociated viral vector serotype 2/5 (rAAV2/5) encoding human alpha-N-acetylgalcosaminidase (NAGLU) plus immunosuppressive therapy. We assessed tolerance, neurocognitive progression, brain growth, NAGLU enzymatic activity in CSF, and specific anti-NAGLU immune response for 30 months after surgery. This trial is registered with EudraCT, number 2012-000856-33, and the International Standard Clinical Trial Registry, number ISRCTN19853672. **FINDINGS:** Of seven eligible children, the four youngest, from France (n=2), Italy (n=1), and Greece (n=1), aged 20, 26, 30, and 53 months, were included between February, 2012, and February, 2014. 125 adverse events were recorded, of which 117 were treatment emergent and included six classified as severe, but no suspected unexpected serious adverse drug reactions were seen. Vector genomes were detected in blood for 2 days after surgery. Compared with the natural history of mucopolysaccharidosis type III syndromes, neurocognitive progression was improved in all patients, with the youngest patient having function close to that in healthy children. Decrease in developmental quotient was -11.0 points in patient one, -23.0 in patient two, -29.0 in patient three, and -17.0 in patient four, compared with -37.7 in the natural history of the disease. NAGLU activity was detected in lumbar CSF and was 15-20% of that in unaffected children. Circulating T lymphocytes that proliferated and produced tumour necrosis factor alpha upon ex-vivo exposure to NAGLU antigens were detectable at 1-12 months and 3-12 months, respectively, but not at 30 months in three of four patients. **INTERPRETATION:** Intracerebral rAAV2/5 was well tolerated and induced sustained enzyme production in the brain. The initial specific anti-NAGLU immune response that later subsided suggested acquired immunological tolerance. The best results being obtained in the youngest patient implies a potential window of opportunity. Longer follow-up is needed to further assess safety outcomes and persistence of improved cognitive development. **FUNDING:** Association Francaise Contre les Myopathies, Vaincre les Maladies Lysosomales, Institut Pasteur, and UniQure.


**BACKGROUND:** Ventilator-associated pneumonia (VAP) in neonates has been associated with high mortality and poor outcome. This study aimed to compare the incidence, risk factors, and outcomes of VAP and non-VAP conditions in neonates. **METHODS:** We performed a prospective cohort study in a neonatal intensive care unit (NICU) in Thailand from January 2014 to December 2014. All neonatal patients who were ventilated more than 48 hours were enrolled. **RESULTS:** There were 128 enrolled patients. The median (inter quartile range) gestational age and birthweight were 35 (30.2, 37.8) weeks and 2380 (1323.8, 3020.0) g. There were 17 VAP patients (19 episodes) and 111 non-VAP ones. The VAP rate was 13.3% or 10.1 per 1000 ventilator days. According to the multivariate analysis, a birthweight less than 750 g [adjusted odds ratio (aOR)=10.75, 95% confidence interval (CI)=2.35-49.16; P=0.002] and sedative medication use [aOR=4.00, 95% CI=1.23-12.50; P=0.021] were independent risk factors for VAP. Compared with the non-VAP group, the median difference in the VAP group yielded a significantly longer
duration of NICU stay (18 days, P=0.001), total length of hospital stay (16 days, P=0.002) and higher hospital costs ($5113, P=0.001). The in-hospital mortality rate in the VAP and non-VAP groups was 17.6% and 15.3% (P=0.73), respectively. CONCLUSIONS: A neonatal birthweight less than 750 g and sedative medication use were independent risk factors for VAP. Our VAP patients experienced a longer duration of both NICU and hospital stay, and incurred higher hospitalization costs. https://www.ncbi.nlm.nih.gov/pubmed/28120236


BACKGROUND: Tolerating higher partial pressures of carbon dioxide (PCO2) in mechanically ventilated extremely low birthweight infants to reduce ventilator-induced lung injury may have long-term neurodevelopmental side effects. This study analyses the results of neurodevelopmental follow-up of infants enrolled in a randomised multicentre trial. METHODS: Infants (n=359) between 400 and 1000 g birth weight and 23 0/7-28 6/7 weeks gestational age who required endotracheal intubation and mechanical ventilation within 24 hours of birth were randomly assigned to high PCO2 or to a control group with mildly elevated PCO2 targets. Neurodevelopmental follow-up examinations were available for 85% of enrolled infants using the Bayley Scales of Infant Development II, the Gross Motor Function Classification System (GMFCS) and the Child Development Inventory (CDI). RESULTS: There were no differences in body weight, length and head circumference between the two PCO2 target groups. Median Mental Developmental Index (MDI) values were 82 (60-96, high target) and 84 (58-96, p=0.79). Psychomotor Developmental Index (PDI) values were 84 (57-100) and 84 (65-96, p=0.73), respectively. Moreover, there was no difference in the number of infants with MDI or PDI <70 or <85 and the number of infants with a combined outcome of death or MDI <70 and death or PDI <70. No differences were found between results for GMFCS and CDI. The risk factors for MDI <70 or PDI <70 were intracranial haemorrhage, bronchopulmonary dysplasia, periventricular leukomalacia, necrotising enterocolitis and hydrocortisone treatment. CONCLUSIONS: A higher PCO2 target did not influence neurodevelopmental outcomes in mechanically ventilated extremely preterm infants. Adjusting PCO2 targets to optimise short-term outcomes is a safe option. TRIAL REGISTRATION NUMBER: ISRCTN56143743. https://www.ncbi.nlm.nih.gov/pubmed/28087725


BACKGROUND: Injury severity induces a proportionate acute metabolic stress response, associated with increased risk of hyperglycemia. We hypothesized that excess caloric delivery (overfeeding) during high stress states would increase hyperglycemia and disrupt response homeostasis. METHODS: Gestational age, daily weight, total daily caloric intake, serum C-reactive protein (CRP), prealbumin, and blood glucose concentrations in all acutely injured premature NICU infants requiring TPN over the past 3 years were reviewed. Injury severity was based on CRP and patients were divided into high (CRP >/=50mg/L) versus low (CRP <50mg/L) stress groups. Glycemic variability was used to measure disruption of homeostasis. RESULTS: Overall sample included N=563 patient days (37 patients; 42 episodes). High stress group pre-albumin levels negatively correlated with CRP levels (R=-0.62, p<0.005). A test of equal variance demonstrated significantly increased high stress glycemic variability (Haratio>1, Pr(F>F)=0.0353). When high stress patients were separated into high caloric intake (>/=70kg/kcal/day) versus low caloric intake (<70kg/kcal/day), maximum serum glucose levels were significantly higher with overfeeding (230.33+/−55.81 vs. 135.71+/−37.97mg/dL, p<0.004). CONCLUSION: Higher injury severity increases induced disruption of response homeostasis in critically ill neonates. TPN-associated overfeeding worsens injury-related hyperglycemia in more severely injured infants. TYPE OF STUDY: Retrospective study. LEVEL OF EVIDENCE: Level II. https://www.ncbi.nlm.nih.gov/pubmed/29550034

OBJECTIVE: The objective is to evaluate the association between antibiotic utilization and neurodevelopmental outcomes at 18 to 21 months’ corrected age among extremely low gestational age neonates without culture-proven sepsis or necrotizing enterocolitis (NEC). STUDY DESIGN: We conducted a retrospective cohort study of infants born between April 2009 and September 2011 at <29 weeks’ gestation and admitted to the neonatal intensive care units contributing data to the Canadian Neonatal Network. Multivariable analysis was performed to examine the primary composite outcome of death or significant neurodevelopmental impairment (sNDI) in infants with various antibiotic utilization rates (AURs). RESULT: There were 1,373 infants who fulfilled our inclusion criteria. Compared with infants in the lowest AUR quartile (Q1), those in the highest quartile (Q4) had higher odds of death or sNDI (adjusted odds ratio [AOR] = 7.44; 95% confidence interval [CI]: 4.55, 12.2) and death (AOR = 39.3; 95% CI: 16.1, 95.9). CONCLUSION: Our results indicate an association between high AUR and a composite outcome of death or adverse neurodevelopmental outcomes at 18 to 21 months’ corrected age.


INTRODUCTION: Availability of objective criteria for predicting successful extubation could avoid unnecessary prolongation of mechanical ventilation and/or inadvertent premature extubation, but the predictors of successful extubation in children are unclear. This study was performed to detect and validate respiratory function predictors of successful extubation in children admitted to the pediatric critical care unit. METHODS: A retrospective chart review from 2010 to 2012 identified 463 patients, who were divided into a derivation cohort (n = 294) and a validation cohort (n = 169). RESULTS: The incidence rate of failed extubation was 5% and 9% in the derivation and validation cohorts, respectively. The optimal cut-off values of crying vital capacity (CVC), peak inspiratory flow rate (PIFR), and maximum inspiratory pressure (MIP) were 17 ml/kg, 3.5 ml/sec/cm, and 50 cmH2O, respectively. The pass rates of CVC, PIFR, and MIP were 54.2%, 92.7%, and 55.5%, respectively. In the validation cohort, the successful extubation rate was 97.9% for patients who passed all 3 respiratory tests, 88.8% for those who passed at least one test, and 66.7% for those who failed all of the tests. Extubation failed in 5 patients who passed all three respiratory tests and failure was due to postoperative respiratory muscle fatigue or upper airway impairment. CONCLUSIONS: We detected and validated predictors of successful extubation in critically ill children. A combination of CVC, PIFR, and MIP may be used to predict successful extubation for critically ill children. It is necessary to pay attention when extubating patients with postoperative respiratory muscle fatigue or upper airway impairment due to disturbance of consciousness and/or glottal edema even if they pass the respiratory function tests.


OBJECTIVE: The study’s objective was to summarize the psychometric evaluation of self-report symptom instruments used in children with cancer younger than 8 years of age. METHODS: We conducted electronic searches of Ovid Medline, EMBASE, PsycInfo, Science Citation, Social Science Citation (Web of Science), and CINAHL. We included studies of children with cancer in which their self-report symptoms had been quantified and in which results were described for those younger than 8 years of age. The search was restricted to publications in English. Two reviewers screened studies and abstracted all data in duplicate. Descriptive analysis of reliability and validity was performed. RESULTS: Thirteen studies were included. Only one study recruited children <8 years alone. Most studies described reliability and validity in a wider age range cohort in which most children were older than 8 years of age. Of the eight studies that evaluated reliability within the younger age group, six raised concerns about poor internal consistency with Cronbach’s alpha <0.7 in at least one dimension. Concerns about test re-test reliability and inter-rater reliability were also observed. None of the studies evaluated validity. CONCLUSIONS: We failed to demonstrate that currently available instruments to measure self-report symptoms are reliable or valid specifically for children with cancer younger than 8 years of age. Development of psychometrically robust instruments for younger children should be a priority.


BACKGROUND: Data are scarce on the long-term clinical outcomes of perinatally HIV-infected children and adolescents receiving antiretroviral therapy (ART) in low/middle-income countries. We assessed the incidence of mortality before (early) and after (late) 6 months of ART and of the composite outcome of new/recurrent AIDS-defining event or death >6 months after ART start (late AIDS/death) and their associated factors. METHODS: Study population was perinatally HIV-infected children (<18 years) initiating ART within the Program for HIV Prevention and Treatment observational cohort (NCT00433030). Factors associated with late AIDS/death were assessed using competing risk regression models accounting for lost to-follow-up and included baseline and time-updated variables. RESULTS: Among 619 children, “early” mortality incidence was 99 deaths per 1000 person-years of follow-up [95% confidence interval (CI): 69 to 142] and “late” mortality 6 per 1000 person-years of follow-up (95% CI 4 to 9). Of the 553 children alive >6 months after ART initiation, median age at ART initiation was 6.4 years, CD4% 8.2%, and HIV-RNA load 5.1 log10 copies/mL. Thirty-eight (7%) children developed late AIDS/death after median time of 3.3 years: 24 died and 24 experienced new/recurrent AIDS-defining events (10 subsequently died). Factors independently associated with late AIDS/death were current age ≥13 years (adjusted subdistribution hazard ratio 4.9; 95% CI: 2.4 to 10.1), HIV-RNA load always ≥400 copies/mL (12.3; 95% CI: 4.0 to 37.6), BMI-z-score always < -2 SD (13.7; 95% CI: 3.4 to 55.7), and hemoglobin < 8 g/dL at least once (4.6; 95% CI: 2.0 to 10.5). CONCLUSIONS: After the initial 6 months of ART, being an adolescent, persistent viremia, poor nutritional status, and severe anemia were associated with poor clinical outcomes. This supports the need for novel interventions that target children, particularly adolescents with poor growth and uncontrolled viremia.


OBJECTIVES: Acute leukemia often causes osteoarthralgia. The aim of this study is characterization of leukemia-associated osteoarthralgia in comparison with juvenile idiopathic arthritis (JIA). METHODS: We retrospectively reviewed clinical records of 31 patients with acute leukemia and 13 patients with articular JIA diagnosed between January 2008 and March 2013. Clinical and laboratory findings at the initial examination were compared among the three groups; 10 leukemia with and 21 leukemia without osteoarthralgia and 13 JIA groups. RESULTS: Eleven of the 31 leukemic patients (35%) had osteoarthralgia before the diagnosis of leukemia. Peripheral leukemic cells were initially absent in 10 of the 31 leukemia patients including three with osteoarthralgia. Platelet counts over 300 x 10⁹/L were common in JIA, but not in osteoarthralgia group. Mean serum lactate dehydrogenase levels were higher in both of the leukemia groups than JIA group but often within normal or near-normal levels in the leukemia groups. Magnetic resonance imaging was examined in three leukemic patients and demonstrated osteomyelitis-like bone marrow edema in two and periarticular infiltration similar to synovitis in one patient. Three leukemic patients with osteoarthralgia showed partial and transient responses to antibiotic therapy. CONCLUSIONS: Leukemia-associated osteoarthralgia is often indistinguishable from rheumatic diseases by imaging and laboratory findings and should be confirmed by bone marrow examination.


STUDY OBJECTIVE: The Verbal Numerical Rating Scale is the most commonly used self-report measure of pain intensity. It is unclear how the validity and reliability of the scale scores vary across children's ages. We aimed to determine the validity and reliability of the scale for children presenting to the emergency department across a comprehensive spectrum of age. METHODS: This was a cross-sectional study of children aged 4 to 17 years. Children self-reported their pain intensity, using the Verbal Numerical Rating Scale and Faces Pain Scale-Revised at 2 serial assessments. We evaluated convergent validity (strong validity defined as correlation coefficient ≥/0.60), agreement (difference between concurrent Verbal Numerical Rating Scale and Faces Pain Scale-Revised scores), known-groups validity (difference in score between children with painful versus nonpainful conditions), responsivity (decrease in score after analgesic administration), and reliability (test-retest at 2 serial assessments) in the total sample and subgroups based on age. RESULTS: We enrolled 760 children; 27 did not understand the Verbal Numerical Rating Scale and were removed. Of the remainder, Pearson correlations were strong to very

Health-related quality of life (HRQOL) is an essential measure to consider when evaluating the full impact of illness in children diagnosed with leukemia. The purpose of the current study was to assess the overall HRQOL and specific functioning subscales of Nepalese children with leukemia using Pediatric Quality of Life Inventory 4.0 Generic Core Scale (PedsQL 4.0), compare self-report with parent proxy report of HRQOL and to identify the determinants affecting HRQOL. After cultural linguistic validation of PedsQL, a descriptive cross-sectional study was conducted on 43 children with leukemia and their parents in B. P. Koirala Memorial Cancer Hospital, Bharatpur via interview schedule. Among the subscales of HRQOL both the child’s self-report and parent proxy report scores were highest in social functioning and lowest in emotional functioning subscale. Intraclass correlation coefficient between proxy reports and self-reports were highest (0.828) in physical functioning and lowest (0.493) in social functioning subscales. Age group was significantly associated with the total score, physical functioning subscale, and emotional functioning subscale of only proxy scores. Leukemic children’s age-specific needs should be addressed properly to improve their overall HRQOL.


Arterial ischemic strokes (AIS) are rare in childhood. Congenital and acquired heart diseases are one of the most important risk factors of AIS in children. OBJECTIVE: Study the outcome of children with heart disease that have suffered AIS and the factors that influence on prognosis. PATIENTS AND METHODS: We evaluated all children with heart disease who had suffered AIS between 2000 and 2014 in our hospital. RESULTS: Seventy-four children with heart disease suffered an arterial ischemic stroke. 20% of them died and 10% had new AIS during the study period. Fifty-two patients were evaluated an average of six years after AIS. According to the Paediatric Stroke Outcome Scale (PSOM), most of the patients had some degree of impairment, mainly in sensorimotor and in cognitive-behavioural areas. The modified Rankin scale (mRS) showed an unfavourable outcome in 70% of patients (including patients that have died). Upper limb was more functionally impaired than lower limb. Strokes in neonatal period and early life were associated with poor prognosis. Size of stroke, cortical and subcortical involvement and basal ganglia stroke were associated with an unfavourable outcome. Fever in the acute phase and hemiparesis at presentation were also poor prognostic factors. Epilepsy at time of evaluation was also associated with unfavourable outcome. On the other hand, a normal electroencephalogram was associated with favourable outcome. CONCLUSIONS: AIS in children with heart disease had an unfavourable outcome, with impairment in different areas. Epilepsy happened in one third of the patients.


Cannabis is used to relieve nausea, trigger weight gain, and reduce pain among adults living with HIV; however, the relationship between its use and medication adherence and management is unclear. Participants (N = 107) were from an ongoing cohort study of community-dwelling HIV+ adults, stratified by cannabis (CB) use: HIV+/CB+ (n = 41) and HIV+/CB- (n = 66). CB+ participants either tested positive in a urine toxicology screen for THC or had a self-reported history of recent use. HIV-status was provided by physician results and/or biomarker assessment. Adherence was measured via the Morisky scale and medication management was assessed via the Medication Management Test-Revised. After adjusting for gender, we found no association between cannabis use group and adherence nor medication management. The amount of cannabis used was also

not associated with measures of adherence and management. Preliminary findings suggest that cannabis use may not adversely influence medication adherence/management among adults living with HIV.


Patient-reported outcomes are increasingly emphasized in clinical trials and population health studies. Our research team developed a smartphone app to track patient-reported outcomes of children with chronic diseases. The purpose of this study is to develop a patient-reported outcome reporting app and evaluate its usability. A multidisciplinary research team including health services researchers, pediatric nurses, and software engineers worked collaboratively in developing the patient-reported outcome app and administration portal. Group discussions and several rounds of feedback and modification were used. Ten pediatric patients with cancer, five parents, and two nurses participated in the usability study. We conducted content analyses in app development and usability evaluation. The app collected demographic information and patient-reported outcomes. Patient-reported outcomes were collected by Chinese versions of pediatric Patient-Reported Outcomes Measurement Information System Short Forms and Patient-Reported Outcomes Measurement Information System Parent Proxy Report Scales for Children. Pediatric patients aged 8 to 17 years and parents with a 5- to 7-year-old pediatric child used different age-appropriate questionnaires. The Web-based administration portal helped to manage demographic information, questionnaires, administrators, and survey-conducting organizations. The users liked the app. All participants felt that this app was easy to use and the interfaces were friendly to children. Nurses thought the administration portal interfaces were simple and the data were convenient to download for further analysis. We conclude that the app and its administration portal meet researchers and clinical nurses’ demand and have potential to promote patient-reported outcomes in assessing quality of life and symptoms of pediatric patients.


OBJECTIVE: To use an objective metric of effort of breathing to determine optimal high flow nasal cannula (HFNC) flow rates in children <3 years of age. STUDY DESIGN: Single-center prospective trial in a 24-bed pediatric intensive care unit of children <3 years of age on HFNC. We measured the percent change in pressurerate product (PRP) (an objective measure of effort of breathing) as a function of weight-indexed flow rates of 0.5, 1.0, 1.5, and 2.0 L/kg/minute. For a subgroup of patients, 2 different HFNC delivery systems (Fisher & Paykel [Auckland, New Zealand] and Vapotherm [Exeter, New Hampshire]) were compared. RESULTS: Twenty-one patients (49 titration episodes) were studied. The most common diagnoses were bronchiolitis and pneumonia. Overall, there was a significant difference in the percent change in PRP from baseline (of 0.5 L/kg/minute) with increasing flow rates for the entire cohort (P < .001) with largest change at 2.0 L/kg/min (-21%). Subgroup analyses showed no significant difference in percent change in PRP from baseline when comparing the 2 different HFNC delivery systems (P = .12). Patients </=8 kg experienced a larger percent change in PRP as HFNC flow rates were increased (P = .001) than patients >8 kg. CONCLUSIONS: The optimal HFNC flow rate to reduce effort of breathing in infants and young children is approximately 1.5-2.0 L/kg/minute with more benefit seen in children </=8 kg.


BACKGROUND: Population-based research to identify underserviced populations and the impact of palliative care (PC) is limited as the validity of such data to identify PC services is largely unknown. OBJECTIVE: To determine the validity of using such data to identify the involvement of specialized pediatric PC teams among children with cancer. DESIGN: Retrospective cohort. SUBJECTS: Ontario children with cancer who died between 2000 and 2012, received care through a pediatric institution with a specialized PC team and a clinical PC database.
MEASUREMENTS: All patients in the clinical databases were linked to population-based health services administrative databases. Six algorithms were created to indicate the use of formal pediatric PC teams based on the record type (physician billings vs. inpatient records vs. both) and number of eligible codes required (≥1 vs. ≥2). Each was validated against the pediatric PC clinical databases. RESULTS: The cohort comprised 572 children; 243 were in the clinical databases. Algorithms using only inpatient records had high specificity (80%-95%) but poor sensitivity (21%-56%). Including physician billings increased sensitivity but lowered specificity. The algorithm with overall best performance required ≥2 physician billing or inpatient diagnosis codes indicating PC [sensitivity 0.79 (95% CI 0.73-0.84), specificity 0.58 (95% CI 0.53-0.64)]. CONCLUSIONS: Health administrative data identifies involvement of specialized pediatric PC teams with good sensitivity but low specificity. Studies using such data alone to compare patients receiving and not receiving specialized pediatric PC are at significant risk of misclassification and potential bias. Population-based PC databases should be established to conduct rigorous population-based PC research.


BACKGROUND: This study evaluated the validity and reliability of the Italian version of the Non-Communicating Children’s Pain Checklist-Postoperative version (I-NCCPC-PV). METHODS: The original NCCPC-PV version was translated into Italian following the guidelines for "the translation, adaptation, and validation of instruments or scales for cross-cultural healthcare research". We tested the Italian NCCPC-PV version (I-NCCPC-PV) in 40 children (3-18 years of age) with severe to profound Intellectual Disability and no verbal communication. Each child’s behavior was observed by a parent or caregiver and by an external observer in a quiet situation and a painful one. They independently assessed the child’s level of pain using the translated Italian version of the NCCPC PV (I-NCCPC-PV). RESULTS: The results from 80 assessments showed that children's behavioral signs differed significantly between painful and calm situations (p < 0.001). The inter-rater reliability was poor in a quiet condition (ICC 0.62) and fair in a painful situation (ICC 0.77). The inter-rater agreement was good in both calm and painful conditions (72.50% and 77.50% respectively). CONCLUSION: The Italian version of the NCCPC-PV (I-NCCPC-PV) can be used for pain assessment in children with Intellectual Disability who lack verbal communication.


Our objective was to conduct a systematic review and meta-analysis for the use of modified (heat-killed or sonicated) probiotics for the efficacy and safety to prevent and treat various diseases. Recent clinical research has focused on living strains of probiotics, but use in high-risk patients and potential adverse reactions including bacteremia has focused interest on alternatives to the use of live probiotics. We searched MEDLINE/PubMed, Embase, Cochrane Central Register of Controlled Trials, CINAHL, Alt Health Watch, Web of Science, Scopus, PubMed, from inception to February 14, 2017 for randomised controlled trials involving modified probiotic strains. The primary outcome was efficacy to prevent or treat disease and the secondary outcome was incidence of adverse events. A total of 40 trials were included (n=3,913): 14 trials (15 arms with modified probiotics and 20 control arms) for the prevention of diseases and 26 trials (29 arms with modified probiotics and 32 control arms) for treatment of various diseases. Modified microbes were compared to either placebo (44%), or the same living probiotic strain (39%) or to only standard therapies (17%). Modified microbes were not significantly more or less effective than the living probiotic in 86% of the preventive trials and 69% of the treatment trials. Modified probiotic strains were significantly more effective in 15% of the treatment trials. Incidence rates of adverse events were similar for modified and living probiotics and other control groups, but many trials did not collect adequate safety data. Although several types of modified probiotics showed significant efficacy over living strains of probiotics, firm conclusions could not be reached due to the limited number of trials using the same type of modified microbe (strain, daily dose and duration) for a specific disease indication. Further research may illuminate other strains of modified probiotics that may have potential as clinical biotherapeutics.

Pain and Symptom Assessment and Relief


BACKGROUND: Systematic assessment of emotional distress is recommended in after care. Yet, it is unclear if parent report may be used as a proxy of child report. The aim of this study was to assess agreements and differences and explore possible moderators of disagreement between child and parent ratings. METHODS: Sixty-two young survivors treated for acute lymphoblastic leukemia (9-18 years) and both parents responded to the Beck Youth Inventory (anxiety and depression) and the distress rating scale on the child’s status. Parents completed the Brief Symptom Inventory-18 on their own psychological status. Systematic analyses of agreement and differences were performed. RESULTS: Mother-child and father-child agreements were fair on anxiety, depression, and distress (median intraclass correlation coefficient = 0.37). Differences between parents and children were medium sized (median d = 0.55) with parents giving higher scores than their children on anxiety, depression, and distress. Mothers reported distress more frequently than fathers (39 vs. 17%) when children reported none. The child being female and lower parental income were associated with lower agreement in fathers when rating child distress. Higher levels of parental psychological symptoms were consistently associated with lower agreement. CONCLUSIONS: Parent-child differences when rating adolescent survivors’ difficulties may be more important than previously thought. Parent report probably cannot be considered as a valid proxy of older child report on such internalized domains as anxiety, depression, or distress in the after-care clinic. Parents’ report is also likely to be influenced by their own mood, a factor that should be corrected for when using their report.


BACKGROUND: Changes in the manner in which medications can be delivered can have significant effects on the quality of care in the acute care setting. OBJECTIVE: The objective of this study was to evaluate the change in three Institute of Medicine quality indicators (timeliness, safety, and effectiveness) in the pediatric emergency department (ED) after the introduction of the Mucosal Atomizer Device Nasal (MADn) for opioid analgesia.

METHODS: This was a retrospective review of patients receiving opioid analgesia for certain conditions over a 5-year period. We compared patients receiving intravenous opioid (IVO) to those receiving intranasal fentanyl (INF). Timeliness outcomes include time from medication order to administration, time from dose to discharge, overall time to analgesia, and ED length of stay. Effectiveness outcomes include change in pain score and frequency of repeat dosing. Safety outcomes were the frequency of reversal agent administration or a documented oxygen desaturation of < 90%. Sensitivity analyses were performed to evaluate the effect of moderate sedation on all three outcomes. RESULTS: During the study period, 1702 patients received opioid analgesia, 744 before and 958 after MADn introduction, of whom, 233 (24%) received INF. After MADn introduction, patients receiving INF had a shorter time to discharge from dose (109 vs. 203 min; p < 0.05) and shorter ED length of stay (168 vs. 267 min; p < 0.05). There was no difference in pain score reduction; however, repeat dosing was less frequent for patients receiving INF (16% vs. 27%). There was no use of reversal medication and no difference in the frequency of oxygen desaturations. When patients undergoing moderate sedation were removed from the analysis, there was no difference in the direction of findings for all three outcomes. CONCLUSIONS: INF is associated with improved timeliness and equivalent effectiveness and safety when compared to IVO in the setting of the pediatric ED.


BACKGROUND: The rate of an unintentional drug overdose involving prescription opioids continues to rise. An understanding of the threshold dose and dose(s) associated with unintentional prescription opioid overdose will help to mitigate this epidemic. OBJECTIVE: The objective of this systematic review is to systematically synthesise and meta-analyse studies on doses of prescription opioids and ascertain the doses of opioids that are associated...
with increased risk of severe opioid poisoning or mortality. DATA SOURCES: A search of PubMed, EMBASE, CINAHL and Web of Science from inception to 16 January 2017 was conducted using search strategies and the MeSH (Medical Subject Headings) terms for studies of adult patients using prescription opioids who experienced an accidental overdose. STUDY SELECTION: Of the 1332 studies identified, 117 were selected for full article review. Ten met the inclusion criteria for qualitative analysis, but only seven studies were meta-analysed. The included studies were in English, and participants met predetermined International Classification of Diseases (ICD) codes. Studies were excluded if they included only paediatric participants or the participants met the ICD code for intentional self-harm. DATA EXTRACTION AND SYNTHESIS: Two researchers elaborated and validated a data extraction form. Data were then independently extracted by both reviewers as per this form. We assessed study quality using the Newcastle-Ottawa Scale (NOS) for non-randomised studies in meta-analyses. We performed a meta-regression using a random-effect model and summarised the results using relative risk (RR) and 95% confidence intervals (CIs). The threshold dose for an unintentional overdose is 20 morphine milligram equivalents (MME)/day. There were higher risks with larger doses: (1) < 20 versus ≥ 21 MME/day: RR 2.81, 95% CI 1.09-7.22, p < 0.001; (2) < 50 versus > 50 MME/day: RR 3.87, 95% CI 2.36-6.33, p < 0.001; (3) < 100 versus > 100 MME/day: RR 4.28, 95% CI 2.61-7.1, p < 0.001; and (4) < 50 versus > 50-100 MME/day: RR 3.09, 95% CI 1.84-5.18, p < 0.001. Heterogeneity was explained by the type of overdose event, inpatient or outpatient status, and length of observation. Type of pain (cancer or non-cancer pain) had no impact on heterogeneity. LIMITATIONS: The definition of exposure in studies included in the meta-analysis was heterogeneous. Some studies defined exposure as the filling of a prescription while others defined exposure as the prescription of an opioid to the patient, and all studies assumed that patients took the prescribed opioid. Medications that may contribute to overdose, such as benzodiazepines and other drugs, were not considered. CONCLUSIONS: A significantly increased risk of inadvertent prescription opioid overdose was found with 20–50 MME/day, with fatality more likely with opioid doses above 50 MME/day, although extensive heterogeneity was found with the dose comparisons. Clinicians should inform patients of this risk and monitor them closely. PROTOCOL REGISTRATION: This protocol was registered with PROSPERO 2017: CRD42017058426.

https://www.ncbi.nlm.nih.gov/pubmed/29498021


BACKGROUND: Neonatal seizures are the most common clinical manifestation of Central Nervous System (CNS) dysfunction and are associated with various neurological sequelae. There are currently no evidence-based guidelines for the management of neonatal seizures and currently used drugs such as phenobarbital, and phenytoin have limited efficacy and potential toxicities. Newer second line anticonvulsants, levetiracetam, has been used in refractory neonatal seizures despite limited data and off-label use. OBJECTIVE: In this review, we will discuss various pharmacological properties of levetiracetam when used in neonatal population. METHODS: A PubMed search for MEDLINE was undertaken to look for studies using the terms "Levetiracetam", AND "Neonates" as key words from year 1995 to January 2017. Relevant articles were selected and information was extracted about pharmacokinetics, pharmacodynamics and clinical uses of levetiracetam in neonates. RESULTS: Levetiracetam is an active, water-soluble S-enantiomer of racemic pyrrolidine acetamide which exerts its antiepileptic action by binding to the synaptic vesicle protein within the brain. Metabolism of levetiracetam does not include the CYP P450 system and it is mainly eliminated through kidneys after rapid absorption. Also, no significant interactions with other drugs have been identified. Unlike other commonly used antiepileptic drugs, levetiracetam is not bound to plasma proteins, thereby, reducing the chances of toxicity and severe, life threatening side effects have not been reported. In fact, it has been shown to prevent neuro-degeneration after hypoxia/ischemia in rodent models of epilepsy. CONCLUSION: Levetiracetam has been emerging as a potential therapeutic option for refractory neonatal convulsions owing to its non-hepatic elimination, linear pharmacokinetics, low protein binding and better safety profile.


Paracetamol (acetaminophen) is the most widely used drug to treat pain or fever in pregnant women or neonates, but its pharmacokinetics (PK) and pharmacodynamics (PD) warrant a focused analysis. During pregnancy, there is an important increase in paracetamol clearance. Consequently, it is reasonable to anticipate that the analgesic effect of paracetamol will decrease faster, whereas higher doses may result in even higher oxidative toxic metabolites. Therefore, most peripartal PD data relate to multimodal analgesia strategies. In neonates,
weight/size is the most relevant covariate of paracetamol PK. This resulted in proposed dosing regimens containing higher doses than currently prescribed in the label for term neonates. Using adequate dosing, paracetamol is a poor procedural analgesic, is effective for mild-to-moderate pain, and has morphine-sparing effects. Short-term safety has been well documented, and there is active research investigating the potential association between paracetamol exposure and atopy, fertility, and neurobehavior.


A framework for defining pain terms such as acute, persistent, prolonged or chronic pain to newborns was derived from the scientific literature on neonatal pain assessments, previous attempts to define chronic pain and the clinical and neurophysiological features of neonatal pain. This novel framework incorporates the temporal features, localising characteristics, and secondary effects of the pain experienced, as well as the behavioural and physiological response patterns of newborns. CONCLUSION: Although not evidence-based, this framework provides an initial starting point for defining commonly used neonatal pain terms. It will require future revision/refinement based on the accumulating evidence for non-acute pain.


Medical marijuana (MM) has become increasingly legal at the state level and accessible to children with serious illness. Pediatric patients with cancer may be particularly receptive to MM, given purported benefits in managing cancer-related symptoms. In this review, we examine the evidence for MM as a supportive care agent in pediatric oncology. We describe the current legal status of MM, mechanism of action, common formulations, and potential benefits versus risks for pediatric oncology patients. We offer suggestions for how providers might approach MM requests. Throughout, we comment on avenues for future investigation on this growing trend in supportive care.


AIM: The neonatal intensive care unit (NICU) provides life-saving medical care for an increasing number of newborn infants each year. NICU care, while lifesaving, does have attendant consequences which can include repeated activation of the stress response and reduced maternal interaction, with possible negative long-term impacts on brain development. Here we present a neuroscientific framework for considering the impact of music on neurodevelopment in the NICU of infants born preterm and evaluate current literature on the use of music with this population to determine what is most reliably known of the physiological effects of music interventions. METHOD: Using online academic databases we collected relevant, experimental studies aimed at determining effects of music listening in infants in the NICU. These articles were evaluated for methodological rigor, ranking the 10 most experimentally stringent as a representative sample. RESULTS: The selected literature seems to indicate that effects are present on the cardio-pulmonary system and behavior of neonates, although the relative effect size remains unclear. INTERPRETATION: These findings indicate a need for more standardized longitudinal studies aimed at determining not only whether NICU music exposure has beneficial effects on the cardio-pulmonary system, but also on the hypothalamic-pituitary-adrenal axis, brain structures, and cognitive behavioral status of these children as well. WHAT THIS PAPER ADDS: Provides a neuroscience framework for considering how music might attenuate stress in neonatal intensive care unit (NICU) infants. Considers how repeated stress may cause negative neurodevelopmental impacts in infants born preterm. Posits epigenetics can serve as a mechanistic pathway for music moderating the stress response.


The transition from a pediatric to adult health care system is challenging for many youths with epilepsy and their families. Recently, the Ministry of Health and Long-Term Care of the Province of Ontario, Canada, created a transition working group (TWG) to develop recommendations for the transition process for patients with epilepsy in the Province of Ontario. Herein we present an executive summary of this work. The TWG was composed of a multidisciplinary group of pediatric and adult epileptologists, psychiatrists, and family doctors from academia and from the community; neurologists from the community; nurses and social workers from pediatric and adult epilepsy programs; adolescent medicine physician specialists; a team of physicians, nurses, and social workers dedicated to patients with complex care needs; a lawyer; an occupational therapist; representatives from community epilepsy agencies; patients with epilepsy; parents of patients with epilepsy and severe intellectual disability; and project managers. Three main areas were addressed: (1) Diagnosis and Management of Seizures; 2) Mental Health and Psychosocial Needs; and 3) Financial, Community, and Legal Supports. Although there are no systematic studies on the outcomes of transition programs, the impressions of the TWG are as follows. Teenagers at risk of poor transition should be identified early. The care coordination between pediatric and adult neurologists and other specialists should begin before the actual transfer. The transition period is the ideal time to rethink the diagnosis and repeat diagnostic testing where indicated (particularly genetic testing, which now can uncover more etiologies than when patients were initially evaluated many years ago). Some screening tests should be repeated after the move to the adult system. The seven steps proposed herein may facilitate transition, thereby promoting uninterrupted and adequate care for youth with epilepsy leaving the pediatric system.


Management of pain is one of the major expectations of children with neurological impairment and their families. The medical literature is poor on this topic accounting for approximately 0.15 % of the publications on pain in general. The objective of the French Pediatric Neurology Society was to review the current knowledge on this topic. Bibliographic research was conducted with PubMed and RefDoc for publications between 1994 and 2014 in French or English. A total of 925 articles were retrieved and 92 were selected for review. Pain is common in this population: a 2-week survey indicated that pain occurs in 50-75 % of children. Pain negatively impacts the quality of life of children and their parents. Children with neurological impairment express their pain with pain expression patterns and specific patterns common to children (change of tone, abnormal movements, spasticity, paradoxical reactions, such as laughter, self-injury or vasomotor dysfunction). Some children with neurological impairment are able to use self-report pain scales. If not, observational measures should be used. Behavioral rating scales specifically designed for this population are more sensitive than others. Scales must be selected according to children’s communication skills, type of pain, and the context. Sometimes behavioral changes are the only expression of pain: any change in sleep, tone, feeding, or mood must suggest pain in this population. Management of pain remains difficult. There are no specific guidelines. Procedural pain management guidelines and the usual analgesic drugs can be used in children with neurological impairment with specific concerns regarding tolerance and side effects. These children are particularly at risk for neuropathic pain. A multidisciplinary approach is helpful, involving physicians, nurses, physiotherapists, psychologists and parents.

https://europepmc.org/abstract/med/29273448


Pain in children is underestimated and undertreated because of lack of pain assessment tools. Pain assessment depends on the cognitive development of the child being tested, clinical context, and pain typology. For children older than age 6 years, pain assessment is based on a self-report. For children younger than age 6 years, behavioral pain scales are needed to assess pain. Numerous pain scales exist. Many are reliable and some are recommended, but all have specific conditions for their use. In this article, we review the available pain scales for children from birth to adolescence. We provide the validity criteria of each pain scale to help caregivers use the adapted tools. We then propose a synthesis of the reliable tools to use based on the pain context. [Pediatr Ann. 2017;46(10):e387-e395.]

OBJECTIVES: Diagnostic labels can help patients better understand their symptoms and can influence providers' treatment planning and patient interactions. Recurrent pain is common in childhood; however, there are various diagnostic labels used. The objective of this study was to evaluate the influence of diagnostic labels on pediatric health care providers' perceptions of pediatric chronic pain patients. MATERIALS AND METHODS: Using an online survey, providers were randomly assigned to 1 of 2 vignette conditions (differing only in diagnostic label provided) and completed questionnaires about their perceptions of the vignette patient. RESULTS: Responses from 58 participants were analyzed. The 2 groups, based on diagnostic conditions used (fibromyalgia and chronic widespread pain) did not differ significantly on general demographics and health care providers' perceptions of the patient. Perceived origin of the pain influenced providers' perceptions; pain of a perceived medical origin was negatively correlated with stigmatization and positively correlated with sympathy. Perceived psychological origin was positively correlated with stigmatization and providers' age. DISCUSSION: Health care providers' perceptions of children's pain are more likely influenced by the presumed etiology rather than the diagnostic label used. Pain believed to be more medically based was associated with more positive reactions from providers (ie, less stigmatization). Older providers in particular perceived the patient more negatively if they believe the pain to be psychologically based. The findings of this pediatric study replicated findings from adult literature on chronic pain, suggesting that children and adults are subject to negative perceptions from health care providers when the providers believe the pain to be psychological in origin. https://www.ncbi.nlm.nih.gov/pubmed/27922842


OBJECTIVES: Fatigue is common among children living with cancer, particularly in advance stages. Little is known about the effectiveness of non-pharmacological approaches to manage this complex and distressing symptom among children. Thus, the present paper aim to critically examine the effectiveness and setting for non-pharmacological interventions to manage fatigue among children with cancer. METHODS: Six electronic databases were screened first in February 2013 and at second instance in March 2015. They include PsycINFO, Medline, EMBASE, CINAHL, Scopus and Cochrane library. All databases were systematically searched for literature on fatigue and cancer, limited to children (as age group) and English language. RESULTS: 1498 articles were identified, of which six were reviewed. Three types of interventions for managing fatigue were identified including (1) complementary and alternative medicine (healing touch/massage therapy), (2) exercise-based interventions and (3) nursing-based interventions. Most interventions were delivered during active treatment and in hospital settings where parents were involved to optimise participation. Despite fatigue scores being lower among intervention groups, no study findings were observed as being statistically significant. CONCLUSION: Fatigue is common among children treated for and living with cancer. The most appropriate setting to deliver non-pharmacological interventions to manage fatigue appears to be in hospital. However, in absence of any strong evidence, professionals need to be cautious about existing non-pharmacological interventions. Future research must adopt more rigorous research designs that are adequately powered using validated measures to identify potential benefits. In addition, researchers may wish to test psychosocial interventions shown to be of benefit in adults. https://www.ncbi.nlm.nih.gov/pubmed/28760817


AIM: Evaluation of comfort and pain in neonates is important for management. Specific signs of persistent pain in neonates remain undefined; few validated clinical tools assess persistent pain. We sought to determine (i) difficulty perceived by staff and parents in assessing comfort/persistent pain in babies, (ii) strategies employed when no clinical tool is used and (iii) variation between clinicians' assessments. METHODS: Parent and staff questionnaires addressed difficulty in assessing pain/comfort in neonates and strategies used in making assessments. RESULTS: A total of 47 of 50 (94%) parents and 83 of 91 (91%) staff participated; 50% of staff reported it was moderately/very difficult to assess persistent pain, and 13% very easy; 75% of parents found it moderately/very easy and 23% difficult to assess their baby's comfort; 15% of parents thought staff found pain

Participants were 17 healthy premature infants, randomly allocated to the intervention group or the control group. METHODOLOGY: A double-blind, randomized controlled trial was conducted in the NICUs of 2 general public hospitals in Andalusia, Spain. RESULTS: The study demonstrated that infants in the intervention group had lower respiratory rates, systolic and diastolic blood pressure, and heart rate compared to the control group. CONCLUSIONS: Listening to relaxing music therapy may improve physiological responses in premature infants. However, further studies are needed to confirm these findings and evaluate the long-term effects of this intervention.

OBJECTIVE: To evaluate the effect of dexmedetomidine infusions in patients with advanced malignancies, advanced heart disease, or after stem cell transplantation (SCT), who during end-of-life care had pain and/or agitation unresponsive to conventional therapies. BACKGROUND: Pediatric patients with intractable advanced malignancies, end-stage congenital heart diseases, or after SCT can suffer a great deal during end of life. Pain, drowsiness, fatigue, irritability, and worrying are experienced frequently, considered distressing, and are strongly associated with reductions in health-related quality-of-life scores. While opioids are the mainstay of analgesic therapy, in some patients, increasing opioid use can be ineffective and can be associated with increasing pain during end of life. Dexmedetomidine, a alpha2-adrenoreceptor agonist with sedative and analgesic properties but without respiratory depressant effects, has been shown to reduce opioid requirement and to facilitate opioid weaning. METHODS: Observational cohort study of consecutive patients treated with dexmedetomidine during end of life in a pediatric intensive care unit (PICU). Primary outcomes included pain scores and morphine-equivalent intake. RESULTS: We identified nine patients (median age 8 [interquartile range: 7-13 years]) who during end of life had received dexmedetomidine infusions. In these patients, dexmedetomidine infusions had a median duration of two days (IQR 1.5-10 days) and were associated with significant (p < 0.001) reductions in pain scores and a trend toward decreasing morphine-equivalent intake. There were no hemodynamic changes requiring vasoactive or anticholinergic agents. CONCLUSIONS: These preliminary findings of beneficial effects of dexmedetomidine support the hypothesis that dexmedetomidine has a role in palliative care of children and adolescents during end of life.

OBJECTIVE: Our aim was to examine and map the consequences of chronic pain in children and adolescents. METHOD: A scoping review was carried out in the international databases (PubMed, SCOPUS, WOS and CINAHL, Cochrane Library) and gray literature. We included documents that addressed psychosocial aspects that influence chronic pain, published in English between 2010 and 2016. We excluded the documents that dealt with pharmacological treatments, chronic pain derived from surgical interventions or where there was no access to full text. 34 of the 716 documents reviewed were included. RESULTS: Studies show that pain is associated with high rates of functional disability, sleep disorders and spectrum depression-anxiety. Young people experience higher rates of victimization and stigmatization, contributing to social isolation, difficulty in meeting academic demands and less opportunity to consume illegal substances. With respect to the family, chronic pain has been associated with poorer family functioning and considerable investment of economic resources. CONCLUSIONS: This Scoping Review shows that functional capacity, sleep, personal development, peer support and family functioning are interesting lines in published works. However, gaps in knowledge are detected in areas such as risk behaviours, the consequences that pain can cause in adulthood and gender inequalities.

BACKGROUND: Premature infants are exposed to high levels of noise in the neonatal intensive care unit (NICU). PURPOSE: This study evaluated the effect of a relaxing music therapy intervention composed by artificial intelligence on respiratory rate, systolic and diastolic blood pressure, and heart rate. METHODS: A double-blind, randomized, controlled trial was conducted in the NICUs of 2 general public hospitals in Andalusia, Spain. Participants were 17 healthy premature infants, randomly allocated to the intervention group or the control group (silence) at a 1:1 ratio. To be included in the study, the subjects were to be 32 to 36 weeks of gestation at birth.
The intervention lasted 20 minutes, 3 times a day for 3 consecutive days, while infants were in the incubator. Infants' heart rate, respiratory rate, and blood pressure were assessed before and after each intervention session. RESULTS: After each session, the respiratory rate decreased in the experimental group (main between-groups effect (F1,13 = 6.73, P = .022, etapartial = 0.34). Across the sessions, the heart rate increased in the control group (main between-groups effect, F1,11 = 5.09, P = .045, etapartial = 0.32). IMPLICATIONS FOR RESEARCH: Future studies can use this music intervention to assess its potential effects in premature infants. IMPLICATIONS FOR PRACTICE: Nurses can apply the relaxing music intervention presented in this study to ameliorate the impact of the stressful environment on premature infants. 


BACKGROUND: Patient satisfaction with pain management is associated with improved patient adherence to medical management and efficient service utilization. Pediatric pain control is challenging, given the inability to elicit reliable histories, particularly in younger patients. Several studies have suggested that communication surrounding pain management can improve satisfaction, although there are limited data describing structured interventions with measurable outcomes. A quality improvement project was conducted to determine if reliably asking families about pain management was associated with improved patient satisfaction with pain management. METHODS: In an academic pediatric hospital, nurse manager rounds were used to invite a conversation about pain management. The question, "Pain management is very important to us. Has your child's pain been well controlled?" was added to the established standard questions asked during nurse manager rounds. Effectiveness was measured using the preexisting Press Ganey survey question, "How well was your child's pain controlled?" Responses were compared between those patients who were and were not exposed to the rounding question. RESULTS: Data for 1,032 patients were used to establish baseline satisfaction scores. In the intervention period, 328 patients received nurse manager rounds and 121 did not. The median of the weighted mean patient survey satisfaction scores were baseline, 91.5%; receiving intervention, 94.2%; and not receiving intervention, 90.0%. Patients who received the intervention reported higher satisfaction with pain management than those who did not (p <0.0001). CONCLUSION: Hospitals seeking to improve satisfaction with pain management should encourage health care providers to reliably discuss pain control with pediatric patients. 


OBJECTIVES: To describe the perceptions of nurses in neonatal units on pain management, meet the educational profile and describe the use of pain assessment tools and non-pharmacological management for treatment. METHODS: Cross-sectional descriptive multicentre study, developed during the months of February to September 2015, in the neonatology services of three hospitals at the Community of Madrid, Spain. Data collection was performed through an ad hoc questionnaire on paper or electronically using Survey Monkey platform. RESULTS: The sample consisted of 142 professionals, with a response rate of 55%; 47.9% (68) confirmed they had received specific training in pain management; 39.5% (56) stated that pain is regularly assessed in the unit; only 43.6% reported using validated scales, the most used being the Premature Infant Pain Profile (PIPP). As for the non-pharmacological management, swaddling and non-nutritive sucking it is the most used, followed by sucrose. Intravenous cannulation was identified as the most painful procedure. CONCLUSIONS: Pain management is in the process of improvement, because of training and because there is little pain assessment using validated scales. The improvement in the use of non-pharmacological management for the relief of pain in minor procedures is noteworthy. 

Synopsis One of the key aspects of good health care for children and young people is the prevention and management of pain. The experience of persistent pain in children and adolescents not only has a major impact on physical, emotional, social, and developmental well-being, but also impacts the broader world, which includes family, school, and social networks. The multidisciplinary pediatric pain clinic adopts a holistic approach to care through a biopsychosocial model. One outcome of an initial pediatric pain clinic review is the creation of a pain management plan that addresses the pharmacological, physical, psychological, and other domains of care. Pediatric pain clinics are improving access by embracing technology through tele-health and internet-based treatment options. Outcome measurement will guide the development of models of care in the future. J Orthop Sports Phys Ther 2017;47(10):806-813. Epub 12 Sep 2017. doi:10.2519/jospt.2017.7355.


Many paediatric patients with cancer experience significant chemotherapy side effects. Predisposition to drug reactions is governed by single nucleotide polymorphisms (SNPs). We performed a systematic review of the literature from 2006 through 2016. Outcomes of interest included patient characteristics, cancer type drug of interest, genes investigated, toxicity identified and genetic polymorphisms implicated. The primary toxicities studied were neurotoxicity cardiotoxicity, osteonecrosis, and thromboembolism and hypersensitivity reactions. The retrieved studies were grouped according to toxicity reported and SNP associations. This review highlights the discoveries to date in pharmacogenomics and paediatric oncology along with highlighting some of the important limitations in the area.


BACKGROUND AND AIMS: No published studies have looked at the dosing and use of rapid onset fentanyl preparations in children. The primary aim of this study was to assess whether there is a correlation between effective dose of rapid onset fentanyl and background oral morphine equivalent analgesia in children less than 18 years old. Secondary objectives included establishing whether there is a correlation between effective dose of rapid onset fentanyl and age and weight. Reported side effects were also reviewed. METHODS: This study is a retrospective case note review of all children less than 18 years old who received rapid onset fentanyl products in a tertiary paediatric oncology centre in England between 2010 and 2015. Correlations were analysed using Spearman’s correlation coefficient as data was non-parametric. RESULTS: Data on 26 children (5-17 yrs; 13-100kg) was analysed. The most common diagnosis in children being given rapid onset fentanyl products was a solid tumour (84.6%). Eleven children used sublingual tablets, 17 used lozenges and one used a fentanyl nasal spray (three patients used two different preparations). The only significant correlation found was between dose of fentanyl lozenge and weight (\(r_s=0.81, p<0.001\)). Very few side effects were reported with the most frequent being nausea (8%) and sleepiness (8%). CONCLUSIONS: Fentanyl lozenges seem to be safe and well tolerated in children as young as five years old, weighing as little as 13kg. Results suggest that children should always be started on the lowest available dose of chosen preparation and that this dose should be titrated according to response. This study demonstrates that there is no correlation between background opioid dose and effective dose of rapid onset fentanyl in children. This mirrors findings of similar studies in adults. There was a strong correlation between effective dose of fentanyl lozenge and weight. This may be in part due to clinicians being more inclined to increase fentanyl lozenge doses as the child is in control of when they have had enough medication. In contrast, buccal tablets are absorbed quickly and the child always receives the full dose, making clinicians more reluctant to titrate the dose. IMPLICATIONS: This article presents initial evidence for feasibility and tolerability of fentanyl lozenges in children as young as five years old, who are on relatively low doses of background opioids. This could be of interest to clinicians who are looking for alternatives to oral opioids to manage breakthrough pain in children with cancer.

BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization guidelines for pharmacological treatments for children’s persisting pain acknowledge that pain in children is a major public health concern of high significance in most parts of the world. While in the past, pain was largely dismissed and was frequently left untreated, views on children’s pain have changed over time, and relief of pain is now seen as important. We designed a suite of seven reviews on chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol as priority areas) in order to review the evidence for children’s pain utilising pharmacological interventions in children and adolescents. As the leading cause of morbidity in children and adolescents in the world today, chronic disease (and its associated pain) is a major health concern. Chronic pain (lasting three months or longer) can arise in the paediatric population in a variety of pathophysiological classifications: nociceptive, neuropathic, idiopathic, visceral, nerve damage pain, chronic musculoskeletal pain, and chronic abdominal pain, and other unknown reasons. Paracetamol (acetaminophen) is one of the most widely used analgesics in both adults and children. The recommended dosage in the UK, Europe, Australia, and the USA for children and adolescents is generally 10 to 15 mg/kg every four to six hours, with specific age ranges from 60 mg (6 to 12 months old) up to 500 to 1000 mg (over 12 years old). Paracetamol is the only recommended analgesic for children under 3 months of age. Paracetamol has been proven to be safe in appropriate and controlled dosages, however potential adverse effects of paracetamol if overdosed or overused in children include liver and kidney failure. OBJECTIVES: To assess the analgesic efficacy and adverse events of paracetamol (acetaminophen) used to treat chronic non-cancer pain in children and adolescents aged between birth and 17 years, in any setting. SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online, MEDLINE via Ovid, and Embase via Ovid from inception to 6 September 2016. We also searched the reference lists of retrieved studies and reviews, and searched online clinical trial registries. SELECTION CRITERIA: Randomised controlled trials, with or without blinding, of any dose and any route, treating chronic non-cancer pain in children and adolescents, comparing paracetamol with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for eligibility. We planned to use dichotomous data to calculate risk ratio and numbers needed to treat, using standard methods where data were available. We assessed GRADE (Grading of Recommendations Assessment, Development and Evaluation) and planned to create a ‘Summary of findings’ table. MAIN RESULTS: No studies were eligible for inclusion in this review. We rated the quality of the evidence as very low. We downgraded the quality of evidence by three levels due to the lack of data reported for any outcome. AUTHORS’ CONCLUSIONS: There was no evidence from randomised controlled trials to support or refute the use of paracetamol (acetaminophen) to treat chronic non-cancer pain in children and adolescents. We are unable to comment about efficacy or harm from the use of paracetamol to treat chronic non-cancer pain in children and adolescents. We know from adult randomised controlled trials that paracetamol, can be effective, in certain doses, and in certain pain conditions (not always chronic). This means that no conclusions could be made about efficacy or harm in the use of paracetamol (acetaminophen) to treat chronic non-cancer pain in children and adolescents.


BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization guidelines for pharmacological treatments for children’s persisting pain acknowledge that pain in children is a major public health concern of high significance in most parts of the world. While in the past, pain was largely dismissed and was frequently left untreated, views on children’s pain have changed over time, and relief of pain is now seen as important. We designed a suite of seven reviews on chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol as priority areas) in order to review the evidence for children’s pain utilising pharmacological interventions in children and adolescents. As the leading cause of morbidity in children and adolescents in the world today, chronic disease (and its associated pain) is a major health concern. Chronic pain (lasting three months or longer) can arise in the paediatric population in a variety of pathophysiological classifications: nociceptive, neuropathic, idiopathic, visceral, nerve damage pain, chronic musculoskeletal pain, and chronic abdominal pain, and other unknown reasons. Opioids are used worldwide for the treatment of pain. They bind to opioid receptors in the central nervous system (mu, kappa, delta, and sigma) and can be agonists, antagonists, mixed agonist-antagonists, or partial agonists. Opioids are generally available in healthcare settings across most high-income countries, but access may be restricted in low- and middle-income
countries. For example, opioids currently available in the UK include: buprenorphine, codeine, fentanyl, hydromorphone, methadone, morphine, oxycodone, and tramadol. Opioids are used in varying doses (generally based on body weight for paediatric patients) by means of parenteral, transmucosal, transdermal, or oral administration (immediate release or modified release). To achieve adequate pain relief in children using opioids, with an acceptable grade of adverse effects, the recommended method is a lower dose gradually titrated to effect in the child. OBJECTIVES: To assess the analgesic efficacy and adverse events of opioids used to treat chronic non-cancer pain in children and adolescents aged between birth and 17 years, in any setting. SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Library, MEDLINE via Ovid, and Embase via Ovid from inception to 6 September 2016. We also searched the reference lists of retrieved studies and reviews, and searched online clinical trial registries. SELECTION CRITERIA: Randomised controlled trials, with or without blinding, of any dose and any route, treating chronic non-cancer pain in children and adolescents, comparing opioids with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for eligibility. We planned to use dichotomous data to calculate risk ratio and number needed to treat, using standard methods. We assessed GRADE (Grading of Recommendations Assessment, Development and Evaluation) and planned to create a ‘Summary of findings’ table. MAIN RESULTS: No studies were eligible for inclusion in this review. We rated the quality of the evidence as very low. We downgraded the quality of evidence by three levels due to the lack of data reported for any outcome. AUTHORS’ CONCLUSIONS: There was no evidence from randomised controlled trials to support or refute the use of opioids to treat chronic non-cancer pain in children and adolescents. We are unable to comment about efficacy or harm from the use of opioids to treat chronic non-cancer pain in children and adolescents. We know from adult randomised controlled trials that some opioids, such as morphine and codeine, can be effective in certain chronic pain conditions. This means that no conclusions could be made about efficacy or harm in the use of opioids to treat chronic non-cancer pain in children and adolescents.


BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization (WHO) guidelines for pharmacological treatments for persisting pain in children acknowledge that pain in children is a major public health concern of high significance in most parts of the world. Views on children's pain have changed over time and relief of pain is now seen as important. In the past, pain was largely dismissed and was frequently left untreated, and it was assumed that children quickly forgot about painful experiences. We designed a suite of seven reviews in chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol as priority areas) to review the evidence for children's pain using pharmacological interventions. As one of the leading causes of mortality and morbidity for children and adolescents in the world today, childhood cancer (and its associated pain) is a major health concern. Specific mortality and morbidity data relating to children are not currently identified. All childhood cancer rates are on the rise; for example, in the USA approximately 10,380 children aged under 15 years were expected to be diagnosed with cancer by the end of 2016. However, with survival rates also increasing, over 80% of paediatric cancer patients are expected to survive for five years or more, thus identifying the need to address pain management in this population. Cancer pain in infants, children, and adolescents is primarily nociceptive pain with negative long term effects. Cancer-related pain is generally caused directly by the tumour itself such as compressing on the nerve or inflammation of the organs. Cancer-related pain generally occurs as a result of perioperative procedures, nerve damage caused by radiation or chemotherapy treatments, or mucositis. However, this review focused on pain caused directly by the tumour itself such as nerve infiltration, external nerve compression, and other inflammatory events. Non-steroidal anti-inflammatory drugs (NSAIDs) are used to treat pain, reduce fever, and for their anti-inflammatory properties. They are commonly used within paediatric pain management. NSAIDs are currently licensed for use in western countries, however not approved for infants aged under three months. Primary adverse effects include gastrointestinal issues and possible renal impairment with long term use. Other adverse effects in children include diarrhoea, headache, nausea, constipation, rash, dizziness, and abdominal pain. OBJECTIVES: To assess the analgesic efficacy, and adverse events, of non-steroidal anti-inflammatory drugs (NSAIDs) used to treat cancer-related pain in children and adolescents aged from birth and 17 years, in any setting. SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online, MEDLINE via Ovid, and Embase via Ovid from inception to 21 February 2017. We also searched the reference lists of retrieved studies and reviews, and searched online clinical trial registries. SELECTION CRITERIA:
Randomised, double-blind trials of any dose, and any route, treating cancer-related pain in children and adolescents, comparing NSAIDs with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for eligibility. We planned to use dichotomous data to calculate risk ratio and number needed to treat for one additional event, using standard methods. We assessed GRADE (Grading of Recommendations Assessment, Development and Evaluation) and planned to create a 'Summary of findings' table. MAIN RESULTS: No studies were eligible for inclusion in this review (very low quality evidence). We downgraded the quality of evidence by three levels due to the lack of data reported for any outcome. AUTHORS’ CONCLUSIONS: There is no evidence from randomised controlled trials that non-steroidal anti-inflammatory drugs (NSAIDs) reduce cancer-related pain in children and adolescents. This means that no reliance or conclusions can be made about efficacy or harm in the use of NSAIDs to treat chronic cancer-related pain in children and adolescents.


BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization guidelines for pharmacological treatments for children’s persisting pain acknowledge that pain in children is a major public health concern of high significance in most parts of the world. While in the past pain was largely dismissed and was frequently left untreated, views on children’s pain have changed over time and relief of pain is now seen as important. We designed a suite of seven reviews on chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol) in order to review the evidence for children’s pain utilising pharmacological interventions. As the leading cause of morbidity in the world today, chronic disease (and its associated pain) is a major health concern. Chronic pain (that is pain lasting three months or longer) can arise in the paediatric population in a variety of pathophysiological classifications (nociceptive, neuropathic, or idiopathic) from genetic conditions, nerve damage pain, chronic musculoskeletal pain, and chronic abdominal pain, as well as for other unknown reasons. Antidepressants have been used in adults for pain relief and pain management since the 1970s. The clinical impression from extended use over many years is that antidepressants are useful for some neuropathic pain symptoms, and that effects on pain relief are divorced and different from effects on depression; for example, the effects of tricyclic antidepressants on pain may occur at different, and often lower, doses than those on depression. Amitriptyline is one of the most commonly used drugs for treating neuropathic pain in the UK. OBJECTIVES: To assess the analgesic efficacy and adverse events of antidepressants used to treat chronic non-cancer pain in children and adolescents aged between birth and 17 years, in any setting. SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online, MEDLINE via Ovid, and Embase via Ovid from inception to 6 September 2016. We also searched the reference lists of retrieved studies and reviews, and searched online clinical trial registries. SELECTION CRITERIA: Randomised controlled trials, with or without blinding, of any dose and any route, treating chronic non-cancer pain in children and adolescents, comparing any antidepressant with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for eligibility. We planned to use dichotomous data to calculate risk ratio and number needed to treat for one additional event, using standard methods. We assessed the evidence using GRADE and created three ‘Summary of findings’ tables. MAIN RESULTS: We included four studies with a total of 272 participants (6 to 18 years of age) who had either chronic neuropathic pain, complex regional pain syndrome type 1, irritable bowel syndrome, functional abdominal pain, or functional dyspepsia. All of the studies were small. One study investigated amitriptyline versus gabapentin (34 participants), two studies investigated amitriptyline versus placebo (123 participants), and one study investigated citalopram versus placebo (115 participants). Due to a lack of available data we were unable to complete any quantitative analysis. Risk of bias for the four included studies varied, due to issues with randomisation and allocation concealment (low to unclear risk); blinding of participants, personnel, and outcome assessors (low to unclear risk); reporting of results (low to unclear risk); and size of the study populations (high risk). We judged the remaining domains, attrition and other potential sources of bias, as low risk of bias. Primary outcomes No studies reported our primary outcomes of participant-reported pain relief of 30% or greater or 50% or greater (very low-quality evidence). No studies reported on Patient Global Impression of Change (very low-quality evidence). We rated the overall quality of the evidence (GRADE rating) as very low. We downgraded the quality of the evidence by three levels to very low because there was no evidence to support or refute. Secondary outcomes All studies measured adverse events, with very few reported (11 out of 272 participants). All but one adverse event occurred in the active treatment groups (amitriptyline, citalopram, and
Chronic pain and its correlates are important problems for adolescents with physical disabilities. These comorbidities of physical disability and chronic pain are not related to depression, anxiety, or insomnia. Evaluation of physical disability status and chronic pain in relation to depressive symptoms, anxiety, or insomnia. INTERPRETATION: Adolescents with physical disabilities experience chronic pain at a significantly higher rate than able-bodied peers, but the prevalence and specific associated factors are unknown. The aims of this study were to determine (1) the prevalence of chronic pain in adolescents with physical disabilities and (2) whether known correlates of chronic pain in the general population are also present in young people both with physical disability and with chronic pain relative to peers. METHOD: We conducted a secondary analysis of cross-sectional nationally representative data from the National Longitudinal Study of Adolescent to Adult Health. Multivariate linear regression analysis was used to identify demographic and psychosocial factors associated with chronic pain. RESULTS: A total of 989 (4.3%) adolescents reported physical disabilities. They had a significantly higher rate of pain (27.2%) compared with able-bodied peers (15.6%, chi(2) = 86.3550, p < 0.001). There was no significant interaction between physical disability status and chronic pain in relation to depressive symptoms, anxiety, or insomnia. INTERPRETATION: Adolescents with physical disabilities experience chronic pain at a significantly higher rate than able-bodied peers, but the comorbidity of physical disability and chronic pain is not related to depression, anxiety, or insomnia. Evaluation of chronic pain and tailored pain interventions need to be developed for this population. WHAT THIS PAPER ADDS: Chronic pain and its correlates are important problems for adolescents with physical disabilities. These
adolescents present with higher rates of chronic pain than other young people. Chronic pain is associated with increased levels of depressive symptoms, anxiety, and insomnia regardless of disability status.  


Ibuprofen is the most widely used non-steroidal anti-inflammatory drug (NSAID) for the treatment of inflammation, mild-to-moderate pain and fever in children, and is the only NSAID approved for use in children aged >/=3 months. Its efficacy and safety profile have led to its increasing use in paediatric care, even without medical prescription. However, an increase of suspected adverse reactions to ibuprofen has been noted in concomitance with the raised, often medically unsupervised, consumption of the drug. The purpose of this work was a critical review of the paediatric literature over the last 15 years on side effects and adverse events associated with ibuprofen, in order to highlight circumstances associated with higher risks and to promote safe and appropriate use of this drug. The literature from 2000 to date demonstrates that gastrointestinal events are rare, but (when they occur) include both upper and lower digestive tract lesions. Dehydration plays an important role in triggering renal damage, so ibuprofen should not be given to patients with diarrhoea and vomiting, with or without fever. Likewise, ibuprofen should never be administered to patients who are sensitive to it or to other NSAIDs. It is contraindicated in neonates and in children with wheezing and persistent asthma and/or during varicella. Most of the analysed studies reported adverse events when ibuprofen was being used for fever symptoms or flu-like syndrome. Ibuprofen should not be used as an antipyretic, except in rare cases. Ibuprofen remains the drug of first choice in the treatment of inflammatory pain in children.  


The aim of this study was to assess the efficacy of self-hypnosis in a therapeutic education program (TEP) for the management of chronic pain in 26 children aged 7 to 17 years. Outcomes of the study were a total or a partial (at least 1) achievement of the therapeutic goals (pain, quality of sleeping, schooling, and functional activity). Sixteen patients decreased their pain intensity, 10 reached all of their therapeutic goals, and 9 reached them partially. Self-hypnosis was the only component of the TEP associated with these improvements. The current study supports the efficacy of self-hypnosis in our TEP program for chronic pain management in children. 


AIM: This study described end-of-life care for children affected by spinal muscular atrophy type 1 (SMA1), which is characterised by progressive muscle weakness and develops in the first six months of life. METHODS: We retrospectively analysed 17 children (13 boys) who attended the University of Padua’s paediatric palliative care centre in Italy from March 2000 to March 2015. All the children received supportive care without proactive respiratory intervention to prolong survival. RESULTS: The median age at admission was 3.57 months, and the median age at death was 6.80 months. The most frequent symptoms were dyspnoea and pain. In the last 72 hours of life, 15/17 children required more intense doses of morphine and/or benzodiazepines for intractable dyspnoea and pain, but deep palliative sedation was not needed. Airway suction to manage secretions and nasogastric tubes was required in all cases. The place of death was previously planned by the parents in all cases - home, hospital or hospice - and 15/17 deaths occurred in that place. We also interviewed 16 of the 17 parents after their child died. CONCLUSION: Our study found that symptom management and psychological support for families were the cornerstones of end-of-life care for children with SMA1.  

BACKGROUND: Pain is a common feature of childhood and adolescence around the world, and for many young people, that pain is chronic. The World Health Organization guidelines for pharmacological treatments for children’s persisting pain acknowledge that pain in children is a major public health concern of high significance in most parts of the world. While in the past pain was largely dismissed and was frequently left untreated, views on children’s pain have changed over time, and relief of pain is now seen as important. We designed a suite of seven reviews on chronic non-cancer pain and cancer pain (looking at antidepressants, antiepileptic drugs, non-steroidal anti-inflammatory drugs, opioids, and paracetamol) in order to review the evidence for children’s pain utilising pharmacological interventions. As the leading cause of morbidity in the world today, chronic disease (and its associated pain) is a major health concern. Chronic pain (that is pain lasting three months or longer) can arise in the paediatric population in a variety of pathophysiological classifications (nociceptive, neuropathic, or idiopathic) from genetic conditions, nerve damage pain, chronic musculoskeletal pain, and chronic abdominal pain, as well as for other unknown reasons. Non-steroidal anti-inflammatory drugs (NSAIDs) are used to treat pain, reduce fever, and for their anti-inflammatory properties. They are commonly used within paediatric pain management. Non-steroidal anti-inflammatory drugs are currently licensed for use in Western countries, however they are not approved for infants under three months old. The main adverse effects include renal impairment and gastrointestinal issues. Common side effects in children include diarrhoea, headache, nausea, constipation, rash, dizziness, and abdominal pain. OBJECTIVES: To assess the analgesic efficacy and adverse events of NSAIDs used to treat chronic non-cancer pain in children and adolescents aged between birth and 17 years, in any setting.

SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online, MEDLINE via Ovid, and Embase via Ovid from inception to 6 September 2016. We also searched the reference lists of retrieved studies and reviews, as well as online clinical trial registries. SELECTION CRITERIA: Randomised controlled trials, with or without blinding, of any dose and any route, treating chronic non-cancer pain in children and adolescents, comparing any NSAID with placebo or an active comparator. DATA COLLECTION AND ANALYSIS: Two review authors independently assessed studies for eligibility. We planned to use dichotomous data to calculate risk ratio and number needed to treat for one additional event, using standard methods. We assessed GRADE and created three ‘Summary of findings’ tables. MAIN RESULTS: We included seven studies with a total of 1074 participants (aged 2 to 18 years) with chronic juvenile polyarthritis or chronic juvenile rheumatoid arthritis. All seven studies compared an NSAID with an active comparator. None of the studies were placebo controlled. No two studies investigated the same type of NSAID compared with another. We were unable to perform a meta-analysis. Risk of bias varied. For randomisation and allocation concealment, one study was low risk and six studies were unclear risk. For blinding of participants and personnel, three studies were low risk and four studies were unclear to high risk. For blinding of outcome assessors, all studies were unclear risk. For attrition, four studies were low risk and three studies were unclear risk. For selective reporting, four studies were low risk, two studies were unclear risk, and one study was high risk. For size, three studies were unclear risk and four studies were high risk. For other potential sources of bias, seven studies were low risk. Primary outcomes Three studies reported participant-reported pain relief of 30% or greater, showing no statistically significant difference in pain scores between meloxicam and naproxen, celecoxib and naproxen, or rofecoxib and naproxen (P > 0.05) (low-quality evidence). One study reported participant-reported pain relief of 50% or greater, showing no statistically significant difference in pain scores between low-dose meloxicam (0.125 mg/kg) and high-dose meloxicam (0.25 mg/kg) when compared to naproxen 10 mg/kg (P > 0.05) (low-quality evidence). One study reported Patient Global Impression of Change, showing ‘very much improved’ in 85% of ibuprofen and 90% of aspirin participants (low-quality evidence). Secondary outcomes All seven studies reported adverse events. Participants reporting an adverse event (one or more per person) by drug were: aspirin 85/202; fenoprofen 28/49; ibuprofen 40/45; indomethacin 9/30; ketoprofen 9/30; meloxicam 18/47; naproxen 44/202; and rofecoxib 47/209 (very low-quality evidence). All seven studies reported withdrawals due to adverse events. Participants withdrawn due to an adverse event by drug were: aspirin 16/120; celecoxib 10/159; fenoprofen 0/49; ibuprofen 0/45; indomethacin 0/30; ketoprofen 0/30; meloxicam 10/147; naproxen 17/285; and rofecoxib 3/209 (very low-quality evidence). All seven studies reported serious adverse events. Participants experiencing a serious adverse event by drug were: aspirin 13/120; celecoxib 5/159; fenoprofen 0/79; ketoprofen 0/30; ibuprofen 4/45; indomethacin 0/30; meloxicam 11/147; naproxen 10/285; and rofecoxib 0/209 (very low-quality evidence). There were few or no data for our remaining secondary outcomes: Carer Global Impression of Change; requirement for rescue analgesia; sleep duration and quality; acceptability of treatment; physical functioning as defined by validated scales; and quality of life as defined by validated scales (very low-quality evidence). We rated the overall quality of the evidence (GRADE rating) for our primary and secondary outcomes as very low because there were limited data from studies and no opportunity for a meta-analysis. AUTHORS’ CONCLUSIONS: We identified only a small number of studies, with insufficient data for analysis. As we could undertake no meta-analysis, we are unable to comment about efficacy or harm from the use of NSAIDs to treat chronic non-cancer pain in children and adolescents. Similarly, we cannot comment on our remaining secondary outcomes: Carer Global Impression of...

BACKGROUND: High rates of depression and anxiety have been consistently reported among patients suffering from chronic pain. Prescription opioids are one of the most common modalities for pharmacological treatment of pain, however in recent years medical marijuana (MM) has been increasingly used for pain control in the US and in several countries worldwide. The aim of this study was to compare levels of depression and anxiety among pain patients receiving prescription opioids and MM. METHODS: Participants were patients suffering from chronic pain treated with prescription opioids (OP, N=474), MM (N=329) or both (OPMM, N=77). Depression and anxiety were assessed using the depression module of the Patient Health Questionnaire (PHQ-9) and the Generalized Anxiety Disorder scale (GAD-7). RESULTS: Prevalence of depression among patients in the OP, MM and OPMM groups was 57.1%, 22.3% and 51.4%, respectively and rates of anxiety were 48.4%, 21.5% and 38.7%, respectively. After controlling for confounders, patients in the OP group were significantly more likely to screen positive for depression (Adjusted Odds Ratio (AOR)=6.18; 95% CI=4.12-9.338) and anxiety (AOR=4.12; CI=3.84-5.71) compared to those in the MM group. Individuals in the OPMM group were more prone for depression (AOR for depression=3.34; CI=1.52-7.34) compared to those in the MM group. LIMITATIONS: Cross-sectional study, restricting inference of causality. CONCLUSIONS: Levels of depression and anxiety are higher among chronic pain patients receiving prescription opioids compared to those receiving MM. Findings should be taken into consideration when deciding on the most appropriate treatment modality for chronic pain, particularly among those at risk for depression and anxiety.


Pediatric opioid and benzodiazepine withdrawal are avoidable complications of pain and sedation management that is well described in the literature. To prevent withdrawal from occurring, practitioners regularly use a steady decrease of pain and sedation medications, also known as a weaning or tapering schedule. The weaning schedule is highly variable based on clinician preference and is usually dependent on the clinician. The purposes of this review are to evaluate the current literature on the process of opioid and benzodiazepine weaning in pediatric patients and to assess the various standardized protocols used to decrease withdrawal occurrences. We conducted a search of the PubMed, MEDLINE, Cochrane Library, Cumulative Index of Nursing and Allied Health (CINAHL), Academic Search Premier, and PsycInfo databases. Studies were included if they described a wean or taper in pediatric patients aged 18 years or younger. Studies describing neonatal abstinence syndrome were excluded from the review. A total of 97 studies published between 2000 and 2014 were retrieved; of those, 15 studies met the inclusion criteria. Studies were evaluated for selection of withdrawal assessment tool, wean protocol summary, preferred weaning agents, benzodiazepine withdrawal, and wean-at-home regimen. The most common opioid-weaning protocol approaches described a 10-20% dose decrease per day. Benzodiazepine weaning was not regularly standardized or described. The use of a standardized opioid-weaning protocol reduced withdrawal rates compared with nonstandardized weaning plans. Benzodiazepine weaning was inconsistently evaluated and may have affected study outcomes. Identified areas of improvement include the use of newer withdrawal assessment tools validated in the older pediatric population and standardized withdrawal assessment and reporting.


A multisite cross sectional study was conducted to examine dyadic friendship features between adolescents with chronic pain and their friends compared to non-pain adolescent friendship dyads and the association of these friendship features with loneliness and depressive symptoms. Participants completed a battery of standardized measures to capture friendship features (friendship quality, closeness, perceived social support from friends) and indices of social-emotional well-being. Sixty-one same sex friendship dyads (122 adolescents) participated; 30
friendship dyads included an adolescent with chronic pain and 52 dyads were female. Adolescents with chronic pain scored significantly higher on measures of loneliness and depressive symptoms compared to all other participants. Hierarchical Multiple Regression analysis revealed that friendship features predicted loneliness and depressive symptoms. Chronic pain predicted loneliness and depressive symptoms above and beyond friendship features. Actor Partner Interdependence Modeling found perceived social support from friends had differing associations on loneliness and depressive symptoms for dyads with a chronic pain member compared to pain-free control dyads. Friendship features were associated with loneliness and depressive symptoms for adolescents but friendship features alone did not explain loneliness and depressive symptoms for adolescents with chronic pain. Further research is needed to understand if pain-related social support improves loneliness and depressive symptoms for adolescents with chronic pain. Furthermore, a more nuanced understanding of loneliness in this population is warranted. Strategies to help adolescents with chronic pain garner needed social support from friends is needed to decrease rates of loneliness to improve long term outcomes.


Little is known about the prevalence, characterization and treatment of pain in children with progressive neurologic, metabolic or chromosomal conditions with impairment of the central nervous system. The primary aims of this study were to explore the differences between parental and clinical pain reporting in children with life-limiting conditions at the time of enrollment into an observational, longitudinal study and to determine if differences in pain experiences were associated with patient- or treatment-related factors. Pain was common, under-recognized and undertreated among the 270 children who enrolled into the "Charting the Territory" study. Children identified by their parents as experiencing pain (n=149, 55%) were older, had more comorbidities such as dyspnea/feeding difficulties, were less mobile with lower functional skills and used analgesic medications more often, compared to pain-free children. Forty-one percent of children with parent-reported pain (21.8% of all patients) experienced pain most of the time. The majority of clinicians (60%) did not document pain assessment or analgesic treatment in the medical records of patients who were experiencing pain. Documentation of pain in the medical record was positively correlated with children receiving palliative care services and being prescribed analgesics, such as acetaminophen, nonsteroidal anti-inflammatory drugs and opioids, as well as the adjuvant analgesics gabapentin and amitriptyline.


INTRODUCTION: Pain in children with intellectual disabilities (ID) is common and complex, yet there is no standard pain training for their secondary caregivers. OBJECTIVES: Determine perceived pain training needs/preferences of children’s respite staff (Phase One) and, use this information combined with extant research and guidelines to develop and pilot a training (Phase Two). METHODS: In Phase One, 22 participants responded to questionnaires and engaged in individual interviews/focus groups about their experiences with pain in children with ID, and perceived training needs/preferences. In Phase Two, 50 participants completed knowledge measures and rated the feasibility of, and their own confidence and skill in, pain assessment and management for children with ID immediately before and after completing a pain training. They also completed a training evaluation. RESULTS: Participants viewed a pain training as beneficial. Their ideal training involved a half-day, multifaceted in person program with a relatively small group of trainees incorporating a variety of learning activities, and an emphasis on active learning. Phase Two results suggested that completion of the 3-3.5 hour pain training significantly increased respite workers’ pain-related knowledge (large effect sizes: r=0.81-0.88), as well as their ratings of the feasibility of, and their own confidence and skill in, pain assessment and management in children with ID (moderate to large effect sizes: r=0.41-0.70). The training was rated favorably. DISCUSSION: Training can positively impact respite workers’ knowledge and perceptions about pain assessment and management. As such, they may be better equipped to care for children with ID in this area.


BACKGROUND: Few clinical trials evaluating the efficacy of oral sweet solutions for procedures in the emergency department (ED) have been published. OBJECTIVES: To compare the efficacy of an oral sucrose solution vs. a placebo in reducing pain in infants undergoing venipuncture without cannulation. METHODS: A randomized, double-blinded clinical trial was conducted in a pediatric ED. Infants 1 to 3 months old were randomly allocated to receive 2 mL of 88% sucrose or 2 mL of placebo, 2 min prior to venipuncture. The outcome measures were the difference in pain levels as assessed by the Face, Legs, Activity, Cry and Consolability Pain Scale (FLACC) and Neonatal Infant Pain Scale (NIPS) scores, crying time, and variations in heart rate. RESULTS: Eighty-two participants were recruited. Data were analyzed for 38 patients from each group (excluding protocol deviations). The mean difference in FLACC scores 1 min post venipuncture compared with baseline was 2.84 +/- .64 (sucrose) vs. 2.71 +/- .62 (placebo) (p = 0.98). For the NIPS score, it was 2.32 +/- .47 (sucrose) vs. 1.63 +/- .49 (placebo) (p = 0.60). The difference in the median crying time was not statistically significant between the two groups: 63.0 +/- 3 (sucrose) vs. 48.5 +/- 5 s (placebo) (p = 0.17). No significant difference was found in participants' heart rates 1 min post venipuncture compared with baseline: 33 +/- 6 (sucrose) vs. 24 +/- 5 beats per minute (placebo) (p = 0.44). CONCLUSIONS: In infants 1 to 3 months of age undergoing simple venipuncture, administration of an oral sweet solution did not statistically decrease pain scores, and participants' heart rate variations and crying time were not significantly changed.


Cannabinoid hyperemesis syndrome (CHS) is an underrecognized diagnosis among adolescents. In the adult literature, it is characterized as nausea, vomiting, and abdominal pain in patients with chronic marijuana use. CHS is often refractory to the standard treatment of nausea and vomiting. Unconventional antiemetics, such as haloperidol, have been successful in alleviating symptoms; however, even 1 dose of haloperidol can lead to grave adverse effects, such as dystonia, extrapyramidal reactions, and neuroleptic malignant syndrome. The use of topical capsaicin cream to treat CHS has been well described in the adult literature. This treatment is cost-effective and is associated with few serious side effects. Here, we describe 2 adolescent patients with nausea, vomiting, and abdominal pain in the setting of chronic cannabis use whose symptoms were not relieved by standard antiemetic therapies, but who responded well to topical capsaicin administration in our pediatric emergency department. We also discuss the pathophysiology behind capsaicin’s efficacy. These are the first reported cases in which capsaicin was successfully used to treat CHS in pediatric patients.


OBJECTIVE: There is a need to identify safe and effective opioid-sparing multimodal alternative treatment strategies and approaches, including topical analgesics, for opioid-experienced chronic pain patients to mitigate the risk of addiction, misuse, and abuse of opioids. METHODS: This subset analysis from a prospective, observational study evaluated changes in opioid use, other concurrent medication use, and pain severity and interference in opioid-experienced patients (OEP) treated with topical analgesics for chronic pain with measures obtained at baseline and 3- and 6-month follow-up. RESULTS: The 3-month opioid-experienced patient (3-month OEP) group included 121 patients who completed baseline and 3-month follow-up assessments; 27 opioid-experienced patients completed baseline and 6-month follow-up assessments (6-month OEP). Demographic characteristics, and mean pain severity and interference scores were similar between groups at baseline. After treatment with topical analgesics, 49% of patients in the 3-month and 56% of patients in the 6-month group reported they had completely discontinued use of opioids. In addition, 31% of patients at the 3-month assessment and 30% at the 6-month assessment reported that they were no longer taking any pain medication. Other concurrent medications decreased by 65% after 3 months, and 74% after 6 months. There were statistically significant decreases from baseline in pain severity and interference scores within the 3- (CI:0.7-1.4, 1.4-2.2) and 6-month (CI:0.7-2.4 (severity); CI:1.2-3.5 (interference)) OEP groups. CONCLUSIONS: Opioid use and other concurrent medications decreased among opioid-experienced chronic pain patients after 3- and 6-months.
of treatment with topical analgesics. Pain severity and interference scores also decreased. The topical analgesics were reported to be effective and safe for the treatment of chronic pain, with randomized controlled trials needed to confirm these findings.


Objective: Virtual reality (VR) is an exciting new technology with almost endless possible uses in medicine. One area it has shown promise is pain management. This selective review focused on studies that gave evidence to the distraction or nondistraction mechanisms by which VR leads to the treatment of pain. Methods: The review looked at articles from 2000 to July 29, 2016, focusing on studies concerning mechanisms by which virtual reality can augment pain relief. The data was collected through a search of MEDLINE and Web of Science using the key words of "virtual reality" and "pain" or "distraction." Results: Six studies were identified: four small randomized controlled studies and two prospective/pilot studies. The search results provided evidence that distraction is a technique by which VR can have benefits in the treatment of pain. Both adult and pediatric populations were included in these studies. In addition to acute pain, several studies looked at chronic pain states such as headaches or fibromyalgia. These studies also combined VR with other treatment modalities such as biofeedback mechanisms and cognitive behavioral therapy. Conclusions: These results demonstrate that in addition to distraction, there are novel mechanisms for VR treatment in pain, such as producing neurophysiologic changes related to conditioning and exposure therapies. If these new mechanisms can lead to new treatment options for patients with chronic pain, VR may have the ability to help reduce opioid use and misuse among chronic pain patients. More studies are needed to reproduce results from prospective/pilot studies in large randomized control studies.

https://academic.oup.com/painmedicine/article-abstract/19/1/151/4100671?redirectedFrom=fulltext


PURPOSE: Opioid analgesics are commonly used to treat vaso-occlusive pain episodes in sickle cell disease (SCD), but comprehensive evidence characterizing opioid use in this patient population is limited. Our objective was to characterize opioid use patterns among SCD patients using a large nationwide database. METHODS: A large, US medical claims database was utilized to identify a cohort of 3882 SCD patients, and characteristics of opioid use were analyzed. Clinical variables including age, gender, medication use, health care utilization, and medical history were evaluated for correlations with opioid use. RESULTS: Forty percent of patients took opioid medications during a 12-month span, and the prevalence of any opioid use was highest for 20 to 29-year-old patients (58%). The median daily opioid dose was 1.85 mg (interquartile range: 0.62-10.68 mg) oral morphine equivalents (OME). While most opioid users took between 0 and 5 mg OME daily, 3% of pediatric patients and 23% of adult patients used more than 30-mg OME daily. High-dose opioid use was associated with older age, hydroxyurea therapy, nonsteroidal anti-inflammatory drug (NSAID) use, and frequent inpatient hospitalizations. In multivariable-adjusted analyses, patients with vaso-occlusive complications such as pain crisis (OR = 3.8, 95% CI 2.7-5.3) and avascular necrosis (AVN) (OR = 3.7, 95% CI 2.7-5.1) were associated with high-dose opioid use. CONCLUSIONS: Our study showed that only 40% SCD patients were on opioid analgesics during a 12-month span. However, a non-trivial number of patients used a much higher dose of opioids despite a relatively low average daily opioid dose among SCD patients, particularly with vaso-occlusive complications.


OBJECTIVES: Controlling seizures in children approaching death can be difficult, and there is a limited evidence base to guide best practice. We compared current practice against the guidance for seizure management produced by the Association of Paediatric Palliative Medicine (APPM). METHODS: Retrospective case note review
of episodes of challenging seizure management in children receiving end-of-life care over a 10-year period (2006-2015) in the south-west region of England. RESULTS: We reviewed 19 admissions, in 18 individuals. Six (33%) had a malignancy, nine (50%) had a progressive neurodegenerative condition and three (17%) had a static neurological condition with associated epilepsy. Thirteen (72%) died in their local hospice, four (22%) at home, and one (6%) in hospital. Seventeen of 19 episodes involved the use of subcutaneous or intravenous midazolam infusion, for a mean of 11 days (range 3-27). There was a wide range of starting doses of midazolam, and 9/17 (53%) received final doses in excess of current dose recommendations. Six individuals received subcutaneous phenobarbital infusions, with four of these (67%) receiving final doses in excess of current dose recommendations. Plans for adjustments of infusion rates, maximal doses or alternative approaches should treatment fail were inconsistent or absent. In 16/18 (88%) cases seizures were successfully controlled prior to the day of the child's death. Staff found the experience of managing seizures at end of life challenging and stressful.

CONCLUSIONS: Pharmacological approaches to seizure management in end-of-life care are variable, often exceeding APPM dose recommendations. Despite this, safe and effective seizure control was possible in all settings.


Pain and distress in the paediatric palliative care population can be very difficult to manage. Clinical scenarios range from the acute management of cancer-related pain at the end of life to the ongoing long-term support of children with complex multimodal pain related to progressive neurological conditions. Understanding the child’s underlying condition, possible causes of pain and their preferred mode of communication are important to the delivery of holistic care. Modification of environmental factors, basic care consideration and non-pharmacological measures have a large role to play, alongside conventional analgesics. Medication may also need to be delivered by novel routes such as transdermal patches, continuous subcutaneous infusion of multiple drugs or transmucosal breakthrough analgesic doses. Two cases are used to illustrate approaches to these clinical problems.


Objective: Early childhood is a time of rapid development, particularly of the central nervous system, and can set a foundation for the entire life course. Complex pain in young children can impact the quality of life through limiting physical and social development, compromising psychological well-being, and disrupting sleep. The aim of this review is to identify the needs of young children who present to a tertiary-level pain service, what services they require, and their treatment outcomes. There are limited data on this vulnerable population, which may be due to small numbers represented and the complexities of pain assessment in this age group. Method: A retrospective chart review recorded demographics, gender, pain location and etiology, treatment, and outcomes of 28 children younger than age eight years attending a pediatric pain clinic over a three-year period. Results: All but two young children had an obvious physical pathology as an explanation for pain; this is in contrast to studies of pain clinics servicing adolescents. A diverse range of conditions, some rare, were identified, requiring a high level of pediatric understanding of the disease process and an ability to work with primary teams with expertise in disease-modifying strategies.


Epidermolysis bullosa comprises a range of conditions characterized by fragile skin with painful blistering induced by minor trauma and friction. The Dowling-Meara variant is a severe form characterized by disseminated painful blistering requiring lifelong skin and wound care. The natural history of the disease is characterized by a chronic course that tends to improve with advancing age. Various multimodal analgesic strategies have been proposed for painful procedures in children with epidermolysis bullosa. In this case report, we describe the use of nitrous oxide for pain control at home of blister treatments in a 4-year-old child with the Dowling-Meara variant.


OBJECTIVE: To evaluate the efficacy and safety of vigabatrin in pediatric epilepsy. METHODS: We retrospectively reviewed patients with epilepsy treated with vigabatrin over a 2-year period at a pediatric tertiary center. We assessed the relationship between seizure frequency, etiology, vigabatrin dose, adverse events, medication discontinuation reasons, and electroencephalography (EEG) characteristics. RESULTS: One hundred three patients followed at Boston Children’s Hospital were treated with vigabatrin and had complete medical records. Within the follow-up interval, 69 (67%) of 103 patients had discontinued vigabatrin therapy. Two patients (1.9%) died during therapy for unknown reasons. Median age at vigabatrin initiation was 8 months (interquartile range [IQR] 5-15). Median starting dose was 48.1 mg/kg per day (IQR 29.8-52.3) with a median target of 100 mg/kg (IQR 81.9-107.9). Median treatment duration was 12.1 months (n = 89, IQR 5.0-22.9) overall, and 13.3 months (IQR 5.2-23.2) for patients who discontinued vigabatrin. The most common reasons for discontinuation were controlled seizures in 31 (43.7%) of 71 and unsatisfactory therapeuetic effect in 23 (32.4%) of 71. Median percent seizure reduction from baseline to first follow-up was 83.3% (IQR 27.4-99.8) and 96.7% (IQR 43.3-100) to last follow-up. Twenty-four (38.7%) of 62 patients with a follow-up posttreatment remained seizure-free. Four patients who had initially achieved seizure freedom relapsed. Patients with structural/metabolic etiology had greater median percent seizure reduction at first follow-up than patients with genetic etiology (98.7% vs. 61.4%, respectively, p = 0.001). Hypsarrhythmia resolved after therapy in 18 of 20 (90%, 95% confidence interval [CI] 70-97) patients with pretreatment hypsarrhythmia, and 2 patients presented with hypsarrhythmia posttreatment. Risk of having hypsarrhythmia was reduced by 32% (95% CI 14.9-49.1) posttreatment. SIGNIFICANCE: Vigabatrin is efficacious in all seizure types and resolved hypsarrhythmia in most patients. In this series with a median treatment duration of 12.1 months, vigabatrin had a good safety profile with a low rate of discontinuation due to nonophthalmologic and ophthalmologic adverse effects.


Death from cancer is often painful. Usually, the pain can be relieved in ways that allow patients to remain awake and alert until the end. Sometimes, however, the only way to relieve pain is to sedate patients until they are unconscious. This method has been called palliative sedation therapy. Palliative sedation therapy is controversial because it can be misunderstood as euthanasia. We present a case in which an adolescent who is dying of leukemia has intractable pain. Experts in oncology, ethics, pain management, and palliative care discuss the trade-offs associated with different treatment strategies.


BACKGROUND: The benefits of patient-reported symptom assessment combined with integrated palliative care are well documented. This study assessed the symptom burden of palliative and curative-intent radiation oncology patients. PATIENTS AND METHODS: Prior to first consultation and at the end of RT, all adult cancer patients planned to receive fractionated percutaneous radiotherapy (RT) were asked to answer the Edmonton Symptom Assessment Scale (ESAS; nine symptoms from 0 = no symptoms to 10 = worst possible symptoms). Mean values were used for curative vs. palliative and pre-post comparisons, and the clinical relevance was evaluated (symptom values >/= 4). RESULTS: Of 163 participating patients, 151 patients (90.9%) completed both surveys (116 curative and 35 palliative patients). Before beginning RT, 88.6% of palliative and 72.3% of curative
patients showed at least one clinically relevant symptom. Curative patients most frequently named decreased general wellbeing (38.6%), followed by tiredness (35.0%), anxiety (32.4%), depression (30.0%), pain (26.3%), lack of appetite (23.5%), dyspnea (17.8%), drowsiness (8.0%) and nausea (6.1%). Palliative patients most frequently named decreased general wellbeing (62.8%), followed by pain (62.8%), tiredness (60.0%), lack of appetite (40.0%), anxiety (38.0%), depression (33.3%), dyspnea (28.5%), drowsiness (25.7%) and nausea (14.2%). At the end of RT, the proportion of curative and palliative patients with a clinically relevant symptom had increased significantly to 79.8 and 91.4%, respectively; whereas the proportion of patients reporting clinically relevant pain had decreased significantly (42.8 vs. 62.8%, respectively). Palliative patients had significantly increased tiredness. Curative patients reported significant increases in pain, tiredness, nausea, drowsiness, lack of appetite and restrictions in general wellbeing. CONCLUSION: Assessment of patient-reported symptoms was successfully realized in radiation oncology routine. Overall, both groups showed a high symptom burden. The results prove the need of systematic symptom assessment and programs for early integrated supportive and palliative care in radiation oncology.


AIM: To determine if differences exist between paediatric intensive care nurses and allied health professionals in empathy, secondary trauma, burnout, pain exposure and pain ratings of self and others. Early and late career differences were also examined. BACKGROUND: Nurses are routinely exposed to patient pain expression. This work context may make them vulnerable to adverse outcomes such as desensitization to patient pain or a compromise in personal well-being. DESIGN: Cross-sectional study. METHODS: Data were collected from a convenience sample of paediatric intensive care nurses (n = 27) and allied health professionals (n = 24), from September 2014-June 2015, at a Canadian health centre. Both groups completed one demographic and three behavioural scales. Participants underwent fMRI while rating the pain of infant and adult patients in a series of video clips. Data were analyzed using parametric and non-parametric methods. fMRI results are reported in a second paper. RESULTS: Nurses were significantly more likely to be exposed to pain at work than allied health professionals and scored significantly higher on dimensions of empathy, secondary trauma and burnout. Nurses scored their own pain and the pain of infant and adult patients, higher than allied health participants. Less experienced nurses had higher secondary trauma and burnout scores than more experienced nurses.

CONCLUSIONS: Paediatric intensive care work demands, such as patient pain exposure, may be associated with nurse’s higher report of empathy and pain in self and others, but also with higher levels of secondary trauma and burnout, when compared with allied health professionals.


Cognitive biases that emphasize bodily harm, injury, and illness could play a role in the maintenance of chronic pain, by facilitating fear and avoidance. Whereas extensive research has established attention, interpretation, and memory biases in adults with chronic pain, far less is known about these same biases in children and adolescents with pain. Studying cognitive biases in attention, interpretation, and memory in relation to pain occurring in youth is important because youth is a time when pain can first become chronic, and when relationships between cognitive biases and pain outcomes emerge and stabilize. Thus, youth potentially offers a time window for the prevention of chronic pain problems. In this article, we summarize the growing corpus of data that have measured cognitive biases in relation to pediatric pain. We conclude that although biases in attention, interpretation, and memory characterize children and adolescents with varying pain experiences, questions regarding the direction, magnitude, nature, and role of these biases remain. We call for independent extension of cognitive bias research in children and adolescents, using well powered longitudinal studies with wide age ranges and psychometrically sound experimental measures to clarify these findings and any developmental trends in the links between cognitive biases and pain outcomes. PERSPECTIVE: This article provides a rationale for the theoretical and practical importance of studying the role of cognitive biases in children and adolescents with chronic pain, which has to date, been relatively understudied. Existing findings are reviewed critically, and recommendations for future research are offered.


STUDY DESIGN: Retrospective comparative study. OBJECTIVE: The aim of this study was to demonstrate that intrathecal morphine (ITM) and oral analgesics provide effective pain control after posterior spinal fusion (PSF) for adolescent idiopathic scoliosis (AIS), and this protocol has a low complication rate so patients can be admitted to a general care floor. SUMMARY OF BACKGROUND DATA: Previous studies have shown that ITM combined with intravenous patient-controlled analgesia or epidural infusion (EPI) provides effective pain control after PSF for AIS. Owing to concerns for respiratory depression, ITM patients were routinely admitted to the intensive care unit (ICU) postoperatively. There are little data on ITM combined with oral analgesics. METHODS: We identified AIS patients aged 10 to 17 years who had undergone PSF. Twenty-eight patients who received ITM were matched to 28 patients who received a hydromorphone EPI. The ITM group received oral oxycodone starting at 16 hours postinjection. The EPI group received oxycodone after the epidural catheter was removed on postoperative day 2. Pain scores, adverse events, and length of stay were recorded. RESULTS: A higher number of EPI patients received fentanyl (11 vs. 3, P = 0.014) in the post-anesthesia care unit (PACU). The ITM group had lower pain scores between PACU discharge and midnight (mean 2.9 vs. 4.2, P = 0.034). Pain scores were similar during the remaining postoperative periods. All ITM patients transitioned to oxycodone without intravenous opioids. Time to ambulation (19.9 vs. 26.5 hours, P = 0.010) and Foley catheter removal (21.3 vs. 41.9 hours, P < 0.001) were earlier in the ITM patients. Length of hospital stay was shorter in the ITM group (3.1 vs. 3.5 days, P = 0.043). Adverse events occurred at similar rates in both groups. CONCLUSION: ITM and oral analgesics provide safe and effective pain control after PSF for AIS. Routine postoperative admission to the ICU is not necessary. LEVEL OF EVIDENCE: 3.


BACKGROUND: Studies addressing physical and psychosocial symptoms among hospitalized children and adolescents with cancer are limited. Understanding commonly occurring symptoms and their associated characteristics across the hospitalization is needed to guide symptom management strategies. OBJECTIVE: This study described the symptom experience of hospitalized children and adolescents with cancer. The study explored the frequencies of individual symptoms and the severity, duration, and associated distress of symptoms during the course of the hospitalization. METHODS: Participants completed the Memorial Symptom Assessment Scale 7-12 during each 12-hour shift of the 3-day/3-night data collection period. RESULTS: Participants were 50 children and adolescents (mean age, 12.6 years; range, 7.1-18.6 years) receiving inpatient chemotherapy. Participants reported a mean of 2.75 symptoms at each assessment point and a mean of 5.42 different symptoms during their hospitalization. Mixed model analyses identified a significant fixed effect for study day, with participants reporting fewer symptoms (F = 8.4, P < .01), less symptom severity (F = 5.81, P < .01), and shorter duration (F = 6.67, P < .01) on day 3 relative to days 1 and 2. A fixed effect for study day was not present for symptom distress. CONCLUSIONS: Children and adolescents receiving inpatient chemotherapy experience multiple physical and psychosocial symptoms of moderate or greater severity and duration throughout the course of their hospitalization. Symptoms of greatest severity may not be those that are most distressing to the patient. IMPLICATIONS FOR PRACTICE: Ongoing assessment that incorporates the multidimensional nature of symptoms is needed. Prioritizing interventions for symptoms that are most distressing to the patient may support a more meaningful, patient-centric approach to care.


PURPOSE: Adolescents and young adults (AYAs) with cancer experience multiple symptoms related to their cancer and its treatment which can negatively impact their development and quality of life. An understanding of the strategies AYAs use to self-manage their symptoms is limited. This study described symptom self-management strategies reported by AYAs with cancer using an iPad-based symptom heuristics tool, the Computerized Symptom Capture Tool. METHODS: The study used a cross-sectional, descriptive design. AYAs' free text responses relating their symptom self-management strategies were explored using qualitative content analysis procedures.
Methadone can cause a prolonged QTc interval that can lead to ventricular arrhythmias. The risk of methadone-induced prolongation of the QTc interval in children and young adults is unknown. The purpose of the study was to determine the frequency of QTc prolongation among pediatric and young adult patients with cancer pain on methadone treatment. METHODS: We retrospectively reviewed data for all patients on methadone during the one treatment. RESULTS: Seventy-two AYAs 13-29 years of age (mean 18.4 years) reported a total of 772 symptom self-management codes for 585 individual symptoms. These codes were organized into 119 distinct categories. These categories were further organized into 16 subthemes and 3 overarching themes: "Things I Take ... or Not" (n = 209 codes), "Physical Care Things I Do" (n = 367 codes), and "Psychosocial Care Things I Do" (n = 132 codes). AYAs frequently reported strategies from all three of the symptom self-management themes to manage individual symptoms; however, "medications" was the most frequently reported strategy. CONCLUSION: AYAs receiving chemotherapy use multiple common, yet uniquely individual symptom self-management strategies. AYAs' reported strategies range from those that involve shared management with a healthcare provider to those that AYAs implement independently. The study provides a foundation for future research to empower AYAs to engage in symptom self-management and to guide healthcare providers as they discuss developmentally relevant and evidence-based symptom self-management strategies.


Cannabis has been employed medicinally throughout history, but its recent legal prohibition, biochemical complexity and variability, quality control issues, previous dearth of appropriately powered randomised controlled trials, and lack of pertinent education have conspired to leave clinicians in the dark as to how to advise patients pursuing such treatment. With the advent of pharmaceutical cannabis-based medicines (Sativex/nabiximols and Epidiolex), and liberalisation of access in certain nations, this ignorance of cannabis pharmacology and therapeutics has become untenable. In this article, the authors endeavour to present concise data on cannabis pharmacology related to tetrahydrocannabinol (THC), cannabidiol (CBD) et al., methods of administration (smoking, vapourisation, oral), and dosing recommendations. Adverse events of cannabis medicine pertain primarily to THC, whose total daily dose-equivalent should generally be limited to 30mg/day or less, preferably in conjunction with CBD, to avoid psychoactive sequelae and development of tolerance. CBD, in contrast to THC, is less potent, and may require much higher doses for its adjunctive benefits on pain, inflammation, and attenuation of THC-associated anxiety and tachycardia. Dose initiation should commence at modest levels, and titration of any cannabis preparation should be undertaken slowly over a period of as much as two weeks. Suggestions are offered on cannabis-drug interactions, patient monitoring, and standards of care, while special cases for cannabis therapeutics are addressed: epilepsy, cancer palliation and primary treatment, chronic pain, use in the elderly, Parkinson disease, paediatrics, with concomitant opioids, and in relation to driving and hazardous activities.


BACKGROUND: Compared with nociceptive pain, neuropathic pain is a challenging diagnosis to make and successfully treat in children with cancer. OBJECTIVE: The objective of this case report was to see whether very-low-dose methadone (VLDM) (defined as <50% of accepted starting analgesic dose of methadone for children) would be an effective strategy to treat refractory neuropathic pain due to vincristine in two children with acute lymphoblastic leukemia. METHODS: This case report is based on the clinical experience and parent-reported outcomes of two children with refractory neuropathic pain who received VLDM. RESULTS: Based on parent/caregiver-reported outcome scores over a one-year period, both children's refractory neuropathic pain syndrome was successfully treated with the addition of VLDM to their pre-existing regimen of gabapentin. Neither child suffered any adverse effects from methadone. CONCLUSIONS: VLDM shows promise as an effective, safe, and inexpensive way to treat refractory neuropathic pain in children with cancer.


INTRODUCTION: A prolonged corrected QT (QTc) interval in pediatric patients is defined as >/=470 msec. Methadone can cause a prolonged QTc interval that can lead to ventricular arrhythmias. The risk of methadone-induced prolongation of the QTc interval in children and young adults is unknown. The purpose of the study was to determine the frequency of QTc prolongation among pediatric and young adult patients with cancer pain on methadone treatment. METHODS: We retrospectively reviewed data for all patients on methadone during the
study period. Qualifying patient data were reviewed to determine whether these patients had an electrocardiogram (ECG) while on methadone. The QTc values for analysis were manually calculated using the standard formula described by Bazett. RESULTS: Twenty-five patients were identified that met eligibility criteria. The median QTc decreased from baseline after initiation of methadone. QTc prolongation occurred in four of 25 (16%) patients and only one patient had a QTc greater than 500 msec. This patient had 17 normal QTc intervals on methadone prior to the prolongation. After resolution of electrolyte abnormalities, six subsequent ECGs on methadone had a normal QTc interval. CONCLUSIONS: Prolongation of the QTc interval is infrequent. The only observed case was transient during multiple comorbid conditions. A prospective study is justified to better understand what role methadone plays as one of many risk factors for prolongation of the QTc interval in children and young adults.


What are the most effective doses of simple oral analgesics such as paracetamol and ibuprofen for pain relief in children? Why can’t I prescribe codeine phosphate for children anymore? Is oral morphine really a safe alternative to codeine phosphate, and if so what dose should I prescribe? These questions are frequently asked by clinicians who wish to give analgesics to children for pain relief. In this article I will address these questions and describe a pragmatic approach for pain relief using oral analgesics based on the best evidence available and my experience as a consultant paediatric anaesthetist.


BACKGROUND: Children and infants with impaired swallow or compromised enteral absorption require alternative routes for administration of analgesia. Recent clinical guidance and practice for paediatric palliative care teams, who often treat such children, supports buccal morphine sulphate as a fast acting, effective and easily administered agent for pain relief. However, a consideration of the physicochemical properties and potency of morphine would suggest that it is not a suitable candidate for delivery via the transmucosal route, raising questions about its use in children and infants. AIM: To explore the permeability of buccal morphine sulphate in an established ex vivo porcine buccal mucosa as a necessary step in examining efficacy for use in children with life-limiting conditions and life-threatening illnesses. DESIGN: A permeation study conducted with morphine sulphate in an ex vivo porcine buccal tissue model. Flux values and pharmacokinetic data were used to calculate the plasma values of morphine that would result following buccal administration in a 20kg child. RESULTS: Results show that the estimated steady state plasma values of morphine sulphate following buccal administration in this model do not achieve minimum therapeutic concentration. CONCLUSION: These data strongly suggest that morphine sulphate is not suitable for buccal administration and that further research is needed to establish its efficacy in relief of pain in children with life-limiting conditions and life-threatening illnesses.


Most children with pain are managed by either acetaminophen or ibuprofen. However, no study has so far investigated if children are prescribed adequate doses of acetaminophen or ibuprofen in emergency department. Aim of this retrospective study was to investigate the prevalence of under-dosage of these drugs in children presenting with pain in emergency department. Children initially prescribed with acetaminophen or ibuprofen for pain management were included. The chi (2) automatic interaction detection method was used considering the percentage variation from the minimum of the appropriate dose as dependent variable while prescribed drug, age, gender, body weight, type of hospital (pediatric or general), and availability of internal guidelines on
pediatric pain management in the emergency department as independent variables. Data on 1471 children managed for pain were available. Under-dosage was prescribed in 893 subjects (61%), of whom 577 were prescribed acetaminophen and 316 ibuprofen. The use of acetaminophen suppositories, body weight <12 kg or >40 kg, and the use of oral ibuprofen identified clusters of children associated with under-dosage prescription. CONCLUSION: Prescription of acetaminophen and ibuprofen was frequently under-dosed. The use of suppositories, lower and higher body weight, and the use of ibuprofen were associated with under-dosage. Under-dosing may reflect prescription of anti-pyretic doses.

CLINICAL TRIAL REGISTRATION: Agenzia Italiana del Farmaco-Observational Study Register (RSO). Registration code: PIERR/1 What is Known: • Pain is frequent in children presented to emergency department. • International recommendations on pain management are often not implemented. What is New: • Acetaminophen and ibuprofen were frequently underdosed in children prescribed for pain in the Italian emergency departments. • Under-dosage may be related to the habit of using acetaminophen and ibuprofen in the recommended range for fever treatment.


OBJECTIVE: Mechanical ventilation (MV) in preterm infants (PTI) causes discomfort. Whether it causes pain is controversial. Meta analysis reviews of published work on PTI during MV have shown no clinically significant impact of opioids on pain scales, and hence not recommended for routine use in neonatal intensive care units (NICUs). Similarly regular use of sedative midazolam is also not recommended. Therefore we hypothesized a downward trend in narcotics and sedatives used in MV of PTI in NICUs. This study aimed to assess trends of sedatives and narcotics use during MV of PTI in Canadian NICUs during 2004-2009. METHODS: PTI born at gestational age (GA) of <35 weeks requiring invasive MV for >24 hours were identified retrospectively from the Canadian Neonatal Network database for 2004-2009. PTI were excluded if moribund on admission, had major congenital anomalies, surgery (except laser eye surgery), necrotizing enterocolitis, chest tube or history of maternal narcotic abuse. PTI were classified according to whether they received any narcotics (morphine, fentanyl, methadone, sufentanil, meperidine, alfentyl and codeine) or sedatives (chloral hydrate, midazolam, lorazepam, phenobarbital, pentobarbital, ketamine and propofol) for >24 consecutive hours during MV. Trends of narcotics and sedatives were assessed using the Cochrane-Armitage Trend test separately for PTI born at <29 and 29-34 weeks of GA. RESULTS: Among 5638 study subjects, 2169 (38.5%) received narcotics and 897 (15.9%) received sedatives. The most common narcotics were morphine (62.2%) and fentanyl (63.8%) and sedatives were phenobarbital (44.9%) and chloral hydrate (44.2%). A significant decreasing trend (P<0.01) in the use of any sedatives during MV was observed in PTI <29 and 29-34 weeks of GA. However, the use of any narcotics during MV increased significantly (P=0.03) among PTI <29 weeks of GA, and no change in trend was detected for PTI born at 29-34 weeks of GA. CONCLUSIONS: The use of sedatives during MV in PTI born at <35 weeks of GA was positively affected, however the narcotics use during MV remained constant for PTI born at 29-34 weeks, and increased in extremely low GA group (less than 29 weeks) suggesting evidence based practice change was not observed during the study period.


Fatigue is among the most common, debilitating, and distressing symptoms associated with chronic condition in pediatric population. The purpose of this study was to identify non-pharmacological fatigue interventions in children and adolescents with cancer. For this, we carried out an integrative review of the literature from January 2000 to December 2016. A comprehensive search of four databases was conducted: Cumulative Index to Nursing and Allied Health Literature, Psychology Information, Medline via PubMed, and Web of Science. Randomized controlled trial, quasi-experimental, case-control and cohort studies were included in this review. Thirteen relevant studies were included for analysis. Seven papers reported positive outcomes for exercise, exercise plus leisure activities, healing touch and acupressure. In another six papers using exercise, exercise plus psychological intervention and massage, no effectiveness was found. Effective management of fatigue in children and adolescents is important but research in this area is limited, so the results of this review should be interpreted cautiously. Future researchers are encouraged to test the effective interventions in homogenous cancer populations and in other groups where fatigue is a common concern.


The case studies are written in this article to illustrate how methadone might be used for pain in the Indian context. These cases might be used for discussion in a multidisciplinary team, or for individual study. It is important to understand that pain requires a multidisciplinary approach as opioids will assist only with physical, i.e. neuropathic and nociceptive pain, but not emotional, spiritual, or relational pain or the pain of immobility. The social determinants of pain were included to demonstrate how emotional, relational, and psychological dimensions of pain amplify the physical aspects of pain. The case studies follow a practical step-wise approach to pain while undergoing cancer treatment, pain toward the end-of-life and needing longer acting opioid. Methadone in children, and methadone in conditions of opioid toxicity or where there is a need for absorption in the proximal intestine cases are included. [https://www.ncbi.nlm.nih.gov/pubmed/29497250](https://www.ncbi.nlm.nih.gov/pubmed/29497250)


AIMS AND OBJECTIVES: To examine empirical studies of musical stimulation and music therapy carried out with preterm infants and their parents published from 2010-2015. BACKGROUND: Prematurity constitutes a global health problem that can impact the development of the preterm infant and the well-being of the parents. Music-based interventions may benefit the infant, parents and their relationship. In our review, we distinguished between musical stimulation and music therapy, as we found no previous studies that had made this distinction. DESIGN: This is a narrative literature review. METHODS: A search was undertaken in PubMed, PsycINFO and Lilacs using the terms “music,” “music therapy,” “singing,” “prematurity” and “preterm.” Thirty studies were included and analysed according to the following categories: (i) aims of the study, (ii) participants, (iii) design, (iv) type of intervention, (v) assessment and measures and (vi) main results. RESULTS: The vast majority of the studies focused on the preterm infants and used an experimental design. Few studies carried out family-centred interventions, despite this having been noted as an important factor in effective interventions. Musical stimulation studies used more recorded music, whereas music therapy studies used more individualised interventions with live music. CONCLUSIONS: Both musical stimulation and music therapy demonstrated significant effects on preterm infants and their parents. However, compared to musical stimulation studies, interventions performed by music therapists provided more individualised care and tended to show greater effects on infants’ physiological and behavioural responses. RELEVANCE TO CLINICAL PRACTICE: Our review showed that music therapy interventions may provide individualised, effective and family-centred care. There is a significant need for these types of interventions in the neonatal intensive care unit (NICU). [https://www.ncbi.nlm.nih.gov/pubmed/28544065](https://www.ncbi.nlm.nih.gov/pubmed/28544065)


Objective: Hyperexcitability of the central nervous system plays an important role in the development and maintenance of chronic pain in adults. This knowledge has led to improved treatment strategies within this population. In children, however, research on the presence of central hyperexcitability is scarce. To further investigate this topic in children with chronic pain, there is a need for a clear literature overview. Design: Systematic review. Methods: The literature search was performed using the electronic databases PubMed and Web of Science. An article was considered eligible if it included children (age two to 12 years) diagnosed with chronic pain. Articles had to report original research outcomes related to central hyperexcitability, and a comparison with a healthy control group was necessary. Characteristics of the study sample, the assessment, and conclusions regarding central hyperexcitability were extracted from each included article. Results: Twelve case-control studies were included with moderate to good methodological quality (510 children with chronic pain and 670 healthy controls). After summarizing the articles’ results on indices of central hyperexcitability, we concluded that secondary hyperalgesia might be present in children with recurrent abdominal pain, juvenile fibromyalgia, and juvenile idiopathic arthritis. Preliminary evidence exists for altered cortical nociceptive processing in children with migraine and recurrent abdominal pain. Conclusions: Based on the results of this review, central hyperexcitability might be present in in several pediatric chronic pain conditions. Further research on other
manifestations of central hyperexcitability (e.g., bottom-up and top-down mechanisms and nociceptive brain changes) is necessary to provide firm evidence about its presence in children with chronic pain.


BACKGROUND: Status epilepticus seizures are distressing events for hospice and palliative care patients. Currently, rectal diazepam is the only abortive therapy approved by the U.S. Food and Drug Administration for seizures occurring out of hospital. However, transmucosal (buccal and intranasal) midazolam hydrochloride is a less expensive, equally effective, and a more socially acceptable alternative. OBJECTIVE: To explore the use of transmucosal midazolam in out-of-hospital hospice patients in the State of Alabama. DESIGN: A cross-sectional survey was used to explore hospice providers’ knowledge and use of transmucosal midazolam in clinical practice within Alabama. Setting Subjects: Hospice providers (physicians, nurses, and administrators) in the State of Alabama (n = 27). MEASUREMENTS: An electronic survey was used to elicit transmucosal midazolam use among hospice providers. RESULTS: Transmucosal midazolam has been documented throughout the literature and reported by expert clinicians as an efficacious, safe, and appropriate pharmaceutical intervention for the abortive treatment of seizures in adult and pediatric out-of-hospital patients. However, barriers to the use of transmucosal midazolam with hospice patients included unfamiliarity with transmucosal route and lack of provider orders. None of the participants reported transmucosal midazolam use in out-of-hospital hospice settings. CONCLUSION: Evidence in the literature supports the use of transmucosal midazolam; however, further research is necessary to understand and address barriers in a more diverse and generalizable population.


BACKGROUND: Dyspnea is one of the most frequent symptoms in children with complex chronic conditions (CCC) requiring palliative care. Although it is a subject of high importance, there has been little research on dyspnea in critically ill children. OBJECTIVE: The purpose of this systematic review was to investigate the prevalence and causes of dyspnea in children with CCC and to identify the current state of research on the measurements, treatments, and the evaluation of therapeutic interventions. METHODS: A systematic literature search for relevant literature from 1990 until the present was performed using the online database PubMed. Information about prevalence, pathophysiological mechanisms, measurement, and treatment of dyspnea was extracted from all 43 eligible publications. RESULTS: The prevalence ranged widely from 17% to 80%. Breathlessness was primarily attributed to a disease-specific pathophysiology. A multidimensional approach has not been reported. Assessment of dyspnea included eight tools using either subjective self- or proxy-ratings or objective measures. Evidence for the effectiveness of various treatment approaches was low. DISCUSSION: The prevalence rates for dyspnea could be generalized across all conditions and patient subgroups. The biopsychosocial-spiritual approach was not addressed by the studies. There is a lack of an adequate and validated measurement tool that can be applied to children of various ages and diagnoses, communication ability, and practicable across different settings. Most found treatment approaches lacked good evidence in children. CONCLUSION: Although the prevalence rate of dyspnea in pediatric palliative care is high, it has been poorly studied.


The neonatal intensive care unit is recognized as a stressful environment; the nature of caring for sick babies with uncertain outcomes and the need to make difficult decisions results in a work place where moral distress is prevalent. According to the prevailing definition, moral distress occurs when the provider believes that what is "done" is not the right course of action, with an element of constraint: the provider has no choice but to act this way. This can lead to adverse outcomes, including burnout and a change of career. Traditionally, moral distress was considered to represent a misuse of power that forced nurses (typically) to provide burdensome treatments they believed not in the patient's best interests. Today, with shared decision-making, it is rare for physicians to act in a purely paternalistic fashion and impose management strategies on a team and parents. However, in the grey zones, it is not unusual for individuals with different values to disagree on a course of treatment. Healthcare
professionals across all disciplines may feel constrained despite there being no identified misuse of power. We argue for a broader understanding of moral distress and an awareness that maladaptive responses to moral distress may result in “transference” of moral distress on to other healthcare professionals and even on to the families of babies for whom we have a duty of care. Strategies for dealing with moral distress exist. An appreciation of these dynamics will enable providers to reduce the negative impacts of moral distress while also using it as a vehicle for constructive discussion and progressive thought that will better serve our patients and our colleagues.


Children represent a vulnerable population in which management of nociceptive pain is complex. Drug responses in children differ from adults due to age-related differences. Moreover, therapeutic choices are limited by the lack of indication for a number of analgesic drugs due to the challenge of conducting clinical trials in children. Furthermore the assessment of efficacy as well as tolerance may be complicated by children’s inability to communicate properly. According to the World Health Organization, weak opioids such as tramadol and codeine, may be used in addition to paracetamol and ibuprofen for moderate nociceptive pain in both children and adults. However, codeine prescription has been restricted for the last 5 years in children because of the risk of fatal overdoses linked to the variable activity of cytochrome P450 (CYP) 2D6 which bioactivates codeine. Even though tramadol has been considered a safe alternative to codeine, it is well established that tramadol pharmacodynamic opioid effects, efficacy and safety, are also largely influenced by CYP2D6 activity. For this reason, the US Food and Drug Administration recently released a boxed warning regarding the use of tramadol in children. To provide safe and effective tramadol prescription in children, a personalized approach, with dose adaptation according to CYP2D6 activity, would certainly be the safest method. We therefore recommend this approach in children requiring chronic or recurrent nociceptive pain treatment with tramadol. In case of acute inpatients nociceptive pain management, prescribing tramadol at the minimal effective dose, in a child appropriate dosage form and after clear instructions are given to the parents, remains reasonable based on current data. In all other situations, morphine should be preferred for moderate to severe nociceptive pain conditions.

https://www.ncbi.nlm.nih.gov/pubmed/29556194


https://www.jpeds.com/article/S0022-3476(17)31625-6/abstract


PURPOSE: Dyspnea is a debilitating symptom commonly experienced by advanced cancer patients that can lead to negative effects on function and quality of life (QOL). The present study aims to determine the relationship between dyspnea and other Edmonton Symptom Assessment System (ESAS) symptoms in palliative cancer patients referred to a radiotherapy clinic. METHODS: The presence and severity of dyspnea was measured using the ESAS. All patients that visited a palliative radiotherapy clinic between 1999 to 2002 and 2006 to 2009 and completed the ESAS were included. ESAS scores and other demographic and clinical information were extracted from a prospectively collected database. Statistical tests including chi-squared tests, Spearman correlations, and multivariate analysis were conducted to explore the relationship between dyspnea, other ESAS items, and other demographic factors. Kaplan-Meier overall survival curves were generated based on dyspnea severity. RESULTS: One thousand three hundred forty-four patients were included in the dyspnea analysis; reported moderate or severe dyspnea. Dyspnea severity was significantly associated with eight other ESAS interference severities (p < 0.001). Upon multivariate analysis, greater severity of dyspnea was significantly related to higher ESAS scores for tiredness, nausea, depression, anxious, drowsiness, and poor appetite (p < 0.05). The actuarial median survival time was 6.57 months (95% CI 5.91-7.29 months). There were highly significant differences in overall survival between those with none, mild, and moderate dyspnea (p < 0.0001). CONCLUSION: Cancer patients often experience dyspnea along with a multitude of other symptoms. Moderate and severe dyspnea should be assessed and optimally managed to reduce functional and QOL debilitations. As presence of increased dyspnea severity is
associated with worse overall survival, interventions should occur at the end of life to reduce symptom burden in palliative patients.  


The objective of this work was to study the agreement between four pain intensity scales when administered electronically: the Numerical Rating Scale-11, the Faces Pain Scale-Revised, the Visual Analogue Scale and the Coloured Analogue Scale. In all, 180 schoolchildren between 12 and 19 years old participated in the study. They had to report the maximum intensity of their most frequent pain using the electronic versions of the four scales. Agreement was calculated using the Bland-Altman method. Results show that the electronic versions of Numerical Rating Scale-11, Coloured Analogue Scale and Visual Analogue Scale can be used interchangeably.  


OBJECTIVES: Pediatric obesity and chronic pain are 2 of the most significant public health crises affecting youth today. Despite the high number of youth experiencing both chronic pain and obesity, little research has been done examining their relationship. This study aims to both replicate and extend this research base. METHODS: A retrospective chart review of 99 patients presenting for evaluation in a pediatric pain clinic was conducted. Demographic information, including patient weight status, and self-report measures completed by both patients and their parents, including the Pain Frequency-Severity-Duration scale, the Functional Disability Inventory, and the Pain Catastrophizing Scale were examined. RESULTS: Abdominal pain was the most frequently reported primary pain diagnosis category, with headache, diffuse musculoskeletal, localized musculoskeletal, and back pain categories reported from greatest to least frequency. Results show that 29% of our sample was obese. Age was related to weight status such that older children were more likely to have a higher body mass index. Among school-aged children, a higher body mass index percentile was associated with greater parent-reported pain catastrophizing. Obese youth had higher parent-reported Functional Disability Inventory scores than those in the normal weight group. Post hoc comparisons identified that this finding was only significant for girls. Further, obese youth were more likely to have a longer pain duration than those classified as normal weight. DISCUSSION: The results of this study add to the growing literature regarding the importance of taking weight status into account when intervening with youth with chronic pain.  


BACKGROUND: Sudden breath-holding episodes during sleep in young children are potentially related to sudden infant death syndrome and other life-threatening events. Additionally, these episodes can negatively affect child's growth and development. CASE PRESENTATION: Here, we present 3 cases of preschool children with similar paroxysmal nocturnal waking events associated with choking that had different etiologies (nocturnal frontal lobe epilepsy, nocturnal gastroesophageal reflux disease, and parasomnia, respectively). CONCLUSIONS: It is important to take into consideration the fact that breath spells during sleep can occur as a rare manifestation of parasomnia due to gastroesophageal reflux or as a symptom of nocturnal frontal lobe epilepsy. Full video electroencephalography, polysomnography, and simultaneous gastric pH monitoring should be used for the differential diagnosis of sleep-related disorders, such as breath spells, in children.  


BACKGROUND: Palliative care is a rapidly evolving area of emergency medicine. With an estimated 5,000 to 10,000 baby boomers per day reaching retirement age, emergency departments (EDs) are treating more patients with chronic and serious disease. Palliative care offers comprehensive care for patients with advanced medical
illness, aims to alleviate suffering and improve quality of life, and plays an important role in caring for these patients in the ED. OBJECTIVES: We sought to increase the emergency physician’s knowledge of and comfort with symptom control in palliative and hospice patients. DISCUSSION: Having the skills to deliver efficient and appropriate palliative and hospice care is imperative for emergency physicians. Palliative care should be considered in any patient suffering from symptoms of a life-limiting illness, whereas hospice care should be considered in the patient with likely <6 months left to live. Palliative care is appropriate earlier in the course of disease, and is appropriate when the practitioner would not be surprised if the patient died in the next 2 years (“The Surprise Question”). This article discusses management in the ED of pain, nausea, dyspnea, agitation, and oral secretions in patients appropriate for hospice and palliative care. CONCLUSION: The need for palliative and hospice care in the ED is increasing, requiring that emergency physicians be familiar with palliative and hospice care and competent in the delivery of rapid symptom management in patients with severe and life-limiting disease.


Cancer-related pain continues to be a significant therapeutic challenge, made more difficult by contemporary opioid use and diversion concerns. Conventional treatment using a tiered approach of nonsteroidal antiinflammatory drugs (NSAIDs), opioids, and adjuvant agents is limited; and alternatives are needed for patients with rapidly progressing pain and those who develop hyperalgesia and tolerance to opioids. Ketamine, an N-methyl-d-aspartate (NMDA) selective antagonist, has historically been used for anesthesia in adult and pediatric populations but has also been investigated for depression, bipolar disorder, and general and postoperative pain management. As an analgesic, low-dose ketamine decreases morphine requirements and rates of nausea and vomiting, suggesting a potentially beneficial role in cancer-related pain. Ketamine is typically administered intravenously and has a rapid onset of action with a relatively short half-life (2-3 hours) but is inconvenient for use in an ambulatory setting. Oral bioavailability is low and erratic, limiting application of this route for chronic use. Intranasal administration has a number of potential advantages, including avoidance of first-pass hepatic metabolism, no need for venous access, ability to repeat doses quickly, and rapid absorption. Although early studies of intranasal ketamine are promising in a number of indications, information is more limited in its use as an adjunct in cancer-related pain. We review the background, rationale, pharmacokinetics, and clinical and safety data using intranasal ketamine as an adjunctive agent and its potential in cancer-related pain.


BACKGROUND: Admission in Pediatric Intensive Care Unit requires management and monitoring of analgesia and sedation, in order to reduce their adverse effects, and to prevent withdrawal syndrome and delirium. The aim of this study was to evaluate the management of analgesia and sedation in critically ill children admitted in the Italian Pediatric Intensive Care Units. METHODS: For this survey we have submitted a telematics questionnaire to 24 nursing coordinators of the Pediatric Intensive Care Units or Neonatal Intensive Care Units admitting critically ill children. RESULTS: Twenty Intensive Care Units (ICUs) replied to the questionnaire. The association of benzodiazepines and opioids was the first choice in 92.8% of analgesic and sedative strategies. Seventy percent of ICUs adopted a protocol for analgesic and sedative drugs used before performing invasive and/or painful procedures in critically ill children. Ninety percent of them followed a protocol for the assessment of pain, 75% adopting the Face, Legs, Activity, Cry, Consolability Scale. Sixty percent of ICUs followed a protocol for sedation, 58% used the Comfort Scale to monitor the level of it. Forty percent adopted a protocol for the withdrawal syndrome, 75% of them monitored the patients with the WAT-1 Scale. Ten percent of Pediatric Intensive Care Units followed a protocol for management of delirium; seventy-five percent did not monitor it. CONCLUSIONS: Despite an increasing sensitivity in pain and sedation management over the last five years, knowledge and monitoring of abstinence syndrome and delirium need to be improved.


Osteoporosis in childhood is uncommon, and it may be secondary to a spectrum of diverse conditions. Idiopathic juvenile osteoporosis is a primary osteoporosis of unknown aetiology present in previously well children and is a
diagnosis of exclusion. We describe a 10-year-old prepubertal boy who presented with back pain of 1-week duration. His spinal X-ray showed generalised loss of vertebral body heights in keeping with osteoporosis. Endocrine and haematological work-up were normal. He was treated with vitamin D supplement and intravenous pamidronate. This case illustrates the general work-up and causes for paediatric osteoporosis, and the management for idiopathic juvenile osteoporosis.


Status epilepticus is common in neonates and infants, and is associated with neuronal injury and adverse developmental outcomes. gamma-Aminobutyric acidergic (GABAergic) drugs, the standard treatment for neonatal seizures, can have excitatory effects in the neonatal brain, which may worsen the seizures and their effects. Using a recently developed model of status epilepticus in postnatal day 7 rat pups that results in widespread neuronal injury, we found that the GABAA agonists phenobarbital and midazolam significantly increased status epilepticus-associated neuronal injury in various brain regions. Our results suggest that more research is needed into the possible deleterious effects of GABAergic drugs on neonatal seizures and on excitotoxic neuronal injury in the immature brain. Ann Neurol 2017;82:115-120.


PURPOSE OF REVIEW: This review seeks to provide an update on the diagnosis, management, and outcome of pediatric delirium. RECENT FINDINGS: Care of patients with delirium depends on correct diagnosis and treatment of its underlying cause. A variety of instruments are available to aid diagnosis. Management of delirium currently depends on atypical antipsychotics, while avoiding agents that may precipitate or exacerbate it. While most critically ill children survive delirium, many children die or have worsening function after their illness. The longer the duration of delirium, the more severe its subsequent problems including postintensive care syndrome and posttraumatic stress disorder. Possible serious long-term consequences emphasize the importance of efforts to improve diagnosis and outcome in critically ill children suffering from delirium.


Americans have embraced a large number of diets in an attempt to manage obesity, improve quality of life, and address specific health problems. Among diets developed to address health problems, the ketogenic diet has had a long and variable history. Developed in the 1920s by a faith healer to help children with epilepsy, this diet induces a state that mimics carbohydrate starvation. As medications became available and effectively addressed seizures, the diet fell out of favor. During the last few decades, researchers and clinicians have learned that it can be useful in children and adults with refractory epilepsy and a variety of other conditions. Once again, pharmacists may encounter patients who are employing dietary management of serious health problems. This very high-fat diet almost eliminates carbohydrates from the patient’s food selection. The result is the substitution of ketone bodies as a source of energy. Today’s ketogenic diet has been modified with scientifically proven adjustments to increase palatability and help with adherence. Effective for some forms of epilepsy, the ketogenic diet also seems to have some utility in Alzheimer’s disease, Parkinson’s disease, and glaucoma, and many
Americans are using it to lose weight. Consultant pharmacists may field questions about this diet, its potential to correct or alleviate health conditions, and its limitations. The article discusses the ketogenic diet’s strengths, limitations, potential mechanisms, and use in a number of conditions with an emphasis on the elderly.


CONTEXT: Legalization of medical marijuana in many states has led to a widening gap between the accessibility and the evidence for cannabinoids as a medical treatment. OBJECTIVE: To systematically review published reports to identify the evidence base of cannabinoids as a medical treatment in children and adolescents. DATA SOURCES: Based on Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines, a search of PubMed, Medline, and the Cumulative Index to Nursing and Allied Health Literature databases was conducted in May 2017. STUDY SELECTION: Searching identified 2743 citations, and 103 full texts were reviewed. DATA EXTRACTION: Searching identified 21 articles that met inclusion criteria, including 22 studies with a total sample of 795 participants. Five randomized controlled trials, 5 retrospective chart reviews, 5 case reports, 4 open-label trials, 2 parent surveys, and 1 case series were identified. RESULTS: Evidence for benefit was strongest for chemotherapy-induced nausea and vomiting, with increasing evidence of benefit for epilepsy. At this time, there is insufficient evidence to support use for spasticity, neuropathic pain, posttraumatic stress disorder, and Tourette syndrome. LIMITATIONS: The methodological quality of studies varied, with the majority of studies lacking control groups, limited by small sample size, and not designed to test for the statistical significance of outcome measures. Studies were heterogeneous in the cannabinoid composition and dosage and lacked long-term follow-up to identify potential adverse effects. CONCLUSIONS: Additional research is needed to evaluate the potential role of medical cannabinoids in children and adolescents, especially given increasing accessibility from state legalization and potential psychiatric and neurocognitive adverse effects identified from studies of recreational cannabis use.


In recent years, the effect of music interventions and music therapy has experienced increased attention in the literature. It has been shown that music has positive effects on cognitive and physical performance, such as concentration and endurance, as well as on psychological parameters, such as anxiety and relaxation. Studies within the context of medicine in particular are increasingly indicating that music may be used as an intervention for relief against anxiety, stress and pain. Music is therefore seen in actual practice as a supplement to conventional pharmacological and non-pharmacological forms of treatment - and the trend is rising. Studies involving music interventions in the field of obstetrics have shown, amongst other things, that music improves the ability to relax during pregnancy and can reduce anxiety. It was also discovered that during childbirth music interventions resulted in a reduction of pain and stress. Music also has the effect of reducing stress, pain and anxiety in expectant mothers during deliveries by caesarean section. This review intends to provide an overview of the literature on music interventions in the field of obstetrics and to give a resume on the current state of research around the topic of music in relation to pregnancy, spontaneous deliveries and caesarean sections. Furthermore, the relevance of music for everyday obstetrics will be illustrated.


PURPOSE: The aim of this study was to explore how children aged 10-18 years describe their neuropathic pain (NP). METHOD: This is a qualitative descriptive study using inductive content analysis. Semi-structured interviews were conducted with eight children, aged 10-18 years with varying diagnoses, who were experiencing NP. RESULTS: All children were able to describe their NP using a variety of strategies, including use of literal and figurative language. While some sensory descriptors commonly reported by adults were used, descriptions of NP pattern and impact were also integral to their narratives. Children were able to differentiate NP from nociceptive pain. Parents clarified and gave context to pain reports. CONCLUSIONS: NP is a complex experience necessitating consideration of the different ways that children describe their symptoms. Involvement of parents is invaluable to
the process of taking a pain history with a child who is being screened for NP. Implications for Rehabilitation The findings of the study may inform the screening process for NP in children to facilitate earlier identification. Clinicians should consider the variety of ways that children may express their NP symptoms and the resulting impact. Clinicians should probe further when children report that symptoms are hard to describe or “weird”. Presence of a parent during the child’s pain assessment may assist with gathering a more complete picture. https://www.ncbi.nlm.nih.gov/pubmed/27684213


INTRODUCTION: Pediatric sickle cell disease, highly prevalent in sub-Saharan Africa, carries great morbidity and mortality risk. Limited resources and monitoring make management of acute vaso-occlusive crises challenging. This study aims to evaluate the efficacy and safety of subdissociative intranasal ketamine as a cheap, readily available and easily administered adjunct to standard pain therapy. We hypothesise that subdissociative, intranasal ketamine may significantly augment current approaches to pain management in resource-limited settings in a safe and cost-effective manner. METHODS AND ANALYSIS: This is a multicentre, randomised, double-blind, placebo-controlled trial enrolling children 4-16 years of age with sickle cell disease and painful vaso-occlusive pain crises. Study sites include two sub-Saharan teaching and referral hospitals with acute intake areas. All patients receive standard analgesic therapy during evaluation. Patients randomised to the treatment arm receive 1 mg/kg intranasal ketamine at onset of therapy, while placebo arm participants receive volume-matched intranasal normal saline. All participants and clinical staff are blinded to the treatment allocation. Data will be analysed on an intention-to-treat basis. Primary endpoints are changes in self-report pain scales (Faces Pain Scale-Revised) at 30, 60 and 120 minutes and rates of adverse events. Secondary endpoints include hospital length of stay, total analgesia use and quality of life assessment 2-3 weeks postintervention. ETHICS AND DISSEMINATION: The research methods for this study have been approved by the Cameroon Baptist Convention Health Board Institutional Review Board (IRB2015-07), the Tanzanian National Institute for Medical Research (NIMR/HQ/R.8a/Vol. IX/2299), Muhimbili National Hospital IRB (MNH/IRB/I/2015/14) and the Tanzanian Food and Drugs Authority (TFDA0015/CTR/0015/9). Data reports will be provided to the Data and Safety Monitoring Board (DSMB) periodically throughout the study as well as all reports of adverse events. All protocol amendments will also be reviewed by the DSMB. Study results, regardless of direction or amplitude, will be submitted for publication in relevant peer-reviewed journals. TRIAL REGISTRATION: ClinicalTrials.Gov, NCT02573714. Date of registration: 8 October 2015. Pre-results. http://bmjopen.bmj.com/content/7/7/e017190


Ibuprofen is a non-steroidal anti-inflammatory drug frequently administered to children of various ages for relief of fever and pain and is approved as an over-the-counter medication in many countries worldwide. Although there are extensive data on its efficacy and safety in children and adults, there are divergent dosing recommendations for analgesia and treatment of fever in infants, especially in the age group between 3 and 6 months of age. In this article, we have assessed the safety and efficacy of ibuprofen use in infants in an attempt to find the optimal method of pain and fever management in this specific age group. Based on the current evidence, short-term use of ibuprofen is considered safe in infants older than 3 months of age having a body weight above 5-6 kg when special attention is given to the hydration of the patient. Ibuprofen should be prescribed based on body weight using a dose of 5-10 mg/kg. This dose can be administered 3-4 times a day resulting in a maximum total daily dose of 30-40 mg/kg. The rectal route has been shown to be less reliable because of erratic absorption, especially in young infants. Since most efficacy and safety data have been derived from trials in infants with fever, future studies should focus on the efficacy of ibuprofen in young infants with pain. https://www.ncbi.nlm.nih.gov/pubmed/28516288
BACKGROUND: Systematic assessment of emotional distress is recommended in after care. Yet, it is unclear if parent report may be used as a proxy of child report. The aim of this study was to assess agreements and differences and explore possible moderators of disagreement between child and parent ratings. METHODS: Sixty-two young survivors treated for acute lymphoblastic leukemia (9-18 years) and both parents responded to the Beck Youth Inventory (anxiety and depression) and the distress rating scale on the child’s status. Parents completed the Brief Symptom Inventory-18 on their own psychological status. Systematic analyses of agreement and differences were performed. RESULTS: Mother-child and father-child agreements were fair on anxiety, depression, and distress (median intraclass correlation coefficient = 0.37). Differences between parents and children were medium sized (median d = 0.55) with parents giving higher scores than their children on anxiety, depression, and distress. Mothers reported distress more frequently than fathers (39 vs. 17%) when children reported none. The child being female and lower parental income were associated with lower agreement in fathers when rating child distress. Higher levels of parental psychological symptoms were consistently associated with lower agreement. CONCLUSIONS: Parent-child differences when rating adolescent survivors’ difficulties may be more important than previously thought. Parent report probably cannot be considered as a valid proxy of older child report on such internalized domains as anxiety, depression, or distress in the after-care clinic. Parents’ report is also likely to be influenced by their own mood, a factor that should be corrected for when using their report.


This study on end-of-life decisions in extremely preterm babies shows that the parents under study experience a multitude of stressors due to the immediate separation after birth, the alienating setting of the neonatal intensive care unit (NICU), the physical distance to the child, medical uncertainties, and upcoming decisions. Even though they are considered to be parents (assigned parenthood), they cannot act as primary caregivers. Instead, they depend on professional instructions for access and care. Embodied parenthood can be experienced only at the end-of-life, that is, during the dying trajectory and after the child’s death. Professionally supporting parents during this compressed process (from assigned and distant to embodied parenthood) contributes fundamentally to their perception of being a family and supports their mourning. This calls for the further establishment of palliative and bereavement care concepts in neonatology.


connection, personal growth, mentor role in mentee growth, and logistics of mentorship. Conclusions: Acting as a peer mentor online is a feasible and rewarding experience that supports the mentor’s own illness self-management, social connection, and personal growth.


BACKGROUND: Limited research has examined the impact of a child’s death from cancer on siblings. Even less is known about how these siblings change over time. OBJECTIVE: This study compared changes in siblings 1 (T1) and 2 (T2) years after the death of a brother or sister from cancer based on bereaved parent and sibling interviews. METHODS: Participants across 3 institutions represented 27 families and included bereaved mothers (n = 21), fathers (n = 15), and siblings (n = 26) ranging from 8 to 17 years old. Participants completed semistructured interviews. Content analysis identified emerging themes and included frequency counts of participant responses. McNemar tests examined differences in the frequency of responses between T1 and T2 data. RESULTS: Participants reported similar types of changes in bereaved siblings at both time points, including changes in sibling relationships, life perspectives, their personal lives, and school performance. A new theme of “openness” emerged at T2. Frequencies of responses differed according to mother, father, or sibling informant. Overall, participants less frequently reported changes at T2 versus T1. Compared with findings in the first year, participants reported greater sibling maturity at follow-up. CONCLUSION: Overall changes in bereaved siblings continued over 2 years with less frequency over time, with the exception of increases in maturity and openness. IMPLICATIONS FOR PRACTICE: Providers can educate parents regarding the impact of death of a brother or sister over time. Nurses can foster open communication in surviving grieving siblings and parents as potential protective factors in families going through their grief.

https://europepmc.org/abstract/med/29489479


BACKGROUND AND OBJECTIVES: This study aimed to examine forms of dyadic coping (DC) as mediators of the association between parents' grief response and dyadic adjustment and to determine whether these indirect effects were moderated by the child's type of death, timing of death, and age. DESIGN: The study design was cross-sectional. METHOD: The sample consisted of 197 bereaved parents. Participants completed the Prolonged Grief Disorder Scale, Revised Dyadic Adjustment Scale, and Dyadic Coping Inventory. RESULTS: Significant indirect effects of parents' grief response on dyadic adjustment were found through stress communication by oneself and by the partner, positive and negative DC by the partner, and joint DC. The timing of death moderated the association between grief response and dyadic adjustment and between joint DC and dyadic adjustment. Grief response was negatively associated with dyadic adjustment only when the death occurred after birth. Grief response was negatively associated with joint DC, which, in turn, was positively associated with dyadic adjustment, when the death occurred both before and after birth. However, the association was stronger in the latter. CONCLUSIONS: Specific forms of DC might be mechanisms through which grief response is associated with dyadic adjustment and should be promoted in clinical practice.


OBJECTIVE: Although the death of a child is a devastating event, recent evidence shows that personal growth is a relevant outcome of parents' grief. This study aimed to examine the factors associated with posttraumatic growth (PTG) and to propose a multidimensional model consisting of sociodemographic, situational, and intrapersonal and interpersonal factors. METHOD: A sample (N = 197; 89.8% female; mean age = 39.44 years) of bereaved parents completed the Post-Traumatic Growth Inventory-Short Form, the 14-Item Resilience Scale, the Continuing Bonds Scale, and the Dyadic Coping Inventory. RESULTS: The final model consisted of sociodemographic, situational, intrapersonal, and interpersonal factors of PTG, which accounted for 36.7% of the variance. Higher levels of PTG were generally associated with female sex, younger age of the child, higher levels of resilience, higher levels of internalized continuing bonds (i.e., internal representation of the child, maintaining psychological proximity), and higher levels of stress communication by the partner (communicating the stress experience and
requesting emotional or practical support). CONCLUSIONS: In clinical practice, health professionals assisting bereaved parents should pay attention to men and parents of older children, who might be at higher risk of difficulties in developing PTG. Additionally, promoting a more internalized bond with the child, resilience and dyadic coping, especially stress communication, can constitute important therapeutic goals. (PsycINFO Database Record


This paper explores the challenges of resolving conflicting feelings around talking with a child about their terminal prognosis. When children are left out of such conversations it is usually done with good intent, with a parent wishing to protect their child from anxiety or loss of hope. There is however growing evidence that sensitive, timely, age appropriate information from those with whom children have a good relationship is helpful both for the child and their family. There is no evidence that involving children in sensitive and timely discussions creates significant problems, rather that withholding information may lead to confusion, frustration, distress and anger. The authors discuss ways in which families can be supported to have these significant conversations with their children.
http://ep.bmj.com/content/102/4/182


Despite the potentially devastating effects of a death on the lives of adolescents, little is known about their help-seeking experiences. We interviewed by telephone 39 bereaved adolescents on their help-seeking experiences. Thematic analysis resulted in three themes: Formal support, Informal support and School-related support. Participants provided a critical appraisal of positive and negative experiences, and noted barriers and facilitators for help-seeking. As adolescents bereaved through suicide may receive less social support, professional help is a much-needed auxiliary. Parental encouragement is important in accessing adequate professional help.


Death of a relative or friend is a potentially disruptive event in the lives of adolescents. To provide targeted help, it is crucial to understand their grief and mental health experiences. Thematic analysis of 39 semistructured telephone interviews yielded two themes: Grieving apart together and Personal growth. High self-reliance and selective sharing were common. Feelings of guilt and “why” questions seemed more pronounced among the suicide bereaved. There was strong evidence of personal growth, increased maturity, and capacity to deal with personal mental health/suicidality. Despite its devastating effects, experiencing a death can be a catalyst for positive mental health.

Arnolds, M., L. Xu, P. Hughes, J. McCoy and W. Meadow (2018). "Worth a Try? Describing the Experiences of Families during the Course of Care in the Neonatal Intensive Care Unit When the Prognosis is Poor.” J Pediatr.

OBJECTIVE: To determine how parents of infants in the neonatal intensive care unit with a poor or uncertain prognosis view their experience, and whether they view their choices as "worth it," regardless of outcome. STUDY DESIGN: Parents of eligible neonates at 2 institutions underwent audiotaped, semistructured interviews while their infants were still in the hospital and then again 6 months to 1 year after discharge or death. Interviews were transcribed and data were analyzed using thematic analysis. Two authors independently reviewed and coded each interview and discrepancies were resolved by consensus. RESULTS: Twenty-six families were interviewed in the initial group and 17 families were interviewed in the follow-up group. The most common themes identified included realism about death (24 families), appreciation for the infant’s care team (23 families), and optimism and hope (22 families). Overall themes were very similar across both centers, and among parents of infants who died and those who survived. Themes of regret, futility, distrust of care team, and infant pain were brought up infrequently or not at all. CONCLUSIONS: No family believed that the care being provided to their infant was

**PURPOSE/OBJECTIVES:** To study the relationship between parental verbal and nonverbal caring behaviors and child distress during cancer-related port access placement using correlational and time-window sequential analyses. **DESIGN:** Longitudinal, observational design. **SETTING:** Children's Hospital of Michigan and St. Jude Children's Research Hospital. **SAMPLE:** 43 child-parent dyads, each with two or three video recordings of the child undergoing cancer-related port placement. **METHODS:** Two trained raters coded parent interaction behaviors and child distress using the Parent Caring Response Scoring System and Karmanos Child Coping and Distress Scale, respectively. Mixed modeling with generalized estimating equations examined the associations between parent interaction behaviors and parent distress, child distress, and child cooperation reported by multiple raters. Time-window sequential analyses were performed to investigate the temporal relationships in parent-child interactions within a five-second window. **MAIN RESEARCH VARIABLES:** Parent caring behaviors, child distress, and child cooperation. **FINDINGS:** Parent caring interaction behaviors were significantly correlated with parent distress, child distress, and child cooperation during repeated cancer port accessing. Sequential analyses showed that children were significantly less likely to display behavioral and verbal distress following parent caring behaviors than at any other time. If a child is already distressed, parent verbal and nonverbal caring behaviors can significantly reduce child behavioral and verbal distress. **CONCLUSIONS:** Parent caring behaviors, particularly the rarely studied nonverbal behaviors (e.g., eye contact, distance close to touch, supporting/allowing), can reduce the child's distress during cancer port accessing procedures. **IMPLICATIONS FOR NURSING:** Studying parent-child interactions during painful cancer-related procedures can provide evidence to develop nursing interventions to support parents in caring for their child during painful procedures.


When a child has a life-limiting illness, parental involvement is amplified, having to respond to the increased needs of the child. Both parents are affected by the illness, yet research has largely under-represented fathers' experiences of their child's illness. Seven fathers were interviewed about their experiences with their child's life-limiting illness. In addition, fathers' attachment strategies were assessed using the Adult Attachment Interview. Narrative analysis was implemented to explore the interviews, and indicators of attachment markers employed in the Adult Attachment Interview were also identified. The dominant themes were found to be 'experience of the diagnosis', 'living with the illness', 'struggling with emotions' and 'relationship with staff'. Within each theme, there were differences which related to the father's attachment strategies. This was particularly evident in parts of their narratives recounting critical moments of threat and anxiety in the course of discovering and adjusting to their child's illness. Importantly, the findings also suggested that the experience for the fathers stressed, and in some cases disrupted, their attachment coping strategies. All fathers told stories of trying to get it right for their children and family. Their experiences of, and adjustment to, the illness were related to their attachment strategies. The clinical implications for health professionals are discussed.

https://pdfs.semanticscholar.org/1543/7d4e79e94361edcc5ca60e281d3667bef3b7.pdf


**PURPOSE:** To conduct a metasynthesis of qualitative research exploring parents' psychosocial experiences during complex and traumatic life transitions related to caring for a child with a life-limiting (LLI) or life-threatening illness (LTI). **BACKGROUND:** Parents' experiences of caring for a child impacted by an LLI or LTI are not clearly understood, and holistic, comprehensive pediatric nursing care for parents who have children with LLIs and LTIs continues to be developed as treatment improves and survival is extended. **REVIEW METHODS:** Predetermined
This paper explores the biopsychosocial and spiritual needs of adolescents and young adults (AYAs) with life-threatening or terminal illnesses. AYA are situated between childhood and adulthood (ages 15-25) and have distinct biopsychosocial and spiritual needs unique to their developmental stage. Having a life-threatening or terminal illness directly challenges normal AYA developmental tasks and identity formation. AYA experience more troubling physical symptoms during the dying process compared to other age groups, which leads to significant psychological distress and an increased need for pharmacological treatments. In general, AYA desire to be fully equipped to serve this population due to aligning professional standards and ability to advocate for holistic care within interdisciplinary teams. Additional research is needed to tailor holistic interventions to meet the needs of this population.
This study evaluated the first interaction (FI) between parents and health care providers at the time of admission of a child in pediatric intensive care unit (PICU), and explored the extent to which parents understood the medical information. This prospective study took place in three French university-affiliated PICUs. Forty-two parents of 30 children were interviewed. The physician and nurse who took care of the child completed a questionnaire. We evaluated parents' comprehension (excellent, fair, or poor) by comparing parents' and physicians' responses to six items: diagnosis, affected organ, reason for hospitalization, prognosis, treatments, and further investigations. Parent-physician FI occurred within 24 h of child’s admission. Two thirds of the parents were dissatisfied to wait before receiving information. Most of the parents had an excellent comprehension of the affected organ (n = 25/28, 89.3%) and prognosis (n = 26/30, 86.7%). Two thirds of the parents understood the reason for hospitalization (n = 18/28, 64.3%) and diagnosis (n = 19/30, 63.3%). Less than half the parents understood child’s treatments (n = 10/30, 33.3%) and further investigations (n = 8/21, 38.1%). When a nurse delivered information on treatment, parental comprehension improved (p = 0.053). CONCLUSION: Parents complained of their wait time before receiving information. Most of them had an excellent comprehension. An improved communication between nurses and physicians is mandatory, and the active participation of nurses to give information to the parents should be encouraged. What is known: * In pediatric intensive care unit, health care providers deliver information to parents on their child’s condition, which fosters the trust between them to build a partnership. * Various guidelines exist to help health care providers communicate with parents in PICU, but never mention the specific time of admission. What is new: * Even though parents could wait before entering the unit, they all received information on their child’s condition within 24 hours after admission. * Parents understood the information well, and nurses improved the parental comprehension of the treatments by reformulating.


INTRODUCTION: Given the particularity of spirituality in the Indian context, models and tools for spiritual care that have been developed in Western countries may not be applicable to Indian palliative care patients. Therefore, we intended to describe the most common signs of spiritual distress in Indian palliative care patients, assess differences between male and female participants, and formulate contextually appropriate recommendations for spiritual care based on this data. METHODS: Data from 300 adult cancer patients who had completed a questionnaire with 36 spirituality items were analyzed. We calculated frequencies and percentages, and we compared responses of male and female participants using Chi-squared tests. RESULTS: Most participants believed in God or a higher power who somehow supports them. Signs of potential spiritual distress were evident in the participants' strong agreement with existential explanations of suffering that directly or indirectly put the blame for the illness on the patient, the persistence of the "Why me?" question, and feelings of unfairness and anger. Women were more likely to consider illness their fate, be worried about the future of their children or spouse and be angry about what was happening to them. They were less likely than men to blame themselves for their illness. The observations on spirituality enabled us to formulate recommendations for spiritual history taking in Indian palliative care. CONCLUSION: Our recommendations may help clinicians to provide appropriate spiritual care based on the latest evidence on spirituality in Indian palliative care. Unfortunately, this evidence is limited and more research is required.


BACKGROUND: Spinal muscular atrophy (SMA) is an inherited neuromuscular disorder and a leading genetic cause of infant death worldwide. However, there is no routine screening program for SMA in the UK. Lack of treatments and the inability of screening tests to accurately predict disease severity are among the key reasons implementation of screening has faltered in the UK. With the recent release of the first therapy for SMA

Few studies have yet examined subgroups among children (aged 8-18) confronted with the death of a close loved one, characterized by different profiles of symptoms of prolonged grief disorder (PGD) and symptoms of bereavement-related posttraumatic stress disorder (PTSD). This study sought to identify such subgroups and socio-demographic and loss-related variables associated with subgroup membership. We used data from 332 children, most of whom (> 80%) were confronted with the death of a parent, mostly (> 50%) due to illness. Latent class analysis revealed three classes of participants: a resilient class (38.6%), a predominantly PGD class (35.2%), and a combined PGD/PTSD class (26.2%). Class membership was associated with self-rated levels of depression and functional impairment, and parent-rated behavioural problems. No significant between-class differences on demographics or loss-related variables were found. The current findings of distinct classes of PGD, and PGD plus PTSD attest to the construct validity of PGD as a distinct disorder, and can inform theory building and the development of diagnostic instruments relevant to children with pervasive distress following loss.


BACKGROUND: It is important to determine the quality of life (QoL) and level of participation in children with Cerebral Palsy (CP). Previous research has used reports from adolescents or caregivers, but there is no evidence that caregivers' reports accurately reflect the experiences of the adolescents they are interested in.

OBJECTIVE/HYPOTHESIS: The aim of this study was to investigate whether a difference was present in the views of the adolescents and their caregivers regarding the participation and the quality of life of adolescents with CP, and to reveal the parameters creating such differences. METHODS: The participation levels and QoL of the adolescents were evaluated separately by the caregiver and the adolescent using the Pediatric Outcomes Data Collection Instrument (PODCI). RESULTS: A statistically significant difference was found in terms of caregivers and adolescents' scores of PODCI upper extremity (Z = -2.560, p = 0.008), transfer&basic mobility (Z = -3.839, p = 0.000), sports/physical functioning (Z = -3.103, p = 0.002), happiness (Z = -2.420, p = 0.016) and global functioning (Z = -3.639, p = 0.001). The children’s scores were statistically significantly higher than caregivers’. It was found that there was a poor consistence in terms of caregivers and adolescents' scores of upper extremity (ICC = 0.373, p = 0.012), transfer/basic mobility (ICC = 0.289, p = 0.016), sport/physical functioning (ICC = 0.359, p = 0.009); moderate consistence in terms of those of global functioning (ICC = 0.421, p = 0.003). CONCLUSION: It was determined that caregivers and children’s answers were not compatible with one another especially in terms of subjective assessments such as happiness and pain, which suggests that the consideration of caregivers or children in the assessment of subjective situations will change the results.


BACKGROUND: Entering the paediatric intensive care unit with a critically ill child is a stressful experience for parents. In addition to fearing for their child’s well-being, parents must navigate both a challenging environment and numerous new relationships with healthcare staff. How parents form relationships with staff and how they...
perceive both their own and the healthcare providers' roles in this early stage of their paediatric intensive care journey is currently unknown. PURPOSE: This paper explores bereaved parents' perceptions of their role and their relationships with healthcare providers when their child is admitted to the intensive care unit, as part of a larger study exploring their experiences when their child dies in intensive care. METHODS: A constructivist grounded approach was utilised to recruit 26 bereaved parents from 4 Australian intensive care units. Parents participated in audio-recorded, semi-structured interviews lasting 90-150min. All data were analysed using the constant comparative analysis processes, supported by theoretical memos. RESULTS: Upon admission, parents viewed healthcare providers as experts, both of their child's medical care and of the hospital system. This expertise was welcomed, with the parent-healthcare provider relationship developing around the child's need for medical care. Parents engaged in 2 key behaviours in their relationships with staff: prioritising survival, and learning 'the system'. Within each of these behaviours are several subcategories, including 'Stepping back', 'Accepting restrictions' and 'Deferring to medical advice'. CONCLUSIONS: The relationships between parents and staff shift and change across the child's admission and subsequent death in the paediatric intensive care unit. However, upon admission, this relationship centres around the child's potential survival and their need for medical care, and the parent's recognition of the healthcare staff as experts of both the child's care and the hospital system.


OBJECTIVE: To explore bereaved parents' judgements of healthcare providers, as part of a larger study examining their perceptions of the death of a child in the paediatric intensive care unit. RESEARCH METHODOLOGY: Constructivist grounded theory. SETTING: Four Australian paediatric intensive care units. MAIN OUTCOME MEASURES: Semi-structured, audio recorded interviews were undertaken with 26 bereaved parents 6-48months after their child's death. Data were transcribed verbatim and analysed using open, focused and theoretical coding and the constant comparative method. FINDINGS: Bereaved parents judged healthcare providers as 'good' or 'poor' based on behaviours they exhibit. 'Good' behaviours were further subdivided by parents into four categories: 'Better than others', 'good', 'very good', and 'fantastic'. Common behaviours identified as 'good' included provision of practical assistance, facilitation of parental presence, and sharing of information. In contrast, the concept of 'poor' had no subdivision: all identified behaviours, including diminishing parental concern, mishandling hope, adopting an unprofessional demeanour, judging the child's worth, and mishandling communication, were equally detrimental. CONCLUSIONS: Findings demonstrate that bereaved parents have clear opinions on what constitutes 'good' and 'poor' behaviours when their child is dying. These judgements provide clear examples for healthcare providers who provide end-of-life care, ensuring they provide high quality care.


PURPOSE: To explore bereaved parents' perspectives of parent and staff roles in the pediatric intensive care unit when their child was dying, and their relationships with healthcare staff during this time. DESIGN AND METHODS: Constructivist grounded theory was used to undertake this study. Semi-structured interviewers were conducted with 26 bereaved parents recruited from four Australian pediatric intensive care units. The constant comparative method, coupled with open, focused, and theoretical coding were used for data analysis. RESULTS: Becoming a team explores the changes that occurred to the parent-healthcare provider relationship when parents realized their child was dying and attempted to become part of their child's care team. When the focus of care changed from 'life-saving' to 'end-of-life', parents' perspectives and desires of their and the healthcare providers' roles changed. Parents' attempted to reconstruct their roles to match their changing perspectives, which may or may not have been successful, depending on their ability to successfully negotiate these roles with healthcare providers. CONCLUSIONS: Findings offer insights into parental understandings of both the parental and healthcare provider roles for parents of dying children in intensive care, and the ways in which the parent-healthcare provider relationships can influence and be influenced by changes to these roles. PRACTICE IMPLICATIONS: Successful parent-healthcare provider relationships require an understanding of the parental and healthcare provider role from the parents' perspective. The meanings of the parental and healthcare provider

AIM: To explore bereaved parents’ interactions with healthcare providers when a child dies in a paediatric intensive care unit. BACKGROUND: Although most children admitted to a paediatric intensive care unit will survive, 2-5% will die during their stay. The parents of these children interact and form relationships with numerous healthcare staff during their child’s illness and death. Although previous studies have explored the parental experience of child death in intensive care generally, the nature of their relationships with healthcare providers during this time remains unknown. DESIGN: This study used a constructivist grounded theory approach. METHODS: Data were collected via semi-structured, audio-recorded interviews with 26 bereaved parents from four paediatric intensive care units over 18 months in 2015-2016. Constant comparative analysis and theoretical memos were used to analyse the data. FINDINGS: The theory “Transitional togetherness” demonstrates the changing nature of the parent-healthcare provider relationship across three key phases of the parents’ journey. Phase one, “Welcoming expertise,” focuses on the child’s medical needs, with the healthcare provider dominant in the relationship. Phase two, “Becoming a team,” centres around the parents’ need to recreate a parental role and work collaboratively with healthcare providers. Finally, “Gradually disengaging” describes the parents’ desire for the relationship to continue after the child’s death as a source of support until no longer needed. CONCLUSIONS: Findings from this study offer valuable insights into the changing nature of the parent-healthcare provider relationship and highlight the key foci of the relationship at each stage of the parental journey. https://www.ncbi.nlm.nih.gov/pubmed/28746800


OBJECTIVE: To examine the correlation between mothers’ participation in infant care in the Neonatal Intensive Care Unit (NICU) and their anxiety and problem-solving skill levels in caregiving. METHODS: The cross-sectional study was conducted with 340 mothers whose babies were in the NICU. Data were collected with a questionnaire, a Participation in Caregiving Observation Form, the State and Trait Anxiety Inventory and the Problem-solving Skills Evaluation Form. Descriptive statistics and correlation analysis were used in the evaluation of the data. RESULTS: The mothers were with their babies an average of 6.28 +/- 2.43 (range: 1-20) times a day, participating in many basic procedures of care. A negative correlation was found between the mothers’ scores on the Participation in Caregiving Observation Form and their State and Trait Anxiety Inventory scores (respectively, r = -0.48, p < 0.001 and r = -0.12, p < 0.05), but a positive correlation was observed between the Problem-solving Process (r = 0.41, p < 0.001) and the Baby Care Skills (r = 0.24, p < 0.001) Subscale scores. CONCLUSIONS: The study revealed that mothers participated in many basic caregiving procedures in the NICU and this participation resulted in reduced state and trait anxiety levels and an improvement in the mothers’ problem-solving skills with respect to baby care and related problems. https://www.ncbi.nlm.nih.gov/pubmed/27937082


Continuing a bond after a loved one’s death is considered typical and healthy. However, such a bond can continue symbolically only if it existed in the first place. What of indirect grievers, those who never knew the decedent? The authors describe bonds between individuals who did not have a living relationship to begin with, a concept referred to as imagined bonds. Forty-nine adults, who had a sibling die that they never knew, were interviewed. This article describes the bonds constructed between participants and the sibling they never knew. The authors compare and contrast the concepts of continuing bonds versus imagined bonds. https://www.ncbi.nlm.nih.gov/pubmed/28140777


roles should be explored with parents of dying children, and supported as much as possible to enable the development of a collaborative relationship. https://www.ncbi.nlm.nih.gov/pubmed/29454506
OBJECTIVE: The objective of this study was to describe and quantify the impact of caring for a child with Dravet syndrome (DS) on caregivers. METHODS: We surveyed DS caregivers at a single institution with a large population of patients with DS. Survey domains included time spent/difficulty performing caregiving tasks (Oberst Caregiving Burden Scale, OCBS); caregiver health-related quality of life (EuroQoL 5D-5L, EQ-5D); and work/activity impairment (Work Productivity and Activity Impairment questionnaire, WPAI). Modified National Health Interview Survey (NHIS) questions were included to assess logistical challenges associated with coordinating medical care. RESULTS: Thirty-four primary caregivers responded, and 30/34 respondents completed the survey. From OCBS, providing transportation, personal care, and additional household tasks required the greatest caregiver time commitment; arranging for child care, communication, and managing behavioral problems presented the greatest difficulty. EuroQoL 5D-5L domains with the greatest impact on caregivers (0=none, 5=unable/extreme) were anxiety/depression (70% of respondents >/=slight problems, 34% >/=moderate) and discomfort/pain (57% of respondents>/=slight problems, 23%>/=moderate). The mean EQ-5D general health visual analogue scale (VAS) score (0=death; 100=perfect health) was 67 (range, 11-94). Respondents who scored <65 were two- to fourfold more likely to report >/=moderate time spent and difficulty managing child behavior problems and assisting with walking, suggesting that children with DS with high degrees of motor or neurodevelopmental problems have an especially high impact on caregiver health. On the WPAI, 26% of caregivers missed >1 day of work in the previous week, with 43% reporting substantial impact (>6, scale=1-10) on work productivity; 65% reported switching jobs, quitting jobs, or losing a job due to caregiving responsibilities. National Health Interview Survey responses indicated logistical burdens beyond the home; 50% of caregivers made >10 outpatient visits in the past year with their child with DS. CONCLUSIONS: Caring for patients with DS exerts physical, emotional, and time burdens on caregivers. Supportive services for DS families are identified to highlight an unmet need for DS treatments.


Families often prefer home care to hospital care, and home-care services for ill children are increasing worldwide with limited knowledge of families’ needs during curative and palliative home care. The aim of this study was to elucidate family members’ lived experience when a sick child received home care from county-based primary healthcare services. A descriptive qualitative design was chosen and 12 families including sick children receiving home care and their mothers, fathers and siblings in the south of Sweden were interviewed between December 2015 and January 2017. The transcribed interviews were analysed using a hermeneutic phenomenological approach. The family members’ lived experience was described in three essential themes: “Strengthening family life” relates to how home care induced freedom and luxury in a strained period of life and supported the family’s everyday life. Usual social activities and relations were maintained as time and energy was saved when receiving home care. “Promoting health” relates to how the family members’ burden of illness decreased as the child’s signs of illness alleviated and the well-being of the whole family increased when the child received care in the home. This provided a peaceful respite for family members’ psychosocial recovery. The third theme, “Creating alliances,” relates to the importance of creating trustful alliances for communicating participation in care. If trustful alliances were not created, parents felt an overwhelming responsibility and family members became anxious. The findings suggest that care in the family’s home is a useful complement to hospital care. Home care should be given with close attention to family members’ needs and conditions, as positive effects of home care might be jeopardised when expectations and possibilities are not successfully shared.


OBJECTIVE: To characterize spiritual beliefs and support provided by maternal-child staff at three academic hospitals. BACKGROUND: Parents in neonatal intensive care units (NICUs) believe that addressing spirituality is
important. The spiritual beliefs and the support provided by NICU staff are currently unknown. METHODS: This prospective study surveyed all maternal-child staff (NICU and Obstetrics): physicians, neonatal nurse practitioners, physician assistants, nurses, respiratory therapists, and social workers. Two validated tools were used: Spiritual Involvement and Beliefs Scale (SIBS) and Spiritual Care Inventory (SCI). SIBS assesses spiritual beliefs and practices, SCI assesses the perception of spiritual care that one provides. Demographic information, including self-identified religious affiliation, was collected. RESULTS: Respondents of 406 completed surveys were mostly nurses, female, white, and Christian. SIBS scores ranged between 21 and 136. Higher SIBS score was associated with Christian religion (p = 0.001) and African American (p = 0.003) and Asian (p = 0.017) race, when controlling for site, age, gender, education, role in the NICU, and years in practice. A high SCI score was also associated with Christian religion (p = 0.01). There was a trend toward an association between SCI and older age (p = 0.051). There was an association between a high SIBS score and higher ratings on both SCI subscales used. DISCUSSION: There is a wide range in spirituality and perceived spiritual support among maternal-child staff. This may coincide with the spiritual needs of families in the NICU.


Cancer is the leading cause of disease-related death for adolescents and young adults (AYAs) in the United States. Parents of AYAs with life-threatening illnesses have expressed the desire to talk to their children about end of life (EOL) care, yet, like caregivers of adult patients, struggle to initiate this conversation. Building Evidence for Effective Palliative/End of Life Care for Teens with Cancer is a longitudinal, randomized, controlled, single-blinded clinical trial aimed at evaluating the efficacy of Family CEntered disease-specific advance care planning (ACP) for teens with cancer (FACE-TC). A total of 130 dyads (260 subjects) composed of AYAs 14-20 years old with cancer and their family decision maker (> = 18 years old) will be recruited from pediatric oncology programs at Akron Children’s Hospital and St. Jude Children’s Research Hospital. Dyads will be randomized to either the FACE-TC intervention or Treatment as Usual (TAU) control. FACE-TC intervention dyads will complete three 60-minute ACP sessions held at weekly intervals. Follow-up data will be collected at 3, 6, 12, and 18 months post-intervention by a blinded research assistant (RA). The effects of FACE-TC on patient-family congruence in treatment preferences, quality of life (QOL), and advance directive completion will be analyzed. FACE-TC is an evidenced-based and patient-centered intervention that considers QOL and EOL care according to the AYA’s representation of illness. The family is involved in the ACP process to facilitate shared decision making, increase understanding of the AYA’s preferences, and make a commitment to honor the AYA’s wishes.


OBJECTIVE: To describe perceived stress and symptoms of depression in fathers of infants admitted to the NICU through 2 months after discharge and to explore associations between fathers’ childhood and current relationships with their own parents and their stress and symptoms of depression. DESIGN: Observational, longitudinal. SETTING: Tertiary care center in northeastern United States. PARTICIPANTS: English-speaking fathers of newborns admitted to the NICU. METHODS: Fathers completed the Parental Stress Scale and the Edinburgh Postnatal Depression Scale (EPDS) at infants’ NICU admissions (Time [T] 1), 3 weeks (T2), discharge (T3), and 2 months after discharge (T4). RESULTS: A total of 146 fathers were enrolled between March 2013 and February 2016. Infants’ mean gestational age at birth was 31.9 weeks, and 88% remained in the NICU for 3 weeks or longer. We found that 12% of fathers reported high stress levels at T1, 8% at T3, and 13% at T4. Overall EPDS scores improved over time (p < .001). From T1 to T4, the proportion of fathers with distress/minor symptoms of depression decreased from 41% to 10% and with symptoms of major depression from 16% to 2%. Statistically significant positive associations were found between fathers’ EPDS scores and the quality of relationships with their fathers (at T1, T2, and T3) and with their mothers (across all time points). CONCLUSION: From admission to 2 months after discharge, stress and symptoms of depression persisted for some fathers of infants admitted to the NICU. Evidence-based strategies to support fathers during and after their infants’ NICU hospitalizations need to be further developed, implemented, and evaluated.

Background: Birth and hospitalization of premature neonates create enormous challenges for the family with serious impacts on parents' mental and emotional health. The present study was designed to explore the experiences of fathers with premature neonates hospitalized in a neonatal intensive care unit (NICU). Materials and Methods: In this interpretative phenomenological study, data were collected using in-depth interviews guided with a semi-structured questionnaire and analyzed by interpretative phenomenological analysis. Totally seven interviews were conducted with six participants. Results: The mean age of the fathers was 32 (23-42) years, and all of the fathers lived with their wives. Experiences of the fathers were categorized into 13 subordinate and three superordinate themes: “abandonment and helplessness” (lack of financial support, lack of informational support, and indignation and distrust toward the hospital staffs); “anxiety and confusion” (family disruption, shock due to the premature birth of the neonate, uncertainty, the loss of wishes, feeling of guilt and blame, and occupational disruption); and “development and self-actualization” (emotional development, spiritual development, independence and self-efficacy, and responsibility). Conclusions: The present study showed that the fathers with premature neonates hospitalized in NICU encounter both positive (development and self-actualization) and negative experiences (lack of financial and informational supports, distrust toward the hospital staffs, family disruption, and occupational disruption). Planning to manage adverse experiences can help fathers to cope with this situation.


OBJECTIVE: While improvements in healthcare have resulted in children with complex and life-threatening conditions living longer, a proportion of them still die. The death of a child puts parents at increased risk for anxiety, depression, and complicated grief. Increasing our understanding of the coping strategies that parents use under such extreme circumstances will enable us to best provide support to families, before and after a child’s death. Our aim herein was to develop a theoretical framework of parental coping. METHOD: Evidence from the literature was employed to develop a theoretical framework to describe parental coping in the context of having a child with a life-limiting illness who is declining and facing eventual death. RESULTS: The reasoning and argument consists of three guiding elements: (1) the importance of approach as well as avoidance (as coping strategies) in the context of managing the extreme emotions; (2) the importance of the social aspect of coping within a family, whereby parents cope for others as well as for themselves; and (3) the importance of a flexible and balanced coping profile, with parents using different coping strategies simultaneously. Central to the proposed framework is that effective coping, in terms of adjustment, is achieved by balancing coping strategies: accessing different coping strategies simultaneously or in parallel with a specific focus on (1) approach and avoidance and (2) coping aimed at self and others. SIGNIFICANCE OF RESULTS: Understanding of parental coping strategies is essential for health professionals in order to support parents effectively.


The death of a child creates especially poignant feelings and extreme stress, distress, and devastation for family members and healthcare providers. In addition, serious or long-term illness forces a reconstruction of our experiences with time and space. In this paper, we report on a long-term ethnographic study of a Pediatric Palliative Care Team (PPCT). Using the concepts of spatiality and temporality; Deleuze's concepts of smooth and striated spaces; Innis's concepts of space and time biases; Foucault's concept of heterotopian space-places with multiple layers of meaning; and a related concept of heterokairoi-moments in time with multiple possibilities—we consider how the PPCT constructs and reconstructs meaning in the midst of chaos, ethical dilemmas, and heartbreaking choices.


The loss of a close friend or relative is always an ordeal. When this loved one is a baby, born or even unborn, a number of specific aspects have been reported by parents and researchers. The specificities of perinatal mourning have been progressively recognized since the 1970s, with increasing literature on this topic. Its complexity should be acknowledged by healthcare professionals who cope with perinatal loss, to allow them to offer adapted familial support. This paper is written by a mother, a founding member of a French nonprofit organization supporting parents in case of a prenatal or postnatal life-limiting disorder (Association SPAMA, soins palliatifs et accompagnement en maternite) with an internet support forum and a neonatologist involved in research with parents after the loss of their baby. It attempts to describe how parents experience this situation and how palliative care provides a source of inspiration to families and helps them give meaning to these situations.


Palliative care for children in pediatric hospitals is a vital part of the network of services supporting children with severe illness. This has been recognized, with a trend over the past decade for an increased number of pediatric palliative care (PPC) services established in pediatric hospitals. The inpatient team is in the unique position of influencing the early identification of children and their families, across the age and diagnostic spectrum, which could benefit from palliative care. These services have an opportunity to influence the integration of the palliative approach throughout the hospital, and in so doing, have the capacity to improve many aspects of care, including altering an increasingly futile and burdensome treatment trajectory, and ensuring improved symptom (physical and psychological) management.


It is known that if one partner wants to talk after the loss of a child, while the other does not, the less satisfied they are with the relationship. The aim of this study was to increase our understanding of parental relationships following the loss of a child. A questionnaire on various aspects of the relationship was sent to 1,027 members of bereavement support organizations for parents who have lost children in Norway. The studied sample (N = 285) consisted of 169 women (59.3%) and 116 men (40.7%), representing 175 couples. Although the participants were generally satisfied with their relationship and felt that it was strengthened, there were challenges of communication and interaction. The loss made the relationship special, and they felt they had learned to know each other on a deeper level. The importance of talking together and communicating thoughts and feelings was emphasized by the parents. Women initiated talk more often and evidenced a greater need to talk. A deterioration of the relationship tended to be related to a lack of understanding, communication, and care. Communication, respect, and understanding were underlined as core factors for perceiving a relationship as positive.


Sibling relationships reflect a unique childhood bond, thus the impact on a sibling when a child is seriously ill or dying is profound. We conducted a prospective, longitudinal, qualitative study over 2 years using interpretive descriptive methodology to understand siblings' perspectives when a brother or sister was dying at home or in hospital. The insights from the 10 siblings revealed complex experiences, both personal and with the ill child, their families, and peers. These experiences were paradoxically sources of strain and of support, revealing the importance of validation and normalization in assisting siblings to successfully navigate the experience.


AIM: To understand parents' experiences and needs during a child's end-of-life care at home and to identify systemic factors that influence its provision. BACKGROUND: A child's end-of-life phase is an extremely difficult
time for the whole family. Parents have specific needs, especially when they care for a dying child at home. 

DESIGN: Concurrent embedded mixed methods design. METHODS: This sub-study of the nationwide survey, ‘Paediatric End-of-Life Care Needs in Switzerland’ (2012-2015) included 47 children who received EOL care at home from 2011-2012. We extracted quantitative data from patients’ medical charts and obtained information via parental questionnaire and then compared parents whose child died at home or in hospital by computing generalized estimation equations. We thematically analysed interviews with parents who provided EOL care at home. RESULTS: Parents created an intimate lifeworld and a sense of normality for the child at home. They constantly balanced the family’s lifeworld with the requirements and challenges posed by the outside world. This work exhausted parents. Parental ‘readiness’ and social support drove EOL care for children at home. Parents needed practical help with housekeeping and had negative experiences when dealing with insurance. In only 34.8% of cases was a child’s EOL home care supported by paediatric palliative care team. CONCLUSION: Paediatric end-of-life care at home is only feasible if parents make extraordinary efforts. If family-centred end-of-life home care is provided by a hospital-based paediatric palliative home care team, which includes paid housekeeping help and psychological support, parents’ needs could be better met. 


Disease-related pain and the consequences of pain in children and families are important areas for further research inquiry. There are limited data on the relationship between children's emotional well-being, their experience of pain, and the strategies they employ to cope with pain within the context of serious medical illnesses. Understanding what strategies chronically ill children use to cope with pain and stress is essential for clinical intervention. This observational study examined the ways that children and adolescents with chronic diseases cope with pain and adapt to illness. The data were collected by self-report measures and projective drawing techniques in a novel way to assess the emotional functioning in a mixed cohort of youth with disease-related pain (oncological, rheumatic, and cystic fibrosis). This protocol was administered to 47 children, aged 7-14, recruited from pediatric wards of hospitals of Trento and Rovereto (Italy). The results showed that coping strategies in youth with disease-related pain differed across diagnoses and with self-representations (as part of the self-concept adaptive mode). We also provide evidence of the applicability of using projective drawing methods in assessing coping in youth with chronic illness and associated pain. 


Most studies that have examined whether a child’s death influences parental relationship stability have used small-scale data sets and their results are inconclusive. A likely reason is that child loss affects not only the risk of parental separation, but also the risk of having another child. Hence parity progression and separation must be treated as two competing events in relation to child loss. The analysis in this paper used Finnish register data from 1971 to 2003, covering over 100,000 married couples whose durations of both first marriage and parenthood could be observed. We ran parity-specific Cox regressions in which process time started from the birth of each additional child. All marriages included women of childbearing age, none of whom had experienced any child death on entering the analysis. We find that child loss only modestly influences the divorce risk, whereas its effect on the risk of parity progression is considerable. 


Objective: The current study examined the effect of stress on sibling conflict during the first year of pediatric cancer treatment. Method: Families (N = 103) included a child with cancer (aged 2-17 years, Mage = 6.46, SD = 3.52) and at least one sibling aged <5 years of the child with cancer (Mage = 8.34, SD = 5.61). Primary caregivers completed monthly questionnaires throughout the first year of treatment assessing five sources of stress (i.e., general life, cancer-related, financial, perceived treatment intensity, and life threat) and level of sibling conflict. Using multilevel modeling, we explored the effects of these stressors on conflict both at the within- and between-family levels to examine if changes in stress resulted in concurrent changes in conflict within an individual family,
and whether greater average stress affected the trajectory of conflict between families, respectively. Results: At the between-family level, higher average levels of cancer-related stress, general life stress, and financial stress were associated with higher sibling conflict at the end of the first year of treatment. Perceived treatment intensity and life threat were not associated with conflict. No stressors were associated with conflict at the within-family level. Conclusions: During pediatric cancer treatment, some stressors may spill over into family relationships and contribute to increases in sibling conflict.


This paper explores the effects of experiencing the death of a sibling on children's developmental outcomes. Recent work has shown that experiencing a sibling death is common and long-term effects are large. We extend understanding of these effects by estimating dynamic effects on surviving siblings' cognitive and socioemotional outcomes, as well as emotional and cognitive support by parents. Using the Children of the National Longitudinal Survey of Youth 1979 (CNLSY79), we find large initial effects on cognitive and noncognitive outcomes that decline over time. We also provide evidence that the effects are larger if the surviving child is older and less prominent if the deceased child was either disabled or an infant, suggesting sensitive periods of exposure. Auxiliary results show that parental investments in the emotional support of surviving children decline following the death of their child.


BACKGROUND: Preterm birth has been linked to increased parental stress, depression, and anxiety. Although the rate of neonatal morbidity and mortality decreases with increasing gestational age, recent research has revealed that there is no threshold age for risk or parental concern. PURPOSE: This study examines parental concern about medical and developmental outcomes of their premature infant. METHODS: Parents of 60 premature infants were surveyed in a follow-up clinic regarding their level of concern about 11 morbidities and their child’s gestation-adjusted age; these were compared with the infant's inpatient chart. "Concern scores" were tallied and compared across gestational age groups and knowledge of gestation-adjusted age using Chi-square tests of independence. FINDINGS: Many parents reported concerns about morbidities that were unsupported by their child's diagnoses. Across parents of extremely, very, and moderate-late preterm children, the mean concern scores were 13.9, 15.7, and 19.7, respectively. Overall, 62% of parents incorrectly reported the gestation-adjusted age of their child. Parents who were correct were significantly more likely to correctly anticipate abnormal developmental patterns (70%) and growth patterns (65%) than those who were incorrect (33% and 31%, respectively). IMPLICATIONS FOR RESEARCH: Future research should focus on whether NICU graduate parental stress levels are directly linked to the severity of their child's condition, and how physicians can help decrease NICU graduate parental stress. IMPLICATIONS FOR PRACTICE: Parental anxiety regarding all gestational age neonatal intensive care unit infant outcomes can be decreased by a thorough explanation of gestation-adjusted age and a discussion of expected prematurity-related issues.


The purpose of this study is to explore bereaved mothers’ 2-year experiences of losing their only child in the 2008 Sichuan earthquake. Taking an interpretative phenomenological approach, this study interviewed six bereaved mothers four times (6 months, 12 months, 18 months, and 24 months) in Dujiangyan area in Sichuan Province. The findings suggest that these mothers’ personal grief experiences evolved: initially, anger toward the cause of their children’s deaths, following despair of meaningless life, guilt and regret, and finally yearning. Although their yearning and missing ebbed after 2 years, these mothers still had unresolved grief. These mothers also faced strained marital relationships and additional pressure from social interactions. This study illuminates that these mothers’ personal grief experiences and their coping strategies corresponded to Chinese family and sociocultural context.
Social support is an important factor that shapes how people cope with illness, and health-related communication among peers managing the same illness (network ties with experiential similarity) offers specialized information, resources, and emotional support. Facebook has become a ubiquitous part of many Americans’ lives, and may offer a way for patients and caregivers experiencing a similar illness to exchange specialized health-related support. However, little is known about the content of communication among people who have coped with the same illness on personal Facebook pages. We conducted a content analysis of 12 months of data from 18 publicly available Facebook pages hosted by parents of children with acute lymphoblastic leukemia, focusing on communication between users who self-identified as parents of pediatric cancer patients. Support exchanges between users with experiential similarity contained highly specialized health-related information, including information about health services use, symptom recognition, compliance, medication use, treatment protocols, and medical procedures. Parents also exchanged tailored emotional support through comparison, empathy, encouragement, and hope. Building upon previous research documenting that social media use can widen and diversify support networks, our findings show that cancer caregivers access specialized health-related informational and emotional support through communication with others who have experienced the same illness on personal Facebook pages. These findings have implications for health communication practice and offer evidence to tailor M-Health interventions that leverage existing social media platforms to enhance peer support for patients and caregivers.


BACKGROUND: Benefit finding, or finding positive outcomes in the face of adversity, may play a role in predicting quality of life (QoL) among caregivers, but mixed results suggest that other factors may moderate this relationship. OBJECTIVE: This study examined demographic and psychosocial moderators of the association between benefit finding and QoL among caregivers of childhood cancer survivors. METHODS: Caregivers of childhood cancer survivors (n = 83) completed measures of benefit finding, QoL, coping, optimism, social support, caregiving demand, posttraumatic stress, and demographics. RESULTS: The relationship between benefit finding and QoL was moderated by caregiver age, marital status, socioeconomic status, geographic location, acceptance and emotion-focused coping, optimism, caregiving demand, and posttraumatic stress. Benefit finding was more strongly related to QoL among caregivers with fewer demographic/psychosocial resources. CONCLUSIONS: Results suggest that finding benefits in the cancer experience may have a greater positive impact for caregivers with relatively fewer demographic and psychosocial resources and may have less of an impact for caregivers with relatively greater resources. Findings further point to the complex nature of QoL among caregivers of childhood cancer survivors. IMPLICATIONS FOR PRACTICE: Results may aid clinicians in identifying caregivers at particular risk for low QoL. They may be counseled to find benefits in their experience or provided with resources to strengthen other factors that impact QoL.


This study explored bereaved mothers’ responses to the death of a child from cancer, with a focus on identifying adaptive and complicated grief reactions. To understand the unique meaning of their loss, in-depth interviews were conducted with 13 mothers at two time points. Interpretative phenomenological analysis-guided by meaning-making theories of loss-revealed five master categories: the perceptions of the child’s life with cancer and death from the disease, changed self-identity, coping style, developing an ongoing relationship to the deceased child, and the postdeath social environment. Each of these master categories and associated subthemes provided insights into the characteristics of the bereaved mothers’ adaptive and complicated grief responses to their loss. Given all the mothers evidenced multiple forms or types of these responses over time, they could not be categorized as adaptive or complicated grievers. However, the varying proportions of each of these responses highlighted differences in overall bereavement adaptation.
Transitioning from paediatric to adult care can be a particularly challenging time for young people with epilepsy and research has shown that there are a range of factors which may influence a young person's ability to successfully cope with this difficult time. The following study aimed to explore the psychosocial characteristics of this transitioning population, as well as investigate how knowledgeable the young person and their parent/carer are of their own condition throughout transition. Young people with epilepsy were recruited from two specialist epilepsy clinics in the North West and allocated to one of three groups; Group 1 pre-transition, Group 2 transitioning, and Group 3 post-transition. Results found that the young person's knowledge increased significantly throughout transition, whilst parent/carer's knowledge decreased. In addition, anxiety was found to be significantly lower in Group 2 (transitioning group) compared to Group 1 (pre-transition) and Group 3 (post-transition) and a number of significant gender differences were also identified across the groups. The study highlights the importance of considering all relevant psychosocial factors, such as anxiety, gender and the degree of knowledge the individual holds of their own condition during the transition process in order to develop psycho-educational programmes and transition pathways.


BACKGROUND AND PURPOSE: In the United States, 57,000 children (newborn to 18 years) die annually. Bereaved parents may rely on religious or spiritual beliefs in their grief. The study’s purpose was to examine differences in parents’ use of spiritual and religious coping practices by gender, race/ethnicity, and religion at 1 and 3 months after infant/ICU death. METHODS: The sample consisted of 165 bereaved parents, 78% minority. The Spiritual Coping Strategies Scale was used to measure religious and spiritual coping practices, separately. One-way ANOVAs indicated that Black non-Hispanic mothers used significantly more religious coping practices at 3 months than White non-Hispanic mothers. Protestant and Catholic parents used more religious coping practices than the "no" and "other" religion groups at 1 and 3 months. Within the 30 mother-father dyads (paired t-tests), mothers reported significantly greater use of religious coping practices at 1 and 3 months and spiritual coping practices at 3 months than fathers. CONCLUSION: Religious coping practices were most commonly used by Black mothers and Protestant and Catholic parents. Within dyads, mothers used more spiritual and religious coping practices than fathers. IMPLICATIONS FOR PRACTICE: These findings are beneficial for healthcare personnel in providing support to bereaved parents of diverse races/ethnicities and religions.


BACKGROUND: Most parents of children with cancer say they want detailed information about their child’s prognosis. However, prior work has been conducted in populations of limited diversity. The authors sought to evaluate the impact of parental race/ethnicity on prognosis communication experiences among parents of children with cancer. METHODS: In total, 357 parents of children with cancer and the children’s physicians were surveyed at Dana-Farber Cancer Institute/Boston Children’s Hospital and Children’s Hospital of Philadelphia. Outcome measures were parental preferences for prognostic information, physician beliefs about parental preferences, prognosis communication processes, and communication outcomes. Associations were assessed by logistic regression with generalized estimating equations to correct for physician clustering. RESULTS: Two hundred eighty-one parents (79%) were white, 23 (6%) were black, 29 (8%) were Hispanic, and 24 (7%) were Asian/other. Eighty-seven percent of parents wanted as much detail as possible about their child’s prognosis, with no significant differences by race/ethnicity (P = .75). However, physician beliefs about parental preferences for prognosis communication varied based on parent race/ethnicity, with physicians considering black and Hispanic parents less interested in details about prognosis than whites (P = .003). Accurate understanding of a less favorable prognosis was greater among white (49%) versus nonwhite parents (range, 20%-29%), although this difference was not statistically significant (P = .14). CONCLUSIONS: Most parents, regardless of racial and ethnic background, want detailed prognostic information about their child’s cancer. However, physicians underestimate
the information needs of black and Hispanic parents. To meet parents’ information needs, physicians should ask about parents’ information preferences before prognosis discussions. Cancer 2017;123:3995–4003. (c) 2017 American Cancer Society.


This paper reviews the theoretical and empirical literature on risk and resilience factors impacting on parental bereavement outcomes following the death of a child with a life-limiting condition. Over the past few decades, bereavement research has focussed primarily on a risk-based approach. In light of advances in the literature on resilience, the authors propose a Risk and Resilience Model of Parental Bereavement, thus endeavouring to give more holistic consideration to a range of potential influences on parental bereavement outcomes. The literature will be reviewed with regard to the role of: (i) loss-oriented stressors (e.g., circumstances surrounding the death and multiple losses); (ii) inter-personal factors (e.g., marital factors, social support, and religious practices); (iii) intra-personal factors (e.g., neuroticism, trait optimism, psychological flexibility, attachment style, and gender); and (iv) coping and appraisal, on parental bereavement outcomes. Challenges facing this area of research are discussed, and research and clinical implications considered.


BACKGROUND: Adolescents commonly experience loss due to death, and perceived closeness to the deceased can often increase the intensity of bereavement. Adolescents and early young adult (AeYA) oncology patients may recall previous losses or experience new losses, possibly of other children with cancer, while coping with their own increased risk of mortality. The bereavement experiences of AeYA patients are not well described in the literature. METHODS AND FINDINGS: This analysis of bereavement sought to describe the prevalence and types of losses, the support following a death, and the impact of loss on AeYAs aged 13-21 years with malignant disease (or a hematologic disorder requiring allogeneic transplant). Participants were receiving active oncologic therapy or had completed therapy within the past 3 years. Participants completed a bereavement questionnaire and inventories on depression, anxiety, and somatization. The cross-sectional study enrolled 153 AeYAs (95% participation), most (88%) of whom had experienced a loss due to death. The most commonly reported losses were of a grandparent (58%) or friend (37%). Peer deaths were predominantly cancer related (66%). Many participants (39%) self-identified a loss as "very significant." As loss significance increased, AeYAs were more likely to report that it had changed their life "a lot/enormously" (P<0.0001), that they were grieving "slowly or never got over it" (P=0.0001), and that they felt a need for more professional help (P = 0.026). Peer loss was associated with increased risk of adverse psychological outcomes (P = 0.029), as was parental loss (P = 0.018). CONCLUSIONS: Most AeYAs with serious illness experience the grief process as slow or ongoing. Peer or parental loss was associated with increased risk of negative mental health outcomes. Given the high prevalence of peer loss, screening for bereavement problems is warranted in AeYAs with cancer, and further research on grief and bereavement is needed in AeYAs with serious illness.


Personal narratives are assumed to be primary sources of the essential meaning of lived experiences of dying. In this study, I analyzed the personal diary of Mirac Fidan, a terminally ill adolescent with advanced cancer who kept a diary until her death at the age of 15. Mirac’s Diary, also published as a book, was subjected to hermeneutic phenomenological narrative analysis. Inferences were drawn regarding the following basic elements: (a) The dynamics in which Mirac lived and (2) her perceptions of herself, her immediate environment, and her experiences. Suffering seems to be the main experience dominating Mirac’s life, which I examined with regard to two dimensions: suffering caused by inevitable factors and suffering caused by preventable/changeable factors. The results suggest that if various causes among contextual factors are neutralized, then the quality of the existential experience determined by the inevitable factors would increase.
OBJECTIVE: Pediatric cancer is highly stressful for parents. The current prospective study examines the impact of several stressors (financial strain, life threat, treatment intensity, treatment-related events, and negative life events) on the trajectory of marital adjustment across the first year following diagnosis. We examined whether average level of stressors across the year was related to (1) levels of marital adjustment at the end of the first year of treatment and (2) the rate of change in marital adjustment. METHOD: One hundred and thirty families of children newly diagnosed with cancer (M age = 6.33 years, SD = 3.61) participated. Primary caregivers provided 12 monthly reports on marital adjustment and stressors. RESULTS: Multilevel models indicated that although marital adjustment was stable across the first year on average, random effect estimates suggested that this was the result of differing trajectories between families (e.g., some increasing and others decreasing). Five individual stress constructs and a cumulative stress composite were then used to predict this variability. Higher average economic strain was related to consistently poorer marital adjustment across time. Higher average frequency of treatment-related events and negative life events were associated with decreasing adjustment over time and lower adjustment at the end of the first year of treatment. Perception of life threat and treatment intensity were not associated with final levels or trajectory of adjustment. Finally, higher cumulative stress was associated with consistently poorer marital adjustment across time. CONCLUSION: Implications for identification of at-risk families are discussed, and importance of delivering tailored interventions for this population.


AIM: This Italian study investigated home-based palliative care for young children and how long it took parents to meet their needs. METHODS: The study population consisted of 33 families with a child under the responsibility of the Veneto Regional Center for Pediatric Palliative Care, northern Italy, who needed medical support in at least two of the following areas: respiratory, feeding, pain and seizures. RESULTS: The children had a mean age of 6.8 +/- 4.7 years. We found that 72% of the patients needed medical devices for feeding, 36% had a tracheostomy and 55% were on mechanical ventilatory support. The children needed an average of five different life-supporting medical appliances, and the time taken to provide for their care increased significantly with each additional appliance (p = 0.016). Their most time-consuming daily needs were feeding (174 minutes) and support when they woke up at night (67 minutes). The average daily time that parents spent taking care of their child amounted to eight hours and 54 minutes per day. CONCLUSION: Parents providing palliative care for children with life-limiting diseases spent an average of nine hours a day caring for them each day and had to maintain an average of five medical appliances.


BACKGROUND: Parents who experience a perinatal loss often leave the hospital with empty arms and no tangible mementos to validate the parenting experience. Opportunities to create parenting experiences with transitional objects exist following the infant’s death. PURPOSE: This article offers suggestions for staff in units where infant loss is possible to best assist parents in optimal grieving through the offering of transitional bereavement objects. METHODS/SEARCH STRATEGY: CINAHL Complete, MEDLINE, and the Cochrane Database of Systematic Reviews were searched using the following key words-perinatal bereavement, grief, perinatal loss, transitional objects, bereavement photography-and the search was limited to 5 years and the English language. FINDINGS/RESULTS: Recommendations exist and are well supported by leading neonatal and perinatal nursing and medicine organizations for the use of transitional objects to facilitate healthy grieving when parents experience perinatal loss. Transitional objects are mementos that validate the meaning of parenthood—even if the physical act of parenting was brief. Nursing and medical staff have significant roles in guiding parents to a healthy state of bereavement and ultimately managing long-term grief. IMPLICATIONS FOR PRACTICE: Transitional objects can be provided by staff that are low-cost or free, such as taking photographs for parents, or they can involve purchased products from perinatal bereavement programs. In the latter case, funding needs are a consideration for
bargaining decisions. IMPLICATIONS FOR RESEARCH: Immediately following a loss, parents experience a brief sense of healing after receiving mementos of their infant. However, further research is needed to assess long-term effects of receiving transitional objects following perinatal loss.


BACKGROUND: Providing care for children with disabilities can negatively influence the physical health and health behaviors of family caregivers. OBJECTIVES/HYPOTHESIS: The study purposes were to compare the prevalence of chronic conditions and health risk behaviors of family caregivers of children with and without disabilities and to examine associations between disability status of children and family caregivers' chronic conditions and health risk behaviors. METHODS: This study compared chronic conditions and health risk behaviors across adult family caregivers of children with a disability (FCG-D) and family caregivers of children without a disability (FCG) living in a U.S. household using 2015 National Health Interview Survey data. Health risk behaviors were defined as heavy drinking, current smoking, physical inactivity, and unhealthy sleep. Multivariable logistic regression was conducted to compare chronic conditions and health risk behaviors between FCG-D and FCG with adjustments for demographic and healthcare coverage covariates. RESULTS: FCG-D showed significantly greater likelihoods of chronic conditions (e.g., asthma, back pain, chronic bronchitis, heart conditions, migraine, and obesity) than FCG. FCG-D also exhibited significantly more smoking and unhealthy sleep. CONCLUSIONS: Family caregivers of children with a disability reported significantly greater likelihoods of various chronic conditions and were more likely to engage in health risk behaviors (smoking and unhealthy sleep). Further study is needed to develop intervention programs for encouraging effective health-promoting behaviors among family caregivers of children with a disability as well as health policies for decreasing health disparities experienced by this population.


Aim: Progress in medical care and technology has led to patients with more advanced illnesses being admitted to the Intensive Care Unit (ICU). The practice of approaching end-of-life (EOL) care decisions and limiting care is well documented in Western literature but unknown in Singapore. We performed a retrospective cohort study to describe the practice of EOL care in patients dying in a Singapore surgical ICU (SICU). The surgical critical care population was chosen as it is unique because surgeons are frequently involved in the EOL process. Methods: All consecutive patients aged 21 and above admitted to the SICU from July 2011 to March 2012, and who passed away in the ICU or within 7 days of discharge from the ICU (to account for transferred patients out of the ICU after end-of-life care decisions were made and subsequently passed away) were included in the study. Results: There were 473 SICU admissions during this period, out of which 53 were included with a mean age of 67.2 +/- 11.1 years. EOL discussions were held in 81.1% of patients with a median time from admission to first discussion at 1 day (IQR 0-2.75) and a median number of ICU discussion of 1 (IQR 1-2). As most patients lacked decision-making capacity (inability to retain and process information secondary to the underlying disease pathology or sedative use), a surrogate was involved: group decision in 27.9%, child in 25.6% and an unclear family nominated member in 20.9%. 28.3% of patients were managed as for full active with resuscitation, 39.6% nonescalation of care, and 32.1% for withdrawal. The main reasons for conservative management (nonescalation and withdrawal of care) were certain death in 52.3%, medical futility with minimal response to maximal care (27.3%), and the presence of underlying malignancy (18.2%). There was no significant difference between race or religion among patients for active or conservative management. Conclusion: 71.7% of patients who passed away in the ICU or within 7 days of discharge from the ICU were managed conservatively. More timely, EOL discussions and better advance care planning may be needed to improve our patient care for patients on conservative management.


BACKGROUND: Experiencing the death of an infant in the neonatal intensive care unit (NICU) affects both families and staff, creating challenges and opportunities for best practices. PURPOSE: This practice-based article describes a comprehensive approach to delivering bereavement services to NICU families, as well as education and support
to NICU staff. METHODS: Bereaved NICU parent and staff survey feedback, including quotes describing individual experiences and suggestions for improved service delivery. RESULTS: Bereaved NICU families and caregivers find meaning and purpose in the act of creating keepsake memories at the time of the infant’s death. Mutual healing takes place with subsequent, individualized follow-up contacts by staff familiar to the bereaved parents over the course of a year. IMPLICATIONS FOR PRACTICE: Those staff involved in the care of a NICU infant and family, during and after the infant’s death, attest to the value in providing tangible keepsakes as well as continuing their relationship with the bereaved parents. An effective administrative infrastructure is key to efficient program operations and follow-through. IMPLICATIONS FOR RESEARCH: Studying different methods of in-hospital and follow-up emotional support for NICU bereaved families. Identifying strategies for staff support during and after NICU infant loss, and the impact a formal program may have on staff satisfaction and retention.


BACKGROUND: Pediatric advance care planning is advocated by healthcare providers because it may increase the chance that patient and/or parent wishes are respected and thus improve end-of-life care. However, since end-of-life decisions for children are particularly difficult and charged with emotions, physicians are often afraid of addressing pediatric advance care planning. AIM: We aimed to investigate parents’ views and needs regarding pediatric advance care planning. DESIGN: We performed a qualitative interview study with parents of children who had died from a severe illness. The interviews were analyzed by descriptive and evaluation coding according to Saldana. SETTING/PARTICIPANTS: We conducted semi-structured interviews with 11 parents of 9 children. Maximum variation was sought regarding the child’s illness, age at death, care setting, and parent gender. RESULTS: Parents find it difficult to engage in pediatric advance care planning but consider it important. They argue for a sensitive, individualized, and gradual approach. Hope and quality of life issues are primary. Parents have many non-medical concerns that they want to discuss. Written advance directives are considered less important, but medical emergency plans are viewed as necessary in particular cases. Continuity of care and information should be improved through regular pediatric advance care planning meetings with the various care providers. Parents emphasize the importance of a continuous contact person to facilitate pediatric advance care planning. CONCLUSION: Despite a need for pediatric advance care planning, it is perceived as challenging. Needs-adjusted content and process and continuity of communication should be a main focus in pediatric advance care planning. Future research should focus on strategies that facilitate parent engagement in pediatric advance care planning to increase the benefit for the families.


OBJECTIVE: Many bereaved siblings have still not come to terms with their grief many years after the loss, but few studies have focused on what can help. The aims of this study were to identify cancer-bereaved adolescents’ and young adults’ ways of coping with grief after loss of a sibling, and examine whether these ways of coping were related to their experience of having worked through their grief. METHOD: This nationwide survey of 174 cancer-bereaved siblings (73% participation rate) is based on one open-ended question about coping with grief (“What has helped you to cope with your grief after your sibling’s death?”) and one closed-ended question about siblings’ long-term grief (“Do you think you have worked through your grief over your sibling’s death?”). The open-ended question was analyzed with content analysis; descriptive statistics and Fisher’s exact test were used to examine the relation between type of coping and siblings’ long-term grief. Result: The siblings described four ways of coping: (1) thinking of their dead brother/sister and feeling and expressing their grief; (2) distracting or occupying themselves; (3) engaging in spiritual and religious beliefs/activities; and (4) waiting for time to pass. One of these categories of coping with grief, namely, engaging in spiritual and religious beliefs and activities, was associated with siblings’ experience of having worked through their grief two to nine years after the loss (p = 0.016). Significance of results Those siblings who had used spirituality, religious beliefs, and activities to cope were more likely to have worked through their grief than those who had not.


Adults who are living with cancer while raising young children are faced with distinct challenges particularly when that cancer is advanced. While the literature examining parental cancer continues to grow, very little has focused on families facing advanced cancer and the father’s perspective is nearly absent. To address these gaps, grounded theory methods were used to study the experiences of 11 fathers living with advanced cancer while raising minor children. The participants were all married with between one and six children living in their household. Semi-structured, in-person interviews revealed concerns for their children permeated the “ordeal” and these fathers described the ongoing challenge of “teeter-tottering between hope and despair.” The fathers used key protective strategies to counterbalance the weight of the barriers to achieve resilience throughout the cancer experience. Primary barriers were characterized as physical impairments, uncertainty, and financial strain. Fathers described relying on flexibility in their roles as fathers, open communication patterns, use of supportive resources, and the ability to find meaning in their experiences as crucial to fostering resilience. Recommendations include interdisciplinary family centered interventions that consider gendered parental roles as well as financial burden. 


OBJECTIVE: To explore whether negative emotions expressed by adolescent cancer survivors during follow-up consultations were associated with potential late effects (persisting disease or treatment-related health problems). METHODS: We video-recorded 66 follow-up consultations between 10 pediatricians and 66 adolescent survivors of leukemia, lymphoma or stem-cell transplantations. In transcripts of the recordings, we identified utterances coded as both 1) expressions of negative emotions (VR-CoDES), and 2) late effect-related discussions. Principles of thematic content analysis were used to investigate associations between the two.

RESULTS: Of the 66 video-recorded consultations, 22 consultations contained 56 (49%) utterances coded as both emotional concerns and discussions of potential late effects. Negative emotions were most commonly associated with late effects such as fatigue (“I'm struggling with not having energy”), psychosocial distress (“When I touch this (scar) I become nauseous”), pain (“I'm wondering how long I am going to have this pain?”), and treatment-related effects on physical appearance (“Am I growing?”). CONCLUSIONS: Negative emotions expressed by adolescent cancer survivors during follow-up consultations were frequently associated with potential late effects. These late effects were not the medically most serious ones, but reflected issues affecting the adolescents’ daily life.

PRACTICE IMPLICATION: Eliciting and exploring patients' emotional concerns serve as means to obtain clinically relevant information regarding potential late effect and to provide emotional support.


OBJECTIVE: To describe bereaved caregivers’ experiences of providing care at home for patients with advanced cancer, while interacting with home care services. METHODS: Caregivers of patients who had completed a 4-month randomized controlled trial of early palliative care versus standard oncology care were recruited 6 months to 5 years after the patient’s death. All patients except one (control) had eventually received palliative care. In semi-structured interviews, participants were asked about their experiences of caregiving. Grounded theory guided all aspects of the study. RESULTS: Sixty-one bereaved caregivers (30 intervention, 31 control) were interviewed, including spouses (33), adult children (19), and other family (9). There were no differences in themes between control and intervention groups. The core category of Taking charge encompassed caregivers’ assumption of active roles in care, often in the face of inadequate formal support. There were 4 interrelated subcategories: (1) Navigating the system-navigating the complexities of the home care system to access resources and supports; (2) Engaging with professional caregivers-interacting with visiting personnel to advocate for consistency and quality of care; (3) Preparing for death-seeking out information about what to expect at the end of life; and (4) Managing after death-managing multiple administrative responsibilities in the emotionally charged period following death. CONCLUSIONS: Caregivers were often thrust into assuming control in order to compensate for deficiencies in formal palliative home care services. Policies, quality indicators, and guidelines are needed to ensure the provision of comprehensive, interdisciplinary home palliative care.


BACKGROUND: Parenting children with life-threatening illness (LTI) and their healthy siblings requires parents to consider their various needs. OBJECTIVE AND METHODS: We conducted a concurrent, cross-sectional mixed-methods study to describe challenges parents face prioritizing tasks and goals for each child with qualitative data, compare parents’ tasks and goals for children with LTI and healthy siblings with quantitative data, and describe parenting in terms of the process of prioritizing tasks and goals for all children in the family. RESULTS: Participants included 31 parents of children with LTI who have healthy siblings and were admitted to a children’s hospital. Qualitative interviews revealed how parents managed children’s needs and their perceptions of the toll it takes. Quantitative data revealed that parents prioritized “making sure my child feels loved” highest for ill and healthy children. Other goals for healthy siblings focused on maintaining emotional connection and regularity within the family and for ill children focused on illness management. Mixed-methods analysis revealed that parents engaged in a process decision making and traded-off competing demands by considering needs which ultimately transformed the meaning of parenting. DISCUSSION: Future research can further examine trade-offs and associated effects, how to support parent problem-solving and decision-making around trade-offs, and how to best offer social services alongside illness-directed care. 


Objective: To examine parent’s knowledge, attitude and psychosocial response regarding their child’s cancer and treatment after initial disease counseling by doctor. Materials and Methods: Structured questionnaire based study of 43 mothers of newly diagnosed pediatric cancer patients undergoing treatment in pediatric oncology division. Mothers received initial counseling regarding their child’s cancer and treatment from the doctor. Questionnaire was administered 2-6 months after initial counseling and mothers self-reported their responses. Results: 83% mothers had school level education only and 84% belonged to lower and middle socio-economic status. More than 80% mothers knew the name of their child’s cancer, type of treatment received by child and approximate duration of treatment. 93% knew regarding painful procedures and 84% mothers reported knowledge about chemotherapy side effects. Hope of cure and satisfaction with treatment were reported by 90% mothers. 81% mothers reported high levels of anxiety and 66% worried regarding painful procedures. As high as 60% of parents were afraid to send their child outside to play and 40% were afraid to send their child to school. 40% mothers wanted more information regarding child’s higher education, married life & fertility. On statistical analysis, mother’s age, educational status or family background did not influence their knowledge and attitude. Conclusion: Relevant information about child’s cancer and treatment can be imparted effectively even to mothers with school level education. This knowledge helps to instill hopeful attitude, confidence and satisfaction in parents. Anxiety and fear related to cancer persists in mothers even after the initial stress period is over. Pain related to injections and procedures is a major concern in parents. Involvement of counselor in the treating team is desirable to overcome these problems. 


BACKGROUND: Parents of children with cancer are susceptible to psychological distress; however, many parents also report posttraumatic growth (PTG). The objective of this study was to explore the variables associated with PTG in parents of children with cancer who were either on treatment or off treatment. METHODS: One hundred and nineteen parents (71 mothers and 48 fathers) of children with cancer completed self-report questionnaires, including the PTG Inventory, Center for Epidemiologic Studies Depression Scale, State-Trait Anxiety Inventory, and Impact of Event Scale-Revised. Demographic data and children’s medical information were also collected. Multivariate linear regression analyses were conducted to investigate the variables associated with PTG. RESULTS: The mean age of participants was 41.4 years (SD = 6). Higher PTG Inventory scores were associated with parents’ lower trait anxiety (P = .028), parents’ sex (female; P = .004), treatment status (within 12 months from treatment end compared with on-treatment; P = .048), surgery (P = .007), and late effects (P = .01). CONCLUSIONS: Parents’ PTG was associated with children’s clinical characteristics, parents’ sex, and parents’ anxiety levels. When dealing
with PTG, the parents' psychological characteristics and children's clinical characteristics should be considered. Particularly for parents with high trait anxiety, it is important to reduce anxiety first before addressing PTG.


OBJECTIVE: To explore recipients' perspectives on the range and origins of their emotional experiences during their 'bad news' consultations. METHODS: Participants were four bereaved families of children who had changed from active treatment to palliative care in paediatric oncology. Data was collected using emotional touchpoint storytelling. The names (descriptors) given to the emotional experiences were linguistically classified. Explanations of their perceived origins were examined using applied thematic analysis. RESULTS: 26 descriptors were given, relating to bodily sensations, affective states, evaluations and cognitive conditions. Three themes were identified in the origins of these experiences - 'becoming aware', 'the changes' and 'being in this situation'. Parents described strong emotional displays during the consultation including physical collapse. These related to the internal process of 'becoming aware'. Three descriptors were given as originating from the clinicians and their delivery of the news - 'supported', 'included', 'trusting'. CONCLUSIONS: Recipients perceive their emotional experiences as mainly originating from the news itself, and perceived consequences of it, rather than its delivery. Strong emotional reactions during the interaction are not necessarily an indicator of ineffectual delivery. PRACTICE IMPLICATIONS: Findings offer a thematic framing that may support and deepen practitioners understanding of recipients' emotional reactions during bad news consultations.


OBJECTIVE: The presence of a child afflicted with a life-threatening illness is a difficult situation for the child's siblings, especially when their own needs are left unmet. The present article describes the first three phases of research involved in the conceptualization, development, and content validation of an initial version of the Inventaire des Besoins de la Fratrie d'Enfants Malades Severement (IBesFEMS) [Needs Inventory for Siblings of Critically ill Children]. METHOD: The first phase of the development of this instrument was conducted using qualitative methodology (focus groups: 6 siblings, 8 parents). The second phase consisted of validating the content of a pool of items developed according to the needs identified in the first phase. Some 21 participants (3 psychometricians, 3 researchers, 9 clinicians, and 6 siblings) evaluated each item for relevance and clarity. Finally, during the third phase, the acceptability and administration procedures of the preliminary version of the instrument were assessed qualitatively by five siblings. RESULTS: The first phase led to production of a typology made up of 43 needs in 10 different environments. The second phase allowed for selection of the items that were clearest and most relevant, based on expert opinion. This procedure gave rise to a first version of the IBesFEMS, which consisted of 48 items. SIGNIFICANCE OF RESULTS: The IBesFEMS appears to be a promising tool for specifically assessing the needs of the adolescent siblings of seriously ill children.


This study explored the development of understanding of death in a sample of 4- to 11-year-old British children and adults (N=136). It also investigated four sets of possible influences on this development: parents' religion and spiritual beliefs, cognitive ability, socioeconomic status, and experience of illness and death. Participants were interviewed using the "death concept" interview that explores understanding of the subcomponents of inevitability, universality, irreversibility, cessation, and causality of death. Children understood key aspects of death from as early as 4 or 5 years, and with age their explanations of inevitability, universality, and causality became increasingly biological. Understanding of irreversibility and the cessation of mental and physical
processes also emerged during early childhood, but by 10 years many children's explanations reflected not an improved biological understanding but rather the coexistence of apparently contradictory biological and supernatural ideas—religious, spiritual, or metaphysical. Evidence for these coexistent beliefs was more prevalent in older children than in younger children and was associated with their parents' religious and spiritual beliefs. Socioeconomic status was partly related to children's biological ideas, whereas cognitive ability and experience of illness and death played less important roles. There was no evidence for coexistent thinking among adults, only a clear distinction between biological explanations about death and supernatural explanations about the afterlife.


Today more and more children are living with complex health care needs, many of these children are living with life limiting and/or threatening conditions, some are medically fragile. To live a childhood these children must live in communities and with their families. In most cases this means the child's carers, their parents, most often their mothers, are required to undertake a great deal of the child's care. During a project on parental coping I became aware of the ways in which parents were restructuring their working lives in order to meet the demands of the nursing and medical care needs of their children. In this paper I relate the stories we discovered in this qualitative study and discuss the tensions between parental and state's responsibility for children, carers and the political and cultural rights and responsibilities pertaining to children's care. I use Margret Urban Walker's ideas of expressive collaborative morality to argue that the care of life limited and life threatened children should be framed in a negotiation between the state and the carers, both informal and professional. That such an agreement should include a covenant to assist parents and siblings when a child dies to recover and adjust to their loss, in recognition of the work they have performed in caring for the child during their child's life and their death.


Bereaved families that collectively make meaning of their grief experiences often function better than those that do not, yet most social work bereavement interventions target individuals rather than family units. In this article, authors describe an innovative social work intervention that employs digital storytelling. This is a narrative technique that combines photography, music, and spoken word to help families bereaved by child death make meaning of their loss and envision a future without their deceased child.


We aimed to explore the predictive value of screening for distress alone, hope alone, or a combination of both. In a multicenter prospective study, 37 English-speaking adolescents and young adults with cancer and 40 parents completed validated instruments at diagnosis ("baseline") and 3-6 months later ("follow-up"). Correlated regression models described associations. Within each instrument, baseline and follow-up scores were associated. However, only a composite hope/distress score predicted all three patient-centered outcomes. Multidimensional screens incorporating positive and negative psychosocial constructs may predict patient-centered outcomes better than isolated, single-construct instruments.


Background/aim: The aim of this study was the determination and prospective follow-up of quality of life, depression, and anxiety in pediatric patients with cancer under chemotherapy, as well as the evaluation of related factors. Materials and methods: Fifty newly diagnosed pediatric cancer patients and their parents were prospectively monitored before, during, and after therapy, and tests were used. Results: Significantly lower quality
of life scores were recorded during treatment, in the group with CNS tumors, in the group receiving chemotherapy plus radiotherapy plus surgery, in the inpatient-only treatment group, in the group receiving treatment for longer than 6 months, and in the group of patients whose diagnosis was delayed for more than 3 months. Total quality of life scores for children and their parents were 82.95 +/- 14.59 vs. 83.61 +/- 14.60 before, 54.69 +/- 16.51 vs. 55.78 +/- 16.05 during, and 83.88 +/- 12.44 vs. 84.19 +/- 13.22 at the end of treatment (P < 0.05). Anxiety and depression scores were significantly higher during treatment, in patients whose diagnoses were delayed for more than 3 months, and among inpatients. Conclusion: The quality of life of a majority of our patients was severely affected, and depression and anxiety were more frequently seen especially during treatment.


BACKGROUND: The psychological and physical health of fathers of children with Autism Spectrum Disorder (ASD) is under-researched. Due to the unique parenting demands, fathers of children with ASD may be at increased risk of experiencing psychological and physical health difficulties compared to fathers of children without disabilities (W/OD) and fathers of children with other long-term disabilities (LTD). What little research there is on fathers of children with ASD is often conducted on small clinical samples, or embeds the experiences of fathers within other groups. AIM: The current study aimed to explore the extent to which fathers of children with ASD experience psychological distress and physical health issues (e.g., general health, smoking, chronic pain) compared to fathers of children W/OD and fathers of children with a LTD. METHOD: From a large, nationally representative sample of children, 159 fathers of children with ASD were identified, along with 45 fathers of children with a LTD and 6578 fathers of children W/OD. RESULTS: The majority of fathers were experiencing good psychological and physical health. Approximately 1 in 6 fathers of children with ASD were experiencing elevated levels of psychological distress and poor global health, and were at significantly greater risk than fathers of children W/OD; although these differences were not found compared to fathers of children with a LTD. CONCLUSIONS: Some fathers of children with ASD may require additional support which not only focuses on their psychological wellbeing but also fathers’ physical health. The current findings encourage health services to check-in with, or reach-out to fathers as they too may require additional support.


There are strong calls from many national and international bodies for there to be a ‘holistic’ and integrated approach to the understanding and management of psychological and physical health needs. Such holistic approaches are characterized by the treatment of the whole person, taking into account mental and social factors, rather than just the symptoms of a disease. Holistic approaches can impact on mental and physical health and are cost-effective. Several psychological interventions have demonstrated efficacy in improving holistic health outcomes, for example Cognitive Behaviour Therapy, Behavioural Therapies and Problem Solving Therapies. They have shown to impact upon a wide range of outcomes, including psychological distress, pain, physical health, medication adherence, and family outcomes. There is increasing recognition that the holistic goals of the child and family should be prioritised, and that interventions and outcomes should reflect these goals. A focus on holistic goals in therapy can be achieved through a combination of personalised goal-based outcomes in addition to symptom-based measures.


BACKGROUND: The medical traumatic stress model is commonly applied to childhood cancer, assuming that the diagnosis of cancer is a traumatic event. However, to the authors' knowledge, little is known regarding what specifically children perceive as stressful about cancer or how it compares with other stressful events more often experienced by children. METHODS: Children with cancer (254 children) and demographically similar peers without a history of serious illness (202 children) identified their most stressful life event as part of a diagnostic
OBJECTIVE: To examine the relationship between the cancer care experiences of adolescents and young adults (AYAs) and their quality of life. METHODS: Two hundred and nine AYAs completed a cross-sectional, self-report survey distributed through the population-based cancer registries in 2 Australian states (New South Wales and Victoria). Eligible AYAs were 15 to 24 years old when diagnosed with any cancer (excluding early-stage melanoma) and were 3 to 24 months post-diagnosis. Questions examined whether particular care experiences occurred for the patient at different points in the cancer care pathway, including diagnosis, treatment, inpatient care, and at the end of treatment. Quality of life was assessed using the Functional Assessment of Cancer Therapy-General scale. RESULTS: Positive experiences of care at diagnosis, during treatment, during inpatient stays, and when finishing treatment were associated with higher functional, emotional, and social well-being. However, these associations generally became nonsignificant when communication and support experiences were included in the model. Inpatient experiences positively influenced emotional well-being over and above the effect of communication and support experiences. CONCLUSIONS: The results suggest that, for most AYAs' quality of life outcomes, positive experiences of age-appropriate communication and emotional support may underpin the effect of positive experiences of care throughout the cancer care pathway. The results support the need for communication and support tailored to an AYA audience, as recognised by recent Australian and international guidelines on the care of AYAs with cancer.


BACKGROUND: The purpose of this study was to explore the differences in illness perception between children with cancer and other chronic diseases. A secondary aim was to examine the similarities and differences between the illness perception of these children and their parents. METHODS: The Revised Illness Perception Questionnaire (IPQ-R) was used to measure the children's and parents' illness perceptions. In this study, 184 children (ages 8-18 years) and their caregivers completed the questionnaires. RESULTS: This study shows that children with cancer feel that they have greater control over their treatment compared to the other two groups. The children's parents have more pessimistic views of the illness than their children. CONCLUSION: Examinations of illness perceptions among paediatric cancer patients and their families are essential in designing psychosocial interventions for these families. The clinical value of our results can help better understand the cancer-specific features of illness perceptions.


OBJECTIVE: The aim of this study was to compare the musculoskeletal pain distribution, quality of life, and the hopelessness level in mothers with disabled children in different ambulation levels. METHODS: This study included a total of 177 mothers (mean age: 36.1 +/- 6.5 years) of children with disabilities. The mothers were divided into 3 different groups according to the ambulation level of their disabled children: Ambulatory children (Group 1), partially ambulatory children (Group 2) and non-ambulatory children (Group 3). Musculoskeletal pain distribution (body diagram) and pain intensity (The Visual Analogue Scale), four quality of life parameters (The Centers for Disease Control and Prevention Health-Related Quality of Life -4 Questionnaire) and hopelessness level (Beck Hopelessness Scale) were evaluated in all mothers. RESULTS: The results of our study showed that musculoskeletal pain was most common (79.1%) in the mothers of disabled children. The frequency and severity of back, shoulder and elbow pain in the mothers, number of activity limitation days and hopelessness level were found to increase significantly as the ambulation level in the child decreased (p< 0.05). CONCLUSIONS: The risk of musculoskeletal pain, participation in daily life and hopelessness level in the mothers increased as the ambulation level of the disabled children decreased.


AIMS AND OBJECTIVES: To explore couples' perceptions of the effects of perinatal loss on their marital relationship, social support and grief 1 year postloss, and analyse what factors changed the severity of their grief. BACKGROUND: Perinatal losses are traumatic events in the lives of families and can have serious long-term consequences for the psychological health of parents and any subsequent children. DESIGN: A prospective follow-up study. METHODS: We recruited, at a teaching hospital in southern Taiwan, a convenience sample of 30 couples whose babies either miscarried or were stillborn. At 1 month (T1), 3 months (T2), 6 months (T3) and 1 year (T4) after the pregnancy loss, all participants completed four questionnaires. To analyse the changing status of their grief and its related factors, we used a generalised estimating equation (GEE) to account for correlations between repeated observations. RESULTS: Postbereavement grief levels fell over the four time-points. Mothers reported feeling more grief than did the fathers. Couples with a history of infertility, no religious beliefs or no living children before the loss felt more grief from a perinatal miscarriage or stillbirth. Furthermore, couples reported more grief if their marital satisfaction level was low, if their socioemotional support from husband's parents was low or if they had never participated in a ritual for their deceased baby. CONCLUSIONS: Six months postloss is the crucial period for bereaved parents after a perinatal loss. Being a mother, having no previous living children and low-level socioemotional support from the husband's parents are significant high-risk factors for a high level of grief 1 year after perinatal death. RELEVANCE TO CLINICAL PRACTICE: We recommend that health professionals increase their ability to identify the factors that psychologically affect postloss grief. Active postloss follow-up programmes should focus on these factors to offer specific support and counselling.

In the US, the non-Hispanic Black infant mortality rate exceeds the rate among non-Hispanic Whites by more than two-fold. To explore factors underlying this persistent disparity, we employed a mixed methods approach with concurrent quantitative and qualitative data collection and analysis. Eighteen women participated in interviews about their experience of infant loss. Several common themes emerged across interviews, grouped by domain: individual experiences (trauma, grieving and counseling; criminalization); negative interactions with healthcare providers and the healthcare system; and broader contextual factors. Concurrently, we estimated the Black infant mortality rate (deaths per 1000 live births) using linked live birth-infant death records from 2010 to 2013 in every metropolitan statistical area in the US. Poisson regression examined how contextual indicators of population health, socioeconomic conditions of the Black population, and features of the communities in which they live were associated with Black infant mortality and inequity in Black-White infant mortality rates across 100 metropolitan statistical areas with the highest Black infant mortality rates. We used principal components analysis to create a Birth Equity Index in order to examine the collective impact of contextual indicators on Black infant mortality and racial inequity in mortality rates. The association between the Index and Black infant mortality was stronger than any single indicator alone: in metropolitan areas with the worst social, economic, and environmental conditions, Black infant mortality rates were on average 1.24 times higher than rates in areas where conditions were better (95% CI = 1.16, 1.32). The experiences of Black women in their homes, neighborhoods, and health care centers and the contexts in which they live may individually and collectively contribute to persistent racial inequity in infant mortality.


Only-child loss parents in China recently gained extensive attention as a newly defined social group. Resilience could be a probable solution out of the psychological dilemma. Using a sample of 185 only-child loss people, this study employed latent class analysis (a) to explore whether different classes of resilience could be identified, (b) to determine socio-demographic characteristics of each class, and (c) to compare the depression and the subjective well-being of each class. The results supported a three-class solution, defined as ‘high tenacity-strength but moderate optimism class’, ‘moderate resilience but low self-efficacy class’ and ‘low tenacity but moderate adaption-dependence class’. Parents with low income and medical insurance of low reimbursement type and without endowment insurance occupied more proportions in the latter two classes. The latter two classes also had a significant higher depression scores and lower subjective well-being scores than high tenacity-strength but moderate optimism class. Future work should care those socio-economically vulnerable bereaved parents, and an elastic economic assistance policy was needed. To develop targeted resilience interventions, the emphasis of high tenacity-strength but moderate optimism class should be the optimism. Moderate resilience but low self-efficacy class should be self-efficacy, and low tenacity but moderate adaption-dependence class should be tenacity.


PURPOSE: Caring for children with acute lymphoblastic leukemia (ALL) is a distressing experience for parents without medical training. The experience can lead to parents’ care burden. This study explored care burden among parents of children with ALL and its related factors. METHODS: A total of 130 parents were surveyed with the Zarit Burden Inventory (ZBI), Perceived Social Support Scale (PSSS), Zung’s Self-rating Anxiety Scale (SAS), Zung’s Self-rating Depression Scale (SDS), Medical Outcome Study Short Form 36 (SF-36), and a study specific demographic information questionnaire. Independent-samples T test, one-way ANOVA, Pearson correlation analysis and multivariate linear regression analysis (stepwise method), and binomial logistic regression were used in data analysis. RESULTS: The mean score of parents’ care burden overall was 37.74 +/- 16.57, 17 (13.08%) had little or no burden, 57 (43.85%) had mild-to-moderate burden, 44 (33.84%) had moderate-to-severe burden, and 12 (9.23%) had severe burden. Regression analyses indicated daily care time, anxiety, general health, average monthly family income, social support, and number of co-caregivers were factors associated with care burden.
These variables accounted for 51% of the variance in care burden. Other demographic information of parents and children, depression, and other dimensions of SF-36 were not related to care burden. The severe burden level was associated the increase risk of emotional distress compared with little or no burden group (OR = 37.500, 95% CI = 4.515-311.348, P = 0.001). CONCLUSION: The results indicated that care burden in parents of children newly diagnosed with ALL is high. Parents with lower levels of care burden tend to have less daily care time, more co-caregivers, higher income, less anxiety, better general health, and social support. Strategies are needed to help reduce parents’ care burden.

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Pediatric palliative care studies often rely on proxy-reported instead of direct child-reported quality of life metrics. The purpose of this study was to longitudinally evaluate quality of life for pediatric patients receiving palliative care consultations and to compare patient-reported quality of life with parent perception of the child’s quality of life across wellness domains. The 23-item PedsQL V4.0 Measurement Model was utilized for ten child and parent dyads at time of initial palliative care consultation, Month 6, and Month 12 to assess for physical, emotional, social, and cognitive dimensions of quality of life as reported independently by the child and by the parent for the child. Findings were analyzed using Bland-Altman plots to compare observed differences to limits of agreement. This study revealed overall consistency between parent- and child-reported quality of life across domains. Physical health was noted to be in closest agreement. At the time of initial palliative care consult, children collectively scored their social quality of life higher than parental perception of the child’s social quality of life; whereas, emotional and cognitive quality of life domains were scored lower by children than by the parental report. At the one year survey time point, the physical, emotional, and social domains trended toward more positive patient perception than proxy perception with congruence between quality of life scores for the cognitive domain. Findings reveal the importance of eliciting a child report in addition to a parent report when measuring and longitudinally trending perceptions on quality of life.


Adolescence, the transition between childhood and adulthood, represents a time of rapid biological, neurocognitive, and psychosocial changes. These changes have important implications for the development and evolution of adolescent spirituality, particularly for adolescents with chronic or life-limiting illnesses. To contribute positively to adolescent spiritual formation, palliative care teams benefit from understanding the normative changes expected to occur during adolescence. This paper provides a narrative review of adolescent spirituality while recognizing the role of religious, familial, and cultural influences on spiritual development during the teenage years. By giving explicit attention to the contextual norms surrounding adolescence and still recognizing each adolescent-aged patient as unique, palliative care teams can help adolescents transition toward meaningful and sustainable spiritual growth. This paper reviews the clinical and research implications relevant to integrating adolescent spiritual health as part of comprehensive palliative care.


"Losing Thomas & Ella" presents a research comic about one father’s perinatal loss of twins. The comic recounts Paul’s experience of the hospital and the babies’ deaths, and it details the complex grieving process afterward, including themes of anger, distance, relationship stress, self-blame, religious challenges, and resignation. A methodological appendix explains the process of constructing the comic and provides a rationale for the use of comics-based research for illness, death, and grief among practitioners, policy makers, and the bereaved.


PURPOSE: Osteogenesis imperfecta (OI) is a chronic, genetic condition frequently described as “brittle bones.” This condition is expressed by low bone density and characterized by frequent fractures with and without trauma. Additional symptoms include pain, altered growth, and challenges with mobility. This experience has a great impact on the daily life of the child diagnosed with OI and their family. With the introduction of bisphosphonate therapy children diagnosed with OI experienced an increase in bone density that included a change in symptoms and improvement in daily functioning. The purpose of this study was to describe the lived experience of children receiving bisphosphonate therapy for osteogenesis imperfecta (OI) and their mothers. DESIGN AND METHODS: A phenomenological study was conducted using interviews with a purposive sample of six children diagnosed with OI and their six mothers (N = 12). Children ranged in age from 6 to 18 years. The Giorgi (2009) methodology was used to discover the meaning of living day to day since initiating the bisphosphonate infusion therapy. RESULTS: Four themes emerged from the synthesis of the meaning units that reflected the experience that bisphosphonate therapy had on daily life with OI. These four themes explicitly described the phenomena being studied and included living daily life in stride; normalcy is living with uncertainty; renewal with infusions; and making choices and living with the consequences. PRACTICE IMPLICATIONS: Nurses must take an active role in developing and promoting family-centered interventions for transition and support.


Inherited cardiomyopathies, including hypertrophic cardiomyopathy (HCM) and dilated cardiomyopathy (DCM), are the most common monogenic cause of cardiac disease and can rarely lead to sudden cardiac death (SCD). They are characterized by incomplete and age-dependent penetrance and are usually initially symptomatic in adulthood yet can present in childhood as well. Over 20 genes have been identified to cause HCM, and more than 40 genes are known to cause DCM. Genetic testing for these genes has been integrated into medical care; however, the psychological impact of genetic testing and the impact of the uncertainty that comes with receiving these results have not been well studied. This study surveyed 90 adult probands and relatives with a personal or family history of cardiomyopathy from a single hospital-based cardiac genetic program to determine the psychosocial impact of genetic testing for cardiomyopathies. Standardized psychological instruments including an adapted Multidimensional Impact of Cancer Risk Assessment (aMICRA), Impact of Event Scale (IES), and Satisfaction with Decision (SWD) scales were utilized. Patients with positive genetic test results had higher scores for intrusive thoughts, avoidance, and distress when compared to those with negative genetic test results and were also more likely to make or plan to make life changes because of the results of their genetic testing. Satisfaction with the decision to undergo genetic testing was similar regardless of genetic test results. The results of this study provide insight into the patient experience of genetic testing for cardiomyopathies and how these experiences are associated with genetic test results and cardiac history.


By employing the phenomenographic approach, the present study explored children’s cognitive understanding of and emotional responses to death and bereavement. Participants included 52 Korean, 16 Chinese, and 16 Chinese American children ages 5-6. Thematic analysis of children’s drawings and open-ended interviews revealed that most children associated death with negative emotions such as fear, anxiety, and sadness. The majority of children used realistic expressions to narrate death. The core themes from their drawings included causes for death, attempts to stop the dying, and situations after death. This study contributes to the literature by targeting young children who have been relatively excluded in death studies and provides evidence in the usefulness of drawings as a developmentally appropriate data collection tool. The findings also enrich our knowledge about children’s understanding of death and bereavement, rooted in the inductive analysis of empirical data with children from culturally diverse backgrounds.


INTRODUCTION: It is well-known that parental stress and coping impacts the well-being of children with serious illness. The current study aimed to evaluate the feasibility and satisfaction of a novel resilience promoting intervention, the Promoting Resilience in Stress Management Intervention for Parents (PRISM-P) among parents of adolescents and young adults with Type 1 diabetes or cancer. Secondary analyses explored the effect of the PRISM-P on parent-reported resilience and distress. METHOD: The PRISM-P includes 4 short skills-based modules, delivered in either 2 or 4 separate, individual sessions. English-speaking parents of adolescents with cancer or Type 1 diabetes were eligible. Feasibility was conservatively defined as a completion rate of 80%; satisfaction was qualitatively evaluated based upon parent feedback regarding intervention content, timing, and format. Resilience and distress were assessed pre- and postintervention with the Connor Davidson Resilience Scale and the Kessler-6 Psychological Distress Scale. RESULTS: Twelve of 24 caregivers of youth with diabetes (50%) and 13 of 15 caregivers of youth with cancer (87%) agreed to participate. Nine of 12 (75%) and 9 of 13 (64%) completed all PRISM-P modules, respectively. Among those who completed the intervention, qualitative satisfaction was high. Parent-reported resilience and distress scores improved after the intervention. Effect sizes for both groups indicated a moderate intervention effect. DISCUSSION: Ultimately, the PRISM-P intervention was well accepted and impactful among parents who completed it. However, attrition rates were higher than anticipated, suggesting alternative or less time-intensive formats may be more feasible. (PsycINFO Database Record https://www.ncbi.nlm.nih.gov/pubmed/28541057)

**Purpose:** Digital technology has the potential to support teenagers and young adults (TYAs) with cancer from the onset of their disease into survivorship. We aimed to establish (1) the current pattern of use of TYA digital technologies within our service-user population, and (2) their preferences regarding digital information and support within the service.

**Methods:** A cross-sectional survey was administered as a paper and online self-completed questionnaire to TYAs aged 13-24 accessing outpatient, inpatient, and day care cancer services at a regional specialist centre over a 4-week period. RESULTS: One hundred two TYAs completed the survey (55.7% male; 39.8% female; 16.7% online; mean age 18.5 years [SD = 3.51]). Of the TYAs, 41.6% rated the importance of digital communication as "essential" to their lives. Half (51.0%) kept in contact with other patients they had not met in person. Respondents wanted to receive clinical information online (66.3%) and use online chat rooms (54.3%). Future online services desired included virtual online groups (54.3%), online counselling or psychological support (43.5%), and receiving (66.3%) and sharing (48.9%) clinical information online. CONCLUSIONS: Young people with cancer are digital natives. A significant subgroup expressed a desire for digital resources from oncology services, though existing resources are also highly valued. Digital resources have potential to improve patient experience and engagement.

**Implications for Cancer Survivors:** There is considerable scope to develop digital resources with which TYAs can receive information and connect with both professionals and fellow patients, following diagnosis, through treatment and survivorship.


Understanding parents' experience of care is essential to develop high-quality perinatal bereavement services. This study aimed at developing a questionnaire to identify parents' needs and record their experience of care. The patient experience questionnaire was developed by professionals and parents, and piloted in a tertiary maternity unit. Responses were received from 58 parents. Sensitivity and kindness of staff and time spent with their baby were ranked as 'very important' by 95% of parents. Care in these areas largely met their needs (90%), although 5% of respondents stated that partners could have been more involved. Between 8% and 15% of respondents did not feel that language used at the diagnosis of fetal death was sensitive, clear and unambiguous. Parents did not always receive written information about their care (5%) or post-mortem (13%). Analysis of bereaved parents' responses identified areas for improvement including greater involvement of partners and a need for timely information. Impact statement What is already known on this subject?: Good quality bereavement care after perinatal death reduces the negative emotional, psychological and social effects for parents. Description of parents' experiences is a potential means to improve the quality of perinatal bereavement care. What do the results of this study add?: Parents' needs and experiences of care after perinatal death were recorded using a patient-experience questionnaire designed by a multi-professional team and parents. Staff behaviour, particularly sensitivity and kindness was highly valued by parents. Giving both verbal and written information could be improved. Training is needed for professionals, particularly those who come into contact with bereaved parents less frequently. What are the implications of these findings for clinical practice and/or further research?: Description of parents' priorities and views can be used to identify areas for improvement in perinatal bereavement care. Parents' views should be regularly sought and used to develop local services in an iterative process.

Introduction: Bereaved children and young people in the UK are ‘hidden mourners’. Sources of data: Review of primary and secondary evidence on childhood bereavement. Area of agreement: Children experience grief that varies according to the circumstance of death and their cognitive ability. Voluntary organizations can be supportive, but provision is patchy and vulnerable to austerity. Areas of concern: Adult-centric denial of the importance and long-term consequences of childhood grief; uncertainty in how best to relate to bereaved children in faiths and in schools. Growing points: Increased awareness of the immediate and long-term consequences of childhood bereavement; even young children can experience loss through death. Areas timely for research: Better knowledge of the numbers of affected children; longitudinal data to track experiences and outcomes; measuring effectiveness of different approaches; identifying risk factors for early intervention in complicated or prolonged grief; the importance of faith and rituals around death; mapping the provision of services to monitor the impact of austerity. Recommendations: ‘Think adult-think child’ means that all staff caring for dying adults should take responsibility for asking what the death means for the children in the family, with schools, primary care and faith organizations having protocols and expertise available to support grieving children; recent catastrophes expose need for agencies to have management plans that focus on vulnerable children and young people.


BACKGROUND: The death of a parent is a highly stressful life event for bereaved children. Several studies have shown an increased risk of mental ill-health and psychosocial problems among affected children. The aims of this study were to systematically review studies about effective support interventions for parentally bereaved children and to identify gaps in the research. METHODS: The review’s inclusion criteria were comparative studies with samples of parentally bereaved children. The focus of these studies were assessments of the effects on children of a bereavement support intervention. The intervention was directed towards children 0-18 years; but it could also target the children’s remaining parent/caregiver. The study included an outcome measure that dealt with effects of the intervention on children. The following electronic databases were searched up to and including November 2015: PubMed, PsycINFO, Cinahl, PILOTS, ProQuest Sociology (Sociological Abstracts and Social Services Abstracts). The included studies were analysed and summarized based on the following categories: type of intervention, reference and grade of evidence, study population, evaluation design, measure, outcome variable and findings as effect size within and between groups. RESULTS: One thousand, seven hundred and-six abstracts were examined. Following the selection process, 17 studies were included. The included studies consisted of 15 randomized controlled studies, while one study employed a quasi-experimental and one study a pre-post-test design. Thirteen studies provided strong evidence with regards to the quality of the studies due to the grade criteria; three studies provided fairly strong evidence and one study provided weaker evidence. The included studies were published between 1985 and 2015, with the majority published 2000 onwards. The studies were published within several disciplines such as psychology, social work, medicine and psychiatry, which illustrates that support for bereaved children is relevant for different professions. The interventions were based on various forms of support: group interventions for the children, family interventions, guidance for parents and camp activities for children. In fourteen studies, the interventions were directed at both children and their remaining parents. These studies revealed that when parents are supported, they can demonstrate an enhanced capacity to support their children. In three studies, the interventions were primarily directed at the bereaved children. The results showed positive between group effects both for children and caregivers in several areas, namely large effects for children’s traumatic grief and parent’s feelings of being supported; medium effects for parental warmth, positive parenting, parent’s mental health, grief discussions in the family, and children’s health. There were small effects on several outcomes, for example children’s post-traumatic stress disorder (PTSD) symptoms, anxiety, depression, self-esteem and behaviour problems. There were studies that did not show effects on some measures, namely depression, present grief, and for the subgroup boys on anxiety, depression, internalizing and externalizing. CONCLUSIONS: The results indicate that relatively brief interventions can prevent children from developing more severe problems after the loss of a parent, such as traumatic grief and mental health problems. Studies have shown positive effects for both children’s and remaining caregiver’s health. Further research is required including how best to support younger bereaved children. There is also a need for more empirically rigorous effect studies in this area.
BACKGROUND: The increasing number of children with life-threatening and life-limiting conditions requires an individualized approach and additional supportive care in hospitals. However, these patients' characteristics and their prevalence in a pediatric tertiary hospital setting have not been systematically analyzed. OBJECTIVE: This study aimed to determine the proportion of hospitalized children who are receiving care for life-threatening diseases with feasible curative treatments and for life-limiting diseases (LLDs) with inevitable premature death as opposed to care for acute or chronic diseases; additionally, it sought to compare patient characteristics, clinical features, and symptoms within these subgroups. DESIGN/SETTING/SUBJECTS: A cross-sectional survey of 208 patients was conducted at a large tertiary pediatric care center through standardized interviews with the responsible medical teams. Patient subgroups were defined as those with acute, chronic, life-threatening, or LLDs. RESULTS: The comparisons of patient subgroups showed distinct differences and revealed that nearly half of all inpatients suffer from life-threatening (20%) or LLDs (27%), with a high proportion of rare diseases (82%). They experienced a high burden of symptoms in all parameters of clinical features, including high demand for medications and nursing care. CONCLUSION: A substantial proportion of pediatric inpatients suffered from life-threatening or LLDs, as well as rare diseases, indicating a high burden of symptoms and a high need for additional care. The results suggest a substantial need to implement pediatric palliative care structures in tertiary care centers for patients in critical and terminal conditions.

CONTEXT: As children with life-limiting illnesses (LLIs) and life-threatening illnesses (LTIs) live longer, challenges to meeting their complex health care needs arise in homes and communities, as well as in hospitals. Integrated knowledge regarding community-based pediatric palliative care (CBPPC) is needed to strategically plan for a seamless continuum of care for children and their families. OBJECTIVES: The purpose of this integrative review article is to explore factors that are associated with the use of CBPPC for U.S. children with LLIs and LTIs and their families. METHODS: A literature search of PubMed, CINAHL, Scopus, Google Scholar, and an ancestry search was performed to identify empirical studies and program evaluations published between 2000 and 2016. The methodological protocol included an evaluation of empirical quality and explicit data collection of synthesis procedures. RESULTS: Forty peer-reviewed quantitative and qualitative methodological interdisciplinary articles were included in the final sample. Patient characteristics such as older age and a solid tumor cancer diagnosis and interpersonal factors such as family support were associated with higher CBPPC use. Organizational features were the most frequently discussed factors that increased CBPPC, including the importance of interprofessional hospice services and interorganizational care coordination for supporting the child and family at home. Finally, geography, concurrent care and hospice eligibility regulations, and funding and reimbursement mechanisms were associated with CBPPC use on a community and systemic level. CONCLUSION: Multilevel factors are associated with increased CBPPC use for children with LLIs or LTIs and their families in the U.S.

PURPOSE: Allogeneic stem cell transplantation may cure approximately 50% of patients, however, a significant part of the other half might benefit from a high-quality palliative care medicine at the end of life. Somatic, psychic and spiritual needs of these patients may differ from those of patients suffering from incurable solid tumours and...
are not comprehensively evaluated so far. METHODS: To address this question, data from charts of 123 patients who have died after allogeneic stem cell transplantation were extracted. In detail, the time line of the clinical course, the symptoms, the administered drugs and other applied procedures were analysed. RESULTS: Approximately one half of the patients, who have died after stem cell transplantation, did not live more than 5 months. Two-thirds of patients died within 14 months after SCT. 28.5% of the patients could not be discharged after transplantation. However, a significant proportion had a low ECOG-score (0-1) prior to death, indicating a high degree of mobility. Major symptoms were weakness, fatigue and need for aid at daily activities. Severe pain, dyspnoea and obstipation, as known from patients suffering from advanced solid tumours, were rare. In consequence, use of opioids seemed to be less frequent than in patients with solid tumours. Measures of intensive care and i.v.-drug administration were applied to a significant proportion of patients. CONCLUSION: The present investigation indicates that the somatic, psychic and spiritual end-of-life-care after allogeneic stem cell transplantation could be optimised. A significant problem for the transplantation team seems to be the realisation of necessity to switch the curative concept into a palliative ambition. Requirements are a subsequent prospectively conducted investigation and an intensification of cooperation between transplant and palliative care teams.


The Open Society Foundation's International Palliative Care Initiative (IPCI) began to support palliative care development in Central and Eastern Europe and the Former Soviet Union in 1999. Twenty-five country representatives were invited to discuss the need for palliative care in their countries and to identify key areas that should be addressed to improve the care of adults and children with life-limiting illnesses. As a public health concern, progress in palliative care requires integration into health policy, education and training of health care professionals, availability of essential pain relieving medications, and health care services. IPCI created the Palliative Care Roadmap to serve as a model for government and/or nongovernment organizations to use to frame the necessary elements and steps for palliative care integration. The roadmap includes the creation of multiple Ministry of Health-approved working groups to address: palliative care inclusion in national health policy, legislation, and finance; availability of essential palliative care medications, especially oral opioids; education and training of health care professionals; and the implementation of palliative care services at home or in inpatient settings for adults and children. Each working group is tasked with developing a pathway with multiple signposts as indicators of progress made. The roadmap may be entered at different signposts depending upon the state of palliative care development in the country. The progress of the working groups often takes place simultaneously but at variable rates. Based on our experience, the IPCI Roadmap is one possible framework for palliative care development in resource constrained countries but requires both health care professional engagement and political will for progress to be made.


OBJECTIVE: To extend previous work and estimate health and social care costs, litigation costs, funeral-related costs, and productivity losses associated with stillbirth in the UK. DESIGN: A population-based cost-of-illness study using a synthesis of secondary data. SETTING: The National Health Service (NHS) and wider society in the UK. POPULATION: Stillbirths occurring within a 12-month period and subsequent events occurring over the following 2 years. METHODS: Costs were estimated using published data on events, resource use, and unit costs. MAIN OUTCOME MEASURES: Mean health and social care costs, litigation costs, funeral-related costs, and productivity costs for 2 years, reported for a single stillbirth and at a national level. RESULTS: Mean health and social care costs per stillbirth were £4191. Additionally, funeral-related costs were £559, and workplace absence (parents and healthcare professionals) was estimated to cost £3829 per stillbirth. For the UK, the annual health and social care costs were estimated at £13.6 million, and total productivity losses amounted to £706.1 million (98% of this cost was attributable to the loss of the life of the baby). The figures for total productivity losses were sensitive to the perspective adopted about the loss of life of the baby. CONCLUSION: This work expands the current intelligence on the costs of stillbirth beyond the health service to costs for parents and society, and yet these additional findings must still be regarded as conservative estimates of the true economic costs. TWEETABLE ABSTRACT: The costs of stillbirth are significant, affecting the health service, parents, professionals, and society. PLAIN LANGUAGE SUMMARY: Why and how was the study carried out? The personal,
social, and emotional consequences of stillbirth are profound. Placing a monetary value on such consequences is emotive, yet necessary, when deciding how best to invest limited healthcare resources. We estimated the average costs associated with a single stillbirth and the costs for all stillbirths occurring in the UK over a 1-year period. What were the main findings? The average cost to the National Health Service (NHS) of care related to the stillbirth and a first subsequent pregnancy was £4191 for each stillbirth. For the UK, this cost was £13.6 million annually. Clinical negligence payments to bereaved parents were estimated at £2.5 million per year. Parents were estimated to spend £1.8 million per year on funerals. The cost of workplace absence as parents cope with the effects of grief was estimated at £2476 per stillbirth. For the UK, this cost was £8.1 million annually. The loss of a baby is also the loss of an individual with the potential to become a valued and productive member of society. The expected value of an adult’s lifetime working hours was taken as an estimate of this productivity loss, and was £213,304 for each stillbirth. The annual cost for all stillbirths was £694 million. We know from parents that the birth of a subsequent child in no way replaces a stillborn baby. We found that 52% of women fall pregnant within 12 months of a stillbirth. From a purely economic perspective concerned only with the number of individuals in society, babies born during this period could potentially replace the productivity losses of the stillborn baby. Adopting this approach, which we understand is controversial and difficult for bereaved parents, the expected productivity losses would be lower, at £333 million. What are the limitations of the work? For some categories, existing data were unavailable and we used clinical opinion to estimate costs. Furthermore, we were unable to quantify some indirect consequences, for example the psychological distress experienced by wider family members. What is the implication for parents? Placing a monetary value on what is for parents a profound personal tragedy may seem unkind. It is, however, unavoidable if we are to provide policy makers with vital information on the wide-ranging consequences that could be prevented through future investments in initiatives to reduce stillbirth.


Approximately 2% of those on the organ transplant list in the UK are children. Early identification of donors and referral to organ donation teams (ODT) has proven to increase both the success rate of gaining consent and the number of organs actually retrieved. To evaluate the practice relating to organ donation for children receiving end-of-life care on a paediatric intensive care unit (PICU) measured against the National Guidelines. All children 0–18 who received their end-of-life care and died on the PICU. A retrospective cohort study of organ donation patterns including referral, approach, consent and donation. This involved a review of case notes on PICU between the years 2009 and 2014. One hundred five deaths were identified and 100 notes were examined and data analysed to ascertain if religion, age and length of stay on PICU impacted on practice. Eighty-six children met the early identification criteria for potential donors, 40 (46.5%) children were referred to the ODT and 33 (38.3%) families were approached regarding donation. Twenty-one (24.4%) families consented to donation. Seventeen donations took place with a total of 41 sets of organs/tissues retrieved. Despite the majority of children meeting early identification for potential donors, many were not being referred. CONCLUSIONS: All children on end-of-life care should be referred for potential organ donation. Organ donation needs to be seen as a priority for hospitals as a part of routine end-of-life care to help increase referral rates and give families the opportunity to donate. Many paediatric deaths are not referred for consideration of organ donation, despite guidelines stating that this process should be standard of care. Further optimization of referral rates may aid in increasing the number of organs available for donation. What is Known: * Shortage of organs continues to be a national problem. * NICE guidelines state that all patients who are on end-of-life care should have the option of organ donation explored. * Required referral both increases the number of donors and organs donated. What is New: * The process of identifying and referring children for paediatric organ donation. * Identifies that children are still not being referred for organ donation. * Organ donation is still not a priority for hospitals.


The application of palliative and hospice care to newborns in the neonatal intensive care unit (NICU) has been evident for over 30 years. This article addresses the history, current considerations, and anticipated future needs for palliative and hospice care in the NICU, and is based on recent literature review. Neonatologists have long managed the entirety of many newborns’ short lives, given the relatively high mortality rates associated with prematurity and birth defects, but their ability or willingness to comprehensively address the continuum of interdisciplinary palliative, end of life, and bereavement care has varied widely. While neonatology service
capacity has grown worldwide during this time, so has attention to pediatric palliative care generally, and neonatal-perinatal palliative care specifically. Improvements have occurred in family-centered care, communication, pain assessment and management, and bereavement. There remains a need to integrate palliative care with intensive care rather than await its application solely at the terminal phase of a young infant’s life-when s/he is imminently dying. Future considerations for applying neonatal palliative care include its integration into fetal diagnostic management, the developing era of genomic medicine, and expanding research into palliative care models and practices in the NICU.


INTRODUCTION: Global scale up of anti-retroviral therapy (ART) has led to expansion of HIV treatment and prevention across sub-Saharan Africa. However, age and gender-specific disparities persist leading to failures in fulfillment of Sustainability Development Goals, including SDG3 (achieving healthy lives and wellbeing for all, at all ages) and SDG5 (gender equality). We assessed ART initiation and adherence, loss to follow-up, all-cause death and early death, according to SDG3 and SDG5 indicators among a cohort of HIV-infected children and adolescents enrolled in care in Dar-es-Salaam, Tanzania METHODS: SDG3 indicators included young (<5 years) and older paediatric children (5 to <10 years), early adolescent (10 to <15 years) and late adolescent (15 to <20 years) age group divisions and the SDG5 indicator was gender. Associations of age group and gender with ART initiation, loss to follow-up and all-cause death, were analysed using Cox proportional hazards regression and with adherence, using generalized estimating equations (GEE) with the Poisson distribution. Associations of age group and gender with early death were analysed, using log-Poisson regression with empirical variance. RESULTS: A total of 18,315 enrollees with at least one clinic visit were included in this cohort study. Of these 7238 (40%) were young paediatric, 4169 (23%) older paediatric, 2922 (16%) early adolescent and 3986 (22%) late adolescent patients at enrolment. Just over half of paediatric and early adolescents and around four fifths of the late adolescents were female. Young paediatric patients were at greater risk of early death, being almost twice as likely to die within 90 days. Males were at greater risk of early death once initiated on ART (HR 1.35, 95% CI 1.09, 1.66)), while females in late adolescence were at greatest risk of late death (HR 2.44 [1.60, 3.74] <0.01). Late adolescents demonstrated greater non-engagement in care (RR 1.21 (95% CI 1.16, 1.26)). Among both males and females, early paediatric and late adolescent groups experienced significantly greater loss to follow-up. CONCLUSION: These findings highlight equity concerns critical to the fulfillment of SDG3 and SDG5 within services for children and adolescents living with HIV in sub-Saharan Africa. Young paediatric and late adolescent age groups were at increased risk of late diagnosis, early death, delayed treatment initiation and loss of continuity of care. Males were more likely to die earlier. Special attention to SDG3 and SDG5 disparities for children and adolescents living with HIV will be critical for fulfillment of the 2030 SDG agenda.


INTRODUCTION: Very little is known about pediatric pain management resource differences. In contrast, disparities in pain management within the adult population are known to exist. This research examined whether significant differences exist between hospitals in the state of New York and what factors impact these pain resources. METHODS: The study was approved by the institutional review board. A questionnaire was sent to the anesthesia/pediatric/pain directors of every hospital in the state of New York via SurveyMonkey. Poverty-enriched areas were identified based on the Census Bureau definition of poverty-enriched areas. The Chi-square test or Fisher exact test was used. Analyses were conducted to compare hospitals with and without a pediatric pain service (PPS) on several hospital characteristics. All analyses were in SAS-V9.4. RESULTS: Of 160 physicians contacted, 40 completed the survey. Twenty-five percent reported that their hospital had a PPS. In these hospitals, 60% were separate from the adult pain service and 90% performed neuraxial but 30% did not offer more specialized nerve blocks. Socioeconomic status in which the hospital is situated did not impact the likelihood of having a PPS. PPSs were significantly more likely to be present in academic centers (p = 0.05) and children’s hospitals (p = 0.01). Rural hospitals were less likely to have a PPS (0%). CONCLUSION: A minority of hospitals have a PPS and disparity exists. The results indicate to us that targeting rural areas and community hospitals for enhancement of PPS would be valuable. Additional teaching of peripheral nerve blocks would also
be valuable.
https://europepmc.org/abstract/med/28181199


PURPOSE OF REVIEW: HIV/AIDS is one of the leading causes of death among adolescents in sub-Saharan Africa and 40% of new HIV infections worldwide occur in this group. HIV testing and counselling (HTC) is the critical first step to accessing HIV treatment. The prevalence of undiagnosed HIV infection is substantially higher in adolescents compared with adults. We review barriers to HTC for adolescents and emerging HTC strategies appropriate to adolescents in sub-Saharan Africa. RECENT FINDINGS: There are substantial individual, health system and legal barriers to HTC among adolescents, and stigma by providers and communities remains an important obstacle. There has been progress made in recent years in developing strategies that address some of these barriers, increase uptake of HTC and yield of HIV. These include targeted approaches focused on provision of HTC among those higher risk of being infected, for example, index-linked HTC and use of screening tools to identify those at risk of HIV. Community-based HIV-testing approaches including HIV self-testing and incentives have also been shown to increase uptake of HTC. SUMMARY: In implementing HTC strategies, consideration must be given to scalability and cost-effectiveness. HTC approaches must be coupled with linkage to appropriate care and prevention services.


BACKGROUND: Accessible information about palliative care available to the public on the Internet is growing. We do not know whether this information is consistent with the current accepted definition of palliative care. AIM: To identify resources on the Internet and social media regarding palliative care and evaluate the information conveyed. DESIGN: A cross-sectional study of “palliative care” search results. SETTING: Top 10 Google websites, top 10 most viewed YouTube videos, and social media platforms, Facebook and Twitter, were searched. RESULTS: The most popular Google websites were mostly from national organizations promoting palliative care, whose definitions of palliative care consistently mention “quality of life” and “relief from symptoms and stress.” None of the websites mentioned children, and 77% cited palliative care as treatment for cancer with less focus on other diseases. No personal stories were included in Google websites, while 60% of YouTube videos included personal stories. Five main themes were generated from 266 YouTube video comments analyzed. The most common theme was emotionality, of which 91% were positive statements. Facebook and Twitter were mostly used by health-care professionals and not the public. CONCLUSIONS: Palliative care resources are mostly positive and consistent with the current definition of palliative care. Major Internet search engines such as Google and YouTube provide valuable insight into information the public receives about palliative care. Future development of Internet resources on palliative care should consider including children and emphasizing palliative care for all life-limiting illnesses.
https://www.ncbi.nlm.nih.gov/pubmed/29552895


INTRODUCTION: The increase in survival of children with severe diseases has led to the rise of children with chronic diseases, sometimes with lifelong disabilities. In 2008, a unit for the specific care of medically complex children (MCC) was created in Hospital La Paz. OBJECTIVES: To describe the work and care activities of this Unit. Patients and methods An analysis was performed on all discharge reports of the Unit between January 2014 and July 2016. RESULTS: The MCC Unit has 6 beds and daily outpatient clinic. A total of 1,027 patients have been treated since the creation of the unit, with 243 from 2014. The median age was 24.2 months (IQ: 10.21-84.25). The large majority (92.59%) have multiple diseases, the most frequent chronic conditions observed were neurological (76.95%), gastrointestinal (63.78%), and respiratory diseases (61.72%). More than two-thirds (69.54%) of MCC are dependent on technology, 53.49% on respiratory support, and 35.80% on nutritional support. Hospital admission rates have increased annually. There have been 403 admissions since 2014, of which 8.93% were readmissions within 30 days of hospital discharge. The median stay during 2014-2016 was 6 days (IQ: 3-14). The occupancy rate has been above 100% for this period. Currently, 210 patients remain on follow-up (86.42%), and 11 children
There is a great need for paediatric palliative care (PPC) services globally, but access to services is lacking in many parts of the world, particularly in resource-poor settings. Globally it is estimated that 21.6 million children need access to palliative care, with 8.2 needing specialist services. PC has been identified as important within the global health agenda e.g., within universal health coverage, and a recent Lancet commission report recognised the need for PPC. However, a variety of challenges have been identified to PPC development globally such as: access to treatment, access to medications such as oral morphine, opioidophobia, a lack of trained health and social care professionals, a lack of PPC policies and a lack of awareness about PPC. These challenges can be overcome utilising a variety of strategies including advocacy and public awareness, education, access to medications, implementation and research. Examples will be discussed impacting on the provision of PPC in resource-poor settings. High-quality PPC service provision can be provided with resource-poor settings, and there is an urgent need to scale up affordable, accessible, and quality PPC services globally to ensure that all children needing palliative care receive it.
palliative care can access it.


CONTEXT: Palliative care patients face legal issues that impact their quality of life. Legal support, embedded in holistic palliative care services, has developed globally over the last decade to address this. OBJECTIVES: This article aims to trace the origins of legal support for palliative care patients, detail models of legal support, and describe achievements and challenges. METHODS: The article draws on years of work in this area and the available literature. RESULTS: Common legal issues include disposing of property and drafting wills, planning for children, dealing with debt and securing social benefits, and addressing discrimination. Diverse approaches to integrating legal support include developing paralegal skills, accessing skilled legal advice, empowering patients and families, and building awareness of rights among health care workers. CONCLUSION: There is robust and growing acceptance of legal support as a key component of holistic palliative care, and many palliative care professionals are identifying and addressing the legal needs they encounter through mediation, guidance on basic rights, or referrals to a lawyer. Addressing legal problems can contribute to peace of mind, well-being, and the health of patients.

Fletcher, S., R. Hughes, S. Pickstock and K. Auret (2018). "Advance Care Planning Discussions with Adolescent and Young Adult Cancer Patients Admitted to a Community Palliative Care Service: A Retrospective Case-Note Audit." J Adolesc Young Adult Oncol 7(1): 112-119.

PURPOSE: Adolescents and young adults (AYA) with cancer are a cohort requiring specialized healthcare models to address unique cognitive and physical challenges. Advance care planning (ACP) discussions likely warrant age-appropriate adaptation, yet, there is little Australian research data available to inform best practice for this group. The goal of this work is to inform future models of ACP discussions for AYA. METHODS: Retrospective medical record audit of AYA patients and an adult comparison group, diagnosed with a malignancy and referred to a community hospice service, in Western Australia, in the period between January 1, 2012 and December 1, 2015. Information was collected regarding end-of-life care discussions, documentation of agreed plan of care, and care received. RESULTS: Twenty-seven AYA and 37 adult medical records were reviewed. Eighteen (66.7%) AYA patients died at home, compared with 19 (51.4%) adults (p = 0.028). Desire to pursue all available oncological therapies, including clinical trials, was documented for 14 (51.9%) AYA patients compared with 9 (24.3%) of the adult group (p = 0.02). Eleven AYA patients (40.7%) received chemotherapy during the last month of life compared with two (5.4%) adults (p = 0.001). CONCLUSIONS: The results indicate that end-of-life care preferences for this unique cohort may differ from those of the adult population and need to be captured and understood. An ACP document incorporating a discussion regarding goals of care, preferred location of care, preference for place of death, and consent to future intervention, including cardiopulmonary resuscitation and prompts for review, could assist in pursuing this objective.


Little is known about specific factors related to chronic pain that need to be considered to support successful transition from pediatric to adult health care settings. This is troubling because 1 in 5 adolescents may experience chronic pain and many will continue to live with pain into adulthood. This paper reviews what is known about successful transition processes for adolescents with various chronic conditions and the unique factors associated with chronic pain and includes a call for further research on transition. Transitioning from the pediatric to the adult health care setting is challenging for adolescents with chronic conditions and their families. Loss to follow-up and negative health outcomes are linked to poor transition processes. Despite studies examining factors associated with successful transition, not all of the findings are transferable to adolescents with chronic pain. We need to support adolescents, young adults, and their parents as they prepare for transition, engage pediatric and adult care providers in care, advocate for system change, and systematically examine the processes that support the successful health care transition of adolescents and young people with chronic pain.
BACKGROUND: Although child mortality is decreasing, more than half of all deaths in childhood occur in children with a life-limiting condition whose death may be expected. AIM: To assess trends in place of death and identify characteristics of children who died in the community after discharge from paediatric intensive care unit. DESIGN: National data linkage study. SETTING/PARTICIPANTS: All children resident in England and Wales when admitted to a paediatric intensive care unit in the United Kingdom (1 January 2004 and 31 December 2014) were identified in the Paediatric Intensive Care Audit Network dataset. Linkage to death certificate data was available up to the end of 2014. Place of death was categorised as hospital (hospital or paediatric intensive care unit) or community (hospice, home or other) for multivariable logistic modelling. RESULTS: The cohort consisted of 110,328 individuals. In all, 7709 deaths occurred after first discharge from paediatric intensive care unit. Among children dying, the percentage in-hospital at the time of death decreased from 83.8% in 2004 to 68.1% in 2014; 852 (0.8%) of children were discharged to palliative care. Children discharged to palliative care were eight times more likely to die in the community than children who died and had not been discharged to palliative care (odds ratio = 8.06 (95% confidence interval = 6.50-10.01)). CONCLUSIONS: The proportion of children dying in hospital is decreasing, but a large proportion of children dying after discharge from paediatric intensive care unit continue to die in hospital. The involvement of palliative care at the point of discharge has the potential to offer choice around place of care and death for these children and families.


At least 8 million children would need specialized pediatric palliative care (PPC) services annually worldwide, and of the more than 42,000 children and teenagers dying annually in the United States, at least 15,000 children would require PPC. Unfortunately, even in resource-rich countries the majority of children dying from serious advanced illnesses are suffering from unrelieved, distressing symptoms such as pain, dyspnea, nausea, vomiting, and anxiety. State of the art treatment and prevention of those symptoms requires employing multi-modal therapies, commonly including pharmacology, rehabilitation, procedural intervention, psychology, and integrative modalities. This article describes the current practice of integrating hypnosis into advanced pain and symptom management of children with serious illness. Three case reports of children living with a life-limiting condition exemplify the effective use of this clinical modality to decrease distressing symptoms and suffering. Hypnosis for pediatric patients experiencing a life-limiting disease not only provides an integral part of advanced symptom management, but also supports children dealing with loss and anticipatory loss, sustains and enhances hope and helps children and adolescents live fully, making every moment count, until death.


OBJECTIVE: To identify statistically significant positive outcomes in pediatric-to-adult transition studies using the triple aim framework of population health, consumer experience, and utilization and costs of care. STUDY DESIGN: Studies published between January 1995 and April 2016 were identified using the CINAHL, Ovid MEDLINE, PubMed, Scopus, and Web of Science databases. Included studies evaluated pre-evaluation and postevaluation data, intervention and comparison groups, and randomized clinic trials. The methodological strength of each study was assessed using the Effective Public Health Practice Project Quality Assessment Tool. RESULTS: Out of a total of 3844 articles, 43 met our inclusion criteria. Statistically significant positive outcomes were found in 28 studies, most often related to population health (20 studies), followed by consumer experience (8 studies), and service utilization (9 studies). Among studies with moderate to strong quality assessment ratings, the most common positive outcomes were adherence to care and utilization of ambulatory care in adult settings. CONCLUSIONS: Structured transition interventions often resulted in positive outcomes. Future evaluations should consider aligning with professional transition guidance; incorporating detailed intervention descriptions about transition planning, transfer, and integration into adult care; and measuring the triple aims of population health, experience, and costs of care.

INTRODUCTION: The creation of paediatric palliative care units (PPCU) could optimise the management of children with palliative focus after admission to a paediatric intensive care unit (PICU). This study describes the clinical and epidemiological characteristics of children referred from PICU to the PPCU of the Autonomous Community of Madrid (CAM). The overall treatment, relapses, re-admissions, and deaths, if occurred, are described. PATIENTS AND METHOD: A retrospective review was performed using the medical records from children transferred from the CAM paediatric intensive care units to the paediatric palliative care unit (1 March 2008–31 January 2015). RESULTS: A total of 41 patients were included (26 male/15 female) with a median age of 33 months (range 1–228). In the follow by the PPCU follow-up, the main approaches were respiratory (invasive ventilation with tracheostomy tube 8/41), nutritional (gastrostomy in 20/41), and pharmacological (anti-epileptics in 29/41 and 34/41 on antibiotic treatment). Hospital re-admission was required by 11/41 patients, with no re-admissions to PICU. Of the 13/41 patients who died, 9/13 was at home, with all of them accompanied by the primary caregivers and family, and only 1/9 with the presence of the home team. CONCLUSIONS: The palliative approach at home is feasible in children, and the integration of PPCU could optimise the comprehensive care of previously critically ill children. It is necessary to achieve an optimal domiciliary care should be achieved, and not just because of patient death. More observational, multicentre and prospective studies are needed to confirm these findings.


Pathfinders is a 10-session program developed in a community setting to creatively address the diverse needs of bereaved children and families, prevent complications of grief and trauma, and promote healthy adaptation. It is an accessible, grief-focused and trauma-informed family systems model that is theory-driven, research-informed, and grounded in practice-based evidence. Pathfinders incorporates principles central to narrative approaches, with a focus on restorative processes for helping children and families stay on track developmentally. This article outlines the structure, process, and content of Pathfinders, including examples of creative interventions used within the program.


It is estimated that 6.3 million children who die annually need pediatric palliative care (PPC) and that only about 10% of them receive the attention they need because about 98% of them live in under-resourced settings where PPC is not accessible. The consultative model and the integrated model of care (IMOC) are the most common strategies used to make PPC available to critically ill children. In the consultative model, the pediatric intensive care unit (PICU) team, the patient, or their family must request a palliative care (PC) consultation with the external PC team for a PICU patient to be evaluated for special care needs. While the consultation model has historically been more popular, issues related to specialist availability, referral timing, staff’s personal biases, misconceptions about PC, and other factors may impede excellent candidates from receiving the attention they need in a timely manner. Contrastingly, in the IMOC, family-centered care, PC tasks, and/or PC are a standard part of the treatment automatically available to all patients. In the IMOC, the PICU team is trained to complete critical and PC tasks as part of normal daily operations. This review investigates the claim that the IMOC is the best model to meet extensive PPC needs in PICUs, especially in low-resource settings; based on an extensive review of the literature, we have identified five reasons why this model may be superior. The IMOC appears to: (1) improve the delivery of PPC and pediatric critical care, (2) allow clinicians to better respond to the care needs of patients and the epidemiological realities of their settings in ways that are consistent with evidence-based recommendations, (3) facilitate the universal delivery of care to all patients with special care needs, (4) maximize available resources, and (5) build local capacity; each of these areas should be further researched to develop a model of care that enables clinicians to provide pediatric patients with the highest attainable standard of health care. The IMOC lays out a pathway to provide the world’s sickest, most vulnerable children with access to PPC, a human right to which
they are entitled by international legal conventions.


PURPOSE: The aim of this article is to provide an overview of our nurse-led transition clinic provided to congenital heart disease patients moving from pediatric into adult care setting. DESCRIPTION OF THE SERVICE: Nurse-led transition clinic was analyzed at various stages of young adult care from an early stage of 12 to 14 years to entering adult setting at 16 years or older. METHODS: Overview of current transition service for young adults being transferred from pediatric into adult services highlights the integral role of clinical nurse specialist as a coordinator of care. RESULTS: The result of the service overview indicates that nurse-led transition service enables patients to build on their knowledge. This is achieved by providing them time and the opportunities to develop an understanding of their condition and the attitudes required to engage with the adult care setting as indicated in the psychology questionnaire from transition day. CONCLUSION: A nurse-led transition clinic enhances long-term care of patients by supporting the young adults and their family/carer through the transition and transfer of the care to promote the young adult's understanding of their condition and to prevent any lost to follow-up.


Although many of the 16,000 children in the United States diagnosed who are with cancer each year could benefit from pediatric palliative care, these services remain underused. Evidence regarding the barriers impeding access to comprehensive palliative care is dispersed in the literature, and evidence specific to pediatric oncology remains particularly sparse. The purpose of the current review was to synthesize the existing literature regarding these barriers and the strategies offered to address them. The authors completed a literature search using the PubMed, Cumulative Index to Nursing and Allied Health Literature (CINAHL), and Web of Science databases. In total, 71 articles were reviewed. Barriers to accessing pediatric palliative care were categorized according to the 4 levels of a modified socioecological model (ie, barriers related to policy/payment, health systems, organizations, and individuals). Major themes identified at each level included: 1) the lack of consistent and adequate funding mechanisms at the policy/payment level, 2) the lack of pediatric palliative care programs and workforce at the health systems level, 3) difficulties integrating palliative care into existing pediatric oncology care models at the organizational level, and 4) the lack of knowledge about pediatric palliative care, discomfort with talking about death, and cultural differences between providers and patients and their families at the individual level. Recommendations to address each of the barriers identified in the literature are included. Cancer 2018. (c) 2018 American Cancer Society.


The end-of-life management of children with diffuse intrinsic pontine glioma (DIPG) is challenging. Families cope with debilitating symptoms and make complex decisions regarding their child’s care. However, there is little evidence guiding palliative care provision for these children. Our objective was to describe the dying trajectory of children with DIPG, their symptoms, the care they require and the end-of-life decisions made for them. This retrospective cohort study analyzed the end-of-life care of 41 consecutive patients with DIPG who died between January 2001 and June 2010. All patients died of disease progression, experiencing a significant symptom burden prior to death. Despite this, the majority of patient days at the end of life were spent at home. However, 60% of patients were hospitalized at least once in their final 3 months, often close to the time of death. A wide range of healthcare professionals were involved, providing a range of medicinal/non-medicinal interventions. Chemotherapy was given to 30% of patients in their final month. Thirty of 33 families approached (91%) agreed to a “Do not resuscitate” order. A small subset of families opted for intensive treatment towards the end of life including cardiopulmonary resuscitation, intensive care admission and mechanical ventilation. Children with DIPG have complex needs and require intensive multidisciplinary support. This paper describes the end-of-life choices made for these children and discusses how these choices influence our institutional model for palliative care. We believe this approach will be useful to clinicians caring for similar patients.
OBJECTIVE: Neonatal ICUs and PICUs increasingly admit patients with chronic critical illness: children whose medical complexity leads to recurrent and prolonged ICU hospitalizations. We interviewed participants who routinely care for children with chronic critical illness to describe their experiences with ICU care for pediatric chronic critical illness. DESIGN: Semi-structured interviews. Interviews were transcribed and analyzed for themes. SETTING: Stakeholders came from five regions (Seattle, WA; Houston, TX; Jackson, MS; Baltimore, MD; and Philadelphia, PA). SUBJECTS: Fifty-one stakeholders including: 1) interdisciplinary providers (inpatient, outpatient, home care, foster care) with extensive chronic critical illness experience; or 2) parents of children with chronic critical illness. INTERVENTIONS: Telephone or in-person interviews. MEASUREMENTS AND MAIN RESULTS: Stakeholders identified several key issues and several themes emerged after qualitative analysis. Issues around chronic critical illness patient factors noted that patients are often relocated to the ICU because of their medical needs. During extended ICU stays, these children require longitudinal relationships and developmental stimulation that outstrip ICU capabilities. Family factors can affect care as prolonged ICU experience leads some to disengage from decision-making. Clinician factors noted that parents of children with chronic critical illness are often experts about their child’s disease, shifting the typical ICU clinician-parent relationship. Comprehensive care for children with chronic critical illness can become secondary to needs of acutely ill patients. Lastly, with regard to system factors, stakeholders agreed that achieving consistent ICU care goals is difficult for chronic critical illness patients. CONCLUSIONS: ICU care is poorly adapted to pediatric chronic critical illness. Patient, family, clinician, and system factors highlight opportunities for targeted interventions toward improvement in care. 


Introduction: Hematologic malignancies (HM) represent the most common neoplasms in childhood. Despite improved overall survival rates, they are still a major contributor to cancer death in children. Aims: To determine the proportion of children with HM in pediatric palliative care (PPC) and to identify the clinical characteristics and symptoms in comparison to children with extracranial solid tumors (non HM patients). Patients and Methods: This study was conducted as a single-center retrospective cohort study of patients in the care of a large specialized PPC team. Results: Fifteen HM and 50 non HM patients were included. Symptoms in which HM patients scored significantly higher than non HM patients were mucositis, difficulty moving, somnolence, fatigue, petechiae and paleness. Blood transfusions were more frequently administered to HM patients, but large external hemorrhage was not observed in any child. A large variety of drugs and appliances were needed by the patients, with morphine being the most frequently prescribed drug. During the study period, a much larger and over the years even increasing number of HM patients (not in the care of the PPC team) died in hospital with an (assumed) curative intent, with two thirds dying in the ICU. Conclusions: Children with HM were referred to outpatient PPC with almost the full clinical picture of advanced leukemia. Noteworthy, the number of children with HM dying at home is decreasing in our center, instead a substantial proportion received high-intensity medical hospital care including novel anticancer therapies. These patients thus seem to be at an increased risk of dying in hospital as the right time to transfer them to palliative care is oftentimes missed.

"To make choices about further education or a career" compared to older individuals (p < 0.05). Conversely, older individuals more frequently cited "To give children a better idea of their risk" (p < 0.002). Sixteen percent of genetic counselors surveyed (6/37) perceived a change in age of testing. All of these respondents had provided HD testing for ten or more years and anecdotaly believed the age at testing has decreased over time. Study results help providers personalize counseling based on patient's age and serve as a starting point for more research into the relationship between age at testing and motivations for testing.


OBJECTIVES: Anticipating case management is considered crucial in pediatric palliative care. In 2012, our children’s university hospital initiated a specialized pediatric palliative care team (PPCT) to deliver inbound and outbound case management for children with life-shortening disease. The aim of this report is to gain insight in the first 9 months of this PPCT. METHODS: Aspects of care during the first 9 months of the PPCT are presented, and comparison is made between patients with malignant disease (MD) and nonmalignant disease (NMD) in a retrospective study design. Insight in the aspects of care of all patients with a life-shortening disease was retrieved from web-based files and the hour registrations from the PPCT. RESULTS: Forty-three children were supported by the PPCT during the first 9 months: 22 with MD with a median of 50 (1-267) days and 29 minutes (4-615) of case management per patient per day and 21 patients with NMD with a median of 79.5 (5-211) days and 16 minutes of case management per day (6-64). Our data show significantly more interprofessional contacts for patients with MD and more in-hospital contacts for patients with NMD. The median number of admission days per patient was 11 (0-22) for MD (44% for anticancer therapy) and 44 (0-303) for NMD (36% for infectious diseases). SIGNIFICANCE OF RESULTS: This overview of aspects of pediatric palliative case management shows shorter but more intensive case management for MD in comparison with NMD. This insight in palliative case management guides the design of a PPCT.


Intensive care facilities are always in demand in the public sector and there is constant competition for beds. Appropriate allocation of children to these resources is based on the ethical principles of distributive justice and beneficence that is determined on the presumed short-term outcome of the acute illness, long-term outcome of the underlying chronic disease and the overall demand for these facilities. At the onset of the HIV epidemic in South Africa, HIV-infected children were refused admission to the paediatric intensive care unit (PICU) on the basis of poor ICU outcomes and the lack of provision of combined antiretroviral therapy (cART) for survivors. The recent significant improvement in outcome in these patients through early recognition and treatment of HIV-related opportunistic infections, the provision of advanced organ support and the routine availability of cART suggests that the previous policy requires review. Ethical principles, the Paediatric Index of Mortality Score for each request, the quality and disability-adjusted life years and cost-effectiveness of care are all important considerations in deciding which patients should be allowed access to these limited and expensive resources. With the improved long-term outcome in HIV-infected children on cART, admission of these cases to a PICU should now be based on the prognosis of the acute illness, as with any other chronic disease such as asthma or diabetes. Withholding and withdrawing advanced life support should accord with standard protocols applied to any condition for which a child is admitted to the PICU.


CONTEXT: Although the early and middle stages of Huntington’s disease (HD) and its complications have been well described, less is known about the course of late-stage illness. In particular, little is known about the population of patients who enroll in hospice. OBJECTIVES: Our goal is to describe the characteristics of patients with HD who enrolled in hospice. METHODS: This is a retrospective cohort study of electronic medical record data
from 12 not-for-profit hospices in the United States from 2008 to 2012. RESULTS: Of the 164,032 patients admitted to these hospices, 101 (0.06%) had a primary diagnosis of HD. Their median age was 57 (IQR 48-65) and 53 (52.5%) were women. Most patients were cared for by a spouse (n = 36, 36.6%) or adult child (n = 20, 19.8%). At the time of admission, most patients were living either at home (n = 39, 38.6%) or in a nursing home (n = 41, 40.6%). All were either bedbound or could ambulate only with assistance. The most common symptom reported during enrollment in hospice was pain (n = 34, 33.7%) followed by anxiety (n = 30, 29.7%), nausea (n = 18, 17.8%), and dyspnea (n = 10, 9.9%). Patients had a median length of stay in hospice of 42 days, which was significantly longer than that of other hospice patients in the sample (17 days), P < 0.001. Of the 101 patients who were admitted to hospice, 73 died, 11 were still enrolled at the time of data analysis, and 17 left hospice either because they no longer met eligibility criteria (n = 14, 13.7%) or because they decided to seek treatment for other medical conditions (n = 3, 3.0%). Of the 73 patients who died while on hospice, most died either in a nursing home (n = 29; 40%) or a hospital (n = 27; 37%). Seventeen patients (23%) died at home. No patient that started in a facility died at home. CONCLUSION: Patients with HD are admitted to hospice at a younger age compared with other patients (57 vs. 76 years old) but have a significant symptom burden and limited functional status. Although hospice care emphasizes the importance of helping patients to remain in their homes, only a minority of these patients were able to die at home.


PURPOSE: Cancer is the leading cause of nonaccidental death among adolescents and young adults (AYAs). High-intensity end-of-life care is expensive and may not be consistent with patient goals. However, the intensity of end-of-life care for AYA decedents with cancer—especially the effect of care received at specialty versus nonspecialty centers—remains understudied. METHODS: We conducted a retrospective, population-based analysis with the California administrative discharge database that is linked to death certificates. The cohort included Californians age 15 to 39 years who died between 2000 and 2011 with cancer. Intense end-of-life interventions included readmission, admission to an intensive care unit, intubation in the last month of life, and in-hospital death. Specialty centers were defined as Children's Oncology Group centers and National Cancer Institute-designated comprehensive cancer centers. RESULTS: Of the 12,938 AYA cancer decedents, 59% received at least one intense end-of-life care intervention, and 30% received two or more. Patients treated at nonspecialty centers were more likely than those at specialty-care centers to receive two or more intense interventions (odds ratio [OR], 1.46; 95% CI, 1.32 to 1.62). Sociodemographic and clinical factors associated with two or more intense interventions included minority race/ethnicity (Black [OR, 1.35; 95% CI, 1.17 to 1.56]; Hispanic [OR, 1.24; 95% CI, 1.12 to 1.36]; non-Hispanic white: reference), younger age (15 to 21 years [OR, 1.36; 95% CI, 1.19 to 1.56]; 22 to 29 years [OR, 1.26; 95% CI, 1.14 to 1.39]; >/= 30 years: reference), and hematologic malignancies (OR, 1.53; 95% CI, 1.41 to 1.66; solid tumors: reference). CONCLUSION: Thirty percent of AYA cancer decedents received two or more high-intensity end-of-life interventions. In addition to sociodemographic and clinical characteristics, hospitalization in a nonspecialty center was associated with high-intensity end-of-life care. Additional research is needed to determine if these disparities are consistent with patient preference.


We must ensure that the 20,000 US children (age 0 to 19 years) who die as a result of serious illness annually receive high-quality end-of-life care. Ensuring high-quality end-of-life care requires recognition that pediatric end-of-life care is conceptually and operationally different than that for adults. For example, in-hospital adult death is considered an outcome to be avoided, whereas many pediatric families may prefer hospital death. Because pediatric deaths are comparatively rare, not all centers offer pediatric-focused palliative care and hospice services. The unique psychosocial issues facing families who are losing a child include challenges for parent decision makers and young siblings. Furthermore, the focus on advance directive documentation in adult care may be less relevant in pediatrics because parental decision makers are available. Health care quality measures provide a framework for tracking the care provided and aid in agency and provider accountability, reimbursement, and educated patient choice for location of care. The National Quality Forum, Joint Commission, and other groups have developed several end-of-life measures. However, none of the current quality measures focus on the unique needs of dying pediatric patients and their caregivers. To evolve the existing infrastructure to
better measure and report quality pediatric end-of-life care, we propose two changes. First, we outline how existing adult quality measures may be modified to better address pediatric end-of-life care. Second, we suggest the formation of a pediatric quality measure end-of-life task force. These are the next steps to evolving end-of-life quality measures to better fit the needs of seriously ill children.


: The Paediatric Emergency Unit at the University Hospital of Wales provides care for over 33 000 children per year. There was no provision in place to support suddenly bereaved families. Staff felt that we could improve the services provided to families who’s children sadly died in the unit. The aim was to provide immediate support for the suddenly bereaved families by working in partnership with local Charity 2 Wish Upon a Star. Together we created a Standard Operating Procedure (SOP) that consisted of a checklist and pathway allowing us to refer relatives for immediate physical and emotional support. The pilot ran for 12 months offering support and counselling tailored to each individual family. The bereavement process begins in the emergency department. When a child had died families were given the to Wish Upon a Star memory boxes which contained physical support including two cuddly elephants, one to follow the patient and one for the parent/carer to keep and foot print kits. As well as written booklets giving practical information on the next steps such as post mortems and funerals- these often very difficult questions to ask and answer. And about the charity itself. Once consent was gained by the staff, the families details were shared with the immediate support worker at the charity who would contact them within 24 hours. The initial 12 months trial had 18 paediatric deaths, 17 of which took up the support offered to them. The informal feedback we gained was positive from both staff and families. Our relationship with the charity has enabled us to feedback to the individual nurse involved, thus improving staff well being. Due to the success of this pathway it has been extended to include young adults up to the age of 25 years. The model has also been rolled out across critical care in UHW and is now being used in all emergency department in Wales. This innovative partnership offers immediate support to bereaved families across Wales which has improved care and services provided by the University Health Board.


BACKGROUND: The field of pediatric palliative oncology is newly emerging. Little is known about the characteristics and illness experiences of children with cancer who receive palliative care (PC). METHODS: A retrospective cohort study of 321 pediatric oncology patients enrolled in PC who died between 2011 and 2015 was conducted at a large academic pediatric cancer center using a comprehensive standardized data extraction tool. RESULTS: The majority of pediatric palliative oncology patients received experimental therapy (79.4%), with 40.5% enrolled on a phase I trial. Approximately one-third received cancer-directed therapy during the last month of life (35.5%). More than half had at least one intensive care unit hospitalization (51.4%), with this subset demonstrating considerable exposure to mechanical ventilation (44.8%), invasive procedures (20%), and cardiopulmonary resuscitation (12.1%). Of the 122 patients who died in the hospital, 44.3% died in the intensive care unit. Patients with late PC involvement occurring less than 30 days before death had higher odds of dying in the intensive care unit over the home/hospice setting compared to those with earlier PC involvement (OR: 4.7, 95% CI: 2.47-8.97, P < 0.0001). CONCLUSIONS: Children with cancer who receive PC experience a high burden of intensive treatments and often die in inpatient intensive care settings. Delayed PC involvement is associated with increased odds of dying in the intensive care unit. Prospective investigation of early PC involvement in children with high-risk cancer is needed to better understand potential impacts on cost-effectiveness, quality of life, and delivery of goal concordant care.


CONTEXT: Early integration of palliative care (PC) in the management of children with high-risk cancer is widely endorsed by patients, families, clinicians, and national organizations. However, optimal timing for PC consultation
is not standardized, and variables that influence timing of PC integration for children with cancer remain unknown. OBJECTIVES: To investigate associations between demographic, disease, treatment, and end-of-life attributes and timing of PC consultation for children with high-risk cancer enrolled on a PC service. METHODS: A comprehensive standardized tool was used to abstract data from the medical records of 321 patients treated at a large academic pediatric cancer center, who died between 2011 and 2015. RESULTS: Gender, race, ethnicity, enrollment on a Phase I protocol, number of high-acuity hospitalizations, and receipt of cardiopulmonary resuscitation were not associated with timing of PC involvement. Patients with hematologic malignancy, those who received cancer-directed therapy during the last month of life, and those with advance directives documented one week or less before death had higher odds of late PC referral (malignancy: odds ratio (OR) 3.24, P = 0.001; therapy: OR 4.65, P < 0.001; directive: OR 4.81, P < 0.0001). Patients who received hospice services had lower odds of late PC referral <30 days before death (OR 0.31, P < 0.001). CONCLUSION: Hematologic malignancy, cancer-directed therapy at the end of life, and delayed documentation of advance directives are associated with late PC involvement in children who died of cancer. Identification of these variables affords opportunities to study targeted interventions to enhance access to earlier PC resources and services for children with high-risk cancer and their families.


BACKGROUND: End-stage renal disease (ESRD) affects nearly 1400 new children each year in the United States. Morbidity and mortality rates remain high for pediatric patients with ESRD, including those that have received a renal transplant. OBJECTIVE: To better understand ESRD patients referred to palliative care, including their physical symptoms, topics discussed, and themes emerging during initial palliative care consultation. DESIGN/SUBJECTS: This study is a retrospective chart review of pediatric ESRD patients who received a palliative care consult. Physical symptoms, core topics, and themes were identified by the interprofessional study team. RESULTS: The study team found 35 patients met inclusion criteria during the study period. The most common standard palliative care metric noted was "complex or time-intensive communication and interdisciplinary social support." Pain was the most common physical symptom addressed with goals of care and communication the most common topics discussed. Themes emerging described the emotional distress of patients and parents as well as prognostic discussions. CONCLUSIONS: This study demonstrates one institution's experience with pediatric ESRD patients undergoing consultation with the pediatric palliative care service. More research is necessary in this population to better describe the best focus for palliative care teams.


Adolescents with life-limiting illnesses have intensive end-of-life trajectories and could benefit from initiation of hospice services. The medical home model, which includes having a usual source of primary care, may help facilitate quality outcomes at the end-of-life for adolescents. The purpose of this study was to determine the relationship between having a usual source of primary care on hospice utilization and end-of-life transitions among adolescents between 15-20 years with a life-limiting illness. A retrospective cohort design used 2007-2010 California Medicaid claims data (n=585). Our dependent variables were hospice utilization (i.e., hospice enrollment, hospice length of stay) and the independent variable was usual source of primary care. Multivariate regression techniques including least squares regression, multivariate logistic regression, and negative binomial regression were used in the analysis of the relationship between usual source of primary care and hospice utilization and end-of-life transitions. Ten percent of our sample utilized hospice services. Having a usual source of primary care was associated with an increase in hospice enrollment, hospice length of stay, and end-of-life transitions. Adolescents with a cancer diagnosis were more likely to enroll in hospice services. For adolescents at the end of life, having a usual source of primary care had a significant impact on hospice enrollment and length.
of stay. This study is among the first to demonstrate a relationship between primary care and hospice use among this vulnerable population.


BACKGROUND: Improvements in care and treatment have led to more young adults with life-limiting conditions living beyond childhood, which means they must make the transition from children's to adult services. This has proved a challenging process for both young adults and service providers, with complex transition interventions interacting in unpredictable ways with local contexts. OBJECTIVES: To explain how intervention processes interact with contextual factors to help transition from children's to adult services for young adults with life-limiting conditions. DESIGN: Systematic realist review of the literature. DATA SOURCES: Literature was sourced from four electronic databases: Embase, MEDLINE, Science Direct and Cochrane Library from January 1995 to April 2016. This was supplemented with a search in Google Scholar and articles sourced from reference lists of included papers. REVIEW METHODS: Data were extracted using an adapted standardised data extraction tool which included identifying information related to interventions, mechanisms, contextual influences and outcomes. Two reviewers assessed the relevance of papers based on the inclusion criteria. Methodological rigor was assessed using the relevant Critical Appraisal Skills Programme tools. RESULTS: 78 articles were included in the review. Six interventions were identified related to an effective transition to adult services. Contextual factors include the need for children's service providers to collaborate with adult service providers to prepare an environment with knowledgeable staff and adequate resources. Mechanisms triggered by the interventions include a sense of empowerment and agency amongst all stakeholders. CONCLUSIONS: Early planning, collaboration between children's and adult service providers, and a focus on increasing the young adults' confidence in decision-making and engaging with adult services, are vital to a successful transition. Interventions should be tailored to their context and focused not only on organisational procedures but on equipping young adults, parents/carers and staff to engage with each other effectively.


Georgia has established the foundational measures for a national palliative care program-policy, education, drug availability, and implementation. Amendments to legislation needed to develop palliative care have been approved. Palliative care has been recognized as a subspecialty in oncology, critical care, internal medicine, and surgery. The National Plan for Palliative Care for 2011-2016 was approved. Opioids, especially oral morphine, are available on a limited basis for patients at home, but oral morphine is not available for patients in the hospital. Prescribing regulations have changed and all physicians are allowed to prescribe and the length of a prescription is now seven days rather than three days previously. Unfortunately, patients and families must still pick up their opioid medications at pharmacies in the police station. Opioids for cancer patients in inpatient units or at home are free. Palliative care education has been incorporated into both undergraduate and postgraduate medical and nursing education and a number of physicians have received specialist training abroad. Palliative Care Standards and Guidelines have been developed; and palliative care services, although insufficient to meet the need, are available for patients at home, as inpatients and a children's hospice opened in 2017.


INTRODUCTION: Despite recent advances in neonatal intensive care in Korea, few studies exist on the end-of-life decisions in newborns. In this study, we sought to examine the status of end-of-life decisions in neonates, changes over time, and affecting factors. METHODS: This is a retrospective study of neonates who died between 2001 and 2015 in the neonatal intensive care unit of Dong-A University Hospital in Busan. The types of end-of-life decisions were divided into active resuscitation, withholding treatment, and withdrawing treatment. The study period was divided into 3 time frames using 5-year intervals to investigate changes over time. To identify the associated factors, we analyzed the demographic and clinical characteristics of the neonates and their parents using the chi(2) test and independent t test. RESULTS: Of the neonatal deaths included in the analysis (n = 222), active resuscitation, withholding treatment, and withdrawing treatment groups accounted for 73.4%, 25.2%, and
1.4% of cases, respectively. When comparing changes over time, between period 1 (2001-2005), 2 (2006-2010), and 3 (2011-2015), the proportion of active resuscitation decreased significantly, from 80.9% to 60.8%, while that of nonactive resuscitation increased significantly from 19.1% to 39.2%. The factors associated with end-of-life decisions were the clinical condition of the neonate at the time of death, rather than general characteristics or socioeconomic factors. CONCLUSIONS: In Korea, changes in the decisions on end-of-life care in neonates are shifting from active resuscitation to nonactive resuscitation based on clinical conditions.


Trisomy 13 (T13) is accompanied by severe complications, and it can be challenging to achieve long-term survival without aggressive treatment. However, recently, some patients with T13 have been receiving home care. We conducted this study to investigate factors related to home health-care transition for patients with T13. We studied 28 patients with T13 born between January 2000 and December 2014. We retrospectively compared nine home care transition patients (the home care group) and 19 patients that died during hospitalization (the discharge at death group). The median gestational age of the patients was 36.6 weeks, with a median birth weight of 2,047 g. Currently, three patients (11%) have survived, and 25 (89%) have died. The home care group exhibited a significantly longer gestational age (38.9 vs. 36.3 weeks, p = 0.039) and significantly larger occipitofrontal circumference Z score (-0.04 vs. -0.09, p = 0.019). Congenital heart defects (CHD) was more frequent in the discharge at death group, with six patients in the home care group and 18 patients in the discharge at death group (67% vs. 95%, p = 0.047), respectively. Survival time was significantly longer in the home care group than in the discharge at death group (171 vs. 19 days, p = 0.012). This study has shown that gestational age, occipitofrontal circumference Z score at birth, and the presence of CHD are helpful prognostic factors for determining treatment strategy in patients with T13.


OBJECTIVES: Every day 43 children are newly diagnosed with cancer. Fortunately, almost 90% of these childhood cancer patients will survive. However, 60-90% of these survivors will experience late effects, health problems that occur months or years after treatment has ended. Late effects could occur as a result of the disease, its treatment, and patient-related factors. The two main objectives of this research are to: 1) Examine the existence of all web-based resources for childhood cancer survivors with acute lymphocytic leukemia which focus on medical and psychological aspects of late effects, and 2) Create an innovative website specifically designed to fill this void.

MATERIALS AND METHODS: A systematic literature review, followed by input from >20 different organizations, resulted in the creation of LEAP3 AHEAD (Late Effects Awareness for Patients, Physicians and the Public; Advancing Health and Eliminating All Disparities), a multi-dimensional website centering on late effects. RESULTS: An extensive review revealed 14 pediatric cancer websites, none of which focused exclusively on late effects. LEAP3 AHEAD is the first interactive website for acute lymphocytic leukemia childhood cancer survivors and families, as well as physicians, and the public to: a) increase awareness about risks, detection, diagnosis, treatment, and prevention of medical and psychological late effects, b) provide suggestions to successfully reintegrate into schools, careers, and socially, and c) present opportunities including camps, scholarships, and pet therapy programs. CONCLUSION: LEAP3 AHEAD is the first national website to provide a comprehensive, accessible, affordable, and multi-dimensional resource for pediatricians, internists, nurse practitioners, psychologists, survivors and their families, as well as the public about late effects.

https://www.ncbi.nlm.nih.gov/m/pubmed/29451924/


OBJECTIVES: To describe health-care spending and utilization for infants discharged from the neonatal intensive care unit (NICU). STUDY DESIGN: Retrospective cohort analysis of 4973 NICU graduates in the Truven MarketScan Medicaid database, with follow-up to the third birthday. Health-care spending and utilization after NICU discharge were assessed. Using logistic regression, we assessed clinical characteristics associated with hospitalization and emergency department (ED) visits. RESULTS: Most (69.5%) post-NICU spending occurred
Participants: Novel but pretested survey tools were administered to 129 patient parent dyads. The aim was to determine the perception of symptom burden early in treatment and assess the feasibility of implementing early palliative care in pediatric oncology patients. OBJECTIVE: To determine the perception of symptom burden early in treatment and assess patients' and parents' needs, attitudes, and perceptions about early palliative care integration in pediatric oncology. The study was conducted at the Children's Hospital of Philadelphia. METHODS: This was a survey-based study conducted in a single institution. Participants were recruited from the oncology inpatient unit. Surveys were administered to patients and their parents before and after the implementation of an intervention called Project ROSE (Reach Out, Soothe, and Embrace). RESULTS: A total of 331 surveys (pre, n = 174, post, n = 157) were analyzed. There was a significant improvement in caregiver spiritual wellbeing and perceptions of care after the intervention. There was no change in the unplanned extubation rate between pre and postimplementation (Wilcoxon rank sum tests compared groups (pre 40.0 [32.0, 44.0] vs post 42.0 [37.5, 45.0] P = .03). CONCLUSION: Project ROSE improved caregiver spiritual wellbeing and perceptions of care, was implemented safely, addresses a need in family centered care of critically ill pediatric patients, and merits consideration for integration into practice.


OBJECTIVES: To evaluate whether a pediatric intensive care unit initiative promoting physical contact between caregiver and patient improves caregiver spiritual wellbeing. The secondary objectives were to evaluate caregiver perceptions of care before and after the initiative and to follow unplanned extubation rate as a marker of safety of the initiative. We hypothesized that caregiver spiritual wellbeing and caregiver perceptions of care would improve with implementation of our physical contact initiative known as Project ROSE (Reach Out, Soothe, and Embrace). STUDY DESIGN: Project ROSE was a practice change initiative promoting physical contact between caregiver and hospitalized child in an academic quaternary care pediatric intensive care unit. Caregivers’ spiritual wellbeing and perceptions of care were surveyed at days 1 and 4, then compared pre- and postimplementation of the unit-wide initiative. Wilcoxon rank sum tests compared groups (pre- and post-Project ROSE). A total of 331 caregivers returned surveys. RESULTS: We analyzed 331 surveys (pre, n = 174/post, n = 157). Caregiver spiritual wellbeing at enrollment (day 1) was no different between groups (P = .47). Caregiver spiritual wellbeing on day 4 was greater in the postintervention group (pre 40.0 [32.0, 44.0] vs post 42.0 [37.5, 45.0] P = .03). Caregiver perceptions of care improved postintervention. There was no change in the unplanned extubation rate between groups. CONCLUSION: Project ROSE improved caregiver spiritual wellbeing and perceptions of care, was implemented safely, addresses a need in family-centered care of critically ill pediatric patients, and merits consideration for integration into practice.


OBJECTIVE: Our aim was to outline a procedure for obtaining a rapid autopsy in order to collect high-quality postmortem tissue for genomic analysis. METHODS: This report details a bi-institutional collaborative effort to coordinate a rapid autopsy for a pediatric patient who had died at home. We discuss the scientific rationale for offering a rapid autopsy to caregivers of pediatric patients as well as parental perspectives on broaching the subject of autopsy. We then review the logistics and coordination involved with planning a rapid autopsy and the sequence of events needed to maximize tissue quality. RESULTS: We report the successful coordination of a rapid autopsy for a patient who died in a hospice setting at her out-of-state home. The time interval from death to the start of the rapid autopsy procedure was 4.5 hours, despite the logistical considerations demanded by the location of the patient. Tumor aliquots and nonneoplastic tissues were successfully snap frozen for downstream genomic studies. SIGNIFICANCE OF RESULTS: Physicians should consider trialing a rapid autopsy program at their institution that could be offered to caregivers of pediatric patients. This case report offers a framework to help clinicians develop their own rapid autopsy programs as well as guidelines to help streamline this process for appropriate candidates going forward.


Importance: Early palliative care integration for cancer patients is now touted as the optimal care model, yet significant barriers often prevent its implementation. A perceived barrier, especially for pediatric oncology patients, is the notion that patients and their families may not need or want palliative care involvement early in the disease trajectory. Objective: To determine the perception of symptom burden early in treatment and assess attitudes toward early integration of palliative care in pediatric oncology patient-parent pairs. Design, Setting, and Participants: Novel but pretested survey tools were administered to 129 patient-parent dyads of hospital-based
pediatric oncology ambulatory clinics and inpatient units between September 2011 and January 2015. All patient participants were aged between 10 and 17 years and were diagnosed as having an oncologic condition 1 month to 1 year before enrollment. Both the patient and the parent in the dyad spoke English, and all participating parents provided written informed consent. A convenience sample was used for selection, with participants screened when otherwise presenting at a participating site. A total of 280 eligible participants were approached for study inclusion, 258 of whom were enrolled in the study (92.1% positive response-rate). Main Outcomes and Measures: Degree of perceived suffering from early symptom-related causes, attitudes toward early palliative care integration, and patient-parent concordance. Statistical analysis included descriptive statistics, calculation of concordance, McNemar test results, and Cochran-Armitage trend test results. Results: Of the 129 patients in the dyads, 68 were boys, and 61 girls; of the 129 parents, 15 were men, and 114 women. Patients reported the following symptoms in the first month of cancer therapy: nausea (n = 109; 84.5%), loss of appetite (n = 97; 75.2%), pain (n = 96; 74.4%), anxiety (n = 77; 59.7%), constipation (n = 69; 53.5%), depression (n = 64; 49.6%), and diarrhea (n = 52; 40.3%). A large proportion of those reporting suffering indicated substantial suffering severity from specific symptoms (ie, a great deal or a lot) including nausea, 52.3% (57 of 109), loss of appetite, 50.5% (49 of 97), constipation 30.4% (21 of 69), pain 30.2% (29 of 96), anxiety 28.6% (22 of 77), depression 28.1% (18 of 64), and diarrhea 23.1% (12 of 52). Few children and parents expressed opposition to early palliative care involvement (2 [1.6%] and 8 [6.2%]) or perceived any detrimental effects on their relationship with their oncologist (6 [4.7%] and 5 [3.9%]), loss of hope (3 [2.3%] and 10 [7.8%]), or therapy interference (3 [2.3%] and 2 [1.6%], respectively). Intradyad concordance was low overall: 26% to 29% for exact concordance and 40% to 69% for agreement within 1 response category. Significant differences in patient-parent attitudes toward aspects of early palliative care included child participants being more likely than their parents (40.3% [n = 52] vs 17.8% [n = 23]) to indicate that palliative care would have been helpful for treating their symptoms (P < .001). Conclusions and Relevance: Pediatric oncology patients experience a high degree of symptom-related suffering early in cancer therapy, and very few patients or parents in this study expressed negative attitudes toward early palliative care. Our findings suggest that pediatric oncology patients and families might benefit from, and are not a barrier to, early palliative care integration in oncology.


PURPOSE: Given childhood cancer survivors’ risk of treatment-induced late effects, long-term follow-up care is recommended. We explored experiences with late effects-related care and preferences for long-term follow-up care among adult survivors of childhood malignant lymphoma in Norway. METHODS: We conducted five focus group interviews with 34 survivors (19 females; 21 Hodgkin/13 non-Hodgkin lymphoma survivors; mean age 39 years; mean time from diagnosis 26 years). Data was analyzed using principles of thematic analysis. RESULTS: Two main themes were identified: (1) the survivors’ experiences with late effects-related care and (2) their preferences for long-term follow-up care. Most of the survivors were dissatisfied with their late effects-related care due to perceptions of poor coordination of healthcare needs in a fragmented system, combined with a perceived lack of knowledge of late effects among themselves and general practitioners (GPs). All survivors valued long-term follow-up care. Oncologists were the preferred care providers, but GPs were considered acceptable providers if they had sufficient knowledge of late effects and routine examinations, short waiting times, and improved GP-oncologist collaboration. CONCLUSIONS: Our results suggest that a shared care model of long-term follow-up care involving specialists, GPs, and the survivors themselves is likely to fulfill several of the currently unmet needs among adult survivors of childhood cancers. Improved patient education about late effects and follow-up care would aid self-management. The survivors’ concerns regarding lack of sufficient knowledge of late effects among GPs suggest a need for improving access to, and dissemination of, information of late effects.


BACKGROUND: A majority of cancer-bereaved siblings report long-term unresolved grief, thus it is important to identify factors that may contribute to resolving their grief. OBJECTIVE: To identify modifiable or avoidable family and care-related factors associated with unresolved grief among siblings two to nine years post loss. DESIGN: This is a nationwide Swedish postal survey. MEASUREMENTS: Study-specific questions and the standardized
instrument Hospital Anxiety and Depression Scale. Primary outcome was unresolved grief, and family and care-related factors were used as predictors. SETTING/PARTICIPANTS: Cancer-bereaved sibling (N = 174) who lost a brother/sister to childhood cancer during 2000-2007 in Sweden (participation rate 73%). Seventy-three were males and 101 females. The age of the siblings at time of loss was 12-25 years and at the time of the survey between 19 and 33 years. RESULTS: Several predictors for unresolved grief were identified: siblings’ perception that it was not a peaceful death (odds ratio (OR): 9.86, 95% confidence interval (CI): 2.39-40.65), limited information given to siblings the last month of life (OR: 5.96, 95% CI: 1.87-13.68), information about the impending death communicated the day before it occurred (OR: 2.73, 95% CI: 1.02-7.33), siblings’ avoidance of the doctors (OR: 3.22, 95% CI: 0.75-13.76), and lack of communication with family (OR: 2.86, 95% CI: 1.01-8.04) and people outside the family about death (OR: 5.07, 95% CI: 1.64-15.70). Depressive symptoms (OR: 1.27, 95% CI: 1.12-1.45) and time since loss (two to four years: OR: 10.36, 95% CI: 2.87-37.48 and five to seven years: OR: 8.36, 95% CI: 2.36-29.57) also predicted unresolved grief. Together, these predictors explained 54% of the variance of unresolved grief. CONCLUSION: Siblings’ perception that it was not a peaceful death and poor communication with family, friends, and healthcare increased the risk for unresolved grief among the siblings.


BACKGROUND: Although the role of pediatric palliative care (PPC) is well described in oncology, neonatal, and pediatric intensive care patients, the involvement of PPC in patients with congenital heart disease (CHD) is not well explored. CHD is a leading cause of neonatal morbidity and can cause ongoing morbidity throughout the course of a child’s life. PPC, with its focus on quality of life and longitudinal care through the course of an illness, could be of benefit to this population. OBJECTIVE: This case description reviews the role of PPC teams in the care of patients with complex CHD. DESIGN: Case study and analysis of three pediatric patients with complex CHD who also received PPC services. CONCLUSIONS: Involvement of PPC teams in patients with complex CHD can be beneficial for both families and caregivers. PPC teams can aid with advance care planning, goal setting, medical decision making, and bereavement support. Further research is needed to better quantify the benefits of PPC teams' involvement in this population.


BACKGROUND: People with chronic diseases experience barriers to managing their diseases and accessing available health services. Patient navigator programs are increasingly being used to help people with chronic diseases navigate and access health services. OBJECTIVE: The objective of this review was to summarize the evidence for patient navigator programs in people with a broad range of chronic diseases, compared to usual care. METHODS: We searched MEDLINE, EMBASE, CENTRAL, CINAHL, PsycINFO, and Social Work Abstracts from inception to August 23, 2017. We also searched the reference lists of included articles. We included original reports of randomized controlled trials of patient navigator programs compared to usual care for adult and pediatric patients with any one of a defined set of chronic diseases. RESULTS: From a total of 14,672 abstracts, 67 unique studies fit our inclusion criteria. Of these, 44 were in cancer, 8 in diabetes, 7 in HIV/AIDS, 4 in cardiovascular disease, 2 in chronic kidney disease, 1 in dementia and 1 in patients with more than one condition. Program characteristics varied considerably. Primary outcomes were most commonly process measures, and 45 of 67 studies reported a statistically significant improvement in the primary outcome. CONCLUSION: Our findings indicate that patient navigator programs improve processes of care, although few studies assessed patient experience, clinical outcomes or costs. The inability to definitively outline successful components remains a key uncertainty in the use of patient navigator programs across chronic diseases. Given the increasing popularity of patient navigators, future studies should use a consistent definition for patient navigation and determine which elements of this intervention are most likely to lead to improved outcomes. TRIAL REGISTRATION: PROSPERO #CRD42013005857.


BACKGROUND: The number of children and young people (CYP) living with life-limiting and life-threatening conditions is rising. Paediatric palliative care is a relatively new aspect of healthcare, the delivery of which is variable, with a wide range of healthcare and voluntary sector providers involved. Policy recommendations are for Specialist Paediatric Palliative Care (SPPC) services to be supported by a physician with specialist training. AIM: To examine the research evidence regarding the distinct benefits of SPPC services, with 'Specialist Paediatric Palliative Care' defined as palliative care services supported by a specialist physician. METHOD: Systematic review of studies of SPPC services published in English from 1980 to 2016. Keyword searches were carried out in medical databases (Cochrane, PubMed, EMBASE, CINAHL and AMED) and a narrative synthesis. RESULTS: Eight studies were identified, most of which were retrospective surveys undertaken within single institutions; three were surveys of bereaved parents and three were medical notes reviews. Together they represented a heterogeneous body of low-level evidence. Cross-cutting themes suggest that SPPC services improve the quality of life and symptom control and can impact positively on place of care and family support. CONCLUSIONS: Current evidence indicates that SPPC services contribute beneficially to the care and experience of CYP and their families, but is limited in terms of quantity, methodological rigour and generalisability. Further research is necessary given the significant workforce and resource implications associated with policy recommendations about the future provision of SPPC and to address the need for evidence to inform the design and delivery of SPPC services. 


INTRODUCTION: The number of children and young people living with life-limiting and life-threatening conditions is rising. Providing high-quality, responsive healthcare for them and for their families presents a significant challenge. Their conditions are often complex and highly unpredictable. Palliative care is advocated for people with life-limiting and life-threatening conditions, but these services for children are highly variable in terms of availability and scope. Little is known about the lived experiences and preferences of children and their families in terms of the palliative care that they do, or do not, receive. This study aims to produce an in-depth insight into the experiences and preferences of such children and families in order to develop recommendations for the future provision of services. The study will be carried out in the West Midlands, UK. METHODS AND ANALYSIS: A qualitative study comprising longitudinal interviews over a 12-month period with children (aged 5-18 years) living with life-limiting or life-threatening conditions and their family members. Data analysis will start with thematic analysis, followed by narrative and cross-case analysis to examine changing experiences and preferences over time, at the family level and within the wider healthcare system. Patient and public involvement (PPI) has informed the design and conduct of the study. Findings will be used to develop recommendations for an integrated model of palliative care for children in partnership with the patient and public involvement (PPI) group. ETHICS AND DISSEMINATION: Ethical approval was granted in September 2016 by the National Health Service Health Research Authority (IRAS ID: 196816, REC reference: 16/WM/0272). Findings will be of immediate relevance to healthcare providers, policy-makers, commissioners and voluntary sector organisations in the UK and internationally. Reports will be prepared for these audiences, as well as for children and their families, alongside academic outputs. 

https://bmjopen.bmj.com/content/8/1/e018266


Introduction: High incidence rates of childhood cancer and the consequent deaths in the Middle East is one of the major reasons for the need for palliative care in these countries. Using the experiences and innovations of the other countries can provide a pattern for the countries of the region and lead to the development of palliative care in children. Therefore, the aim of this study is to compare the status of pediatric palliative care in Egypt, Lebanon, Jordan, Turkey, and Iran. Materials and Methods: This is a comparative study in which the information related to pediatric palliative care system in the target countries (from 2000 to 2016) has been collected, summarized, and classified by searching in databases, such as “PubMed, Scopus, Google scholar, Ovid, and science direct.” Results: Palliative care in children in the Middle East is still in its early stages and there are many
obstacles to its development, namely, lack of professional knowledge, inadequate support of policy-makers, and lack of access to opioids and financial resources. Despite these challenges, providing services at the community level, support of nongovernmental organizations (NGOs), using trained specialists and multi-disciplinary approach is an opportunity in some countries. Conclusion: Considering the necessity of the development of pediatric palliative care in the region, solutions such as training the human resources, integrating palliative care programs into the curriculum of the related fields, establishing facilitating policies in prescription and accessibility of opioids, providing the necessary support by policy-makers, doing research on assessment of palliative care quality, as well as NGOs' participation and public education are suggested.


As the demand for pediatric palliative care (PC) increases, data suggest that Latino children are less likely to receive services than non-Latino children. Evidence on how to best provide PC to Latino children is sparse. We conducted a narrative review of literature related to PC for Latino children and their families in the United States. In the United States, Latinos face multiple barriers that affect their receipt of PC, including poverty, lack of access to health insurance, language barriers, discrimination, and cultural differences. Pediatric PC research and clinical initiatives that target the needs of Latino families are sparse, underfunded, but essential. Education of providers on Latino cultural values is necessary. Additionally, advocacy efforts with a focus on equitable care and policy reform are essential to improving the health of this vulnerable population.


Many of the 23 million individuals with heart failure (HF) worldwide receive daily, unpaid support from a family member or friend. Although HF and palliative care practice guidelines stipulate that support be provided to family caregivers, the evidence base to guide care for this population has not been comprehensively assessed. In order to appraise the state-of-the-science of HF family caregiving and recommend areas for future research, the aims of this review were to summarize (1) how caregivers influence patients, (2) the consequences of HF for caregivers, and (3) interventions directed at HF caregivers. We reviewed all literature to December 2015 in PubMed and CINAHL using the search terms "heart failure" AND "caregiver." Inclusion criteria dictated that studies report original research of HF family caregiving. Articles focused on children or instrument development or aggregated HF with other illnesses were excluded. We identified 120 studies, representing 5700 caregivers. Research on this population indicates that (1) caregiving situations vary widely with equally wide-ranging tasks for patients to help facilitate their health behaviors, psychological health and relationships, and quality of life (QoL); (2) caregivers have numerous unmet needs that fluctuate with patients' unpredictable medical status, are felt to be ignored by the formal healthcare system, and can lead to distress, burden, and reduced QoL; and (3) relatively few interventions have been developed and tested that effectively support HF family caregivers. We provide recommendations to progress the science forward in each of these areas that moves beyond descriptive work to intervention development and clinical trials testing.


AIM: To report the trend in end-of-life health services (HS) utilization among cancer patients treated in a large Australian academic cancer center over a 12-year period. METHODS: This is a retrospective study of cancer patients treated at the Peter MacCallum Cancer Centre (PMCC), who had documented death between January 2002 and December 2013. Using administrative and billing database, we report on the utilization of different categories of HS within two weeks of death: diagnostic investigations (pathology and radiology), inpatient and outpatient services, and potentially futile interventions (PFI, which include radiotherapy, chemotherapy and surgery). RESULTS: Of the 27 926 "active" cancer patients in the study (i.e. those with medical contact at PMCC in
the last year of life), 6368 (23%) had documented HS utilization within two weeks of death. 11% and 9% had pathology and radiology investigations respectively, 14% had outpatient clinic appointments, and 7% had hospital admissions. There were 2654 patients (10%) who had PFI within two weeks of death - 2198 (8%) had radiotherapy, 287 (1%) chemotherapy and 267 (1%) surgery. We observed peak HS and PFI utilization in 2004, which then dropped to its lowest in 2009/2010. CONCLUSION: Experience in an Australian cancer center suggests approximately one in four “active” cancer patients had HS utilization, and one in ten had PFI, within two weeks of death. The implementation of palliative care guidelines may reduce some of these potentially wasteful and futile interventions.


OBJECTIVES: Several barriers have been identified as preventing or delaying access to children’s palliative care services. The aim of this study is to further explore such barriers from palliative care professionals’ perspective from two London boroughs. METHODS: Qualitative-five children's palliative care professionals' perceptions were obtained from semi-structured interviews. RESULTS: Three themes emerged: availability and adequacy of child palliative care (e.g., unreliability of services), obstacles to accessing palliative care (e.g., logistical challenges), and cultural values and family priorities. CONCLUSION: These findings contribute to the equal opportunities dialogue in this sector and the need for future research to address the challenges identified.


AIMS AND OBJECTIVES: To report parent and professional perspectives of step-down care in assisting the transition from hospital to home, within one children’s hospice in a constituent country of the United Kingdom. BACKGROUND: In recent years, increasing numbers of children-dependent on long term assisted ventilation have been noted. Meeting the complex physical, emotional and social needs of the child and family is challenging. Many of these children spend extended periods in hospital even when medically stable. DESIGN: This was a qualitative study using an inductive, semantic analytic approach within a realist epistemology. METHODS: Data collection was carried out in 2013. Interviews took place with parents (n = 5) and focus groups with professionals (n = 26) who had experience of step-down care. RESULTS: Multiple benefits of step-down in the hospice were clear. Both sets of accounts suggested that for children and families life was "on hold" in hospital. Hospice was considered a home-like environment where the child and family could "live again". Parents reflected that, in hospice they were "living, not existing" while professionals highlighted hospice as nurturing and empowering the whole family, promoting the child’s development while safely meeting their clinical needs. CONCLUSIONS AND RELEVANCE TO CLINICAL PRACTICE: The study highlights a number of crucial benefits to the child and family both in the immediate and longer terms. The collective perspectives therefore endorse hospice as a potential viable choice for these children and their families during the always difficult, usually protracted transition from hospital to home.


Providing end-of-life care to children with cancer is most ideally achieved by initiating palliative care at the time of diagnosis, advocating for supportive care throughout the treatment trajectory, and implementing hospice care during the terminal phase. The guiding principles behind offering palliative care to pediatric oncology patients are the prioritization of providing holistic care and management of disease-based symptoms. Pediatric hematology-oncology nurses and clinicians have a unique responsibility to support the patient and family unit and foster a sense of hope, while also preparing the family for the prognosis and a challenging treatment trajectory that could result in the child’s death. In order to alleviate potential suffering the child may experience, there needs to be an emphasis on supportive care and symptom management. There are barriers to implementing palliative care for children with cancer, including the need to clarify the palliative care philosophy, parental acknowledgement and acceptance of a child’s disease and uncertain future, nursing awareness of services, perception of availability, and a shortage of research guidance. It is important for nurses and clinicians to
have a clear understanding of the fundamentals of palliative and end-of-life care for pediatric oncology patients to receive the best care possible.


A positive youth development perspective focuses on recognizing psychosocial strengths and providing social environments that contribute to the development of these in children and adolescents. Bereavement camps can provide such an environment as they help children cope with the death of someone close. The purpose of this study was to observe bereavement camps through the lens of positive youth development to determine the applicability of the eight features of positive developmental settings for describing bereavement camps (safety, appropriate structure, supportive relationships, opportunities to belong, positive norms, support for efficacy, skill building opportunities, integration of family and community). Observational notes were recorded by researchers during on-site visits to three different weekend bereavement camps. Results identified how each element of positive developmental settings was exemplified in either typical camp activities or bereavement-focused activities. For example, assigning campers to cabin groups based on age and gender provided opportunities to belong, and giving campers a comfort object and a big buddy provided supportive relationships. Findings were used to create a positive developmental settings observation checklist for use by bereavement camp practitioners to assess the extent to which each camp provides the requisite elements for promoting positive youth development.


PURPOSE: This study examined the provision of palliative care and related decision-making in Swiss pediatric oncology settings. The aim was to determine if and when children who died from cancer received palliative care, whether there were differences by cancer diagnosis, and inclusion of children in decision-making regarding palliative care. METHODS: Using a standardized data extraction form, a retrospective review of medical records of deceased pediatric patients was conducted. The form captured information on demographics, diagnosis, relapse(s), treatments, decision-making during palliative care, and circumstances surrounding a child’s death. RESULTS: For 170 patients, there was information on whether the child received palliative care. Among those, 38 cases (22%) did not receive palliative care. For 16 patients, palliative care began at diagnosis. The mean duration of palliative care was 145 days (Mdn = 89.5, SD = 183.4). Decision to begin palliative care was discussed solely with parent(s) in 60.9% of the cases. In 39.1%, the child was involved. These children were 13.6 years of age (SD = 4.6), whereas those not included were 7.16 years old (SD = 3.9). Leukemia patients were less likely to receive palliative care than the overall sample, and patients with CNS neoplasms received palliative care for a longer time than other patients. CONCLUSIONS: There are still high numbers of late or non-referrals, and even children older than 12 years were not involved in decision-making regarding palliative care. These results do not align with international organizational guidelines which recommend that palliative care should begin at diagnosis.


BACKGROUND: Ninety-eight percent of children needing palliative care live in low- and middle-income countries (LMICs), and almost half of them live in Africa. In contrast to the abundance of data on populations in high income countries, the current data on populations in LMICs is woefully inadequate. This study aims to identify and summarize the published literature on the need, accessibility, quality, and models for palliative care for
children in LMICs. METHODS: A scoping review was performed following the method of Arksey and O’Malley. Systematic searches were conducted on PubMed and Google Scholar using the main keywords, ‘children AND palliative care OR terminal care OR hospice OR end of life AND developing countries OR LMICs.’ Additional publications were obtained by handsearching. Papers were only included if they reported on the need, accessibility, quality, and models for palliative care for children in LMICs. RESULTS: Fifteen papers met the inclusion criteria for review. Of these, 10 assessed need, seven examined availability and/or accessibility, one assessed quality, and one examined the models. We found an urgent need for palliative care, particularly in the training for health workers and improving poor availability and/or accessibility to palliative care in terms of factors such as medication and bereavement support. The best practice models demonstrated feasibility and sustainability through cooperation with governments and community organizations. The quality of pain management and emotional support was lower in LMICs compared to HICs. CONCLUSION: Although we found limited evidence in this review, we identified common challenges such as the need for further training for health workers and greater availability of opioid analgesics. While efforts to change the current systems and laws applying to children in LMICs are important, we should also tackle underlying factors including the need to raise awareness about palliative care in public health and improve the accuracy of data collection.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5702244/


A substantial number of children cared for by pediatric palliative care physicians have progressive non-malignant conditions. Some elements of their care overlap with care for children with cancer while other elements, especially prognosis and trajectory, have nuanced differences. This article reviews the population, physical-emotional and social concerns, and trajectory.


Diffuse intrinsic pontine gliomas (DIPGs) are rare but devastating brain tumors that occur primarily in children. These gliomas have poor prognoses and present options focus on palliation of symptoms and prolongation of life. Here, we present a case of a 16-year-old female diagnosed with a DIPG whose age group has been mostly left out of discussions regarding psychosocial support options. This report is meant to start a conversation about the different support options available at our institution that have shown promising results in the literature for palliative care applications. These options can include camps for patients with brain tumors, psychological counseling, the Ronald McDonald House, and other psychosocial programs. Many of these programs can be tailored to meet the specific needs of adolescent and young adult (AYA) patients and will hopefully be integrated into a comprehensive palliative care regimen in future studies.


PURPOSE OF REVIEW: Pediatric palliative oncology (PPO) is an emerging field that integrates the principles of palliative care early into the illness trajectory of children with cancer. PPO providers work with interdisciplinary clinicians to provide optimal medical and psychosocial care to children with cancer and their families. Ongoing advances in the field of pediatric oncology, including new treatment options for progressive cancers, necessitate the early integration of palliative care tenets including holistic care, high-quality communication, and assessment and management of refractory symptoms. RECENT FINDINGS: Research in this emerging field has expanded dramatically over the past several years. This review will focus on advancements within several key areas of the field, specifically regarding investigation of the communication needs and preferences of patients and families, exploration of educational initiatives and interventions to teach PPO principles to clinicians, study of patient-reported and parent-reported tools to better assess and manage refractory symptoms, and development of novel models to integrate palliative care within pediatric oncology. SUMMARY: Research findings in the field of PPO, concurrent with advances in the treatment of pediatric cancer, may help improve survival and quality of life for children with cancer.

Adolescent and young adult (AYA) oncology patients experience many physical and psychological symptoms at the end of life (EOL); however, data on these experiences for AYA patients who have undergone hematopoietic cell transplantation (HCT) remains sparse. We sought to investigate the characteristics of AYA patients aged 15-25 years who received allogeneic HCT and subsequently died while inpatient at our institution between the years 2008 and 2014. A standardized data extraction tool was used to collect information about patient demographics, treatment and symptoms. We found that during this time frame, 34 AYA patients had received HCT and died while inpatient at our institution, 23 (68%) of whom died because of treatment-related complications. Compared with non-HCT AYA oncology patients (n=35), patients who received HCT (n=34) were more likely to have died in the intensive care unit (71% vs 23%, P<.0001) and to have received mechanical ventilation (68% vs 17%, P<0.0001) or hemodialysis (53% vs 0%, P<0.0001) in the last 30 days of life. These findings demonstrate that AYA patients who receive allogeneic HCT receive intensive EOL treatment, suggesting that these patients may benefit from early integration of expert interdisciplinary services to prospectively assess and manage distressing symptoms.


AIM: We compared neonatal deaths and end-of-life decisions in a neonatal intensive care unit (NICU) and paediatric intensive care unit (PICU) in a Dutch tertiary children's hospital. SUBJECTS: All 235 full-term infants who died within 28 days of life between 2003 and 2013 in the NICU (n = 199) and PICU (n = 36) were retrospectively studied. RESULTS: The median length of stay was three days in the NICU and seven days in the PICU (p = 0.003). The main reasons for NICU stays were asphyxia (52.8%) and congenital malformations (42.2%), and in the PICU, they were congenital malformations (97.2%) and primarily cardiac problems (83.3%, p < 0.001). The median age of death was three days in the NICU and eight days in the PICU (p < 0.001), and mortality despite full intensive care treatment was 4.0% and 25.0%, respectively. Intensive treatment was discontinued because of poor survival chances in 25.1% of NICU and 52.8% of PICU cases (p < 0.001), and care was redirected because of expected poor quality of life in 70.9% and 22.2%, respectively. CONCLUSION: Differences between the age at death and end-of-life decisions were found between full-term infants in the NICU and PICU in the same children's hospital. Underlying disorders and doctors’ attitudes may have played a role.


Solanke, F., A. Colver and H. McConachie (2018). "Are the health needs of young people with cerebral palsy met during transition from child to adult health care?" Child Care Health Dev.

BACKGROUND: The transition from child to adult health care is a particular challenge for young people with cerebral palsy, who have a range of needs. The measurement of reported needs, and in particular unmet needs, is one means to assess the effectiveness of services. METHODS: We recruited 106 young people with cerebral palsy, before transfer from child services, along with their parents to a 3-year longitudinal study. Reported needs were measured with an 11-item questionnaire covering speech, mobility, positioning, equipment, pain, epilepsy, weight, control of movement, bone or joint problems, curvature of the back, and eyesight. Categorical principal component analysis was used to create factor scores for bivariate and regression analyses. RESULTS: A high level of reported needs was identified particularly for control of movement, mobility, and equipment, but these areas were generally being addressed by services. The highest areas of unmet needs were for management of pain, bone or joint problems, and speech. Analysis of unmet needs yielded two factor scores, daily living health care and medical care. Unmet needs in daily living health care were related to severity of motor impairment and to attending nonspecialist education. Unmet needs tended to increase over time but were not significantly (p > .05) related to whether the young person had transferred from child services. CONCLUSIONS: Reporting of unmet needs can indicate where service development is required, and we have shown that the approach to measurement can be improved. As the number of unmet health needs at the start of transition is considerable, unmet health needs after transition cannot all be attributed to poor transitional health care. The range and

BACKGROUND: Improving child survival for HIV-infected children remains an important health agenda. We present progress regarding care and treatment services to HIV infected children in Tanzania. METHODS: The National AIDS Control Programme Care and Treatment (CTC 2) database was used to obtain information of all children aged 0-14 years enrolled in the HIV Care and Treatment Program between January 2011 and December 2014. We assessed eligibility for ART, time from enrolment to ART initiation, nutritional status, and mortality using Kaplan-Meier methods. RESULTS: A total of 29,531 (14,304 boys and 15,227 girls) ART-naive children aged 0-14 years were enrolled during the period, approximately 6700 to 8000 children per year. The male to female ratio was 48:50. At enrolment 72% were eligible for ART, 2-3% of children were positive for TB, and 2-4% were severely malnourished. Between 2011 and 2014, 2368 (8%) died, 9243 (31%) were Lost to Follow-up and 17,920 (61%) were on care or ART. The probability of death was 31% (95% CI 26-35), 43% (40-47), 52% (49-55) and 61% (58-64) by 1.2, 5 and 10 years of age, respectively. The hazard of death was greatest at very young ages (<2 years old), and decreased sharply by 4 years old. Children who were on ART had around 10-15% higher survival over time.

CONCLUSIONS: Significant progress has been made regarding provision of paediatric HIV care and treatment in Tanzania. On average 7000 children are enrolled annually, and that approximately two thirds of children diagnosed under the age of 2 years were initiated on ART within a month. Provision of ART as soon as the child is diagnosed is the biggest factor in improving survival. However we noted that i) most children had advanced disease at the time of enrolment ii) approximately two-thirds of children were missing a baseline CD4 measurement and only 35% of children had either a CD4 count or percentage recorded, indicating limited access to CD4 testing services, and iii) 31% were lost to follow-up (LTFU). These challenges need to be addressed to improve early detection, enrolment and retention of HIV-infected children into care and improve documentation of services offered.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5547461/


OBJECTIVES: Our aim in this study was to understand usage patterns of pediatric palliative care (PPC) consultation and associations with end-of-life preparation among pediatric patients who are deceased. METHODS: We reviewed 233 pediatric mortalities. Data extraction from the electronic health record included determination of PPC consultation by using Current Procedural Terminology codes. Diagnoses were identified by International Classification of Disease codes and were classified into categories of life-threatening complex chronic conditions (LT-CCCs). Data analysis included Student’s t test, Wilcoxon rank test, Fisher’s exact test, chi(2) test, and multivariable logistic regression. RESULTS: The overall PPC consultation rate for pediatric patients who subsequently died was 24%. A PPC consultation for patients admitted to the pediatric ward and PICU was more likely than for patients cared for in the NICU (31% vs 12%, P < .01) and was more likely for those with an LT-CCC (40% vs 10%, P < .01), particularly malignancy (65% vs 35%, P < .01). Also noted were increased completion of Physician Orders for Life-Sustaining Treatment forms (8 vs 0, P < .01) and increased documentation of mental health disorders (60% vs 40%, P = .02). CONCLUSIONS: Our findings suggest that PPC consultation for patients in the pediatric ward and PICU is more likely among patients with a greater number of LT-CCCs, and is associated with increased Physician Orders for Life-Sustaining Treatment preparation and documentation of mental health disorders. Patients at risk to not receive PPC consultation are those with acute illness and patients in the NICU.


BACKGROUND: Children receiving hospice and palliative care (HPC) differ from adults in important ways. Children are more likely to have rare diagnoses, less likely to have cancer, have longer lengths of stay on hospice, and are
more likely to be technology dependent than adults. The National Consensus Project (NCP) in Palliative Care established domains of quality for HPC, but these domains have not been evaluated for applicability in children. OBJECTIVES: This study aims to establish consensus stakeholder-prioritized domains of high-quality pediatric home-based hospice and palliative care (HBHPC). DESIGN: Mixed methods design. SETTING/SUBJECTS: Providers from the Ohio Pediatric Palliative Care and End-of-life Network. MEASUREMENTS: Using a modified Delphi technique, providers were surveyed regarding the NCP quality domains for HPC. RESULTS: There was strong consensus on the applicability of each domain to the participants’ practices (median scores ranged from 0.97 to 1.0 with interquartile ranges = 0). Consensus on the rank importance of the eight domains was not achieved. Qualitative data included challenges with NCP domain 3 (Psychological and Psychiatric Aspects of Care). It was recommended that titles should remain consistent with adult standards, but domain definitions should be broadened for pediatric HBHPC. Continuity and coordination of care should be added as a ninth domain of quality in pediatric HBHPC. CONCLUSIONS: All eight NCP domains were validated in pediatric HBHPC. A ninth domain, Continuity and Coordination of Care, was also added. Ranking the domains was not recommended as consensus indicated weighting them as equally integrated standards. Future studies are needed to evaluate parent- and patient-prioritized domains of quality in pediatric HBHPC and to validate and map pediatric-specific indicators to these domains.


Background: Up to 90 % of patients with congenital heart disease (CHD) now reach adulthood. To avoid lapses in care during the change from pediatric to adult care, a nurse-led transition program (TP) was implemented at a Swiss University Hospital. Aim: This study explored the experiences and expectations of adolescents with CHD and their parents regarding a nurse-led TP. Method: This qualitative study used an interpretive, phenomenological approach. Individual interviews were conducted with seven adolescent CHD patients in the transition period and their parents (six mothers, two fathers). Analysis followed an iterative process. Results: For most study participants, the transfer from pediatric to adult medicine as part of the TP went smoothly. They experienced the TP positively. Patients valued the provision of a constant contact person to provide CHD-related information; parents welcomed the support of an informed, neutral clinician for their children. To varying degrees, adolescents were willing to take over self-responsibility; conversely, parents found it difficult to turn their responsibility over to their children. Parents wished to give the adolescent as much time as needed to act responsibility on their own. Conclusions: A transition program is a key element for establishing a continuous care in adolescents with a chronic disease. It facilitates the parents' process of allowing their youths to assume increasing responsibility for their own health.


Paediatric palliative care is no longer restricted to patients with cancer and has been extended to patients with other chronic conditions, such as cystic fibrosis or neuromuscular disorders. This review focused on the current state of palliative care for children and adolescents with chronic kidney disease (CKD). We assessed the literature on CKD published up to August 2017. All the papers, except one from 1996, were published this century. This review discusses the role that palliative care plays in the process of decision-making and explores the possibilities of implementing palliative care into the routine therapy of affected patients and providing support for their families. Offering early palliative care as an integral part of the kidney, supportive care provided by the nephrology care team is both necessary and feasible for patients with CKD. As a minimum, a specialised palliative care team should be involved in patients with multiple comorbidities, in conservative treatment scenarios and in acute life-threatening complications. Further studies and guidelines are required to improve the care of patients with CKD and their families. CONCLUSION: Supportive palliative care should be implemented into the routine care of patients with life-limiting kidney disease.

https://www.ncbi.nlm.nih.gov/pubmed/29220099

BACKGROUND: In 2007, the European Association of Palliative Care (EAPC) provided a comprehensive set of recommendations and standards for the provision of adequate pediatric palliative care. A number of studies have shown deficits in pediatric palliative care compared to EAPC standards. In Germany, pediatric palliative care patients can be referred to specialized outpatient palliative care (SOPC) services, which are known to enhance quality of life, e.g. by avoiding hospitalization. However, current regulations for the provision of SOPC in Germany do not account for the different circumstances and needs of children and their families compared to adult palliative care patients. The “Evaluation of specialized outpatient palliative care (SOPC) in the German state of Hesse (ELSAH)” study aims to perform a needs assessment for pediatric patients (children, adolescents and young adults) receiving SOPC. This paper presents the study protocol for this assessment (work package II).

METHODS/DESIGN: The study uses a sequential mixed-methods study design with a focus on qualitative research. Data collection from professional and family caregivers and, as far as possible, pediatric patients, will involve both a written questionnaire based on European recommendations for pediatric palliative care, and semi-structured interviews. Additionally, professional caregivers will take part in focus group discussions and participatory observations. Interviews and focus groups will be tape- or video-recorded, transcribed verbatim and analyzed in accordance with the principles of grounded theory (interviews) and content analysis (focus groups). A structured field note template will be used to record notes taken during the participatory observations. Statistical Package for Social Sciences (SPSS, version 22 or higher) will be used for descriptive statistical analyses. The qualitative data analyses will be software-assisted by MAXQDA (version 12 or higher). DISCUSSION: This study will provide important information on what matters most to family caregivers and pediatric patients receiving SOPC. The results will add valuable knowledge to the criteria that distinguish SOPC for pediatric from SOPC for adult patients, and will provide an indication of how the German SOPC rule of procedure can be optimized to satisfy the special needs of pediatric patients. TRIAL REGISTRATION: Internet Portal of the German Clinical Trials Register ( www.germanctr.de , DRKS-ID: DRKS00012431).


BACKGROUND: Parents of children with a life-limiting disease have to rely on themselves at home while adequate paediatric palliative care is lacking. In several countries, paediatric palliative care teams are introduced to ensure continuity and quality of care and to support the child and the family. Yet, little is known about how parents experience such multidisciplinary teams. AIM: To obtain insight into the support provided by a new paediatric palliative care team from the parents’ perspective. DESIGN: An interpretative qualitative interview study using thematic analysis was performed. SETTING/PARTICIPANTS: A total of 47 single or repeated interviews were undertaken with 42 parents of 24 children supported by a multidisciplinary paediatric palliative care team located at a university children’s hospital. The children suffered from malignant or non-malignant diseases. RESULTS: In advance, parents had limited expectations of the paediatric palliative care team. Some had difficulty accepting the need for palliative care for their child. Once parents experienced what the team achieved for their child and family, they valued the team’s involvement. Valuable elements were as follows: (1) process-related aspects such as continuity, coordination of care, and providing one reliable point of contact; (2) practical support; and (3) the team members’ sensitive and reliable attitude. As a point of improvement, parents suggested more concrete clarification upfront of the content of the team’s support. CONCLUSION: Parents feel supported by the paediatric palliative care team. The three elements valued by parents probably form the structure that underlies quality of paediatric palliative care. New teams should cover these three valuable elements.


BACKGROUND: Telephone surveys are intended to reduce attrition in longitudinal studies. For paediatric chronic pain patients, the comparability of pain-related information gathered using telephone interviews and postal surveys remain unknown. Furthermore, it remains unknown how social desirability may influence answers. METHODS: To compare data from telephone interviews and postal surveys, a randomized cross-over design with two measure points 2 weeks apart and four conditions (combinations of telephone interviews (T) and postal surveys (P): P-T, T-P, P-P, T-T) was conducted in a sample of N = 323 paediatric chronic pain patients. RESULTS: In the inter-group comparison, pain-related information did not differ between telephone interviews and postal surveys except for the information on pain location (back and extremities). Agreement measures of the intra-group comparisons suggest substantial to excellent agreements for all items and did not differ between the groups. The internal consistency of a disability scale was excellent for both assessment modes; the number of missing values did not differ. Participation rate was higher for telephone interviews compared to the postal surveys. Across both time points, attrition was lowest for the groups without a switch in assessment mode compared to the groups with a switch in assessment mode. Except for pain-related school absence, no effect of social desirability occurred. CONCLUSIONS: Telephone interviews are a useful method to achieve a high response rate. Pain locations should be asked for separately and not in an open question when interviewing children and adolescents on the telephone. SIGNIFICANCE: Telephone interviews are a good method to achieve a high response rate and obtain valid data in studies with paediatric chronic pain patients.


OBJECTIVES: Pediatric subspecialty care, including multidisciplinary palliative care, tends to be located in urban academic centers or children’s hospitals. Telehealth provides the opportunity to care for patients who would otherwise not be able to access services. We present cases wherein telehealth was used to provide counseling services to patients who would not have been able to receive this service. METHODS: We discuss cases of telehealth use for patient and family counseling in the setting of palliative care and bereavement follow-up. Patients who live a great distance from the hospital with limited access to services were followed by a hospital-based pediatric palliative care team. Patients and families gave feedback after use of telehealth for counseling services. RESULTS: Counseling through telehealth by our hospital-based palliative care social worker was successful for all parties involved: patient, family, and social worker. CONCLUSIONS: Telehealth helps relieve disparity in access to services and care, which is particularly problematic in pediatrics and mental health. For the patients in this case series, it was an effective modality to receive counseling services and meet needs that otherwise would not have been addressed.


Pediatric Neurocritical Care diagnoses account for a large proportion of intensive care admissions. Critical care survivors suffer high rates of long-term morbidity, including physical disability, cognitive impairment, and psychosocial dysfunction. To address these morbidities in Pediatric Neurocritical Care survivors, collaboration between Pediatric Neurology and Pediatric Critical Care created a multidisciplinary follow-up clinic providing specialized evaluations after discharge. Clinic referrals apply to all Pediatric Neurocritical Care patients regardless of admission severity of illness. Here, we report an initial case series, which revealed a population that is heterogenous in age, ranging from 1 month to 18 years, and in diagnoses. Traumatic brain injuries of varying severity as well as neuroinfectious and inflammatory diseases accounted for the majority of referrals. Most patients (87%) seen in the clinic had morbidities identified, requiring ongoing evaluation and expansion of the clinic. Cognitive and psychological disturbance were seen in over half of patients at the initial clinic follow-up. Sleep disturbances, daytime fatigue, headache or chronic pain, and vision or hearing concerns were also common at initial follow-up. Data from this initial population of clinic patients reiterates the need for specialized follow-up care, but also highlights the difficulties related to providing this comprehensive care and evaluating interventions to improve outcomes.


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