When a child’s life is expected to be short, there’s no time to waste. Together for Short Lives is here to make sure the 49,000 seriously ill children and their families across the UK can make the most of every moment they have together, whether that’s for years, months or only hours. We stand alongside families, supporting them to make sure they get the vital care and help that they need.

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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.

Services include:

- postal loans
- obtaining journal articles
- literature searching
- current awareness bulletin
- advice on accessing NHS electronic resources.

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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Clinical and Ethical Decision Making


**BACKGROUND:** Very preterm birth (24 to < 32 week’s gestation) is a major public health issue due to its prevalence, the clinical and ethical questions it raises and the associated costs. It raises two major clinical and ethical dilemma: (i) during the perinatal period, whether or not to actively manage a baby born very prematurely and (ii) during the postnatal period, whether or not to continue a curative treatment plan initiated at birth. The Wallonia-Brussels Federation in Belgium counts 11 neonatal intensive care units.

**METHODS:** An inventory of key practices was compiled on the basis of an online questionnaire that was sent to the 65 neonatologists working in these units. The questionnaire investigated care-related decisions and practices during the antenatal, perinatal and postnatal periods, as well as personal opinions on the possibility of standardising and/or legislating for end-of-life decisions and practices. The participation rate was 89% (n = 58).

**RESULTS:** The results show a high level of homogeneity pointing to overall agreement on the main principles governing curative practice and the gestational age that can be actively managed given the current state of knowledge. There was, however, greater diversity regarding principles governing the transition to end-of-life care, as well as opinions about the need for a common protocol or law to govern such practices.

**CONCLUSION:** Our results reflect the uncertainty inherent in the complex and diverse situations that are encountered in this extreme area of clinical practice, and call for qualitative research and expert debates to further document and make recommendations for best practices regarding several “gray zones” of end-of-life care in neonatology, so that high quality palliative care may be granted to all neonates concerned with end-of-life decisions.


Dying adolescents presenting for palliative procedures have complicated developmental and ethical issues, especially when reconsidering do-not-resuscitate orders. Though the American Academy of Pediatrics has guidelines, there is limited information in the literature on how to take care of these patients. We describe the case of a 14-year-old patient presenting to the interventional radiology suite for management of superior vena cava syndrome. The patient’s goals of treatment were elucidated through a comprehensive care team consisting of the procedural and oncology teams. Effective communication with the patient and family was paramount for success.


Decisions regarding whether or not to pursue experimental therapies or life-sustaining medical treatment of children with life-limiting illness can be a significant source of distress and conflict for both families and health care providers. This article reviews the concepts of parental permission (consent), assent, and emerging capacity and how they relate to decision-making for minors with serious illness. Decision-making capacity for adolescents is discussed generally and in the context of emotionally charged situations pertaining to the end of life. Strategies for minimizing conflict in situations of disagreement between children and families are provided.

Anoxic brain injury in children is a rare and devastating occurrence. Families are shocked by the unexpected nature of their child's neurologic injury, which may be the result of a sudden and prolonged cardiac arrest. Organ donation in these children is subject to much discussion and controversy. Recently, we encountered three pediatric patients with anoxic brain damage who progressed to brain death within a few days of admission. Pediatric palliative care was involved from the time of arrival to the hospital in all the patients. The team served as a critical conduit to support families and helped in managing end-of-life decisions including organ donation. All three families consented to organ donation. We discuss here the patients, the palliative care involvement, and the factors responsible for successful donation.

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. Further optimization of referral rates may aid in increasing 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.

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 consultants 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.


The determination of death by neurological criteria remains controversial scientifically, culturally, and legally, worldwide. In the United Kingdom, although the determination of death by neurological criteria is not legally codified, the Code of Practice of the Academy of Medical Royal Colleges is customarily used for neurological (brainstem) death determination and treatment withdrawal. Unlike some states in the US, however, there are no provisions under the law requiring accommodation of and respect for residents’ religious rights and commitments when secular conceptions of death based on medical codes and practices conflict with a traditional concept well-grounded in religious and cultural values and practices. In this article, we analyse the medical, ethical, and legal issues that were generated by the recent judgement of the High Court of England and Wales in Re: A (A Child) [2015] EWHC 443 (Fam). Mechanical ventilation was withdrawn in this case despite parental religious objection to a determination of death based on the code of practice. We outline contemporary evidence that has refuted the reliability of tests of brainstem function to ascertain the two conjunctive clinical criteria for the determination of death that are stipulated in the code of practice: irreversible loss of capacity for consciousness and somatic integration of bodily biological functions. We argue that: (1) the tests of brainstem function were not properly undertaken in this case; (2) the two conjunctive clinical criteria set forth in the code of practice cannot be reliably confirmed by these tests in any event; and (3) absent authentication of the clinical criteria of death, the code of practice (in fact, although implicitly rather than explicitly) wrongly invokes a secular definition of death based on the loss of personhood. Consequently, the moral obligation of a pluralistic society to honor and respect diverse religious convictions to the greatest extent possible is being violated. Re A (A Child) is contrasted with the US case of Jahi McMath in which the court accommodated parental religious objection to the determination of neurological death codified in the Uniform Determination of Death Act. We conclude that the legal system in the United Kingdom should not favour a secular definition of death over a definition of death that is respectful of religious values about the inviolability and sanctity of life. We recommend the legal recognition of religious accommodation in death determination to facilitate cultural sensitivity and compassionate care to patients and families in a pluralistic society.


Much of the commentary in the wake of the Charlie Gard litigation was aimed at apparent shortcomings of the law. These include concerns about the perceived inability of the law to consider resourcing issues, the vagueness of the best interests test and the delays and costs of having disputes about potentially life-sustaining medical treatment resolved by the courts. These concerns are perennial ones that arise in response to difficult cases. Despite their persistence, we argue that many of these criticisms are unfounded. The first part of this paper sets out the basic legal framework that operates when parents seek potentially life-sustaining treatment that doctors believe is against a child’s best interests, and describes the criticisms of that framework. The second part of the paper suggests an alternative approach that would give decision-making power to parents, and remove doctors’ ability to unilaterally withhold or withdraw life-sustaining treatment that they regard is futile. This proposal is grounded in several values that we argue should guide these regulatory choices. We also contend that the best interests test is justifiable and since the courts show no sign of departing from it, the focus should be on how to better elucidate the underlying
values driving decisions. We discuss the advantages of our proposed approach and how it would address some of the criticisms aimed at the law. Finally, we defend the current role that the judiciary plays, as an independent state-sanctioned process with a precedent-setting function.


OBJECTIVE: A personal reflection on the changing landscape with regard to case mix, care, and staffing and how mortality and expectations have evolved over the past 30 years in a multidisciplinary Pediatric Critical Care Unit in a Quaternary level academic institution in Canada.

CONCLUSIONS: Many of the preventable deaths have been prevented with Public Health initiatives. Death now is increasingly in complex patients with complicated treatment regimes in a society that has increasingly unrealistic expectations of what modern medicine can do. Many of these complex children do not die but are dependent on our technology and skill set-something we are often ill prepared for.


OBJECTIVE: To develop and validate the Test of Ethics Knowledge in Neonatology (TEK-Neo) with good internal consistency reliability, item performance, and construct validity that reliably assesses interprofessional staff and trainee knowledge of neonatal ethics. STUDY

DESIGN: We adapted a published test of ethics knowledge for use in neonatology. The novel instrument had 46 true/false questions distributed among 7 domains of neonatal ethics: ethical principles, professionalism, genetic testing, beginning of life/viability, end of life, informed permission/decision making, and research ethics. Content and correct answers were derived from published statements and guidelines. We administered the voluntary, anonymous test via e-mailed link to 103 participants, including medical students, neonatology fellows, neonatologists, neonatology nurses, and pediatric ethicists. After item reduction, we examined psychometric properties of the resulting 36-item test and assessed overall sample performance.

RESULTS: The overall response rate was 27% (103 of 380). The test demonstrated good internal reliability (Cronbach alpha = 0.68), with a mean score of 28.5 +/- 3.4 out of the maximum 36. Participants with formal ethics training performed better than those without (30.3 +/- 2.9 vs 28.1 +/- 3.5; P = .01). Performance improved significantly with higher levels of medical/ethical training among the 5 groups: medical students, 25.9 +/- 3.7; neonatal nurses/practitioners, 27.7 +/- 2.7; neonatologists, 28.8 +/- 3.7; neonatology fellows, 29.8 +/- 2.9; and clinical ethicists, 33.0 +/- 1.9 (P < .0001).

CONCLUSIONS: The TEK-Neo reliably assesses knowledge of neonatal ethics among interprofessional staff and trainees in neonatology. This novel tool discriminates between learners with different levels of expertise and can be used interprofessionally to assess individual and group performance, track milestone progression, and address curricular gaps in neonatal ethics.


OBJECTIVE: To consider whether and how family members and clinicians discuss end of life during paediatric palliative care consultations.

METHODS: Nine naturally occurring paediatric palliative care consultations were video recorded and analysed using conversation analytic methods.

ANALYSIS: Focusing on three consultations in which end of life was treated as a certain outcome, analysis explored ways in which end of life was made either implicit or explicit within these consultations. Our analysis suggests that end of life was made explicit when: 1) ancillary to the current focus of discussion, 2) in relation to someone else’s child, or 3) specifically relevant to the local context of the discussion. More commonly, in all other instances in the data, end of life was made implicit during discussions relating to this matter. CONCLUSION: This preliminary research indicates that the local context of a conversation can influence how end of life is mentioned and discussed.
PRACTICE IMPLICATIONS: Clinicians often are encouraged to promote honest and ‘open’ discussions about end of life. Our findings show that it is not necessary to explicitly mention end of life in order to discuss it.


Figueroa Gray, M., E. J. Ludman, T. Beatty, A. R. Rosenberg and K. J. Wernli (2018). "Balancing Hope and Risk Among Adolescent and Young Adult Cancer Patients with Late-Stage Cancer: A Qualitative Interview Study." J Adolesc Young Adult Oncol.

PURPOSE: Previous studies have called for further research to explore adolescent and young adult (AYA) decision-making in the context of advanced cancer to understand the perspectives of this understudied population. We conducted a qualitative study with patients and providers to better understand the decision-making experience of AYA patients with advanced stages of cancer.

METHODS: Semistructured qualitative telephone interviews were conducted from April 2016 to October 2016. English-speaking AYAs and healthcare providers were recruited through the social media sites Twitter and Facebook. AYAs were eligible if they were aged 18-39 years at diagnosis and self-reported having metastatic cancer; any provider who worked with AYAs with metastatic cancer was eligible. Researchers with expertise in qualitative methods conducted inductive thematic content analysis of transcribed interviews. The analyzed data were used to formulate recommendations for clinicians.

RESULTS: Twelve AYA patients with self-reported stage IV cancer and five clinicians who care for AYAs with advanced stages of cancer were enrolled and shared their experience about AYA medical decision-making. Four primary themes emerged: (1) AYAs describe receiving unclear prognosis, (2) AYAs balance concepts of hope and risk, (3) AYAs choose aggressive treatment options, and (4) AYAs want support facing mortality. Recommendations for clinicians include clear communication about prognosis and side effects and concerted efforts to elicit patient values.

CONCLUSION: AYA patients and clinicians provided insights into the experiences and decision-making processes of AYA patients choosing to continue or discontinue treatment and into the areas for improvement in patient-centered oncology care. Taken together, these data provide important suggestions for clinicians caring for this vulnerable population.


AIM: When parents-to-be are faced with a terminal prenatal diagnosis, they are confronted with the decision either to continue the pregnancy or to terminate it at an advanced stage. This difficult decision is intimately affected by the experience of the inevitability of loss, and ethical dilemmas posed in this usually completely unexpected situation. Studies indicate that perinatal child loss due to lethal foetal anomalies is a major life event and a source of serious psychological issues, which can last for many years after the experience. Moreover, it has been shown that care for bereaved parents, if guided by their needs, can ease their burden, regardless of whether they choose to end or continue the pregnancy. The aim of this study is to inspect current practices of counselling and support of affected families and develop practical guidelines for health and social professionals involved.

METHODS: A sample of 32 parents in the German-speaking part of Switzerland was investigated between December 2012 and March 2014. Semi-structured problem-centred interviews were conducted, transcribed verbatim and thematically analysed.

RESULTS: 4 main time periods and 6 themes were identified by participants ranging from diagnosis until birth: "shock", "choices and dilemmas", "taking responsibility", "still being pregnant", "saying goodbye/letting go" and "planning the future". However, findings reflect critical points of care and showed gaps not only between emphasising time periods but also between affected parents’ and involved professionals’ views. This article reports the findings from the parents.

CONCLUSION: This study provided new insights into parental responses when they are confronted with a fatal prenatal diagnosis. The results contribute towards rethinking current practices in midwifery and other healthcare during pregnancy, birth and puerperium as well as the palliative care of the child.

https://www.ncbi.nlm.nih.gov/pubmed/28129658

The end-of-life litigation involving Alfie Evans (9 May 2016 - 28 April 2018) from Liverpool, England, who suffered from an incurable and degenerative neurological condition was extraordinary. It emerged in the shadow of comparable but not as extensive litigation enabled by crowdfunding in relation to Ashya King and Charlie Gard. Although Alfie’s parents lost repeatedly in the High Court, the Court of Appeal and the Supreme Court of England, as well as before the European Court of Human Rights, they persisted in bringing more legal challenges. The public relations campaign on their behalf at times was threatening and accusatory of the clinicians and of Alder Hey Hospital. Both persons employed at the Christian Legal Centre, which represented the parents at times, and medical practitioners from Europe who participated in forensic assessments behaved unethically. There are many lessons to be learned from the Alfie Evans saga. If we are to maintain morale and commitment among those who provide paediatric clinical services to the very ill and the dying, they must be protected from the public relations and litigation campaigns deployed by those purporting to represent the Alfie Evans family, and better non-adversarial methods need to be constructed as a matter of urgency to resolve matters involving disagreements about the treatment of terminally ill children.


This paper argues that Charlie Gard’s parents should have been the decision-makers about their son’s best interests and that determination of Charlie’s best interests depended on a moral decision about which horn of a profound moral dilemma to choose. Charlie’s parents chose one horn of that moral dilemma and the courts, like Charlie Gard’s doctors, chose the other horn. Contrary to the first UK court’s assertion, supported by all the higher courts that considered it, that its judgement was ‘objective’, this paper argues that the judgement was not and could not be ‘objective’ in the sense of objectively correct but was instead a value judgement based on the judge’s choice of one horn of the moral dilemma. While that horn was morally justified so too was the horn chosen by the parents. The court could and should have avoided depriving the parents of their normal moral and legal right and responsibility to decide on their child’s best interests. Instead, this paper argues that the court should have acknowledged the lawfulness of both horns of the moral dilemma and added to its judgement that Charlie Gard’s doctors were not legally obliged to provide treatment that they believed to be against their patient’s best interests the additional judgement that Charlie’s parents could lawfully transfer his care to other doctors prepared to offer the infant a trial of the experimental treatment requested by his parents.


The public often believes that parents have a right to make medical decisions about their child. The idea that, in respect of children, doctors should do what parents tell them to do is problematic on the face of it. The effect of such a claim would be that a doctor who acted deliberately to harm a child would be making a morally correct decision, providing only that it is what the child’s parents said they wanted. That is so obviously nonsense that it cannot be what people who claim it actually mean. In this paper, I suggest that the claim actually represents either or both of two misunderstandings. It can be a result of wrongly appealing to the principle of respect for autonomy, or a belief that doctors are not committed to acting in the interests of the child. In this paper, I show that, while neither belief is entirely justified, there are elements of truth in both. I argue that if ethically correct decisions are those that are directed to improving the quality of a child’s existence, then neither parents nor doctors are in a position to make ethically correct decisions about a child except in discussion with one another. Where such discussion is not possible, I suggest there should be a national Children’s Interests Panel to agree on the child’s interests. The panel should include, but not be limited to, paediatricians and lawyers and its decisions should be legally binding on all parties.


Patient race/ethnicity affects health care utilization, provider trust, and treatment choice. It is uncertain how these influences affect pediatric care. We performed a systematic review (PubMed, Scopus, Web of Science, PsycINFO, Cochrane, and Embase) for articles examining race/ethnicity and parental treatment decision-making, adhering to PRISMA methodology. A total of 9200 studies were identified, and 17 met inclusion criteria. Studies focused on treatment decisions concerning end-of-life care, human papillomavirus vaccination, urological surgery, medication regimens, and dental care. Findings were not uniform between studies; however, pooled results showed (1) racial/ethnic minorities tended to prefer more aggressive end-of-life care; (2) familial tradition of neonatal circumcision influenced the decision to circumcise; and (3) non-Hispanic Whites were less likely to pursue human papillomavirus vaccination but more likely to complete the vaccine series if initiated. The paucity of studies precluded overarching findings regarding the influence of race/ethnicity on parental treatment decisions. Further investigation may improve family-centered communication, parent engagement, and shared decision-making.


An advance care plan (ACP) is the record of a discussion between an individual (where possible), their professional care givers and those close to them about their future care. When performed well, the process provides all those involved with the opportunity to talk honestly about the future allowing children and their families to retain autonomy and to influence how they are looked after. While this may represent a difficult area of practice for healthcare professionals, both staff and families appear to benefit when the process is fully informed and the child and family are actively involved. This article is enriched by the insight of two bereaved parents, who have engaged actively with the process of advance care planning. As a multidisciplinary writing team, we aim to share our experiences, in the context of recent national guidance, on the use of ACPs.


BACKGROUND: Our objective was to evaluate children with metabolic diseases in paediatric palliative home care (PPC) and the process of decision-making. This study was conducted as single-centre retrospective cohort study of patients in the care of a large specialized PPC team.

RESULTS: Between 01/2013 and 09/2016, 198 children, adolescents and young adults were in the care of our PPC team. Twenty-nine (14.6%) of these patients had metabolic conditions. Median age at referral was 2.6 years (0-24), median duration of care 352 days (3-2248) and median number of home visits 13 (1-80). Most patients are still alive (16; 55.2%). Median number of drugs administered was 5 (range 0-12), antiepileptics were given most frequently. Symptom burden was high in all children with metabolic disorders at referral and remained high throughout care. Predominant symptoms were gastrointestinal, respiratory and neurologic symptoms. Children with metabolic conditions, who were referred to PPC younger than 1 year of age had a shorter period of care and died earlier compared to those children, who were referred to PPC later in their lives (older than 10 years of age). Eleven (37.9%) of the children initially had no resuscitation restrictions and 7 (53.8%) of those who died, did so on ICU.

CONCLUSIONS: About 15% of children with life-limiting conditions in PPC present with metabolic diseases. Symptom burden is high with neurologic, respiratory and gastrointestinal symptoms being the most frequent and most of those being difficult to treat. In these children, particular attention needs to be addressed to advance care planning.


Pediatric advance care planning seeks to ensure end-of-life care conforming to the patients/their families’ preferences. To expand our knowledge of advance care planning and "medical orders for life-sustaining treatment" (MOLST) in pediatric palliative home care, we determined the number of patients with MOLST, compared MOLST between the four "Together for Short Lives" (TfSL) groups and analyzed, whether there was a relationship between the content of the MOLST and the patients’ places of death. The study was conducted as a single-center retrospective analysis of all patients of a large specialized pediatric palliative home care team (01/2013-09/2016). MOLST were available in 179/198 children (90.4%). Most parents decided fast on MOLST, 99 (55.3%) at initiation of pediatric palliative home care, 150 (83.4%) within the first 100 days. MOLST were only changed in 7.8%. Eighty/179 (44.7%) patients decided on a Do Not Attempt Cardio-Pulmonary Resuscitation (DNACPR) order, 58 (32.4%) on treatment limitations of some kind and 41 (22.9%) wished for the entire spectrum of life-sustaining measures (Full Code). Most TfSL group 1 families wanted DNACPR and most TfSL group 3/4 parents Full Code. The majority (84.9%) of all DNACPR patients died at home/hospice. Conversely, all Full Code patients died in hospital (80% in an intensive care setting). The circumstances of the childrens’ deaths can therefore be predicted considering the content of the MOLST. Regular advance care planning discussions are thus a very important aspect of pediatric palliative home care.


BACKGROUND: An increasing number of young people are living with life-limiting conditions. Current research about advance care planning for young people indicates differing experiences for those involved. Understanding how far young people are engaged in their own advance care plan is important to shape future practice and facilitate young people’s wishes. AIM: To identify and assess the current evidence to determine the barriers and facilitators to the engagement of young people in their own advance care planning process.

DESIGN: A systematic narrative synthesis according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. Study quality was assessed using a quality assessment framework previously used in similar research.

DATA SOURCES: CINAHL Complete, MEDLINE, PubMed and PsycINFO were searched for articles published between 1 January 1990 and 31 October 2017. Grey literature was searched using Google Scholar and Open Grey.

RESULTS: Most studies related to the engagement of young people were conducted in hospitals or other institutions. Research reported not only the aim to include young people in their own advance care planning but also potential barriers to engagement. Barriers include poor communication, conflict within relationships of those in the planning process and patchy education and training for healthcare professionals. Some existing studies are characterised by a lack of rigorous, high-quality research, limiting their impact.

CONCLUSION: Irrespective of setting, engagement of young people would benefit their advance care planning. More detailed, high-quality research is needed to understand the extent of the barriers to young people’s engagement in their own advance care plan and how to facilitate their involvement.


Law’s processes are likely always to be needed when particularly intractable conflicts arise in relation to the care of a critically ill child like Charlie Gard. Recourse to law has its merits, but it also imposes costs, and the courts’ decisions about the best interests of such children appear to suffer from uncertainty, unpredictability and insufficiency. The insufficiency arises from the courts’ apparent reluctance to enter into the ethical dimensions of such cases. Presuming that such reflection is warranted, this article explores alternatives to the courts, and in particular the merits of specialist ethics support services, which appear to be on the rise in the UK. Such specialist services show promise, as they are less formal and adversarial than the courts and they appear capable of offering expert ethical advice. However, further research is needed into such services - and into generalist ethics support services - in order to gauge whether this is indeed a promising development.


BACKGROUND: In 2005, the Israeli parliament passed the "law of dying patients" legalizing life and death decisions (do not resuscitate) in patients with life expectancy less than 6 months.

OBJECTIVE: To determine whether ethnic and religious backgrounds (both religion and religiosity) influence neonatologists' attitudes in simulated clinical situations and opinions about the new law.

DESIGN/METHODS: Prospective design, using standard questionnaire sent to all 155 board-certified practising Israeli Neonatologists. The questionnaire sought demographic and descriptive data, personal opinions regarding four simulated cases, and opinions about five statements regarding variables that may influence decision-making. Statistical analyses were by stepwise backward regression analysis, linear regression, and Kruskal-Wallis tests, wherever indicated.

RESULTS: Sixty-nine percent of the neonatologists replied, representing 27 NICUs out of the 29 NICUs in Israel. Most neonatologists would respect the wish of the family as long as it would be within the limits of the law or their personal beliefs. In stepwise regression analysis, religion, religiosity, age, gender, experience, or country of training did not influence significantly the neonatologists’ opinions or their decisions in simulated practice. Most neonatologists felt that Ethical Committees had no role in NICUs and were seldom consulted. Most felt that likelihood of severe handicap was critical in decision-making. Issues related to treatment cost of a handicapped or dying infant, as well as impact of a handicapped infant on family's well-being, were not deemed critical.

CONCLUSION: Israeli neonatologists appear to be a relatively homogeneous group in end-of-life decisions, regardless of their ethnic, religious, or religiosity background.


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.


As part of the invited supplement on Death and Dying in the PICU, we reviewed ethical, cultural, and social considerations for the bedside healthcare practitioner prior to engaging with children and families in decisions about limiting therapies, withholding, or withdrawing therapies in a PICU. Clarifying beliefs and values is a necessary prerequisite to approaching these conversations. Striving for medical consensus is important. Discussion, reflection, and ethical analysis may determine a range of views that may reasonably be respected if professional disagreements persist. Parental decisional support is recommended and should incorporate their information needs, perceptions of medical uncertainty, child’s condition, and their role as a parent. Child’s involvement in decision making should be considered, but may not be possible. Culturally attuned care requires early examination of cultural perspectives before misunderstandings or disagreements occur. Societal influences may affect expectations and exploration of such may help frame discussions. Hospital readiness for support of social media campaigns is recommended. Consensus with family on goals of care is ideal as it addresses all parties’ moral stance and diminishes the risk for superseding one group’s value judgments over another. Engaging additional supportive services early can aid with understanding or resolving disagreement. There is wide variation globally in ethical permissibility, cultural, and societal influences that impact the clinician, child, and parents. Thoughtful consideration to these issues when approaching decisions about limitation or withdrawal of life-sustaining therapies will help to reduce emotional, spiritual, and ethical burdens, minimize misunderstanding for all involved, and maximize high-quality care delivery.


BACKGROUND: Perinatal and paediatric autopsy rates are at historically low levels with declining uptake due to dislike of the invasiveness of the procedure, and religious objections particularly amongst Muslim and Jewish parents. Less invasive methods of autopsy including imaging with and without tissue sampling have been shown to be feasible alternatives. We sought to investigate attitudes including religious permissibility and potential uptake amongst members of the Muslim and Jewish communities in the United Kingdom.

METHODS: Semi-structured interviews with religious and faith-based authorities (n = 16) and bereaved parents from the Jewish community (n = 3) as well as 10 focus groups with community members (60 Muslim participants and 16 Jewish participants) were conducted. Data were analysed using thematic analysis to identify key themes.

FINDINGS: Muslim and Jewish religious and faith-based authorities agreed that non-invasive autopsy with imaging was religiously permissible because it did not require incisions or interference with the body. A minimally invasive approach was less acceptable as it still required incisions to the body, although in those circumstances where it was required by law it was more acceptable than a full autopsy. During focus group discussions with community members, the majority of participants indicated they would potentially consent to a non-invasive autopsy if the body could be returned for burial within 24 hours, or if a family had experienced multiple fetal/pregnancy losses and the information gained might be useful in future pregnancies. Minimally invasive autopsy was less acceptable but around half of participants might consent if a non-invasive autopsy was not suitable, with the exception of the Jewish Haredi community who unanimously stated they would decline this alternative.

CONCLUSIONS: Our research suggests less invasive autopsy offers a viable alternative to many Muslim and Jewish parents in the UK who currently decline a full autopsy. The findings may be of importance to other countries with significant Muslim and/or Jewish communities as well as to other religious communities where concerns around autopsy exist. Awareness-raising amongst religious leaders and community members will be important if these methods become routinely available.


Fetuses at low gestational age limit of viability, neonates with life threatening or life limiting congenital anomalies and deteriorating acutely ill newborn babies in intensive care, pose taxing ethical questions on whether to forego or stop treatment and allow them to die naturally. Although there is essentially no ethical difference between end of life decision between neonates and other children and adults, in the former, the fact that we are dealing with a new life, may pose greater problems to staff and parents. Good communication skills and involvement of all the team and the parents should start from the beginning to see which treatment can be foregone or stopped in the best interests of the child. This article deals with the importance of clinical ethics to avoid legal and moral showdows and discusses accepted moral practice in this difficult area.

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 prognostic disclosure were not found to be significantly associated with more accurate prognostic understanding.

CONCLUSIONS: The results of the current study demonstrate no evidence that disclosure is associated with anxiety, depression, or decreased hope. Communication processes may increase peace of mind, trust, and hope. It remains unclear how best to enhance prognostic understanding. Cancer 2018;124:1232-41. (c) 2017 American Cancer Society.


When healthcare professionals feel constrained from acting in a patient's best interests, moral distress ensues. The resulting negative sequelae of burnout, poor retention rates, and ultimately poor patient care are well recognized across healthcare providers. Yet an appreciation of how particular disciplines, including physicians, come to be "constrained" in their actions is still lacking. This paper will examine how the application of shared decision-making may contribute to the experience of moral distress for physicians and why such distress may go under-recognized. Appreciation of these dynamics may assist in cross-discipline sensitivity, enabling more constructive dialogue and collaboration.


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 case of Charlie Gard, an infant who was hospitalized in England due to a mitochondrial DNA depletion syndrome that led to an epileptic encephalomyopathy, was highly publicized. Though Charlie’s parents lobbied for him to receive experimental nucleoside replacement therapy as a desperate effort to save him, this request was denied, and after a lengthy legal battle, he died in late July 2017. We discuss the ethical considerations and consequences of this case.

Background: This manuscript reviews unique aspects of end of life decision-making in pediatrics.

Methods: A narrative literature review of pediatric end of life issues was performed in the English language.

Results: While a paternalistic approach is typically applied to children with life-limiting medical prognoses, the cognitive, language, and physical variability in this patient population is wide and worthy of review. In end of life discussions in pediatrics, the consideration of a child’s input is often not reviewed in depth, although a shared decision-making model is ideal for use, even for children with presumed limitations due to age. This narrative review of end of life decision-making in pediatric care explores nomenclature, the introduction of the concept of death, relevant historical studies, limitations to the shared decision-making model, the current state of end of life autonomy in pediatrics, and future directions and needs. Although progress is being made toward a more uniform and standardized approach to care, few non-institutional protocols exist. Complicating factors in the lack of guidelines include the unique facets of pediatric end of life care, including physical age, paternalism, the cognitive and language capacity of patients, subconscious influencers of parents, and normative values of death in pediatrics.

Conclusions: Although there have been strides in end of life decision-making in pediatrics, further investigation and research is needed in this field.


Pediatric palliative care is a field which focuses on caring for and treating the symptoms and distress typically associated with life-limiting illness. Integrative medicine is supported by evidence and aims to heal the whole person, including all aspects of one’s lifestyle. Therapies offered by integrative medicine often empower patients and families, allowing for a sense of control. This review addresses the merging of integrative medicine philosophy and modalities with the care given to children with life-limiting illness. We review an introduction to integrative medicine, trends in its incorporation in the healthcare setting, application to patients receiving palliative care and the management of specific symptoms. A case study is offered to illustrate these principles.


BACKGROUND: In Japan, owing to the progress in medical technology, more children with congenital life-threatening conditions survive than ever before. Nurses who care for these children may also influence decision making in difficult situations.

AIM: We aimed to describe Japanese nurses’ approach toward medical decision making when caring for families of infants with congenital life-threatening conditions. Frequently, these nurses must care for and support parents with ambivalent feelings. DESIGN: Qualitative descriptive study.

METHODS: Participants were recruited from neonatal intensive care unit and paediatric wards at a university hospital in Japan from June to July 2016. Data were collected using semi-structured interviews and qualitatively analysed.

RESULTS: Japanese nurses described these situations as “decision making regarding the child’s medical care” and “daily life at hospitals.” The themes included support of parents and the choices made about their children, nurses giving or holding their opinions about care choices, or withholding their opinions during decision-making events. The narratives included 5 focus areas: the parents; the children, the family as a whole; relationship between families and health care providers; and the effect of the clinical environment on the children and families.

CONCLUSION: When caring for children with life-threatening congenital conditions, nurses should develop supportive relationships with parents and contribute to the clinical decision-making process with empathy and based on the most current research data.

BACKGROUND: Little is known about how decision-making conversations occur during pediatric intensive care unit (PICU) family conferences (FCs).

OBJECTIVE: Describe the decision-making process and implementation of shared decision making (SDM) during PICU FCs. DESIGN: Observational study.

SETTING/SUBJECTS: University-based tertiary care PICU, including 31 parents and 94 PICU healthcare professionals involved in FCs.

MEASUREMENTS: We recorded, transcribed, and analyzed 14 PICU FCs involving decision-making discussions. We used a modified grounded theory and content analysis approach to explore the use of traditionally described stages of decision making (DM) (information exchange, deliberation, and determining a plan). We also identified the presence or absence of predefined SDM elements.

RESULTS: DM involved the following modified stages: information exchange; information-oriented deliberation; plan-oriented deliberation; and determining a plan. Conversations progressed through stages in a nonlinear manner. For the main decision discussed, all conferences included a presentation of the clinical issues, treatment alternatives, and uncertainty. A minority of FCs included assessing the family’s understanding (21%), assessing the family’s need for input from others (28%), exploring the family’s desired decision-making role (35%), and eliciting the family’s opinion (42%).

CONCLUSIONS: In the FCs studied, we found that DM is a nonlinear process. We also found that several SDM elements that could provide information about parents’ perspectives and needs did not always occur, identifying areas for process improvement.


Sorin, G., R. Vialet and B. Tosello (2018). "Formal procedure to facilitate the decision to withhold or withdraw life-sustaining interventions in a neonatal intensive care unit: a seven-year retrospective study." BMC Palliat Care 17(1): 76.

BACKGROUND: Neonatal deaths are often associated with the complex decision to limit or withdraw life-sustaining interventions (LSIs) rather than therapeutic impasses. Despite the existence of a law, significant disparities in clinical procedures remain. This study aimed to assess deaths occurring in a Neonatal Intensive Care Unit (NICU) and measure the impact of a traceable Limitation or Withdrawal of Active Treatment (LWAT) file on the treatment of these newborns.

METHODS: In this monocentric retrospective study, we reviewed all consecutive neonatal deaths occurring during two three-year periods among patients in the NICU at the North Hospital of Marseille: cohort 1 (from 2009 to 2011 without the LWAT file) and cohort 2 (from 2013 to 2015 after introduction of the LWAT file). Newborns included were: gestational age over 22 weeks, birth weight over 500 g, and admission and death in the same NICU. Deaths were categorized according to the classification described by Verhagen et al.: 1) children who died despite cardiopulmonary resuscitation (CPR) (no withholding nor withdrawing of LSIs), (2) children who died while the ventilator, without CPR (no withdrawing of LSIs, but CPR withheld), (3) children who died after LSIs were withdrawn, or (4) LSIs were withheld.

RESULTS: 193 deaths were analyzed: 77 in cohort 1 and 116 in cohort 2. 50% of deaths followed the decision to limit or stop life-sustaining interventions. The mean age at death did not differ between the two cohorts (p = 0.525). An increase in the mortality rate after life-sustaining interventions were withdrawn was observed. The number of multidisciplinary decision meetings was statistically higher in cohort 2 (32.5% versus 55.2% p = 0.002), which were most often prompted due to neurological pathologies, with an increase in parental advice concerning the management of their child (p = 0.026). Even if the introduction of this file did not have an effect on patient age at death, it was significantly associated with a better understanding of end-of-life conditions (p = 0.019), including medication used to sedate and comfort the patient.

CONCLUSIONS: Introduction of the LWAT file seems imperative to develop a personalized healthcare strategy for each child and situation.

Decisions about whether to withdraw or withhold life-sustaining medical treatment from children give rise to complex and value-laden judgments. While recourse to the courts is uncommon, judicial decisions provide an important source of guidance for the children (where they can participate), families and health and medical professionals involved in these decisions. Yet, there has been remarkably little consideration of the Australian jurisprudence on this issue. This article addresses that gap by undertaking the first comprehensive analysis of all publicly available Australian cases that consider whether or not it is in a child’s best interests to receive life-sustaining treatment. A total of 25 cases were located and the judicial consideration of best interests was thematically analysed. Key considerations (to varying degrees) when assessing best interests included the likelihood of treatment curing or improving the child’s health, medical views about diagnosis, prognosis and treatment and the child’s and parents’ views and wishes. The article concludes that the law requires greater certainty and transparency in decision-making. Given the significance of these cases, judgments should describe the factors that the court considers relevant and important, and those that are less influential, as well as the weight ascribed to those various factors and the reasoning that underpins an assessment that treatment is or is not in a child’s best interests.


OBJECTIVES: To evaluate parental decisions following a prenatal diagnosis of trisomy 13 (T13) or trisomy 18 (T18), prenatal counseling received, and pregnancy outcomes. STUDY DESIGN: Single-center, retrospective cohort study of families with a prenatal diagnosis of T13 or T18 from 2000 to 2016. RESULTS: Out of 152 pregnancies, 55% were terminated. Twenty percent chose induction with palliative care, 20% chose expectant management, 2% chose full interventions, and 3% were lost to follow-up. Counseling was based on initial parental goals, but most women were given options besides termination. Women who chose expectant management had a live birth in 50% of the cases. Women who chose neonatal interventions had a live birth in 100% of the cases, but there were no long-term survivors. CONCLUSIONS: The majority of women who continue their pregnancy after a fetal diagnosis of T13 or T18 desire expectant management with palliative care. A live birth can be expected at least half of the time.
Education, Research and Professional Issues

This volume covers aspects of sudden infant and early childhood death, ranging from issues with parental grief, to the most recent theories of brainstem neurotransmitters. It also deals with the changes that have occurred over time with the definitions of SIDS (sudden infant death syndrome), SUDI (sudden unexpected death in infancy) and SUDIC (sudden unexpected death in childhood). The text will be indispensable for SIDS researchers, SIDS organisations, paediatric pathologists, forensic pathologists, paediatricians and families, in addition to residents in training programs that involve paediatrics. It will also be of use to other physicians, lawyers and law enforcement officials who deal with these cases, and should be a useful addition to all medical examiner/forensic, paediatric and pathology departments, hospital and university libraries on a global scale. Given the marked changes that have occurred in the epidemiology and understanding of SIDS and sudden death in the very young over the past decade, a text such as this is very timely and is also urgently needed.


Metaiodobenzylguanidine, a guanithidine analog, labeled with (123)I and (131)I, is used for imaging and therapy of neuroblastosomas and various neural crest tumors like paragangliomas, pheochromocytomas, medullary cancer of thyroid and carcinoids since the past three to four decades. In this review article, we shall revisit metaiodobenzylguanidine as a radiopharmaceutical and its various applications in neural crest tumors.


Palliative care is patient- and family-centered care that enhances quality of life throughout the illness trajectory and can ease the symptoms, discomfort, and stress for children living with life-threatening conditions and their families. This paper aims to increase nurses’ and other healthcare providers’ awareness of selected recent research initiatives aimed at enhancing life and decreasing suffering for these children and their families. Topics were selected based on identified gaps in the pediatric palliative care literature. Published articles and authors’ ongoing research were used to describe selected components of pediatric palliative nursing care including (I) examples of interventions (legacy and animal-assisted interventions); (II) international studies (parent-sibling bereavement, continuing bonds in Ecuador, and circumstances surrounding deaths in Honduras); (III) recruitment methods; (IV) communication among pediatric patients, their parents, and the healthcare team; (V) training in pediatric palliative care; (VI) nursing education; and (VII) nurses’ role in supporting the community. Nurses are in ideal roles to provide pediatric palliative care at the bedside, serve as leaders to advance the science of pediatric palliative care, and support the community.


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 intracacies of adolescence that can improve the social aspects of their lives.

BACKGROUND: It is important not to ignore the impact of parental cancer on children, and this is where oncology and palliative care nurses can play a key role, providing support to parents as a regular aspect of oncological nursing care.

OBJECTIVES: This study explored the experience, needs, and confidence of nurses working in acute cancer services when supporting parents with cancer who have dependent children.

METHODS: Two focus group interviews were conducted with oncology and palliative care nurses in 1 acute hospital trust in the south of England.

RESULTS: Nurses described how they identified with their patients as a parent themselves. This identification with patients added to the emotionally charged context of care and resulted in nurse avoidance of the troubling issue of dependent children. Nurses identified the importance of peer support with regular opportunities to reflect on practice when dealing with issues relevant to parents and children.

CONCLUSIONS: Oncology and palliative care nurses take a reactive approach to family centred care, taking their cue from patients to initiate or request support for their children.

IMPLICATIONS FOR PRACTICE: Guidance was needed on children's developmental stages and how to communicate with children of different ages. In addition, guidance was needed on assessing family needs and access to up to date resources. To enable nurses to engage with the issue of children, strategies of peer support and further educational opportunities need to be implemented.


BACKGROUND: It is known that information regarding the quality of life of a patient is central to pediatric palliative care. This information allows professionals to adapt the care and support provided to children and their families. Previous studies have documented the major areas to be investigated in order to assess the quality of life, although it is not yet known what operational criteria or piece of information should be used in the context of pediatric palliative care. The present study aims to: 1) Identify signs of quality of life and evaluation methods currently used by professionals to assess the quality of life of children with cancer receiving palliative care. 2) Collect recommendations from professionals to improve the evaluation of quality of life in this context.

METHODS: We selected a qualitative research design and applied an inductive thematic content analysis to the verbal material. Participants included 20 members of the Department of Hematology-Oncology at CHU Sainte-Justine from various professions (e.g. physicians, nurses, psychosocial staff) who had cared for at least one child with cancer receiving palliative care in the last year.

RESULTS: Professionals did not have access to pre-established criteria or to a defined procedure to assess the quality of life of children they followed in the context of PPC. They reported basing their assessment on the child's non-verbal cues, relational availability and elements of his/her environment. These cues are typically collected through observation, interpretation and by asking the child, his/her parents, and other members of the care. To improve the assessment of quality of life professionals recommended optimizing interdisciplinary communication, involving the child and the family in the evaluation process, increasing training to palliative care in hematology/oncology, and developing formalized measurement tools.

CONCLUSION: The formulation of explicit criteria to assess the quality of life in this context, along with detailed recommendations provided by professionals, support the development of systematic measurement strategy. Such a strategy would contribute to the development of common care goals and further facilitate communication between professionals and with the family.

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.


AIMS: The study aim was to determine the direct and indirect relations of the five-factor model of personality traits and work stress with professional quality of life in neonatal nurses.

BACKGROUND: Neonatal intensive care nursing has positive and negative effects on neonatal nurses’ psychological well-being. Although individual and situational factors interact to influence professional quality of life, there have been few studies of these relationships in neonatal nurses.

DESIGN: A cross-sectional study conducted in 2016.

METHODS: Self-report questionnaires were used to measure professional quality of life (burnout, secondary traumatic stress and compassion satisfaction), five-factor model of personality traits (neuroticism, agreeableness, extraversion, conscientiousness and openness) and work stress (role ambiguity, role conflict and role overload).

RESULTS: One hundred and forty (34%) of 405 eligible neonatal nurses provided the data. After controlling for work stress, neuroticism and agreeableness were related to burnout, neuroticism was related to secondary traumatic stress, and extraversion was related to compassion satisfaction. Work stress controlled for personality traits was related to burnout and secondary traumatic stress, but not to compassion satisfaction. Neuroticism moderated the effect of work stress on secondary traumatic stress and agreeableness and openness moderated the effect of work stress on compassion satisfaction. Work stress mediated the effect of neuroticism and extraversion on burnout and the effects of extraversion and conscientiousness on compassion satisfaction.

CONCLUSION: Strategies to reduce work stress may not lessen burnout and secondary traumatic stress or increase compassion satisfaction in neonatal nurses who are prone to high neuroticism, low agreeableness and low extraversion.


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.


ABSTRACT

Objective: Compassion fatigue, burnout, and vicarious traumatization are prominent topics in the current literature on the impact of the rewarding but challenging work of healthcare professionals who care for patients with life-limiting illnesses. The positive effects of caregiving constitute a newly emerging outcome that has been relatively unexplored in the pediatric literature, and yet they may play an important role in contributing to the satisfaction and well-being of the healthcare professionals who care for children who have a life-limiting illness.

METHOD: This paper reports the results of a secondary analysis of qualitative interview transcripts that explored the experiences of hospital-based pediatric healthcare providers caring for children with varied life-limiting illnesses. In-depth qualitative interviews were conducted with 25 healthcare professionals (9 social workers, 8 nurses, and 8 physicians). The majority of participants were women (80%), with an age range between 20 and 60 years, and most (84%) had the experience of caring for more than 15 dying children. Thematic analysis was conducted using interpretive description and constant comparison.

RESULTS: Every healthcare professional interviewed experienced personal growth as a result of their providing care for dying children. Three dimensions of personal growth were most consistently reported: (1) new or altered life perspectives, (2) enhanced personal resources, and (3) benevolence.

SIGNIFICANCE OF RESULTS: A deeper understanding of the phenomenon of personal growth could help healthcare organizations to implement innovative approaches that would counterbalance compassion fatigue, and thereby enhance both healthcare provider well-being and child and family outcomes.


BACKGROUND: Mechanically ventilated children are prone to pneumonia due to immobilization and lack of laryngeal (cough) reflex and swallowing. Nurses are directly responsible for many clinical approaches used to prevent ventilator-associated pneumonia.

OBJECTIVE: The research objective is to determine the effectiveness of the nurse education program on the performance of nurses in providing oral care for mechanically ventilated children.

METHODS: This quasi-experimental pretest-posttest design was conducted on 100 nurses (50 in each of the control and intervention groups) in pediatric intensive care units (PICU) in Tehran, 2015. The research tools included a demographic form and three checklists for evaluation of performance according to the clinical practice guidelines for the oral health status of children in PICU. Before intervention, the performance of nurses in both groups was observed at three stages and three different shifts, using an observational checklist. After one month, their performance was observed again with the same checklist at three stages and three different shifts in the PICU. The training was done in four 40-50 minute sessions in a workshop with a 4-week follow-up. The Chi-square test,
Fisher’s exact test, paired t-test, independent t-test, and regression analysis comprised the tools used to analyze the data.

**FINDINGS:** The mean performance scores of nurses before the education program in the intervention and control groups were 42.8 (+/-18.5) and 48.7 (+/-15.7), respectively. These scores improved to 68.6 (+/-31.4) and 48.6 (+/-15.4) four weeks after the intervention (p < 0.001).

**CONCLUSION:** The performance of nurses in providing oral care for mechanically ventilated children improved after the intervention. It is recommended to implement this program for all nurses, regardless of their ward or specialty, based on the clinical practice guidelines. The periodic refreshing in-service training program should be provided to nurses in PICU in order to enhance their performance in providing oral care.


Growing evidence of the association between health professionals’ well-being and patient and organisational outcomes points to the need for effective staff support. This paper reports a brief survey of the UK’s children’s cancer Principal Treatment Centres (PTCs) regarding staff support systems and practices. A short on-line questionnaire, administered in 2012-2013, collected information about the availability of staff support interventions which seek to prevent work-related stress among different members of the multi-disciplinary team (MDT). It was completed by a member of staff with, where required, assistance from colleagues. All PTCs (n = 19) participated. Debriefs following a patient death was the most frequently reported staff support practice. Support groups were infrequently mentioned. There was wide variability between PTCs, and between professional groups, regarding the number and type of interventions available. Doctors appear to be least likely to have access to support. A few Centres routinely addressed work-related stress in wider staff management strategies. Two Centres had developed a bespoke intervention. Very few Centres were reported to actively raise awareness of support available from their hospital’s Occupational Health department. A minority of PTCs had expert input regarding staff support from clinical psychology/liaison psychiatry.


Early-life epilepsies represent a group of many individually rare and often complex developmental brain disorders associated with lifelong devastating consequences and high risk for early mortality. The quantity and quality of evidence needed to guide the evaluation and treatment to optimize outcomes of affected children is minimal; most children are treated within an evidence-free practice zone based solely on anecdote and lore. The remarkable advances in diagnostics and therapeutics are implemented haphazardly with no systematic effort to understand their effects and value. This stands in stark contrast to the evidence-rich practice of the Children’s Oncology Group, where standard of care treatments are identified through rigorous, multicenter research studies, and the vast majority of patients are treated on protocols developed from that research. As a consequence, overall mortality for childhood cancers has declined from approximately 90% in the 1950s to approximately 20% today. The situations of these 2 rare disease specialties are contrasted, and some suggestions for moving early-life epilepsy onto a fast track for success are offered. Chief amongst these is that early-life epilepsy should be treated with the same urgency as pediatric cancer. The best diagnostics and evidence-based treatments should be used in a systematic fashion right from the start, not after the child and family have been subjected to the ravages of the disorder for months or years. This will require unity and cooperation among physicians, researchers, and institutions across state and national borders.


Hypoplastic left heart syndrome is a type of congenital heart disease characterized by underdevelopment of the left ventricle, outflow tract, and aorta. The condition is fatal if aggressive palliative operations are not undertaken, but even with the complete 3-staged surgical palliation, there is significant morbidity because of progressive and ultimately intractable right ventricular failure. For this reason, there is interest in developing novel therapies for the management of right ventricular dysfunction in patients with hypoplastic left heart syndrome. Stem cell therapy may
represent one such innovative approach. The field has identified numerous stem cell populations from different tissues (cardiac or bone marrow or umbilical cord blood), different age groups (adult versus neonate-derived), and different donors (autologous versus allogeneic), with preclinical and clinical experience demonstrating the potential utility of each cell type. Preclinical trials in small and large animal models have elucidated several mechanisms by which stem cells affect the injured myocardium. Our current understanding of stem cell activity is undergoing a shift from a paradigm based on cellular engraftment and differentiation to one recognizing a primarily paracrine effect. Recent studies have comprehensively evaluated the individual components of the stem cells’ secretomes, shedding new light on the intracellular and extracellular pathways at the center of their therapeutic effects. This research has laid the groundwork for clinical application, and there are now several trials of stem cell therapies in pediatric populations that will provide important insights into the value of this therapeutic strategy in the management of hypoplastic left heart syndrome and other forms of congenital heart disease. This article reviews the many stem cell types applied to congenital heart disease, their preclinical investigation and the mechanisms by which they might affect right ventricular dysfunction in patients with hypoplastic left heart syndrome, and finally, the completed and ongoing clinical trials of stem cell therapy in patients with congenital heart disease.


This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of unusual cancers of childhood. It is intended as a resource to inform and assist clinicians who care for cancer patients. It does not provide formal guidelines or recommendations for making health care decisions. This summary is reviewed regularly and updated as necessary by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of the National Cancer Institute (NCI). The summary reflects an independent review of the literature and does not represent a policy statement of NCI or the National Institutes of Health (NIH).


This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of retinoblastoma. It is intended as a resource to inform and assist clinicians who care for cancer patients. It does not provide formal guidelines or recommendations for making health care decisions. This summary is reviewed regularly and updated as necessary by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of the National Cancer Institute (NCI). The summary reflects an independent review of the literature and does not represent a policy statement of NCI or the National Institutes of Health (NIH).


This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of childhood non-Hodgkin lymphoma. It is intended as a resource to inform and assist clinicians who care for cancer patients. It does not provide formal guidelines or recommendations for making health care decisions. This summary is reviewed regularly and updated as necessary by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of the National Cancer Institute (NCI). The summary reflects an independent review of the literature and does not represent a policy statement of NCI or the National Institutes of Health (NIH).


This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of childhood central nervous system embryonal tumors. It is intended as a resource to inform and assist clinicians who care for cancer patients. It does not provide formal guidelines or recommendations for making health care decisions. This summary is reviewed regularly and updated as necessary.
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This PDQ cancer information summary for health professionals provides comprehensive, peer-reviewed, evidence-based information about the treatment of neuroblastoma. It is intended as a resource to inform and assist clinicians who care for cancer patients. It does not provide formal guidelines or recommendations for making health care decisions. This summary is reviewed regularly and updated as necessary by the PDQ Pediatric Treatment Editorial Board, which is editorially independent of the National Cancer Institute (NCI). The summary reflects an independent review of the literature and does not represent a policy statement of NCI or the National Institutes of Health (NIH).


This PDQ cancer information summary has current information about the treatment of childhood Ewing sarcoma. It is meant to inform and help patients, families, and caregivers. It does not give formal guidelines or recommendations for making decisions about health care. Editorial Boards write the PDQ cancer information summaries and keep them up to date. These Boards are made up of experts in cancer treatment and other specialties related to cancer. The summaries are reviewed regularly and changes are made when there is new information. The date on each summary ("Date Last Modified") is the date of the most recent change. The information in this patient summary was taken from the health professional version, which is reviewed regularly and updated as needed, by the PDQ Pediatric Treatment Editorial Board.


This PDQ cancer information summary has current information about the treatment of childhood soft tissue sarcoma. It is meant to inform and help patients, families, and caregivers. It does not give formal guidelines or recommendations for making decisions about health care. Editorial Boards write the PDQ cancer information summaries and keep them up to date. These Boards are made up of experts in cancer treatment and other specialties related to cancer. The summaries are reviewed regularly and changes are made when there is new information. The date on each summary ("Date Last Modified") is the date of the most recent change. The information in this patient summary was taken from the health professional version, which is reviewed regularly and updated as needed, by the PDQ Pediatric Treatment Editorial Board.


This PDQ cancer information summary has current information about the treatment of unusual cancers of childhood. It is meant to inform and help patients, families, and caregivers. It does not give formal guidelines or recommendations for making decisions about health care. Editorial Boards write the PDQ cancer information summaries and keep them up to date. These Boards are made up of experts in cancer treatment and other specialties related to cancer. The summaries are reviewed regularly and changes are made when there is new information. The date on each summary ("Date Last Modified") is the date of the most recent change. The information in this patient summary was taken from the health professional version, which is reviewed regularly and updated as needed, by the PDQ Pediatric Treatment Editorial Board.


ABSTRACT
Objective: Our aims were to report an analysis of the concept of cultural competency and to explore how the cultural competency of the palliative care workforce impacts the holistic care of young people with palliative care needs from South Asian cultures.

METHOD: Using keywords, we searched the online databases MEDLINE, CINAHL, ScienceDirect, and PubMed from January of 1990 through to December of 2016. Some 1543 articles were retrieved, and inclusion and exclusion criteria were applied. A total of 38 papers were included in the concept analysis. The data were analyzed using Coad’s (2002) adapted framework based on Rodgers’s (1989) evolutionary concept analysis, focusing on the attributes, antecedents, consequences, and related terms in relation to culturally competent care. A model case of culturally competent care was also constructed.

RESULTS: The literature provides evidence that the concept of culturally competent care is a complex one, which is often expressed ambiguously. In addition, there is a paucity of research that involves service users as experts in defining their own needs and assessing their experiences related to cultural care.

SIGNIFICANCE OF RESULTS: Cultural care should be integral to holistic patient care, irrespective of a person’s race or ethnicity. There is an urgent need to involve young BAME patients with palliative care needs and their families in the development of a robust tool to assess cultural competency in clinical practice.

EBN engages readers through a range of online social media activities to debate issues important to nurses and nursing. EBN Opinion papers highlight and expand on these debates.


Pancreatoblastoma is a rare paediatric malignant neoplasm. The treatment of choice is complete surgical resection. However, it is often unresectable due to its large size, local infiltration or distant metastasis. Since the condition is rare, there is currently no standard treatment regimen. We outline the case of a 4-year-old child who presented with abdominal pain and distention, together with an enlarged liver and elevated serum alpha-fetoprotein levels. Imaging studies showed the presence of an abnormal pancreatic tumour and multiple nodular lesions in the liver, the biopsies from which led to a diagnosis of pancreatoblastoma. In this case, the patient received cycles of neoadjuvant chemotherapy, combining cisplatin and doxorubicin. The patient subsequently underwent scheduled surgery in which the primary pancreatic lesion was resected, obtaining a circumscribed and nodular specimen measuring 7 x 6 cm and weighing 150 g. Given the extent of the metastasis, the child is currently awaiting a liver transplant.


Developments in managing CF continue to drive dramatic improvements in survival. As newborn screening rolls-out across Europe, CF centres are increasingly caring for cohorts of patients who have minimal lung disease on diagnosis. With the introduction of mutation-specific therapies and the prospect of truly personalised medicine, patients have the potential to enjoy good quality of life in adulthood with ever-increasing life expectancy. The landmark Standards of Care published in 2005 set out what high quality CF care is and how it can be delivered throughout Europe. This underwent a fundamental re-write in 2014, resulting in three documents; center framework, quality management and best practice guidelines. This document is a revision of the latter, updating standards for best practice in key aspects of CF care, in the context of a fast-moving and dynamic field. In continuing to give a broad overview of the standards expected for newborn screening, diagnosis, preventative treatment of lung disease,
nutrition, complications, transplant/end of life care and psychological support, this consensus on best practice is expected to prove useful to clinical teams both in countries where CF care is developing and those with established CF centres. The document is an ECFS product and endorsed by the CF Network in ERN LUNG and CF Europe.


Dravet Syndrome is a devastating childhood epilepsy disorder with high incidence of premature death plus comorbidities of ataxia, circadian rhythm disorder, impaired sleep quality, autistic-like social-interaction deficits and severe cognitive impairment. It is primarily caused by heterozygous loss-of-function mutations in the SCN1A gene that encodes brain voltage-gated sodium channel type-1, termed NaV1.1. Here I review experiments on mouse genetic models that implicate specific loss of sodium currents and action potential firing in GABAergic inhibitory interneurons as the fundamental cause of Dravet Syndrome. The resulting imbalance of excitatory to inhibitory neurotransmission in neural circuits causes both epilepsy and co-morbidities. Promising therapeutic approaches involving atypical sodium channel blockers, novel drug combinations, and cannabidiol give hope for improved outcomes for Dravet Syndrome patients.


This study aims to examine perceived challenges, including knowledge, skills, self, and work environment, of professionals in providing pediatric palliative care (PPC) in Hong Kong and the differences in perceived challenges between groups. A total of 680 pediatric doctors and nurses participated in the survey. They tended to perceive the provision of PPC as difficult and considered "advanced skills" (those dealing with death-related issues) challenging. Findings indicate that nurses, professionals who are less experienced, do not have children, and have not received palliative care training perceived a higher level of challenges in providing PPC. Implications for training and support are discussed.


Leigh syndrome (LS) is an inherited mitochondrial encephalopathy associated with gene mutations of oxidative phosphorylation pathway that result in early disability and death in affected young children. Currently, LS is incurable and unresponsive to many treatments, although some case reports indicate that supplements can improve the condition. Many novel therapies are being continuously tested in pre-clinical studies. In this review, we summarize the genetic basis of LS, current treatment, pre-clinical studies in animal models and the management of other mitochondrial diseases. Future therapeutical strategies and challenges are also discussed.


OBJECTIVES: To evaluate the existing body of evidence to determine the current state of knowledge regarding the perspectives of the following groups: (1) children with cancer, (2) family caregivers, and (3) healthcare professionals, about symptoms, as well as factors that may influence the symptom reports. METHODS: A systematic search was performed for all types of studies that included the perspectives of at least two groups of participants’ symptom reports. Children included anyone younger than 19 years of age who was diagnosed with any type of cancer. Electronic searches were conducted in five English databases and four Chinese databases. The appraisal of methodological quality was conducted using the GRADE criteria. Data were extracted into matrix tables. RESULTS: Thirty-three studies were included. The pediatric oncology symptoms reported by children, family caregivers, and healthcare professionals were synthesized. Findings suggested that family caregivers’ symptom reports were more closely aligned with children’s reports than with the healthcare professionals’ reports. Influencing factors on the different symptom reports included the children’s diagnosis, symptom characteristics, social-demographic factors, and family caregivers’ psychosocial status. CONCLUSIONS: Children with cancer should be the primary reporters for their symptoms. When there are reporters other than the children, the potential discrepancy between the different perspectives needs to be carefully considered.
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.

Effective communication is central to children, young people, and their families’ experiences of health care. Most patient complaints in developed health care systems result from ineffective communication, including inadequate information provision, not feeling listened to, failure to value patients concerns, and patients not feeling involved in care decisions. Advanced communication skills training is now embedded within cancer care policy in the United Kingdom and now features prominently within cancer education in many countries. Here, we share findings from a research evaluation of an advanced communication skills training program dedicated to health professionals caring for children and young people with cancer. We evaluated participants’ (n = 59) perceptions of the program, impact on their skills, knowledge, competence, and confidence. An appreciative inquiry design was adopted; data included interviews, precourse-postcourse evaluations, e-mail blog survey, and 360-degree reflective work records. The framework approach underpinned data analysis and triangulation of data sets. Key findings highlighted good and poor practice in health professionals’ engagement with children, young people, and their families; the purpose of communicating effectively was not always consistent with collaborative working. Attending a program helped participants expand their knowledge of communication theories and strategies. Participants valued using simulated scenarios to develop their skills and were keen to use their new skills to enhance care delivery. Our emphasis within this evaluation, however, remained on what was communicated, when and how, rather than to what effect. The impact of programs such as these must now be evaluated in terms of patient benefit.

Education is integral to the development of children’s palliative care (CPC) globally; thus, the International Children’s Palliative Care Network (ICPCN) developed a training programme including face-to-face and e-learning programmes to increase access to CPC. A review of ICPCN’s e-learning programmes was undertaken in April/May 2018. At the time of writing, there are seven courses available, with more scheduled to be released in the near future. All courses are available in English, with some available in other languages, including Mandarin, Czech and Dutch. Between May 2016 and April 2018, 1501 individuals accessed the courses from 96 countries (39% nurses, 28% doctors). English was the prevalent language used (74%), followed by Spanish (8.5%). To date, over 3106 participants have accessed the e-learning programme from 124 countries. An evaluation in 2015/16 found that >80% of respondents said the
courses were clear, understandable, rated them highly and found them useful. Some 75% of respondents reported improved knowledge, skills and change in attitude, while 61% reported a change in practice. The ICPCN e-learning platform is an innovative way of improving knowledge and understanding of CPC, thereby increasing the accessibility and availability of CPC.


BACKGROUND: Individuals with cystic fibrosis (CF) face the challenges of managing a chronic, progressive disease. While palliative care is a standard of care in serious illnesses, there are no guidelines for its incorporation into CF care. Patients with CF, caregivers, and CF care providers may lack knowledge about palliative care and perceive barriers to integrated care.

OBJECTIVES: To: 1) explore knowledge and perceptions of palliative care among patients with CF, caregivers, and CF care providers; 2) solicit opinions about incorporating palliative care into routine CF care; and 3) solicit recommendations for CF-specific palliative care education for patients and caregivers.

METHODS: We conducted semi-structured interviews with adult patients with CF, parents of adolescents with CF, and CF care providers to assess knowledge and perceptions of palliative care. Discussion included suggestions for palliative care education and integration into CF care. The sample was characterized using summary statistics. Key themes were identified using qualitative content analysis.

RESULTS: Ten patients with CF, ten parents, and eight CF care providers participated. Many had minimal knowledge of palliative care and endorsed the association with end of life as a barrier to palliative care, but after learning more about palliative care, thought it could be helpful, and should be introduced earlier.

CONCLUSIONS: In this single center study, many patients with CF, caregivers, and providers lacked knowledge about palliative care. These findings warrant replication in a larger, multisite study to inform palliative care educational interventions as a step toward consistent integration of palliative care into routine CF care.


BACKGROUND: The death of a child before or shortly after birth is frequently preceded by an end-of-life decision (ELD). Population-based studies of incidence and characteristics of ELDs in neonates and infants are rare, and those in the foetal-infantile period (> 22 weeks of gestation - 1 year) including both neonates and stillborns, are non-existent. However, important information is missed when decisions made before birth are overlooked. Our study protocol addresses this knowledge gap.

METHODS: First, a new and encompassing framework was constructed to conceptualise ELDs in the foetal-infantile period. Next, a population mortality follow-back survey in Flanders (Belgium) was set up with physicians who certified all death certificates of stillbirths from 22 weeks of gestation onwards, and infants under the age of a year. Two largely similar questionnaires (stillbirths and neonates) were developed, pilot tested and validated, both including questions on ELDs and their preceding decision-making processes. Each death requires a postal questionnaire to be sent to the certifying physician. Anonymity of the child, parents and physician is ensured by a rigorous mailing procedure involving a lawyer as intermediary between death certificate authorities, physicians and researchers. Approval by medical societies, ethics and privacy commissions has been obtained.

DISCUSSION: This research protocol is the first to study ELDs over the entire foetal-infantile period on a population level. Based on representative samples of deaths and stillbirths and applying a trustworthy anonymity procedure, the research protocol can be used in other countries, irrespective of legal frameworks around perinatal end-of-life decision-making.


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.


Purpose of Review: Candida infections of the central nervous system (CNS) are a life-threatening complication of invasive infections that most often affect vulnerable groups of patients, including neonates and children with primary immunodeficiency disorders (PID). Here, we review the currently known risk factors for CNS candidiasis, focusing predominantly on the PID caused by biallelic mutations in CARD9. Recent Findings: How the CNS is protected itself against fungal invasion is poorly understood. CARD9 promotes neutrophil recruitment and function, and is the only molecule shown to be critical for protection against CNS candidiasis in humans thus far. Summary: Fundamental insights into the pathogenesis of CNS candidiasis gained from studying rare CARD9-deficient patients has significant implications for other patients at risk for this disease, such as CARD9-sufficient neonates. These findings will be important for the development of adjunctive immune-based therapies, which are urgently needed to tackle the global burden of invasive fungal diseases.


OBJECTIVES: To describe the consequences of workplace stressors on healthcare clinicians in PICU, and strategies for personal well-being, and professional effectiveness in providing high-quality end-of-life care. DATA SOURCES: Literature review, clinical experience, and expert opinion.

STUDY SELECTION: A sampling of foundational and current evidence was accessed. DATA SYNTHESIS: Narrative review and experiential reflection.

CONCLUSIONS: The well-being of healthcare clinicians in the PICU influences the day-to-day quality and effectiveness of patient care, team functioning, and the retention of skilled individuals in the PICU workforce. End-of-life care, including decision making, can be complicated. Both are major stressors for PICU staff that can lead to adverse personal and professional consequences. Overresponsiveness to routine stressors may be seen in those with moral distress, and underresponsiveness may be seen in those with compassion fatigue or burnout. Ideally, all healthcare professionals in PICU can rise to the day-to-day workplace challenges—responding in an adaptive, effective manner. Strategies to proactively increase resilience and well-being include self-awareness, self-care, situational awareness, and education to increase confidence and skills for providing end-of-life care. Reactive strategies include case conferences, prebriefings in ongoing preidentified situations, debriefings, and other postevent meetings. Nurturing a culture of practice that acknowledges the emotional impacts of pediatric critical care work and celebrates the shared experiences of families and clinicians to build resilient, effective, and professionally fulfilled healthcare professionals thus enabling the provision of high-quality end-of-life care for children and their families.


Spinal muscular atrophy [SMA] is the most common genetic cause of childhood mortality, primarily from the most severe form SMA type 1. It is a severe, progressive motor neurone disease, affecting the lower brainstem nuclei and the spinal cord. There is a graded level of severity with SMA children from a practical viewpoint described as "Non-sitters", "Sitters" and less commonly, "Ambulant" correlating with SMA Type 0/Type 1, Type 2 and Type 3 respectively. Children with SMA Type 0 have a severe neonatal form whilst those with SMA Type 1 develop hypoventilation, pulmonary aspiration, recurrent lower respiratory tract infections, dysphagia and failure to thrive before usually succumbing to respiratory failure and death before the age of 2years. The recent introduction of the antisense oligonucleotide nusinersen into clinical practice in certain countries, following limited trials of less than two years duration, has altered the treatment landscape and improved the outlook considerably for SMN1 related SMA. Approximately 70% of infants appear to have a clinically significant response to nusinersen with improved motor
function. It appears the earlier the treatment is initiated the better the response. There are other rarer genetic forms of SMA that are not treated with nusinersen. Clinical expectations will change although it is unclear as yet what the extent of response will mean in terms of screening initiatives [e.g., newborn screening], "preventative strategies" to maintain respiratory wellbeing, timing of introduction of respiratory supports, and prolonged life expectancy for the subcategory of children with treated SMA type 1. This article provides a review of the strategies available for supporting children with respiratory complications of SMA, with a particular emphasis on SMA Type 1.


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<.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 overweigh 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.


Among diseases affecting skeletal muscle, muscular dystrophy is one of the most devastating and complex disorders. The term ‘muscular dystrophy’ refers to a heterogeneous group of genetic diseases associated with a primary muscle defect that leads to progressive muscle wasting and consequent loss of muscle function. Muscular dystrophies are accompanied by numerous clinical complications and abnormalities in other tissues that cause extreme discomfort in everyday life. The fact that muscular dystrophy often takes its toll on babies and small children, and that many patients die at a young age, adds to the cruel character of the disease. Clinicians all over the world are facing the same problem: they have no therapy to offer except for symptom-relieving interventions. Patients, their families, but also clinicians, are in urgent need of an effective cure. Despite advances in genetics, increased understanding of molecular mechanisms underlying muscle disease, despite a sweeping range of successful preclinical strategies and relative progress of their implementation in the clinic, therapy for patients is currently out of reach. Only a greater comprehension of disease mechanisms, new preclinical studies, development of novel technologies, and tight collaboration between scientists and physicians can help improve clinical treatment. Fortunately, inventiveness in research is rapidly extending the limits and setting new standards for treatment design. This review provides a synopsis of muscular dystrophy and considers the steps of preclinical and clinical research that are taking the muscular dystrophy community towards the fundamental goal of combating the traumatic disease.


Niemann-Pick Type C (NPC) is a progressive and life-limiting autosomal recessive disorder caused by mutations in either the NPC1 or NPC2 gene. Mutations in these genes are associated with abnormal endosomal-lysosomal trafficking, resulting in the accumulation of multiple tissue-specific lipids in the lysosomes. The clinical spectrum of NPC disease ranges from a neonatal rapidly progressive fatal disorder to an adult-onset chronic neurodegenerative disease. The age of onset of the first (beyond 3 months of life) neurological symptom may predict the severity of the disease and determines life expectancy. NPC has an estimated incidence of ~1:100,000 and the rarity of the disease translates into misdiagnosis, delayed diagnosis, and barriers to good care. For these reasons, we have developed clinical guidelines that define standard of care for NPC patients, foster shared care arrangements between expert centres and family physicians, and empower patients. The information contained in these guidelines was obtained through a systematic review of the literature and the experiences of the authors in their care of patients with NPC. We adopted the Appraisal of Guidelines for Research & Evaluation (AGREE II) system as method of choice for the guideline development process. We made a series of conclusive statements and scored them according to level of evidence, strengths of recommendations, and expert opinions. These guidelines can inform care providers, care funders, patients and their carers of best practice of care for patients with NPC. In addition, these guidelines have identified gaps in the knowledge that must be filled by future research. It is anticipated that the implementation of these guidelines will lead to a step change in the quality of care for patients with NPC irrespective of their geographical location.


Genetic testing and counseling have become integral to the timely control of heritable cancers, like the childhood eye cancer retinoblastoma. This study aimed to determine attitudes, knowledge, and experiences related to retinoblastoma genetics among survivors and parents of children with retinoblastoma in Kenya. This qualitative study used focus groups as the primary data collection method. Study settings were Kenyatta National Hospital and Presbyterian Church of East Africa Kikuyu Hospital. Thematic analysis was used to identify key themes. Thirty-one individuals participated in five focus groups. Two main concepts emerged: (1) the origins of retinoblastoma are unclear, and (2) retinoblastoma is associated with significant challenges. The lack of clarity surrounding the origins of retinoblastoma was linked to limited knowledge of retinoblastoma genetics, and limited genetic counseling delivery and uptake. The challenges associated with retinoblastoma were discussed in terms of the impact of the diagnosis on individuals and families, and unmet healthcare needs related to the diagnosis. Next steps will incorporate these findings to develop evidence-informed and accessible cancer genetic services in Kenya.


INTRODUCTION: Pain in children with intellectual disabilities (ID) is common and complex, yet there is no standard pain training for their secondary caregivers (ie, respite staff).

OBJECTIVES: Determine perceived pain training needs/preferences of children’s respite staff (phase 1) and, use this information combined with extant research and guidelines to develop and pilot a training (phase 2).

METHODS: In phase 1, 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 2, 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 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 2 results suggested that completion of the 3 to 3.5-hour pain training significantly increased respite workers’ pain-related knowledge (effect sizes: r=0.81 to 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 (effect sizes: r=0.41 to 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.


OBJECTIVES: To describe practical considerations related to discussions about death or possible death of a critically ill child.

DATA SOURCES: To describe practical considerations related to discussions about death or possible death of a critically ill child.

DATA EXTRACTION: Not available.

DATA SYNTHESIS: Narrative and experiential review were used to describe the following areas benefits and potential adverse consequences of conversations about risk of death and the timing of, preparation for, and conduct of conversations about risk of death. CONCLUSIONS: Timely conversations about death as a possible outcome of PICU care are an important part of high-quality ICU care. Not all patients "require" these conversations; however, identifying patients for whom conversations are indicated should be an active process. Informed conversations require preparation to provide the best available objective information. Information should include distillation of local experience, incorporate the patients’ clinical trajectory, the potential impact(s) of alternate treatments, describe possible modes of death, and acknowledge the extent of uncertainty. We suggest the more factual understanding of risk of death should be initially separated from the more inherent value-laden treatment recommendations and decisions. Gathering and sharing of collective knowledge, conduct of additional investigations, and time can increase the factual content of risk of death discussions. Timely and sensitive delivery of this best available knowledge then provides foundation for high-quality treatment recommendations and decision-making.


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 inexistent. 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.


Caring for a child in hospital who is approaching death, in the terminal phase, requires a focus on caring for the physical, emotional, and spiritual needs of the child and family. Health professionals caring for these children and families may need to shift their focus from a treatment-focused approach aimed at cure or maintaining life to a comfort-focused approach. The Comfort Care Case (CCC) is a collection of resources designed for use in hospital to ease suffering and facilitate comfort within a pediatric end-of-life (EOL) context. The resources are intended to support the child, the family, and the health professionals involved in EOL care. This article describes the development, implementation, and education associated with the CCC in a tertiary pediatric hospital.


AIM: To synthesize qualitative research examining the experience of critical care nurses caring for a dying child. BACKGROUND: Caring for a dying child remains one of the most difficult aspects of nursing, potentially leading to personal and professional distress. A thorough understanding of this experience for critical care nurses allows for improved delivery of care and support for the nurse. DESIGN: A qualitative evidence synthesis was undertaken, informed by Thomas and Harden’s thematic synthesis methodology. DATA SOURCES: Studies were retrieved from CINAHL Plus, Scopus, OVID Medline, and Embase, alongside hand-searching reference lists in February 2016. REVIEW METHODS: Two reviewers independently assessed each study using a multistep screening process and performed critical appraisal of each included study. Data were extracted onto a predeveloped tool and analysed using thematic analysis. RESULTS: There is a blurred line between the role of the nurse as a person or a professional while caring for the child and family throughout hospitalization and during and after the death. Each stage of care involves tasks and emotions that highlight the changing dominance of the nurse as either a person or professional. CONCLUSION: Personal, interpersonal, and contextual factors affect delivery of care and impact of the death of the child on the critical care nurse. Reviewing individual and institutional practices could improve provision of care, interprofessional collaboration, and support provided to staff involved.


Spinal muscular atrophy (SMA) is a degenerative motor neurone disorder causing progressive muscular weakness. Without assisted ventilation or novel therapies, most children with SMA type 1 die before the second year of life due to respiratory failure as the respiratory muscles and bulbar function are severely affected. Active respiratory treatment (mechanically assisted cough, invasive or non-invasive ventilation) has improved survival significantly in recent decades, but often at the cost of becoming ventilator dependent. The advent of a new oligonucleotide based therapy (Nusinersen) has created new optimism for improving motor function. However, the long-term effect on respiratory function is unclear and non-invasive respiratory support will remain an important part of medical management in patients with SMA. This review summarises the existing knowledge about sleep-disordered breathing and respiratory failure in patients with SMA, especially type 1, as well as the evidence of improved outcome and survival in patients treated with non-invasive or invasive ventilation. Practical considerations and ethical concerns are delineated with discussion on how these may be affected by the advent of new therapies such as Nusinersen.

https://www.ncbi.nlm.nih.gov/pubmed/30396824


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 3/46/3000 (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.


OBJECTIVE: To study the value of Pediatric Early Warning Score (PEWS) in identifying the condition of critically ill children.

METHODS: A total of 120 children who were transferred to the pediatric intensive care unit (PICU) from the general ward during hospitalization or admitted to the PICU after emergency treatment in the Xiangya Hospital of Central South University from January to December, 2016 were enrolled as the PICU group. The other 120 children who were admitted to the general ward in the hospital were used as the control group. According to the disease type, the PICU group was further divided into two subgroups: respiratory/circulatory system diseases (n=55) and nervous/other system diseases (n=65). The PEWS score on admission was recorded, and the receiver operating characteristic (ROC) curve was used to analyze the value of PEWS in evaluating patients' condition.

RESULTS: The PICU group had a significantly higher PEWS score than the control group (P<0.05). The respiratory/circulatory system disease subgroup had a significantly higher PEWS score than the nervous/other system disease subgroup (P<0.05). In predicting whether the child was admitted to the PICU, PEWS had a sensitivity of 85%, a specificity of 95%, and an area under the ROC curve (AUC) of 0.951 (95% confidence interval: 0.923-0.980) at the optimal cut-off value of 3.5 (PEWS score). The AUC of PEWS was 0.768 in the nervous/other system disease subgroup and 0.968 in the respiratory/circulatory system disease subgroup. The mortality rate of children with a PEWS score of >6, 4-6 and </=3 was 40%, 21% and 0 respectively (P<0.001).

CONCLUSIONS: PEWS can well identify disease severity in critically ill children, and it has different sensitivities in children with different varieties of diseases. PEWS has a good value in predicting children's prognosis.


BACKGROUND: End-of-life (EOL) care intensity is known to vary by secular and geographic patterns. US physicians receive less aggressive EOL care than the general population, presumably the result of preferences shaped by workplace experience with EOL care.

OBJECTIVE: We investigated occupation as a source of variation in EOL care intensity.

METHODS: Across 4 states, we identified 660 599, nonhealth maintenance organization Medicare beneficiaries aged >/=66 years who died between 2004 and 2011. Linking death certificates, we identified beneficiaries with prespecified occupations: nurses, farmers, clergy, mortuary workers, homemakers, first-responders, veterinary workers, teachers, accountants, and the general population. End-of-life care intensity over the last 6 months of life was assessed using 5 validated measures: (1) Medicare expenditures, rates of (2) hospice, (3) surgery, (4) intensive care, and (5) in-hospital death.

RESULTS: Occupation was a source of large variation in EOL care intensity across all measures, before and after adjustment for sex, education, age-adjusted Charlson Comorbidity Index, race/ethnicity, and hospital referral region. For example, absolute and relative adjusted differences in expenditures were US$9991 and 42% of population mean expenditure ( P < .001 for both). Compared to the general population on the 5 EOL care intensity measures, teachers (5 of 5), homemakers (4 of 5), farmers (4 of 5), and clergy (3 of 5) demonstrated significantly less aggressive care. Mortuary workers had lower EOL care intensity (4 of 5) but small numbers limited statistical significance.

CONCLUSION: Occupations with likely exposure to child development, death/bereavement, and naturalistic influences demonstrated lower EOL care intensity. These findings may inform patients and clinicians navigating choices around individual EOL care preferences.
BACKGROUND: Good end-of-life care planning is vital to ensure optimal care is provided for patients and their families. Two key factors are open and honest advance care planning conversations between the patient (where possible), family, and health care professionals, focusing on exploring what their future wishes are; and the development of an advance care plan document. However, in paediatric and neonatal settings, there has been little research to demonstrate how advance care planning conversations take place. This study explored health care professionals' views and experiences of paediatric advance care planning in hospitals, community settings and hospices.

METHODS: A qualitative methodology was employed using purposive sampling of health care professionals involved in the end-of-life care for children aged 0-18 years known to the hospital palliative care team, and had died at least three months before, but less than 18 months prior to the study. Ethics committee approval was obtained for the study. Located in the North of England, the study involved three hospitals, a children's hospice, and community services. Data were collected using semi-structured, digitally recorded, telephone interviews. All interviews were transcribed verbatim and subjected to thematic analysis.

RESULTS: Twenty-one health care professionals participated, including generalist paediatric staff as well as specialist palliative care staff. Two themes were generated from the study: The timing of planning conversations, including waiting for the relationship with the family to form; the introduction of parallel planning; avoiding a crisis situation. Secondly, supporting effective conversations around advance care planning, including where to have the conversation; introducing the conversation; and how to approach the topic encompassing the value of advance care planning and documentation for families.

CONCLUSION: The timing of when to start the advance care planning conversations remains an issue for health care professionals. The value of doing it in stages and considering the environment where the conversations are held was noted. Timely planning was seen as vital to avoid difficult conversations at a crisis point and for co-ordination of care. Good advance care planning is to provide the best person-centred care for the child and experience for the family.

PURPOSE: This study aimed (1) to examine the current status of psychological distress experienced by neonatal intensive care unit (NICU) nurses in supporting bereaved families, (2) to identify the factors associated with psychological distress, and (3) to understand the professional characteristics of nurses experiencing high psychological distress by comparing the study results with those of pediatricians.

METHODS: We sent questionnaires to 64 NICUs. The psychological distress of nurses was classified into two groups based on the frequency of psychological distress experienced and analyzed using the chi2 test and Fisher's exact test. A multiple logistic regression analysis was used to investigate the factors related to psychological distress.

RESULTS: Of the 384 nurse respondents, 190 (49.5%) reported having supported bereaved families, 169 of who were included in the analysis. A total of 123 nurses (72.8%) reported high levels of psychological distress. Our study revealed that the use of coping methods is associated with high psychological distress. The comparison with pediatricians revealed that nurses were significantly more likely to be female and had fewer years of working experience. Nurses were also significantly more likely to use coping methods and to experience high psychological distress.

CONCLUSION: Clarifying the coping methods for psychological distress in supporting bereaved families may be necessary, and nurses need to identify appropriate coping methods. In nursing education, information on psychological distress related to children’s deaths and bereavement care should be conveyed from the early stage and nurses must obtain preliminary knowledge. The creation of a bereavement follow-up system is recommended.

OBJECTIVES: To examine the circumstance of death in the PICU in the setting of ongoing curative or life-prolonging goals. DATA SOURCES: Multidisciplinary author group, international expert opinion, and use of current literature.

DATA SYNTHESIS: We describe three common clinical scenarios when curative or life-prolonging goals of care are pursued despite a high likelihood of death. We explore the challenges to providing high-quality end-of-life care in this setting. We describe possible perspectives of families and ICU clinicians facing these circumstances to aid in our understanding of these complex deaths. Finally, we offer suggestions of how PICU clinicians might improve the care of children at the end of life in this setting.

CONCLUSIONS: Merging curative interventions and optimal end-of-life care is possible, important, and can be enabled when clinicians use creativity, explore possibilities, remain open minded, and maintain flexibility in the provision of critical care medicine. When faced with real and perceived barriers in providing optimal end-of-life care, particularly when curative goals of care are prioritized despite a very poor prognosis, tensions and conflict may arise. Through an intentional exploration of self and others’ perspectives, values, and goals, and working toward finding commonality in order to align with each other, conflict in end-of-life care may lessen, allowing the central focus to remain on providing optimal support for the dying child and their family.


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’.


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: Palliative care physicians often assist with pain management in children with cancer, but little is known about how they use long-acting opioids for chronic pain with these patients.

OBJECTIVE: To determine the practices, attitudes, and beliefs of palliative care physicians toward the use of long-acting opioids in children with advanced cancer. DESIGN: An electronic survey was sent to all members of The American Academy of Pediatrics (AAP) Section of Hospice and Palliative Medicine (SOHPM) and those identified as physicians who provide palliative care to children on the AAP SOHPM LISTSERV(R).

RESULTS: The response rate to the survey was 62% (116/188). A majority (66% [77/116]) of physicians are board certified in both pediatrics and hospice and palliative medicine. This represents 28% of all board-certified pediatric palliative care physicians. Most palliative care physicians report comfort in using long-acting opioids in children (84-94%), with the exception of long-acting hydromorphone (37%). Physicians perceived methadone as least costly (3%) but associated it with a higher perceived family resistance (51%). As compared with pediatric palliative care fellowship-trained physicians, nonpediatric fellowship-trained physicians perceived titration of oxycodone ER and morphine ER to be easier (p = 0.06, p = 0.07) and less likely to agree that the main reason for starting methadone is that the existing formulations of other long-acting opioids are unsuitable for children (p = 0.05).

CONCLUSIONS: Most physicians who provide palliative care to children are comfortable using opioids but there is significant variation in the level of comfort with different opioids. This information will be helpful in developing targeted education for palliative care providers.


According to Professor Basil T. Darras, Professor of Neurology (Pediatrics) at Harvard Medical School and Director of the Spinal Muscular Atrophy (SMA) Program at Boston Children's Hospital in Boston (MA, USA), the diagnosis of SMA type I is clinical and is based on detailed general physical and neurological examinations. SMA type I remains the most common genetic disease resulting in death in infancy and is really devastating for the child, the parents, as well as the medical professionals with the privilege of caring for patients with SMA and their parents. The proposed management options include: i) no respiratory support; ii) non-invasive ventilation; and iii) tracheotomy with mechanical ventilation. Deciding, which option is the best, is indeed a very personal decision. The optimal clinical care should be extremely mindful of parents' wishes and management goals with regard to the quality of life. Since the end of 2016 in the USA, and recently in Europe, there exists the possibility of accessing a novel treatment drug for SMA, namely Nusinersen. This antisense oligonucleotide is administered intrathecally and increases the production of the fully functional SMN protein, thus improving motor function, the quality of life and survival. Among the ongoing clinical trials, oral treatment with RG7916, a small molecule SMN2 splicing modifier, appears to be really promising. Gene therapy using viral vectors is expected to offer an ‘one and done’ therapy and possibly a cure, if administered early in life, before any symptoms appear. It is really interesting that viruses, which at the moment are the cause of death of children with SMA, if genetically modified, may be used for their treatment.


PURPOSE: To qualitatively explore neonatal intensive care nurses’ experiences with end-of-life photography as part of their bereavement support work with families.

DESIGN AND METHODS: An Interpretive Phenomenological Analysis with data collected through a focus group (n=6) and one semi-structured interview (n=1) with neonatal nurses from a Level 3/4 NICU in a Canadian pediatric hospital.

RESULTS: Participants’ comfort with EOL photography developed over time through exposure to bereavement scenarios and positive experiences with families. Participants experienced a feeling of pressure to balance the photography with clinical responsibilities and find the right time to introduce photography while being sensitive to family experiences. Participants experienced EOL photography as something tangible to give families and were satisfied knowing the images might play an important role in the family’s healing after the NICU.
CONCLUSIONS: All participants had come to value EOL photography as a positive and meaningful part of their work with bereaved families. Identified challenges related to balancing the practice with the unpredictable flow and demands of critical care and to developing an appreciation for and comfort with the photography as part of their healing and the families’ healing.

PRACTICAL IMPLICATIONS: Findings contribute insight into care-provider experience that can inform best practices, training, and staff support for palliative and bereavement work in neonatal and pediatric settings. The findings suggest a need to support nurses emotionally and clinically in carrying out this photography as part of their care for families.


BACKGROUND: Education sessions about palliative care among teenagers are uncommon in developed countries. However, very little is known either about the impact of this type of intervention or about how this age-group perceives its impact. The purpose of this study was therefore to (i) implement an education program about palliative care among teenagers and (ii) to investigate the impact of the program on the participants.

METHODS: An action-research study was conducted at a local community parish in Portugal in November 2015. An education programme was purposively built about palliative care, using active educational strategies adapted for teenagers. Quantitative and qualitative techniques and instruments were used for data collection: questionnaire; reflective diaries; interviews and written testimony. The program had three stages: preparation; intervention; and evaluation. Qualitative data were analysed using thematic content analysis; quantitative data were analysed descriptively.

RESULTS: 69 people (47 teenagers) participated in the education program. Findings show that the education program contributed to creating awareness about palliative care. Both the teenagers and other participants assessed the education program positively. At the end of the program, teenagers had a constructive message about palliative care.

CONCLUSIONS: The education-intervention contributed to create awareness about palliative care among the participant teenagers, who ended the program with a positive message about palliative care. Based on our findings, the following policy implications can be drawn: (1) Further research is needed to evaluate the effect of education programs about palliative care among younger age groups (teenagers and children), particularly in relation to the changing of attitudes toward palliative care. (2) Education about palliative care should be promoted to local communities, involving all age groups, to foster involvement, participation and empowerment. (3) Compassionate communities should be promoted to enhance the health and wellbeing of all citizens at the end of their life.


On March 29, 2018, blinatumomab (Blincyto, Amgen) received an accelerated expanded approval for the treatment of adult and pediatric patients with B-cell precursor acute lymphoblastic leukemia (ALL) who are in first or second complete remission (CR) and have minimal residual disease (MRD). Blinatumomab was first approved for use in adult patients (in December 2014) and later in pediatric patients (in September 2016) with relapsed or refractory Philadelphia chromosome (Ph)-negative B-cell precursor ALL; the approval was expanded in July 2017 to include patients with Ph-positive disease. The agent is a bispecific CD19-directed CD3 T-cell engager.


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.


Pediatric consultation-liaison clinicians are well positioned to provide support, guidance, and systemic recommendations about how to help medical clinicians cope with the stresses of working with dying children. Interventions to support sustainability in this work need to occur at the institutional and team-based levels as well as in individual practice. Shared clinical work around challenging cases provides opportunities to engage with medical clinicians about their difficult experiences and provide reflection and support. Psychiatry services may also be in a role of advocating for institutionally based interventions that can help their medical colleagues.


Effective communication with parents is a very important skill for pediatricians especially in a neonatal setup. The authors analyzed non-verbal communication of medical caregivers during counseling sessions. Recorded videos of counseling sessions from the months of March-April 2016 were audited. Counseling episodes were scored using Non-verbal Immediacy Scale Observer Report (NIS-O). A total of 150 videos of counseling sessions were audited. The mean (SD) total score on (NIS-O) was 78.96(7.07). Female counseled sessions had significantly higher proportion of low scores (p < 0.001). No video revealed high score. Overall 67(44.67%) sessions revealed low total score. This reflects an urgent need to develop strategies to improve communication skills in a neonatal unit. This study lays down a template on which other Neonatal intensive care units (NICUs) can carry out gap defining audits.


Despite the continuous improvement of pediatric palliative care, medical professionals still face various barriers regarding its implementation; our aim was to investigate this question in Hungarian pediatric oncology practice. Structured interviews were carried out in person with physicians from the Hungarian Pediatric Oncology Group (n = 22). Codes were generated inductively with the aid of Atlas.ti 6.0 software. Most physicians placed the palliative care discussion at the end of curative treatment (n = 21) and preferred to conduct it in a team setting (n = 18), mainly in the presence of a psychologist. Preparing parents for the child's death can occur during the palliative care discussion (n = 3), in the child's final days/h (n = 6), gradually (n = 10), or never (n = 3). There are words consciously utilized and avoided during this discussion, with the word "death" proving to be the most ambivalent (utilized n = 5, avoided n = 6).

CONCLUSIONS: There is no widely accepted unified practice among pediatric oncologists concerning the implementation of palliative care in Hungary. Despite the international recommendation, the common practice of timing is still at the end of curative treatment. Physicians rely on multidisciplinary teamwork, where the psychologist's role is the most prominent in this discussion. What is Known: * There is an international consensus that palliative care should commence at the diagnosis of a pediatric malignant disease regardless of illness outcome. * Barriers to the early implementation of palliative care in pediatric oncology involve resource-based and attitudinal factors. What is New: * In Hungary, where pediatric oncologists are sole decision-makers, early implementation of palliative care is rare. * There is a strong preference among physicians for working within a team, while also asserting that presence of team members may decrease the level of intimacy.
OBJECTIVE: To evaluate research priority setting approaches in childhood chronic diseases and to describe the priorities of stakeholders including patients, caregivers/families and health professionals.

DESIGN: We conducted a systematic review of MEDLINE, Embase, PsycINFO and CINAHL from inception to 16 October 2016. Studies that elicited stakeholder priorities for paediatric chronic disease research were eligible for inclusion. Data on the prioritisation process were extracted using an appraisal checklist. Generated priorities were collated into common topic areas.

RESULTS: We identified 83 studies (n=15 722). Twenty (24%) studies involved parents/caregivers and four (5%) children. The top three health areas were cancer (11%), neurology (8%) and endocrine/metabolism (8%). Priority topic areas were treatment (78%), disease trajectory (48%), quality of life/psychosocial impact (48%), disease onset/prevention (43%), knowledge/self-management (33%), prevalence (30%), diagnostic methods (28%), access to healthcare (25%) and transition to adulthood (12%). The methods included workshops, Delphi techniques, surveys and focus groups/interviews. Specific methods for collecting and prioritising research topics were described in only 60% of studies. Most reviewed studies were conducted in high-income nations.

CONCLUSIONS: Research priority setting activities in paediatric chronic disease cover many discipline areas and have elicited a broad range of topics. However, child/caregiver involvement is uncommon, and the methods often lack clarity. A systematic and explicit process that involves patients and families in partnership may help to inform a more patient and family-relevant research agenda in paediatric chronic disease.


Studies indicate research ethics committee (REC) approval and clinician gatekeeping are two key barriers in recruiting children and young people (CYP) with life-limiting conditions (LLCs) and life-threatening illnesses (LTIs) and their families to research. OBJECTIVES: To explore the reported experiences, difficulties and proposed solutions of chief investigators (CIs) recruiting CYP with LLCs/LTIs and families in the UK. METHODS: 61 CIs conducting studies with CYP with LLCs/LTIs and their families, identified from the UK National Institute of Health Research portfolio, completed an anonymous, web-based questionnaire, including both closed and open-ended questions. Descriptive statistics and inductive and deductive coding were used to analyse responses. RESULTS: UK CIs cited limitations on funding, governance procedures including Research and Development, Site-Specific and REC approval processes, and clinician gatekeeping as challenges to research. CIs offered some solutions to overcome identified barriers such as working with CYP and their families to ensure their needs are adequately considered in study design and communicated to ethics committees; and designing studies with broad inclusion criteria and developing effective relationships with clinicians in order to overcome clinician gatekeeping.

CONCLUSIONS: Many of the challenges and solutions reported by UK CIs have applicability beyond the UK setting. The involvement of clinicians, patients and their families at the inception of and throughout paediatric palliative care research studies is essential. Other important strategies include having clinician research champions and increasing the visibility of research. Further research on the perspectives of all stakeholders, leading to mutually agreed guidance, is required if care and treatment are to improve.

EBN engages readers through a range of Online social media activities to debate issues important to nurses and nursing. EBN Opinion papers highlight and expand on these debates.


BACKGROUND: Research found that low levels of professional confidence and personal comfort among neonatal clinicians regarding palliative care may indicate a lack of competence and hesitancy to offer neonatal palliative care services.

PURPOSE: This study evaluated the factors associated with the confidence and comfort levels of neonatal clinicians providing neonatal palliative care.

METHODS: A cross-sectional survey and questionnaire were used to investigate the confidence and comfort levels of neonatal clinicians regarding neonatal palliative care.

RESULTS: Research subjects included 154 neonatal clinicians. Clinicians’ confidence in providing neonatal palliative care was significantly impacted by age, marital status, years of professional experience (p < 0.05), and prior palliative care training. Comfort levels were significantly impacted by educational degree, marital status, and years of working experience. Clinicians with a supportive workplace reported increases in both professional confidence (r = 0.286, p < 0.001) and personal comfort (r = 0.521, p < 0.001).

CONCLUSION: Research reveals the importance of neonatal palliative education and suggests further development of interdisciplinary neonatal palliative care teams to improve clinicians’ professional confidence and personal comfort.


Background: Medication errors (MEs) in neonates are frequent and associated with increased potential for harm compared with adults. The effect of learning from reported MEs is potentially lacking due to underreporting, lack of feedback and missing actions to improve medication safety. A new approach involving positive recognition of current and future strategies may facilitate greater exploration of how to improve medication safety in neonates. We aimed to explore current and potential future practices to prevent MEs in neonatal intensive care units (NICUs).

Methods: Focus group interviews of physicians and nurses were conducted at three Danish NICUs. Participants were included if they had at least 1 month of working experience and provided direct patient care. A semistructured interview guide involving three questions was used: (a) how do you feel about discussing prevention of MEs? (b) how do you currently prevent MEs from occurring? and (c) how can we become better at preventing MEs in the future? Content analysis was used to identify themes in the interviews.

Results: Participants commented that MEs still occur and that action must be taken to improve medication safety. Current practices to prevent MEs involved technology, procedures, education, skills and hospital pharmacy services. Potential future practices to prevent MEs included customizing the computerized physician order entry systems to support optimal prescribing, standardizing the double-check process, training of calculation skills and teamwork and increased use of hospital pharmacy services.

Conclusions: Several current and potential future practices to reduce MEs in NICUs were identified, highlighting the complexity of MEs. Our findings support an interdisciplinary multifaceted intervention involving both technical and nontechnical elements to improve medication safety in NICUs.

Invasive ventilation is often necessary for the treatment of newborn infants with respiratory insufficiency. The neonatal patient has unique physiological characteristics such as small airway caliber, few collateral airways, compliant chest wall, poor airway stability, and low functional residual capacity. Pathologies affecting the newborn’s lung are also different from many others observed later in life. Several different ventilation modes and strategies are available to optimize mechanical ventilation and to prevent ventilator-induced lung injury. Important aspects to be considered in ventilating neonates include the use of correct sized endotracheal tube to minimize airway resistance and work of breathing, positioning of the patient, the nursing care, respiratory kinesiotherapy, sedation and analgesia, and infection prevention, namely, the ventilator-associated pneumonia and nosocomial infection, as well as prevention and treatment of complications such as air leaks and pulmonary hemorrhage. Aspects of ventilation in patients under ECMO (extracorporeal membrane oxygenation) and in palliative care are of increasing interest nowadays. Online pulmonary mechanics and function testing as well as capnography are becoming more commonly used. Echocardiography is now a routine in most neonatal units. Near infrared spectroscopy (NIRS) is an attractive tool potentially helping in preventing intraventricular hemorrhage and periventricular leukomalacia. Lung ultrasound is an emerging tool of diagnosis and can be of added value in helping monitoring the ventilated neonate. The aim of this scientific literature review is to address relevant aspects concerning the respiratory care and monitoring of the invasively ventilated newborn in order to help physicians to optimize the efficacy of care.


Objective: Nursing is a rewarding but also challenging profession. Nurses are at risk for burnout and premature exit from the profession, which is detrimental to them, their patients, and the healthcare system. There are few studies examining the unique correlates of burnout in nurses working with pediatric populations. The current 2-study project used mixed-methods (qualitative and then quantitative) analysis to explore burnout in nurses working in an inpatient unit with youth with chronic pain.

Method: Study I participants included all of the 32 nurses who worked in an inpatient pediatric unit, which admits patients with chronic pain. Qualitative analyses of focus groups were used to extract themes. These themes were examined via a quantitative battery completed by 41 nurses from 2 inpatient pediatric units with youth with chronic pain.

Results: The themes were burnout, moral distress, negative beliefs about chronic pain, barriers to pain management, fear of losing compassion, coworker support as a coping method, time worked in the unit, professional self-efficacy, and negative views of the hospital environment. Quantitative results supported most of the qualitative findings, and taken together, the findings supported a model of burnout in nurses working with youth with chronic pain.

Conclusions: Conclusions We integrated qualitative and quantitative findings to develop a model of nurse burnout. This model provides a framework for evaluating and targeting burnout in nurses working with pediatric patients with chronic pain.


INTRODUCTION: Our aims were (1) to explore the prevalence of burnout syndrome (BOS) and posttraumatic stress disorder (PTSD) in a sample of Spanish staff working in the paediatric intensive care unit (PICU) and compare these rates with a sample of general paediatric staff and (2) to explore how resilience, coping strategies, and professional and demographic variables influence BOS and PTSD.

MATERIALS AND METHODS: This is a multicentre, cross-sectional study. Data were collected in the PICU and in other paediatric wards of nine hospitals. Participants consisted of 298 PICU staff members (57 physicians, 177 nurses, and 64 nursing assistants) and 189 professionals working in non-critical paediatric units (53 physicians, 104 nurses, and 32 nursing assistants). They completed the Brief Resilience Scale, the Coping Strategies Questionnaire for healthcare providers, the Maslach Burnout Inventory, and the Trauma Screening Questionnaire.
RESULTS: Fifty-six percent of PICU working staff reported burnout in at least one dimension (36.20% scored over the cut-off for emotional exhaustion, 27.20% for depersonalisation, and 20.10% for low personal accomplishment), and 20.1% reported PTSD. There were no differences in burnout and PTSD scores between PICU and non-PICU staff members, either among physicians, nurses, or nursing assistants. Higher burnout and PTSD rates emerged after the death of a child and/or conflicts with patients/families or colleagues. Around 30% of the variance in BOS and PTSD is predicted by a frequent usage of the emotion-focused coping style and an infrequent usage of the problem-focused coping style.

DISCUSSION AND CONCLUSIONS: Interventions to prevent and treat distress among paediatric staff members are needed and should be focused on: (i) promoting active emotional processing of traumatic events and encouraging positive thinking; (ii) developing a sense of detached concern; (iii) improving the ability to solve interpersonal conflicts, and (iv) providing adequate training in end-of-life care.


In contrast to many of the malignant tumors that occur in the central nervous system in adults, the management, responses to therapy, and future perspectives of children with malignant lesions of the brain hold considerable promise. Within the past 5 years, remarkable progress has been made with our understanding of the basic biology of the molecular genetics of several pediatric malignant brain tumors including medulloblastoma, ependymoma, atypical teratoid rhabdoid tumour, and high grade glioma/diffuse intrinsic pontine glioma. The recent literature in pediatric neuro-oncology was reviewed, and a summary of the major findings are presented. Meaningful sub-classifications of these tumors have arisen, placing children into discrete categories of disease with requirements for targeted therapy. While the mainstay of therapy these past 30 years has been a combination of central nervous system irradiation and conventional chemotherapy, now with the advent of high resolution genetic mapping, targeted therapies have emerged, and less emphasis is being placed on craniospinal irradiation. In this article, the present and future perspective of pediatric brain malignancy are reviewed in detail. The progress that has been made offers significant hope for the future for patients with these tumors.


This article elaborates on how neonatologists and perinatologists might conceive of prognosis as an intervention with outcomes relevant to patients, families, and society at large and highlights aspects of this important area of practice requiring further study.


Children in foster care are considered a "vulnerable population" in clinical care and research, with good reason. These children face multiple medical, psychological, and social risks that obligate the child welfare and healthcare systems to protect them from further harms. An unintended consequence of the "vulnerable population" designation for children in foster care is that it may impose barriers on tracking and studying their health that creates gaps in knowledge that are key to their receipt of medical care and good outcomes. These gaps in knowledge have implications for justice, beneficence, and maleficence and serve to undermine "protection" of this population. Here we review the challenges of research regarding children in foster care, particularly medically complex children, and offer specific recommendations to include children in foster care in medical research.


BACKGROUND: Hurler syndrome (MPS IH), the severe, neurodegenerative form of type one mucopolysaccharidosis, is associated with rapid neurocognitive decline during toddlerhood and multi-system dysfunction. It is now standardly treated with hematopoietic cell transplantation (HCT), which halts accumulating disease pathology and prevents early death. While norm-based data on developmental functioning in untreated children have previously demonstrated neurocognitive decline, advances in methodology for understanding the cognitive functioning of children with neurodegenerative diseases have highlighted that the previous choice of scores to report results was not ideal. Specifically, the lowest possible norm-based score is 50, which obscures the
complete range of cognitive functioning at more advanced stages of neurodeterioration. To a set of cognitive data collected on a sample of untreated children, we applied a modern method of score analysis, calculating a developmental quotient based on age equivalent scores, to reveal the full range of cognitive functioning beneath this cutoff of 50, uncovering new information about the rapidity of decline and the profound impairment in these children.

RESULTS: Among 39 observations for 32 patients with untreated Hurler syndrome, the full array of cognitive functioning below 50 includes many children in the severely to profoundly impaired range. The loss of skills per time unit was 14 points between age 1 and 2. There was a very large range of developmental quotients corresponding to the norm-based cutoff of 50.

CONCLUSIONS: This report enables clarification of functioning at levels that extend beneath the floor of 50 in previous work. At the dawn of newborn screening and amidst a proliferation of new therapies for MPS I, these data can provide crucial benchmark information for developing treatments, particularly for areas of the world where transplant may not be available.


When a child dies suddenly and unexpectedly, we, as professionals, have a wide range of duties and obligations that must be fulfilled. Statutory requirements may place constraints on what we can do, when we need to do it, and how we can go about it. At the heart of it all, however, there remains a bereaved family, for whom the worst thing imaginable has just happened. As one bereaved mother put it: "Words may hurt me or make me angry, but I have lost my child, so don’t flatter yourself - nothing that you say will actually make the situation worse". Nevertheless, as the quotes at the start of this chapter highlight, parents’ experiences following the death of their child vary enormously, and the way we respond to them can make a considerable difference. The way we respond can make a difference also to the outcome of an investigation. Identifying an unusual medical cause of death, or uncovering the circumstances of a tragic accident or a case of intentional filicide is more likely with a thorough, systematic investigation, conducted with sensitivity and respect, than with one carried out carelessly or in a haphazard or aggressive manner. In most jurisdictions, the sudden unexpected death of an infant or child requires the case to be referred to a coroner, medical examiner, or procurator fiscal. In England, for example, a coroner is obliged to conduct an investigation into violent or unnatural deaths, deaths where the cause is unknown, and deaths which occur in custody or otherwise in state detention (1). Coroner’s officers, or police officers acting on behalf of the coroner, will need to carry out an investigation into the causes and circumstances of the death. Where there are concerns about parenting, or the possibility of abuse or neglect, there may be other children in the family who need protection, necessitating the involvement of children’s social care services. Nevertheless, the reality is that in the majority of cases the child’s death will be from natural causes, whether or not we are able to ascertain the actual cause. Therefore, health practitioners will need to carry out full investigations to look for possible medical causes, including infectious or genetic causes, which may have wider implications for the family or community. While infant mortality has fallen dramatically over the past decades, and continues to fall across the world, every death should still be seen as a tragedy, and we should do all we can to further reduce mortality rates and to reduce the risks of future child deaths. In order to do this effectively, we need to learn lessons - from each individual child’s death, and from the patterns of children’s deaths in any area - to identify potentially modifiable factors, and to take effective action to improve childcare, health and welfare services, and support for families. Through all this, all professionals will need to respond sensitively to the family in those awful, early stages of grief - in coming to terms with the reality of their child’s death, in coping with the practical arrangements that need to be made, in breaking the news to family and friends. Bearing in mind these varying and, at times, potentially conflicting obligations, five primary aims of our response to unexpected infant deaths can be defined (2, 3): 1. to establish, as far as is possible, the cause or causes of the infant’s death; 2. to identify any potential contributory or modifiable factors; 3. to provide ongoing support to the family; 4. to ensure that all statutory obligations are met; 5. to learn lessons, in order to reduce the risks of future infant deaths. Underpinning this are three fundamental principles of practice which support a positive response: a thorough, systematic approach to investigation, a sensitive approach to supporting families, and an attitude of collaboration and learning (Figure 5.1).


BACKGROUND:: Education and training for interdisciplinary pediatric providers requires training in principles of palliative and end-of-life (EOL) care. The experiences of bereaved parents can inform and enhance palliative care educational curricula in uniquely powerful and valuable ways. The objective of this study is to present an innovative palliative care educational program facilitated by trained bereaved parents who serve as volunteer educators in local and national palliative care educational forums and to describe how incorporation of bereaved parents in these educational forums affects participant comfort with communication and management of children at the EOL.

METHODS:: Parent educators underwent both general and session-specific training and participated in debriefings following each session. Survey tools were developed or adapted to determine how bereaved parent educators affected participant experiences in 3 different educational forums. Pre- and postsession surveys with incorporation of retrospective preprogram assessment items to control for response shift were used in the evaluation of institutional seminars on pediatric palliative and EOL care and role-play-based communication training sessions. Results from feedback surveys sent to attendees were used to appraise the participants’ experience at the international oncology symposium.

RESULTS:: Involvement of trained parent educators across diverse, interdisciplinary educational forums improved attendee comfort in communicating with, and caring for, patients and families with serious illness. Importantly, parent educators also derive benefit from involvement in educational sessions with interdisciplinary clinicians.

CONCLUSIONS:: Integration of bereaved parents into palliative and EOL care education is an innovative and effective model that benefits both interdisciplinary clinicians and bereaved parents.


The care of pediatric patients with cancer and their families is complex and rapidly evolving. Despite significant advances in outcomes, symptoms of the disease and complications of therapy continue to cause suffering that may improve with the involvement of pediatric palliative care (PPC) services. This descriptive study responds to the observation of great variability in PPC utilization within pediatric oncology. Data collected from 156 health care professionals (nurses, advanced practice professionals, and physicians) from a statewide hematology alliance evaluates the knowledge, beliefs, and perceived barriers to PPC involvement. Data analysis reveals significant variability when comparing respondents from professional roles and practice environments. Despite progress in PPC availability, care delivery remains incongruent with current recommendations. Knowledge gained from this study emphasizes the important role for all health care providers in advocating for support of PPC programs, educating the public, and committing to intentional involvement of PPC services while caring for pediatric oncology patients.


Snyder-Robinson syndrome, also known as spermine synthase deficiency, is an X-linked intellectual disability syndrome (OMIM #390583). First described by Drs. Snyder and Robinson in 1969, this syndrome is characterized by an asthenic body habitus, facial dysmorphism, broad-based gait, and osteoporosis with frequent fractures. We report here a pediatric autopsy of a 4 year old male with a history of intellectual disability, gait abnormalities, and osteoporosis. Despite significant advances in outcomes, symptoms of the disease and complications of therapy continue to cause suffering that may improve with the involvement of pediatric palliative care (PPC) services. This descriptive study responds to the observation of great variability in PPC utilization within pediatric oncology. Data collected from 156 health care professionals (nurses, advanced practice professionals, and physicians) from a statewide hematology alliance evaluates the knowledge, beliefs, and perceived barriers to PPC involvement. Data analysis reveals significant variability when comparing respondents from professional roles and practice environments. Despite progress in PPC availability, care delivery remains incongruent with current recommendations. Knowledge gained from this study emphasizes the important role for all health care providers in advocating for support of PPC programs, educating the public, and committing to intentional involvement of PPC services while caring for pediatric oncology patients.

Perinatal palliative care is an emerging area of health care. To date, no published tools assess health-care provider’s knowledge and level of comfort in providing such care. A 2-phase study was undertaken to develop and implement a survey to evaluate the self-assessed competency, attitudes, and knowledge of health-care providers working in perinatal palliative care. Phase 1 included a review of the literature and appraisal of palliative and death-related instruments to inform the initial draft of the Perinatal Palliative Care Survey (PPCS). Twenty-four Canadian pediatric palliative care specialists critiqued the PPCS, establishing its face and content validity. Phase 2 involved administering the PPCS at 4 sites across Canada, resulting in 167 responses from nurses, physicians, and midwives. The majority of participants responded that they possessed a degree of comfort in providing perinatal palliative care, particularly with assessing pain (76%), managing pain (69%), assessing other symptoms (85%), and managing other symptoms (78%). Two areas where participants level of confidence or extreme confidence was diminished included having conversations with families about the possibility of their infant dying (55%) and knowing and accessing community palliative care resources (32%). Responses in the knowledge section identified gaps related to opioid use, pharmacological interventions for breathlessness, pain behaviors, and tolerance developed to opioids and sedatives. Eighty-six percent of respondents stated that if education about palliative care was made available, they would participate with priority topics identified as communication with families (75%), managing other symptoms (78%). Two areas where participants level of confidence or extreme confidence was


In recent years, many different DNA mutations underlying the development of refractory epilepsy have been discovered. However, genetic diagnostics are still not routinely performed during presurgical evaluation and reports on epilepsy surgery outcome for patients with genetic refractory epilepsy are limited. We aimed to create an overview of the literature on seizure outcome following epilepsy surgery in patients with different genetic causes of refractory epilepsy. We systematically searched PubMed and Embase prior to January 2017 and included studies describing treatment outcome following epilepsy surgery in patients with genetic causes of epilepsy. We excluded studies in which patients were described with epilepsy due to Tuberous Sclerosis Complex or Sturge-Weber syndrome (since this extensive body of research has recently been described elsewhere) and articles in which surgery was aimed to be palliative. We identified 24 eligible articles, comprising a total of 82 patients who had undergone surgery for (mainly childhood-onset) refractory epilepsy due to 15 different underlying genetic causes. The success rate of surgery varied widely across these different genetic causes. Surgery was almost never effective in patients with epilepsy due to mutations in genes involved in channel function and synaptic transmission, whereas surgery was significantly more successful regarding seizure control in patients with epilepsy due to mutations in the mTOR pathway. Patients with a lesion on MRI tended to have higher seizure freedom rates than those who were MRI-negative. Although the evidence is still scarce, this systematic review suggests that studying genetic variations in patients with refractory epilepsy could help guide the selection of surgical candidates.


Context: Patients' need for comfort at the end of their lives is rarely fulfilled. The comfort of patients at the end of their lives, especially children, is affected by nurses’ understanding of what comfort means.

Aims: This research aims to explore and to understand the meaning of children's comfort at the end of their life for nurses. Setting and Design: The research applied descriptive qualitative phenomenology design.

Subject and Methods: The study was conducted at Jakarta. Nurses who have experience in caring the child at the end of their life were in-depth interview with an open-ended question. Data were then analyzed using the Colaizzi method. Results: This research identified six themes: striving to reduce children’s suffering, realizing what children wanted, observing the children felt comfortable in their family’s acceptance of their condition, facing internal and external conflict, experiencing mixed feelings knowing the children’s condition, and requiring support from all parties.

Conclusion: Nurses should provide information regarding children's end of life conditions to the family, to achieve family acceptances. Eventhough it was hard situation and rose internal conflict to nurses. It was found that children also felt comfortable at the end-of-life when they did not experience any suffering, and their wishes were granted. Subsequently, the nurses did not have mixed feelings when the children died. Therefore, evaluation of the training
effectiveness that has been given to the nurses should be done to fulfill the need of the child's comfort at the end-of-life.


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: In 2017, the Ohio Pediatric Palliative Care and End-of-Life Network (OPPEN) published nine domains of high-quality care for pediatric home-based hospice and palliative care (HBHPC). Eight domains established by the National Consensus Project (NCP) were validated for pediatric HBHPC, and a ninth domain of "Continuity and Coordination of Care" was added.

OBJECTIVE: The aim of this study was to establish definition criteria for each of these domains.

DESIGN AND SETTING: Using a modified Delphi technique, providers from the OPPEN were surveyed regarding definitions drawn from the NCP domain criteria. For the ninth domain, new definition criteria were generated de novo based on qualitative responses.

RESULTS: Definition criteria were established for the nine domains of quality in HBHPC previously identified. In the course of analysis, Bereavement Care was established as a 10th domain of quality, and definition criteria generated.

CONCLUSIONS: This is the first study to define domains of quality for pediatric HBHPC, and the second to leverage the infrastructure of a pediatric HPC statewide consortium toward this work. Future studies are needed to establish parent and patient-prioritized domains of quality in pediatric HBHPC, and to map indicators validated in pediatrics to these domains.


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: In hospital, staff need to routinely monitor patients to identify those who are seriously ill, so that they receive timely treatment to improve their condition. A Paediatric Early Warning System is a multi-faceted socio-technical system to detect deterioration in children, which may or may not include a track and trigger tool. It functions to monitor, detect and prompt an urgent response to signs of deterioration, with the aim of preventing morbidity and mortality. The purpose of this study is to develop an evidence-based improvement programme to optimise the effectiveness of Paediatric Early Warning Systems in different inpatient contexts, and to evaluate the feasibility and potential effectiveness of the programme in predicting deterioration and triggering timely interventions.

METHODS: This study will be conducted in two district and two specialist children's hospitals. It deploys an Interrupted Time Series (ITS) design in conjunction with ethnographic cases studies with embedded process evaluation. Informed by Translational Mobilisation Theory and Normalisation Process Theory, the study is underpinned by a functions based approach to improvement. Workstream (1) will develop an evidence-based improvement programme to optimise Paediatric Early Warning System based on systematic reviews. Workstream (2) consists of observation and recording outcomes in current practice in the four sites, implementation of the improvement programme and concurrent process evaluation, and evaluation of the impact of the programme. Outcomes will be mortality and critical events, unplanned admission to Paediatric Intensive Care (PICU) or Paediatric High Dependency Unit (PHDU), cardiac arrest, respiratory arrest, medical emergencies requiring immediate assistance, reviews by PICU staff, and critical deterioration, with qualitative evidence of the impact of the intervention on Paediatric Early Warning System and learning from the implementation process.

DISCUSSION: This paper presents the background, rationale and design for this mixed methods study. This will be the most comprehensive study of Paediatric Early Warning Systems and the first to deploy a functions-based approach to improvement in the UK with the aim to improve paediatric patient safety and reduce mortality. Our findings will inform recommendations about the safety processes for every hospital treating paediatric in-patients across the NHS.

Ensuring optimal nutrition is vital in critically ill children and enteral feeding is the main route of delivery in intensive care. Feeding intolerance is the most commonly cited reason amongst pediatric intensive care unit healthcare professionals for stopping or withholding enteral nutrition, yet the definition for this remains inconsistent, nebulous, and entirely arbitrary. Not only does this pose problems clinically, but research in this field frequently uses feeding intolerance as an endpoint and the heterogeneity in this definition makes the comparison of studies difficult and meta-analysis impossible. We reviewed the use of, and definitions of, the term feed intolerance in pediatric intensive care research papers in the last 20 years. Gastric residual volume remains the most common factor used to define feed intolerance, despite the lack of evidence for this. Healthcare professionals would benefit from further education to improve their awareness of the limitations of the markers to define feeding intolerance, and the international PICU community needs to agree a consistent definition of this phenomenon to improve consistency in both practice and research.

Conclusion: This paper will provide a narrative review of the definitions of, evidence for, and markers of feeding intolerance in critically ill children. What is Known?: * Feeding intolerance is a commonly cited reason amongst pediatric intensive care unit healthcare professionals for stopping or withholding enteral nutrition. * There is no agreed definition for feeding intolerance in critically ill children. What is New?: * This paper provides an up to date review of the definitions of, evidence for, and markers of feeding intolerance in critically ill children. * Despite no evidence, gastric residual volume continues to drive clinical bedside decisions about enteral feeding and feeding tolerance.

CONTEXT: Children have limited access to hospice care: few existing hospice programs have dedicated pediatric teams, and adult hospice providers feel inadequately trained to care for children.

OBJECTIVES: The aim of this study was to increase access to pediatric hospice care by empowering adult hospice providers to care for children through a comprehensive education program. Education empowers providers by changing their attitudes from inadequacy to confidence.

METHODS: The authors developed a two-day education program to train interdisciplinary teams of adult hospice providers in pediatric care. The curriculum consists of 13 modules to improve participants’ knowledge, skills, and attitudes. Ninety-three providers across the U.S. learned via multiple teaching methods including lectures, role plays by professional actors, interviews of bereaved parents, and self-reflections. Learning was evaluated with assessments before, immediately after, and six months after the program. Responses were compared using a one-sided analysis of variation with a significance level of alpha <0.05.

RESULTS: Participants improved their knowledge in 12 of 13 modules. Self-reported confidence levels with pediatric care improved significantly in all 13 modules (P < 0.05). After this program, 79% of providers reported feeling better prepared to care for pediatric hospice patients. Qualitative data reinforced that learners felt more prepared to care for pediatric patients.

CONCLUSION: A two-day, high-intensity low-cost community-based education program can improve adult providers’ knowledge of and skill level with pediatric care, leading to a change in attitude from fear to confidence. This model has the potential to increase access to pediatric hospice care as it uses existing adult hospice infrastructure.

CONCLUSION: A two-day, high-intensity low-cost community-based education program can improve adult providers’ knowledge of and skill level with pediatric care, leading to a change in attitude from fear to confidence. This model has the potential to increase access to pediatric hospice care as it uses existing adult hospice infrastructure.


BACKGROUND: Lack of pediatric palliative care (PPC) training impedes successful integration of PPC principles into pediatric oncology.

OBJECTIVES: We examined the impact of an enhanced implementation of the Education in Palliative and End-of-Life Care for Pediatrics (EPEC(R))-Pediatrics curriculum on the following: (1) knowledge dissemination; (2) health professionals’ knowledge; (3) practice change; and (4) quality of PPC.

DESIGN: An integrated knowledge translation approach was used with pre-/posttest evaluation of care quality. Setting/Subjects/Measurements: Regional Teams of 3-6 health professionals based at 15 pediatric oncology programs in Canada became EPEC-Pediatrics Trainers who taught the curriculum to health professionals (learners) and implemented quality improvement (QI) projects. Trainers recorded the number of learners at each education session and progress on QI goals. Learners completed knowledge surveys. Care quality was assessed through surveys with a cross-sectional sample of children with cancer and their parents about symptoms, quality of life, and care quality plus reviews of deceased patients’ health records.

RESULTS: Seventy-two Trainers taught 3475 learners; the majority (96.7%) agreed that their PPC knowledge improved. In addition, 10/15 sites achieved practice change QI goals. The only improvements in care quality were an increased number of days from referral to PPC teams until death by a factor of 1.54 (95% confidence interval [CI] = 1.17-2.03) and from first documentation of advance care planning until death by a factor of 1.50 (95% CI = 1.06-2.11), after adjusting for background variables.

CONCLUSION: While improvements in care quality were only seen in two areas, our approach was highly effective in achieving knowledge dissemination, knowledge improvement, and practice change goals.


BACKGROUND: Caring for a child near the end of life (EOL) can be a stressful experience. Resident physicians are often the frontline providers responsible for managing symptoms, communicating difficult information, and pronouncing death, yet they often receive minimal education on EOL care.

OBJECTIVE: To develop and implement an EOL curriculum and to study its impact on resident comfort and attitudes surrounding EOL care.

DESIGN: Kern’s 6-step approach to curriculum development was used as a framework for curriculum design and implementation. SETTING/PARTICIPANTS: Categorical and combined pediatric residents at a large quaternary care children’s hospital were exposed to the curriculum.

MEASUREMENTS: A cross-sectional survey was distributed pre- and postimplementation of the curriculum to evaluate its impact on resident comfort and attitudes surrounding EOL care.

RESULTS: One-hundred twenty-six (49%) of 258 residents completed the preimplementation survey, and 65 (32%) of 201 residents completed the postimplementation survey. Over 80% of residents reported caring for a dying patient, yet less than half the residents reported receiving prior education on EOL care. Following curriculum implementation, the percentage of residents dissatisfied with their EOL education fell from 36% to 14%, while the percentage of residents satisfied with their education increased from 14% to 29%. The postimplementation survey identified that resident comfort with communication-based topics improved, and they sought additional training in symptom management.

CONCLUSIONS: The implementation of a longitudinal targeted multimodal EOL curriculum improved resident satisfaction with EOL education and highlighted the need for additional EOL education.


PURPOSE: Children with severe neurological disabilities are at an increased risk of acute, life-threatening events. We assessed physicians’ attitudes when making decisions in these situations.
METHODS: We surveyed physicians in pediatric intensive care, neurology, and rehabilitation units in Swiss hospitals. The questionnaire explored participants’ attitudes toward life-threatening situations in two scenarios: a child with profound intellectual and multiple disabilities (PIMD) and an infant with spinal muscular atrophy (SMA) type I.

RESULTS: The participation rate was 55% (52/95). There was a consensus favoring non-invasive ventilation and comfort care as well as avoiding tracheostomy and invasive ventilation. For the child with PIMD, 61% of participants opposed cardiopulmonary resuscitation (CPR), 51% for the child with SMA. Physicians with over 20 years of experience were significantly more opposed to providing CPR than less experienced colleagues.

CONCLUSIONS: Physicians held different views, influenced by personal factors. This highlights the importance of standardizing multidisciplinary processes toward approaching these complex situations.


PURPOSE: We explored pediatricians’ practices and attitudes concerning end-of-life discussions (EOLds) with pediatric patients with cancer, and identified the determinants of pediatricians’ positive attitude toward having EOLds with pediatric patients.

METHODS: A multicenter questionnaire survey was conducted with 127 pediatricians specializing in the treatment of pediatric cancer.

RESULTS: Forty-two percent of participants reported that EOLds should be held with the young group of children (6-9 years old), 68% with the middle group (10-15 years old), and 93% with the old group (16-18 years old). Meanwhile, 6, 20, and 35% of participants answered that they "always" or "usually" discussed the incurability of the disease with the young, middle, and old groups, respectively; for the patient’s imminent death, the rates were 2, 11, and 24%. Pediatricians’ attitude that they "should have" EOLds with the young group was predicted by more clinical experience (odds ratio [OR] 1.077; p = 0.007), more confidence in addressing children’s anxiety after EOLd (OR 1.756; p = 0.050), weaker belief in the demand for EOLd (OR 0.456; p = 0.015), weaker belief in the necessity of the EOLd for children to enjoy their time until death (OR 0.506; p = 0.021), and weaker belief in the importance of maintaining a good relationship with the parents (OR 0.381; p = 0.025).

CONCLUSIONS: While pediatricians nearly reached consensus on EOLds for the old group, EOLds with the young group remain a controversial subject. While pediatricians who supported EOLds believed in their effectiveness or necessity, those who were against EOLds tended to consider the benefits of not engaging in them.


BACKGROUND: In children with profound intellectual and multiple disabilities (PIMD), discussions about end-of-life decisions (EoLds) are comparatively common. Nurses play a crucial role in the care for these children, yet their involvement in EoLD discussions is largely unknown. The objective of this research was to investigate the involvement in the hospital of nurses in discussions with parents and physicians about EoLds for children with PIMD.

METHOD: In a retrospective, qualitative study, we conducted semi-structured interviews with the nurses of 12 children with PIMD for whom an EoLD was made within the past 2 years.

RESULTS: Parents primarily discuss EoLds with nurses before and after the meeting with the physician. Nurses who were involved in EoL discussions with parents and physicians assisted them by giving factual information about the child and by providing emotional support. Some nurses, especially nurses from ID-care services, were not involved in EoL discussions, even if they had cared for the child for a long period of time. Some of the nurses had moral or religious objections to carrying out the decisions.

CONCLUSION: Most nurses were not involved in EoL discussions with parents and physicians in the hospital. Excluding nurses from EoL discussions can cause them moral distress. The involvement of nurses in EoL discussions for children with PIMD should be improved, especially by involving nurses from ID-care services. Because these nurses are usually familiar with the child, they can be valuable sources of information about the child’s quality of life.


AIMS AND OBJECTIVES: To explore aspects related to the fulfilment of the role of nurses in palliative sedation.

BACKGROUND: Palliative sedation demands knowledge and a proper attitude for maintaining comfort, preserving dignity and contributing to a peaceful death. In some developed countries, nurses have a well-established role in palliative sedation. However, studies on their role and its fulfilment are limited, particularly in the developing world.

DESIGN: An exploratory, mixed, qualitative and quantitative study was conducted. A self-administered questionnaire was used to examine the level of knowledge of palliative sedation and the level of confidence in skills and knowledge about palliative sedation. Also, focus groups were conducted to explore the emotional impact and the perceived role of nurses.

METHODS: Forty-one nurses from three advanced-care hospitals with palliative care units in Colombia completed the questionnaire. Also, four focus groups were conducted with 22 participants selected from the first phase.

RESULTS: A high level of knowledge regarding palliative sedation was found, but the level of confidence in skills was higher than the confidence in knowledge. The participants expressed their belief that their knowledge was derived from experience but believed that it was not enough to fulfil their role with confidence. A negative emotional impact about the patients’ condition was found. For some, it served as motivation to provide better care. For others, it was difficult to face, especially when assisting children. They also expressed satisfaction and gratification about providing relief from suffering through sedation.

CONCLUSIONS: The role of nursing is essential in palliative sedation. Although the nurses’ knowledge is adequate, it primarily derives from experience and not from formal training, which impacts on their perceived confidence and their distress.

RELEVANCE TO CLINICAL PRACTICE: Formal training for the optimal fulfilling of the nursing role in palliative sedation is crucial to provide better end-of-life care, particularly in developing countries.

Epidemiology and Pathology


OBJECTIVES: Although substantial reductions in under-5 mortality have been observed during the past 35 years, progress in the Eastern Mediterranean Region (EMR) has been uneven. This paper provides an overview of child mortality and morbidity in the EMR based on the Global Burden of Disease (GBD) study.

METHODS: We used GBD 2015 study results to explore under-5 mortality and morbidity in EMR countries.

RESULTS: In 2015, 755,844 (95% uncertainty interval (UI) 712,064-801,565) children under 5 died in the EMR. In the early neonatal category, deaths in the EMR decreased by 22.4%, compared to 42.4% globally. The rate of years of life lost per 100,000 population under 5 decreased 54.38% from 177,537 (173,812-181,463) in 1990 to 80,985 (76,308-85,876) in 2015; the rate of years lived with disability decreased by 0.57% in the EMR compared to 9.97% globally.

CONCLUSIONS: Our findings call for accelerated action to decrease child morbidity and mortality in the EMR. Governments and organizations should coordinate efforts to address this burden. Political commitment is needed to ensure that child health receives the resources needed to end preventable deaths.


Sudden unexpected death in epilepsy (SUDEP) is a rare in children; the risk of SUDEP in children is up to 10-fold less than adults. Herein, we report a case of SUDEP in a 14-year-old boy. The post-mortem findings in neuropathological examination in SUDEP are not pathognomonic. Tongue and lip bites marks are only an indication of a seizure before death. Basically, there are no lesions that could explain the incidence of seizures before death. However, post-mortem examination is mandatory in order to determine the diagnosis of SUDEP. Autopsy, histopathological, and toxicologic examinations and a proper medical history of epilepsy are required to come to diagnosis of SUDEP. This case report further demonstrates the importance of medicolegal autopsy in allegedly dead victims.


Background

Stroke is the fifth leading cause of death in young individuals globally. Data on the burden of sudden death by stroke are sparse in the young.

Aims The aim of this study was to report mortality rates, cause of death, stroke subtype, and symptoms in children and young adults who suffered sudden death by stroke.

Methods We conducted a retrospective, nationwide study including all deaths within Danish borders between 2000-2009 and 2007-2009 in persons aged 1-35 years and 36-49 years, respectively. Two physicians identified all sudden death cases through review of all death certificates. All available autopsy reports and records from hospitals and general practitioners were retrieved and a neurologist identified all sudden death by stroke cases. Results Of the 14,567 deaths in the 10-year period, there were 1,698 sudden death cases, of which 52 (3%) were sudden death by stroke. There was a male predominance (56%) and the median age was 33 years. The incidence of sudden death by stroke in individuals aged 1-49 years was 0.19 deaths per 100,000 person-years. Stroke was hemorrhagic in 94% of cases, whereof subarachnoid hemorrhage was the cause of death in 63% of cases. Seventeen (33%) cases contacted the healthcare system because of neurological symptoms, whereof one was suspected of having a stroke (6%).

Conclusions Sudden death by stroke in children and young adults occurs primarily due to hemorrhagic stroke. We report a high frequency of neurological symptoms prior to sudden death by stroke. Increased awareness among healthcare professionals towards stroke symptoms in children and young adults may lead to earlier detection of stroke, and thereby potentially lowering the incidence of sudden death by stroke.

Retinoblastoma can present in 1 or both eyes and is the most common intraocular malignancy in childhood. It is typically initiated by biallelic mutation of the RB1 tumor suppressor gene, leading to malignant transformation of primitive retinal cells. The most common presentation is leukocoria, followed by strabismus. Heritable retinoblastoma accounts for 45% of all cases, with 80% being bilateral. Treatment and prognosis of retinoblastoma is dictated by the disease stage at initial presentation. The 8th Edition American Joint Committee on Cancer (AJCC) TNM/H (tumor, node, metastasis, heritable trait) staging system defines evidence-based clinical and pathological staging for overall prognosis for eye(s) and child. Multiple treatment options are available in 2018 for retinoblastoma management with a multidisciplinary team, including pediatric ocular oncology, medical oncology, radiation oncology, genetics, nursing, and social work. Survival exceeds 95% when disease is diagnosed early and treated in centers specializing in retinoblastoma. However, survival rates are less than 50% with extraocular tumor dissemination. We summarize the epidemiology, genetics, prenatal screening, diagnosis, classification, investigations, and current therapeutic options in the management of retinoblastoma.


A salivary gland anlage tumour (SGAT) is a very rare type of benign tumour that usually presents in early infancy with respiratory distress which is exacerbated upon feeding. We report a full-term male neonate who was referred to the Al Nahdha Hospital, Muscat, Oman, in 2015 with severe neonatal respiratory distress due to a nasopharyngeal obstruction immediately after birth. Computed tomography and magnetic resonance imaging revealed a well-circumscribed mass in the nasopharynx, without intracranial extension. Histopathological analysis of the lesion confirmed a diagnosis of SGAT. Following excision of the tumour, the postoperative period was uneventful. No recurrence was observed over the next two years. This case report highlights the importance of the early recognition of this extremely rare and potentially life-threatening, yet easily curable, condition.


OBJECTIVE: Our goal is to highlight the prenatal diagnosis and management of central nervous system (CNS) anomalies through sharing our clinic’s experience.

MATERIAL AND METHODS: We evaluated prenatal findings and postnatal outcomes of neonates who had a CNS anomaly diagnosis in our clinic over a ten-year period. A total of 183 cases with various CNS anomalies were included in the study. Birth or termination preferences of mothers were recorded in all cases, and postnatal diagnosis concordance and prognosis after surgical procedures were evaluated in mothers who chose to continue the pregnancy.

RESULTS: The mean maternal age was 28.2+-5.5 years, mean gravida was 2.2+-1.3, and the mean gestational age at diagnosis was 30.5+-5.5 weeks. Seventy five out of 183 (41%) patients chose to terminate their pregnancy. Twenty babies (26.6%) in the termination of pregnancy group had additional anomalies. A hundred and eight patients gave birth at our institution. Mean birth weight was 3060+-647.5 g, mean gestational week at delivery was 37.9+-1.7 weeks, and mean Apgar score (5th minute) was 8.8+-/2.3. Four neonates died at the postpartum first day. The postnatal diagnosis of 60 of the 108 (55.5%) patients who gave birth was concordant with the prenatal diagnosis, and 32 of the 108 (29.6%) babies underwent surgical interventions.

CONCLUSION: CNS anomalies have broad spectrum and variable prognoses. This study highlights the limitations of prenatal diagnoses, and the need for parents to have this information in order to determine the course of their pregnancy and prepare themselves for the postnatal challenging treatment/rehabilitation process.
BACKGROUND: Data on pediatric HIV in Peru are limited. The National Institute of Child Health (Instituto Nacional de Salud del Niño; INSN) cares for the most HIV-infected children under the age of 18 years in the country. We describe the outcomes of children seen at INSN’s HIV clinic over the 10 years when antiretroviral therapy and prevention of mother-to-child transmission (PMTCT) interventions became available in 2004.

METHODS: We conducted a retrospective review of INSN HIV clinic patients between 2003 and 2012. Deidentified data were collected and analyzed. RESULTS: A total of 280 children were included: 50.0% (140/280) were male; 80.0% (224/280) lived in metropolitan Lima. Perinatal transmission was the mode of HIV infection in 91.4% (256/280) of children. Only 17% (32/191) of mothers were known to be HIV-infected at delivery; of these mothers, 41% (13/32) were receiving antiretroviral therapy at delivery. 72% (23/32) delivered by Cesarean section and 47% (15/32) of their infants received antiretroviral prophylaxis. Median age at HIV diagnosis for all children was 35.7 months (interquartile range 14.5-76.8 months), and 67% (143/213) had advanced disease (clinical stage C). After HIV diagnosis, the most frequent hospitalization discharge diagnoses were bacterial pneumonia, chronic malnutrition, diarrhea, anemia and tuberculosis. Twenty-four patients (8.6%) died at a median age of 77.4 months.

CONCLUSIONS: Most cases of pediatric HIV were acquired via perinatal transmission; few mothers were diagnosed before delivery; and among mothers with known HIV status, PMTCT was suboptimal even after national PMTCT policy was implemented. Most children were diagnosed with advanced disease. These findings underscore the need for improving early pediatric HIV diagnosis and treatment, as well as PMTCT strategies.

INTRODUCTION: Congenital anomalies (CAs) represent one of the main causes of foetal death, infant mortality and morbidity, and long-term disability. CAs have been object of systematic registration activity for a long-time in many geographical areas in Europe and worldwide. CAs are often associated with disabilities of different types and severity, including the developed Countries worldwide. According to the World Health Organization (WHO), each year approximately 3.2 million of children worldwide are born with a CA and approximately 300,000 newborns with a diagnosis of birth defect die within the first 28 days of life. In Europe, CAs are the leading cause of perinatal mortality: the European Surveillance of Congenital Anomalies (EUROC AT) network estimated a perinatal mortality associated with CAs of 9.2 per 10,000 births in 2008-2012. In Italy, the Ministry of Health estimates that, on the average of 500,000 births each year, about 25,000 present at least one CA. Moreover, approximately 25% of infant mortality is due to CAs and about 50% of infant mortality is attributable to perinatal mortality, almost always of prenatal origin. Regarding long-term survival, a recent population study conducted between 1985 and 2003 in the UK estimated a 20.5-year survival of 85.5% of children born with at least one CA. According to the Centre for Disease Control and Prevention, approximately 3.3% of live births in the United States have a severe birth defect. Since CAs represent a significant public health issue, an effective primary prevention strategy should be a priority for public policies and healthcare system. Regarding aetiology, although in many cases the cause is still unknown, it has been hypothesized that CAs may be developed during the first trimester of pregnancy as a result of hereditary polygenic defects or of a gene-environment interaction. The aetiology is predominantly multifactorial, caused by complex interactions between genes and environment, which modify the normal embryo-foetal development, especially during the organogenesis phase. In particular, environmental factors (e.g., chemical toxicants, infection agents, maternal disease, and exogenous factors) can have preconceptional mutagenic action, postconceptional teratogenic effects, periconceptional endocrine disruption or epigenetic action. Regarding genetic causes, there are genetic chromosomal aberrations or dysgeneses. Furthermore, socioeconomic factors affect reproductive health by differentiating the exposure to the other risk factors as well as the access to prevention measures. In recent years, the importance of the environment as a major factor of reproductive risk has been highlighted. An individual may be exposed to pollutants present in the workplace and the population may be exposed to multiple sources of environmental contamination of water, soil, and air matrices. Pregnant women and the developing foetus are particularly sensitive to the effects of environmental exposure.

OBJECTIVE: The aim of the present working paper is to produce an updated review of the epidemiological evidence on the risk of CAs associated with environmental exposures, socioeconomic, and main individual risk factors, such as cigarette smoking and alcohol consumption, according to the approach proposed by Pirastu et al. 2010 in the framework of the SENTIERI Project (the Italian Epidemiological Study of Residents in National Priority Contaminated Sites).
DESIGN AND METHODS: Literature search was carried out in PubMed, following the SENTIERI project criteria to evaluate evidence, by selecting articles in English or Italian language published from 2011 to 2016 regarding human studies. For this review, descriptive and analytical epidemiological studies (cohort, case-control, cross-sectional, and ecological), systematic reviews, and metanalyses reporting association estimates between the outcome and at least one of the risk factors were selected. As in Pirastu et al., the sources of environmental exposure have been classified into four macrocategories: industries, mines, landfills, and incinerators. The sources of individual exposure considered were: active and passive cigarette smoking, alcohol consumption, socioeconomic status (SES), occupational and environmental exposures related to air pollutants from vehicular traffic only. The obtained results were assessed according to the evaluation criteria on the epidemiological evidence related to the association between the outcome and exposures predefined and published by the SENTIERI working group (WG). For the evidence assessment, the SENTIERI WG criteria favoured firstly primary sources and quantitative metanalyses, secondly, consistency among sources. The evaluation of the epidemiological evidence for the association between outcome and the exposure has been classified into three categories: sufficient (S), limited (L), inadequate (I).

RESULTS: Industries: during the period under review, six single studies evaluating the association between industrial sites exposure and the risk of CAs were found. The epidemiological evidence of association between outcome and exposure has been considered limited. Mines: from the bibliographic research, three single studies investigating possible cause-effect relationship between maternal residential proximity to mines and the risk of CAs have been collected providing inadequate epidemiological evidence. Landfills: during the period under review, one systematic review and one literature review evaluating the causal associations between maternal residential proximity to landfills and CAs were identified. The epidemiological evidence is limited and concerns almost exclusively sites containing industrial or hazardous waste. Incinerators: a systematic review has been selected; it concludes that the evidence for the association between maternal residential proximity to incinerators and CAs are inadequate. Cigarette smoking: the literature search identified eight systematic reviews with metanalysis, five multicentre studies, and ten single studies assessing the causal association between maternal and/or paternal exposure to smoking and the risk of CAs in the offspring providing sufficient evidence for a causal association between maternal exposure to cigarette smoke and the risk of congenital heart defects, oro-facial clefts, neural tube defects, and gastrointestinal malformations. Alcohol: three systematic reviews with metanalysis, two metanalyses, one multicentre study, and four single studies were collected for the period under review. The acquired literature has provided limited epidemiological evidence for associations between alcohol consumption and CAs in the nervous system, particularly for anencephaly and spina bifida. Socioeconomic status: the evidence of an association with socioeconomic factors was inadequate due to an insufficient number of studies selected during the period under consideration. Occupational exposure: the literature search collected one metanalysis, eight multicentre studies, and five single studies. The epidemiological evidence for associations between paternal occupational exposure to solvents and neural tube defects and between maternal pesticide exposure and gold-facial clefts were judged limited. Air pollution: two systematic reviews with metanalyses, two multicentre studies, and nine single studies were selected by literature search; the epidemiological evidence for a causal association between air pollutants exposure and the risk of CAs is still to be considered limited.

CONCLUSIONS: For future epidemiological studies, a better exposure assessment, using in particular more accurate spatial measurements or models, a standardized case definition, a larger sample and more accurate control of the recognized or presumed confounding variables are needed.


BACKGROUND: Southeast Asia is undergoing a transition from infectious to chronic diseases, including a dramatic increase in adult cancers. Childhood cancer research in Thailand has focused predominantly on leukemias and lymphomas or only examined children for a short period of time. This comprehensive multisite study examined childhood cancer incidence and survival rates in Thailand across all International Classification of Childhood Cancer (ICCC) groups over a 20-year period.

METHODS: Cancer cases diagnosed in children ages 0-19 years (n = 3574) from 1990 to 2011 were extracted from five provincial population-based Thai registries, covering approximately 10% of the population. Descriptive statistics of the quality of the registries were evaluated. Age-standardized incidence rates (ASRs) were calculated using the Segi world standard population, and relative survival was computed using the Kaplan-Meier method. Changes in incidence and survival were analyzed using Joinpoint Regression and reported as annual percent changes (APC).

RESULTS: The ASR of all childhood cancers during the study period was 98.5 per million person-years with 91.0 per million person-years in 1990-2000 and 106.2 per million person-years in 2001-2011. Incidence of all childhood cancers increased significantly (APC = 1.2%, P < 0.01). The top three cancer groups were leukemias, brain tumors,
and lymphomas. The 5-year survival for all childhood cancers significantly improved from 39.4% in 1990-2000 to 47.2% in 2001-2011 (P < 0.01).

CONCLUSIONS: Both childhood cancer incidence and survival rates have increased, suggesting improvement in the health care system as more cases are identified and treated. Analyzing childhood cancer trends in low- and middle-income countries can improve understanding of cancer etiology and pediatric health care disparities.


Bryant, V. A. and N. J. Sebire (2018). Natural Diseases Causing Sudden Death in Infancy and Early Childhood. SIDS Sudden Infant and Early Childhood Death: The Past, the Present and the Future. J. R. Duncan and R. W. Byard. Adelaide (AU), University of Adelaide Press (c) 2018 The Contributors, with the exception of which is by Federal United States employees and is therefore in the public domain.

Global childhood mortality rates in the under-5s were 44 per 1,000 live births in 2013, ranging from 2.3 in Singapore to 152.5 in Guinea-Bissau (Western sub-Saharan Africa), with rates of 4.9 per 1,000 live births in the United Kingdom (UK) (1). In England and Wales there are >5,000 deaths annually in children aged 0-19 years (2) from an estimated population of 12.9 million in this age group (3). Around 3,000 of these deaths are in infants (less than 1 year) with the majority having known serious medical conditions; such deaths are hence "expected". Most are due to perinatal and immaturity-related conditions, which account for around 40% of cases, followed by congenital anomalies. Many of these deaths occur in the early (less than 7 days) or late neonatal (7 to 27 days) period (2). The next most commonly affected age group is adolescents, who account for around 1,000 deaths annually, with more than half being due to external, non-natural causes (2). Unexpected death occurring in an apparently healthy infant is termed "sudden unexpected death in infancy (SUDI)" and refers to such a presentation in an infant 7-365 days of age. According to most definitions, unexpected deaths in infants under 7 days of age are excluded from the SUDI category, and instead have been termed "sudden unexpected early neonatal death (SUEND)". All cases of SUDI and SUEND require investigation to determine the cause of death. In England and Wales such cases are referred to Her Majesty’s Coroner (HMC), who will direct a post-mortem examination by a specialist pediatric pathologist. The primary rationale of the post-mortem examination, including its components and ancillary investigations, is to diagnose or exclude those natural (and non-natural) causes of death which are identifiable and to allow a specific cause of death to be provided (the specific details of the autopsy procedure are detailed in Chapter 24). Whilst many cases will subsequently be found to have died from previously unrecognized medical conditions, such as congenital anomalies or acquired natural diseases, a significant number will remain unexplained despite a complete autopsy including ancillary investigations (microbiology, virology, radiology, and metabolic studies). These cases are referred to as "unexplained SUDI", "unascertained", or "sudden infant death syndrome (SIDS)" according to the precise circumstances of the case and local practice, these terms by definition being diagnoses of exclusion. Excluding SIDS cases and neonatal deaths (0-27 days), for infants and children in England and Wales the most common acquired causes of natural deaths are neoplasms, diseases affecting the neurological, cardiovascular or respiratory systems, and infections (2) (Figure 25.1). It is likely that >50% of these cases may occur in infants and children with known life-limiting conditions. However, similar to in infancy, sudden unexpected death in childhood (SUDC; >1 year) also occurs, albeit less frequently than SUDI, with cases referred to the Coroner in the same manner. Following investigation, unexplained SUDC is less common than SIDS but remains a significant proportion of all childhood deaths; in England and Wales, for example, there were 212 registered SIDS cases compared to 27 unexplained SUDC cases in 2014 (4). However, globally, accurate figures regarding the proportions of explained and unexplained deaths following autopsy are difficult to establish. This is, in part, due to wide variability in the death certification process, making epidemiological evaluation unreliable (5), and a lack of large population-based studies, in particular those investigating SUDC. A recent review identified 24 published studies investigating 25 cohorts (17 in infants, 4 including both infants and children, and 4 children only) from 11 different countries; following full investigation the cause of death was found in 9-67% of SUDI and 22-86% of SUDC cases (6). In the same study, infection was reported as the commonest explanation for death overall in SUDI (52% of all the cases reported across studies) and variably reported in individual studies to account for between 15-86% of the explained cases. Of the studies in children >1 year, 36-68% of explained deaths were due to infectious causes (6).

https://www.ncbi.nlm.nih.gov/pubmed/30035967


Background: Neurocognitive impairment in survivors of childhood cancer may be associated with direct neurotoxicity, as well as indirect effects of systemic health complications. We evaluated associations among treatment exposures, chronic health conditions, and neurocognitive outcomes in adult survivors of childhood cancer.
Methods: Participants included 5507 adult survivors of childhood cancer (47.1% male; mean [SD] age = 31.8 [7.6] years at evaluation; 23.1 [4.5] years postdiagnosis) in the Childhood Cancer Survivor Study who completed a self-report measure of neurocognitive function. Cardiac, pulmonary, and endocrine chronic health conditions were graded using the National Cancer Institute Common Terminology Criteria for Adverse Events (version 4.03). Structural equation modeling was used to examine a priori hypothesized causal pathways among cancer treatment, subsequent chronic health conditions, and neurocognitive outcomes. Multivariable models were used to estimate relative risk for associations of treatments and chronic conditions on neurocognitive function. All statistical tests were two-sided.

Results: One-third of survivors with a grade 2 or higher chronic condition reported impairments in task efficiency and memory. In addition to direct effects of cranial radiation, path analyses and multivariable models demonstrated direct effects of cardiopulmonary (beta = 0.10, P = .002; relative risk [RR] = 1.27, 95% confidence interval [CI] = 1.12 to 1.44) and endocrine (beta = 0.07, P = .04; RR = 1.14, 95% CI = 1.02 to 1.28) conditions on impaired task efficiency. We identified similar effects of cardiopulmonary condition on memory (P = .01) and emotional regulation (P = .01). Thoracic radiation was associated with impaired task efficiency (P = .01) and emotional regulation (P = .01) through endocrine morbidity.

Conclusions: Non-neurotoxic exposures, such as thoracic radiation, can adversely impact survivors’ neurocognitive function through chronic conditions. Management of chronic diseases may mitigate neurocognitive outcomes among aging survivors of childhood cancer.


We demonstrated the pattern in presentation of primary intracranial tumors in a population-based cohort of patients aged 0-24 years identified from the National Cancer Registry for England, using linked medical records from primary care and hospitals. We used generalized additive models to estimate temporal changes in presentation rates. Borderline and malignant tumors presented at a similar rate in primary care (6.4 and 6.6 consultations per 100 patients each month) and in hospital (3.4 and 3.6). Benign tumors presented earlier but less frequently (rate = 4.4 and rate ratio = 0.75, 95% CI = 0.60-0.93, in primary care; rate = 2.6 and rate ratio = 0.83, 95% CI = 0.77-0.89, in hospital). Many tumors began presenting shortly before their diagnosis, but less aggressive tumors were likely to present earlier in primary care. Earlier detection of less aggressive tumors in primary care may reduce the risk of complications and morbidity among survivors.


CLINICAL CHARACTERISTICS: The dystrophinopathies cover a spectrum of X-linked muscle disease ranging from mild to severe that includes Duchenne muscular dystrophy, Becker muscular dystrophy, and DMD-associated dilated cardiomyopathy (DCM). The mild end of the spectrum includes the phenotypes of asymptomatic increase in serum concentration of creatine phosphokinase (CK) and muscle cramps with myoglobinuria. The severe end of the spectrum includes progressive muscle diseases that are classified as Duchenne/Becker muscular dystrophy when skeletal muscle is primarily affected and as DMD-associated dilated cardiomyopathy (DCM) when the heart is primarily affected. Duchenne muscular dystrophy (DMD) usually presents in early childhood with delayed motor milestones including delays in walking independently and standing up from a supine position. Proximal weakness causes a waddling gait and difficulty climbing stairs, running, jumping, and standing up from a squatting position. DMD is rapidly progressive, with affected children being wheelchair dependent by age 12 years. Cardiomyopathy occurs in almost all individuals with DMD after age 18 years. Few survive beyond the third decade, with respiratory complications and progressive cardiomyopathy being common causes of death. Becker muscular dystrophy (BMD) is characterized by later-onset skeletal muscle weakness. With improved diagnostic techniques, it has been recognized that the mild end of the spectrum includes men with onset of symptoms after age 30 years who remain ambulatory even into their 60s. Despite the milder skeletal muscle involvement, heart failure from DCM is a common cause of morbidity and the most common cause of death in BMD. Mean age of death is in the mid-60s. DMD-associated DCM is characterized by left ventricular dilation and congestive heart failure. Females heterozygous for a DMD pathogenic variant are at increased risk for DCM.

DIAGNOSIS/TESTING: The diagnosis of a dystrophinopathy is established in a proband with the characteristic clinical findings and elevated CK concentration and/or identification of a hemizygous pathogenic variant in DMD.
on molecular genetic testing in a male and of a heterozygous pathogenic variant in DMD on molecular genetic testing in a female. Females may present with a classic dystrophinopathy or may be asymptomatic carriers.

MANAGEMENT: Treatment of manifestations: ACE inhibitors are used with or without beta blockers for cardiomyopathy in both DMD and BMD phenotypes. Congestive heart failure is treated with diuretics and oxygen as needed; cardiac transplantation is offered to persons with severe dilated cardiomyopathy and BMD with limited or no clinical evidence of skeletal muscle disease. Scoliosis is treated with bracing and surgery. Corticosteroid therapy improves muscle strength and function for individuals with DMD between ages five and 15 years; the same treatment is used in BMD, although the efficacy is less clear. Prevention of secondary complications: Evaluation by a pulmonologist and cardiologist before surgeries; pneumococcal and influenza immunizations annually; nutrition assessment; physical therapy to promote mobility and prevent contractures; sunshine and a balanced diet rich in vitamin D and calcium to improve bone density and reduce the risk of fractures; weight control to avoid obesity. Surveillance. For males with DMD or BMD: annual or biannual evaluation by a cardiologist beginning at the time of diagnosis; monitoring for scoliosis; baseline pulmonary function testing before wheelchair dependence; frequent evaluations by a pediatric pulmonologist. For heterozygous females: cardiac evaluation at least once after the teenage years. Agents/circumstances to avoid: Botulinum toxin injections; succinylcholine and inhalational anesthetics because of susceptibility to malignant hyperthermia or malignant hyperthermia-like reactions. Evaluation of relatives at risk: Early identification of heterozygous females who are at increased risk for cardiomyopathy and, thus, need routine cardiac surveillance and prompt treatment.

GENETIC COUNSELING: The dystrophinopathies are inherited in an X-linked manner. The risk to the sibs of a proband depends on the genetic status of the mother. Heterozygous females have a 50% chance of transmitting the DMD pathogenic variant in each pregnancy. Sons who inherit the pathogenic variant will be affected; daughters who inherit the pathogenic variant are heterozygous and may have a range of clinical manifestations. Males with DMD usually do not reproduce. Males with BMD or DMD-associated DCM may reproduce: all of their daughters are heterozygotes; none of their sons inherit their father’s DMD pathogenic variant. Carrier testing for at-risk females and prenatal testing or preimplantation genetic diagnosis for pregnancies at increased risk are possible if the DMD pathogenic variant in the family is known.


BACKGROUND: Despite recent therapeutic advancements, Wilms tumour (WT) presents remarkable survival variations. We explored mortality and survival patterns for children (0-14 years) with WT in 12 Southern and Eastern European (SEE) countries in comparison with the United States of America (USA).

METHODS: A total of 3966 WT cases (0-14 years) were registered by a network of SEE childhood cancer registries (N:1723) during available registration periods circa 1990-2016 and surveillance, epidemiology, and end results program (SEER) (N:2243; 1990-2012); mortality data were provided by the respective national statistical services. Kaplan-Meier curves and Cox proportional hazards models were used to assess the role of age, sex, year of diagnosis, urbanisation and Human Development Index (HDI) on overall survival (OS).

RESULTS: Persisting regional variations shape an overall 78% 5-year OS in the participating SEE countries, lagging behind the USA figure (92%, p=0.001) and also reflected by higher SEE mortality rates. Worth mentioning is the gradually escalating OS in SEE (hazard ratio [HR]5-year increment:0.67, 95% confidence interval [CI]:0.60, 0.75) vs. a non-significant 10% improvement in the SEER data, which had a high starting value. OS differentials [two-fold less favourable among children aged 10-14 years, boys and those living in rural SEE areas (HR:1.37; CI:1.10-1.71) or countries with inferior HDI (2-3-fold)] were minimal in the USA.

CONCLUSIONS: Children with WT residing in SEE countries do not equally enjoy the substantial survival gains, especially for those living in rural areas and in lower HDI countries. Noteworthy are steep and sizeable survival gains in SEE along with the newly presented Greek data pointing to achievable survival goals in SEE despite the financial crisis.


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.


Nonalcoholic fatty liver disease (NAFLD) has become the dominant form of chronic liver disease in children and adolescents with the increasing prevalence of obesity worldwide. NAFLD represents a wide spectrum of conditions, ranging from fatty liver - which generally follows a benign, non-progressive clinical course - to non-alcoholic steatohepatitis, a subset of NAFLD that may progress to cirrhosis and end-stage liver disease or liver carcinoma. The underlying pathophysiological mechanism of "pediatric" NAFLD remains unclear, although it is strongly associated with obesity and insulin resistance. In this review we provide a general overview on the current understanding of NAFLD in children and adolescents, which underpins practice, enabling early diagnosis and appropriate therapeutic intervention for this life-threatening liver disease.


Primary bone cancers include osteosarcoma, Ewing sarcoma, and chondrosarcoma. They account for less than 1% of diagnosed cancers each year and are associated with significant morbidity and mortality. Timely diagnosis is challenging because of late patient presentation, nonspecific symptoms that mimic common musculoskeletal injuries, and low suspicion by physicians. Plain radiography is the preferred diagnostic test. Radiographic suspicion of a bone malignancy should prompt quick referral to a cancer center for multidisciplinary care. Osteosarcoma, the most common bone cancer, most often occurs in children and adolescents. It typically develops in the metaphysis of long bones, specifically the distal femur, proximal tibia, and proximal humerus. Metastasis to the lungs is common. Use of neoadjuvant and adjuvant chemotherapy, in combination with surgery, has improved survival rates to nearly 80% for patients with localized disease, and 90% to 95% of patients do not require limb amputation. Ewing sarcoma is the second most common bone cancer and is similar to osteosarcoma in terms of presenting symptoms, age at occurrence, and treatment. Prognosis for osteosarcoma and Ewing sarcoma depends on the presence of metastasis, which lowers the five-year survival rate to 20% to 30%. Chondrosarcoma is the rarest bone cancer, primarily affecting adults older than 40 years. Survival rates are higher because most of these tumors are low-grade lesions.


PURPOSE: To describe the clinical features of electric powered indoor/outdoor wheelchair users with a muscular dystrophy, likely to influence optimal prescription; reflecting features of muscular dystrophies, conditions secondary to disability, and comorbidities impacting on equipment provision.

METHODS: Cross-sectional retrospective case note review of recipients of electric powered indoor/outdoor wheelchairs provided by a specialist regional wheelchair service. Data on demography, diagnostic/clinical, and wheelchair prescription were systematically extracted. RESULTS: Fifty-one men and 14 women, mean age 23.7
Forty had Duchenne muscular dystrophy, 22 had other forms of muscular dystrophy, and three were unclassified. Twenty-seven were aged under 19. Notable clinical features included problematic pain (10), cardiomyopathy (5), and ventilatory failure (4). Features related to disability were (kypho)scoliosis (20) and edema/cellulitis (3) whilst comorbidities included back pain (5). Comparison of younger with older users revealed younger users had more features of muscular dystrophy affecting electric powered chair provision (56%) whilst older users had more comorbidity (37%). Tilt-in-space was prescribed for 81% of users, specialized seating for 55% and complex controls for 16%.

CONCLUSIONS: Muscular dystrophy users were prescribed electric powered indoor/outdoor chairs with many additional features reflecting the consequences of profound muscle weakness. In addition to facilitating independence and participation, electric powered indoor/outdoor chairs have major therapeutic benefits. Implications for rehabilitation Powered wheelchairs have therapeutic benefits in managing muscular dystrophy pain and weakness. The use of specialized seating needs careful consideration in supporting progressive muscle weakness and the management of scoliosis. Pain, discomfort, pressure risk, and muscle fatigue may be reduced by use of tilt-in-space.


OBJECTIVE: To identify perinatal mortality risk factors in the Southern Zone of Tigray, northern Ethiopia.

METHODS: The present unmatched case-control study included data from 20 health facilities; stillbirths and neonatal deaths were included as a case group and patients with neonates who survived until discharge or day 7 postpartum were included as a control group. Perinatal mortality risk factors were investigated using bivariate and multivariate logistic regression analyses.

RESULTS: There were 126 and 252 patients included in the case and control groups, respectively. Prematurity (adjusted odds ratio [AOR] 12.2; 95% confidence interval [CI] 3.46-43.17; P<0.001), delivery weight below 2500 g (AOR 11.5; 95% CI 3.16-42.36; P<0.001), and fewer prenatal visits (AOR 5.4; 95% CI 0.80-36.63; P=0.028) were determinants of perinatal mortality. Partograph use (AOR 0.2; 95% CI 0.08-0.48; P<0.001) and seeking labor care at the start of labor (AOR 0.1; 95% CI 0.01-0.96; P=0.010) were protective. Short childbirth interval (<2 years) (AOR 2.2; 95% CI 1.03-5.09; P=0.013), distance to facility (AOR 3.7; 95% CI 1.56-9.02; P=0.007), and lack of iron supplementation (AOR 3.3; 95% CI 1.16-9.76; P=0.021) were also predictors of perinatal mortality.

CONCLUSION: Perinatal mortality was linked to prematurity and low delivery weight. Interventions including partograph and auscultation, as well as subsidizing transport and iron supplementation, could help in this region.


We studied the etiology of pediatric acute encephalitis/encephalopathy (pAEE) using epidemiological data obtained from a nationwide survey in Japan. Two-step questionnaires were sent to the pediatric departments of hospitals throughout the country in 2007, querying the number of the cases during 2005-2006 as the first step, and asking for the details of clinical information as the second step. In all, 636 children with pAEE (age </= 15 years) were enrolled. For the known etiology of pAEE (63.5% of the total cases), 26 microbes and 2 clinical entities were listed, but the etiology of 36.5% remained unknown. Influenza virus (26.7%), exanthem subitum (12.3%), and rotavirus (4.1%) were the most common, and the incidence of pAEE peaked at the age of 1 year. This trend was common among all etiologies. Among the neurological symptoms observed at the onset of pAEE, seizures were observed more often in patients aged </= 3 years, although abnormal speech and behavior were also common in older children. Undesirable outcomes (death and neurological sequelae) occurred at high rates in patients with any known etiology other than mycoplasma. In conclusion, these findings provide comprehensive insight into pAEE in Japan.


This report presents final 2016 data on the 10 leading causes of death in the United States by age, sex, race, and Hispanic origin. Leading causes of infant, neonatal, and postneonatal death are also presented. This report
supplements "Deaths: Final Data for 2016," the National Center for Health Statistics’ annual report of final mortality statistics.


AIM: The primary objective of this study was to describe demographics and end-of-life treatments of children with cancer at a government tertiary cancer center in India.

METHODS: A retrospective review was undertaken of medical charts of all children younger than 18 years, who died as inpatients while undergoing treatment at the pediatric oncology department between April and September 2016. Data were collected on demographics, diagnosis, treatments, survival, palliative care involvement, and symptoms at end of life.

RESULTS: There were 44 pediatric oncology patients who died in the hospital during the study period. The most frequent diagnoses were hematological malignancies (n = 29). Tumor-specific treatment was given to 38/44 (86%) patients in the last 30 days of life, and 13 patients in the last day of life or 1 day before. Of all deaths, 23/44 (52%) occurred within 30 days of admission to the pediatric ward and 34/44 (77%) within 90 days. Of the 44 patients, 25 (57%) were referred to palliative care. The median number of days between referral and death was 14 (0-78) days. Frequent symptoms documented were bleeding (11/44), dyspnea (10/44), pain (7/44), seizures (7/44), and delirium (5/44), with each patient having one or more of these symptoms. Only patients with a palliative care referral received opioid analgesics or benzodiazepines at the end of life.

CONCLUSIONS: This study highlights the demographics of suffering, death, and end-of-life care in children with cancer at a government tertiary cancer center in India.


BACKGROUND: As advances in neonatal intensive care increase the survival of extremely premature infants, the at-risk population for necrotizing enterocolitis (NEC) continues to rise. Although racial health disparities in preterm births have been well documented, large-scale studies exploring racial differences in NEC outcomes are lacking. Here, we conduct a study of racial health disparities in NEC using a nationally representative multicenter cohort.

STUDY DESIGN: Infants <1500 g birth weight and <30 weeks gestational age admitted in the first week after birth to neonatal intensive care units in the Pediatrix Medical group from 1997 to 2015 were included. Multivariable logistic regression was used to determine the adjusted odds ratio (AOR) of risk factors related to NEC and associated mortality.

RESULTS: Of the 126,089 (45% non-Hispanic White, 27% non-Hispanic Black, and 19% Hispanic) infants who met the inclusion criteria, 8796 (7%) developed NEC. On multivariable analysis, non-Hispanic Black and Hispanic infants had higher odds of developing NEC (AOR 1.31, 95% confidence interval (CI) [1.24-1.39], p < 0.001 and AOR 1.30 [1.21-1.39], p < 0.001, respectively). Among infants with NEC, mortality was higher in non-Hispanic Black and Hispanic infants compared to non-Hispanic White infants (AOR 1.35 [1.15-1.58], p < 0.001 and AOR 1.31 [1.09-1.56], p = 0.003, respectively).

CONCLUSION: Our study demonstrates that non-Hispanic Black and Hispanic infants are significantly more likely to be diagnosed with NEC. In addition, non-Hispanic Black and Hispanic infants have higher odds of death after NEC compared to non-Hispanic White infants. Further studies are necessary to investigate the etiology of these health disparities and to test interventions to improve these health outcomes.


BACKGROUND: For many childhood cancers, survival is lower among non-Hispanic blacks and Hispanics in comparison with non-Hispanic whites, and this may be attributed to underlying socioeconomic factors. However,
prior childhood cancer survival studies have not formally tested for mediation by socioeconomic status (SES). This study applied mediation methods to quantify the role of SES in racial/ethnic differences in childhood cancer survival.

METHODS: This study used population-based cancer survival data from the Surveillance, Epidemiology, and End Results 18 database for black, white, and Hispanic children who had been diagnosed at the ages of 0 to 19 years in 2000-2011 (n = 31,866). Black-white and Hispanic-white mortality hazard ratios and 95% confidence intervals, adjusted for age, sex, and stage at diagnosis, were estimated. The inverse odds weighting method was used to test for mediation by SES, which was measured with a validated census-tract composite index.

RESULTS: Whites had a significant survival advantage over blacks and Hispanics for several childhood cancers. SES significantly mediated the race/ethnicity-survival association for acute lymphoblastic leukemia, acute myeloid leukemia, neuroblastoma, and non-Hodgkin lymphoma; SES reduced the original association between race/ethnicity and survival by 44%, 28%, 49%, and 34%, respectively, for blacks versus whites and by 31%, 73%, 48%, and 28%, respectively, for Hispanics versus whites (log hazard ratio total effect - log hazard ratio direct effect)/log hazard ratio total effect).

CONCLUSIONS: SES significantly mediates racial/ethnic childhood cancer survival disparities for several cancers. However, the proportion of the total race/ethnicity-survival association explained by SES varies between black-white and Hispanic-white comparisons for some cancers, and this suggests that mediation by other factors differs across groups.


BACKGROUND: Cystic fibrosis (CF) is the most common inherited disease in Caucasians, affecting around 10,000 individuals in the UK today. Prognosis has improved considerably over recent decades with ongoing improvements in treatment and care. Providing up-to-date survival predictions is important for patients, clinicians and health services planning.

METHODS: Flexible parametric survival modelling of UK CF Registry data from 2011 to 2015, capturing 602 deaths in 10,428 individuals. Survival curves were estimated from birth; conditional on reaching older ages; and projected under different assumptions concerning future mortality trends, using baseline characteristics of sex, CFTR genotype (zero, one, two copies of F508del) and age at diagnosis.

FINDINGS: Male sex was associated with better survival, as was older age at diagnosis, but only in F508del non-homozygotes. Survival did not differ by genotype among individuals diagnosed at birth. Median survival ages at birth in F508del homozygotes were 46years (males) and 41years (females), and similar in non-homozygotes diagnosed at birth. F508del heterozygotes diagnosed aged 5 had median survival ages of 57 (males) and 51 (females). Conditional on survival to 30, median survival age rises to 52 (males) and 49 (females) in homozygotes. Mortality rates decreased annually by 2% during 2006-2015. Future improvements at this rate suggest median survival ages for F508del homozygous babies of 65 (males) and 56 (females).

INTERPRETATION: Over half of babies born today, and of individuals aged 30 and above today, can expect to survive into at least their fifth decade.

RESEARCH IN CONTEXT: Evidence before this study We searched PubMed with terms "(cystic fibrosis survival) and (projection OR model OR registry OR United Kingdom OR UK)" to identify relevant studies on survival estimates for individuals with cystic fibrosis (CF). We also considered the most recent annual report from the UK Cystic Fibrosis Registry (Cystic Fibrosis Trust, 2016), a review by Buzzetti and colleagues (2009), the chapter on Epidemiology of Cystic Fibrosis by MacNeill (2016), the study of MacKenzie and colleagues (2014), and references therein. There have been many studies of factors associated with survival in CF; most have focused on identifying risk factors, and only a few have presented estimated survival curves, which are the focus of this work. The most recent study of survival in the UK is by Dodge and colleagues (2007), who used data obtained from CF clinics and the national death register, and gave an estimate of survival for babies born in 2003. We found no previous studies that have obtained detailed information on survival using UK Cystic Fibrosis Registry data. Jackson and colleagues obtained survival estimates for the US and Ireland using registry data (Jackson et al., 2011). MacKenzie and colleagues used US Cystic Fibrosis Foundation Patient Registry data from 2000 to 2010 to project survival for children born and diagnosed with CF in 2010, accounting for sex, genotype and age at diagnosis (MacKenzie et al., 2014). Previous studies on estimated survival in CF have become out of date or have not accounted for the full range of patient characteristics available at birth. Few have presented conditional survival estimates (Dodge et al., 2007). Added value of this study This is the first study to yield detailed survival statistics using the UK Cystic Fibrosis Registry, which is one of the largest national CF registries outside of the US and has almost complete coverage of the UK CF
population. The primary goal was to leverage the long-term follow-up of the nearly complete UK CF population available in the Registry for the purposes of producing accurate, precise predictions in the modern era of CF care. Estimates are presented from birth and conditional on survival to older ages. These are the first conditional estimates in CF to also account for genotype, sex and age at diagnosis, which were each included in the modelling using a flexible approach. Projections are also provided under different scenarios based on downward trends in mortality rates. Our use of flexible parametric survival models is novel in this field, and our approach could be used to provide modern survival statistics for other chronic diseases and disorders. Implications of all the available evidence Our estimates of future survival in CF under a range of different scenarios are based on data on nearly all individuals living with the disease in the UK in recent times, reflective of a modern era of care, and are most appropriate for the families of babies being born in the present day with CF. Conditional estimates inform patients who have already reached an older age, and their clinicians. Over half of babies born today, and of individuals aged 30 years and above alive today, can expect to survive into their fifth decade. Insights based on our survival projections can be used to inform future needs in CF health care provision.


OBJECTIVE: Pediatric intramedullary spinal cord ependymomas represent a rare central nervous system neoplasm with few available data regarding incidence and outcomes. To this end, large population-based studies are needed to assess the epidemiology and survival risk factors associated with these tumors in the hope of better understanding these tumors as well as improving outcomes. This retrospective study was undertaken to explore factors that may influence survival in pediatric patients with intramedullary spinal cord ependymomas.

METHODS: Using the SEER (Surveillance Epidemiology and End Results) database, a prospective cancer registry, we retrospectively assessed survival in histologically confirmed spinal ependymomas in patients 17 years of age and younger. Survival was described with Kaplan-Meier curves, and a multivariate regression analysis was used to assess the association of several variables with survival, controlling for confounding variables. RESULTS: Invasive tumor extension (P < 0.001) was associated with decreased survival, whereas gross total resection (P = 0.028) correlated with better rates of survival. Age, gender, tumor size, tumor extension, the use and sequence of radiation therapy, or use of chemotherapy were not found to have a statistically significant association with survival outcomes.

CONCLUSIONS: Invasive ependymomas occurring in the spine have a worse prognosis, whereas higher tumor grades do not clearly show worse rates of survival. Early diagnosis and surgery seem to be associated with improved survival and outcomes, whereas radiation therapy and chemotherapy have an unclear role.


Langerhans cell histiocytosis (LCH) is an inflammatory neoplasia of myeloid precursor cells driven by mutations in the mitogen-activated protein kinase pathway. When disease involves the skin, LCH most commonly presents as a seborrheic dermatitis or eczematous eruption on the scalp and trunk. Evaluation for involvement of other organ systems is essential, because 9 of 10 patients presenting with cutaneous disease also have multisystem involvement. Clinical manifestations range from isolated disease with spontaneous resolution to life-threatening multisystem disease. Prognosis depends on involvement of risk organs (liver, spleen, and bone marrow) at diagnosis, particularly on presence of organ dysfunction, and response to initial therapy. Systemic treatment incorporating steroids and cytostatic drugs for at least one year has improved prognosis of multisystem LCH and represents the current standard of care.


Background: The mortality burden in children aged 5-14 years in the WHO European Region has not been comprehensively studied. We assessed the distribution and trends of the main causes of death among children aged 5-9 years and 10-14 years from 1990 to 2016, for 51 countries in the WHO European Region.
Methods: We used data from vital registration systems, cancer registries, and police records from 1980 to 2016 to estimate cause-specific mortality using the Cause of Death Ensemble model.

Findings: For children aged 5-9 years, all-cause mortality rates (per 100 000 population) were estimated to be 46.3 (95% uncertainty interval [UI] 45.1-47.5) in 1990 and 19.5 (18.1-20.9) in 2016, reflecting a 58.0% (54.7-61.1) decline. For children aged 10-14 years, all-cause mortality rates (per 100 000 population) were 37.9 (37.3-38.6) in 1990 and 20.1 (18.8-21.3) in 2016, reflecting a 47.1% (43.8-50.4) decline. In 2016, we estimated 10 740 deaths (95% UI 9970-11 542) in children aged 5-9 years and 10 279 deaths (9652-10 897) in those aged 10-14 years in the WHO European Region. Injuries (road injuries, drowning, and other injuries) caused 4163 deaths (3820-4540; 38.7% of total deaths) in children aged 5-9 years and 4468 deaths (4162-4812; 43.5% of total) in those aged 10-14 years in 2016. Neoplasms caused 2161 deaths (1872-2406; 20.1% of total deaths) in children aged 5-9 years and 1943 deaths (1749-2101; 18.9% of total deaths) in those aged 10-14 years in 2016. Notable differences existed in cause-specific mortality rates between the European subregions, from a two-times difference for leukaemia to a 20-times difference for lower respiratory infections between the Commonwealth of Independent States (CIS) and EU15 (the 15 member states that had joined the European Union before May, 2004). Interpretation: Marked progress has been made in reducing the mortality burden in children aged 5-14 years over the past 26 years in the WHO European Region. More deaths could be prevented, especially in CIS countries, through intervention and prevention efforts focusing on the leading causes of death, which are road injuries, drowning, and lower respiratory infections. The findings of our study could be used as a baseline to assess the effect of implementation of programmes and policies on child mortality burden. Funding: WHO and Bill & Melinda Gates Foundation.


BACKGROUND: Late-infantile neuronal ceroid lipofuscinosis type 2 (CLN2) disease, characterised by rapid psychomotor decline and epilepsy, is caused by deficiency of the lysosomal enzyme tripeptidyl peptidase 1. We aimed to analyse the characteristics and rate of progression of CLN2 disease in an international cohort of patients.

METHODS: We did an observational cohort study using data from two independent, international datasets of patients with untreated genotypically confirmed CLN2 disease: the DEM-CHILD dataset (n=74) and the Weill Cornell Medical College (WCMC) dataset (n=66). Both datasets included quantitative rating assessments with disease-specific clinical domain scores, and disease course was measured longitudinally in 67 patients in the DEM-CHILD cohort. We analysed these data to determine age of disease onset and diagnosis, as well as disease progression—measured by the rate of decline in motor and language summary scores (on a scale of 0-6 points)—and time from first symptom to death.

FINDINGS: In the combined DEM-CHILD and WCMC dataset, median age was 35.0 months (IQR 24.0-38.5) at first clinical symptom, 37.0 months (IQR 35.0 -42.0) at first seizure, and 54.0 months (IQR 47.5-60.0) at diagnosis. Of 74 patients in the DEM-CHILD dataset, the most common first symptoms of disease were seizures (52 [70%]), language difficulty (42 [57%]), motor difficulty (30 [41%]), behavioural abnormality (12 [16%]), and dementia (seven [9%]). Among the 41 patients in the DEM-CHILD dataset for whom longitudinal assessments spanning the entire disease course were available, a rapid annual decline of 1.81 score units (95% CI 1.50-2.12) was seen in motor-language summary scores from normal (score of 6) to no function (score of 0), which occurred over approximately 30 months. Among 53 patients in the DEM-CHILD cohort with available data, the median time between onset of first disease symptom and death was 7.8 years (SE 0.9) years.

INTERPRETATION: In view of its natural history, late-infantile CLN2 disease should be considered in young children with delayed language acquisition and new onset of seizures. CLN2 disease has a largely predictable time course with regard to the loss of language and motor function, and these data might serve as historical controls for the assessment of current and future therapies.

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In Germany, every child with a life-limiting condition suffering from symptoms that cannot sufficiently be controlled is eligible by law for specialized pediatric palliative home care (SPPHC). It is the aim of this study to describe the demographic and clinical characteristics of children referred to SPPHC and to compare patients with cancer and non-cancer conditions. The prospective multicenter study includes data on 75 children (median age 7.7 years, 50.7% male). The majority had non-cancer conditions (72%). The most common symptoms were cognitive impairment, somatic pain, impairment in communication or swallowing difficulties. Swallowing difficulties, seizures, and spasticity occurred significantly more often in non-cancer patients ($p < 0.01$). Cancer patients received antiemetics significantly more often (permanent and on demand) than non-cancer patients ($p < 0.01$). Significantly more non-cancer patients had some type of feeding tube (57.3%) or received oxygen (33.3%) ($p < 0.01$). Central venous catheters had been fitted in 20% of the patients, mostly in cancer patients ($p < 0.001$). Tracheostomy tubes (9.3%) or ventilation (14.7%) were only used in non-cancer patients. In conclusion, patients referred to SPPHC are a diverse cohort with complex conditions including a large range of neurologically originating symptoms. The care of pediatric palliative care patients with cancer is different to the care of non-cancer patients.


OBJECTIVES: To evaluate mortality in adolescents and young adult patients with chronic diseases followed in a Latin American tertiary hospital.

METHODS: A cross-sectional retrospective study was performed in a tertiary/academic hospital in the state of Sao Paulo, Brazil. Death occurred in 529/2850 (18.5%) adolescents and young adult patients with chronic diseases, and 25/529 (4.7%) were excluded due to incomplete medical charts. Therefore, 504 deaths were evaluated.

RESULTS: Deaths occurred in 316/504 (63%) of early adolescent patients and in 188/504 (37%) of late adolescent/young adult patients. Further comparisons between early adolescents ($n=316$) and late adolescent/young adult patients ($n=188$) with pediatric chronic diseases at the last hospitalization showed that the median disease duration (22.0 [0-173] vs. 43.0 [0-227] months, $p<0.001$) was significantly lower in early adolescents vs. late adolescent/young adult patients. The median number of previous hospitalizations was significantly lower in the former group (4.0 [1-45] vs. 6.0 [1-52], $p<0.001$), whereas the last hospitalization in intensive care unit was significantly higher (60% vs. 47%, $p=0.003$). Regarding supportive measures, palliative care was significantly lower in the younger group compared to the older group (33% vs. 43%, $p=0.02$). The frequencies of renal replacement therapy (22% vs. 13%, $p=0.02$), vasoactive agents (65% vs. 54%, $p=0.01$), and transfusion of blood products (75% vs. 66%, $p=0.03$) were significantly higher in the younger group. The five most important etiologies of pediatric chronic diseases were: neoplasias (54.2%), hepatic diseases/transplantation (10%), human immunodeficiency virus (5.9%), and childhood-onset systemic lupus erythematosus and juvenile idiopathic arthritis (4.9%). Autopsy was performed in 58/504 (11%), and discordance between clinical and postmortem diagnoses was evidenced in 24/58 (41.3%).

CONCLUSIONS: Almost 20% of deaths occurred in adolescents and young adults with distinct supportive care and severe disease patterns. Discordance between clinical diagnosis and autopsy was frequently observed.


OBJECTIVE: To provide a systematic evaluation of the broad clinical variability in Friedreich ataxia (FRDA), a multisystem disorder presenting mainly with afferent ataxia but also a complex phenotype of nonataxia symptoms.

METHODS: From the large database of the European Friedreich's Ataxia Consortium for Translational Studies, 650 patients with genetically confirmed FRDA were included. Detailed data of medical history documentation, questionnaires, and reports on clinical features were analyzed to provide in-depth description of the clinical profile and frequency rates of phenotypical features with a focus on differences between typical-onset and late-onset...
FRDA. Logistic regression modeling was used to identify predictors for the presence of the most common clinical features.

RESULTS: The most frequent clinical features beyond afferent ataxia were abnormal eye movements (90.5%), scoliosis (73.5%), deformities of the feet (58.8%), urinary dysfunction (42.8%), cardiomyopathy and cardiac hypertrophy (40.3%), followed by decreased visual acuity (36.8%); less frequent features were, among others, depression (14.1%) and diabetes (7.1%). Most of these features were more common in the typical-onset group compared to the late-onset group. Logistic regression models for the presence of these symptoms demonstrated the predictive value of GAA repeat length on the shorter allele and age at onset, but also severity of ataxia signs, sex, and presence of neonatal problems.

CONCLUSIONS: This joint European effort demonstrates the multisystem nature of this neurodegenerative disease encompassing most the central nervous, neuromuscular, cardiologic, and sensory systems. A distinct and deeper knowledge of this rare and chronic disease is highly relevant for clinical practice and designs of clinical trials.


BACKGROUND/AIMS: Duchenne Muscular Dystrophy (DMD) has childhood onset, primarily affects males, and is usually fatal before the age of 40 years. Previous studies have indicated that this X-linked condition is more prevalent in whites than in blacks, but those were based on active surveillance, and limited to smaller populations and younger ages.

METHODS: US death data were used to calculate mortality rates by race and ethnicity, with MD as either the underlying or multiple cause of death (MCD). Poisson approximation was used for confidence intervals; chi-square was used to compare rates.

RESULTS: From 2006 to 2015, there were 3,256 deaths in males <40 years with MD as MCD, and 71% of these were aged 15-29 years. For whites, the average annual death rate was 0.43/100,000, which was significantly higher (p < 0.0001) that that of blacks (0.28), American Indian/Alaska Natives (0.20), and Asian/Pacific Islanders (0.21). The rate for non-Hispanic whites (0.46) was significantly higher (p < 0.0001) than the rates for Hispanic whites (0.31), Hispanic blacks (0.07), and non-Hispanic blacks (0.29). CONCLUSION: Since DMD is the primary cause of deaths in young males with MD, mortality rates are a reasonable proxy for the relative difference in racial prevalence. It appears that DMD is significantly more common in white males than in males of other races.


Sudden unexpected death in epilepsy (SUDEP) in children, although rare, needs critical attention given the tragic nature and devastating consequences for families and caregivers. True incidence is unknown and risk factors are not completely understood, more so in children compared with adults. A focused narrative review of available studies on paediatric SUDEP was undertaken to comprehend its risk factors and to develop strategies to recognise and where possible modify SUDEP risk and ultimately reduce incidence. We reviewed 16 population-based studies from various settings. We found overlapping risk factors from different studies. The prime risk factor is uncontrolled seizures. This review supports the view that children entering adolescence with optimal seizure control could be a key aspect in reducing adult mortality related to SUDEP. Ideally, clinicians would want to be able to predict prospective, individualised SUDEP risk, which is challenging due to a myriad of risk factors and an inherent non-homogeneous paediatric epilepsy population. Nevertheless, an adequate evidence base exists as evidenced by this review to support information giving and communication to support young people with epilepsy and their families in being active partners in recognising and reducing their SUDEP risk. More work particularly in the form of prospective studies and registries are needed to further clarify true incidence which may have been previously underestimated and to update risk factors.


Our objectives were to determine whether procedural pain and glucose exposure are associated with altered structural and functional brain development differently in preterm males and females, and neurodevelopment at 18-month corrected age. Fifty-one very preterm neonates (22 males; median [interquartile range] gestational age 27.6 [2.0] weeks) underwent 3 serial scans including T1-weighted and resting-state functional magnetic resonance imaging (MRI) at median postmenstrual weeks: 29.4, 31.9, and 41.1. Thalamus, basal ganglia, and total brain volumes were segmented. Functional resting-state MRI data were extracted from the independent-components maps. Pain was operationalized as the total number of neonatal intensive care unit-administered invasive procedures. Neurodevelopmental outcomes at 18-month corrected age were assessed with the Bayley Scales of Infant Development, second edition. Generalized estimating equations assessed the association of pain and glucose exposure with brain structural and functional development. More invasive procedures were independently associated with slower growth of thalamic (P < 0.001), basal ganglia (P = 0.028), and total brain volumes (P = 0.001), particularly in females. Similar relationships were observed between glucose exposure and brain volumes. Functional connectivity between thalamus and sensorimotor cortices was negatively associated with number of invasive procedures. Greater procedural pain and higher glucose exposure were related to poorer neurodevelopmental outcomes. These findings suggest that structural and functional brain development is vulnerable to procedural pain. Glucose used for analgesia does not appear to mitigate the adverse impact of pain on brain development. The vulnerability of brain development in females towards early pain is distinct from other neonatal morbidities. The link between pain and glucose with neurodevelopment suggests that these factors have long-lasting impact.


Background Bacterial infections account for a significant proportion of neonatal and infant mortality globally. We aimed to identify predictors of death in infants with probable serious bacterial infection (PSBI) defined as signs/symptoms of possible serious bacterial infection along with baseline C-reactive protein (CRP) >/=12 mg/l. Methods We did a secondary analysis using the data collected from 700 infants with PSBI who participated in a randomized controlled trial in India in which zinc or placebo was given in addition to the standard antibiotics. Logistic regression was used to estimate the associations between relevant variables and death within 21 days. Results Those infants who were fed cow's milk or formula before the illness episode had 3.7-fold (95% confidence interval (CI) 1.5-9.3) and 5.3-fold (95% CI 2.0-13.6) higher odds of death, respectively. Lethargy (odds ratio (OR) 2.4, 95% CI 1.1-5.4) and CRP (OR 1.9, 95% CI 1.1-3.3) were also independent predictors of death. In the model including only clinical features, female gender (OR 2.25, 95% CI 1.0-5.0), abdominal distention (3.7, 95% CI 1.1-12.3), and bulging fontanelle (5.8, 95% CI 1.1-30.5) were also independent predictors for death. Conclusion Formula or cow milk feeding prior to the illness, lethargy at the time of presentation, and high serum CRP levels predicted death in infants with PSBI.

https://www.ncbi.nlm.nih.gov/pubmed/29166376


OBJECTIVES: To estimate the magnitude of mortality and loss to follow-up and describe predictors of mortality among HIV-infected children in Guinea-Bissau. METHODS: Retrospective follow-up study among HIV-infected children under 15 years of age at the largest HIV-clinic in Guinea-Bissau from 2006-2016. A multivariate Cox proportional hazards model was used to identify predictors of mortality. RESULTS: Of 525 children were included in the analysis: 371 (70.7%) with HIV-1, 17 (3.2%) with HIV-2, 25 (4.8%) with HIV-1/2, and 112 (21.3%) with HIV of unknown type. At diagnosis, the median age was 3.5 years, 44.7% met the WHO criteria for severe immunodeficiency by age based on CD4 cell count, and 59.4% were underweight. The median follow-up time was 6 months. Despite the availability of antiretroviral treatment, the mortality rate was 10.4 deaths per 100 person-years of follow-up. Within the first year of follow-up, 11.0% died, 3.1% were transferred and 38.8% were lost to follow-up, leaving 47.1% in follow-up. Severe immunodeficiency (adjusted hazard ratio (aHR) = 2.52, 95% CI: 1.22-5.21) and underweight (aHR = 3.14, 95% CI: 1.40-7.02) were independent predictors of mortality. CONCLUSIONS: This study reveals a high rate of early mortality and loss to follow-up among HIV-infected children in Guinea-Bissau. Initiatives to improve patient retention are urgently needed.


OBJECTIVE: The objective of this study was to assess whether in-hospital morbidity or mortality differed by race/ethnicity for preterm neonates admitted to the neonatal intensive care unit (NICU).

STUDY DESIGN: In a retrospective cohort study, preterm infants, < 37 weeks, were admitted to the NICU from 1994 to 2009. Exclusions included structural anomalies and aneuploidy. Primary outcome was in-hospital mortality (IHM). Secondary outcomes were respiratory distress syndrome (RDS), interventricular hemorrhage (IVH), necrotizing enterocolitis (NEC), and retinopathy of prematurity (ROP). Sub-analysis of very preterm (VPT) infants, < 28 weeks, was performed. Five racial/ethnic groups (REGs) were compared: White, Black, Hispanic, Asian, and Mixed. Associations were modeled by logistic regression. White neonates (WNs) were the referent group. Unadjusted and adjusted odds ratios and 95% confidence intervals for remaining REGs were reported. p value was significant at 5% for overall tests and at Bonferroni-corrected level < 0.0125 for between-race comparisons with WNs.

RESULTS: Four thousand nine hundred fifty-five preterm neonates were identified; 153 were excluded leaving 4802 for analysis. After controlling covariates that were chosen a priori, there was no difference across REGs for IHM (all between-race comparison p values > 0.0125). There was a significant difference in RDS among Black neonates (BNs) (aOR 0.57, 95% CI 0.45-0.73; p < 0.001) and Hispanic neonates (HNs) (aOR 0.67, 95% CI 0.50-0.89; p = 0.005) compared to WNs. The risk of ROP was significantly different across REGs with HNs having a 70% increase in ROP (aOR 1.70, 95% CI 1.15-2.49; p = 0.008) and Mixed neonates (MNs) experiencing a 55% reduction (aOR 0.45, 95% CI 0.29-0.68; p < 0.001) compared to WNs. There was no difference in IVH or NEC across REGs (all p values > 0.0125). In the VPT cohort sub-analysis, BNs experienced a significant 59% reduction in IHM compared to WNs (BNs aOR 0.41, 95% CI 0.22-0.73; p = 0.003). MNs experienced a 46% reduction in ROP compared to WNs (aOR 0.54, 95% CI 0.35-0.81; p = 0.004). There was no difference in RDS, IVH, or NEC in very preterm infants across REGs (all between comparison p values > 0.0125). CONCLUSION: In preterm neonates, in-hospital mortality does not significantly differ across racial and ethnic groups. However, in very preterm infants, in-hospital mortality for Black neonates is improved. There are morbidity differences (RDS, ROP) seen among racial/ethnic groups.


Background: Allergic disease is suspected to play a role in the development of childhood acute lymphoblastic leukemia (ALL). Studies conducted over the last several decades have yielded mixed results.

Methods: We examined the association between allergy, a common immune-mediated disorder, and ALL in the California Childhood Leukemia Study (CCLS), a case-control study of 977 children diagnosed with ALL and 1,037 matched controls (1995-2015). History of allergies in the first year of life was obtained from interviews, mainly reported by mothers. Logistic regression analyses were conducted to estimate ORs and 95% confidence intervals (CIs), controlling for birth order, daycare attendance, and mode of delivery. In addition, we conducted meta-analyses with data from the CCLS and 12 published studies and employed a new method to estimate between-study heterogeneity (R_b).

Results: Overall, no associations were observed between childhood ALL risk and specific allergy phenotypes or any allergy, as a group. However, having any allergy was associated with an increased risk of ALL among the youngest study participants. In the meta-analysis random-effects models, reduced odds of ALL were associated with hayfever (metaOR = 0.65; 95% CI, 0.47-0.90); however, restricting the analysis to studies that used medical records for assessment of allergy or recently published studies led to null or attenuated results.

Conclusions: Overall, our findings do not support a clear association between allergy and childhood ALL. Impact: The degree to which epidemiologic studies can inform the relationship between allergies and risk of childhood ALL is limited by R_b. Cancer Epidemiol Biomarkers Prev; 27(10); 1142-50. (c)2018 AACR.


Decisions about whether to withdraw or withhold life-sustaining medical treatment from children give rise to complex and value-laden judgments. While recourse to the courts is uncommon, judicial decisions provide an important source of guidance for the children (where they can participate), families and health and medical professionals involved in these decisions. Yet, there has been remarkably little consideration of the Australian jurisprudence on this issue. This article addresses that gap by undertaking the first comprehensive analysis of all publicly available Australian cases that consider whether or not it is in a child’s best interests to receive life-sustaining treatment. A
total of 25 cases were located and the judicial consideration of best interests was thematically analysed. Key
considerations (to varying degrees) when assessing best interests included the likelihood of treatment curing or
improving the child’s health, medical views about diagnosis, prognosis and treatment and the child’s and parents’
views and wishes. The article concludes that the law requires greater certainty and transparency in decision-making.
Given the significance of these cases, judgments should describe the factors that the court considers relevant and
important, and those that are less influential, as well as the weight ascribed to those various factors and the
reasoning that underpins an assessment that treatment is or is not in a child’s best interests.


of Mortality 3 and Paediatric Logistic Organ Dysfunction 2 Scores in Critically Ill Children." Ann Acad Med

INTRODUCTION: The Paediatric Index of Mortality 3 (PIM 3) and Paediatric Logistic Organ Dysfunction 2 (PELOD 2)
scores were recently revised. We aimed to assess the performance of these scores in a contemporary cohort of
critically ill children.

MATERIALS AND METHODS: This is a single-centre prospective study conducted in a multidisciplinary paediatric
intensive care unit (PICU). Consecutive PICU admissions over 1 year were included and admission PIM 3 and
PELOD 2 scores were calculated. The performance of each of the scores was evaluated by calculating the area
under the curve (AUC) of the receiver operating characteristic (ROC) and the Hosmer-Lemeshow goodness-of-fit test
for the outcome of PICU mortality.

RESULTS: A total of 570 patient admissions were eligible for this study. The median age of patients was 3.1
(interquartile range [IQR]: 0.4, 8.9 years). Overall median PIM 3 and PELOD 2 scores were 1.2 (IQR: 0.4, 3.2) % and
4 (IQR: 2, 7), respectively. The overall mortality rate was 35/570 (6.1%). The PIM 3 and PELOD 2 scores had good
discrimination for mortality (AUCs 0.88 [95% confidence interval (CI) 0.85, 0.91] and 0.86 [95% CI 0.83, 0.89],
respectively). Goodness-of-fit was satisfactory for both scores. Higher PIM 3 and PELOD 2 scores were also
associated with decreasing ventilator and PICU-free days.

CONCLUSION: PIM 3 and PELOD 2 scores are robust severity of illness scores that are generalisable to a
contemporary cohort of critically ill children in Singapore.

Outcomes and Instruments


BACKGROUND: Early discharge of very low birth weight infant (VLBW) in low resource settings is inevitable but to minimize mortality of these infants after discharge we need to identify the death attributes.

METHOD: A prospective cohort was conducted among 190 VLBW infants discharged from Mulago Special Care Baby Unit (SCBU) with discharge weight of < 1500 g over an 8 months period. These infants were followed up with the aims of determining the proportion dead 3 months after discharge, identifying factors associated and possible causes of death. Relevant data were captured, transferred in to STATA and imported to SPSS 12.0.1 for analysis. To determine factors associated with mortality bi-variable and multivariable regressions were conducted. A p-value of < 0.05 was considered significant and 95% confidence interval was used.

RESULTS: Of the enrolled infants 164 (86.3%) completed follow up. The median gestational age of study participants was 32 weeks (range 26-35 weeks), the mean discharge weight was 1119 g (range 760-1470 g), and 59.8% were small for gestational age (SGA). During follow up 32 (19.5%) infants died. Infants discharged with weight of < 1200 g accounted for 81.2% of the deaths. Majority of the deaths (68.7%) occurred in the first month after discharge. Factors independently associated with mortality were discharge weight < 1000 g (OR 3.10, p 0.015) and not being SGA (OR 3.54, p 0.019). The main causes of death were presumed sepsis 50.0% and suspected cot death (25.0%).

CONCLUSION: Mortality after hospital discharge among VLBW infants is high. Discharge at weight < 1200 g may not be a safe practice. Measures to prevent sepsis and suspected cot death should be addressed prior to considering early discharge of these infants.


INTRODUCTION: Neuropathic pain (NP) can cause substantial suffering and, therefore, it must be diagnosed and treated promptly. Diagnosis of NP can be difficult and if made by an expert pain physician is considered the gold standard, however where expert help may not be easily available, screening tools for NP can be used. The painDETECT questionnaire (PD-Q) is a simple screening tool and has been widely used in several languages. We developed an Arabic version of PD-Q and tested its validity and reliability.

METHODS: The original PD-Q was translated into the Arabic language by a team of experts. The translated version of the PD-Q was administered to the study population, which included patients having moderate to severe pain for at least three months. Reliability of the Arabic version was evaluated by an intra-class-correlation coefficient (ICC) between pre- and post-measures and Cronbach’s alpha values. Validity was measured by receiver operating characteristic (ROC) curve. Expert pain physician diagnosis was considered as the gold standard for comparing the diagnostic accuracy.

RESULTS: A total of 375 patients were included in the study, of which 153 (40.8%) patients were diagnosed with NP and 222 [59.2%] patients had nociceptive pain. The ICC between pre- and post-PD-Q scale total scores for the overall sample, NP group, and NocP group was 0.970 (95% CI, 0.964-0.976), 0.963 (95% CI, 0.949-0.973), and 0.962 (95% CI, 0.951-0.971), respectively. The Cronbach’s alpha values for the post-assessment measures in the overall sample, NP group, and nociceptive pain group, were 0.764, 0.684, and 0.746, respectively. The area under the ROC curve was 0.775 (95% CI, 0.725-0.825) for the PD-Q total score. CONCLUSION: The Arabic version of the PD-Q showed good reliability and validity in the detection of NP component in patients with chronic pain.


BACKGROUND: Cancer stage at diagnosis is crucial for assessing global efforts to increase awareness of childhood cancer and improve outcomes. However, consistent information on childhood cancer stage is absent from population cancer registries worldwide. The Toronto Childhood Cancer Stage Guidelines, compiled through an international consensus process, were designed to provide a standard framework for collection of information on stage at diagnosis of childhood cancers. We aimed to assess the feasibility of implementing the Toronto Guidelines within a national population cancer registry.

METHODS: We did a population-based registry study using data from the Australian Childhood Cancer Registry and included data from children aged 0-14 years diagnosed between Jan 1, 2006, and Dec 31, 2010 with one of 16 childhood cancers listed in the Toronto Guidelines (acute lymphoblastic leukaemia, acute myeloid leukaemia, Hodgkin’s lymphoma, non-Hodgkin lymphoma, neuroblastoma, Wilms’ tumour, rhabdomyosarcoma, non-rhabdomyosarcoma soft tissue sarcoma, osteosarcoma, Ewing’s sarcoma, retinoblastoma, hepatoblastoma, testicular cancer, ovarian cancer, medulloblastoma, and ependymoma). We extracted data from medical records, and assigned stage according to the Tier 1 criteria (basic) and Tier 2 criteria (more detailed, requiring data from cytology, imaging, and other diagnostic tests, where available) using computer algorithms derived from the Toronto Guidelines. Additionally, expert reviewers independently assigned Tier 2 stage to a random subsample of 160 cases (ten per malignancy type). Feasibility of the guidelines was assessed on the percentage of cases that could be staged, agreement between stage assigned by the algorithms and the expert reviewers, and the mean time (min) taken to collect the required data.

FINDINGS: We obtained data for 1412 eligible children. Stage could be assigned according to Tier 2 criteria for 1318 (93%) cases, ranging from 48 (84%) of 57 cases of non-rhabdomyosarcoma soft tissue sarcoma to 46 (100%) cases of hepatoblastoma. According to Tier 1 criteria, stage could be assigned for 1329 (94%) cases, ranging from 131 (87%) of 151 cases of acute myeloid leukaemia to 46 (100%) cases of hepatoblastoma. By contrast, stage at diagnosis was recorded by the treating physician for 555 (39%) of the 1412 cases. The computer algorithm assigned the same stage as did one or more independent expert reviewers in 155 (97%) of the 160 cases assessed. The mean time taken to review medical records and extract the required data was 18.0 min (SD 9.5 per case).

INTERPRETATION: The Toronto Guidelines provide a highly functional framework that can be used to assign cancer stage at diagnosis using data routinely available in medical records for most childhood cancers. Data on staging have the potential to inform interventions targeting improved diagnosis and survival. FUNDING: Cancer Australia.


With increasing numbers of children being diagnosed with neurodevelopmental disorders (NDDs) attention has been drawn to these children’s physical health. We aimed to identify the prevalence of defined physical problems (epilepsy, migraine, asthma, cancer, diabetes, psoriasis, lactose intolerance, celiac disease, diarrhea, constipation, daytime enuresis, encopresis) in a nationwide population of 9- and 12-year-old twins subdivided into those with and without indications of NDDs. Parents of 28,058 twins participated in a well-validated telephone interview regarding their children’s mental health and answered questions about their physical problems. The results indicate a high rate of physical problems in children with NDDs, particularly in those with indications of the presence of combinations of several NDDs.


To measure the impact of a novel interactive inpatient pediatric pain management solution integrating our hospital’s electronic health record system, the nurse communication phones, and the pharmacy dispensing system, we assessed parent and nurse perspectives on the tool’s potential value, benefits, and challenges. A mixed-methods approach with survey instruments containing closed-ended and open-ended questions was administered to 30 parents and 59 nurses (66% and 23% response rate respectively). Overall, parents were more satisfied with the interactive technology experience (90%) compared to nurses (50%) with both indicating timely reassessments of pain being the most valuable feature. Qualitative analysis of nurses’ responses yielded 6 themes for technology benefits and 12 for challenges. While patient-interactive technology solutions appear well-received particularly by parent end-users for pediatric hospital pain management, nurse training and interface improvements may result in higher efficacy, ultimately empowering parents/patients, promoting patient engagement and satisfaction.

OBJECTIVES: Implement a novel pain-management interface that is used to bring real-time, patient-reported pain assessments to the inpatient television and evaluate the impact of implementation on the pain-management clinical workflow, patient engagement, and nursing pain reassessments.

METHODS: We developed a pain-management tool interfacing 4 stand-alone technologies: a television-based, interactive patient care system; electronic health record system; nursing call system; and pharmacy inventory-management system. The workflow is triggered when pain medications are dispensed by sending an automatic pain assessment rating question via the patient’s television at a predefined time. To measure the effects of implementation, we calculated patient and/or parent use rates and pain reassessment timely documentation rates. Data were extracted from the electronic health record for a period of 22 months and covered pre- and postimplementation.

RESULTS: A total of 56,931 patient records were identified during the study period, representing 2,447 unique patients. In total, 608 parents and/or patients reported their pain through the tool. Use rates were 6.5% for responding to the pain rating prompt and 13.3% for the follow-up prompt, in which additional nonpharmacologic strategies to eliminate pain were offered. A modest increase was found in the mean timely documentation rates on the basis of nursing documentation standards (26.1% vs 32.8%, a percentage increase of 25.7%; P < .001) along with decreased median time to pain reassessment documentation (29 minutes versus 25 minutes, a percentage decrease of 13.8%; P < .001).

CONCLUSIONS: With this novel tool, we offer a potentially scalable approach in supporting the pain-management clinical workflow, integration of technologies, and promoting of patient and/or parent engagement in the inpatient setting.


BACKGROUND: Gentle ventilation with optimal oxygenation is integral to prevention of chronic lung disease in the extremely low birth-weight (ELBW) infant. Various types of noninvasive ventilation are used in neonatal intensive care units worldwide. Bubble continuous positive airway pressure (BCPAP) has been in use in newborn intensive care since 1975.

PURPOSE: To synthesize the current evidence on the use of BCPAP in the ELBW infant and its relationship to outcomes, particularly morbidity and mortality. METHODS/SEARCH STRATEGIES: A literature review was completed using PubMed, EMBASE, CINAHL, and Cochrane with a focus on BCPAP use in the ELBW population.

FINDINGS/RESULTS: No study found was exclusive to the ELBW population. All studies ranged from ELBW to full-term neonates. Studies supported the use of BCPAP in the ELBW, demonstrating decreased incidence of chronic lung disease and barotrauma through the use of oscillation and permissive hypercapnia.

IMPLICATIONS FOR PRACTICE: Literature supports the use of nasal bubble CPAP in the ELBW population. Barriers such as septal erosion, pneumothorax, inconsistent pressures, and air in the abdomen were identified and management recommendations were provided.

IMPLICATIONS FOR RESEARCH: Studies are needed comparing outcomes of nasal bubble CPAP use with other forms of CPAP in the ELBW infant, comparison of prongs to mask for nasal bubble CPAP, and comparing interventions to recommend optimal care bundles to prevent nasal septum injuries.


BACKGROUND: Sickle cell disease is a common inherited hemoglobinopathy and is associated with high morbidity and mortality. Vasculo-occlusive crises commonly occur in individuals with SCD that results in high morbidity due to end-organ ischemia and infarction. These include splenic infarction, pulmonary involvement, acute chest syndrome, and orbital compression syndrome. Ocular manifestations of SCD include anterior segment ischemia, secondary
Primary brain tumors are a leading cause of cancer-related morbidity and mortality in children. Glioblastoma (GBM) is a high-grade astrocytoma that occurs in both children and adults and is associated with a poor prognosis. Despite extensive study in recent years, the clinical management of these tumors has remained largely unchanged, consisting of surgical resection, conventional chemotherapy, and radiotherapy. Although the etiology and genomic drivers in GBM are diverse, constitutional mismatch repair-deficiency (CMMRD) syndrome is a rare, recessively inherited disease with a predisposition to gliomagenesis. CMMRD results from biallelic mutations in one of the mismatch repair genes including mutL homolog 1 (MLH1), mutS homolog 2 (MSH2), mutS homolog 6 (MSH6), and post-meiotic segregation increased 2 (PMS2). In this report, we present the case of a 5-year-old female with GBM and CMMRD due to an MSH6 homozygous c.1883G>A mutation consistent with CMMRD. Given her CMMRD status, she was treated with nivolumab (3 mg/kg every 2 weeks for 36 weeks) and showed a 60% reduction in tumor size, improved clinical symptoms, and an ongoing durable response lasting 10 months to date. Our study highlights a durable response to the ICPI nivolumab in a pediatric patient with recurrent/refractory CMMRD-associated GBM. We show that incorporating genomic and/or molecular testing for CMMRD into routine pediatric oncology clinical care can identify a subset of patients likely to benefit from ICPI. KEY POINTS: Constitutional mismatch repair-deficiency (CMMRD) syndrome, alternatively known as biallelic mismatch repair deficiency syndrome, occurs in subset of pediatric cancer patients, including those with primary brain tumors. Patients from Arab and other developing countries are predicted to have higher incidence of CMMRD due to high prevalence of consanguinity. Integration of molecular and/or genomic testing into routine clinical care for pediatric cancer patients is important to identify patients with CMMRD syndrome. Patients with CMMRD-associated cancers may show increased responsiveness to immune checkpoint inhibitors. To the authors’ knowledge, this is the first report in the Arab world of a durable response to immune checkpoint inhibitors in a pediatric glioblastoma patient.
muscular dystrophy (DMD) alters P2RX7 signaling in both muscle and inflammatory cells and inhibition of this receptor resulted in a significant attenuation of muscle and non-muscle symptoms in DMD (mdx) mouse model. As P2RX7 is an attractive target in a range of human diseases, specific antagonists have been developed. Yet, these will require lengthy safety testing in the pediatric population of Duchenne muscular dystrophy (DMD) patients. In contrast, Nucleoside Reverse Transcriptase Inhibitors (NRTIs) can act as P2RX7 antagonists and are drugs with an established safety record, including in children. We demonstrate here that AZT (Zidovudine) inhibits P2RX7 functions acting via the same allosteric site as other antagonists. Moreover, short-term AZT treatment at the peak of disease in DMD (mdx) mice attenuated the phenotype without any detectable side effects. Recovery was evident in the key parameters such as reduced sarcolemma permeability confirmed by lower serum creatine kinase levels and IgG influx into myofibres, decreased inflammatory cell numbers and inflammation markers in leg and heart muscles of treated mice. Moreover, this short-term therapy had some positive impact on muscle strength in vivo and no detrimental effect on mitochondria, which is the main side-effect of Nucleoside Reverse Transcriptase Inhibitors (NRTIs). Given these results, we postulate that AZT could be quickly re-purposed for the treatment of this highly debilitating and lethal disease. This approach is not constrained by causative DMD mutations and may be effective in alleviating both muscle and non-muscle abnormalities.


BACKGROUND: We examined the effect of genetic syndromes and extracardiac (GS/EC) anomalies on single-ventricle (SV) palliation with focus on hospital and interstage death and progression toward subsequent palliation stages.

METHODS: First-stage palliation was performed in 530 neonates with SV: Norwood in 284 (53%), shunt in 173 (33%), and band in 73 (14%). Outcomes were compared between those with GS/EC anomalies (121 [23%]) and without GS/EC anomalies (409 [77%]). Regression analyses were adjusted for other risk factors (age, sex, prematurity, weight, SV anomaly, and first-stage palliation operation).

RESULTS: GS/EC anomalies varied among SV defects (range, 3% for double-inlet left ventricle to 100% for atrial isomerism). Patients with GS/EC anomalies required significantly longer durations of mechanical ventilation and intensive care unit and hospital stay. Although patients had comparable rates of extracorporeal membrane oxygenation (13% vs 11%, p = 0.552) and unplanned reoperation (16% vs 11%, p = 0.189), hospital mortality was higher in patients with GS/EC anomalies (24% vs 12%, p = 0.0008). After discharge, patients with GS/EC anomalies had higher interstage death, with lower progression to Glenn (60% vs 77%, p = 0.002) and lower 10-year survival (56% vs 76%, p < 0.001). After adjustment for other risk factors, GS/EC anomalies significantly affected survival in almost all subgroups of patients.

CONCLUSIONS: The presence of GS/EC anomalies varies among SV anomalies and is associated with additional risk factors such as prematurity and low weight. After adjusting for other risk factors, GS/EC anomalies are associated with prolonged recovery after first-stage palliation and increased hospital and interstage death, with subsequently fewer patients progressing toward Glenn shunt. The increased death risk in those patients is highest in the first 6 months and persists for 2 to 3 years after first-stage palliation, suggesting the need for more vigilant monitoring and outpatient care in these high-risk patients.


BACKGROUND: PPAR-delta is a transcription factor which has crucial roles in stimulating oligodendroglial differentiation and myelination and its activation was also shown to differentiate malignant C6 glioma cells into oligodendrocytes.

OBJECTIVE: One of the ligands of PPAR-delta is erucic acid (EA), an edible omega-9 fatty acid consumed more by Asian populations and exists highly in Chinese womens milk. There exist epidemiological evidence that pediatric brain tumor incidence is among the lowest in the Chinese population. EA is also an ingredient of Lorenzo’s oil used against adrenoleukodystrophy, a pediatric demyelinating disease. EA was inappropriately assumed as a strong cardiotoxin based on Spanish oil syndrome, caused by toxic-aniline dye refined rapeseed oil. In this study, we studied whether EA is capable to block growth of C6 glioma cells and modify cardiotoxicity of doxorubicin.
MATERIALS AND METHODS: We studied effects of EA on the 3-dimensional appearance of the adherent cells, soft agar colony formation and S-phase in the 3-dimensional spheroids in C6 glioma cell cultures. We also investigated the effects of EA on hepatic and cardiac toxicity of doxorubicin.

RESULTS: EA decreased in vitro growth of C6 glioma cells at therapeutically achievable concentrations. EA effects were more prominent in 3D-assays (soft agar colonies and spheroids) and induced cell fusions in monolayer cultures. EA decreased S-phase inhibitory potency of doxorubicin (DOX), yet augmented its efficacy to induce a senescent morphology (as assessed by scanning electron microscopy) in monolayer and to increase iNOS and eNOS expression in spheroids. In our study, EA reduced DOX-induced necrosis in mice heart and liver and induced healthier morphology of heart mitochondria (as assessed by transmission electron microscopy); yet intercalated disks (ID) were more disturbed with DOX + EA.

CONCLUSIONS: Both the antitumor and cardiac effects of EA may associate with the cell-to-cell contact mechanisms. Combining systemic EA with intrathecal DOX-chemotherapy via Ommaya reservoirs may reduce DOX concentrations in systemic circulation, hinder toxic interactions with EA and induce selective kill of glioma cells.


BACKGROUND: To develop an empirically derived, reliable and valid measure of grief in adolescents, aged 12-18 years old.

METHODS: An online survey comprising 59 items derived from a qualitative study of 39 bereaved adolescents, the Hogan Inventory of Bereavement Children and Adolescents (HIB), the Depression, Anxiety and Stress Scales (DASS-21), the Multidimensional Scale of Perceived Social Support (MSPSS), and a series of death- and mental health-related questions, targeted adolescents bereaved when aged 12-18 years, with 176 adolescents (80.6% girls) completing the survey. RESULTS: Factor Analysis of the 59-items resulted in a final solution, the Adolescent Grief Inventory (AGI) comprised of 40 items and 6 factors: Sadness, Self-blame, Anxiety and Self-harm, Shock, Anger and Betrayal, and Sense of Peace, with indices of good fit (RMSEA=0.057, CFI=0.952, TLI=0.948). There was strong evidence of convergent (HIB) and divergent (MSPSS) validity. Adolescents bereaved by suicide scored higher on Self-blame, Anger and Betrayal while those with a history of suicidal behaviour or having a mental health diagnosis scored higher overall than those who had not.

LIMITATIONS: Study limitations include the self-selected, mostly female, sample, a high proportion of participants with a mental health and self-harm history, and reliance on self-reported data. CONCLUSIONS: The AGI is a novel, comprehensive and valid measure of grief in adolescents. It can be used broadly, including with bereaved adolescents at-risk of mental health ramifications.


OBJECTIVE: To study self-reported pain early in the disease course of juvenile idiopathic arthritis (JIA) as predictor of long-term disease outcomes.

METHODS: Consecutive cases of JIA with disease onset 1997-2000 from defined geographical areas of Norway, Sweden, Finland and Denmark were prospectively enrolled in this population-based cohort study. Self-reported, disease-related pain was measured on a 10 cm visual analogue scale (VAS pain). Inclusion criteria were a baseline visit with pain score six months after disease onset, followed by an eight-year study visit. Remission was defined according to Wallace preliminary criteria. Functional disability was measured by Childhood Health Assessment Questionnaire (CHAQ) and Child Health Questionnaire (CHQ-PF50) if age <18 and Health Assessment Questionnaire (HAQ) if age >/=18 years. Damage was scored using the Juvenile Arthritis Damage Index (JADI).

RESULTS: The final study cohort consisted of 243 participants, and 120 (49%) had oligoarticular onset. At baseline 76% reported VAS pain >0 compared to 57% at eight-year. Half of those who reported baseline pain also reported pain at eight-year, but at a lower intensity. Compared to no pain, higher pain intensity at baseline predicted more pain at eight-year, more functional disability, more damage and less remission off medication. Baseline pain predicted more use of DMARDs/biologics during the disease course. Participants with oligoarticular JIA reporting pain at baseline were more likely to develop extended oligoarticular or other unfavorable JIA categories.
CONCLUSION: Early self-reported, disease-related pain among children and adolescents with JIA is common and seems to predict persistent pain and unfavorable long-term disease outcomes. This article is protected by copyright. All rights reserved.


BACKGROUND: The Faces Pain Scale-revised (FPS-r) has been developed as an interval scale. For other pain measurement instruments, several studies found evidence for and against an interval level of measurement.

OBJECTIVES: The primary aim of the current study was to evaluate the scale properties of the FPS-r using an item response theory approach. DESIGN: Secondary analysis of published data. SETTING: Three studies; Study 1 and study 2: One university hospital; Study 3: international pain registry.

PARTICIPANTS: Study 1: n = 246, female: 41%, age: 11-18 years, 3 pain items; Study 2: n = 240, female: 43%, age: 11-18 years, 9 pain items; Study 3: n = 2266, female: 41%, age: 4-18 years, 3 pain items. METHODS: The rating scale model (interval scale), the graded response model (no interval scale, ordered response categories) and the partial credit model (no interval scale) were used to scale the data.

RESULTS: In all three studies, the rating scale model was outperformed by the graded response model or the partial credit model in terms of model fit. Overlapping response categories were found in items associated with less pain. Response category widths were wider for categories associated with low pain intensity and smaller for categories associated with high pain intensities. Smallest response categories were 1%-67% smaller compared to the widest response category of the same item.

CONCLUSION: According to these findings, the interval scale properties of the FPS-r may be questioned. Item response theory methods may help to solve the problem of missing linearity in pain intensity ratings using FPS-r.


INTRODUCTION: Atypical teratoid/rhabdoid tumor (AT/RT) of the central nervous system is characterized by SMARCB1/INI deletion or mutation in the long arm of chromosome 22 11(22q11.2), also resulting in loss of nuclear expression of INI1 protein immunohistochemically. AT/RT tumors usually occur in children below 3 years. The tumor is usually seen in the cerebellum or the cerebrum, with an extremely rare incidence in the spinal cord.

MATERIALS AND METHODS: We report a rare case of AT/RT in a 6-year-old boy who had a primary spinal cord lesion in the thoracolumbar junction. Pathology revealed loss of nuclear staining of INI1 immunohistochemically. AT/RT tumors usually occur in children below 3 years. The tumor is usually seen in the cerebellum or the cerebrum, with an extremely rare incidence in the spinal cord.

RESULTS AND DISCUSSION: We reviewed the literature on all children with spinal cord AT/RT. The review showed that the cervical region is the most common location of origin, especially in younger children. Reported cases were treated with a combination of surgery, systemic and intrathecal chemotherapy, and radiation therapy, and a survival time of 18 months represented the best outcome. Overall mean survival time was 10 months.


BACKGROUND: Sickle cell disease is an inherited blood disorder that affects over 100,000 Americans. Sickle cell disease-related complications lead to significant morbidity and early death. Evidence supporting the feasibility, acceptability, and efficacy of self-management electronic health (eHealth) interventions in chronic diseases is growing; however, the evidence is unclear in sickle cell disease.
OBJECTIVE: We systematically evaluated the most recent evidence in the literature to (1) review the different types of technological tools used for self-management of sickle cell disease, (2) discover and describe what self-management activities these tools were used for, and (3) assess the efficacy of these technologies in self-management.

METHODS: We reviewed literature published between 1995 and 2016 with no language limits. We searched MEDLINE, EMBASE, CINAHL, PsycINFO, and other sources. We followed the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. Two independent reviewers screened titles and abstracts, assessed full-text articles, and extracted data from articles that met inclusion criteria. Eligible studies were original research articles that included texting, mobile phone-based apps, or other eHealth interventions designed to improve self-management in pediatric and adult patients with sickle cell disease.

RESULTS: Of 1680 citations, 16 articles met all predefined criteria with a total of 747 study participants. Interventions were text messaging (4/16, 25%), native mobile apps (3/16, 19%), Web-based apps (5/16, 31%), mobile directly observed therapy (2/16, 13%), internet-delivered cognitive behavioral therapy (2/16, 13%), electronic pill bottle (1/16, 6%), or interactive gamification (2/16, 13%). Interventions targeted monitoring or improvement of medication adherence (5/16, 31%); self-management, pain reporting, and symptom reporting (7/16, 44%); stress, coping, sleep, and daily activities reporting (4/16, 25%); cognitive training for memory (1/16, 6%); sickle cell disease and reproductive health knowledge (5/16, 31%); cognitive behavioral therapy (2/16, 13%); and guided relaxation interventions (1/16, 6%). Most studies (11/16, 69%) included older children or adolescents (mean or median age 10-17 years; 11/16, 69%) and 5 included young adults (>/=18 years old) (5/16, 31%). Sample size ranged from 11 to 236, with a median of 21 per study: <20 in 6 (38%), >/=20 to <50 in 6 (38%), and >50 participants in 4 studies (25%). Most reported improvement in self-management-related outcomes (15/16, 94%), as well as high satisfaction and acceptability of different study interventions (10/16, 63%).

CONCLUSIONS: Our systematic review identified eHealth interventions measuring a variety of outcomes, which showed improvement in multiple components of self-management of sickle cell disease. Despite the promising feasibility and acceptability of eHealth interventions in improving self-management of sickle cell disease, the evidence overall is modest. Future eHealth intervention studies are needed to evaluate their efficacy, effectiveness, and cost effectiveness in promoting self-management in patients with sickle cell disease using rigorous methods and theoretical frameworks with clearly defined clinical outcomes.


Purpose: T-cell acute lymphoblastic leukemia (T-ALL) is one of the most common malignancies associated with T-lymphocytes, accounting for 10 to 15 percent of ALL cases in children and 25 percent in adults. Innovative therapeutic approaches that overcome ineffective treatments on tumor cells may be a potential source of improvement in therapeutic approaches. Suppression of gene expression at transfusion level is one of the important strategies in gene therapy. The expression of PTPN22 and miR-181 genes in all types of hematologic malignancies increases and is likely to contribute to the survival and death of cells by affecting a variety of signaling pathways. The purpose of this study was to determine the role of PTPN22 inhibition by siRNA, and alteration in miR-181a and miR-181b in Jurkat cell line. Methods: Jurkat cells were transfected with 80 pmol of siRNA to inhibit PTPN22. After that, expression of PTPN22 mRNA and transcript levels of miR-181a and miR-181b were measured with Real-time PCR after 48hrs.

Results: Experiments demonstrated that siRNA transfection resulted in significant downregulation of PTPN22 mRNA after 48 hrs in 80 pmol dose of siRNA. Moreover, transcript levels of both miR-181a and miR-181b was decreased after transfection.

Conclusion: PTPN22, miR-181a and miR-181b might be involved in progression of Jurkat cells and targeting these molecules by RNAi might confer promising tool in treatment of T-ALL.


Background: The diagnosis and management of pediatric melanomas is challenging given the presence of both melanomas and histologically aggressive Spitz tumors of undetermined biological significance (S-UBS) in this age group. Study objectives were to examine: factors leading to diagnostic delays, therapy side effects and patient outcomes in these diagnostic groups.
Methods: A retrospective case review was performed using The University of Michigan’s pediatric oncology database over a 13-year timespan. Patients referred to our clinic for consideration of interferon therapy due to a diagnosis of a stage III melanoma or aggressive appearing S-UBS with significant lymph node involvement were included.

Results: We found two major causes of diagnosis delay: patients with amelanotic lesions misdiagnosed as having a wart and cases reviewed by non-expert pathologists upfront. The side effects from interferon therapy requiring dose adjustments included neutropenia, thrombocytopenia and mood disturbances. There was wide variability in surveillance scan utilization, therefore leading to variability in patient radiation exposure. Unlike melanoma patients, none of the S-UBS patients experienced disease progression or death.

Conclusions: This study highlights the challenges with the initial clinical diagnosis and pathological sub-categorization of the pediatric S-UBS/melanoma spectrum of skin lesions and emphasizes the role of expert pathology review upfront. Earlier diagnosis could spare patients from more extensive interventions, metastatic spread or adverse outcomes in this patient population. This study is limited due to its retrospective, single-institution perspective.


BACKGROUND: Traumatic brain injury (TBI) is the leading cause of death and long-term disability among injured children. Early feeding has been shown to improve outcomes in adults, with some similar evidence in children with severe TBI. We aimed to examine the current practice of initiation of enteral nutrition in children with TBI and to evaluate the risk factors associated with delayed initiation of enteral nutrition.

METHODS: This retrospective, multicenter study used the Pediatric Trauma Assessment and Management Database including all children with head trauma discharged from five pediatric intensive care units (PICU) at pediatric trauma centers between January 1, 2013 and December 31, 2013. We compared demographics, injury and procedure data, time to initiation of nutrition, and injury and illness severity scores between patients who received enteral nutrition early (</= 48 h) and late (> 48 h). Fisher’s exact and Mann-Whitney U tests compared discrete and continuous variables. Univariate and multivariable analyses evaluated variables associated with delayed initiation of feeding. Outcomes of interest included mortality, complications, ventilator days, hospital and ICU length of stay, and functional status at ICU discharge.

RESULTS: In the 416 patients in the study, the overall mortality was 2.6%. The majority of patients (83%; range 69-88% between five sites, p = 0.0008) received enteral nutrition within 48 h of PICU admission. Lower Glasgow Coma Scale scores and higher Injury Severity Score (ISS) were independently associated with delayed initiation of enteral nutrition. Delayed enteral nutrition was independently associated with worse functional status at PICU discharge (p = 0.02) but was not associated with mortality or increased length of stay.

CONCLUSIONS: Children with severe TBI and higher ISS were more likely to have delayed initiation of enteral nutrition. Delayed enteral nutrition was an independent risk factor for worse functional status at ICU discharge for the entire cohort, but not for the severe TBI group.


Introduction: Cerebral palsy (CP) is the most common cause of physical disability in children and is often associated with secondary musculoskeletal pain. Cerebral palsy is a heterogeneous condition with wide variability in motor and cognitive capacities. Although pain scales exist, there remains a need for a validated chronic pain assessment tool with high clinical utility for use across such a heterogeneous patient population with and without cognitive impairment.

Objectives: The purpose of this study was an initial assessment of several psychometric properties of the 12-item modified brief pain inventory (BPI) pain interference subscale as a proxy-report tool in a heterogeneous sample of children with CP with and without cognitive impairment. Methods: Participants (n = 167; 47% male; mean age = 9.1 years) had pain assessments completed through caregiver report in clinic before spasticity treatment (for a subgroup, the modified BPI was repeated after procedure). To measure concurrent validity, we obtained pain intensity ratings (Numeric Rating Scale of pain) and pain intensity, duration, and frequency scores (Dalhousie Pain Interview).
Results: Modified BPI scores were internally consistent (Cronbach alpha = 0.96) and correlated significantly with Numeric Rating Scale intensity scores ($rs = 0.67, P < 0.001$), Dalhousie Pain Interview pain intensity ($rs = 0.65, P < 0.001$), pain frequency ($rs = 0.56, P = 0.02$), and pain duration scores ($rs = 0.42, P = 0.006$). Modified BPI scores also significantly decreased after spasticity treatment (pretest [scored 0-10; $3.27 +/- 2.84$], posttest [2.27 +/- 2.68]; $t(26) = 2.14$, 95% confidence interval [0.04-1.95], $P = 0.04$).

Conclusion: Overall, the modified BPI produced scores with strong internal consistency and that had concurrent validity as a proxy-report tool for children with CP.


Ornithine transcarbamylase deficiency (OTCD) disrupts the metabolic pathway responsible for converting nitrogenous waste to urea, allowing for excretion. When impaired, ammonia levels accumulate in the blood resulting in severe, sometimes life-threatening toxicities. Abnormalities of the urea cycle are often inherited, though there are some rarer acquired forms. We describe two cases of acquired OTCD in pediatric patients with fibrolamellar hepatocellular carcinoma (FL-HCC). We detail its presentation and management, explore potential underlying pathophysiology, and propose a practice change to optimize care of FL-HCC patients.


OBJECTIVES: We assessed neuroactive medication use in critically ill children who require neurological consultation and evaluated the associations between administration of these medications and continuous electroencephalography (cEEG) utilization and seizure frequency.

METHODS: We evaluated exposure to sedatives, analgesics, anesthetics, and paralytics in consecutive patients (0 days to 18 years) for whom neurological consultation was requested in three intensive care units (ICUs) [neonatal (NICU), pediatric (PICU), and cardiothoracic (PCTU)] at one children’s hospital. We assessed cEEG usage and seizure incidence in relation to drug exposure.

RESULTS: From November 2015 to November 2016, 300 consecutive patients were evaluated (93 NICU, 139 PICU, and 68 PCTU). Ninety-seven (32%) were receiving $\geq$1 sedative infusion at the time of consultation [NICU 7 (8%), PICU 50(36%), PCTU 40 (68%)]; 91 (30%) received $\geq$1 paralytic agent within the preceding 24 hours. Continuous electroencephalography was performed more often for patients treated with sedative infusions (81 of 97 versus 133 of 203, $P = 0.001$) and paralytic medications (80 of 91 versus 134 of 209, $P < 0.001$) within 24 hours preceding consultation than those who were not. Sixty-eight of 214 (32%) had electrographic seizures (65 of 68 within initial 24 hours of monitoring); seizures were less common among patients who had received sedative infusions (18 of 81 versus 51 of 133, $P = 0.014$). In multivariable analysis of seizure likelihood, only younger age was associated with increased risk ($P = 0.037$).

CONCLUSIONS: Critically ill infants and children are frequently treated with sedatives, anesthetics, analgesics, and paralytics. Neuroactive medications limit bedside neurological assessments and, in this cohort, were associated with increased cEEG usage. Our data underscore the need to study the effect of these medications on clinical care and long-term outcomes.


AIM: We aimed to identify early predictors of intractable epilepsy, intellectual disability (ID) and autism spectrum disorders (ASD) in the cohort of TSC patients initially diagnosed with cardiac rhabdomyomas (CR).

METHOD: Over the period of twelve years we prospectively obtained clinical, neuropsychological, electrophysiological and neuroimaging data in a group of 22 TSC patients (9 females, 13 males) with the
pre/perinatal diagnosis of CR, included to the study at the time of diagnosis. Afterwards, we statistically determined variables associated with ID, ASD and intractable epilepsy.

RESULTS: Development of ID was predicted by severe epilepsy (a higher number of anti-epileptic drugs used), a higher number of dysplastic lesions on MRI, and abnormal background activity on EEG (p < 0.05). Predictors of ASD included early developmental delay, abnormal background activity on EEG at the end of follow-up and a higher number of areas with dysplastic features on MRI (p < 0.05). Intractable epilepsy was associated with a higher number of areas with dysplastic features on MRI, ID and with TSC2 genotype.

CONCLUSION: Adverse mental and clinical outcome was associated with intractable epilepsy and the severe anatomical brain involvement; therefore, our centre developed a tailored protocol for early identification of TSC patients at a higher risk of developing intractable epilepsy with its deleterious effect on cognitive outcome.


Vitamin K deficiency bleeding (VKDB) is a life-threatening condition and can be found in children as early as neonatal period with early onset intracranial hemorrhage (ICH). Here, we reported a 1-year-old boy who initially presented with intracranial hemorrhage secondary to vitamin K deficiency since 3 months of age and later found to have XL-CGD which was complicated by malabsorption due to severe vaccine-associated mycobacterial disease.


Fulminant meningococcemia is a relatively rare life-threatening disease caused by Neisseria meningitidis. The clinical presentation is varied, but, when associated with myocarditis, it carries a particularly poor prognosis. We report a case of a patient with fulminant meningococcemia who subsequently developed severe myocardial dysfunction and successfully recovered within a period of 7 days of hospitalization. A 15-year-old girl presented with headache, fever, body ache for 1 day and few ecchymotic rash over her body for 3 hours. Blood cultures confirmed infection with N. meningitidis. After 2 days in the hospital, the patient developed dyspnea, elevated jugular venous pressure and shock. The patient was managed with intravenous ceftriaxone, furosemide and norepinephrine. Over the next 4 days the patient rapidly improved. Meningococcemia complicated by myocarditis has an extremely poor prognosis with high mortality. Our case suggests that recovery from a severe myocardial dysfunction can occur rapidly within a few days. Prompt recognition and management in this case might have contributed to the patient’s rapid recovery from myocarditis.


BACKGROUND: The Child Perceptions Questionnaire (CPQ) belongs to a set of questionnaires measuring Child Oral Health Quality of Life (COHQOL). The CPQ is used to collect the perceptions of children on the impact of oral diseases on their quality of life. This cross-sectional study was aimed to translate the CPQ8-10 into French language and evaluate its psychometric properties.

METHODS: The translation process complied with international recommendations. The final French version was tested on children aged 8-10 years old attending consultations in a Parisian public hospital and divided into three groups: children with oral-facial clefts, children with dental anomalies linked to a rare disease other than clefts and children presumed to be healthy and without anomalies. The internal consistency relating to the reliability of CPQ8-10 was evaluated by Cronbach’s alpha. The intra-class correlation was used to measure reproducibility at the test-retest level. Construct validity was evaluated by Spearman’s correlation and tested using factor analysis. The discriminant validity was assessed using Kruskall Wallis test. Criterion validity was calculated using Spearman’s correlation.

RESULTS: One hundred seventy-six children participated in this study. During the translation process, minor changes were made. The French version showed good reliability with a Cronbach’s alpha of 0.81 for the total scale.
The ICC of the test-retest was excellent (=0.90) demonstrating good reproducibility. The construct validity was acceptable with a statistically significant correlation between the scores of the French-CPQ8-10 and the evaluation of oral health (r = 0.381 and p < 0.001) and its impact on oral health quality of life (r = 0.363 and p < 0.001). The loading weights obtained in the Exploratory Factor Analysis showed that this model revealed seven factors with eigenvalue greater than 1, explaining the 63.89% of the cumulative variance. The differences observed between the scores of the study groups revealed good discriminant validity. Criterion validity was supported by significant association between CPQ scores and pain.

CONCLUSION: The French-CPQ8-10 is reliable and valid for use with the children of this age group.


Pediatric Central Nervous System (CNS) neoplasms are the second most prevalent tumors of childhood. Further on, prognosis of this type of neoplasms still remain poor and the comprehenshion of the etiology and pathogenesis of the disease still remains scarce. Several reports have identified microRNAs as significant molecules in the development of central nervous system tumors and propose that they might compose key molecules underlying oncogenesis. In a previous study we have identified several miRNAs, common to different subtypes of pediatric embryonal CNS malignancies as well as, we have identified miRNAs that manifest significant dynamics with respect to their expression and the neoplastic subtype. Overall, 19 tumor cases from children diagnosed with embryonal brain tumors were investigated. As controls, children who suffered a sudden death underwent autopsy and were not present with any brain malignancy were used (13 samples of varying localization). Our experimental approach included microarrays covering 1211 miRNAs, which appeared to manifest tumor-specific dynamics. In conclusion, it appeared that certain miRNAs are neoplasm specific and in particular, their expression manifests linear dynamics. Thus, the investigation of miRNA expression in pediatric embryonal brain tumors might contribute towards the discovery of tumor-specific miRNA signatures, which could potentially afford the identification of gene-specific biomarkers related to diagnosis, prognosis and patient targeted therapy, as well as help us understand oncogenetic dynamics.


Background Although the survival of pediatric cancer has increased dramatically in the last decades, the survival of refractory, relapsed, and metastatic cases is still dismal. The combination of irinotecan and temozolomide has shown activity against refractory/relapsed pediatric solid tumors.

Method Thirty-four children with refractory/relapsed solid tumors who had previously been heavily pretreated and who were given vincristine, irinotecan, and temozolomide as third- or further line chemotherapy during 2004-2015 were evaluated. Results Patients were diagnosed with Ewing sarcoma (n = 15), rhabdomyosarcoma (n = 8), neuroblastoma (n = 8), osteosarcoma (n = 2), and Wilms’ tumor (n = 1). Thirty patients presented with disease progression on therapy and the other four presented with relapsing. A total of 141 cycles were administered. Radiotherapy was used in 17 patients and surgery in 4 as local therapy. Among all patients, 6 had complete response, 3 had partial response, 14 had stable disease, and 11 had progressive disease. The objective response was 26.4% (complete response + partial response) and median survival duration was six months. The first and second year overall survival rates were 22.3% and 16.8%. The objective response in Ewing sarcoma patients was 40%. Diarrhea was the most common toxicity and 14 (10%) courses were associated with grade 3-4 diarrhea.

Conclusions In heavily pretreated patients with refractory/relapsed solid tumors, the vincristine, irinotecan, and temozolomide regimen seemed promising in Ewing sarcoma patients and was well tolerated.


Subacute sclerosing panencephalitis (SSPE) is a progressive neurodegenerative disease which affects children and young adults, caused by a persistent infection of defective measles virus. IFN-lambda5 (IL-28A, IL-28B and IL-29) are a group of cytokines mediating antiviral responses. It has been shown that IL-29 levels are significantly higher in infected cells with defective measles virus. IL-29 expression is thought to be regulated at post-transcriptional level.
and miRNA-548 family targets the 3'UTR of the IFNL1 gene. Impaired immune system has an important role as well as viral factors in SSPE. The aim of our study investigates whether IL-28B, IL-29 levels and gene polymorphisms contribute to the damaged immune response leading to the development of SSPE. Also possible association of miRNA-548 family with IL-29 and SSPE is explored. Frequencies of rs12979860, rs8099917, rs30461, serum levels of IL-28B, IL-29 and expression levels of miR-548b, miR-548c, miR-548i are determined at 64 SSPE patients and 68 healthy controls. Serum IL-29 levels are statistically significant higher in SSPE patients. Allele frequencies of rs8099917 are statistically significant higher in SSPE patients and resulted G allele is found to increase 2.183-fold risk of SSPE. The expression levels of miR-548b-5p, miR-548c-5p and miR-548i are found to be statistically significant higher in SSPE patients. Dramatically increased level of IL-29 seen in patient group indicates that the elevated miR-548 expression is compensatory result of the over-activated immune system response. Further studies referred to IL28, IL29 and related miRNA’s will be enlightened the pathogenesis of SSPE.


INTRODUCTION: DBS is initially used for treatment of essential tremor and Parkinson’s disease in adults. In 1996, a child with severe life-threatening dystonia was offered DBS to the internal globus pallidus (GPI) with lasting efficacy at 20 years. Since that time, increasing number of children benefited from DBS.

PATIENTS AND METHODS: We retrospectively evaluated our database of patients who underwent DBS from 2011 to 2017. All patients <\= 17 years of age at the time of implantation of DBS were included in this series. Subjective Benefit Rating Scale (SBRS), Hoehn Yahr Scale (HYS), Fahn Marsden Rating Scale (FMRS), Clinical Global Impressions Scales (CGI), and Yale Global Tic Severity Scale (YGT) were used to evaluate clinical outcome. RESULTS: Between May 2014 and October 2017, 11 children underwent DBS procedure in our institution. Six of them were female and five of them were male. Mean age at surgery was 11.8 +/- 4.06 years (range 5-17 years). In our series, four patients had primary dystonia (PDY) (36.3%), three patients had secondary dystonia (SDY) (27.2%), two patients had JP (18.1%), and two patients had Tourette Syndrome (TS) (18.1%). Two JP patients underwent bilateral STN DBS while the other nine patients underwent bilateral GPI DBS. SBRS scores were 1.75 +/- 0.5 for patients with PDY, 3 +/- 0 for patients with JP, 2.5 +/- 0.7 for patients with TS, and 2 +/- 1 for patients with SDY. Mean FMRS reduction rate was 40.5 for patients with dystonia. Significant improvement was also defined in patients with TS and JP after DBS. None of the patients experienced any intracerebral hemorrhage or other serious adverse neurological effect related to the DBS. Wound complications occurred in two patients.

CONCLUSION: There are many literatures that support DBS as a treatment option for pediatric patients with medically refractory neurological disorders. DBS has replaced ablative procedures as a treatment of choice not only for adult patients, but also for pediatric patients. Wound-related complications still remain the most common problem in pediatric patients. Development of smaller and more flexible hardware will improve quality of children’s life and minimize wound-related complications in the future.


OBJECTIVE: Cancer mortality is a leading cause of disease-related death in the adolescent and young adult (AYA) population. Compared with older and younger patients, AYA patients often experience worse cancer-specific outcomes. Here, we compare AYA and pediatric overall survival (OS) in the most common pediatric extracranial solid tumors.

MATERIALS AND METHODS: Using the US Surveillance, Epidemiology, and End Results database, we studied patients (age, 0 to 39 y) diagnosed with Ewing sarcoma, neuroblastoma, osteosarcoma, rhabdomyosarcoma, and Wilms tumor.

RESULTS: A total of 12,375 patients (age, 0 to 39 y) were diagnosed between 1973 and 2010 (8247 pediatric and 4128 AYA patients). AYA patients with rhabdomyosarcoma and Ewing sarcoma were more likely to present with metastatic disease. OS was significantly worse in the AYA cohort for all tumor types (P<0.001) with the exception of osteosarcoma (P=0.29). Across 2 treatment time periods (1973 to 1989 and 1990 to 2010), there was significant improvement in 5-year OS in all tumor types with the exception of rhabdomyosarcoma; however, AYA patients continued to experience worse OS in the modern treatment cohort with the exception of osteosarcoma patients. There was no improvement in OS among AYA patients with Ewing sarcoma, neuroblastoma, rhabdomyosarcoma, or Wilms tumor over the 2 treatment eras.
CONCLUSIONS: For the most common pediatric extracranial solid tumors, AYA patients experience significantly worse OS compared with pediatric patients. Although improvements in therapy have led to gain in survival for pediatric patients, with the exception of osteosarcoma, AYA experienced no increase in survival over the study period. This investigation demonstrates the importance for further research in the AYA population.


Although progressive cardiac dysfunction is the leading cause of death in patients with Duchenne muscular dystrophy (DMD), their cardiac function measured by conventional echocardiography has been generally interpreted as normal at a young age. We aimed to determine whether two-dimensional speckle tracking echocardiography (STE) or tissue Doppler imaging (TDI) could be used for early identification and detection of cardiac dysfunction in young patients with DMD. Thirteen pediatric patients (mean age, 9.69 +/- 2.2 years) with DMD and 26 age-matched healthy children (mean age, 9.65 +/- 2.2 years) were included in the study. All patients were examined via conventional echocardiography, TDI, and STE. Standard echocardiographic measurements of left ventricular (LV) systolic and diastolic function were obtained. Myocardial velocities including peak-systolic and early- and late-diastolic myocardial velocities were calculated in longitudinal direction in the interventricular septum, using TDI. Speckle tracking analyses were performed by acquiring apical four-, three-, and two-chamber views with the highest possible frame rates. Conventional parameters were similar between the two groups, but heart rates were higher in patients with DMD than in controls. The results of LV diastolic function evaluated using TDI showed that annular peak velocity during early diastole (e'; 10.9 +/- 1.7 vs. 14.6 +/- 1.7 cm/s), e'/a' ratio (2.0 +/- 0.5 vs. 3.0 +/- 0.5), E/e' ratio (9.4 +/- 1.4 vs. 7.3 +/- 0.8), and myocardial performance index (0.46 +/- 0.05 vs. 0.36 +/- 0.06) of the mitral septal annulus among patients with DMD differed significantly from those of healthy children. A significant decrease in global longitudinal systolic strain was found in patients with DMD (~ 16.6 +/- 3.7 vs. -21.2 +/- 2.1), with a marked decrease in the LV basal inferolateral and basal inferior walls. In young patients with DMD who have global normal systolic function, reductions in systolic deformation parameters as well as reduced early diastolic myocardial velocities can be detected particularly in the basal inferolateral LV walls. The prognostic significance of these findings warrants further longitudinal follow-up.


PURPOSE: The paper presents a long-term follow-up study of VNS patients, analyzing seizure outcome, medication changes, and surgical problems.

METHOD: 74 adults with VNS for 10 to 17 years were evaluated yearly as: non-responder - NR (seizure frequency reduction <50%), responder - R (reduction>/=50% and <90%), and 90% responder - 90R (reduction>/=90%). Delayed R or 90R (>4years after surgery), patients with antiepileptic medication changes and battery or complete system replacement were identified. Statistical analysis of potential outcome predictors (age, seizure duration, MRI, seizure type) was performed.

RESULTS: The rates of R and 90R related to the patients with outcome data available for the study years 1, 2, 10, and 17 were for R 38.4%, 51.4%, 63.6%, and 77.8%, and for 90R 1.4%, 5.6%, 15.1%, and 11.1%. The absolute numbers of R and 90R increased until years 2 and 6. Antiepileptic therapy was changed in 62 patients (87.9%). There were 11 delayed R and four delayed 90R, with medication changes in the majority. At least one battery replacement was performed in 51 patients (68.9%), 49 of whom R or 90R. VNS system was completely replaced in 7 patients (9.5%) and explanted in 7 NR (9.5%). No significant predictor of VNS outcome was found.

CONCLUSIONS: After an initial increase, the rate of R and 90R remains stable in long-term follow-up. The changes of antiepileptic treatment in most patients potentially influence the outcome. Battery replacements or malfunctioning system exchange reflect the patient’s satisfaction and correlate with good outcomes.


Objective: Psychological intervention is widely recognized as an integral part of the recovery process from pediatric chronic pain, but service acquisition is often limited by resource barriers. The aim of this study was to assess the feasibility, acceptability, and satisfaction of a brief, structured, skills-based, group intervention designed expressly to address gaps in service delivery. Exploratory outcomes were also assessed.

Method: Adolescents with chronic pain (n = 102; ages 10-17 years) and their mothers (n = 105) completed self-report questionnaires at baseline, 1-week, 1-month, and 3-month posttreatment.

Results: This study demonstrated feasibility, and overall high acceptability and satisfaction among adolescents and parents. Exploratory analyses within this nonrandomized design suggest that adolescents demonstrate improvement in functionality (p = .0012), depression symptoms (p < .0001), and pain catastrophizing (p < .0001) by 1-month posttreatment and continued making gains over time. Parents made significant changes in parenting practices (p-values < .01) and in their beliefs about their adolescent’s ability to manage pain (p < .001) by 1-week posttreatment and continued making gains over time.

Conclusions: This brief intervention is both feasible and acceptable. Although small effect sizes were found for all outcome measures, parents and adolescents made significant gains postintervention. In the absence of a direct comparison group, we cannot determine if these improvements are exclusively attributable to the intervention. Future research will be needed to understand the degree to which this brief intervention may effectively enhance the attainment of evidence-based psychoeducation and cognitive behavioral skills that are known to foster adaptive parent and adolescent responses to chronic pain.


Spinal muscular atrophy (SMA) is one of the most common childhood onset neurodegenerative disorders in global health whereby novel biomarkers and therapeutic targets are sorely needed. SMA is an autosomal recessive genetic disorder resulting in degeneration of alpha-motor neurons in the brain stem and spinal cord that leads to mortality in infants worldwide. In majority of the patients, SMA is caused by homozygous deletion of the SMN1 gene. The clinical spectrum of the SMA displays, however, large person-to-person variations where the underlying mechanisms are poorly understood. We report in this study transcriptomics insights gleaned from patients with the severe type I (GM03813 and GM09677) and the mild type III. Pathway enrichment and functional analysis showed that especially extracellular matrix (ECM), synapse organization, and ECM receptor interaction pathways were affected. Among the neural ECM components, hyaluronan and proteoglycan link protein (HAPLN1), which is a key triggering molecule of the perineuronal net (PNN), was significantly downregulated in type I fibroblasts compared to type III. PNN is a specialized form of neural ECM around the neuronal cell bodies and dendrites in the central nervous system. In addition, we evaluated the PNN expression in vitro in a model established by SMN silencing in the PC12 rat pheochromocytoma cell line which can be differentiated into neurons with nerve growth factor treatment. In this neuronal in vitro model, we found that HAPLN1 showed a significant 50% decrease. Our results describe the association between PNN elements, especially HAPLN1, and SMA pathophysiology for the first time. These observations collectively inform future translational research on SMA for discovery of novel molecular targets for diagnostics and precision medicine innovation.


BACKGROUND: Pineoblastomas are rare, malignant embryonal tumors that have a relatively higher incidence and a poorer prognosis in children. Owing to the rarity of these tumors, there is a paucity of data on associated prognostic factors. We used the Surveillance, Epidemiology, and End Results (SEER) database to evaluate prognostic factors for pineoblastomas with the aim of improving tumor management.

METHODS: Data from all pediatric patients (age <=17 years) diagnosed with pineoblastoma between 1990 and 2013 were extracted from the SEER-18 registry database. Survival was described with Kaplan-Meier curves. The Cox proportional hazards model was used for both univariate and multivariate analyses. A nomogram was established for predicting 1-, 3-, and 5-year overall survival (OS) in patients with pineoblastoma.

RESULTS: Age >5 years (P = 0.004) and radiotherapy treatment (P = 0.000) were associated with better rates of survival. Gross total resection (P = 0.054) also was correlated with better prognosis, whereas tumor size >30 mm in maximum diameter (P = 0.025) was associated with poorer outcome. A nomogram was established based on the
results of the Cox model and was validated by a concordance index (C-index) of 0.767 (95% confidence interval, 0.698-0.836) and calibration plots.

CONCLUSIONS: Our results show that the impact of tumor extension is not defined. OS is better in older children treated by radiotherapy, and gross total resection also appears to result in increased survival. A nomogram was built to predict 1-, 3-, and 5-year OS for these patients.


OBJECTIVES: Fetal endoscopic tracheal occlusion (FETO) is offered to fetuses with congenital diaphragmatic hernia (CDH) and severe lung hypoplasia to promote lung growth and may secondarily affect left heart growth. The effects of FETO on left heart hypoplasia (LHH) are not described post-CDH repair.

METHODS: A retrospective analysis was performed for fetuses with left-sided CDH who underwent FETO and severity-matched controls from 2007 to 2016 at our institution. Echocardiographic, ultrasound, and MRI data were reviewed. Left heart dimensions were assessed prenatally and postnatally. Primary clinical outcome evaluated was death. RESULTS: Twelve FETO patients and 18 controls were identified. Fetal LHH was noted in both groups and worsened after FETO. Postnatal mitral valve dimensions were larger in the FETO group pre-CDH repair (P = .03). Post-CDH repair, mitral valve and left ventricular dimensions were not significantly different between groups (P = .79 and P = .63 respectively) while FETO aortic valve dimensions were smaller (P = .04). Extracorporeal membrane oxygenation use was lower in the FETO group. No associations were found between left heart dimensions and outcomes.

CONCLUSION: Although increased lung growth was seen after FETO, fetal LHH persisted with relative normalization seen post-repair. Persistent LHH post-FETO could be secondary to a small contribution of pulmonary venous return to the fetal left heart and increased intrathoracic pressures post-FETO.


Diverse behavioral cues have been proposed to be useful cues in infant pain assessment, but there is a paucity of evidence on the basis of formal psychometric evaluation to establish their validity for this purpose. We aimed to examine 2 widely used coding systems, the Neonatal Facial Coding System (NFCS) and the Modified Behavior Pain Scale (MBPS), by examining their factor structures with confirmatory factor analysis using a large archival data set. The results indicated that an item-reduced NFCS scale with 3 items produced a 1-factor pain model that maintained the good psychometric properties of the 7-item scale. In addition, it was found that MBPS also has challenging internal consistency, with items that are weakly correlated as well as highly redundant. One item of the MBPS may be able to construct the pain of equal quality or potentially improve its psychometric properties. Redefinition of the MBPS with cry as a sole indicator was suggested. This analysis provides 2 new iterations of the NFCS and MBPS scales. These revised measures improve the feasibility of both measures and increase their potential for clinical use because less time is required for their administration.

PERSPECTIVE: This article presents new iterations of the NFCS and MBPS scales. These revised measures improve the internal consistency of the measures, feasibility of use of the tools in research settings, and the efficiency of the coding process. The revised tools could also improve the feasibility of coding within clinical settings.


BACKGROUND: The Neonatal Infant Pain Scale and the Premature Infant Pain Profile have been used widely in neonatal intensive care units for pain assessment. AIM: This study reports the evaluation and validation of these scales in full-term newborns who were hospitalized in two Greek neonatal intensive care units. Evaluation and validation of the Neonatal Infant Pain Scale and the Premature Infant Pain Profile in full-term newborns who were hospitalized in two Greek neonatal intensive care units.
**MATERIALS AND METHODS:** This is a cross-sectional study. Two neonatal intensive care units at a large General Children’s Hospital in Greece. A total of 81 full-term newborns. This cross-sectional study was conducted in two neonatal intensive care units at a large General Children’s Hospital in Greece. We studied 81 full-term newborns, who were exposed to various painful routine procedures. A single measurement was taken from each neonate. Two observers were present during each procedure and evaluated pain using both the Neonatal Infant Pain Scale and Premature Infant Pain Profile. Internal consistency coefficient Cronbach’s alpha, internal class agreement coefficient, and kappa factor were appropriately measured.

**RESULTS:** The weighting of the Neonatal Infant Pain Scale and Premature Infant Pain Profile pointed out an excellent coherence between the two scales and agreement among the researchers. The internal consistency coefficient Cronbach’s alpha was >.8 and the internal class agreement coefficient was >.98 for both scales, which indicates an excellent consistency between scales. The kappa factor for Neonatal Infant Pain Scale was >.73 and for the Premature Infant Pain Profile it was >.6, which indicates a significant agreement among investigators.

**CONCLUSIONS:** The Neonatal Infant Pain Scale and Premature Infant Pain Profile were successfully adjusted in Greek standards with reliability between the scales and among the researchers. Moreover, they constitute reliable tools for the evaluation of neonatal procedural pain in full-term newborns in Greece.


**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.


**OBJECT:** To compare the occurrence of surgery-related complications in patients with childhood-onset focal epilepsy operated on in the paediatric or in the adult age. To investigate risk factors for surgery-related complications in the whole cohort, with special attention to age at surgery and severe morbidity.

**METHODS:** A cohort of 1282 patients operated on for childhood-onset focal epilepsy was retrospectively analysed. Occurrence of surgery-related complications, including a severely complicated course (SCC: surgical complication requiring reoperation and/or permanent neurological deficit and/or death), was compared between patients operated on in the paediatric age (<16 year-old; 452 cases) and, respectively, in adulthood (>/=16 year-old; 830 cases). The whole cohort of patients was also evaluated for risk factors for a SCC.

**RESULTS:** At last contact (median follow-up 98 months), 74.5% of patients were in Engel’s class I (78.0% of children and 73.0% of adults). One hundred patients (7.8%) presented a SCC (6.4% for children and 8.6% for adult patients). Postoperative intracranial haemorrhages occurred more frequently in adult cases. At multivariate analysis,
increasing age at operation, multilobar surgery, resections in the rolandic/perirolandic and in insulo-opercular regions were independent risk factors for a SCC.

CONCLUSIONS: Surgery for childhood-onset focal epilepsy provides excellent results on seizures and an acceptable safety profile at any age. Nevertheless, our results suggest that increasing age at surgery is associated with an increase in odds of developing severe surgery-related complications. These findings support the recommendation that children with drug-resistant, symptomatic (or presumed symptomatic) focal epilepsy should be referred for a surgical evaluation as early as possible after seizure onset.


BACKGROUND: The need for paediatric palliative care (PPC) globally is great yet there is limited evidence of the quality or outcomes of the care provided. The lack of an outcome measure for PPC has been consistently cited as one reason for the lack of robust evidence in the field. Thus recommendations have been made for the development of locally relevant, validated tools to measure outcomes for children.

METHODS: This paper reviews relevant outcomes and quality measures in PPC, the current state of science on outcome measurement for children and young people (CYP) with life-limiting and life-threatening conditions and the development of the African Children’s Palliative Outcome Scale (C-POS). Lessons learnt from the past are presented before looking ahead at the need for future developments in outcome measures in PPC. A narrative review was undertaken and authors have drawn upon reflective insights from their collective experiences.

RESULTS: Outcomes can be measured in a variety of ways, and due to the multi-dimensional nature of PPC, outcomes can be complex and hard to measure. Whilst there are a variety of outcome measures for use in adult palliative care, a similar range of tools does not exist in PPC. Literature reviews have confirmed the absence of a multi-dimensional PPC outcome measurement tool. Following on from their success in developing an outcome scale for adults in Africa, the African Palliative Care Association (APCA) have developed a multi-dimensional outcome tool for PPC-the African C-POS. Tool development and validation followed the COSMIN guidance. The draft C-POS consists of 12 questions, 8 in Section A for the child, and 4 in Section B for the parents/carers. The tool has been developed across eight African countries and is the first specifically designed, multi-dimensional outcome measure for PPC. Lessons have been learnt in the development of outcome scales in palliative care, including those specifically for PPC such as: undertaking research in PPC; the definition of PPC; if you ask a child what their concerns are they will tell you; do you use child and or proxy report? do you have different tools for different ages? what methods of scoring should be used? is it an outcome tool, an assessment tool or both? the length of the outcome measure; the length of time it takes to develop; and, it won’t be perfect. Whilst progress has occurred through the development of the C-POS there is still a long way to go in the development of outcome measures for PPC. Future developments include: finalization and publication of the African C-POS; utilization of the C-POS in clinical practice, research and audit; collation and review of data sets; and the development of C-POS in different settings.

CONCLUSIONS: The measurement of outcomes in PPC is an imperative. Whilst there are challenges in developing outcome tools and utilizing them in practice, these should not prevent us from advancing the field. The development of the first outcome measure for PPC the African C-POS is a key milestone in the ongoing development and utilization of outcome measures for PPC.


AIM: To determine the timing and modes of death of children admitted to a pediatric critical care unit (PICU) of a tertiary care center after an out-of-hospital cardiac arrest (OHCA).

METHODS: This is a retrospective descriptive study at a tertiary care PICU of all consecutive patients <18 years old who received >/=1 min of chest compressions, had return of spontaneous circulation (ROSC) for >/=20 min, and were admitted to the PICU after an OHCA. Modes of death were classified as brain death (BD), withdrawal due to neurologic prognosis (W/D-neuro), withdrawal for refractory circulatory failure (W/D-RCF), and re-arrest without ROSC (RA).

RESULTS: 191 consecutive patients were admitted to the PICU from February 2005 to May 2013 after an OHCA. Eighty-six(45%) patients died prior to discharge: BD in 47%(40/86), W/D-neuro in 34%(29/86), W/D-RCF in
9% (8/86), and RA in 9% (8/86). Time to death was longer for patients with W/D-neuro: 4 days [1, 5] and BD 4 days [1, 5](p < 0.01) as opposed to those with W/D-RCF (1 day[1, 2]) and RA(1 day[0.5, 1]). Of patients who underwent W/D-neuro, 9/29(31%) died within 3 days of PICU admission and 20/29(69%) >/=3 days. Of patients who died after W/D-neuro, 12/29(41%) received therapeutic hypothermia, 27/29(93%) underwent EEG monitoring, 21/29(72%) had a brain CT, and 13/29(45%) had a brain MRI. All MRIs showed signs of hypoxic-ischemic injury.

CONCLUSION: Neurologic injury was the most common mode of death post-resuscitation care OHCA after in a tertiary care center PICU. Neurologic prognostication impacts the outcome of a large proportion of patients after OHCA, and further studies are warranted to improve its reliability.


OBJECTIVES: The objective of this study was to evaluate contemporary clinical outcomes and identify triggers for arrhythmias or sudden death in an international cohort of Timothy Syndrome (TS) patients including those with novel TS-associated CACNA1C mutations.

BACKGROUND: TS is an extremely rare genetic disorder of the L-type cardiac channel Cav1.2 encoded by CACNA1C. The syndrome is characterized by multisystem abnormalities consisting of QT prolongation, congenital heart defects, syndactyly, facial dysmorphism, and neurological symptoms.

METHODS: Patients diagnosed with TS between January 1, 1994, and April 1, 2016, from 12 international tertiary care pediatric centers were included in this retrospective study. Data were gathered via survey from the patients' electrophysiologists.

RESULTS: Seventeen patients diagnosed with TS were identified. Length of follow-up was 4.9 years (range 3.0 to 19.0 years). Mean QTc was 640 ms (range 500 to 976 ms). All patients were treated with beta-blockers; 13 patients (76%) were also treated with an implantable defibrillator. Eleven patients experienced an episode of aborted cardiac arrest, 6 associated with general anesthesia and 2 with hypoglycemia. Four patients died suddenly due to ventricular fibrillation, 2 of whom had associated hypoglycemia.

CONCLUSIONS: This study shows that mortality in TS patients is due to multifactorial mechanisms, which include ventricular arrhythmias, pulseless electrical activity, and hypoglycemia. A simple nomenclature for ongoing studies of TS and related syndromes is described. A worldwide prospective registry is needed for continued exploration of this syndrome.


Background: The objective was to evaluate the reliability and validity of the self-report Symptom Screening in Pediatrics Tool (SSPedi) from the perspective of children with cancer and pediatric hematopoietic stem cell transplant (HSCT) recipients.

Methods: In this multicenter study, respondents were children age eight to 18 years who had cancer or had received HSCT, and their parents. Two different child respondent populations were targeted. More symptomatic respondents were receiving active treatment for cancer, admitted to the hospital, and expected to be in the hospital three days later. Less symptomatic respondents were in maintenance therapy for acute lymphoblastic leukemia or had completed cancer therapy. Children completed SSPedi and then responded to validated self-report measures of mucositis, nausea, pain, and global quality of life. Children in the more symptomatic group repeated SSPedi and a global symptom change scale three days later. Parent proxy-report was optional. Reliability was evaluated using intraclass correlations while convergent validity was evaluated using Spearman correlations.

Results: Of 502 children enrolled, 302 were in the more symptomatic group and 200 were in the less symptomatic group. Intraclass correlation coefficients were 0.88 (95% confidence interval [CI] = 0.82 to 0.92) for test-retest reliability and 0.76 (95% CI = 0.71 to 0.80) for inter-rater reliability. The mean difference in SSPedi scores between more and less symptomatic groups was 7.8 (95% CI = 6.4 to 9.2). SSPedi was responsive to change in global symptoms. All hypothesized relationships among measures were observed.
Conclusions: SSPedi is a self-report symptom bother tool for children with cancer and HSCT recipients that is reliable, valid, and responsive to change. SSPedi can be used for clinical and research purposes. Future work should focus on integration into care delivery.


BACKGROUND: The Single Ventricle Reconstruction trial randomised neonates with hypoplastic left heart syndrome to a systemic-to-pulmonary-artery shunt strategy. Patients received care according to usual institutional practice. We analysed practice variation at the Stage II surgery to attempt to identify areas for decreased variation and process control improvement.

METHODS: Prospectively collected data were available in the Single Ventricle Reconstruction public-use database. Practice variation across 14 centres was described for 397 patients who underwent Stage II surgery. Data are centre-level specific and reported as interquartile ranges across all centres, unless otherwise specified. RESULTS: Preoperative Stage II median age and weight across centres were 5.4 months (interquartile range 4.9-5.7) and 5.7 kg (5.5-6.1), with 70% performed electively. Most patients had pre-Stage-II cardiac catheterisation (98.5-100%). Digoxin was used by 11/14 centres in 25% of patients (23-31%), and 81% had some oral feeds (68-84%). The majority of the centres (86%) performed a bidirectional Glenn versus hemi-Fontan. Median cardiopulmonary bypass time was 96 minutes (75-113). In aggregate, 26% of patients had deep hypothermic circulatory arrest >10 minutes. In 13/14 centres using deep hypothermic circulatory arrest, 12.5% of patients exceeded 10 minutes (8-32%). Seven centres extubated 5% of patients (2-40) in the operating room. Postoperatively, ICU length of stay was 4.8 days (4.0-5.3) and total length of stay was 7.5 days (6-10).

CONCLUSIONS: In the Single Ventricle Reconstruction Trial, practice varied widely among centres for nearly all perioperative factors surrounding Stage II. Further analysis may facilitate establishing best practices by identifying the impact of practice variation.


OBJECTIVES: To systematically investigate the relationship between motor and non-motor symptoms, and health-related quality of life (HR-QoL) in children and young adults with dystonia. METHODS: In this prospective observational cross-sectional study, 60 patients (6-25 years) with childhood-onset dystonia underwent a multidisciplinary assessment of dystonia severity (Burke-Fahn-Marsden Dystonia Rating Scale, Global Clinical Impression), motor function (Gross Motor Function Measure, Melbourne Assessment of Unilateral Upper Limb Function), pain (visual analogue scale), intelligence (Wechsler Intelligence Scale), executive functioning (Behavior Rating Inventory of Executive Function) and anxiety/depression (Child/Adult Behavior Checklist). Measures were analyzed using a principal component analysis and subsequent multiple regression to evaluate which components were associated with HR-QoL (Pediatric Quality of life Inventory) for total group, and non-lesional (primary) and lesional (secondary) subgroups.

RESULTS: Patients (29 non-lesional, 31 lesional dystonia) had a mean age of 13.6+/-.5.9 years. The principal component analysis revealed three components: 1) motor symptoms; 2) psychiatric and behavioral symptoms; and 3) pain. HR-QoL was associated with motor symptoms and psychiatric and behavioral symptoms (R^2=0.66) for the total sample and lesional dystonia, but in the non-lesional dystonia subgroup only with psychiatric and behavioral symptoms (R^2=0.51).

CONCLUSIONS: Non-motor symptoms are important for HR-QoL in childhood-onset dystonia. We suggest a multidisciplinary assessment of motor and non-motor symptoms to optimize individual patient management.


Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening hyperinflammatory syndrome requiring aggressive immunosuppressive therapy. Following 2 large international studies mainly targeting pediatric patients with familial disease and patients without underlying chronic or malignant disease, the HLH-94 protocol is recommended as the standard of care when using etoposide-based therapy by the Histiocyte Society. However, in clinical practice, etoposide-based therapy has been widely used beyond the study inclusion criteria, including older patients and patients with underlying diseases (secondary HLH). Many questions remain around these extended indications and published reports do not address several practical issues. To tackle these concerns, the HLH Steering Committee of the Histiocyte Society decided to issue guidance for use of the HLH-94 protocol. The group convened in a structured consensus finding process to define recommendations that are based largely on expert opinion backed up by available data from the literature. The recommendations address all main elements of HLH-94 including corticosteroids, cyclosporin, etoposide, intrathecal therapy, and hematopoietic stem cell transplantation (HSCT) and consider various forms of HLH and all age groups. Aspects covered include indications, applications, dosing, side effects, duration of therapy, salvage therapy, and HSCT. These recommendations aim to provide a framework to guide treatment decisions in this severe disease.


OBJECTIVES: This study sought to characterize risk in children with Wolff-Parkinson-White (WPW) syndrome by comparing those who had experienced a life-threatening event (LTE) with a control population.

BACKGROUND: Children with WPW syndrome are at risk of sudden death. METHODS: This retrospective multicenter pediatric study identified 912 subjects <=21 years of age with WPW syndrome, using electrophysiology (EPS) studies. Case subjects had a history of LTE: sudden death, aborted sudden death, or atrial fibrillation (shortest pre-excited RR interval in atrial fibrillation [SPERRI] of <=250 ms or with hemodynamic compromise); whereas subjects did not. We compared clinical and EPS data between cases and subjects.

RESULTS: Case subjects (n = 96) were older and less likely than subjects (n = 816) to have symptoms or documented tachycardia. Mean age at LTE was 14.1 +/- 3.9 years of age. The LTE was the sentinel symptom in 65%, consisting of rapidly conducted pre-excited atrial fibrillation (49%), aborted sudden death (45%), and sudden death (6%). Three risk components were considered at EPS: SPERRI, accessory pathway effective refractory period (APERP), and shortest paced cycle length with pre-excitation during atrial pacing (SPPCL), and all were shorter in cases than in control subjects. In multivariate analysis, risk factors for LTE included male sex, Ebstein malformation, rapid anterograde conduction (APERP, SPERRI, or SPPCL <=250 ms), multiple pathways, and inducible atrial fibrillation. Of case subjects, 60 of 86 (69%) had >=2 EPS risk stratification components performed; 22 of 60 (37%) did not have EPS-determined high-risk characteristics, and 15 of 60 (25%) had neither concerning pathway characteristics nor inducible atrioventricular reciprocating tachycardia.

CONCLUSIONS: Young patients may experience LTE from WPW syndrome without prior symptoms or markers of high-risk on EPS.


INTRODUCTION: Impossibility to place a gastrostomy and failed gastroesophageal reflux surgery with unsafe swallow are the main indications to Feeding Jejunostomy (FJ) in children. The aim of this study is to quantify the incidence of complications associated with FJ.

MATERIALS AND METHODS: A retrospective review of patients who had surgically inserted FJ between January 2009 and August 2013 at our institution was conducted. Data were obtained from medical records, operative notes, and radiology database, focusing on complications.

RESULTS: A total of 19 patients, average age 39.6 months (3-168 months), were treated during the study period. Indications to FJ were gastroesophageal reflux disease (GERD) associated with unsafe swallow in 12, esophageal
atresia in 5, and foregut dysmotility in 2. Seventeen FJ were inserted via laparotomy and 2 were laparoscopically assisted. In all cases, a serosal tunnel on the antimesenteric border was fashioned. No intraoperative complications were recorded. Tube dislodgement/blockage occurred on an average of 0.48 times per month in 18 out of 19 patients. The average radiation dose received for tube reinsertion/manipulation was 3.316 mSv/year/patient (0-10.66). Major postoperative complications occurred in 7 out of 19. After an average follow-up of 21 months, two have abandoned the use of FJ due to poor tolerance and three have fully weaned off. Two patients died due to unrelated causes.

CONCLUSION: FJ, as an alternative means for enteral feeding, may require multiple readmissions and exposure to radiological procedures. The high risk of severe complications should be considered when offering this procedure.

https://www.ncbi.nlm.nih.gov/pubmed/28561131


Objective: To conduct a systematic review of pain anxiety, pain catastrophizing, and fear of pain measures psychometrically established in youth with chronic pain. The review addresses three specific aims: (1) to identify measures used in youth with chronic pain, summarizing their content, psychometric properties, and use; (2) to use evidence-based assessment criteria to rate each measure according to the Society of Pediatric Psychology (SPP) guidelines; (3) to pool data across studies for meta-analysis of shared variance in psychometric performance in relation to the primary outcomes of pain intensity, disability, generalized anxiety, and depression.

Methods: We searched Medline, Embase, PsycINFO, and relevant literature for possible studies to include. We identified measures studied in youth with chronic pain that assessed pain anxiety, pain catastrophizing, or fear of pain and extracted the item-level content. Study and participant characteristics, and correlation data were extracted for summary and meta-analysis, and measures were rated using the SPP evidence-based assessment criteria.

Results: Fifty-four studies (84 papers) met the inclusion criteria, including seven relevant measures: one assessed pain anxiety, three pain catastrophizing, and three fear of pain. Overall, five measures were rated as "well established." We conducted meta-analyses on four measures with available data. We found significant positive correlations with the variables pain intensity, disability, generalized anxiety, and depression.

Conclusion: Seven measures are available to assess pain anxiety, pain catastrophizing, and fear of pain in young people with chronic pain, and most are well established. We present implications for practice and directions for future research.


AIM: People with cerebral palsy (CP) are often unable to express pain owing to cognitive or speech impairments. Reports that rely on observation can be inaccurate, because behaviours such as grimacing, common in people with spastic CP, resemble pain expressions. We examined preliminary validity and reliability of the revised Face, Legs, Activity, Cry, and Consolability (r-FLACC) scale in people with spastic CP.

METHOD: Forty-eight young people and adults (35 females, 13 males; mean [SD] age 29y 2mo [13y]) were video-recorded during a standard examination, rating their pain (0-10) afterwards. Two raters completed the r-FLACC using the video recordings. Intermter reliability was assessed with an unconditional cross-classified random-effects model and item response theory approach; Pearson correlations measured agreement between raters and participants.

RESULTS: Mean (SD) participant (n=48) pain scores were 2.48 (2.5) and mean (SD) r-FLACC scores were 1.46 (1.68). There was moderate agreement between raters (intraclass coefficient 0.41 and 0.57 respectively) but low agreement between participants and raters (r=0.26). There were no significant effects for raters (lay observers, nurses, physicians, and inexperienced raters).

INTERPRETATION: Results provide mixed support for the interrater reliability of the r-FLACC in people with spastic CP. WHAT T

His paper ADDS: The revised Face, Legs, Activity, Cry, and Consolability (r-FLACC) scale can be reliably used by experts and lay raters for people with spastic cerebral palsy (CP). Support is mixed for interrater reliability of the r-FLACC scale used with people with spastic CP.

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 72.9% (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 2.47 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.


Importance: Treatment delay for seizures can lead to longer seizure duration. Whether treatment delay is associated with major adverse outcomes, such as death, remains unknown. Objective: To evaluate whether untimely first-line benzodiazepine treatment is associated with unfavorable short-term outcomes.

Design, Setting, and Participants: This multicenter, observational, prospective cohort study included 218 pediatric patients admitted between June 1, 2011, and July 7, 2016, into the 11 tertiary hospitals in the United States within the Pediatric Status Epilepticus Research Group. Patients, ranging in age from 1 month to 21 years, with refractory convulsive status epilepticus (RCSE) that did not stop after the administration of at least 2 antiseizure medications were included. Patients were divided into 2 cohorts: those who received the first-line benzodiazepine treatment in less than 10 minutes and those who received it 10 or more minutes after seizure onset (untimely). Data were collected and analyzed from June 1, 2011, to July 7, 2016.

Main Outcomes and Measures: The primary outcome was death during the related hospital admission. The secondary outcome was the need for continuous infusion for seizure termination. Multivariate analysis of mortality controlled for structural cause, febrile RCSE, age, and previous neurological history (including previous RCSE events). Use of continuous infusions was additionally adjusted for generalized RCSE, continuous RCSE, and 5 or more administrations of antiseizure medication.

Results: A total of 218 patients were included, among whom 116 (53.2%) were male and the median (interquartile range) age was 4.0 (1.2-9.6) years. The RCSE started in the prehospital setting for 139 patients (63.8%). Seventy-four patients (33.9%) received their first-line benzodiazepine treatment in less than 10 minutes, and 144 (66.1%) received untimely first-line benzodiazepine treatment. Multivariate analysis showed that patients who received untimely first-line benzodiazepine treatment had higher odds of death (adjusted odds ratio [AOR], 11.0; 95% CI, 1.43 to infinity; \( P = .02\)), had greater odds of receiving continuous infusion (AOR, 1.8; 95% CI, 1.01-3.36; \( P = .047\)), had longer convulsive seizure duration (AOR, 2.6; 95% CI, 1.39-4.88; \( P = .003\)), and had more frequent hypotension (AOR 2.3; 95% CI, 1.16-4.63; \( P = .02\)). In addition, the timing of the first-line benzodiazepine treatment was correlated with the timing of the second-line (95% CI, 0.64-0.95; \( P < .001\)) and third-line antiseizure medications (95% CI, 0.25-0.78; \( P < .001\)).
Conclusions and Relevance: Among pediatric patients with RCSE, an untimely first-line benzodiazepine treatment is independently associated with a higher frequency of death, use of continuous infusions, longer convulsion duration, and more frequent hypotension. Results of this study raise the question as to whether poor outcomes could, in part, be prevented by earlier administration of treatment.


Objective: To investigate the prevalence and characteristics of neonates with life-limiting or life-threatening conditions who receive care focused exclusively on comfort.

Methods: Retrospective chart review of all newborn infants admitted to a level III perinatal center within a 5 year period.

Results: 1,777 of 9,878 infants (18.0%) had life-limiting or life-threatening conditions. 149 (1.5% of all neonates) were categorized as comfort care patients with death being anticipated within hours to weeks. 34.2% of comfort care patients suffered from conditions specific to the neonatal period, 28.9% were preterm infants at the limit of viability, and 22.8% were patients with congenital complex chronic conditions. In 80.5% of all comfort care patients treatment goals were re-directed toward a comfort-care-only regimen only once that life-prolonging therapies were demonstrated to be unhelpful. 136/149 comfort care patients (91.3%) died in hospital, while 13 (8.7%) were discharged home or into a hospice. Median age at death for comfort care patients was 3 days after birth (interquartile range 1-15.5 days), and delivery room death immediately after birth occurred in 37 patients (27.2%).

Conclusions: The vast majority of neonatal comfort care patients died in the hospital during the first week of life. However, almost one in 10 comfort care patients were discharged to home or hospice, suggesting that planning transition out of the NICU should be routinely discussed for all infants receiving comfort care.


OBJECTIVES: To demonstrate the efficacy of laser photobiomodulation (PBM) compared to that of placebo on severe oral mucositis (OM) in pediatric oncology patients. The primary objective was the reduction of OM grade (World Health Organization [WHO] scale) 7 days after starting PBM. Secondary objectives were reduction of pain, analgesic consumption, and incidence of side effects.

METHODS: One hundred and one children with WHO grade > 2 chemotherapy-induced OM were enrolled in eight Italian hospitals. Patients were randomized to either PBM or sham treatment for four consecutive days (days +1 to +4). On days +4, +7, and +11, OM grade, pain (following a 0-10 numeric pain rating scale, NRS) and need for analgesics were evaluated by an operator blinded to treatment.

RESULTS: Fifty-one patients were allocated to the PBM group, and 50 were allocated to the sham group. In total, 93.7% of PBM patients and 72% of sham patients had OM grade < 3 WHO on day +7 (P = 0.01). A significant reduction of pain was registered on day +7 in the PBM versus sham group (NRS 1 [0-3] vs. 2.5 [1-5], P < 0.006). Reduced use of analgesics was reported in the PBM group, although it was not statistically significant. No significant adverse events attributable to treatment were recorded.

CONCLUSIONS: PBM is a safe, feasible, and effective treatment for children affected by chemotherapy-induced OM, as it accelerates mucosal recovery and reduces pain.


Cirrhotic Cardiomyopathy (CCM), a comorbidity of end-stage cirrhotic liver disease, remains uncharacterized in children, largely due to a lack of an established pediatric definition. The aim of this retrospective cohort analysis is to derive objective 2-dimensional echocardiographic (2DE) criteria to define CCM associated with biliary atresia (BA), or
family relationships. Resulted in decreased maternal anxiety, improved maternal positive reframing, and improved communication and infants with single-ventricle disease experience significant depression and anxiety prenatally. Early palliative care family relationships scores (effect size 0.46 and 0.41, respectively). In conclusion, these data show that mothers of positive reframing scores (p=0.03), and a positive change in PedsQL Family Impact Module communication and State-Trait Anxiety Index scores (-7.6 versus 0.3 in standard care, p=0.02), higher postnatal Brief Cope Inventory scores, and statistically significant differences between study groups. The early palliative care group had a decrease in prenatal to postnatal depression and anxiety scores (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.


Objectives: Supervised exercise interventions during inpatient care are feasible. The objective was to evaluate the usability of activity trackers and centralised monitoring to conduct a home-based exercise intervention during cancer treatment. The primary endpoint and confirmatory analysis was achievement of individual goals for daily steps, compared (A) in the intervention group (IG) over time and (B) between the IG and control group (CG). Secondary endpoints included achievement of goals for active minutes and effects on motor performance and health-related quality of life (hrQoL).

Methods: Forty patients treated for paediatric cancer (14.7+/-3.9 years) were included. The IG received a 6-8 week intervention during acute treatment (T1) and a 2-week intervention in transition to aftercare (T2). The CG only received the intervention at T2. Baseline tests to assess motor performance and physical activity were conducted prior to every intervention.

Results: In the primary confirmatory analysis, the IG significantly improved achievement of individual step goals (p=0.04) whereas group analyses did not reveal significant differences. Achievement of active minutes remained low (p=0.23). IG scored higher in hrQoL than CG (p<0.01) and percentage of children scoring below normative value in strength tests was higher in CG. Of all participants, 94% rated the intervention as meaningful and 80% as motivational.

Conclusions: Results of this study indicate that this intervention for home stays with centralised supervision is feasible and leads to increased achievement of individual step goals. Despite the positive effects on hrQoL, further strategies are needed to increase positive effects on motor performance.


Children with single-ventricle disease experience high mortality and complexity. 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.
Management of hypoplastic left heart syndrome (HLHS) is resource intensive. Heath care systems are pressured to provide value to patients by improving outcomes while decreasing costs. A single-center retrospective cohort of infants with HLHS who underwent Norwood procedure or hybrid Norwood from 2004 to 2014 and survived to first outpatient follow up were studied. The primary outcome was total cost through 12 months with a sub-analysis of patients with 60 months of data. Costs were calculated using internal cost accounting system and reported by cost center. Of the 152 HLHS patients identified, 69 met inclusion criteria. Stage I hospitalization (n = 69), with a median length of stay 34 days [interquartile range (IQR) 24-58 days], resulted in a median cost of $203,817 (IQR $136,236-272,453). Of survivors at 12 months (n = 55), the median cost was $369,393 (IQR $216,289-594,038) generated in part by a median of 67 (40-126 days) hospitalized days during that year. A subgroup analysis of patients who reached 60 months of age (n = 29) demonstrated a median total cost of $391,812 (IQR $293,801-577,443) and a median of 74 lifetime hospitalized days (IQR 58-116 days). High cost centers included intensive care (41%), non-ICU hospital (17%), operative services (11%), catheterization lab (9%), and pharmacy (9%). Using multiple regression analysis, significant drivers of cost included reoperation, length of hospitalization, low birthweight, and use of ECMO. Costs related to HLHS management are driven both by care-related complications such as surgical re-intervention and patient factors such as low birth weight.


BACKGROUND/OBJECTIVES: Probiotics are living microorganisms that confer a health benefit on the host when administered. This systematic review and meta-analysis investigates the efficacy and safety of probiotics in adult and paediatric patients diagnosed with cancer.

METHODS: A systematic review and meta-analysis was undertaken (PROSPERO registration: CRD42016050252). Randomised controlled trials (RCT), identified through screening multiple databases were included for analysis of efficacy. Non-randomised controlled trials and case reports were included for safety analysis. Outcomes included the reduction in the incidence and severity of diarrhoea, and adverse events. Where possible, data were combined for meta-analysis using a random-effects model. Planned subgroup analyses were not possible through marked heterogeneity of study characteristics.

RESULTS: Twenty one studies (N = 2982 participants) were included for assessment of efficacy. Probiotics may reduce the incidence of diarrhoea in patients with cancer [odds ratio (OR) = 0.52, 95% confidence interval (CI) 0.34-0.78, 95% prediction interval (PI) 0.3-0.92, I-sq 36.9%, 5 studies] and the duration of pyrexia [standardised mean difference 0.39 days, 95% CI 0.35-0.43, I-sq 0.01%, 5 studies]. Twenty five studies (N = 2242) were included in the safety analysis. Five case reports showed probiotic-related bacteraemia/fungaemia/positive blood cultures. Definitions and reporting of adverse events were variable and inconsistent.

CONCLUSIONS: There remain insufficient studies to assess the true effect of probiotics in people with cancer. Meta-analysis suggests probiotics may be beneficial but further studies are still required. Improved reporting of outcomes and adverse events in clinical trials are required to improve accuracy and confidence of conclusions drawn in future updates.
does USP6 independently induce activation of the IFN signaling mediators, JAK1 and STAT1, but it also renders Ewing sarcoma cells exquisitely responsive to exogenous IFNs, potentiating activation of STAT1 and STAT3. Furthermore, IFNbeta (a type I IFN) induces apoptosis specifically in USP6-positive but not USP6-negative Ewing sarcoma cells. Finally, apoptosis is mediated through the proapoptotic ligand TRAIL, which is synergistically induced by type I IFN and USP6.

Implications: These findings provide the first insights into USP6 functions in a clinically relevant malignant entity, and raise the possibility of using IFN for targeting USP6-positive Ewing sarcoma. Mol Cancer Res; 1-10. (c)2018 AACR.


Dystonic storm or status dystonicus is a life-threatening hyperkinetic movement disorder with biochemical alterations due to the excessive muscle contractions. The medical management can require pediatric intensive care unit admission and a combination of medications while the underlying trigger is managed. Severe cases may require general anesthesia and paralytic agents with intubation and may relapse when these drugs are weaned. Deep brain stimulation of the globus pallidum has been reported to terminate dystonic storm in several pediatric cases. We present a 10-year-old boy with a de novo GNAO1 mutation-induced dystonic storm who required a 2-month pediatric intensive care unit admission and remained refractory to all medical treatments. Deep brain stimulation was performed under general anesthetic without complication. His dyskinetic movements stopped with initiation of stimulation. He was discharged from the pediatric intensive care unit after 4 days. We present prospectively evaluated changes in dystonia symptoms and quality of life for a patient with GNAO1 mutation treated with deep brain stimulation.


OBJECTIVES: To determine, in preschool- and school-aged children with cerebral palsy (CP): (i) the prevalence of sleep disorders, including disorders of initiation and maintenance of sleep, and (ii) the association between child characteristics and sleep disorders.

METHODS: Children with CP aged 3-12 years were recruited from neurology clinics and a provincial CP registry. Caregivers completed the Sleep Disturbance Scale for Children (SDSC) and a questionnaire on sleep-related characteristics. Children’s medical information was collected from the registry and hospital records.

RESULTS: 150 children with CP (mean age +/- standard deviation: 6.9 +/- 2.9 years) completed the study (66 preschool-and 84 school-aged children). An abnormal total score on the SDSC was found in 20.7% of children (10.6% and 28.6% in preschool-and school-aged children, respectively). Overall, 44.0% of children had one or more sleep disorder (24.2% and 59.5% in preschool-and school-aged children, respectively), as determined by subscales of the SDSC. The most common sleep problem, disorders of initiation and maintenance of sleep, was found in 26.0% of children (18.2% of preschool- and 32.1% of school-aged children, respectively). Pain was the strongest predictor of having an abnormal total score and disorders of initiation and maintenance of sleep, with odds ratios (95% confidence intervals) of 6.5 (2.2-18.9) and 3.4 (1.3-9.3), respectively, adjusted for age group and degree of motor impairment.

CONCLUSIONS: Sleep disorders are prevalent in children with CP, with higher frequencies in school-aged as compared to preschool-aged children. Health care professionals caring for this population should routinely inquire about sleep problems and pain.


AIMS AND OBJECTIVES: To compare and evaluate the reliability, validity, feasibility, clinical utility, and nurses’ preference of the Premature Infant Pain Profile-Revised, the Neonatal Pain, Agitation, and Sedation Scale, and the Neonatal Infant Acute Pain Assessment Scale used for procedural pain in ventilated neonates.
BACKGROUND: Procedural pain is a common phenomenon but is undermanaged and underassessed in hospitalised neonates. Information for clinician selecting pain measurements to improve neonatal care and outcomes is still limited. DESIGN: A prospective observational study was used.

METHODS: A total of 1,080 pain assessments were made at 90 neonates by two nurses independently, using three scales viewing three phases of videotaped painful (arterial blood sampling) and nonpainful procedures (diaper change). Internal consistency, inter-rater reliability, discriminant validity, concurrent validity and convergent validity of scales were analysed. Feasibility, clinical utility and nurses’ preference of scales were also investigated.

RESULTS: All three scales showed excellent inter-rater coefficients (from 0.991-0.992) and good internal consistency (0.733 for the Premature Infant Pain Profile-Revised, 0.837 for the Neonatal Pain, Agitation, and Sedation Scale and 0.836 for the Neonatal Infant Acute Pain Assessment Scale, respectively). Scores of painful and nonpainful procedures on the three scales changed significantly across the phases. There was a strong correlation between the three scales with adequate limits of agreement. The mean scores of the Neonatal Pain, Agitation, and Sedation Scale for feasibility and utility were significantly higher than those of the Neonatal Infant Acute Pain Assessment Scale, but not significantly higher than those of the Premature Infant Pain Profile-Revised. The Neonatal Pain, Agitation, and Sedation Scale was mostly preferred by 55.9% of the nurses, followed by the Neonatal Infant Acute Pain Assessment Scale (23.5%) and the Premature Infant Pain Profile-Revised (20.6%).

CONCLUSIONS: The three scales are all reliable and valid, but the Neonatal Pain, Agitation, and Sedation Scale and the Neonatal Infant Acute Pain Assessment Scale perform better in reliability. The Neonatal Pain, Agitation, and Sedation Scale appears to be a better choice for frontier nurses to assess procedural pain in ventilated neonates based on its good feasibility, utility and nurses’ preference.

RELEVANCE TO CLINICAL PRACTICE: Choosing a valid, reliable, feasible and practical measurement is the key step for better management of procedural pain for ventilated newborns. Using the right and suitable tool is helpful to accurately identify pain, ultimately improve the neonatal care and outcomes.


BACKGROUND: Wilms tumor (WT) is the most common renal tumor in children. We describe the outcomes for patients with WT that metastasized to bone (WTBM) to assist in decision making for these uncommon patients.

PROCEDURE: We retrospectively reviewed the research records of patients identified with WTBM from the National Wilms Tumor Study (NWTS 1-5) database. We then related overall survival (OS) to histology, chemotherapy, radiation therapy to bone, location of metastasis, and when bone metastasis presented.

RESULTS: Thirty-eight of 8609 patients enrolled on NWTS 1-5 (0.44%) developed bone metastasis. Bone metastasis most commonly first occurred at progression or relapse (29/38, 76%). Five of thirty-eight survived (13%) with the 5-year OS following presentation of bone metastasis of 14.3% (95% CI: 2.7-25.8%). The primary cause of death was tumor (29/33, 88%). Of those who died, the median survival time was 10.9 months (range 0.49-61.4). Four of nine (44%) patients presenting at diagnosis and 3% (1/29) of patients presenting in progression or relapse survived (P = 0.0075). Nineteen percent (5/26) of patients with favorable histology and 0% (0/12) with anaplastic histology survived (P = 0.16). Of the five survivors, median follow-up was 14 years (range 6.7-23.8). Radiation to metastatic bone sites was recorded in three of five survivors. No consistent chemotherapeutic approach appeared to be associated with disease outcome.

CONCLUSION: Bone metastasis is rare in patients with WT, occurring more commonly in progression or relapse than at initial diagnosis. Patients with WTBM have poor prognosis. We could not identify a consistent chemotherapeutic strategy associated with survival.


Glioblastoma multiforme (GBM) is the most aggressive intracranial tumor and diffusely infiltrates the surrounding brain tissue. Despite their malignant nature, extraneural metastases from glioblastomas are rare with an estimated incidence of <2%. We present a case of a 9-year-old boy with exophytic brainstem GBM who developed cervical node metastases. He had undergone gross total excision of GBM in January 2017. Histopathological examination
confirmed the diagnosis of glioblastoma multiforme. The patient underwent chemotherapy and radiotherapy as per hospital protocol. He developed hydrocephalus after 3 months, which required ventriculoperitoneal shunt. Two more months later, he developed drowsiness and was found to have shunt dysfunction causing hydrocephalus and multiple enlarged cervical lymph nodes. Cerebrospinal fluid diversion and neck node biopsy were performed for the patient but he died. The histopathological examination of the neck node biopsy revealed metastases from glioblastoma. We report this case to create awareness regarding possibility of extraneural metastases even in pediatric brainstem glioblastoma.


RATIONALE: Holoprosencephaly is a structural malformation of the brain that results from the complete or incomplete noncleavage of the forebrain of the embryo into 2 hemispheres. We report a severe case of alobar holoprosencephaly diagnosed at 38 weeks, associated with cebaophyaly, microcephaly, and craniosynostosis.

PATIENT CONCERN: The main knowledge added by this case is the late ultrasound diagnosis and chromosomal analysis that revealed a very rare abnormality (45X/46,XX/47,XX) with mosaicism at chromosome 18.

DIAGNOSES: Investigation of the mother revealed nothing remarkable from clinical point of view and on laboratory tests. Ultrasonography identified a fetal biometry appropriate for gestational age, except for the head biometry and abdominal circumference, that were appropriate for less than the fifth percentile. Microcephaly, a large midline monovenetle, absent midline structures, cleft lip, cebaophyaly (hypotelorism, single-nostril nose), ethmocephaly (hypotelorism, interorbital proboscis) and craniosynostosis, were also present. Fetal magnetic resonance imaging of fetus revealed an absent midline structure, a central monovenetle, abnormal corpus calosum, and abnormal gyri.

INTERVENTIONS: A cesarean section at 38 weeks was indicated for fetal bradycardia and a female baby was delivered, with Apgar score 6, weight 2290g. After birth, the diagnosis of the fetus confirmed holoprosencephaly with facial anomalies and demonstrated repeated tonic-clonic seizure, severe respiratory failure, cyanosis, decreased muscle tone, palor, and apnea. Laboratory examination of the newborn revealed acidosis and a prolonged of prothrombin time. The neonate was treated for severe respiratory distress syndrome, with immediate intubation and resuscitation. Vitamin K, fresh frozen plasma, and antibiotics were also administered.

OUTCOMES: After delivery, exitus of the fetus occurred at 3 days and 18hours due to massive pulmonary hemorrhage. LESSONS: We described a case of alobar holoprosencephaly diagnosed at 38 weeks of gestation and associated with a rare chromosomal abnormality (45X/46,XX/47,XX) with mosaicism at chromosome 18. Emotional implications could have been less severe if the patient underwent regular ultrasonography allowing a diagnosis in the first or early second trimester.


OBJECTIVE: To identify factors impacting overall survival (OS) in children, adolescents, and young adults with high-risk renal tumors (HRRTs).

METHODS: The National Cancer Database was queried for patients 30 years old diagnosed with anaplastic Wilms tumors (AWT), clear cell sarcoma of the kidney (CCSK), or rhabdoid tumor of the kidney (RTK) between 2004 and 2013. Demographic, clinical, and OS data were abstracted. OS between groups was compared with a Kaplan-Meier curve. Univariate and multivariate survival analyses were performed.

RESULTS: A total of 349 patients were identified meeting criteria; 133 (38.1%) AWT, 120 (34.4%) CCSK, and 96 (27.5%) RTK. Patients with RTK were less likely to undergo surgery than those with AWT or CCSK (77.1% vs 94% vs 99%, P < .001) and less likely to receive chemotherapy (84.4% vs 96.2% vs 95%, P = .013) or radiation (52.1% vs 81.2% vs 86.7%, P < .001). Estimated 5-year OS was 76.1% (95% confidence interval [CI] 67.9-84.4) for AWT, 92.7% (95% CI 87.4-97.9) for CCSK, and 33.5% (95% CI 23.1-43.9) for RTK (P < .001). On multivariate analysis, AWT (HR 3.372, P = .032) and RTK histology (HR 12.595, P < .001) were significantly associated with worse OS, while receiving radiation (HR 0.43, P = .006) was associated with improved OS. LN positivity, margin status, and undergoing surgery were not. Analyzing the HRRTs individually, for AWT, undergoing surgery was associated with OS (HR 0.308, P = .031). For RTK, factors associated with OS included undergoing surgery (HR 0.209, P = .007) and radiation (HR 0.411, P = .008).
CONCLUSION: Within the HRRTs, RTK is associated with worse outcomes than either AWT or CCSK. Receiving radiation is significantly associated with improved outcomes, and surgery is important for those with AWT and RTK.


BACKGROUND: Data regarding health care resource utilization (HRU) in early childhood among children with congenital heart disease (CHD) are scarce. Therefore, we sought to describe the extent of HRU incurred among children with CHD in the first 5 years of life.

METHODS: This population-based retrospective cohort study included all children born between January 2005 and March 2014 in Alberta, Canada. We linked inpatient, outpatient, practitioner claims, and drug dispensing databases with vital statistics (birth and death registries).

RESULTS: In the first year of life, the cumulative hospitalization rate per 100 children was 335 (95% confidence interval: 312-360) for single ventricle (SV) children, 200 (194-206) for moderate-complex CHD, and 152 (149-156) for simple CHD vs 109 (108-109) among children without CHD (P < 0.001). The ambulatory-care visit rate per 100 children was 4871 (4780-4963) for SV, 2278 (2258-2299) for moderate-complex, and 1416 (1405-1426) for simple CHD vs 246 (246-247) for children without CHD (P < 0.001). The rates of physician claims and drug dispensing also demonstrated similar patterns. The median total hospitalization length of stay during the first year of life was 54 days (interquartile range: 26-95) in SV, 15 (4-39) in moderate-complex, and 6 (2-26) in simple CHD compared with 2 (1-3) among children without CHD (P < 0.001). These differences remained throughout the first 5 years of life, with children with CHD having consistently higher hospitalization rates and emergency department visit rates in every year of age compared with children without CHD.

CONCLUSIONS: Cumulative HRU is high among children with CHD in the first 5 years of life and increases with increasing CHD severity. Improving survival of SV lesions will require increasing resource allocation to this group.


Pontocerebellar hypoplasia type 1 (PCH1) is a major cause of non-5q spinal muscular atrophy (SMA). We screened 128 SMN1-negative SMA patients from Bulgaria for a frequent mutation -p.G31A in EXOSC3, and performed a literature review of all genetically verified PCH1 cases. Homozygous p.G31A/EXOSC3 mutation was identified in 14 Roma patients, representing three fourths of all our SMN1-negative Roma SMA cases. The phenotype of the p.G31A/EXOSC3 homozygotes was compared to the clinical presentation of all reported to date genetically verified PCH1 cases. Signs of antenatal onset of disease present at birth were common in all PCH1 sub-types except in the homozygous p.D132A/EXOSC3 patients. The PCH1 sub-types with early death (between ages 1 day and 17 months), seen in patients with p.G31A/EXOSC3 or SLC25A46 mutations have a SMA type 1-like clinical presentation but with global developmental delay, visual and hearing impairment, with or without microcephaly, nystagmus and optic atrophy. Mutations with milder presentation (homozygous p.D132A/EXOSC3 or VRK1) may display additionally signs of upper motor neuron impairment, dystonia or ataxia and die at age between 5 and 18 years. Other EXOSC3 mutations and EXOSC8 cases are intermediate - SMA type 1-like presentation, spasticity (mostly in EXOSC8) and death between 3 months and 5 years. There is no correlation between neurological onset and duration of life. We add marble-like skin and congenital laryngeal stridor as features of PCH1. We show that imaging signs of cerebellar and pontine hypoplasia may be missing early in infancy. EMG signs of anterior horn neuronopathy may be missing in PCH1 patients with SLC25A46 mutations. Thus, there is considerable phenotypic variability in PCH1, with some cases being more SMA-like, than PCH-like. Detailed clinical evaluation and ethnicity background may guide genetic testing and subsequent genetic counseling.


improve diagnosis and facilitate early intervention, we previously developed a newborn screening assay based on newly identified plasma bile acid biomarkers. Because the newborn screen had been validated using dried blood spots (DBS) from already diagnosed NPC1 patients, an unanswered question was whether the screen would be able to detect individuals with NPC1 at birth.  

METHODS: To address this critical question, we obtained the newborn DBS for already diagnosed NPC1 subjects (n=15) and carriers (n=3) residing in California, New York, and Michigan states that archive residual DBS in biorepositories. For each of the DBS, we obtained two neighbor controls - DBS from patients born on the same day and in the same hospital as the NPC1 patients and carriers. 3beta,5alpha,6beta-trihydroxycholanic acid (bile acid A) and trihydroxycholanic acid glycine conjugate (bile acid B) were measured in the DBS using a liquid chromatography-tandem mass spectrometry (LC-MS/MS) assay.

RESULTS: Bile acid B, the more specific biomarker for which the fully validated DBS assay was developed, was detected in 8/15 NPC1 patients, and elevated above the cut-off in 2/15 patients (the two samples with the shortest storage time). Bile acid B was detected in 2/2, 6/10, and 0/7 NPC1 samples that have been stored for <10.5 years, 13-20 years, and >20 years, respectively, indicating that the glycine conjugate is detectable in DBS but may have reduced long-term stability compared with bile acid A, the precursor trihydroxycholanic acid, which was elevated in 15/15 NPC1 subjects, but not in carriers and controls.

CONCLUSIONS: These results demonstrate that newborn screening for NPC1 disease is feasible using bile acid biomarkers.


BACKGROUND: Pain in adolescents with cancer is common and negatively impacts health-related quality of life. The Pain Squad+ smartphone app, capable of providing adolescents with real-time pain management support, was developed to enhance pain management using a phased approach (ie, systematic review, consensus conference and vetting, iterative usability testing cycles). A 28-day Pain Squad+ pilot was conducted with 40 adolescents with cancer to evaluate the feasibility of implementing the app in a future clinical trial and to obtain estimates of treatment effect.

OBJECTIVE: The objective of our nested qualitative study was to elucidate the perceptions of adolescents with cancer to determine the acceptability and perceived helpfulness of Pain Squad+, suggestions for app improvement, and satisfaction with the pilot study protocol.

METHODS: Post pilot study participation, telephone-based, semistructured, and audio-recorded exit interviews were conducted with 20 adolescents with cancer (12-18 years). All interviews were transcribed and independently coded by 2 study team members. Content analysis was conducted to identify data categories and overarching themes.

RESULTS: Five major themes comprising multiple categories and codes emerged. These themes focused on the acceptability of the intervention, acceptability of the study, the perceived active ingredients of the intervention, the suitability of the intervention to adolescents’ lives, and recommendations for intervention improvement.

CONCLUSIONS: Overall, Pain Squad+ and the pilot study protocol were acceptable to adolescents with cancer. Suggestions for intervention and study improvements will be incorporated into the design of a future randomized clinical trial (RCT) aimed at assessing the effectiveness of Pain Squad+ on adolescents with cancer health outcomes.


PURPOSE: The present study aimed to evaluate progression and prognosis according to the palliation method used in neonates and early infants aged 3 months or younger who were diagnosed with pulmonary atresia with ventricular septal defect (PA VSD) or tetralogy of Fallot (TOF) with severe pulmonary stenosis (PS) in a single tertiary hospital over a period of 12 years.

METHODS: Twenty with PA VSD and 9 with TOF and severe PS needed initial palliation. Reintervention after initial palliation, complete repair, and progress were reviewed retrospectively. RESULTS: Among 29 patients, 14 patients underwent right ventricle to pulmonary artery (RV-PA) connection, 11 palliative BT shunt, 2 central shunt, and 2 ductal stent insertion. Median age at the initial palliation was 13 days (1-98 days). Additional procedure for
pulmonary blood flow was required in 5 patients; 4 additional BT shunt operations and 1 RV-PA connection. There were 2 early deaths among patients with RV-PA connection, one from RV failure and the other from severe infection. Finally, 25 patients (86%) had a complete repair. Median age of total correction was 12 months (range, 2-31 months). At last follow-up, 2 patients had required reintervention after total correction; 1 conduit replacement and 1 right ventricular outflow tract (RVOT) patch enlargements.

CONCLUSION: For initial palliation of patients with PA VSD or TOF with severe PS, not only shunt operation but also RV-PA connection approach can provide an acceptable outcome. To select the most proper surgical strategy, we recommend thorough evaluation of cardiac anomalies such as RVOT and PA morphologies and consideration of the patient’s condition.


Several phenomena in contemporary perinatology create challenges for analyzing pregnancy outcomes. These include recent increases in iatrogenic delivery at late preterm and early term gestation, which are incongruent with the belief that stillbirth and neonatal death risks decrease exponentially with advancing gestational age. Perinatal epidemiologists have also puzzled over the paradox of intersecting birthweight-specific and gestational age-specific perinatal mortality curves for decades. For example, neonatal mortality rates among preterm infants of women who smoke are substantially lower than neonatal mortality rates among preterm infants of non-smoking women, whereas the reverse pattern occurs at term gestation. This mortality crossover is observed across several contrasts (for example, women with hypertensive disorders of pregnancy vs. normotensive women, older vs. younger women, twins vs. singletons) and outcomes (stillbirth, neonatal death, sudden infant death syndrome and cerebral palsy), and irrespective of how advancing “maturity” is defined (birthweight or gestational age). One approach proposed to address and explain these unexpected phenomena is the fetuses-at-risk model. This formulation involves a reconceptualization of the denominator for perinatal outcome rates from births to surviving fetuses. In this overview of the fetuses-at-risk model, we discuss the central tenets of the births-based and the fetuses-based formulations. We also describe the extension of the fetuses-at-risk approach to outcomes into and beyond the neonatal period and to a multivariable adaptation. Finally, we provide a substantive context by discussing biological mechanisms underlying the fetuses-at-risk model and contemporary obstetric phenomena that are better understood from that model than from one based on births.


Pain scales using faces are commonly used tools for assessing pain in children capable of communicating. However, some children require other types of pain scales because they have difficulties in understanding faces pain scales. The goal of this study was to develop and validate the “Pain Block” concrete ordinal scale for 4- to 7-year-old children. This was a multicenter prospective observational study in the emergency department. Psychometric properties (convergent validity, discriminative validity, responsivity, and reliability) were compared between the “Pain Block” pain scale and the Faces Pain Scale-Revised (FPS-R) to assess the validity of the “Pain Block” scale. A total of 163 children (mean age, 5.5 years) were included in this study. The correlation coefficient between the FPS-R and the Pain Block scale was 0.82 for all participants which increased with age. Agreement between the 2 pain scales was acceptable, with 95.0% of the values within the predetermined limit. The differences in mean scores between the painful group and nonpainful group were 3.3 (95% confidence interval, 2.6-4.1) and 3.8 (95% confidence interval, 3.1-4.6) for FPR-S and Pain Block, respectively. The pain scores for both pain scales were significantly decreased when analgesics or pain-relieving procedures were administered (difference in Pain Block, 2.4 [1.4-3.3]; and difference in FPS-R, 2.3 [1.3-3.3]). The Pain Block pain scale could be used to assess pain in 4- to 7-year-old children capable of understanding and counting up to the number 5, even if they do not understand the FPS-R pain scale.


OBJECTIVE: Factors predicting survival over time after pediatric intensive care unit (PICU) admissions are not fully understood. The primary aim of the current study was to investigate whether multiple admissions (MADM) compared to single PICU admissions (SADM) were associated with poor survival over time after being admitted to PICU
facilities. Our secondary aim was to investigate if the presence of a complex chronic condition (CCC) would further impair prognosis.

DESIGN: A closed cohort of all children up to 16 years of age admitted to the three PICUs in Sweden between 2008 and 2010 was prospectively collected and followed until 2012, providing survival data for at least one but up to four years of follow-up.

SETTING: Three Swedish tertiary referral centers for pediatric intensive care and extracorporeal membrane oxygenation (ECMO) care were used.

PATIENTS: In total, 3,688 Swedish children with 5,019 PICU admissions were included.

INTERVENTIONS: No interventions were conducted.

MEASUREMENTS: An extensive data set was recorded, including up to four-year survival information following first PICU admission. The patients were assigned to seven admission diagnostic groups, which were then divided into SADM or MADM groups. The difference in survival over time and mortality rates (MR) and mortality rate ratios (MRR) were calculated. SADM and MADM groups with and without an existing CCC were formed. The difference in survival over time between groups was calculated.

MAIN RESULTS: A highly significant difference in survival over time was noted between SADM and MADM patients (p<0.0001), which was intensified by the presence of a CCC. MADM patients with a CCC had the worst outcome, while SADM patients without a CCC had the best outcome. MADM patients with no CCC demonstrated decreased survival over time compared to SADM patients with a CCC. Survival over time was statistically worsened for patients with MADM compared to SADM for the following admission diagnostic groups: Cardiovascular, Gastrointestinal/Penal, Respiratory, Neurological, and Miscellaneous. The mortality rate (deaths/patient year of follow-up) during the time of follow-up was 0.023 for SADM and 0.062 for MADM patients. The mortality rate ratio (MRR) between these groups was 2.69.

CONCLUSION: Compared to single admissions, multiple admissions to PICU were associated with a significant decrease in survival over time in some but not all diagnostic groups. Regarding our secondary aim, we found that when the presence of a CCC is factored into the survival analysis, survival over time is further impaired.


Sudden unexplained cardiac death (SUCD) can occasionally occur in nonelderly patients with epilepsy, psychiatric disorders, or no medical history. This study was conducted to aim to analyze whether values of the biomarkers for heart failure are associated with the SUCD. Serum concentrations of N-terminal probrain natriuretic peptide, high-sensitivity C-reactive protein (hs-CRP), and tumor necrosis factor alpha were analyzed in 57 nonelderly patients with SUCD who was diagnosed at medicolegal autopsy. The subjects were divided into 3 subgroups according to the medical history: (1) epilepsy, (2) psychiatric disorders, and (3) no specific medical history. The results showed that serum hs-CRP levels were significantly high in patients with epilepsy (P = 0.01) or psychiatric disorders (P = 0.01) as compared with the controls. Also, significantly high concentrations of hs-CRP were observed in psychiatric patients with schizophrenia, compared with the controls (P = 0.003) or the other psychiatric diseases (P = 0.01). The level of N-terminal probrain natriuretic peptide and tumor necrosis factor alpha did not show a significant difference between the SUCD and the controls. These results might suggest the association between high serum hs-CRP levels and the potential impairment of the cardiac function before the fatal event.


OBJECTIVE: There has been controversy regarding the beneficial effects of calcium on myocardial contractility and the harmful effects on myocardial cells, especially in children. The aim of this study was to investigate an association between ionized calcium concentration (iCa) and outcomes for pediatric patients after cardiac surgery.
DESIGN: A retrospective, single-center study from May 2013 to December 2014. SETTING: Referral high-volume pediatric cardiac center in a tertiary teaching hospital.

PATIENTS: Patients <72 months old with congenital heart disease who underwent palliative or definitive surgery.

INTERVENTIONS: None.

MEASUREMENTS AND MAIN RESULTS: The association between pH-corrected iCa within 24 hours after surgery and intensive care unit (ICU) length of stay (ILOS) was defined as the primary outcome. The highest iCa (iCamax), lowest iCa (iCamin), and time-weighted average iCa (iCaave) were stratified and compared with the outcomes. The authors reviewed 5,468 ionized calcium measurements from 357 consecutive pediatric patients during the study period. One patient died at postoperative day 34 in the ICU. Significant differences in ILOS were observed among patients after cardiopulmonary bypass (CPB) according to iCaave, iCamax, and iCamin but not among patients without CPB. Patients with CPB and an iCaave value of 1.31-to-1.40 mmol/L, 1.41-to-1.50 mmol/L, 1.51-to-1.60 mmol/L, and >1.60 mmol/L stayed in the ICU for 7 (IQR 4-10) days, 8 (IQR 6-16) days, 10 (IQR 8-14) days, and 19 (IQR 12-38) days, respectively, which was significantly longer than the ILOS of 5 (4-8) days for patients with an iCaave of 1.21-to-1.30 mmol/L. Even after adjustment for other predictors of ILOS using multivariable analyses, there were significant relationships of ILOS with iCaave and iCamin values of >1.50 mmol/L among patients with CPB.

CONCLUSIONS: Higher iCa within 24 hours after congenital cardiac surgery using CPB was independently associated with longer LOS in the ICU.


BACKGROUND: There is little research on number of planned home deaths. We need information about factors associated with home deaths, but also differences between planned and unplanned home deaths to improve end-of-life-care at home and make home deaths a feasible alternative. Our aim was to investigate factors associated with home deaths, estimate number of potentially planned home deaths, and differences in individual characteristics between people with and without a potentially planned home death.

METHODS: A cross-sectional study of all decedents in Norway in 2012 and 2013, using data from the Norwegian Cause of Death Registry and National registry for statistics on municipal health and care services. We defined planned home death by an indirect algorithm-based method using domiciliary care and diagnosis. We used logistic regressions models to evaluate factors associated with home death compared with nursing home and hospital; and to compare unplanned home deaths and potentially planned home deaths.

RESULTS: Among 80,908 deaths, 12,156 (15.0%) were home deaths. A home death was most frequent in ‘Circulatory diseases’ and ‘Cancer’, and associated with male sex, younger age, receiving domiciliary care and living alone. Only 2.3% of home deaths were from ‘Dementia’. In total, 41.9% of home deaths and 6.3% of all deaths were potentially planned home deaths. Potentially planned home deaths were associated with higher age, but declined in ages above 80 years for people who had municipal care. Living together with someone was associated with more potentially planned home deaths for people with municipal care.

CONCLUSION: There are few home deaths in Norway. Our estimations indicate that even fewer people than anticipated have a potentially planned home death.


Recurrent 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 I² 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.


BACKGROUND: Exacerbation of hyperkinesia is a life-threatening complication of dyskinetic movement disorders, which can lead to multi-organ failure and even to death. GNAO1 has been recently identified to be involved in the pathogenesis of early infantile epileptic encephalopathy and movement disorders. Patients with GNAO1 mutations can present with a severe, progressive hyperkinetic movement disorder with prolonged life-threatening exacerbations, which are refractory to most anti-dystonic medication.

OBJECTIVE: The objective was to investigate the evolution of symptoms and the response to deep brain stimulation of the globus pallidus internus (GPI-DBS) in patients with different GNAO1 mutations.

METHODS: We report six patients presenting with global motor retardation, reduced muscle tone and recurrent episodes of severe, life-threatening hyperkinesia with dystonia, choreoathetosis, and ballism since early childhood. Five of them underwent GPI-DBS.

RESULTS: The genetic workup revealed mutations in GNAO1 for all six patients. These encompass a new splice site mutation (c.723+1G>T) in patient 1, a new missense mutation (c.610G>C; p.Gly204Arg) in patient 2, a heterozygous mutation (c.625>T; p.Arg209Cys) in patients 3 and 4, and a heterozygous mutation (c.709G>A; p.Glu237Lys) in patients 5 and 6. By intervention with GPI-DBS the severe paroxysmal hyperkinetic exacerbations could be stopped in five patients. One patient is still under evaluation for neuromodulation.

CONCLUSION: In complex movement disorders of unsolved etiology clinical WES can rapidly streamline pathogenic genes. We identified two novel GNAO1 mutations. GPI-DBS can be an effective and life-saving treatment option for patients with GNAO1 mutations and has to be considered early.


PurposeTo determine feasibility and utility of newborn screening for spinal muscular atrophy (SMA) in New York State.

MethodsWe validated a multiplex TaqMan real-time quantitative polymerase chain reaction assay using dried blood spots for SMA. From January 2016 to January 2017, we offered, consented, and screened 3,826 newborns at three hospitals in New York City and tested newborns for the deletion in exon 7 of SMN1.

ResultsNinety-three percent of parents opted in for SMA screening. Overall the SMA carrier frequency was 1.5%. We identified one newborn with a homozygous SMN1 deletion and two copies of SMN2, which strongly suggests the severe type 1 SMA phenotype. The infant was enrolled in the NURTURE clinical trial and was first treated with Spinraza at age 15 days. She is now age 12 months, meeting all developmental milestones, and free of any respiratory issues.

ConclusionOur pilot study demonstrates the feasibility of population-based screening, the acceptance by families, and the benefit of newborn screening for SMA. We suggest that SMA be considered for addition to the national recommended universal screening panel.


PURPOSE: Treatment strategies in palliation of pediatric cancer remain a significant challenge. In this study, we aimed to assess the efficacy and safety of a short course of hypofractionated radiation therapy (RT) for metastatic or recurrent childhood tumors.

METHODS AND MATERIALS: A total of 104 lesions in 62 pediatric patients with metastatic or recurrent cancer were treated with a short hypofractionation schedule (>1 but /<5 fractions; />3 Gy per fraction) between 2007 and 2017 in our institution. The primary endpoint was local control (LC). Other endpoints included treatment response, overall survival, progression-free survival, and toxicity. Toxicities were assessed using the Common Terminology Criteria for Adverse Events v.4.0.

RESULTS: The most common histologies were neuroblastoma, comprising 50 of the 104 lesions (48.1%); osteosarcoma, 17 lesions (16.4%); and Ewing sarcoma, 13 lesions (12.5%). A median total dose of 24 Gy was delivered in a median of 5 fractions. Of 104 lesions, 26 (25.0%) were treated with stereotactic body radiation therapy, 24 (23.1%) with intensity modulated RT, and 48 (46.2%) with 2-dimensional RT or 3-dimensional conformal RT. A complete or partial response was observed in 63 (60.6%) of lesions, and stable disease was observed in 34 (32.7%). At a median follow-up of 8.7 months, 21 local failures occurred (20.2%). The 1- and 2-year LC rates were 74% and 68%, respectively. LC was better for tumors without previous irradiation (83% vs 57% with previous RT; P = .004). LC rates did not differ between RT techniques or total biologically effective dose with alpha/beta ratio of 10 (BED10) (/>=30 vs >30 Gy). At the time of analysis, 38 deaths in the cohort of 62 patients (61.3%) were recorded. The 1-year progression-free survival and overall survival rates were 31% and 44%, respectively. Incidence of any grade >/=3 toxicity was 6.7% (7 of 104). No grade 5 events occurred.

CONCLUSIONS: A short hypofractionation scheme yields effective disease control and treatment response with a favorable side effect profile. Select pediatric patients with symptomatic metastases or recurrent disease can be considered for a short course of palliative RT.


Appropriate pain measurement relies on the use of valid, reliable tools. The aim of this study was to determine and compare the psychometric properties of 3 self-reported pain scales commonly used in the pediatric emergency department (ED). The inclusion criteria were children aged 6 to 17 years presenting to the ED with a musculoskeletal injury and self-reported pain scores >/=30 mm on the mechanical Visual Analogue Scale (VAS). Self-reported pain intensity was assessed using the mechanical VAS, Faces Pain Scale-Revised (FPS-R), and Colour Analogue Scale (CAS). Convergent validity was assessed by Pearson correlations and the Bland-Altman method; responsiveness to change was assessed using paired sample t tests and standardized mean responses; and reliability was estimated using relative and absolute indices. A total of 456 participants were included, with a mean age of 11.9 years +/- 2.7 and a majority were boys (252/456, 55.3%). Correlations between each pair of scales were 0.78 (VAS/FPS-R), 0.92 (CAS). Convergent validity was assessed by Pearson correlations and the Bland-Altman method; responsiveness to change was assessed using paired sample t tests and standardized mean responses; and reliability was estimated using relative and absolute indices. A total of 456 participants were included, with a mean age of 11.9 years +/- 2.7 and a majority were boys (252/456, 55.3%). Correlations between each pair of scales were 0.78 (VAS/FPS-R), 0.92 (CAS), and 0.79 (CAS/FPS-R). Limits of agreement (95% confidence interval) were -3.77 to 2.33 (VAS/FPS-R), -1.74 to 1.75 (VAS/CAS), and -2.21 to 3.62 (CAS/FPS-R). Responsiveness to change was demonstrated by significant differences in mean pain scores among the scales (P < 0.0001). Intraclass correlation coefficient and coefficient of repeatability estimates suggested acceptable reliability for the 3 scales at, respectively, 0.79 and +/- 2.29 (VAS), 0.82 and +/- 2.07 (CAS), and 0.76 and +/- 2.82 (FPS-R). The scales demonstrated good psychometric properties for children with acute pain in the ED. The VAS and CAS showed a strong convergent validity, whereas FPS-R was not in agreement with the other scales.


BACKGROUND: Status dystonicus (SD) is a life-threatening complication in which episodes of dystonic movements become increasingly frequent and severe, requiring urgent hospital admission, and can lead to respiratory, metabolic, and bulbar complications. Pharmacologic treatment has been the mainstay management for this complication; however, many refractory patients will still require further treatment. Deep brain stimulation (DBS) is an established therapeutic strategy that has been used for dystonia, and now it has been proposed to be used for SD.

METHODS: In this case series, we describe our experience with early DBS placement in 5 patients with SD to control symptoms that are refractory to pharmacologic therapy. In addition, we present a literature review of this therapy in the treatment of SD. RESULTS: Before discharge, symptomatic relief (decrease of dystonic movements
and resolution of abnormal postures) was evidenced in all patients with a median of 3 days (interquartile range, 1-7) after surgery was performed. A follow-up Unified Dystonia Rating Scale score and Burke-Fahn-Marsden rating scale motor subscale score, at 6 months after hospital discharge with values being inferior to 20 and 30, respectively, for all cases. None of the patients had a recurrence of SD in the last follow-up period.

CONCLUSIONS: DBS surgery is a suitable, versatile, reversible and adequate therapy in the treatment of SD that is refractory to initial pharmacologic treatment.


BACKGROUND: Pediatric high-grade brainstem gliomas are aggressive tumors with dismal prognoses. Large-scale studies are needed to further characterize these tumors and determine factors influencing cancer-specific mortality and survival at varying time points.

METHODS: We used the SEER (Surveillance Epidemiology and End Results) database to conduct a population-based study of pediatric patients with histologically confirmed anaplastic astrocytoma or glioblastoma tumors located within the brainstem. Multivariate analyses incorporating patient demographics, tumor characteristics, and treatments were used to determine predictors of cancer-specific mortality and survival at 6 months, 9 months, 1 year, and 2 years.

RESULTS: We included 154 patients from the SEER database: 72 patients with anaplastic astrocytoma (47%) and 82 (53%) with glioblastoma. Median survival for the entire cohort was 10.0 months. Glioblastoma histology, developmental stage, and large tumor size were significantly associated with cancer-specific mortality. Six-month, 9-month, 1-year, and 2-year survival was 75%, 57%, 42%, and 20%, respectively. Glioblastoma histology was associated with worsened survival at 6 months (odds ratio [OR], 0.19; P = 0.0081), 9 months (OR, 0.18; P < 0.001), 1 year (OR, 0.19; P < 0.001), and 2 years (OR, 0.14; P = 0.0056). Radiation therapy was associated with improved survival at 6 (OR, 8.53; P < 0.001) and 9 months (OR, 3.58; P = 0.0012) and 9 months (OR, 3.58; P = 0.0012) and 9 months (OR, 3.58; P = 0.0012) but not at 1 or 2 years. Radiation therapy was associated with improved survival in glioblastoma (9.0 vs. 3.0 months; P < 0.001).

CONCLUSIONS: This population-based study showed that glioblastoma histology is associated with a poor prognosis in pediatric patients with high-grade brainstem gliomas. Regardless of histology, radiation therapy improved survival at 6 and 9 months but not long-term.


AIM: Sudden unexpected death in infancy (SUDI) rates for Maori and Pacific infants remain higher than for other ethnic groups in New Zealand and bed-sharing is a major risk factor when there is smoking exposure in pregnancy. Sleep space programmes and Pepi-Pod baby beds require evaluation.

METHODS: Two hundred and forty Maori and Pacific women and infants were randomised 1:1, to the Pepi-Pod sleep space programme, or to a control group with ‘usual care’. When infants were under 2 weeks of age, baseline interviews occurred, followed up by interviews at 2 and 4 months of age to assess safe sleep knowledge, infant care practices and Pepi-Pod use and acceptability. All participants were offered a New Zealand Standard approved portable cot.

RESULTS: At baseline, 25% of babies did not have a baby bed. Knowledge of smoking and bed-sharing as SUDI risks improved at follow-up in both groups. One quarter regularly bed-shared at follow-up in both groups. Intention to bed-share was a strong predictor of subsequent behaviour. Pepi-Pods were regularly used by 46% at 2 months and 16% at 4 months follow-up.

CONCLUSIONS: Bed-sharing and knowledge improvement were similar irrespective of group. It is likely that the impact of the intervention was reduced because the control group received better support than ‘usual care’ and all participants had a baby bed. New Zealand SUDI rates have declined since sleep space programmes have been available. Sleep space programmes should be prioritised for those with modifiable SUDI risk.


OBJECTIVE: The objective of this research was to evaluate a cohort of children with both autism spectrum disorder (ASD) and drug-resistant epilepsy (DRE) after epilepsy surgery to determine predictors of best outcome.

METHODS: Retrospective chart review was done for 29 children ages 2 to 18 years with ASD and DRE who had neurosurgical intervention for seizure management over 15 years at one institution. All subjects had at least 1 year of follow-up. Data abstraction included demographic information, seizure diagnosis, treatment, investigations, surgical intervention, neuropsychological assessment, and outcome. Statistical analysis software (SAS) was used for statistical analysis. Engel classification was used to assess seizure outcome.

RESULTS: Fifteen subjects had resective surgery. Fourteen had palliative surgery with vagal nerve stimulator (VNS) insertion (13) and corpus callosotomy (1). Of the 29 subjects, 35% had class I outcome (all in the resective group). When combining all subjects (resective and palliative), 66% of subjects benefited with class I-III outcomes. In the total cohort, age at time of surgery was significant, with class I outcome more frequently seen in the younger age group when compared with classes II-IV (p=0.01).

CONCLUSION: A subset of children with ASD can benefit from resective surgery, and for those who are not candidates, a VNS can offer significant improvements in seizure control.


Introduction: Seventy percent of an estimated 10 million children less than five years of age in developing countries die each year of acute respiratory infections, diarrhoea, measles, malaria, malnutrition or a combination of these conditions. Children living with Human immunodeficiency virus (HIV) are at risk of diarrhoea because of drug interactions with antiretroviral therapy and bottle feeding. This may be aggravated by malnutrition and other infectious diseases which are frequent in children living with HIV. Objective: to evaluate treatment interventions for diarrhoea in HIV infected and exposed children.

Methods: A comprehensive search was conducted on 02 June 2016 to identify relevant studies for inclusion. We included randomised controlled trials of HIV infected or exposed children under 15 years of age with diarrhoea. Two authors independently selected studies for inclusion, assessed risk of bias (RoB) and extracted data using a pre-designed data extraction form. Results: We included two studies (Amadi 2002 and Mda 2010) that each enrolled 50 participants. The RoB was assessed as low-risk for both included studies. There was no difference in clinical cure and all-cause mortality between nitazoxanide and placebo for cryptosporidial diarrhoea in Amadi 2002. In Mda 2010, there was a reduction in duration of hospitalisation in the micronutrient supplement group (P < 0.005) although there was no difference in all-cause mortality.

Conclusion: There is low certainty evidence on the effectiveness of nitazoxanide for treating cryptosporidial diarrhoea and micronutrient supplementation in children with diarrhoea. Adequately powered trials are needed to assess micronutrients and nitazoxanide, as well as other interventions, for diarrhoea in HIV-infected and exposed children.


BACKGROUND: The Non-Communicating Children Pain Check List-Revised (NCCPC-R) is a clinical assessment tool used to assess and measure pain in children aged 3 to 18 years, with mental and intellectual disabilities, incapable to speaking. AIM: Aim of our study was to test the validity and reliability of the Italian version of the NCCPC-R in children with cognitive impairment, in order to obtain a valid tool for pain assessment in these children.

DESIGN: Prospective observational study. SETTING: Paediatric Outpatient of Physical and Rehabilitative Medicine Department, and Paediatrics, Child Neurology and Psychiatry Department, Sapienza University, Rome.

POPULATION: 55 non-communicating children, with severe intellectual disability, aged 3-18 years.

METHODS: The guidelines for "translation, adaptation, and validation of instruments or scales for cross-cultural healthcare research" were used to translate the scale, which was administered by the parents/caregivers twice for 2
consecutive days, in association with NRS (Numerical Rating Scale). The reliability of the scale was evaluated using the intra and inter-class correlation coefficient (ICCs); Cronbach alpha coefficient was used to test the internal validity of the scale; "Receiver Operating Characteristic" (ROC) curves were used to compare pain-free scores with pain scores, determining threshold scores; Pearson correlation between NCCPC-R and NRS values was measured.

RESULTS: The InterCC between the first and the second interviewer at T0 was 0.97, the IntraCC of the first interviewer at T0-T1 was 0.89, showing a high correlation; the Cronbach alpha coefficient at T0 was 0.97, showing a high scale's validity. Pearson correlation between NRS and NCCPC-R values at T0 was 0.54 showing a medium level of agreement (p<0.0001). AUC (area under the curve) obtained by ROC curve at T0 was 0.807 (p=0.001), with sensitivity 95.2 and specificity 55.6, while a T1 AUC was 0.814 (p<0.001), with sensitivity 86.49, specificity 78.57.

CONCLUSIONS: The Italian version of NCCPC-R scale could be used to assess pain in non-communicating patients with mental and intellectual disabilities, showing a good correlation when compared to the NRS. CLINICAL

REHABILITATION IMPACT: The use of NCCPC-R scale in daily life made parents/caregivers able to discriminate the presence/absence of pain in non-communicating children, based on the scores obtained with the questionnaire.


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.


Aortic aneurysms are a rare condition in children. Wiskott-Aldrich syndrome is a primary immunodeficiency characterized by infections, thrombocytopenia, and eczema. Aortitis and aneurysm formation seem to be progressive in patients with Wiskott-Aldrich syndrome. The risk of death from aneurysmal rupture in patients with Wiskott-Aldrich syndrome is high and surgery is required for resection of aneurysms. We report a case where a successful resection of a descending thoracic aneurysm. We present a-12 year-old child with this syndrome who underwent a one-stage descending aortic aneurysm repair under continuous visceral perfusion.Histologic examination showed the presence of an aortitis withgranulomatous inflammatory response and multinucleated cells.


PURPOSE: Though high numbers of children with cerebral palsy experience chronic pain, it remains under-recognized. This paper describes an evaluation of implementation supports and adoption of the Chronic Pain
Assessment Toolbox for Children with Disabilities (the Toolbox) to enhance pain screening and assessment practices within a pediatric rehabilitation and complex continuing care hospital.

METHODS: A multicomponent knowledge translation strategy facilitated Toolbox adoption, inclusive of a clinical practice guideline, cerebral palsy practice points and assessment tools. Across the hospital, seven ambulatory care clinics with cerebral palsy caseloads participated in a staggered roll-out (Group 1: exclusive CP caseloads, March-December; Group 2: mixed diagnostic caseloads, August-December). Evaluation measures included client electronic medical record audit, document review and healthcare provider survey and interviews.

RESULTS: A significant change in documentation of pain screening and assessment practice from pre-Toolbox (<2%) to post-Toolbox adoption (53%) was found. Uptake in Group 2 clinics lagged behind Group 1. Opportunities to use the Toolbox consistently (based on diagnostic caseload) and frequently (based on client appointments) were noted among contextual factors identified. Overall, the Toolbox was positively received and clinically useful.

CONCLUSION: Findings affirm that the Toolbox, in conjunction with the application of integrated knowledge translation principles and an established knowledge translation framework, has potential to be a useful resource to enrich and standardize chronic pain screening and assessment practices among children with cerebral palsy. Implications for Rehabilitation It is important to engage healthcare providers in the conceptualization, development, implementation and evaluation of a knowledge-to-action best practice product. The Chronic Pain Toolbox for Children with Disabilities provides rehabilitation staff with guidance on pain screening and assessment best practice and offers a range of validated tools that can be incorporated in ambulatory clinic settings to meet varied client needs. Considering unique clinical contexts (i.e., opportunities for use, provider engagement, staffing absences/turnover) is required to optimize and sustain chronic pain screening and assessment practices in rehabilitation outpatient settings.


OBJECTIVE: We previously developed the paper-based Symptom Screening in Pediatrics Tool (SSPedi) designed for paediatric cancer symptom screening. Objectives were to evaluate and refine the electronic mobile application (app) of SSPedi using the opinions of children with cancer.

METHODS: Participants were children 8-18 years of age with cancer. Participants completed electronic SSPedi on their own and then responded to semistructured questions to determine whether they found electronic SSPedi easy or difficult to complete and understand, understood and liked the app features (audio and animation), and understood previously difficult to understand concepts with the introduction of a help menu. After each group of 10 children, responses were reviewed to determine whether modifications were required.

RESULTS: 20 children evaluated electronic SSPedi. None found electronic SSPedi difficult to complete or understand. All children understood the app features and each of the 4 more difficult to understand concepts after using the help menu. 19 of 20 children thought the app was a good way to communicate with doctors and nurses.

CONCLUSIONS: We finalised an electronic version of SSPedi that is easy to use and understand with features specifically designed to facilitate child self-report. Future work will evaluate the psychometric properties of electronic SSPedi.


OBJECTIVE: Niemann-Pick disease type C (NP-C) is a rare, autosomal recessive, neurodegenerative disease associated with a wide variety of progressive neurological manifestations. Miglustat is indicated for the treatment of progressive neurological manifestations in both adults and children. Since approval in 2009 there has been a vast growth in clinical experience with miglustat. The effectiveness of miglustat has been assessed using a range of measures.

METHODS: Comprehensive review of published data from studies of cellular neuropathological markers and structural neurological indices in the brain, clinical impairment/disability, specific clinical neurological manifestations, and patient survival.
RESULTS: Cranial diffusion tensor imaging and magnetic resonance spectroscopy studies have shown reduced levels of choline (a neurodegeneration marker), and choline/N-acetyl aspartate ratio (indicating increased neuronal viability) in the brain during up to 5 years of miglustat therapy, as well as a slowing of reductions in fractional anisotropy (an axonal/myelin integrity marker). A 2-year immunoassay study showed significant reductions in CSF-calbindin during treatment, indicating reduced cerebellar Purkinje cell loss. Magnetic resonance imaging studies have demonstrated a protective effect of miglustat on cerebellar and subcortical structure that correlated with clinical symptom severity. Numerous cohort studies assessing core neurological manifestations (impaired ambulation, manipulation, speech, swallowing, other) using NP-C disability scales indicate neurological stabilization over 2-8 years, with a trend for greater benefits in patients with older (non-infantile) age at neurological onset. A randomized controlled trial and several cohort studies have reported improvements or stabilization of saccadic eye movements during 1-5 years of therapy. Swallowing was also shown to improve/remain stable during the randomized trial (up to 2 years), as well as in long-term observational cohorts (up to 6 years). A meta-analysis of dysphagia - a potent risk factor for aspiration pneumonia and premature death in NP-C - demonstrated a survival benefit with miglustat due to improved/stabilized swallowing function.

CONCLUSIONS: The effects of miglustat on neurological NP-C manifestations has been assessed using a range of approaches, with benefits ranging from cellular changes in the brain through to visible clinical improvements and improved survival.

https://www.ncbi.nlm.nih.gov/pubmed/30111334

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TRIAL REGISTRATION NUMBER: N/A.


OBJECTIVE: Different feeding strategies have been suggested to improve growth and survival of infants with hypoplastic left heart syndrome following stage 1 palliation. The study objective was to assess hospital mortality following stage 1 palliation among infants with hypoplastic left heart syndrome who had two feeding modalities, gastrostomy tube vs no gastrostomy tube.

DESIGN: Retrospective study design.

SETTING: Multicenter pediatric health information system database. PATIENT: About 4287 patients with hypoplastic left heart syndrome who underwent stage 1 Norwood procedure from 2004 through 2013. Infants who had gastrostomy tube with or without fundoplication procedure were identified and their clinical characteristics were compared.

INTERVENTION: None.

OUTCOMES MEASURES: The primary outcome was discharge hospital mortality following stage 1 palliation.

RESULTS: About 1214 patients who underwent stage 1 palliation had gastrostomy tube placement prior to hospital discharge. About 881 only had this procedure, while 333 patients also underwent fundoplication. Infants who had a gastrostomy tube placement vs no gastrostomy procedure had longer hospital stay, but significantly lower hospital mortality (5% vs 19%, P < .001). Hospital mortality was lower in infants who had only gastrostomy vs gastrostomy with fundoplication procedure (4% vs 8%, P = .004). In the multivariable analysis, gastrostomy procedure was associated with a higher likelihood of survival to hospital discharge (HR: 0.06, CI [0.04, 0.1]), whereas additional fundoplication procedure increased the risk of mortality (HR: 2.77, CI [1.52, 5.04]).

CONCLUSIONS: The gastrostomy procedure did not place infants with hypoplastic left heart syndrome at higher risk of mortality. These infants should be considered for gastrostomy tube placement if they had persistent difficulty in oral feeding following stage 1 palliation.


PURPOSE: To evaluate the consequences of using nebulized drugs in patients subjected to noninvasive ventilation (NIV) with total face mask (TFM) and helmet.

DESIGN: A descriptive analytical study of a prospective patient cohort was carried out.

AMBIT: Pediatric intensive care unit (PICU) of a tertiary hospital. PATIENTS: Consecutive sampling was used to include all patients admitted to the PICU and requiring NIV with helmet or TFM over a period of 29 months. No patients were excluded.

INTERVENTIONS: Nebulized treatment was added according to medical criteria.

VARIABLES OF INTEREST: Independent variables were age, sex, diagnosis, disease severity, ventilation parameters and nebulized drugs (if administered). Secondary outcomes were duration and failure of NIV, and length of PICU stay.

RESULTS: The most frequent diagnoses were bronchiolitis (60.5%) and asthma (23%). Patients received NIV for a median of 43h. Nebulized drugs were administered in 40% of the cases during NIV, and no adverse effects were registered. Using Bayesian statistics, the calculated probability of suffering an adverse effect was 1.3% with helmet and 0.5% with TFM (high density 95% probability intervals). Patients with helmet and nebulized therapy were in more
serious condition than those who did not receive nebulization; nevertheless, no differences were observed regarding
the need to change to bilevel modality. With TFM, PICU stay was shorter for the same degree of severity (p=0.033),
and the NIV failure rate was higher in patients who did not receive inhaled drugs (p=0.024).

CONCLUSIONS: The probability of suffering an adverse effect related to nebulization is extremely low when using a
helmet or TFM. Inhaled therapy with TFM may shorten PICU stay in some patients.


Severity Scale for Assessment of Nausea in Children with Abdominal Pain-Related Functional
Gastrointestinal Disorders." Children (Basel) 5(6).

The objective of this study was to develop a pediatric measure of chronic nausea severity, the Nausea Severity Scale
(NSS), and evaluate its reliability and validity in youth with abdominal pain-related functional gastrointestinal disorders
(AP-FGID). Pediatric patients (aged 11(-)17 years-old, n = 236) presenting to an outpatient clinic for evaluation of
abdominal pain completed the NSS, Children's Somatization Inventory (CSI), Functional Disability Inventory (FDI),
Abdominal Pain Index (API), Patient-Report Outcomes Measurement Information System (PROMIS), Anxiety and
Depression Scales and the Pediatric Rome III Questionnaire for FGIDs. The NSS demonstrated good concurrent,
discriminant, and construct validity, as well as good internal consistency. One-third (34%) of AP-FGID patients
reported experiencing nausea "most" or "every day" in the previous two weeks. The severity of nausea was higher in
females than males and correlated significantly with the severity of somatic symptoms, functional disability, anxiety,
and depression. The NSS is a valid and reliable measure of nausea in children with AP-FGID.


PI3K/Akt/GSK3 Pathway Under Chronic Neurodegenerative Conditions Triggered by Perinatal Asphyxia." Front Pharmacol 9: 335.

Perinatal asphyxia (PA) remains as one of the most important causes of short-term mortality, psychiatric and
neurological disorders in children, without an effective treatment. In previous studies we have observed that the
expression of different neurodegenerative markers increases in CA1 hippocampal area of 4-months-old male rats
born by cesarean section and exposed for 19 min to PA. We have also shown that a late treatment with 17beta
estradiol (daily dose of 250 mug/kg for 3 days) was able to revert the brain alterations observed in those animals.
Based on these previous results, the main aim of the present study was to explore the mechanism by which the
estrogenic treatment is involved in the reversion of the chronic neurodegenerative conditions induced by PA. We
demonstrated that estradiol treatment of adult PA exposed animals induced an increase in estrogen receptor (ER)
alpha and insulin-like growth factor receptor (IGF-1R) protein levels, an activation of the phosphatidylinositol 3-
kinase/Akt/glycogen synthase kinase 3 beta/beta-catenin signaling pathway and an increase in Bcl-2/Bax ratio in the
hippocampus in comparison to PA exposed animals treated with vehicle. Taking together, our data suggest that the
interaction between ERalpha and IGF-IR, with the subsequent downstream activation, underlies the beneficial effects
of estradiol observed in late treatment of PA.


Scaravilli, V., A. Zanella, V. Ciceri, M. Bosatra, C. Flandoli, A. La Bruna, S. Sosio, R. Parini, S. Gasperini, A.

BACKGROUND: Complications are common during anesthesia for patients with mucopolysaccharidoses. San
Gerardo Hospital (Italy) is a reference center for mucopolysaccharidoses with a dedicated pediatric anesthesia
service.

AIMS: This study aims to evaluate the safety of anesthesia for mucopolysaccharidoses patients, describe their
anesthetic management at our institution, and assess risk factors for complications.

METHODS: The anesthetic charts of mucopolysaccharidoses patients admitted from January 1999 to December
2014 were retrospectively analyzed. We retrieved patients' demographics; location and type of the procedure;
anesthetic approach airway management and occurrence of difficult intubation and complications and outcome at
hospital discharge. A generalized linear mixed model was performed to assess risk factors for complications and
difficult intubation.

RESULTS: Fifty-four consecutive children were included. The anesthetic charts of 232 procedures (52% radio-
diagnostics, 15% orthopedics, 15% ear-nose-throat surgery, 10% neurosurgery, and 8% general surgery) were
analyzed. Each patient underwent a median of 4 (1-6) procedures. The median age at the first procedure was 2 (1-5), and overall age was 5 (2-8) years old. One hundred and twenty-five (54%) procedures were performed in remote locations. General anesthesia was utilized for 100 (43%) procedures. No death was recorded. Twenty-one (9%) procedures had respiratory complications. Remote location anesthesia was associated with increased risk for complications (odds ratio 5.405 [1.355-28.571], P = .016). All planned intubations (n = 65) were successful. Nineteen (29%) of those were defined difficult. All emergency intubations (n = 3) failed and were rescued by laryngeal mask airways. Older age was associated with an increased risk of difficult intubation (OR 1.200 [1.019-1.436], P = .028).

CONCLUSION: Patients with mucopolysaccharidoses are at high risk for anesthesia-related complications. Remote location anesthesia is associated with increased risk for complications, and older age is associated with increased risk for difficult intubation.


PURPOSE: Pain is prevalent and affects functioning and quality of life of children with cerebral palsy (CP). However, pain in CP is under recognized. The International Classification of Functioning, Disability and Health (ICF) guides the selection of comprehensive chronic pain assessment tools. Our objectives were to identify measures addressing pain in children with CP, characterize the content of each measure using the ICF, and identify gaps and overlaps.

MATERIALS AND METHODS: Measures were identified from: (1) a systematic review of outcome measures (1998-2012) and (2) a scan of chronic pain measures (2013-2015). Included measures were those published in English, used in children and youth with CP, and contained an item/domain addressing pain. Constructs of the measures were linked to the ICF.

RESULTS: Overall, 31 measures addressing chronic pain in CP were included. Considerable variability was found in the degree to which their content represented the ICF. Most of pain measures address pain intensity and pain location (body functions) as opposed to functional impact of pain (activities and participation).

CONCLUSIONS: Functional dimensions are poorly represented in pain measures. Our findings may guide the selection of measures for research and clinical needs for comprehensive chronic pain management in children with CP. Implications for Rehabilitation Chronic pain is prevalent among children with cerebral palsy and significantly interfere with functional activities. To effectively manage chronic pain in children with cerebral palsy, measures capturing functional-based information need to be part of routine chronic pain assessment.


The neuronal ceroid lipofuscinoses are a class of inherited neurodegenerative diseases characterized by the accumulation of autofluorescent storage material. The most common neuronal ceroid lipofuscinosis has juvenile onset with rapid onset blindness and progressive degeneration of cognitive processes. The juvenile form is caused by mutations in the CLN3 gene, which encodes the protein CLN3. While mouse models of Cln3 deficiency show mild disease phenotypes, it is apparent from patient tissue- and cell-based studies that its loss impacts many cellular processes. Using Cln3 deficient mice, we previously described defects in mouse brain endothelial cells and blood-brain barrier (BBB) permeability. Here we expand on this to other components of the BBB and show that Cln3 deficient mice have increased astrocyte endfeet area. Interestingly, this phenotype is corrected by treatment with a commonly used GAP junction inhibitor, carbenoxolone (CBX). In addition to its action on GAP junctions, CBX has also been proposed to alter lipid microdomains. In this work, we show that CBX modifies lipid microdomains and corrects membrane fluidity alterations in Cln3 deficient endothelial cells, which in turn improves defects in endocytosis, caveolin-1 distribution at the plasma membrane, and Cdc42 activity. In further work using the NIH Library of Integrated Network-based Cellular Signatures (LINCS), we discovered other small molecules whose impact was similar to CBX in that they improved Cln3-deficient cell phenotypes. Moreover, Cln3 deficient mice treated orally with CBX exhibited recovery of impaired BBB responses and reduced autofluorescence. CBX and the compounds identified by LINCS, many of which have been used in humans or approved for other indications, may find therapeutic benefit in children suffering from CLN3 deficiency through mechanisms independent of their original intended use.

BACKGROUND: New prognostic markers are needed to identify patients with Ewing sarcoma (EWS) and osteosarcoma unlikely to benefit from standard therapy. We describe the incidence and association with outcome of circulating tumour DNA (ctDNA) using next-generation sequencing (NGS) assays.

METHODS: A NGS hybrid capture assay and an ultra-low-pass whole-genome sequencing assay were used to detect ctDNA in banked plasma from patients with EWS and osteosarcoma, respectively. Patients were coded as positive or negative for ctDNA and tested for association with clinical features and outcome.

RESULTS: The analytic cohort included 94 patients with EWS (82% from initial diagnosis) and 72 patients with primary localised osteosarcoma (100% from initial diagnosis). ctDNA was detectable in 53% and 57% of newly diagnosed patients with EWS and osteosarcoma, respectively. Among patients with newly diagnosed localised EWS, detectable ctDNA was associated with inferior 3-year event-free survival (48.6% vs. 82.1%; p = 0.006) and overall survival (79.8% vs. 92.6%; p = 0.01). In both EWS and osteosarcoma, risk of event and death increased with ctDNA levels.

CONCLUSIONS: NGS assays agnostic of primary tumour sequencing results detect ctDNA in half of the plasma samples from patients with newly diagnosed EWS and osteosarcoma. Detectable ctDNA is associated with inferior outcomes.

https://www.ncbi.nlm.nih.gov/pubmed/30131550


Objective Nocturnal hypoventilation (NH) is a complication of respiratory involvement in neuromuscular disorders (NMD) that can evolve into symptomatic daytime hypercapnia if not treated proactively with non-invasive ventilation. This study aimed to assess whether NH can be detected in the absence of other signs of nocturnal altered gas exchange.

Methods We performed nocturnal transcutaneous coupled (tc) pCO2/SpO2 monitoring in 46 consecutive cases of paediatric-onset NMD with a restrictive respiratory defect (forced vital capacity < 60%). Nocturnal hypoventilation was defined as tcPCO2 > 50 mmHg for > 25% of recorded time, and hypoxemia as tcSpO2 < 88% for > 5 minutes. Daytime symptoms and bicarbonate were recorded after overnight monitoring.

Results Twenty-nine of 46 consecutive patients showed NH. Twenty-three patients did not have nocturnal hypoxemia and 18 were clinically asymptomatic. In 20 patients, PaCO2 in daytime blood samples was normal. Finally, 13/29 patients with NH had isolated nocturnal hypercapnia without nocturnal hypoxia, clinical NH symptoms, or daytime hypercapnia.

Conclusions Paediatric patients with NMD can develop NH in the absence of clinical symptoms or significant nocturnal desaturation. Therefore, monitoring of NH should be included among nocturnal respiratory assessments of these patients as an additional tool to determine when to commence non-invasive ventilation.


OBJECTIVE: Sisom is an interactive computer software program that allows children to rate the severity of their cancer symptoms. The study objectives were to describe the usability of Sisom in terms of ease of use, usefulness, and aesthetics and to offer suggestions for improvement.

METHOD: A multisite, descriptive study was conducted to describe the usability of Sisom. A purposive sample of children, ages 6 to 12 years, being treated for cancer was recruited. English- and French-speaking children completed the eight tasks in Sisom recorded using Morae software and provided input via an audiotaped interview. Data were downloaded, transcribed verbatim, and analyzed descriptively.
RESULTS: Thirty-four children with varying cancers participated. The majority of children liked Sisom and found it easy to use, found it to be helpful in expressing their symptoms, and were satisfied with the aesthetics. Some children provided suggestions for improvement to optimize Sisom use in Canada.

CONCLUSIONS: Children’s positive responses and desire to use Sisom again suggest that future research should be directed toward implementing and evaluating its effectiveness in a variety of settings.


AIM: To describe the clinical course for children with severe physical disability (SPD) in the 2 years prior to their death and to identify whether these children had palliative care involvement and advance care planning prior to death. To investigate whether there is a difference between children with progressive (PSPD) and non-progressive (NPSPD) aetiologies of SPD.

METHODS: A retrospective cohort analysis of 48 children with SPD who died between 1 January 2013 and 1 January 2015 at The Royal Children’s Hospital, Melbourne. Clinical charts were reviewed to collect data about the type of SPD, frequency and duration of hospital admissions, duration of palliative care involvement (if any) and presence of an advance care plan.

RESULTS: The majority of children were admitted in the 6 months before their death, and over a third were admitted to the intensive care unit. There was a significant increase in the frequency of hospital admissions as the study cohort approached death (P = 0.003). The majority of children with SPD were offered a referral to a palliative care service, with referrals more likely in children with PSPD (90%) compared to children with NPSPD (57%). While approximately 60% of children in each cohort had an advance care plan, there was a trend towards this being formalised earlier in children with PSPD (P = 0.09).

CONCLUSION: The increase in hospital admissions prior to death in children with SPD suggests an opportunity for greater consistency in offering advanced care planning and palliative care, especially to those with NPSPD.


PURPOSE: Data on end-of-life care practices in Asia are scarce. This study aimed to analyze the clinical factors associated with the recommended premedication protocol for mechanical ventilation withdrawal, in Taiwan.

METHODS: A total of 135 terminally ill patients who had mechanical ventilation withdrawn between 2013 and 2016 from a single medical center in Taiwan were enrolled. A premedication protocol of morphine and midazolam intravenous bolus was routinely recommended for the patients before mechanical ventilation withdrawal. Receipt of opioids and/or benzodiazepines during the withdrawal process was defined as full (both), partial (1 drug), and no (none) adherence. The clinical factors relevant to the adherence of recommended premedication protocol for mechanical ventilation withdrawal were analyzed.

RESULTS: Overall, 126 (93.3%) patients died, 8 (5.9%) patients were transferred to other institutions for further care, and 1 (0.7%) patient was discharged to home after mechanical ventilation withdrawal. The median survival time was 45 minutes, and 102 (75.6%) patients died within 1 day after the withdrawal process. The full, partial, and no adherence rates for premedication guideline were 17.8%, 40.0%, and 42.2%, respectively. The main diagnosis of cancer, receipt of hospice care, and preservation of spontaneous respiration were independent variables associated with the partial or full adherence to the premedication protocol.

CONCLUSION: Our data show that adherence to the premedication protocol for mechanical ventilation withdrawal in terminally ill patients was inadequate in Taiwan. Promoting hospice care and educating medical personnel in the compassionate withdrawal of mechanical ventilation, especially in patients with noncancer disease, are warranted.

BACKGROUND: Prior studies identifying symptom clusters used a symptom-centered approach to demonstrate the relationship among symptoms. Latent profile analysis (LPA) is a patient-centered approach that classifies individuals from a heterogeneous population into homogeneous subgroups, helping prioritize interventions to focus on clusters with the most severe symptom burden.

OBJECTIVE: The aim of this study was to use LPA to determine the best-fit models and to identify phenotypes of severe symptom distress profiles for adolescents with cancer who are undergoing treatment and in survivorship.

METHODS: We used estimated means generated by the LPA to predict the probability of an individual symptom occurring across on- and off-treatment groups for 200 adolescents with cancer.

RESULTS: The 3-profile solution was considered the best fit to the data for both on- and off-treatment groups. Adolescents on treatment and classified into the severe profile were most likely to report distress in appetite, fatigue, appearance, nausea, and concentration. Adolescents off treatment and classified into the severe profile were most likely to report distress in fatigue, pain frequency, and concentration.

CONCLUSIONS: Latent profile analysis provided a cluster methodology that uncovered hidden profiles from observed symptoms. This made it possible to directly compare the phenotypes of severe profiles between different treatment statuses.

IMPLICATIONS FOR PRACTICE: The co-occurring 13-item Symptom Distress Scale symptoms found in the severe symptom distress profiles could be used as items in a prespecified severe symptom distress cluster, helping evaluate a patient’s risk of developing varying degrees of symptom distress.


INTRODUCTION: Adequate postoperative analgesia in pediatric patients in the intensive care unit (ICU) matters, since untreated pain is associated with negative outcomes. Compared to routine postoperative patients, children undergoing hypothermia (HT) or extracorporeal membrane oxygenation (ECMO), or recovering after cardiac surgery likely display non-maturational differences in pharmacokinetics (PK) and pharmacodynamics (PD). These differences warrant additional dosing recommendations to optimize pain treatment. Areas covered: Specific populations within the ICU will be discussed with respect to expected variations in PK and PD for various analgesics. We hereby move beyond maturational changes and focus on why PK/PD may be different in children undergoing HT, ECMO or cardiac surgery. We provide a stepwise manner to develop PK-based dosing regimens using population PK approaches in these populations. Expert opinion: A one-dose to size-fits-all for analgesia is suboptimal, but for several commonly used analgesics the impact of HT, ECMO or cardiac surgery on average PK parameters in children is not yet sufficiently known. Parameters considering both maturational and non-maturational covariates are important to develop population PK-based dosing advices as part of a strategy to optimize pain treatment.


OBJECTIVES: This study tested the hypothesis that longer duration of any type of respiratory support is associated with an increased rate of death or neurodevelopmental impairment (NDI) at 18-22 months.

METHODS: Retrospective cohort study using the Generic Database of NICHD Neonatal Research Network from 2006 to 2010. Infants were born at <27 weeks gestational age with birth weights of 401-1000 g. Respiratory support received during initial hospitalization from birth was characterized as follows: no support, only invasive support, only non-invasive support or mixed invasive, and non-invasive support. The primary outcome was death after 24 h of life or NDI at 18-22 months corrected age.

RESULTS: In a cohort of 3651 infants, 1494 (40.9%) died or had NDI. Cumulative respiratory support of any type beyond 60 days was associated with the likelihood of death or NDI. Infants who only received invasive support had the highest rate (89.1%), followed by those received mixed support (26.1%). Infants who received only non-invasive support had the lowest rate (7.7%). When compared to the only non-invasive support group, both invasive [OR 62.7
(95%CI 25.7, 152.6) and mixed [OR 6.1 (95%CI 2.6, 14.4)] support groups were significantly more likely to die or have NDI.

CONCLUSION: Prolonged respiratory support, whether invasive or non-invasive, is associated with increased odds of a poor outcome. The proportion of infants with a poor outcome increased in a dose dependent manner by two factors: the cumulative duration of respiratory support beyond 60 days, and the extent to which invasive support is provided.


Background: Compared with full-term infants, very preterm infants are more vulnerable to injury and long-term disability and are at high risk of death. The predictive value of ultrasound and imaging on the neurodevelopment is one of the hot topics. This study aimed to investigate the relationship between cranial ultrasound (cUS) variables and neurodevelopmental outcomes of very preterm infants.

Methods: Totally 129 very preterm infants (gestational age </=28 weeks) in neonatal intensive care unit of Hunan Children’s Hospital between January 2012 and November 2014 were included in this retrospective study. Serial cUS (weekly before discharge and monthly after discharge) was performed on the infants until 6 months or older. Magnetic resonance imaging (MRI) was performed on the infants at approximately the term-equivalent age. The mental developmental index (MDI) and psychomotor developmental index (PDI) were followed up until the infants were 24 months or older. The relationship between brain injury and MDI/PDI scores was analyzed. Results: The consistency rate between cUS and MRI was 88%. At the first cUS, germinal matrix hemorrhage (GMH) Grades 3 and 4, hospitalization duration, and weight are significantly correlated with MDI/PDI and prognosis (MDI: odds ratio [OR] = 8.415, 0.982, and 0.042, P = 0.016, 0.000, and 0.004; PDI: OR = 7.149, 0.978, and 0.012, P = 0.025, 0.000, and 0.000, respectively). At the last cUS, gestational age, extensive cystic periventricular leukomalacia (c-PVL), and moderate and severe hydrocephaly are significantly correlated with MDI (OR = 0.292, 60.220, and 170.375, P = 0.004, 0.000, and 0.000, respectively). Extensive c-PVL and moderate and severe hydrocephaly are significantly correlated with PDI (OR = 76.861 and 116.746, P = 0.003 and 0.000, respectively).

Conclusions: Very premature infants with GMH Grades 3 and 4, short hospitalization duration, and low weight have low survival rates and poorly developed brain nerves. Cerebral palsy can result from severe cerebral hemorrhage, moderate and severe hydrocephaly, and extensive c-PVL. The sustained, inhomogeneous echogenicity of white matter may suggest subtle brain injury.

Pain and Symptom Assessment and Relief


Internet-delivered cognitive-behavioral therapy (iCBT) is a promising treatment for chronic pain among youth, but effect sizes are small, and strategies aimed at enhancing treatment effects are needed. Participants’ engagement with the program may be an important factor in determining treatment outcomes. The primary aim of the current study was to examine the relationship between treatment engagement and treatment outcomes. Secondly, we sought to characterize participant engagement in an iCBT program for adolescents with chronic pain and their parents. Participants included 134 adolescents randomized to the intervention arm of a controlled trial examining iCBT for chronic pain. Overall engagement with the intervention by adolescents and parents was high. Parental engagement (number of modules completed by parents and number of parent logins) predicted adolescent activity limitations change scores at post-treatment. Contrary to our expectation, adolescent treatment engagement was not predictive of treatment outcomes. Results indicate that parental engagement with the program may be an important predictor of treatment outcomes. Further research is needed to better understand influences of treatment engagement on outcomes in iCBT for youth.


AIMS: To identify and synthesize evidence regarding the knowledge and attitudes of nurses, and barriers and facilitators to effective pain assessment and management in infants and children.

BACKGROUND: Pain among children is a common experience. Relief from pain is a fundamental human right, yet hospitalized children continue to experience unrelieved pain. Provision of effective pain management is an integral part of the nurse’s role.

METHODS: Guided by Whittemore & Knaff’s five-stage framework, primary peer-reviewed studies published in English between 2000 and 2018 were searched using CINAHL, PubMed, ProQuest, PsyCINFO and Scopus. The initial search yielded 292 papers. Twenty-seven papers were included in this review: quantitative (n = 18), qualitative (n = 5) and mixed-methods (n = 4).

RESULTS: Findings showed that nurses internationally have poor knowledge and attitudes of basic pain assessment and management principles. Barriers to effective pain management include the absence of pain education and assessment tools, parents’ reluctance to report pain and insufficient prescription of analgesia by physicians. Facilitators for the effective management of pain include parental participation in care, trusting and respectful relationships between nurses and children, and adequate nurse-patient ratios.

CONCLUSION: The review findings suggest a need to improve education for nurses, doctors and the patients’ family in relation to paediatric pain management, communication and interprofessional collaborations. There is a need to maximize facilitators and overcome barriers, such as those identified in this review, to ensure the quality of paediatric pain management.

IMPLICATIONS FOR NURSING AND HEALTH POLICY: Nursing and health policy should mandate the prioritization of paediatric pain management and the clinical roles and responsibilities of the interdisciplinary team members. Undergraduate, postgraduate and in-service education for nurses and other health professionals should also address paediatric pain management. In-service education on paediatric pain management should be compulsory for all health professionals caring for children.


PURPOSE: The use of celiac plexus block (CPB) for abdominal pain has been extensively reported in adults. However, pediatric literature is limited to three single case reports and a series of three cases. This study evaluated the effectiveness of CPB in children and young adults (aged 8-20 years) with abdominal malignancies.

METHODS: Pain outcomes after CPB were evaluated in four children and young adults with cancer. Mean daily pain score (PS, 0-10) and morphine consumption (intravenous morphine equivalent daily [MED], mg/kg/day) before and after CPB were used to assess effectiveness.

RESULTS: Mean daily PS reduced after CPB in all patients. In one patient, this reduction was sustained up to 6 months of follow-up, and analgesics were discontinued 1 week after CPB. The other three patients had limited survival (6, 16, and 37 days) after CPB. One patient had a PS of 0 over the last few days of life, but the MED was escalated from 0.74 before the block to 5.4 mg/kg/day at the end of life. In the other two patients, MED was lower during the first week after CPB than that before CPB (4.55 vs. 1.59 and 2.88 vs. 1.51 mg/kg/day, respectively). As these two patients had disease progression during their last days of life, the MED was increased to 4.75 and 263.9 mg/kg/day, respectively.

CONCLUSIONS: Our results suggest that CPB may contribute to reducing PS and MED. We observed the use of CPB rather late in the disease trajectory.


The use of bisphosphonates for pain control in children with cancer is not extensively studied. We retrospectively evaluated 35 children with cancer treated with intravenous bisphosphonates for pain management at a single institution from 1998 through 2015. We analyzed pain scores and opioid and adjuvant medication consumption before bisphosphonate administration, daily for 2 weeks, and at 3 and 4 weeks after administration. We also determined the time interval between diagnosis and first administration of bisphosphonates and duration of life after bisphosphonate administration. Mean pain scores were 2.45 (+/-2.96) and 0.75 (+/-1.69) before and 14 days after bisphosphonate administration, respectively (P = .25), and morphine equivalent doses of opioids were 5.52 (+/-13.35) and 5.27 (+/-9.77), respectively (P = .07). Opioid consumption was significantly decreased at days 4 to 8, days 11 to 12, and week 3 after first bisphosphonate administration. The median duration of life after first bisphosphonate administration was 80 days, indicating its use late in the course of treatment. Bisphosphonates did not significantly improve pain outcomes at 2 weeks, but opioid consumption was reduced at several time points during the first 3 weeks. The use of bisphosphonates earlier in the course of pediatric oncological disease should be evaluated in prospective investigations.


PURPOSE: The aim of this trial was to investigate the efficacy and safety of transcutaneous vagal nerve stimulation (t-VNS) in the palliative treatment of drug resistant epileptic patients ineligible for surgery.

METHODS: Twenty adult patients received four hours of t-VNS per day for six months (T1), followed by a two-month washout period (T2). The frequency and type of seizures recorded at T1 and T2 were compared with those occurring in the three months preceding study entry (T0). Responders (>30% reduction in the total number of seizures) subsequently received two hours of t-VNS per day for further six months (T3). All patients underwent electroencephalography (EEG) and completed the Quality of Life in Epilepsy questionnaire at baseline and T1.

RESULTS: At T1 six patients were considered responders. In these patients, at T3 the average reduction in seizure frequency was 60% compared to T0 (p=0.043), and 51% compared to T2 (p=0.043). Responders had more often seizures with falls (5 of 6; 83.3%) compared with non-responders (3 of 14; 21.4%) (p=0.010) and t-VNS reduced their frequency by a percentage ranging from 47.5 to 100%. There was no change in responders' EEG findings after stimulation. At the end of the trial, three responders continued t-VNS, one implanted VNS.

CONCLUSIONS: t-VNS had no or minimal side effects and significantly reduced seizures in about one third of the enrolled patients. Further studies should be planned to assess whether t-VNS is a suitable tool to predict the efficacy of implanted VNS.


AIMS: To determine the effects of sponge baths and swaddled bathing on premature infants’ vital signs, oxygen saturation levels, crying times, pain, and stress levels.

METHODS: This study was a clinical trial with a crossover design. Data were conducted in the neonatal intensive care unit of a public hospital in Denizli, Turkey. A total of 35 premature infants, who were born at 33-37 weeks gestation with a birth weight <1,500 g, were enrolled in the study. Two bathing methods were applied at 3-day intervals. Vital signs and oxygen saturation levels were measured before and at minutes 1, 5, 15, 30 after bathing. Infants’ bathing was video recorded to assess pain and stress behaviors. The pain and stress behaviors of infants were evaluated by independent observers. A significance level of .05 was used for all statistical analyses.

RESULTS: There were statistically significant differences between bathing methods on vital signs, oxygen saturation levels, and crying times. Levels of stress and pain according to bathing type were significantly higher in the sponge bath condition (p < .05).

LINKING EVIDENCE TO ACTION: Swaddled bathing has a positive effect on the infant’s vital signs, oxygen saturation levels, crying time, and level of stress and pain compared to the sponge bath condition. Swaddled bathing is a harmless and safe nursing practice.


Epidermolysis bullosa is a rare blistering skin disorder that is challenging to manage because skin fragility and repeated wound healing cause itching, pain, limited mobility, and recurrent infections. Cannabidiol, an active cannabinoid found in cannabis, is postulated to have antiinflammatory and analgesic effects. We report 3 cases of self-initiated topical cannabidiol use in patients with epidermolysis bullosa in an observational study. One patient was weaned completely off oral opioid analgesics. All 3 reported faster wound healing, less blistering, and amelioration of pain with cannabidiol use. Although these results demonstrate promise, further randomized, double-blind clinical trials are necessary to provide scientific evidence of our observed benefits of cannabidiol for the treatment of epidermolysis bullosa.


BACKGROUND: The provision of pediatric palliative care in Asia Pacific varies between countries and availability of essential medications for symptoms at the end of life in this region is unclear.

OBJECTIVE: To determine medications available and used in the management of six symptoms at the end of life among pediatric palliative care practitioners in Asia Pacific. To identify alternative pharmacological strategies for these six symptoms if the oral route was no longer possible and injections are refused.

DESIGN AND SETTING: An online survey of all Asia Pacific Hospice Palliative Care Network (APHN) members was carried out to identify medications used for six symptoms (pain, dyspnea, excessive respiratory secretions, nausea/vomiting, restlessness, seizures) in dying children. Two scenarios were of interest: (1) hours to days before death and (2) when injectables were declined or refused.

RESULTS: There were 54 responses from 18 countries. Majority (63.0%) of respondents were hospital based. About half of all respondents were from specialist palliative care services and 55.6% were from high-income countries. All respondents had access to essential anaglesics. Several perceived that there were no available drugs locally to treat the five other commonly encountered symptoms. There was a wide variation in preferred drugs for treating each symptom that went beyond differences in drug availability or formulations.
CONCLUSION: Future studies are needed to explore barriers to medication access and possible knowledge gaps among service providers in the region, so that advocacy and education endeavors by the APHN may be optimized.


BACKGROUND AND METHODS: Despite advances in health care, the majority of children undergoing cancer treatment experience pain, particularly in the home setting. Mobile health tools provide a promising avenue to deliver pain management education and information to parents of children receiving cancer treatment. The current study describes the development and formative evaluation of a novel intervention, Cancer-Tailored Intervention for Pain and Symptoms (C-TIPS), which provides empirically-based pharmacological and non-pharmacological pain management information and coping skills training to parents of pediatric cancer patients. C-TIPS is a web-based application including a tailoring algorithm, customization tools, guided diaphragmatic breathing training, relaxation practice, and educational material (COPE modules). Thirty parents of children undergoing chemotherapy treatment for cancer participated in this initial mixed methods pilot study. Participants completed quantitative measures assessing their stress and relaxation ratings and satisfaction with C-TIPS. Formative evaluation and qualitative data were collected using individual and group interviews.

RESULTS: Parents reported high satisfaction with both the educational and skills training modules of C-TIPS (p<0.001). Parent self-reported stress significantly reduced (p=0.004) and relaxation increased (p=0.05) following participation with the skills training module.

CONCLUSIONS: C-TIPS is a feasible and well-received web-based intervention that promises to improve pain management in children undergoing cancer treatment, improve stress management in parents, and increase parents' knowledge and understanding of their child's cancer treatment. Results from the current study will help make improvements to C-TIPS in preparation for a randomized-controlled trial of this innovative program.


BACKGROUND: Rural Tennessee, especially rural East Tennessee has seen a dramatic increase in rates of controlled drug prescriptions and controlled drug overdose deaths in recent years. However, little is known about the individual decisions to prescribe or continue prescriptions with relation to addiction concerns.

OBJECTIVES: The purpose of this study was to learn more about what factors lead to physicians' prescribing control drugs for non-cancer pain through the use of focus groups.

METHODS: A qualitative study, using focus groups, in five family medicine clinics in East Tennessee and Southwest Virginia. The investigators used a semi-structured interview guide designed to facilitate group discussions about prescription drug abuse and misuse.

RESULTS: There were four main themes identified by the focus groups: (1) prescribers' changing prescribing patterns over time; (2) factors that influence controlled drug prescribing; (3) use and barriers to using state prescription drug monitoring programs (PDMPs); (4) prescribing controlled drugs to women of childbearing age. Each theme had several subthemes.

CONCLUSIONS: The balance between treating the patient's symptoms and causing potential harm is a challenge. The patient's pain cannot be ignored, but the potential harm of opioid therapy is not taken lightly. As the public health concern of prescription drug abuse in rural Appalachia continues to spread, prescribers are aware of their connection to the problem, and ultimately the solution.


Objectives: The aim of this study was to conduct a randomized, controlled comparison of outcomes associated with parent/nurse-controlled analgesia (PNCA), with and without a basal (background) opioid infusion, with intravenous (IV) opioids intermittently administered by a nurse on an "as needed" basis (IV PRN) for postoperative pain management in children with developmental delay (DD).

Methods: Participants included children with DD expected to require IV opioids for at least 24 postoperative hours. Patients were randomized to one of three groups: PNCA with a basal infusion, PNCA without a basal infusion, or IV PRN opioids. Demographics, pain scores, opioid consumption, and frequency of side effects were collected beginning 12 hours after emerging from anesthesia to decrease the impact of anesthetic agents on outcomes.

Results: The 81 participants (median = 12.0, 9.0-15.0 years) were primarily Caucasian (74%) males (58%), with severe DD (69%) having spinal surgery (41%). The proportion of patients in each group with pain scores ≤3 vs ≥4 revealed no between-group differences in any epoch (P = 0.09-0.27). Patients in the PNCA with a basal group consumed significantly more opioid (median = 0.03 mg/kg/h morphine equivalents, 0.02-0.03 mg/kg/h) than the PNCA without a basal infusion. No difference was found between the PNCA without a basal (median = 0.01 mg/kg/h morphine equivalents, 0.00-0.02 mg/kg/h) and the PRN groups (median = 0.01 mg/kg/h morphine equivalents, 0.01-0.02 mg/kg/h). There were no statistically significant differences in side effects, with the exception that more children in the PNCA group required supplemental oxygen (P = 0.05).

Conclusions: Results suggest there may be no advantage to PNCA over PRN opioids in this patient population after the first 12 postoperative hours with regard to pain scores, opioid consumption, or side effects.


A high prevalence of sleep problems exists in children and adolescents with life-limiting conditions (LLC) and severe psychomotor impairment (SPMI). This study aimed to compare the impacts of various child-related (pain, epilepsy, repositioning, medical care) and environment-related (light, noise, TV/radio, open door) factors on sleep in this vulnerable population. Data were obtained through the &"Sleep Questionnaire for Children with Severe Psychomotor Impairment" (&quot;SNAKE&quot;) by proxy assessment. n = 212 children (mean age: 10.4 years) were included in the analyses. Logistic and linear regression models were used to compare child- and environment-related factors against the global rating of children's sleep quality, five SNAKE scales, children's sleep duration, and sleep efficacy. Pain increased the risk of sleeping poorly four-fold (OR (odds ratio) = 4.13; 95% CI (confidence interval): 1.87(-)9.13) and predicted four sleep problems as assessed by the SNAKE. Children who needed to reposition during the night were at three times greater risk of sleeping poorly (OR = 3.08; 95% CI: 1.42(-)6.69). Three of the five SNAKE scales were predicted through nocturnal repositioning. Repositioning and epilepsy predicted a reduced sleep duration and low sleep efficacy. None of the environment-related factors exhibited statistically significant results. This study emphasizes the urgent need for reliable pain detection in the context of sleep disturbances in severely ill children.


BACKGROUND: Pediatric epilepsy, including treatment-resistant forms, has a major effect on the quality of life, morbidity, and mortality of affected children. Interest has been growing in the use of medical cannabis as a treatment for pediatric epilepsy, yet there has been no comprehensive review of the benefits and harms of cannabis use in this population. In this systematic review, we will search for, synthesize, and assess the published and gray literature in order to provide usable and relevant information to parents, clinicians, and policy makers.

METHODS: We will perform a living systematic review of studies involving the use of cannabis to treat pediatric epilepsy. We will search the published and gray literature for studies involving children with any type of epilepsy taking any form of cannabis. Studies will be selected for inclusion by two independent reviewers. The primary outcome is seizure freedom. Secondary outcomes are seizure frequency, quality of life (child, caregiver), quality and quantity of sleep, status epilepticus, tonic-clonic seizures, death (all-cause, sudden unexpected death in epilepsy),
gastrointestinal adverse events (diarrhea, vomiting), and visits to the emergency room. The quality of each included study will be assessed. If data are sufficient in quantity and sufficiently similar, we will conduct pairwise random-effects meta-analysis. We will repeat the literature search every 6 months to identify studies published after the previous search date. Sequential meta-analysis will be performed as necessary to update the review findings.

DISCUSSION: Our review aims to provide a comprehensive and up-to-date summary of the available evidence to inform decisions about the use of cannabis in children with treatment-resistant epilepsy. The results of this review will be of use to parents, clinicians, and policy makers as they navigate this rapidly evolving area.

SYSTEMATIC REVIEW REGISTRATION: PROSPERO CRD42018084755.


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.


Nearly 20 years ago, standards were established for hospitals to assess and treat pain in all patients. Research continues to demonstrate evolving trends in the measurement and effective treatment of pain in children. Behavioral research demonstrating long-lasting effects of inadequate pain control during childhood supports the concepts of early and adequate pain control for children suffering from painful conditions in the acute care setting. The authors discuss pain concepts, highlighting factors specific to the emergency department, and include a review of evidence for pharmacologic and nonpharmacologic treatments.


Parents wish to reduce their child’s pain during medical procedures but may not know how to do so. We systematically reviewed the literature on parents’ experiences and information needs related to managing their child’s pain for common medical procedures. Of 2678 records retrieved through database searching, 5 were included. Three additional records were identified by scanning reference lists. Five studies were qualitative, and 3 were quantitative. Most took place in North America or Europe (n = 7) and described neonatal intensive care unit experiences (n = 5). Procedures included needle-related medical procedures (eg, venipuncture, phlebotomy, intravenous insertion), sutures, and wound repair and treatment, among others. Generally, parents desired being present during procedures, wanted to remain stoic for their child, and thought that information would be empowering and relieve stress but felt unsupported in taking an active role. Supporting and educating parents may empower them to lessen pain for their children while undergoing medical procedures.


The Lidocaine 5% plaster is licensed for the symptomatic relief of neuropathic pain associated with post-herpetic neuralgia in adult patients over 18 years of age. Studies in adults also demonstrate efficacy of Lidocaine 5% plasters in other neuropathic pain conditions. Case reports and experience suggested efficacy of Lidocaine 5% plasters in children and adolescents with localised neuropathic pain. Initiated by the Pain in Children Special Interest Group
BACKGROUND: Cerebral palsy occurs in up to 2.1 of every 1000 live births and encompasses a range of motor problems and movement disorders. One commonly occurring movement disorder amongst those with cerebral palsy is dystonia: sustained or intermittent involuntary muscle spasms and contractions that cause twisting, repetitive movements and abnormal postures. The involuntary contractions are often very painful and distressing and cause significant limitations to activity and participation. Oral medications are often the first line of medical treatment for dystonia. Trihexyphenidyl is one such medication that clinicians often use to treat dystonia in people with cerebral palsy.

OBJECTIVES: To assess the effects of trihexyphenidyl in people with dystonic cerebral palsy, according to the World Health Organization’s (WHO) International Classification of Functioning, Disability and Health (ICF) domains of impairment, activity and participation. We also assessed the type and incidence of adverse effects in people taking the drug. SEARCH METHODS: We searched CENTRAL, MEDLINE, Embase, eight other databases and two trials registers in May 2017, and we checked reference lists and citations to identify additional studies.

SELECTION CRITERIA: We included randomised controlled trials comparing oral trihexyphenidyl versus placebo for dystonia in cerebral palsy. We included studies in children and adults of any age with dystonic cerebral palsy, either in isolation or with the associated movement disorders of spasticity, ataxia, chorea, athetosis and/or hypotonia. We included studies regardless of whether or not the study authors specified the method used to diagnose dystonia in their study population. Primary outcomes were change in dystonia and adverse effects. Secondary outcomes were: activity, including mobility and upper limb function; participation in activities of daily living; pain; and quality of life.

DATA COLLECTION AND ANALYSIS: We used standard methodological procedures expected by Cochrane.

MAIN RESULTS: We identified one study, which was set in Australia, that met the inclusion criteria. This was a randomised, double-blind, placebo-controlled, cross-over trial in 16 children (10 boys and 6 girls) with predominant dystonic cerebral palsy and a mean age of 9 years (standard deviation 4.3 years, range 2 to 17 years). We considered the trial to be at low risk of selection, performance, detection, attrition, reporting and other sources of bias. We rated the GRADE quality of the evidence as low. We found no difference in mean follow-up scores for change in dystonia as measured by the Barry Albright Dystonia Scale (BADS), which assesses eight body regions for dystonia on a 5-point scale (0 = none to 4 = severe), resulting in a total score of 0 to 32. The BADS score was 2.67 points higher (95% confidence interval (CI) -2.55 to 7.90; low-quality evidence), that is, worse dystonia, in the treated group. Trihexyphenidyl may be associated with an increased risk of adverse effects (risk ratio 2.54, 95% CI 1.38 to 4.67; low-quality evidence). There was no difference in mean follow-up scores for upper limb function as measured by the Quality of Upper Extremity Skills Test, which has four domains that collectively assess 36 items (each scored 1 or 2) and produces a total score of 0 to 100. The score in the treated group was 4.62 points lower (95% CI -10.98 to 20.22; low-quality evidence), corresponding to worse function, than in the control group. We found low-quality evidence for improved participation (as represented by higher scores) in the treated group in activities of daily living, as measured by three tools: 18.86 points higher (95% CI 5.68 to 32.03) for the Goal Attainment Scale (up to five functional goals scored on 5-point scale -2 = much less than expected to +2 = much more than expected), 2.91 points higher (95% CI 1.01 to 4.82) for the satisfaction subscale of the Canadian Occupational Performance Measure (COPM); satisfaction with performance in up to five problem areas scored on a 10-point scale (1 = not satisfied at all to 10 = extremely satisfied), and 2.24 points higher (95% CI 0.64 to 3.84) for performance subscale of the COPM (performance in up to five problem areas scored on a 10-point scale (1 = not able to do to; 10 = able to do extremely well)). The study did not report on pain or quality of life.

AUTHORS’ CONCLUSIONS: At present, there is insufficient evidence regarding the effectiveness of trihexyphenidyl for people with cerebral palsy for the outcomes of: change in dystonia, adverse effects, increased upper limb...
function and improved participation in activities of daily living. The study did not measure pain or quality of life. There is a need for larger randomised, controlled, multicentre trials that also examine the effect on pain and quality of life in order to determine the effectiveness of trihexyphenidyl for people with cerebral palsy.


Limited knowledge exists of current pain management practices and supporting guidelines in Jordanian pediatric intensive care units. To determine the current pain management practices and the availability and content of practice guidelines in Jordanian pediatric intensive care units, we conducted a cross-sectional and multisite survey of four pediatric intensive care units in Jordan. A questionnaire was developed and orally administered over the phone or in person to head nurses or their nominees to capture pain management practices and the existence and content of guidelines. All units had written pain management guidelines that included pain assessment, documentation, and management. All four units used one or more pain assessment tools. In three units, pain management was considered multidisciplinary and routinely discussed on unit rounds. In two units, continuous infusion of intravenous opioids was used as well as sedatives and neuromuscular blockers for most ventilated patients. In the two other units, continuous intravenous infusion of opioids was not used and only sedatives were administered for patients on mechanical ventilation. In two units, there were no specific guidelines on the use of nonopioid analgesics, patient-controlled anesthesia, or the management of postoperative pain. No unit used an opioid or sedative withdrawal assessment tool or had pain management guidelines on the use of topical anesthetic agents or sucrose. Pain management practices and guidelines varied across the four units, suggesting that there is an opportunity for improvement in pain management in pediatric intensive care units in Jordan.


BACKGROUND: This was a subgroup analysis of age group, dexamethasone use, and very highly emetogenic chemotherapy (VHEC) use from a randomised, multicentre, double-blind, Phase 3 study of oral aprepitant in paediatric subjects.

METHODS: Subjects aged 6 months to 17 years scheduled to receive chemotherapeutic agents associated with at least moderate risk for emesis were randomly assigned to receive either aprepitant plus ondansetron (aprepitant regimen) or placebo plus ondansetron (control regimen); both could be administered with or without dexamethasone. This secondary analysis evaluated subjects stratified by pre-specified age groups, dexamethasone use, and VHEC use. The primary endpoint of this analysis was the proportion of subjects who experienced complete response (CR) during the delayed phase.

RESULTS: CR rates in the delayed phase were numerically higher with the aprepitant than the control regimen across all age categories, and reached significance for subjects aged 12-17 years (51% vs. 10%; P < 0.0001). In subjects receiving dexamethasone, CR was twice as high for the aprepitant versus control regimen in the 6 months to <2 year group (50% vs. 25%) and significantly higher in the 12-17 year group (40% vs. 7%, P < 0.05). CR was also significantly higher with aprepitant in the 6 months to <2 year and 12-17 year age groups who received VHEC. Similar proportions of subjects experiencing at least one adverse event were seen in both regimens across age categories.

CONCLUSION: A 3 day aprepitant regimen seemed effective and safe for prevention of chemotherapy-induced nausea and vomiting in paediatric subjects across subgroups (ClinicalTrials.gov NCT01362530).


INTRODUCTION: Pruritis after burn is one of the most common chronic complaints in burn survivors. Pruritus is often indistinguishable from neuropathic pain. There is a paucity of studies reporting the use of gabapentin and pregabalin to treat both pruritus and neuropathic pain. The purpose of this current study is to explore and document the effect of gabapentin and pregabalin in children and adolescent burn survivors.
METHODS: A retrospective review of charts and pharmacy records of gabapentin and pregabalin dispensed to control pruritus and/or pain was conducted for burn survivors up to 20 years of age. Data collected included medication doses, age and weight of patients, presence of neuropathic pain and pruritus, reported response to medication, and side effects of these medications. 136 individuals who received gabapentin, pregabalin, or both medications are included in the study. 112 received only gabapentin, none received only pregabalin, and 24 received both. All results are documented in mean+/−standard deviation (s.d.) dose/kg/day. 104 individuals experienced pruritus exclusively, two experienced neuropathic pain exclusively, and 30 experienced both. Use of medications was considered effective if the individuals reported pruritus or pain relief from the medication. The medication was considered safe if the individuals did not experience adverse side effects warranting discontinuation of the drugs. Medications were continued with dose adjustments if an individual reported minor side effects such as sedation or hyperactivity.

RESULTS: The average effective dose mg/kg/day for gabapentin and pregabalin was calculated for each of the three age groups (≤5years, 6-12 years, and >12years). The average effective dose of gabapentin was 23.9+/−10.3mg/kg/day for children ≤5years, 27.0+/−15.3mg/kg/day for children 6-12 years, and 34.1+/−15.7mg/kg/day for children >12years. The average effective dose of pregabalin was 6.5+/−3.5mg/kg/day for children 6-12 years and 4.7+/−1.6mg/kg/day for children >12years. One 5-year-old child received 3.7mg/kg/day of pregabalin. Note that for all patients in this study, pregabalin was added after an inadequate response to gabapentin. For individuals receiving both gabapentin and pregabalin, the maximum gabapentin failure dose for pruritus was 32.8+/−18.0mg/kg/day and for both pain and pruritus was 28.1+/−18.3mg/kg/day. For individuals treated with only gabapentin, 91.4% had an adequate response for pruritus, 100% for neuropathic pain, and 43.3% for both pruritus and pain. 100% of individuals treated with both gabapentin and pregabalin had an adequate response for pruritus and 88.2% had an adequate response for both pruritus and pain. Gabapentin was associated with hyperactivity in two individuals, and sedation in one individual. One individual reported nausea, vomiting, and headaches when taking both medications; this resolved when gabapentin was discontinued. One individual reported sedation while taking both medications.

CONCLUSION: Gabapentin and pregabalin are effective in relieving pruritus and neuropathic pain in most burn survivors. In some instances, these medications can be given together. Few individuals reported side effects.


A complex motor disorder is a combination of various types of abnormal movements that are associated with impaired quality of life (QOL). Current therapeutic options are limited. We studied the efficacy, safety, and tolerability of medical cannabis in children with complex motor disorder. This pilot study was approved by the institutional ethics committee. Two products of cannabidiol (CBD) enriched 5% oil formulation of cannabis were compared: one with 0.25% delta-9-tetrahydrocannabinol (THC) 20:1 group, the other with 0.83% THC 6:1 group. Patients aged 1 to 17 years (n = 25) with complex motor disorder were enrolled. The assigned medication was administered for 5 months. Significant improvement in spasticity and dystonia, sleep difficulties, pain severity, and QOL was observed in the total study cohort, regardless of treatment assignment. Adverse effects were rare and included worsening of seizures in 2 patients, behavioral changes in 2 and somnolence in 1.


RATIONALE: Pantotenate kinase-associated neurodegeneration (PKAN) is a rare autosomal recessive disease. Progressive motor symptoms such as dystonia and spasticity begin in childhood and relentlessly become incapacitating later in life. Treatments including anticholinergics and iron chelation are usually ineffective. Botulinum toxin type A (BoNT-A) is effective for adult patients with dystonia or spasticity.

PATIENT CONCERNS: We reported a 10-year-old female patient with advanced PKAN, manifesting as generalized dystonia and spasticity.

DIAGNOSIS: The patient was diagnosed with PKAN by a pediatric neurologist.

INTERVENTIONS: The patient received BoNT-A injection.

OUTCOMES: The effect was obvious at four weeks after the injection, with an improvement of 25% in Barry-Albright Dystonia Scale and 4% in Functional Independence Measure for Children score. Furthermore, there was a 3.8% reduction in Parenting Stress Index Short Form score.
and 8.3% improvement in Pain and Impact of Disability domain in the score of Cerebral Palsy Quality of Life for
Children.

LESSONS: BoNT-A injection was effective to improve functional independence and to alleviate stress of caregivers in
the patient with advanced PKAN.


Management Strategies Identified by Children With Cancer Using Draw-and-Tell Interviews." Oncol Nurs
Forum 45(3): 290-300.

PURPOSE: This cross-sectional study described how school-aged children with cancer represent their symptoms
and associated characteristics using draw-and-tell interviews.

PARTICIPANTS & SETTING: 27 children aged 6-12 years receiving treatment for cancer at the Cancer
Transplant Center at Primary Children’s Hospital, a tertiary pediatric hospital in Salt Lake City, Utah.

METHODOLOGIC APPROACH: Children participated in draw-and-tell interviews while completing drawings
depicting days when they felt well and days when they felt sick. Children’s drawings and accompanying explanations
were analyzed qualitatively.

FINDINGS: Children’s drawings related symptoms and the strategies children used to self-manage those symptoms.
Nausea, fatigue, pain, and sadness were the most frequently reported symptoms. Strategies to manage symptoms
most often included physical and psychosocial care strategies.

IMPLICATIONS FOR NURSING: Children with cancer were able to relate detailed descriptions of their symptoms
and symptom self-management strategies when presented with developmentally sensitive approaches. Healthcare
providers are well positioned to integrate arts-based approaches to symptom assessment and to support children in
implementing their preferred strategies to alleviate symptoms.


resources for health professionals: a systematic review with subset meta-analysis of educational

Online educational interventions are increasingly developed for health professionals and students, although graduate
and undergraduate medical curricula often contain limited information about how to assess and manage pain. This
study reviews the literature on the effectiveness of pain-related online educational resources. Studies were identified
through a search of Medline, PsychnFO, Web of Science, CINAHL, PubMed, Scopus, Cochrane Library, Google
Scholar, and OpenGrey databases. Search terms included 3 concept blocks: (1) type of intervention-online
education, computer-based, e-learning, web-based, and internet-based; (2) population-pediatrician, physician,
nurse, psychologist, and medical; and (3) outcome-pain*. Thirty-two studies (13 randomised controlled trials, 5
nonrandomised controlled trials, and 14 single-group pre-post studies) were included. Ten provided data for
inclusion in a series of between-groups meta-analyses. After intervention, participants receiving online instruction had
significantly greater knowledge compared with those receiving training as usual/alternative training (Hedges’ g =
0.80, 95% confidence interval [CI]: 0.12-1.49), and students had significantly greater skills compared with students
receiving training as usual (g = 1.34, CI: 0.38-2.30). No significant differences were found for
confidence/competence (g = 0.02, CI: -0.79 to 0.84) or attitudes/beliefs (g = 0.16, CI: -0.48 to 0.79). Although online
educational resources show promise in improving learner knowledge, considerable heterogeneity exists between
studies in quality, design, educational content, and outcomes. Furthermore, methodologically robust RCTs are
required to establish the effectiveness of online educational interventions and a greater understanding of the key
features of successful online resources, including cognitive interactivity. Few studies assessed health outcomes for
patients, remaining a major priority for future investigations.


Neonatal transport is a highly specialized medical service that shifts critically ill neonates between hospitals for on-
going care. In other words, it is an extension of the Neonatal Intensive Care Unit (NICU), which provides intensive
care to critical ill neonates during transport. Furthermore, pain assessment and management is a crucial element
during neonatal transport. However despite significant advances over the last 20 years in relation to our
understanding of pain mechanisms in the neonates, the immediate long and short term consequences of neonatal pain along with proliferation of pain assessment measures, there continues to be reports of neonates in a variety of settings suffering needlessly from acute, prolonged, persistent and chronic pain. The central focus of the present review article is to put light on the existing challenges accompanying neonatal pain assessment during transport.


BACKGROUND: Inter-patient variability in response to opioids is well known but a comprehensive definition of its pathophysiological mechanism is still lacking and, more importantly, no studies have focused on children. The STOP Pain project aimed to evaluate the risk factors that contribute to clinical response and adverse drug reactions to opioids by means of a systematic review and a clinical investigation on paediatric oncological patients.

METHODS: We conducted a systematic literature search in EMBASE and PubMed up to the 24th of November 2016 following Cochrane Handbook and PRISMA guidelines. Two independent reviewers screened titles and abstracts along with full-text papers; disagreements were resolved by discussion with two other independent reviewers. We used a data extraction form to provide details of the included studies, and conducted quality assessment using the Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies.

RESULTS: Young age, lung or gastrointestinal cancer, neuropathic or breakthrough pain and anxiety or sleep disturbance were associated to a worse response to opioid analgesia. No clear association was identified in literature regarding gender, ethnicity, weight, presence of metastases, biochemical or hematological factors. Studies in children were lacking. Between June 2011 and April 2014, the Italian STOP Pain project enrolled 87 paediatric cancer patients under treatment with opioids (morphine, codeine, oxycodone, fentanyl and tramadol).

CONCLUSIONS: Future studies on cancer pain should be designed with consideration for the highlighted factors to enhance our understanding of opioid non-response and safety. Studies in children are mandatory.

TRIAL REGISTRATION: CRD42017057740


BACKGROUND: Methadone is an attractive medication for treating children with advanced cancer with pain as it is the only long-acting opioid available as a liquid. However, it is not frequently used due to concerns about potential toxicities and side effects.

OBJECTIVE: Evaluate the efficacy and safety of methadone as the first long-acting opioid in children with advanced cancer. DESIGN: Retrospective chart review of 52 consecutive patients referred to Pediatric Supportive Care for pain management started on methadone as their first long-acting opioid. Data collected at baseline, follow-up visits #1 (F1) and #2 (F2) included child and parent-reported outcomes for various physical and psychological symptoms, opioid side effects and other clinical data. Symptoms were rated on a 0 (not at all) to 4 (a lot) scale.

RESULTS: Pain (mean +/- standard deviation [SD]) scored by the child was 3.6 (+/-0.6)/4 at baseline and 1.8 (+/-1.1)/4 at F1 (p < 0.0001). Compared to baseline, pain scored by the child at F2 was 1.2 (+/-1.3)/4 (p < 0.0001). Pain scored by the parent was 3.5 (+/-0.7)/4 at baseline and 1.4 (+/-1.3)/4 at F1 (p < 0.0001). Compared to baseline, pain scored by the parent at F2 was 1.0 (+/-1.2)/4 (p < 0.0001). Thirty-three (70%) patients at F1 and 23 (79%) patients at F2 did not need a change in dose of methadone. No cardiac arrhythmias or opioid neurotoxicity was observed.

CONCLUSIONS: Initiation of methadone was effective and safe as the first long-acting opioid in children with pain.

AIM: With increasing survival rates in paediatric malignancies, the quality-of-life of children during hospitalisation should be given more attention. We aimed to identify factors associated with psychological and psychosomatic symptoms (PPS) that required medication among children hospitalised for treatment of malignancies.

METHODS: We retrospectively analysed data of 190 patients aged 2-18 years old. They were diagnosed with malignant diseases and admitted for treatment at St. Luke’s International Hospital between 2003 and 2013. Patients were considered as having PPS if they were prescribed psychotropic agents during hospitalisation.

RESULTS: Of the 190 patients, 56 (30%) were prescribed psychotropic agents for PPS. Types of PPS included insomnia in 21 (38%), anxiety in 11 (20%), and others conditions (psychogenetic nausea, agitation, delirium, depression). The most prescribed psychotropic agents were etizolam for 34 cases (61%), followed by diazepam and risperidone. The multivariable analyses confirmed statistically significant independent associations for haematopoietic stem cell transplantation (HSCT) (odds ratio (OR), 5.21; 95% confidence interval (CI), 1.77-15.35), older age (12-18 years vs. 2-5 years, OR, 3.74; 95% CI, 1.04-10.00), and opioid use (OR, 7.15; 95% CI, 2.36-21.69).

CONCLUSIONS: Older age at admission, undergoing HSCT, and those given opioids were found to be risk factors for PPS among children with malignancies. Appropriate preventive measures against PPS may be warranted for patients with these risk factors.


Background: Numerous physical, psychological, and emotional benefits have been attributed to marijuana since its first reported use in 2,600 BC in a Chinese pharmacopoeia. The phytocannabinoids, cannabidiol (CBD), and delta-9-tetrahydrocannabinol (Delta9-THC) are the most studied extracts from cannabis sativa subspecies hemp and marijuana. CBD and Delta9-THC interact uniquely with the endocannabinoid system (ECS). Through direct and indirect actions, intrinsic endocannabinoids and plant-based phytocannabinoids modulate and influence a variety of physiological systems influenced by the ECS.

Methods: In 1980, Cunha et al. reported anticonvulsant benefits in 7/8 subjects with medically uncontrolled epilepsy using marijuana extracts in a phase I clinical trial. Since then neurological applications have been the major focus of renewed research using medical marijuana and phytocannabinoid extracts.

Results: Recent neurological uses include adjunctive treatment for malignant brain tumors, Parkinson’s disease, Alzheimer’s disease, multiple sclerosis, neuropathic pain, and the childhood seizure disorders Lennox-Gastaut and Dravet syndromes. In addition, psychiatric and mood disorders, such as schizophrenia, anxiety, depression, addiction, postconcussion syndrome, and posttraumatic stress disorders are being studied using phytocannabinoids.

Conclusions: In this review we will provide animal and human research data on the current clinical neurological uses for CBD individually and in combination with Delta9-THC. We will emphasize the neuroprotective, antiinflammatory, and immunomodulatory benefits of phytocannabinoids and their applications in various clinical syndromes.


BACKGROUND: Hospitalized children continue to experience inadequate pain management. Children in the rural hospital setting may be at risk due to unique challenges experienced by Registered Nurses (RNs) in this context.

OBJECTIVES: To understand the experience of pain care from RNs who work in rural hospitals with inpatient pediatric patients.

DESIGN: Qualitative description that used semi-structured interviews to explore RNs' inpatient pediatric pain care experiences.

PARTICIPANTS: RNs who: 1) worked directly with pediatric in-patients; 2) spoke English; 3) and who worked in rural Northern Ontario. Hospital sites were selected based on population density, from one province in Canada. To reduce heterogeneity, only sites with dedicated pediatric beds were eligible (n=9).

METHODS: This qualitative descriptive study used semi-structured interviews over Skype and telephone. Data were analyzed using inductive content analysis.
RESULTS: Ten participants were recruited from seven sites. Five main categories were identified, with one category that influenced all other categories. Rural RNs needed to practice as generalists as they care for many types of patients. Resource challenges included a lack of specialist expertise and educational opportunities. Pediatric pain was not perceived as a priority within their organizations. Most participants perceived there were no explicit standards for pain care. Moving forward the adoption of built in assessments in electronic documentation was suggested as a solution to standard pain care.

CONCLUSIONS: Opportunity exists to improve pediatric pain management, however, without a systematic approach that considers the rural context, pain care for children will continue to be based on individual’s beliefs and knowledge.


This case-control study evaluated interventions for bronchiolitis in relation to time in the pediatric intensive care unit (PICU) during a 16-year surveillance period. Together, 105 infants aged < 12 months were treated for bronchiolitis in the PICU, and for them, we selected 210 controls admitted for bronchiolitis closest to cases. We collected data on treatments in the PICU, at the ward and in the emergency department for three periods: years 2000-2005, 2006-2010, and 2011-2015. Median hospital length of stay for PICU patients were 7 days (interquartile range 5-12), 5 days (4-8) and 8 days (4-12.5, p = 0.127), respectively. By time, the use of inhaled beta-agonist (68 vs. 44 vs. 38%, p = 0.019) and systemic corticosteroids (29 vs. 15 vs. 5%, p = 0.019) decreased, but that of racemic adrenaline (59 vs. 78 vs. 84%, p = 0.035) and hypertonic saline (0 vs. 0 vs. 54%, p < 0.001) inhalations increased in the PICU. Similar changes were seen at the ward. In the PICU, non-invasive ventilation therapies increased significantly, but intubation rates did not decline.

CONCLUSION: Beta-agonists and systemic corticosteroids were used less by time in intensive care for infant bronchiolitis, but the use of hypertonic saline and racemic adrenaline increased, though their effectiveness has been questioned. What is Known: * Until now, studies have shown which treatments do not work in bronchiolitis, and so, there is no consensus how infants with bronchiolitis should be treated. In particular, there is no consensus on different interventions in intensive care for bronchiolitis. What is New: * During 2000-2015, treatments with inhaled beta-agonists and systemic corticosteroids decreased but treatments with racemic adrenaline and hypertonic saline inhalations increased in intensive care for bronchiolitis. Similar changes were seen at the ward. Though non-invasive ventilation therapies increased, the intubation rate did not decline.


OBJECTIVE: To report on the population of infants receiving a tracheostomy, identify acute post-tracheostomy clinical decompensations, and seek predictive markers associated with acute complications following the placement of a tracheostomy.

STUDY DESIGN: Retrospective deidentified clinical data was provided by the Infant Pulmonary Data Repository at Children’s Mercy Hospital, Kansas City. Data from infants undergoing tracheostomy from January 1, 2008 through September 30, 2016 were divided into one of two study groups based on clinical correlations: (1) no acute decompensations within 72 hours post-tracheostomy or (2) acute clinical decompensation defined as sustained escalation of respiratory care within the 72 hours following tracheostomy.

RESULTS: Thirty-four percent of infants undergoing tracheostomy during this period developed acute post-tracheostomy clinical decompensations. Elevated pre-tracheostomy positive end expiratory pressure, mean airway pressure, and echocardiogram findings suggestive of pulmonary hypertension (PH) or ventricular dysfunction were associated with acute post-tracheostomy clinical decompensations. Additionally acute post-tracheostomy clinical decompensation was associated with higher rate of death prior to discharge.

CONCLUSION: Infants requiring higher respiratory support and infants with PH or ventricular dysfunction are at risk of acute post-tracheostomy clinical decompensation, thus identifying these patients may lead to better pre-tracheostomy counseling and potentially targeted treatments to decrease this risk.


PURPOSE: Chemotherapy-induced nausea and vomiting (CINV) affects quality of life for patients with cancer undergoing chemotherapy. We aimed to assess the effect of lorazepam with granisetron on CINV in children with acute lymphoblastic leukemia (ALL).

METHODS: We reviewed the records of 71 consecutive patients with newly diagnosed ALL who received chemotherapy including vincristine, anthracycline, and systemic steroids between January 2011 and December 2016 in our hospital. The number of chemotherapy cycles reviewed was 164. All patients received granisetron as CINV prophylaxis.

RESULTS: Nausea was observed in 51/71 patients (72%) and 93/164 cycles (57%). Vomiting was observed in 47/71 patients (66%) and 79/164 cycles (48%). Age and gender distribution were not significantly different between patients who received lorazepam at the initiation of the chemotherapy cycle (LZP group, n = 30) and those who did not receive lorazepam (non-LZP group, n = 134). There were no significant differences in the incidence of CIN and CIV between the LZP group and non-LZP group (CIN, 67% vs. 57%, P = 0.31; CIV, 53% vs. 47%, P = 0.98). In multivariate logistic regression, female gender and older age (> 5 years) were significant risk factors for CIV (female, odds ratio (OR) 2.5, 95% confidence interval (CI) 1.3-5.0, P = 0.007; older age, OR 2.5, CI 1.3-4.8, P = 0.008).

CONCLUSIONS: We found no beneficial effect of providing lorazepam as adjuvant antiemetic for prevention of CINV in children with ALL.

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Generalized severe junctional epidermolysis bullosa is a rare mechanobullous skin disorder that is uniformly fatal. We present the case of an infant who received palliative pain management and ultimately proportionate palliative sedation. However, because of the extent of the patient's skin disease, we were unable to provide palliative medication through parenteral routes. We discuss the provision of enteral palliative sedation, including the pharmacology, and creative use of medications to achieve sufficient palliation in this difficult and unique situation.


BACKGROUND: Total pain is a concept that approaches pain holistically: physically, psychologically, socially, and spiritually. Any individual may experience pain in each domain at a different level. This is the case report of an adolescent who suffered from total pain and how his healthcare team and peers helped to relieve it.

CASE PRESENTATION: A 15-years-old Thai male was diagnosed with recurrent T-cell lymphoma and readmitted to hospital. He was admitted to an adult ward and suffered from pain due to his disease and from the fear of being alienated. As a result, he had an existential crisis. His parents felt unsure whether they or the patient should make the medical decisions and advance care plan.

CONCLUSIONS: This case report emphasises the importance of total pain assessment in the relief of total pain in an adolescent whose needs are different from both children and adults. It also highlights the role of medical decision-making in adolescents and the importance of the social support of peers in the alleviation of pain.


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.


PURPOSE: Epileptic seizures complicate the management of childhood brain tumours. There are no published standards for clinical practice concerning risk factors, treatment selection or strategies to withdraw treatment with antiepileptic drugs (AED). METHOD: we undertook a case note review of 120 patients with newly diagnosed brain tumours, referred to a regional paediatric cancer service.

RESULTS: data was available on 117/120 (98%) children <18 years: median age at tumour presentation was 8.1 years (IQR 25 degrees -75 degrees ): 3.6-12.7, median follow up was 33 months (IQR 25 degrees -75 degrees : 24-56), and 35/117 (29%) experienced seizures. A cortical tumour location was associated with the highest risk of seizures (OR: 7.1; CI 95% 2.9-17.3). At a median follow up of 24 months (IQR25 degrees -75 degrees : 15-48), 22/35 (63%) with seizures, had a single seizure episode, 15/35 (43%) were seizure free (SF) on AEDs, 13/35 (37%) were SF off AEDs, and 7/35 (20%) experienced continuing epileptic seizures. Overall 34/35 (97%) were treated with AEDs after a seizure, of whom 12/35 (35%) withdrew from AED medication, and although 4/35 (12%) had seizure relapse, all were after further acute events. The median duration of AED before withdrawal was 11 months (IQR25 degrees -75 degrees : 5-14 months), and the median follow up after withdrawal was 15 months (IQR25 degrees -75 degrees : 5-34 months).

CONCLUSIONS: Seizures affect about 1/3rd of children and young people presenting with and being treated for brain tumours particularly when the tumour is in the cerebral cortex. The low risk of recurrent seizures after AED treatment justifies consideration of early withdrawal of AED after seizure control.


The mucopolysaccharidosis (MPS) constitutes a heterogeneous group of rare genetic disorders caused by enzymatic deficiencies that lead to the accumulation of glycosaminoglycans. Several types of MPS are described, historically numbered from I to IX. Clinical observations strongly suggest the presence of chronic pain in patients with all types of MPS. There are few data in the literature on the evaluation and management of pain in these patients, a fact that can compromise the quality of life even more. Professionals with extensive experience in the care for patients with MPS held a meeting in April 2017 to discuss and propose recommendations for the evaluation and management of pain in patients with MPS in Latin America. This article summarizes the content of the discussions and presents the recommendations produced at the meeting. Patients with MPS present joint, bone, and muscle pain, as well as entrapment syndromes (spinal, optic nerve, carpal tunnel). The panel suggests the use of the following instruments for pain assessment: Face, Legs, Activity, Cry and Consolability Scale for children of up to four years of age and patients unable to communicate their pain; Child Health Assessment Questionnaire Scale; Facial Pain Scale and Numerical Pain Scale for patients of five to <18 years of age; Brief Pain Inventory and Short Form Health Survey 36 scales for patients aged 18 years or older. Based on the scores verified in these scales, the panel proposes pharmacological interventions for pain relief in this population of patients.

Purpose: Children with medical complexity experience frequent pain. But it can be challenging to recognize unexpected behaviors as pain related, especially in the absence of self-report. Often these children undergo extensive workups aimed at diagnosing the sources of pain. This study had three objectives: to describe the signs and symptoms parents of nonverbal children with medical complexity found worrisome, to describe the sources of pain in these children, and to describe nursing pain assessment practices in this population.

**DESIGN AND METHODS:** Retrospective chart review was used to identify the initial presenting symptoms, sources of pain, and nursing documentation for 46 children with medical complexity who were admitted with a chief concern of pain to a 395-bed pediatric teaching hospital in the northeastern United States.

RESULTS: Irritability, pain, feeding intolerance, and "not acting like herself [or himself]" were common parent-reported symptoms that prompted further evaluation. On average, five diagnostic studies were taken to identify a source of pain, and four specialty services were consulted during the admission. Nursing assessments of pain were documented approximately every three hours; the mean pain intensity score documented was 1.1 out of 10. The discharge diagnoses included infection (including urinary tract infection), seizures, constipation, chronic pain, failure to thrive, dehydration, and subdural hematoma.

CONCLUSIONS: The discharge diagnoses covered a wide range. A systematic approach to pain evaluation could help to ensure that the diagnostic process is both thorough and efficient. Common childhood ailments such as constipation or urinary tract infection, as well as other causes, must be considered when diagnosing pain in this population. Practice implications include consulting parents regarding changes in a child’s behavior.

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Objectives: To test the efficacy of a brief behavioral pain management strategy (The ABCDs of Needle Pain Management), delivered via video, on infants’ and toddlers’ pain scores and on parental soothing behavior.

Methods: This was a double-blind, parallel trial design. Parent-child dyads (N = 128) were recruited before their child’s 6-month (infant) or 18-month (toddler) vaccination in a pediatric clinic and randomly assigned to watch a 5-min treatment video or a placebo video. The primary outcome was the Modified Behavior Pain Scale (Taddio et al., Journal of Pain and Symptom Management, 10, pp. 456-463, 1995), coded during four epochs (Pain Reactivity, Pain Regulation 1 min, Pain Regulation 2 min, and Pain Regulation 3 min) after the last vaccination needle. Secondary analyses examined parental use of distraction, rocking, and physical comforting over this same time period.

Results: Results demonstrated a treatment effect for toddlers (18-month-olds) for the Pain Regulation 1 (d = 0.84) and Pain Regulation 2 (d = 0.76) postvaccination scores. Secondary analyses found differences in parental rocking and physical comforting between treatment conditions and between age-groups (d’s = 0.37-0.54).

Conclusions: The ABCD pain management strategy delivered via video was an effective way to reduce toddler pain after vaccination and increase parental use of rocking and physical comforting. The treatment effect was not demonstrated with infants.

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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.

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OBJECTIVES: To assess the effect of 3 musical interventions, as compared to no music, on the physiological response of healthy newborns undergoing painful medical procedures (Guthrie test and/or intramuscular antibiotic injections).

METHODS: Prospective study of 80 full-term newborns, aged 1 to 3 days, randomly allocated to exposure to Mozart’s Sonata for two pianos K.448, Beethoven’s Moonlight Sonata, heartbeat sound recordings (70bpm) or no music. Pain perception (evaluated using the Neonatal Infant Pain Scale), heart rate and oxygen saturation were measured 10 min before (T0), during (T1), 10 (T2) and 20 (T3) minutes after the interventions.

RESULTS: Infants who were exposed to the three music interventions displayed a significant reduction in heart rate and in pain perception and an increase in oxygen saturation, as compared to the control group, which showed less modifications on stress measurements after painful medical procedures (F(3,76)=6.40, p=.001, partial eta(2)=0.20).

CONCLUSIONS: Exposure to music and heartbeat sound recordings changes short-term physiological parameters in healthy newborns undergoing potentially painful procedures. The similar effect shown by the 3 interventions might be explained by the common characteristics of the sound shared by the various tracks. Further research is needed to investigate the impact of different types of music used in intervention, in order to develop guidelines and include music as a part of evidence-based strategies to promote the outcome for neonates.


BACKGROUND: The management of neuropathic pain and pain related to bone vaso-occlusive crises in sickle cell disease remains challenging in children. Lidocaine 5% patches are recommended in adults for neuropathic pain treatment, but they are not recommended in children. The purpose of this study was to assess the efficacy and tolerance of lidocaine 5% patches in pediatric inpatients.

METHODS: This prospective, multicenter, single-arm, phase II study aimed to assess the use of lidocaine 5% patches in 6- to 21-year-old pediatric patients suffering from neuropathic pain or superficial bone vaso-occlusive crises. Patches were applied on the painful area for 12 hours a day. The primary endpoint was the proportion of inpatients with significant pain relief defined as a decrease of at least 2 points on the visual analog pain scale (VAS) measured at 12 hours after patch placement over at least 2 consecutive days.

RESULTS: The 12-hour VAS score decreased by at least 2 points over 2 consecutive days in 48.6% of patients 95% unilateral confidence interval (33.8%). Only 7.7% of patients experienced grade 1 or grade 2 toxicities.

CONCLUSION: Although lidocaine 5% patches decreased the pain’s intensity in nearly half of the enrolled patients with an excellent tolerance, the efficacy endpoint was not reached. Further studies should consider a more refined selection of the experimental population to assess the efficacy of lidocaine 5% patches in the pediatric population.


Experiencing pain is the greatest contributor to a reduced quality of life in children with cerebral palsy (CP). The presence of pain is quite common (~60%) and increases with age. This leads to missed school days, less participation, and reduced ambulation. Despite these alarming consequences, strategies to relieve the pain are absent and poorly studied. Moreover, it is difficult to evaluate pain in this group of children, especially in cases of children with cognitive deficits, and tools for pain evaluation are often inadequate. Botulinum toxin has been shown to alleviate pain in a variety of disorders and could potentially have an analgesic effect in children with CP as well. Even though most of the studies presented here show promising results, many also have limitations in their methodology as it is unlikely to capture all dimensions of pain in this heterogeneous group using only one assessment tool. In this review, we present a new way of examining the analgesic effect of botulinum toxin in children with CP using a variety of pain scores.


Persistent pain is a global health care issue affecting more than 30% of children and young people. mHealth applications delivered using smartphones, are an innovative method to engage children in pain self-management. This article outlines the evidence concerning the development, implementation, and evaluation of mHealth apps for these children in terms of feasibility, acceptability, and impact on important pain outcomes such as quality of life and health care utilization.

https://www.ncbi.nlm.nih.gov/pubmed/30126292


BACKGROUND: Epilepsy affects about 50 million people worldwide, nearly a quarter of whom have drug-refractory epilepsy. People with drug-refractory epilepsy have increased risks of premature death, injuries, psychosocial dysfunction, and a reduced quality of life.

OBJECTIVES: To assess the efficacy and tolerability of clonazepam when used as an add-on therapy for adults and children with refractory focal or generalised onset epileptic seizures, when compared with placebo or another antiepileptic agent. SEARCH METHODS: We searched the following databases on 14 September 2017: Cochrane Epilepsy Group Specialized Register, Cochrane Central Register of Controlled Trials (CENTRAL) via the Cochrane Register of Studies Online (CRSO), MEDLINE (Ovid 1946 to 14 September 2017), ClinicalTrials.gov, and the WHO International Clinical Trials Registry Platform (ICTRP).

SELECTION CRITERIA: Double-blind randomised controlled studies of add-on clonazepam in people with refractory focal or generalised onset seizures, with a minimum treatment period of eight weeks. The studies could be of parallel or cross-over design.

DATA COLLECTION AND ANALYSIS: Two review authors independently selected studies for inclusion, extracted relevant data, and assessed trial quality. We contacted study authors for additional information.

MAIN RESULTS: No double-blind randomised controlled trials met the inclusion criteria.

AUTHORS’ CONCLUSIONS: There is no evidence from double-blind randomised controlled trials for or against the use of clonazepam as an add-on therapy for adults and children with refractory focal or generalised onset epileptic seizures.


Pain is an unpleasant subjective experience. At present, clinicians are using self-report or pain scales to recognise and monitor pain in children. However, these techniques are not efficient to observe the pain in children having cognitive disorder and also require highly skilled observers to measure pain. Using these techniques it is also difficult to choose the analgesic drug dosages to the patients after surgery. Thus, this conceptual work explains the demand for automatic coding techniques to evaluate pain and also it documents some evidence of techniques that act as an alternative approach for objectively determining pain in children. In this review, some good indicators of pain in
children are explained in detail; they are facial expressions from an RGB image, thermal image and also feature from well proven physiological signals such as electrocardiogram, skin conductance, body temperature, surgical pleth index, pupillary reflex dilation, analgesia noception index, photoplethysmography, perfusion index etc.


BACKGROUND: Patients with Lennox-Gastaut syndrome, a rare, severe form of epileptic encephalopathy, are frequently treatment resistant to available medications. No controlled studies have investigated the use of cannabidiol for patients with seizures associated with Lennox-Gastaut syndrome. We therefore assessed the efficacy and safety of cannabidiol as an add-on anticonvulsant therapy in this population of patients.

METHODS: In this randomised, double-blind, placebo-controlled trial done at 24 clinical sites in the USA, the Netherlands, and Poland, we investigated the efficacy of cannabidiol as add-on therapy for drop seizures in patients with treatment-resistant Lennox-Gastaut syndrome. Eligible patients (aged 2-55 years) had Lennox-Gastaut syndrome, including a history of slow (<3 Hz) spike-and-wave patterns on electroencephalogram, evidence of more than one type of generalised seizure for at least 6 months, at least two drop seizures per week during the 4-week baseline period, and had not responded to treatment with at least two antiepileptic drugs. Patients were randomly assigned (1:1) using an interactive voice response system, stratified by age group, to receive 20 mg/kg oral cannabidiol daily or matched placebo for 14 weeks. All patients, caregivers, investigators, and individuals assessing data were masked to group assignment. The primary endpoint was percentage change from baseline in monthly frequency of drop seizures during the treatment period, analysed in all patients who received at least one dose of study drug and had post-baseline efficacy data. All randomly assigned patients were included in the safety analyses. This study is registered with ClinicalTrials.gov, number NCT02224690.

FINDINGS: Between April 28, 2015, and Oct 15, 2015, we randomly assigned 171 patients to receive cannabidiol (n=86) or placebo (n=85). 14 patients in the cannabidiol group and one in the placebo group discontinued study treatment; all randomly assigned patients received at least one dose of study treatment and had post-baseline efficacy data. The median percentage reduction in monthly drop seizure frequency from baseline was 43.9% (IQR -69.6 to -1.9) in the cannabidiol group and 21.8% (IQR -45.7 to 1.7) in the placebo group. The estimated median difference between the treatment groups was -17.21 (95% CI -30.32 to -4.09; p=0.0135) during the 14-week treatment period. Adverse events occurred in 74 (86%) of 86 patients in the cannabidiol group and 59 (69%) of 85 patients in the placebo group; most were mild or moderate. The most common adverse events were diarrhoea, somnolence, pyrexia, decreased appetite, and vomiting. 12 (14%) patients in the cannabidiol group and one (1%) patient in the placebo group withdrew from the study because of adverse events. One patient (1%) died in the cannabidiol group, but this was considered unrelated to treatment.

INTERPRETATION: Add-on cannabidiol is efficacious for the treatment of patients with drop seizures associated with Lennox-Gastaut syndrome and is generally well tolerated. The long-term efficacy and safety of cannabidiol is currently being assessed in the open-label extension of this trial.

FUNDING: GW Pharmaceuticals.


PURPOSE: The purpose of this integrative review was to identify, review, synthesize, and analyze the current literature related to pain management in the pediatric palliative care population from infancy through adolescence.

METHODS: The literature was searched for the terms palliative, pediatric, and pain in PubMed, PsycINFO, Cumulative Index to Nursing and Allied Health Literature (CINAHL) Complete, and Google Scholar. The search was limited to papers in English that had been published from January 1, 2005, to December 31, 2016.

RESULTS: These searches resulted in 918 articles, of which 29 met inclusion criteria. These 29 articles were reviewed and reported. Four broad themes emerged: patient and family experience, pain assessment, pharmacological pain management, and nonpharmaceutical interventions.

CONCLUSIONS: Gaps in current research have been identified, such as investigating pediatric pain scales for the palliative care population and new complementary and alternative medical therapy and other interventions. More
research is needed to bring innovative pain management interventions to the attention of pediatric caregivers. CLINICAL RELEVANCE: A better understanding of current research on pain in the pediatric palliative care population can improve patient care and lead to better research in this specialty field.


Antiepileptic medications, and valproate principally, are commonly prescribed teratogens. There is significant concern that we are not doing enough to educate clinicians and potential parents about the risks of valproate in pregnancy. There is clear advice from the Medicines and Healthcare products Regulatory Agency and the International League Against Epilepsy about the risks of valproate exposure in utero Reviews and guidelines that are focused on fetal risk, however, fall short in being able to fully replicate the complexity of a real clinical decision. Valproate is certainly life-changing if your child is one of the 10% with a major malformation or 30-40% with a neurodevelopmental disorder, but valproate is also potentially life-saving in the context of ensuring the best possible seizure control for some mothers with epilepsy. There are significant knowledge gaps regarding the risks to mothers who elect to take another drug, or to mother and baby if she comes off medication entirely. We also should be doing more to reduce rates of sudden unexpected death in epilepsy (SUDEP), which is recognised as a key target when evaluating all maternal deaths.


PURPOSE: Chemotherapy-induced peripheral neuropathy (CIPN) is a common side effect of chemotherapy, in need of effective treatment. Preliminary data support the efficacy of scrambler therapy (ST), a noninvasive cutaneous electrostimulation device, in adults with CIPN. We test the efficacy, safety, and durability of ST for neuropathic pain in adolescents with CIPN.

PATIENTS AND METHODS: We studied nine pediatric patients with cancer and CIPN who received ST for pain control. Each patient received 45-min daily sessions for 10 consecutive days as a first step, but some of them required additional treatment.

RESULTS: Pain significantly improved comparing Numeric Rate Scale after 10 days of ST (9.22 +/- 0.83 vs. 2.33 +/- 2.34; P < 0.001) and at the end of the optimized cycle (EOC) (9.22 +/- 0.83 vs. 0.11 +/- 0.33, P < 0.001). The improvement in quality of life was significantly reached on pain interference with general activity (8.67 +/- 1.66 vs. 3.33 +/- 2.12, P < 0.0001), mood (8.33 +/- 3.32 vs. 2.78 +/- 2.82, P < 0.0005), walking ability (10.00 vs. 2.78 +/- 1.22, P < 0.0001), sleep (7.56 +/- 2.24 vs. 2.67 +/- 1.41, P < 0.001), and relations with people (7.89 +/- 2.03 vs. 2.11 +/- 2.03, P < 0.0002; Lansky score 26.7 +/- 13.2 vs. 10 days of ST 57.8 +/- 13.9, P < 0.001; 26.7 +/- 13.2 vs. EOC 71.1 +/- 16.2, P < 0.001).

CONCLUSION: Based on these preliminary data, ST could be a good choice for adolescents with CIPN for whom pain control is difficult. ST caused total relief or dramatic reduction in CIPN pain and an improvement in quality of life, durable in follow-up. It caused no detected side effects, and can be retrained successfully. Further larger studies should be performed to confirm our promising preliminary data in pediatric patients with cancer.


OBJECTIVES: Pain is a common and distressing symptom of pediatric cancer, as reported by both children and their parents. Increasingly, children with cancer are cared for as outpatients, yet little is known about how parents manage their cancer-related pain. The aim of the current study was to examine pain prevalence and characteristics, and the pharmacological, physical, and psychological pain management strategies used by parents to manage their child's cancer pain.

MATERIALS AND METHODS: In total, 230 parents and caregivers (89% mothers) of children (mean age=8.93 y, SD=4.50) with cancer currently in treatment or who are survivors completed an online survey about their child's pain in the preceding month.
RESULTS: Results indicated that children with cancer who were on active treatment and who were posttreatment experienced clinically significant levels of pain. Parents reported using more physical and psychological strategies than pharmacological strategies to manage their child's pain. The most frequently used physical/psychological strategy was distraction and acetaminophen was the most frequently administered pain medication. Parents' confidence in managing their child’s pain was inversely associated with both how much pain they perceived their child had, and also whether they had given any pain medication.

DISCUSSION: The results of this study suggest that despite parents’ use of pain management strategies, management of cancer-related pain continues to be a problem for children during treatment and into survivorship.


The main remit of the European Society for Paediatric Anaesthesiology (ESPA) Pain Committee is to improve the quality of pain management in children. The ESPA Pain Management Ladder is a clinical practice advisory based upon expert consensus to help to ensure a basic standard of perioperative pain management for all children. Further steps are suggested to improve pain management once a basic standard has been achieved. The guidance is grouped by the type of surgical procedure and layered to suggest basic, intermediate, and advanced pain management methods. The committee members are aware that there are marked differences in financial and personal resources in different institutions and countries and also considerable variations in the availability of analgesic drugs across Europe. We recommend that the guidance should be used as a framework to guide best practice.


Hypercukaesthesia is a rare neurogenetic disorder characterized by startle. Accurate diagnosis of this notorious mimicker of epilepsy is important to prevent life-threatening apnoea. We report a novel case of concomitant GLRA1-related hyperkplexia and myoclonic epilepsy. A toddler with daily paroxysms of head drops and falls presented with epileptic myoclonus on EEG, however, whole-exome sequencing revealed hyperkplexia-related GLRA1 mutation. The boy eventually developed spells induced by noise and surprise. All his spells remitted upon treatment with clonazepam. Paediatricians and paediatric neurologists should be aware of this possible mixed presentation in order to appropriately tailor medication regimens and treatment goals. [Published with video sequence on www.epilepticdisorders.com].


The death of a newborn infant is one of the most devastating situations a mother could experience. The aim of this study was to understand bereavement and its associated meanings as lived and experienced by the mothers who lost their newborn infants in the intensive care units of hospitals in Jordan. Data were generated using semistructured face-to-face interviews with 12 mothers who had the experience. A qualitative phenomenological approach was used for data analysis. Three main themes emerged from the analysis: (1) longing and grieving, as natural emotional responses to the loss; (2) adaptive work of coping, as the mothers internalized meanings to cope with their loss; and (3) moving forward but with a scar, as the mothers moved on with their lives while they carried the unforgettable memories of the newborns’ death experience. Bereavement support services should be considered vitally important as soon as the news of a newborn’s death is delivered to the mother. Palliative care nurses and other health care providers should give careful attention to the meanings that the mothers attached to the loss and support each bereaved mother’s spiritual values and effective coping mechanisms.

INTRODUCTION: We aimed to determine the benefits/efficacy of mindfulness-based interventions (MBIs) implemented among adolescents with chronic diseases in clinical settings.

METHODS: An electronic search of PubMed, CINAHL, and PsycINFO databases was conducted in November 2017 to identify studies in which mindfulness was the primary intervention delivered for adolescents with chronic diseases to improve psychological and physical health.

RESULTS: Nineteen eligible studies were included in this review. Fifteen studies included adolescents with psychiatric or pain disorders, and four included adolescents with a chronic physical disorders. Psychological outcomes and pain were examined in most studies with effect sizes for MBIs ranging from small to large.

DISCUSSION: MBI studies conducted in clinical settings mainly engaged adolescents with psychiatric or pain disorders. The effectiveness of MBIs on improving psychological outcomes were inconsistent. Large randomized trials are needed to examine the effectiveness of MBIs and should expand to include adolescents with chronic physical diseases.

The purpose of this study was to describe the impact of peer support on post-traumatic stress disorder in parents who have experienced the death of a child, the factors associated with the parents’ post-traumatic stress reactions and the parents’ experiences of peer support. The research data comprise the responses of parents who participated in a family weekend organised by Child Death Families Finland (KAPY). The data were collected 2 weeks before (n = 110) and 2 weeks after (n = 73) the family weekend by a questionnaire consisting of items designed to identify the relevant background variables and the Impact of Event Scale-Revised (IES-R), a self-report measure for assessing post-traumatic stress disorder (PTSD). Statistical methods were applied in the data analysis. No statistically significant differences were observed in the parents’ stress reactions 2 weeks before and 2 weeks after the family weekend. The stress reactions, on the other hand, had a statistically significant association with the self-perceived health of the parents, the age at which their child had died and the time that had elapsed since the death. Two-thirds of the parents regarded the peer support provided during the family weekend as supportive or very supportive. The parents also regarded the support provided during the weekend as important, although the support had no statistically significant impact on their stress reactions. One can draw the conclusion that the parents experienced the family weekend and the peer support provided during it as supportive.


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


Febrile neutropenia requires prompt assessment and antibiotic administration and is the most common reason for unexpected hospital admission in pediatric oncology. Parents are expected to be vigilant and "drop everything" to take their child to their nearest hospital for assessment if fever occurs. Delays in antibiotic administration are associated with poorer outcomes; however, delays are common. Our aim was to understand and describe the lived experience of parents of children with cancer who received treatment for fever with confirmed/suspected neutropenia. We used descriptive phenomenological concepts to undertake and analyze interviews with parents, who were asked to describe their recent experience of hospitalization in Queensland, Australia. Nine participants were interviewed. Five children were treated in the tertiary treating center and four were treated in smaller regional towns. Three main categories were identified that shaped and characterized parents' experiences: being heard, confidence in capabilities of health care professionals, and living with anticipated distress and uncertainty. Parents' experiences were related to the level they needed to advocate for their child's care across all themes. Familiarity with health care professionals increased confidence and improved parents' experiences. Maintaining vigilance and managing the child and family's response to an unexpected admission had a substantial negative effect on parents. Understanding parents' experiences and perceptions of the management of febrile neutropenia adds to the current body of knowledge and offers potential new insights to improve clinical practice.


BACKGROUND: Defensiveness is one of the strategies that children with cancer use against psychosocial difficulties, yet it remains unclear what factors may impact children's use of defensiveness.

OBJECTIVE: The aim of this study was to explore the psychological adjustment, including use of defensive behaviors, in children who may or may not be told about the diagnosis of cancer.
METHODS: A total of 58 children and 51 mothers participated in the study. Children answered questionnaires about defensiveness, anxiety, and depression, whereas mothers completed a questionnaire of anxiety, depression, and stress and a question about disclosure of cancer to child.

RESULTS: Significant differences were found in the defensiveness and depression scores between children with full disclosure and those with no disclosure about their cancer. Although children's adjustment has been directly related to that of their mother’s adjustment, we did not find this variable to be a predictor of child’s adjustment. The child’s defensiveness scores were a strong predictor of child’s anxiety and eventually for a high risk for depression.

CONCLUSIONS: Mothers’ emotional strains and lack of open disclosure about cancer significantly affect the psychosocial well-being of children.

IMPLICATIONS FOR PRACTICE: It is essential for nurses to assess the emotional adjustment and defensiveness strategies that children with cancer use. We believe that nurses caring for children with cancer have a professional responsibility to identify and understand defensive behaviors and other characteristics of psychosocial distress and advocate for psychological interventions that will help mothers and their children cope with cancer.


Objective: The majority of problems and symptoms occur in the gastrointestinal system in children with cancer. Parents have difficulty in coping with the nutritional problems and changing routines of children and need support in this respect. This study aimed to assess the nutritional problems of children with cancer and the information needs of their parents.

Methods: This descriptive study was performed among children with cancer aged 3-18 years and their parents (n = 69). The data were collected through a data collection form developed by the researchers based on the literature.

Results: The most prominent nutritional problems experienced by children were loss of appetite (85.5%), nausea (84.1%), vomiting (81.2%), fatigue (79.7%), and mucositis (66.7%). According to the parents, the factors causing these nutritional problems in children were physiological factors (100%) and the foods given to children in the hospital (65.2%). The parents mostly needed information about food-drug interactions (58.0%), food-disease interactions (52.2%), foods that children with neutropenia should avoid or should eat (neutropenic diet) (46.4%), and frequency of nutritional intake (36.2%).

Conclusions: This study has shown that most children experience at least one nutritional problem, and the parents need comprehensive and regular information about nutrition. Pediatric oncology nurses have a significant responsibility in the evaluation, education, and monitoring of these children.


Despite the frequent co-occurrence of hypermobile Ehler-Danlos syndrome (hEDS) and pathological anxiety, little is known about the psychosocial and health implications of such comorbidity. Our aim was to explore the association between high levels of anxiety and psychosocial (catastrophizing, kinesiophobia, somatosensory amplification, social support and functioning), health (pain, fatigue, BMI, tobacco/alcohol use, depression, diagnosis delay, general health), and sociodemographic factors in people with hEDS. In this cross-sectional study, 80 hEDS patients were divided into two groups according to self-reported anxiety levels: low and high. Psychosocial, sociodemographic and health variables were compared between the groups. Forty-one participants reported a high level of anxiety (51.2%). No differences were found in the sociodemographic variables between high-anxious and low-anxious patients. The percentage of participants with severe fatigue and high depressive symptomatology was significantly higher in the high-anxious group (80.5 vs 56.4; 26.8 vs 12.8%, respectively). High-anxious hEDS patients also showed
significantly higher levels of pain catastrophizing, somatosensory amplification as well as a poorer social functioning and general health. Multivariate analyses showed that somatosensory amplification, pain catastrophizing and poor social functioning are variables that increase the probability of belonging to the high-anxious group. Despite limitations, this first study comparing high-anxious versus low-anxious hEDS patients with respect to health aspects, highlight the importance of considering the psychosocial factors (many susceptible to modification), to improve the adjustment to this chronic condition and provide support to those affected through a biopsychosocial approach.


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.


CONTEXT: Supporting patients' spiritual needs is central to palliative care. Adolescents and young adults (AYAs) may be developing their spiritual identities; it is unclear how to navigate conversations concerning their spiritual needs.

OBJECTIVES: To 1) describe spiritual narratives among AYAs based on their self-identification as religious, spiritual, both, or neither and 2) identify language to support AYAs' spiritual needs in keeping with their self-identities.

METHODS: In this mixed-methods, prospective, longitudinal cohort study, AYAs (14-25 years old) with newly diagnosed cancer self-reported their "religiousness" and "spirituality." One-on-one, semistructured interviews were conducted at three time points (within 60 days of diagnosis, six to 12 months, and 12-18 months later) and included queries about spirituality, God/prayer, meaning from illness, and evolving self-identity. Post hoc directed content analysis informed a framework for approaching religious/spiritual discussions.

RESULTS: Seventeen AYAs (mean age 17.1 years, SD = 2.7, 47% male) participated in 44 interviews. Of n = 16 with concurrent survey responses, five (31%) self-identified as both "religious and spiritual" and "spirituality." One-on-one, semistructured interviews were conducted at three time points (within 60 days of diagnosis, six to 12 months, and 12-18 months later) and included queries about spirituality, God/prayer, meaning from illness, and evolving self-identity. Post hoc directed content analysis informed a framework for approaching religious/spiritual discussions.

CONCLUSION: AYA self-identities evolve during the illness experience. When words such as "religion" and "spirituality" do not fit, explicitly exploring hopes, worries, meaning, and changing life perspectives may be a promising alternative.


The concept of quality of life (QoL) is used in consultations to plan the care and treatment of children and young people (CYP) with brain tumors (BTs). The way in which CYP, their parents, and their health care professionals (HCP) each understand the term has not been adequately investigated. This study aimed to review the current qualitative research on CYP, parents’ and clinicians’ concepts of QoL for CYP with BTs using meta-ethnography. Six studies were found, which reflected on the concept of QoL in CYP with BTs; all explored the CYP’s perspective and one study also touched upon parents’ concept. A conceptual model is presented. Normalcy (a “new normal”) was found to be the key element in the concept. This study calls for a conception of QoL, which foregrounds normalcy over the more common health-related quality of life (HRQoL) and the need to understand the concept from all perspectives and accommodate change over time.


This paper explores the biopsychosocial and spiritual needs of adolescents and young adults (AYA) 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 informed and involved in the health care decision-making process, leading to ethical dilemmas when the AYA is a minor and their wishes differ from the wishes of their legal guardian(s). Social workers are especially well-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.


BACKGROUND: Adolescents living with incurable cancer require ongoing support to process grief, emotions, and information as disease progresses including treatment options (phase 1 clinical trials and/or hospice/palliative care). Little is known about how adolescents become ready for such discussions.

OBJECTIVE: The purpose of this study was to explore the process of adolescent readiness for end-of-life preparedness discussions, generating a theoretical understanding for guiding clinical conversations when curative options are limited.

METHODS: We explored 2 in-depth cases across time using case-study methodology. An a priori conceptual model based on current end-of-life research guided data collection and analysis. Multiple sources including in-depth adolescent interviews generated data collection on model constructs. Analysis followed a logical sequence establishing a chain of evidence linking raw data to study conclusions. Synthesis and data triangulation across cases and time led to theoretical generalizations. Initially, we proposed a linear process of readiness with 3 domains: a cognitive domain (awareness), an emotional domain (acceptance), and a behavioral domain (willingness), which preceded preparedness.

RESULTS: Findings led to conceptual model refinement showing readiness is a dynamic internal process that interacts with preparedness. Current awareness context facilitates the type of preparedness discussions (cognitive or emotional). Furthermore, social constraint inhibits discussions.

CONCLUSIONS: Data support theoretical understanding of the dynamism of readiness. Future research that validates adolescent conceptualization will ensure age-appropriate readiness representation.

IMPLICATIONS FOR PRACTICE: Understanding the dynamic process of readiness for engaging in end-of-life preparedness provides clinician insight for guiding discussions that facilitate shared decision making and promote quality of life for adolescents and their families.


OBJECTIVES: The present study aims to examine relationships between parental behavior and cognition and treatment outcomes in children enrolled in an intensive interdisciplinary pain rehabilitation program.

RESEARCH METHOD: 670 consecutive referrals of children with chronic pain were enrolled in a clinical database registry from 2009 to 2014. Participants and their parents completed measures of physical and psychosocial functioning, and pain-related severity ratings. Data were taken at three time points: admission (N = 670), discharge (N = 504), and 6-month posttreatment (N = 110), although only complete data from 82 participants was used for final analyses.

RESULTS: Both children and parents alike reported significant improvement in functioning, both at discharge and 6 months posttreatment. Parent functioning showed weak to moderate associations with child functioning, with stronger correlations at 6 months posttreatment. Regression analyses demonstrated that changes in parent functioning predicted child functioning and report of pain at 6 months.

CONCLUSIONS: Parents are an integral part of a child’s pain experience and associated disability. An improvement in parent functioning in the course of chronic pain rehabilitation is linked with functional gains in the child. Future research and clinical programming should target the role of parents in pediatric chronic pain interventions in order to optimize both child and family functioning. (PsycINFO Database Record


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.


THEORETICAL PRINCIPLES: Pediatric oncology nurses are particularly vulnerable to emotional distress. Responsible for the oversight of a child’s care, these nurses sustain close interactions with multiple patients and families over time, many of whom are coping with life-limiting diagnoses. The world of pediatric oncology nurses is one where tragedy is routinely witnessed thus demanding self-care and healing across a continuum.

PHENOMENON ADDRESSED: The aim of this article is to outline and review the emotional sequelae of pediatric oncology nurses’ work and to suggest interventions to support well-being in light of prolonged caregiving. Three major categories that are addressed include the aspects of clinical practice that influence caregiving, the risks of burnout, compassion fatigue, moral distress and grief, and interventions to counteract these phenomena.

RESEARCH LINKAGES: Future-nursing research should focus upon the development of validated, psychometrically sound measurement tools to assess nurse-specific variants of burnout, compassion fatigue, moral distress, and nurse grief. Qualitative research should investigate the relationship between personal variables, workplace and team characteristics, age and experience, and their influence on the predominance of burnout, compassion fatigue, moral distress, and nurse grief. Lastly, the phenomena of resiliency demands further study.


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.


When a child dies in the intensive care unit, many bereaved parents want relationships with their child’s health care staff to continue in the form of follow-up care. However, the nature of these relationships and how they change across the parents’ bereavement journey is currently unknown. This article explores early and ongoing relationships between parents and health care staff when a child dies in intensive care. Constructivist grounded theory methods were used to recruit 26 bereaved parents from four Australian pediatric intensive care units into the study. Data were collected via audio-recorded, semistructured interviews and analyzed using the constant comparative methods and theoretical memoing. Findings show that these relationships focus on Gradually disengaging, commonly moving through three phases after the child dies: Saying goodbye, Going home, and Seeking supports. These findings provide guidance to health care staff on what families need as they leave the intensive care unit and move through bereavement.


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 patient 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 (by $>/=6, scale=1-10$) 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.


Parents of infants hospitalized in a neonatal intensive care unit (NICU) experience increased anxiety and stress, which may persist after discharge. The rationale and design of a randomized clinical trial assessing the impact of a 1-year, post-discharge, peer support intervention (parent navigation) on parental mental health and infant health care utilization is described. Qualitative methods guided the adaptation of an existing parent support program to target emotional and resource-related needs of NICU families. Approximately 300 parent-infant dyads were enrolled at discharge and randomized to either receive a care notebook (control group) or a parent navigator and a care notebook (intervention group). We aim to determine if the parent navigator intervention: 1) increases self-efficacy and decreases stress in parents, 2) decreases overall levels of anxiety and depression in parents, 3) decreases infant hospitalizations and emergency department visits, and 4) increases adherence to infant vaccination recommendations during 1 year of follow-up. Standardized, self-reported psychological scales to assess parent depression, anxiety, self-efficacy and social support were administered at baseline (NICU discharge) and at 1-week, 1-, 3-, 6- and 12-month intervals. Infant immunization status and health care utilization during the study period were also assessed. This paper reviews challenges and successes during implementation. If this intervention improves outcomes, NICUs may choose to provide similar parent navigation interventions for infants and families transitioning from the NICU to home. This study was registered with ClinicalTrials.gov (NCT02643472) on December 31, 2015.


BACKGROUND: Emergency baptism remains an important emotional and spiritual element for many parents of critically ill infants in the neonatal unit. There is no published data available as to which neonates are baptised and their outcomes.

OBJECTIVES: To evaluate trends, outcomes and characteristics of newborn infants baptised over a 15-year period in an Irish maternity hospital. METHODS: Retrospective study of infants baptised in University Maternity Hospital Limerick (UMHL) over a 15-year period. Patients were identified from the ‘register of baptisms’ for the years 2002-2016.

RESULTS: A total of 354 neonates were identified and further information was available for 341. We observed a gradual decline of emergency baptisms over the 15-year period. A total of 114 (32.2%) infants were term and 199 (56.2%) preterm. A total of 288 infants (81.5%) were baptised by Catholic priest, 61 (17.3%) by staff member, 1 (0.3%) by family member and in 3 cases (0.9%) the person baptising was unrecorded. Day of baptism varied from 1 to 88 with a mean age of 4.6 days. A total of 113 (31.9%) neonates died after baptism. Majority of infants baptised were preterm and low birth weight, with predominance of extremely low birth weight (ELBW) who also had proportionately higher mortality 47 (47.5%) following the baptism.

CONCLUSION: Emergency baptism remains an important element in the spiritual care of the critically ill newborn infants and their families. Maternity hospitals and neonatal units should have access to emergency baptism service or other equivalent ‘spiritual blessings’ as appropriate to the faiths followed by the family, especially in an emerging multi-faith population.


Siblings of children with life limiting conditions (LLC) are an important part of the broader family system and require consideration in the holistic care of the family. There can be considerable variation in the functioning and adjustment of these siblings. The current paper explores the resilience paradigm, particularly in the context of siblings of children with LLC and serious medical conditions. The potential impact of children living with a seriously ill brother or sister will be overviewed, and a range of functional outcomes considered. Factors contributing to sibling resilience are
detailed, including individual, family, and broader external and social factors. Given the limited research with siblings of children with LLC, literature has also been drawn from the siblings of children with serious and/or chronic medical conditions. Implications for clinical practice and future research are considered. Pediatric palliative care services may be well placed to contribute to this body of research as they have commonly extended relationships with the families of children with LLC, which span across the child's disease trajectory.

https://www.ncbi.nlm.nih.gov/pubmed/30012977


Two bereaved mothers recount how they made meaning after the deaths of their children, recounting how opportunities to tell their stories in medical settings enabled them to construct narratives that promoted resilience and a sense of control. Pediatric palliative care can be conceived as opening space for patients and guardians to tell their stories outside of the specifics of illness, so medical teams can work to accommodate families' values and goals, thereby initiating the process of meaning making. Viewing videos of parent stories enables medical trainees to enhance their communications skills, empathy, and compassion.

https://www.ncbi.nlm.nih.gov/pubmed/30219225


BACKGROUND: Fear of Progression (FoP) is a commonly reported psychological strain in parents of children with cancer. This expert survey investigates how professionals in pediatric oncology estimate the burden and consequences of FoP in parents and how they assess and treat parental FoP.

METHOD: N=77 professionals in pediatric oncology (members and associates of the Psychosocial Association in Paediatric Oncology and Haematology, PSAPOH) were examined in an online survey with a self-developed questionnaire. Data were analyzed via descriptive statistics and qualitative content analysis.

RESULTS: Three of four experts in clinical practice were (very) often confronted with parental FoP which was associated with more negative (e.g., psychosomatic reactions, reduced family functioning) than positive (e.g., active illness processing) consequences. N=40 experts indicated that they mainly assess parents' anxiety via clinical judgment (72.5%) and/or according to ICD-10/DSM-5 diagnostic criteria (37.5%), whereas standardized methods such as psycho-oncological questionnaires (12.5%) were applied less often. Only n=6 experts named a specific diagnostic approach to assess parental FoP. The most common treatment approaches for FoP were supportive counseling (74.0%), psychotherapy (59.7%) and/or relaxation techniques (55.8%).

DISCUSSION: Parental FoP is frequently perceived by experts in clinical practice. A standardized diagnostic procedure would increase comparability of diagnostic judgments and harmonize treatment indications.


BACKGROUND: This study explored mothers’ perspectives of the experiences and impact on themselves and their family when their child has a life-limiting neurodevelopmental disability.

METHODS: Twelve mothers were interviewed and topics included mothers' experiences of caring, the impact on themselves and their family of care provision, and the management of day-to-day life. Data were analysed using thematic analysis.

RESULTS: Four themes were identified. "Starting Out" relates to mothers’ experiences of the birth of their child and the aftermath. "Keeping the Show on the Road" describes the strategies families employ to manage daily life to day and the resources they use. "Shouldering the Burden" describes the range of physical, psychological, and social consequences of the situation for mothers and the family. "The Bigger Picture" relates to the world outside the family and how this is navigated.

CONCLUSIONS: Findings suggest mothers’ overall experiences are characterized by a constant struggle, with evidence of negative impacts on family life, though there is also evidence of resilience and coping. Implications regarding the provision of services are discussed.

This study aimed to understand, through the bias of everyday life, the phenomenon of maternal mourning. It is a qualitative pilot study on three bereaved mothers. Semi-structured interviews were used for data collection and content analysis was performed using typical categories of daily life. It was possible to observe significant changes in the mothers’ daily lives after their children’s deaths; however, they created strategies that minimized such impacts. Losing a child leads to reconstructing one’s own history and identity. It is believed that daily life is an important bias, and thus its study could help us understand this phenomenon.


ABSTRACT 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.


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 ‘right thing’, (2) act on 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.

PURPOSE: To examine bereaved parents' physical, mental, and social health during the first 6 months after their child's (<12 years) death from a life-threatening illness.

BACKGROUND AND SIGNIFICANCE: Bereaved parents have higher mortality and morbidity rates when compared to nonbereaved parents. Acute illnesses, hospitalizations, and medication changes are highest in the first 6 months. An understanding of bereaved parents' health risk indicators can help inform development of health promotion and disease prevention measures.

Methods and Analysis: A prospective descriptive study examined 8 parent dyads. Parents completed health surveys (Patient-Reported Outcomes Measurement Information System-global, social, and sleep; Brief Symptom Inventory [BSI] 18), which are used to assess parents' health at 3 and 6 months after their child's death. Demographic data included a medical history, hospital or emergency department visits, and smoking and alcohol intake. Descriptive statistics were used to compare parents' scores to US general population scores.

FINDINGS: Mothers' and fathers' physical, mental, and sleep health scores were typically within 1 to 2 standard deviations of the population norms. However, their social health scores were as low as 3 standard deviations and all parents' scores were below population norms. Four (25%) of the 16 parents had new diagnosis during the first 6 months. Based on the BSI-18, 3 parents had their scores above population cutoffs, which warranted a need for further clinical evaluation.

CONCLUSIONS: Health data highlight the "at-risk" health status of bereaved parents. Further validation of these data is required to support the development of health promotion and disease prevention programs.


Background: Families of children at the worst end of the congenital heart disease endure a significant burden which is often not clearly delineated in the clinical literature. We examined the greatest concerns of parents whose children have a Fontan circulation.

Methods: Parents (N=107) of children in the Australian and New Zealand Fontan Registry completed online surveys with open-ended and closed questions. A qualitative method approach incorporating thematic analyses was used.

Results: The greatest concerns for parents of a child with a Fontan circulation were centered on fear of death for their child and psychosocial well-being, followed by lesser themes around anti-coagulation use, pregnancy and financial burdens.

Conclusions: Fear of death and the psychological well-being of their children were the main parental concerns. It highlights the need to clearly communicate information on outcomes to families, and the need for family-focused psychological interventions to improve the psychosocial functioning of both parents and young people.


BACKGROUND: Limited knowledge exists of parents' perceptions and experiences of children's hospices and how these contribute to the varied access and uptake of services. AIM: This study aimed to explore parents’ perspectives and experiences of a hospice, to understand the barriers and/or facilitators to accessing a hospice, and what characteristics parents wanted from hospice provision.

METHODS: A two-phase qualitative study underpinned by a constructivist grounded theory methodology was employed. Phase 1 used focus groups to collect data from parents of children already accessing the hospice (n=24). Phase 2 used in-depth semistructured interviews with parents of children who did not use the hospice (n=7) and with parents who had previous experience of using a hospice (n=7).

RESULTS: A grounded theory of place bonding was developed which illustrates the cognitive journey taken by parents of children with life-limiting conditions considering/receiving hospice care for their child.
CONCLUSIONS: Finding a place where they belonged and felt at ‘home’ made the decision to accept help in caring for their child with a life-limiting condition more acceptable. The theory of place bonding offers children’s hospices a new perspective from which to view how parents access, accept and build relationships at the hospice.

https://www.ncbi.nlm.nih.gov/pubmed/30108042


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.


OBJECTIVE: Our aim was to explore bereaved siblings' positive and negative memories and experiences of their brother’s or sister’s illness and death. METHOD: In our nationwide Swedish study, 174 of 240 (73%) bereaved siblings participated, and 70% responded to two open-ended statements, which focused on siblings' positive and negative memories and experiences of illness and death. The data were analyzed using systematic text condensation.

RESULTS: The bereaved siblings’ responses were categorized into four different themes: (1) endurance versus vulnerability, (2) family cohesion versus family conflicts, (3) growth versus stagnation, and (4) professional support versus lack of professional support. The first theme expressed endurance as the influence that the ill siblings’ strong willpower, good mood, and stamina in their difficult situation had on healthy siblings, whereas vulnerability was expressed as the feeling of emptiness and loneliness involved with having an ill and dying sibling. In the second theme, family cohesion was expressed as the bonds being strengthened between family members, whereas family conflicts often led siblings to feel invisible and unacknowledged. In the third theme, most siblings expressed the feeling that they grew as individuals in the process of their brother’s or sister’s illness and death, whereas others experienced stagnation because of the physical and mental distress they bore throughout this time, often feeling forgotten. In the last theme-professional support-most siblings perceived physicians and staff at the hospital as being warm, kind, and honest, while some siblings had negative experiences.

SIGNIFICANCE OF RESULTS: The study shows that bereaved siblings can have positive memories and experiences. The significance of the positive buffering effect on bereaved siblings’ own endurance, personal growth, family cohesion, and social support should be noted. This knowledge can be valuable in showing healthcare professionals the importance of supporting the siblings of children with cancer throughout the cancer trajectory and afterwards into bereavement.


BACKGROUND: Globally, an estimated eight million children could benefit from palliative care each year. Effective communication about children with life-limiting conditions is well recognized as a critical component of high-quality pediatric palliative care.

OBJECTIVE: To synthesize existing qualitative research exploring healthcare users’ experiences of communicating with healthcare professionals about children with life-limiting conditions.

DESIGN: The results of a systematic literature search were screened independently by two reviewers. Raw data and analytic claims were extracted from included studies and were synthesized using thematic analysis methods for systematic reviews.

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DATA SOURCES: MEDLINE, PubMed, CINAHL, Embase, PsycINFO, Scopus, Web of Science, ProQuest, and ScienceDirect were searched for articles published in English between 1990 and May 2017.

RESULTS: This review included 29 studies conducted across 11 countries and involving at least 979 healthcare users (adults [n = 914], patients [n = 25], and siblings [n = 40]). The four domains of communication experience identified through thematic synthesis are: Information, Emotion, Collaboration, and Relationship. Although included studies were from a range of settings and diverse populations, further research is needed to explore whether and how domains of communication experience differ across settings and populations. In particular, further research about children’s palliative care experiences is needed.

CONCLUSIONS: Healthcare users typically value communication with healthcare professionals: that (1) is open and honest, (2) acknowledges emotion, (3) actively involves healthcare users, and (4) occurs within established and trusting relationships.


OBJECTIVES: Support from healthcare professionals in a PICU is highly valuable for parents of dying children. The way they care for the patients and their families affects the parents’ initial mourning process. This study explores what interaction with hospital staff is meaningful to parents in existential distress when their child is dying in the PICU.

DESIGN: Qualitative interview study. SETTING: Level 3 PICU in the Erasmus University Medical Center-Sophia Children’s Hospital, Rotterdam, and the Netherlands.

SUBJECTS: Thirty-six parents of 20 children who had died in this unit 5 years previously.

INTERVENTIONS: Parents participated in audio-recorded interviews in their own homes. The interviews were transcribed and analyzed using qualitative methods.

MEASUREMENTS AND MAIN RESULTS: Parents’ narratives of their child’s end-of-life stage in the PICU bespeak experiences of estrangement, emotional distancing, and loneliness. Significant moments shared with hospital staff that remained valuable even after 5 years primarily involved personal connectedness, reflected in frequent informational updates, personal commitment of professionals, and interpersonal contact with doctors and nurses.

CONCLUSIONS: Parents whose children died in the PICU value personal connectedness to doctors and nurses when coping with existential distress. Medical and nursing training programs should raise awareness of parents’ need for contact in all interactions but especially in times of crisis and apprehension.


Fardell, J. E., C. E. Wakefield, P. Patterson, A. Lum, R. J. Cohn, S. A. Pini and U. M. Sansom-Daly (2018). "Narrative Review of the Educational, Vocational, and Financial Needs of Adolescents and Young Adults with Cancer: Recommendations for Support and Research." J Adolesc Young Adult Oncol 7(2): 143-147.

Adolescents and young adults (AYAs) with cancer have unique needs around education and vocation during and after treatment. This narrative review series aims at documenting the unique needs of AYAs from the current literature and at providing recommendations to inform an update of the Australian National Service Delivery Framework for AYAs with Cancer. AYAs with cancer may experience impairments to cognitive, physical, and psychological functioning and health, which can adversely affect their academic grades, peer relationships, and likelihood of entering the workforce. Treatment expenses and time off work can stifle AYAs' financial independence from their parents. The combined effect of disrupted education, vocation, and financial dependence can reduce AYAs' sense of identity. Although support is available in some countries, support efficacy is yet to be clearly established. Continued research is required to deliver successful education and work reintegration programs that build the confidence of AYAs with cancer to achieve their best. Educational and vocational support, as well as financial advice, may improve AYAs' financial security and quality of life during survivorship.


OBJECTIVE: To assess the influence of resiliency and stress on parental perspectives of the future quality of life (QOL) of neonatal intensive care unit (NICU) newborns at high risk of neurodevelopmental disability.

STUDY DESIGN: We conducted a prospective multicenter questionnaire study. Perspectives from parents of newborns at high risk of disability as per neonatal follow-up criteria were compared with a low-risk group consisting of parents of all other NICU newborns. Parental anxiety and resiliency, measured using Brief Symptom Inventory and Sense of Coherence scales, respectively, were associated with QOL projections.

RESULTS: Parents returned 129 (81%) questionnaires. Parents considering their newborn as currently sicker were more stressed (P = .011) and worried about future physical (P < .001) and mental (P < .001) health, QOL (P < .001), coping (P = .019), and financial (P < .001) and emotional (P = .002) impact on the family. Overall, there was no difference between parents of high-risk and low-risk newborns on QOL projections. Almost all parents projected a good future QOL. Less resilient parents projected more pain (P = .04), more financial (P = .019), and emotional (P = .031) impact on their family, and were 10 times more likely to predict that their newborn would remain chronically ill.

CONCLUSIONS: Parental projection of future QOL of NICU newborns is not associated with risk of disability. Most parents predict overall a good future QOL and focus more on familial impact. The Sense of Coherence scale may be used in clinical settings to identify less resilient parents.


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 aim was to compare quality of life (QoL) among children and adolescents with different stages of chronic kidney disease (CKD) and determine factors associated with changes in QoL.

DESIGN: Cross-sectional.

SETTING: The Kids with CKD study involved five of eight paediatric nephrology units in Australia and New Zealand.

PATIENTS: There were 375 children and adolescents (aged 6-18 years) with CKD, on dialysis or transplanted, recruited between 2013 and 2016. MAIN

OUTCOME MEASURES: Overall and domain-specific QoL were measured using the Health Utilities Index 3 score, with a scale from -0.36 (worse than dead) to 1 (perfect health). QoL scores were compared between CKD stages using the Mann-Whitney U test. Factors associated with changes in QoL were assessed using multivariable linear and ordinal logistic regression.

RESULTS: QoL for those with CKD stages 1-2 (n=106, median 0.88, IQR 0.63-0.96) was higher than those on dialysis (n=43, median 0.67, IQR 0.39-0.91, p<0.001), and similar to those with kidney transplants (n=135, median 0.83, IQR 0.59-0.97, p=0.4) or CKD stages 3-5 (n=91, 0.85, IQR 0.60-0.98). Reductions were most frequent in the domains of cognition (50%), pain (42%) and emotion (40%). The risk factors associated with decrements in overall QoL were being on dialysis (decrement of 0.13, 95% CI 0.02 to 0.25, p=0.02), lower family income (decrement of 0.10, 95% CI 0.03 to 0.15, p=0.002) and short stature (decrement of 0.09, 95% CI 0.01 to 0.16, p=0.02).

CONCLUSIONS: The overall QoL and domains such as pain and emotion are substantially worse in children on dialysis compared with earlier stage CKD and those with kidney transplants.
INTRODUCTION: Adolescents diagnosed with cancer have to temporarily stop pursuing several vital needs and making plans for the future. The Youth Project in Milan, Italy, focuses on such issues in young cancer patients’ personal lives. Uncertainty about the future had already emerged in several artistic projects designed to help these patients voice their emotions.

METHODS: We describe a project revolving around the question “What shall I do when I grow up?” Twenty-seven 15- to 26-year-old patients wrote a brief account of what they hoped to do as adults. Using theatrical costumes, they then dressed up in their chosen role for a photo shoot with a well-known professional photographer. The results were published in one of Italy’s main national dailies.

RESULTS: Participants reacted differently: some lightheartedly, others more seriously. They often expressed the wish to help others, desiring to return the support and protection they had received as patients. Another common theme concerned their need for normality.

CONCLUSION: By writing about their dreams, these patients gave their doctors a glimpse of their inner world. The project proved an important complement to more conventional forms of psychosocial support and interaction with young patients.


Objective: To examine hope and its components of agency and pathways as predictors of anxiety and depressive symptoms in children receiving cancer treatment.

Methods: Sixty patients (mean standard deviation age = 13.3 (2.7); 57% male) completed Snyder’s Hope Scales, the Children’s Depression Inventory, and the State-Trait Anxiety Inventory at diagnosis and 3 month intervals for 1 year following pediatric cancer diagnosis. Parents also completed Snyder’s Hope Scales. Linear mixed-effect regression was used to assess hope’s role in longitudinal models of symptoms of depression and anxiety.

Results: Agency was a significant predictor of between-patient differences and within-patient changes in symptoms of depression and anxiety. Neither patient pathways nor either component of parent hope was predictive of symptoms of depression or anxiety. Patients who were more likely to have depressive symptoms at baseline were older, diagnosed with leukemia, and non-Hispanic White as opposed to Hispanic. Patient demographics were not predictive of anxiety.

Conclusions: Patient agency is a potential target for intervention to prevent or reduce anxiety and depressive symptoms following pediatric cancer diagnosis.


BACKGROUND: Many parents report a strong desire to take on information-giving roles, and believe they are best positioned to discuss their child’s illness with their child. Healthcare professionals have a supporting role to reduce the burden on parents who feel responsible for conveying information to their child and other family members.

OBJECTIVE: To examine parents’ and healthcare professionals’ perceptions of roles in receiving and communicating information when a child is diagnosed with and treated for acute lymphoblastic leukaemia.

DESIGN, SETTING AND PARTICIPANTS: We used the principles of a grounded theory approach. This was a single site study, recruiting from a principal children’s cancer treatment centre in the United Kingdom. The sample included parents of children receiving and completed treatment for acute lymphoblastic leukaemia (n=28), and healthcare professionals (n=34).
METHODS: Methods included individual interviews, face-to-face and telephone, focus groups, and an online forum.

FINDINGS: Communication ‘touch points’ are many over the course of a child’s cancer journey. We describe often ‘mismatched’ communication encounters where those seeking information and those providing information have different goals. Healthcare professionals in the encounter have expertise at the outset while parents have less expertise, but this expertise grows over time and this can increase the perceptions of this ‘mismatch’ and create different challenges.

CONCLUSIONS: Considered in the context of middle range transition theory, we might suggest that parental foreground (seeking information directly) and background (passive actors) roles are the result of differing levels of uncertainty, and depend on the situation and preferences and child and family needs that may present differently over time in different contexts. Our work contributes to the emerging consensus that communication is more than a core set of skills that healthcare professionals just need to learn: clear specifications of mutual roles, responsibilities and a shared understanding of goals is also essential.


AIMS: To analyze the narratives of illness blogs created by parents of children with cancer.

BACKGROUND: The profound effects of the childhood cancer experience on family members and the turn to the Internet by parents for help in the process are gaining research attention.

DESIGN: The qualitative study design involved secondary narrative analysis of 14 illness blogs: 9 by the parents of children with neuroblastoma and 5 by the parents of children with leukemia. Daily blog entries were analyzed as individual units of illness experience expression and in relation to one another to identify thematic and linguistic similarities.

METHODS: The initial analysis of these illness blogs resulted in identification of the quest for balance as a primary theme. Narratives in parents’ childhood cancer illness blogs illustrated themes of performance. During this initial analysis, however, the author repeatedly asked, "Why are they writing this? And why publish this?" A second analysis of the data answered these questions of why parents blog about the experience. RESULTS: Narrative analysis resulted in the discovery of 6 main reasons that parents wrote and published the childhood cancer experience online: to report, explain, express, reflect, archive, and advocate.

CONCLUSION: The analysis suggests that incorporation of parent writing may improve family--provider communication, enhance the family-health care professional relationship, enhance safety by preventing medical errors, improve reporting of clinical trial data such as adverse events, and improve satisfaction.


BACKGROUND: Parents of children diagnosed with cancer may experience decision regret about cancer treatment decisions and dissatisfaction with the perceived clarity in information received from their child’s providers.

OBJECTIVE: The aim of this study was to describe parental perspectives about receiving an early palliative care and end-of-life (PC/EOL) communication intervention titled ‘Communication Plan: Early through End of Life Intervention’ (COMPLETE) from an interprofessional team of physician and registered nurse providers.

METHODS: Ten parents participated in semistructured interviews after receiving the COMPLETE intervention. The COMPLETE intervention included 3 sessions delivered shortly after diagnosis and at the next 2 cancer treatment evaluations. Sessions of COMPLETE focused on early PC/EOL care discussions at diagnosis and after tumor response evaluations with their child’s providers.

RESULTS: Results included 2 theme categories: (1) COMPLETE nurtures realistic hope and meaningful dialogue by parents connecting with healthcare providers as a dyad, and (2) benefits of COMPLETE helped parents to make informed decisions. In addition, there were offered suggestions to improve COMPLETE.

CONCLUSION: The COMPLETE intervention provided a unique mechanism to foster early discussions about PC/EOL options between parents and an interprofessional team during the first 6 months of the child’s cancer
treatment. Future study is needed using a randomized clinical control-group design to evaluate COMPLETE with a large sample of parents.

IMPLICATIONS FOR PRACTICE: Findings provide promising evidence of parents' preference and receptivity to receive early information about PC/EOL care options for a child with a brain tumor with a poor prognosis. The COMPLETE intervention provided a mechanism to help encourage parental consideration of realistic hoped-for goals for their child's condition and care.


BACKGROUND: Hopes of parents of children with serious illness play an important role in decision-making and coping. Little is known about how parent hopes change over time. We describe the changes in parent hopes across multiple domains and time intervals, examine hopes in a subgroup of parents whose child died, and explore the maintenance of domains over time.

METHODS: In a mixed-methods prospective cohort study on decision-making, parents of seriously ill children reported demographic characteristics and hopes at baseline and reported any changes in hopes at 4-, 8-, 12-, 16-, and 20-month follow-up visits. Hopes were coded into 9 domains. Hope changes and domain changes were identified for each parent at each visit.

RESULTS: One hundred and ninety-nine parents of 158 patients most often reported hopes in the domains of quality of life (75%), physical body (69%), future well-being (47%), and medical care (34%). Hope percentages increased over time for quality of life (84%), future well-being (64%), and broader meaning (21%). The hope domains reported by parents of children who died were similar to the rest of the sample. The majority of parents who completed 5 to 6 follow-up visits changed at least 1 domain. At the individual parent level, some domains revealed considerable change over time, whereas other domains were stable among a subset of parents.

CONCLUSIONS: The specific hopes and overall areas of hope of parents of seriously ill children vary over time, although most hopes fall within 4 major areas. Accordingly, clinicians should regularly check with parents about their current hopes.


BACKGROUND: The past decade saw the establishment of pediatric intensive care units (PICU) across China. This occurred in the context of increasing private shares of medical costs. Payment schemes have not kept pace with the increased availability and demand. As a result a substantial number of parents, in the face of financial constraints, choose to withdraw the medical care of children even when recovery is expected.

OBJECTIVE: We set out to describe the experience of one PICU in Changsha, an industrialized city near the center of the country with a population of 7.3 million.

RESULTS: During the two-year period 883 patients were admitted to the PICU. One hundred one (11%) patients died during their hospital stay. Of these 69 (68%) died after parents elected to withdraw care. A large proportion (33 out of 69 48%) cited economic factors as a reason behind the decision. Compared with the non-withdrawal group the cases had lower disease severity at admission and on the day of death. On the day of death 34% in the withdrawal group had lower disease severity than at admission, showing clinical improvement. The mean hospital charge for the ICU stay was RMB35,000 (~$5600).

CONCLUSION: A substantial proportion of patients in a Chinese urban PICU died after parents chose to withdraw their care in the face of financial hardship, even while some were showing clinical improvement. The society has an obligation, and, likely, an economic incentive, to share this burden.


Within Western cultural traditions, the idea that parents should talk about the death of their child with each other is deeply rooted. However, across bereaved parent couples there are wide variations in communication about their
grief with each other. In this study, we explored the experiences of bereaved couples related to the process of talking and not talking. We used a thematic coding approach to analyze 20 interviews with 26 bereaved parents (11 interviewed as couples, four as individuals). Four main meanings emerged out of our analysis: not talking because of the inadequacy and pointlessness of words in grief, not talking as a way to regulate emotions in daily life, not talking as an expression of a personal, intimate process, and not talking because the partner has the same loss but a different grief process. In addition, we found that the process of talking and not talking can partly be understood as an emotional responsive process on an intrapersonal and interpersonal level. In this process partners search for a bearable distance from their own grief and their partner’s, and attune with their relational context. A better understanding of this process is sought in a dialectical approach, emphasizing the value of both talking and not talking in a tense relationship with each other. Implications for clinical work are described.


BACKGROUND: Healthy lifestyle choices, including participation in regular physical activity, may improve health outcomes in survivors of childhood cancer. We aimed to evaluate the efficacy of a web-delivered physical activity intervention among adolescent survivors to increase moderate to vigorous physical activity (MVPA) and improve fitness and neurocognitive and health-related quality of life (HRQoL) over 24 weeks.

PROCEDURE: This randomized controlled trial was conducted among survivors (aged >/=11 to <15 years) treated at a single institution. Participants were randomized to either a physical activity intervention delivered over the internet or a control group. The intervention group received educational materials, an activity monitor, and access to an interactive website designed to motivate increased physical activity via rewards; the control group received an activity monitor and educational materials. Physical activity, fitness, and neurocognitive and HRQoL outcomes were assessed at baseline and at 24 weeks. Mean changes were compared between groups using paired t-tests.

RESULTS: Of the 97 survivors enrolled, 78 completed the study; the mean age was 12.7 (standard deviation 1.1), 80% were White, and 55.1% were female. Fifty-three survivors were assigned to the intervention and 25 to the control group. While survivors in the intervention group increased, and those in the control group decreased (4.7 +/- 119.9 vs. -24.3 +/- 89.7 min) weekly MVPA, this difference was not significant (P = 0.30). However, hand grip strength, number of sit-ups and pushups, neurocognitive function, and HRQoL outcomes improved in the intervention, but not in the control group.

CONCLUSIONS: An interactive, rewards-based intervention designed to increase MVPA is feasible in adolescent survivors of childhood cancer.


OBJECTIVE: To evaluate the ability of the Perinatal Grief Intensity Scale (PGIS) when used within 8 weeks of perinatal loss to predict intense anxiety and severe depression symptoms in women 3 months later (Time 2 [T2]).

DESIGN: Prospective survey. SETTING: Participants were recruited from hospitals in Louisville, KY and via the Internet.

PARTICIPANTS: Women (N = 103) who experienced perinatal loss.

METHODS: Data were collected using the PGIS, Beck Anxiety Inventory, and the Center for Epidemiologic Studies Depression Scale. We used logistic regression, odds ratios, and receiver operating characteristic curve analysis.

RESULTS: The PGIS had 97.9% sensitivity and 29.6% specificity to predict severe depression symptoms and 95.2% sensitivity and 58.2% specificity to predict intense anxiety at T2. A baseline PGIS score greater than or equal to 3.53 predicted severe depression symptoms (odds ratio = 1.82, 95% confidence interval [CI] [1.46, 2.18], p = .014) and intense anxiety (odds ratio = 1.43, 95% CI [1.07, 1.82], p = .029) at T2. The receiver operating characteristic curves of the PGIS suggest the PGIS performs well at predicting (screening positive) for severe depression symptoms (area under the curve = 0.86, 95% CI [0.79, 0.94], p < .001) and intense anxiety (area under the curve = 0.86, 95% CI [0.78, 0.93], p < .001) after perinatal loss.
CONCLUSION: The PGIS accurately predicted intense anxiety and severe depression symptoms 3 to 5 months after perinatal loss. This instrument may help health care providers identify women who need further mental health evaluation after perinatal loss.


PURPOSE: Symptom burden in children with cancer who are less than 8 years old is not well understood. Our research focuses on identifying how to structure a self-report instrument for younger children. Our aim was to describe how children with cancer, aged 4-7 years, express their symptoms through drawings.

METHODS: Children were asked to make drawings of a day when they were "feeling bad or not good". Content of 18 children’s drawings was analyzed.

RESULTS: Four themes were established: physical symptoms, emotions, location and miscellaneous. Most of the drawings illustrated specific symptoms important to this age group, while also facilitating our understanding of how children with cancer view their symptoms.

CONCLUSION: Having children draw pictures may help initiate communication regarding how they feel, and develop rapport between the interviewer and children.


Aim: To assess parents' ability to express their concerns and hopes for the future in their children with disability and assess their children’s disability as well as to analyse these data for consistency.

Method: Parents of 162 children with spina bifida, spinal muscular atrophy, muscular disorders, cerebral palsy, visual impairment, hearing impairment, mental disability, or disability following brain tumours were asked to freely express their concerns and hopes for the future and to assess disability in their own children by employing a set of 26 International Classification of Functioning, Disability and Health, Children and Youth Version (ICF-CY) body function (b) codes and activity and participation (d) codes. A grounded theory approach was employed to systematize parents’ expressions of concerns and hopes; then, parents scored qualifiers on a 5-step qualitative Likert scale. Parents assessed their children’s disability in the same way using the ICF-CY 5-step qualifier scale.

Results: Altogether, 119 parents freely expressed their concerns and hopes, and 101 of them also assessed their children's disability using the 26 ICF-CY codes. A total of 475 expressions of concern and hopes (issues) were expressed and categorized into 34 areas of concern and hopes (subsections). The most frequently mentioned issues were education; understanding, goodwill, and communication between parents; and community support. Qualitative data on both 5-step qualifier scales showed good reliability. Rasch analysis maps on concerns and hopes for children as well as on the ICF-CY assessment demonstrated good alignment and a clinically relevant progression from the least to the most disabled children.

Conclusion: Parents can express valid and reliable data on their concerns and hopes for the future and can reliably assess disability in their own children.


Siblings of children with cancer often experience negative feelings, and art intervention can help them express their feelings and overcome hardships. This study aimed to develop an Art Intervention Program to improve the psychological adaptation of siblings of children with cancer and to evaluate its effects. Participants were seventeen 7- to 10-year-old siblings of children with cancer. The program comprised 12 sessions conducted once a week. The effects of the intervention were assessed in terms of self-esteem, anxiety, depression, and problem behavior. Self-esteem significantly improved after intervention compared with pretest. Children showed improved scores in externalizing problems and total behavior problems; however, anxiety and depression did not change. The study results indicated that the art intervention program helped improve the self-esteem and helped reduce somatic symptoms, aggressiveness, externalizing problems, and emotional instability among problem behaviors of siblings of
children with cancer. However, the study was limited by its small sample size and the lack of a control group. Therefore, the study design allows no firm conclusions, and a randomized controlled trial is needed to investigate the effectiveness of the program.


The death of a child is a heart-wrenching experience that can have a significant impact on parents, siblings, and families while also often having ripple effects throughout the child's community. Pediatric loss has an impact on family structure and dynamics, individual identity formation, and conceptualization as well as professional practice. This article explores bereavement after a child’s death through the lens of the family, the parent, the sibling, the forgotten grievers, and the provider.


BACKGROUND: To the authors' knowledge, health-related quality of life (HRQOL) outcomes are not well described in patients with medulloblastoma. The use of proton radiotherapy (RT) may translate into an improved HRQOL. In the current study, the authors report long-term HRQOL in patients with proton-treated pediatric medulloblastoma.

METHODS: The current study was a prospective cohort HRQOL study of patients with medulloblastoma who were treated with proton RT and enrolled between August 5, 2002, and October 8, 2015. Both child report and parent-proxy report Pediatric Quality of Life Inventory (PedsQL) surveys were collected at baseline during RT and annually thereafter (score range on surveys of 0-100, with higher scores indicating better HRQOL). Patients were dichotomized by clinical/treatment variables and subgroups were compared. Mixed-model analysis was performed to determine the longitudinal trajectory of PedsQL scores. The Student t test was used to compare long-term HRQOL measures with published means from a healthy child population.

RESULTS: Survey data were evaluable for 116 patients with a median follow-up of 5 years (range, 1-10.6 years); the median age at the time of diagnosis was 7.6 years (range, 2.1-18.1 years). At baseline, children reported a total core score (TCS) of 65.9, which increased by 1.8 points annually (P<.001); parents reported a TCS of 59.1, which increased by 2.0 points annually. Posterior fossa syndrome adversely affected baseline scores, but these scores significantly improved with time. At the time of last follow-up, children reported a TCS of 76.3, which was 3.3 points lower than that of healthy children (P = .09); parents reported a TCS of 69, which was 11.9 points lower than that of parents of healthy children (P<.001). Increased follow-up time from diagnosis correlated with improved HRQOL scores.

CONCLUSIONS: HRQOL scores appear to increase over time after treatment in children treated with proton RT for medulloblastoma but remain lower compared with those of parent-proxy reports as well as published means from a healthy normative sample of children. Additional follow-up may translate into continued improvements in HRQOL. Cancer 2018. (c) 2018 American Cancer Society.


This article describes a family that had to deal with grief from losing a family member. They sought help for 13-yr-old Katie’s prolonged pain following a sprain. The family’s unresolved grief over their second child’s passing continued to emerge, peeping through the small talk, the wry smiles, the daily efforts to move and do, despite physical pain. As the demands of Katie’s rehabilitation stripped away reserves of politeness and energy, the family’s emotional pain became more fully uncovered. The author describes the 6-week course of physical and occupational therapies, pain medicine management, and individual and family behavioral health support. In this period of family crisis, the author was their partner, coach, advocate, and confidante. (PsycINFO Database Record


This study investigated the relationship between the role of the surviving parent in the child’s grieving process, the continuing bond with the deceased parent and biopsychosocial functioning and active grief in adulthood. A survey of 135 adults, parentally bereaved in childhood, indicated that the surviving parent’s role in facilitating the grieving process promoted a positive continuing bond with the deceased in childhood as well as general functioning in adulthood. The continuing bond with the deceased had a weak association with both better general functioning and relational active grief.


OBJECTIVE: To describe the trajectory of patient and caregiver mental health from diagnosis through the first year of treatment for pediatric cancer and assess whether rates of clinically relevant symptoms were elevated compared with norms. We examined mean levels of internalizing and externalizing symptoms and posttraumatic stress symptoms (PTSS) in children with cancer, and depression, anxiety, and PTSS in caregivers during the first year of treatment; the proportion of patients and caregivers that scored in the clinical range at each time point; and the typical trajectory of symptoms in patients and caregivers and whether trajectories differed between individuals.

METHOD: Families (N = 159) of children newly diagnosed with cancer (Mage = 5.6 years; range = 2-18 years) participated in a short-term prospective study. Primary caregivers provided monthly reports of their own and their children’s psychological adjustment.

RESULTS: On average, children were well-adjusted. However, compared with norms, there was a higher than expected proportion of children with clinically relevant internalizing symptoms around the time of diagnosis. On average children’s symptoms declined over time, though variability was observed. Caregivers were less well-adjusted on average, with a high proportion reporting clinically relevant symptoms over time for depression and anxiety. Caregiver symptoms also declined over time, though considerable variability was observed.

CONCLUSION: Although most children remain well-adjusted during the first year of treatment, many caregivers experience clinically relevant symptoms of psychological distress. Implications for development of interventions targeting at-risk patients and caregivers are discussed. Identifying processes that predict between-family variability in trajectories of psychopathology is an important next step. (PsycINFO Database Record


PURPOSE: Although childhood traumatic experiences are recognized as important determinants for adolescent psychiatric health in general, our objective was to explore the specific influence of childhood bereavement on the stress resilience development trajectory.

METHODS: In this national register-based cohort study, we identified 407,639 men born in Sweden between 1973 and 1983, who underwent compulsory military enlistment examinations in late adolescence, including measures of psychological stress resilience. We defined exposure as loss of a first-degree family member in childhood, and estimated relative risk ratios (RRRs) for reduced (moderate or low), compared with high, stress resilience with 95% confidence intervals (CIs) using multinomial logistic regression.

RESULTS: Loss of a parent or sibling in childhood conferred a 49% increased risk of subsequent low stress resilience (RRR, 1.49, 95% CI, 1.41-1.57) and an 8% increased risk of moderate stress resilience (RRR, 1.08, 95% CI, 1.03-1.13) in late adolescence. There was also a graded increase in risk with increasing age at loss; teenagers were at higher risk for low resilience (RRR, 1.64, 95% CI, 1.52-1.77) than children aged 7-12 (RRR, 1.47, 95% CI, 1.34-1.61) and <=6 years (RRR, 1.16 95% CI, 1.02-1.32). The excess risk was observed for all causes of death, including suicide and unexpected deaths as well as deaths due to other illnesses. The associations remained after exclusion of parents with a history of hospitalization for psychiatric diagnoses.

CONCLUSIONS: The long-term consequences of childhood bereavement may include lower stress resilience in late adolescence.


The well-being of parents is essential to the well-being of children with life-limiting illness. Parents are vulnerable to a range of negative financial, physical, and psychosocial issues due to caregiving tasks and other stressors related to the illness of their child. Pediatric palliative care practitioners provide good care to children by supporting their parents in decision-making and difficult conversations, by managing pain and other symptoms in the ill child, and by addressing parent and family needs for care coordination, respite, bereavement, and social and emotional support. No matter the design or setting of a pediatric palliative care team, practitioners can seek to provide for parent needs by referral or intervention by the care team.


ABSTRACT

Objective: The working ages (25-65 years) are a period when most people have significant work, financial, and family responsibilities. A small proportion of working age people will face an expected premature death from cancer or other life-limiting illness. Understanding the impact an expected premature death has on this population is important for informing support. The current study set out to summarize research describing the effects that facing an expected premature death has on employment, financial, and lifestyle of working age people and their families.

METHOD: A systematic review using narrative synthesis approach. Four electronic databases were searched in July 2016 for peer-reviewed, English language studies focusing on the financial, employment, and lifestyle concerns of working age adults living with an advanced life-limiting illness and/or their carers and/or children.

RESULTS: Fifteen quantitative and 12 qualitative studies were included. Two-thirds (n = 18) were focused on cancer. All studies identified adverse effects on workforce participation, finances, and lifestyle. Many patients were forced to work less or give up work/retire early because of symptoms and reduced functioning. In addition to treatment costs, patients and families were also faced with child care, travel, and home/car modification costs. Being younger was associated with greater employment and financial burden, whereas having children was associated with lower functional well-being. Changes in family roles were identified as challenging regardless of diagnosis, whereas maintaining normalcy and creating stability was seen as a priority by parents with advanced cancer. This review is limited by the smaller number of studies focusing on the needs of working age people with nonmalignant disease.

SIGNIFICANCE OF RESULTS: Working age people facing an expected premature death and their families have significant unmet financial, employment, and lifestyle needs. Comparing and contrasting their severity, timing, and priority for people with nonmalignant conditions is required to better understand their unique needs.


The identification of cancer-related fatigue as a clinical problem in pediatric oncology is an important phenomenon, and there are limited number of studies about raising the awareness of pediatric oncology patients and their parents. Fatigue-related education for patients and their parents before and during cancer treatment reduces the fatigue levels of patients. This study aims to analyze the effect of fatigue-related education for pediatric oncology patients aged 7-12 and their parents on their fatigue and quality of life. This study was conducted with 80 children with cancer and their parents who were assigned to either the control group (n = 40) or the experimental group (n = 40). The experimental group received a fatigue-related educational program. The data were collected three times: prior to the program, 3 months later, and 6 months afterwards. Multidimensional variance analysis, the Bonferroni adjusted t test and regression analysis were used to analyze the data. A significant difference was found among the experimental and the control group for total mean scores and the mean scores of subdimensions of the Scale for the Assessment of Fatigue-Child Form in terms of the interactions of group, time, and group*time (p < 0.05). Significant differences were found among the experimental and control groups' mean scores on the Scale for the Quality of Life-Child and Parents Form in terms of the interactions of group, time, and group*time (p < 0.05). Fatigue-related education is an effective education model as a way to reduce fatigue and increase the quality of life of children with cancer. The use of fatigue-related education by nurses in pediatric oncology clinics will have positive effects on children and their parents.


BACKGROUND: Of the nearly 500,000 children in foster care, several hundred children die each year. Their quality of life at end of life is a matter of their foster care experience.

OBJECTIVES: The purpose of this study was to investigate whether serious illness was associated with foster care placement outcomes.

METHODS: US foster care data from 2005 to 2015 were used. Children who were younger than 18 years with residence in the United States were included. Serious illness (i.e., physical health, mental/behavioral health, developmental disabilities) was measured via the foster care files. Two foster care placement outcomes were created (i.e., type of placement, placement instability). Using multinomial and logistic regressions, the influence of serious illness on placement outcomes was evaluated while controlling for demographic, geographic, prior trauma, and foster care support characteristics.

RESULTS: Fifty-seven percent of the children were placed with nonrelatives, 27% in group homes/institutions, and 17% with relatives. Twenty-eight percent experienced placement instability. Serious illness was significantly associated with nonrelative (relative risk ratio [RRR] = 1.97; 95% confidence interval [CI] = 1.58-2.45) and group home/institution placement (RRR = 2.67; 95% CI = 2.09-3.40). Serious illness was not significantly related to placement instability. Children with serious illness were no more likely than their peers to experience multiple foster care placements.

CONCLUSIONS: Foster care youth at end of life were more likely to be placed with nonrelatives or in group homes/institutions. They also did not experience the disruption and stress of being moved to multiple foster homes while seriously ill.


Objective: Although many siblings experience distress after a child’s cancer diagnosis, their psychosocial functioning is seldom assessed in clinical oncology settings. One barrier to systematic sibling screening is the lack of a validated, sibling-specific screening instrument. Thus, this study developed sibling-specific screening modules in English and Spanish for the Psychosocial Assessment Tool (PAT), a well-validated screener of family psychosocial risk.

Methods: A purposive sample of English- and Spanish-speaking parents of children with cancer (N = 29) completed cognitive interviews to provide in-depth feedback on the development of the new PAT sibling modules. Interviews were transcribed verbatim, cleaned, and analyzed using applied thematic analysis. Items were updated iteratively according to participants’ feedback. Data collection continued until saturation was reached (i.e., all items were clear and valid).

Results: Two sibling modules were developed to assess siblings’ psychosocial risk at diagnosis (preexisting risk factors) and several months thereafter (reactions to cancer). Most prior PAT items were retained; however, parents recommended changes to improve screening format (separately assessing each sibling within the family and expanding response options to include *sometimes*), developmental sensitivity (developing or revising items for ages 0-2, 3-4, 5-9, and 10+ years), and content (adding items related to sibling-specific social support, global assessments of sibling risk, emotional/behavioral reactions to cancer, and social ecological factors such as family and school).

Conclusions: Psychosocial screening requires sibling-specific screening items that correspond to preexisting risk (at diagnosis) and reactions to cancer (several months after diagnosis). Validated, sibling-specific screeners will facilitate identification of siblings with elevated psychosocial risk.


This study explored how Danish students experienced returning to school following parental bereavement. Eighteen focus group interviews were conducted with 39 participants aged 9 to 17. All participants had experienced the loss of a primary caregiver. Data collection was divided into two phases. In Phase I, 22 participants from four grief groups
were interviewed 4 times over the course of a year. During Phase II, confirmatory focus groups were undertaken with the 17 participants. This article explores the findings related to ideas and suggestions made by the students about how the Danish school response could be improved to better meet their needs. The presentation of data is divided into seven themes, which are: Desired school response; Desired support from teachers; Desired boundaries between students and teachers; Desired collaboration; Desired support from peers; Desired rules and structure, and; Desires related to gifts and rituals. Study findings indicate that most students want to be included and have a say when the school plans how to respond to their loss. Students further highlight a need for teacher support when having to reconnect with the class; a need for set rules in relation to leaving the class when feeling sad, and; a need for schools to see the loss as a life-changing event, and grief as something that does not simply disappear after a few months. The article concludes by discussing the ways in which the recommendations provided by the participants can be incorporated into a modern revision of Danish school bereavement response plans.


Purpose Communication about prognosis affects decisions patients and family members make about cancer care, and most patients say they want to know about their chances of cure. We sought to evaluate experiences with prognosis communication among adolescents and young adults (AYAs) with cancer. Patients and Methods We surveyed 203 AYAs with cancer age 15 to 29 years (response rate, 74%) treated at Dana-Farber Cancer Institute and their oncologists. Patients were approached within 6 weeks of diagnosis and asked to report on their prognosis communication preferences and experiences, their beliefs about likelihood of cure, and psychosocial outcomes of communication, such as trust (using an item from the Trust in Physician Scale), peace of mind (using select items from the Functional Assessment of Chronic Illness Therapy-Spiritual Well-Being Scale), and anxiety and depression (using the Hospital Anxiety and Depression Scale). Oncologists were asked to report the patient’s likelihood of cure. Results Most patients (83%, 167 of 203 patients) considered prognostic information to be extremely or very important. Patients who reported having received more extensive prognostic disclosure had higher odds of trust in the oncologist (odds ratio [OR], 1.30; 95% CI, 1.01 to 1.67; P = .05), peace of mind (OR, 2.13; 95% CI, 1.29 to 3.51; P = .002), and hope related to physician communication (OR, 1.27; 95% CI, 1.01 to 1.59; P = .04), after adjusting for patient sex, age, race or ethnicity, prognosis, and diagnosis. Disclosure was also associated with lower distress related to knowing about prognosis (OR, 0.65; 95% CI, 0.44 to 0.95; P = .03). However, a majority of patients (62%) reported prognostic estimates that exceeded those reported by physicians (McNemar P < .001). Conclusion Most AYAs with cancer value receiving prognostic information, which is positively associated with aspects of well-being. However, most overestimate chances of cure relative to oncologists, highlighting the importance of efforts to improve communication with this young population.


Context: Pain experienced by children can adversely affect their growth and development. Pain is a major health problem for cancer patients and remains an unresolved problem.

Aim: To know how the experiences of mothers managing their children’s pain during palliative care following cancer diagnosis.

Background: Pain experienced by children can adversely affect their growth and development.

Subject and Methods: Using qualitative methods within a descriptive phenomenological approach, in-depth interviews were conducted with parents (mostly mothers) of eight children diagnosed with cancer. The data were collected using the snowball sampling method.

Results: Participants experienced in managing the pain of children with cancer. Analysis of the results identified 8 themes: the dimensions of pain experienced by children undergoing palliative care; mothers’ physical and psychological responses; mothers’ emotional responses; barriers encountered by mothers when taking care of their child at home; mothers’ interventions to reduce their child’s pain; mothers’ efforts to distract their child from pain; giving encouragement when the child is in pain; and mothers’ efforts and prayers to make their child comfort.

Conclusion: It can be concluded that the child’s pain is the main cause of mothers’ stress and pressure and also affects the daily lives of mothers and children. Along with the most effective intervention, nurses need to provide mothers and children with adequate information about cancer pain.

This paper argues that there is a cultural taboo against the public recognition and expression of perinatal grief that hinders parents' ability to mourn and their psychological adjustment following a loss. It is proposed that this cultural taboo is recreated within the therapy relationship, as feelings of grief over a perinatal loss are minimized or avoided by the therapist and parent or patient. Importantly, it is suggested that if these cultural dynamics are recognized within the therapy relationship, then psychotherapy has the immense opportunity to break the taboo by validating the parent's loss as real and helping the parent to mourn within an empathic and affect-regulating relationship. Specifically, it is suggested that therapists break the cultural taboo against perinatal grief and help parents to mourn through: acknowledging and not pathologizing perinatal grief reactions, considering intrapsychic and cultural factors that impact a parent's response to loss, exploring cultural reenactments within the therapy relationship, empathizing with the parent's experience of loss and of having to grieve within a society that does not recognize perinatal loss, coregulating the parent's feelings of grief and loss, and helping patients to create personally meaningful mourning rituals. Lastly, the impact of within and between cultural differences and therapist attitudes on the therapy process is discussed.


OBJECTIVE To identify the socioeconomic variables which influence the families of the child with cancer. METHODS Quantitative, descriptive, correlational research with 128 families of children with cancer. Three instruments were used: "Questionnaire assessing the impact on the family of children with cancer", "Social Support Satisfaction Scale", and "Graffar Scale". RESULTS Families report increased economic spending due to the disease, with the displacements to hospital and medication. The loss of income by one of the parents also exacerbates the economic impact of the disease. Families with greater support needs and lower social support present greater economic impact. CONCLUSION The social support assumes an important role in the decrease of the economic needs incited by the disease. Nurses must identify the economic needs of the families and become part of the support network of them, being a source of support so they can strengthen themselves in caring.


BACKGROUND: Communication is key in optimizing medical care when a child is approaching end of life (EOL). Research is yet to establish best practices for how medical teams can guide intrafamily communication (including surviving siblings) when EOL care is underway or anticipated for a pediatric patient. While recommendations regarding how medical teams can facilitate communication between the medical team and the family exist, various barriers may prevent the implementation of these recommendations. OBJECTIVE: This review aims to provide a summary of research-to-date on family and medical provider perceptions of communication during pediatric EOL care.

DESIGN: Systematic review.

RESULTS: Findings from a review of 65 studies suggest that when a child enters EOL care, many parents try to protect their child and/or themselves by avoiding discussions about death. Despite current recommendations, medical teams often refrain from discussing EOL care with pediatric patients until death is imminent for a variety of reasons (e.g., family factors and discomfort with EOL conversations). Parents consistently report a need for honest complete information, delivered with sensitivity. Pediatric patients often report a preference to be informed of their prognosis, and siblings express a desire to be involved in EOL discussions.

CONCLUSIONS: Families may benefit from enhanced communication around EOL planning, both within the family and between the family and medical team. Future research should investigate a potential role for medical teams in supporting intrafamily communication about EOL challenges and should examine how communication between medical teams and families can be facilitated as EOL approaches.

PURPOSE: This study aimed to explore health-related information needs of adolescent and young adults (AYAs) and their parent-carers and to examine demographic and clinical variables associated with unmet information needs, including patient activation.

METHODS: In a national cross-sectional study, 196 Australian AYAs diagnosed with cancer between 15 and 25 years and within 24 months of diagnosis and 204 parent-carers reported on total and unmet needs for cancer and health-related information. Fifty-one percent of AYAs were male, 81% had completed treatment and 86% were treated in adult hospitals.

RESULTS: AYAs and parents reported high levels of total need for information. The mean number of unmet needs was 5.63 and 6.82 for AYAs and parents, respectively. AYAs reported the highest unmet needs in relation to their cancer (e.g. late effects and cancer recurrence, and having children in the future). The highest unmet parent information needs were related to medical information about their child as well as information on financial issues for their children and themselves. Unmet information need was associated with psychological distress (posttraumatic stress symptoms) for AYAs and parents. Patient activation was negatively associated with unmet information needs for AYAs. Demographic and treatment variables were not significantly associated with information needs.

CONCLUSION: These findings indicate the importance of information needs for AYAs and their carers. The association between patient activation and information needs suggests that promoting young people’s engagement with healthcare is a key opportunity within AYA care. Parent information needs and associated emotional distress additionally highlight the importance of family-centered care.


Illness blogs are the online narrative expression of the experience of illness and its treatment. The purpose of the present research was to explore, describe, and analyze blog narratives created by parents during their child’s cancer experience in the hope that knowledge generated would amplify the voices of these vulnerable families. The study aimed to answer this question: What themes are evident in illness blogs created by a parent when a child has cancer? The purposive sample of 14 parent blogs included publicly accessible, English language narratives that contained descriptions of life with a child who had undergone treatment for acute lymphocytic leukemia (ALL; five blogs analyzed) or neuroblastoma (nine blogs analyzed) in the previous 5 years or who was currently undergoing treatment for these types of cancer. Analysis resulted in discovery of new knowledge of the uncertainty inherent in daily family life during illness and treatment. The parents’ vivid depictions of the quest for balance while living with prolonged uncertainty during the illness experience suggested new ways to understand experiences of parents of children with cancer.


BACKGROUND: There are no published studies on notification of death by a next of kin to the treating medical staff.

AIM: To explore the content and circumstances of death notifications by next of kin to the treating medical staff in a palliative home care unit.

DESIGN: A cross-sectional study that combines qualitative and quantitative analysis.

SETTING: Assessment of 153 telephone death notifications by a next of kin to the treating medical staff.

RESULTS: The qualitative analysis of death notifications revealed 2 themes: direct and indirect death notifications. In direct notifications, death was portrayed by the notifier in direct and specific words such as death, the patient has died, or the patient is not alive. Indirect notifications included nonspecific or general descriptions of death such as breath cessation, it ended, or it’s over or finished. Direct notifications tended to include specific requests from the medical staff and expressed acceptance and closure, while indirect notifications tended to include more general requests and expressed more panic, distress, or doubt in death. Although spouses were more likely to serve as the primary caregiver, the children or other family members were more likely to notify the treating staff. In 30% of the notifications, there was an element of doubt or uncertainty. Emotions were expressed in 20% of the notifications. Cessation of breathing was the most common physical sign mentioned.
CONCLUSION: Medical staff members who receive notifications of death should expect and be prepared for the expression of varied emotions and doubts as an integral part of the notification.


Despite the high rate of infant mortality in Ghana, few studies have explored the maternal experience of infant loss and the perinatal grieving process. As part of a larger study that interviewed 153 mothers with a sick infant, this 1-year follow-up study reinterviewed eight mothers from the original cohort whose infant died since the study began. Mothers were queried about mental health, coping, and cultural issues related to the loss. Mothers were often discouraged from speaking or thinking about the death due to fear of psychological harm and impact on fertility. Primary coping mechanisms involved seeking support within the community and accepting the loss as God’s will. Mothers desired more communication from health-care providers at the time of death. Despite the cultural norm of silent acceptance in the face of perinatal loss, intense maternal grief and desire to mourn may allow more opportunities for health-care workers to support bereaved mothers.


Background: The responsibility of breaking bad news (BBN) to patients is one of the most difficult tasks of a medical profession.

Aim: The current study aimed to investigate the preferences of mothers of children with cancer about BBN.

Materials and Methods: In this cross-sectional study was conducted in Mashhad during years of 2016, 62 mothers of children with cancer at Dr-Sheikh hospital were recruited by convenience sampling and completed a questionnaire including demographic data and 20 questions about the mothers’ preferences to BBN. Data displayed as percent by SPSS V20 software.

Results: Mothers preferred that BBN conducted by their child’s doctor (93.5%), with an emotional and compassionate way (83.9%), and in a private setting (90.3%). Be told completely about the process of diagnosis (98.4%), meet people with similar conditions (83.9%), receive psychological (85.5%), and religious (79%) support after getting bad news, being in touch with a close relative (82.3%) and applying another term-like malignancy instead of cancer (95.5%).

Conclusion: We tried providing helpful information for developing national guidelines about how to breaking news in Iran, by doing this study.


Context: Parents’ despair and feelings of grief, as well as communication and coordination that is less than adequate between the parents and the palliative team, can affect the provision of a qualified palliative care plan for children and their families.

Aims: This study aims to explore the parents’ experience in caring for children with cancer under palliative care condition.

Setting and Design: The research applied descriptive qualitative phenomenology design.

Subjects and Methods: The study was conducted at Jakarta, Bogor, and Bekasi. Parents who caring their child with cancer under palliative care were in-depth interviewed with open-ended question. Data were then analyzed using the Colaizzi method.

Results: This study resulted in two themes, still hoping for a miracle of God and always being surrendered while under palliative care. Parents still hope for a miracle of God to keep having their children during palliative care. The
forms of surrendering exhibited by the parents in this study are believing in God, praying to God, saying thanks to God, relying on God, and preparing to face the deaths of their children.

Conclusion: This study shows that during palliative care, parents cannot be separated from their relationship with God. Therefore, nurses are vital to the continuous assessment of parents’ spiritual needs and to the facilitation of need fulfillment involving family and religious figures.


Social media as an effective source of information and support among parents and other caregivers of children with cancer has not been explored. The purpose of this cross-sectional study was to describe caregivers’ reasons for using social media, social media sites used, and predictors of social media usage. This study sample included 215 caregivers (96% parents) of children with cancer receiving cancer-related care at a tertiary children’s hospital in the Intermountain West. Most of caregivers (74%) reported using social media in relation to their child’s cancer and reported using social media to provide and receive support and information about their child’s diagnosis or treatment. Our findings suggest that social media could be a delivery platform for future interventions seeking to meet the informational and emotional needs of caregivers of children with cancer. An awareness of how parents and caregivers of children receiving cancer-related treatment use social media can help nurses understand their ongoing informational and emotional needs. Nurses can also support parents and caregivers in selecting reputable sources of support that are accessible via social media.


OBJECTIVES: To examine nurses’ experiences of prognosis-related communication (PRC) with parents of children with cancer.

SAMPLE AND SETTING: Cross-sectional, correlational study in the pediatric oncology setting involving 316 members of the Association of Pediatric Hematology/Oncology Nurses.

METHODS AND VARIABLES: Online survey regarding individual nurse factors, PRC, interprofessional collaboration, moral distress, and perceived quality of care.

RESULTS: Nurses strongly agreed that prognostic disclosure is critical for decision making, but they are challenged in determining their role. Nurses with more years of experience and training in PRC, those working in an outpatient setting, and those with higher levels of nurse-physician collaboration reported more positive experiences with PRC. Positive experiences with PRC and collaboration were significantly associated with higher nurse-perceived quality of care and reduced nurse moral distress.

IMPLICATIONS FOR NURSING: Nurses should work to be active participants in the process of PRC by collaborating with physician colleagues. When nurses sense that prognostic discussions have been absent or unclear, they should feel confident in approaching physician colleagues to ensure parent understanding and satisfaction with communication.


OBJECTIVES: To describe the health outcomes of bereaved parents and identify practical strategies for critical care providers as they support and provide anticipatory guidance to bereaved parents.

DATA SOURCES: PubMed and PsycInfo databases with search terms of bereavement, grief, with parent and pediatric or child complimented by personal experience and knowledge.

STUDY SELECTION: We sought studies describing parental health outcomes and needs of bereaved parents after the death of their child.
DATA SYNTHESIS: A narrative literature review was performed and framed from the perspective of PICU providers who care for bereaved parents. We aimed to describe experiences of grief and short- and long-term consequences and approaches to the care of parents and families in the immediate period and in the months after the death of a child.

CONCLUSIONS: The death of a child is a traumatic experience that can put parents at risk for adverse mental and physical health during bereavement. Health professionals working in PICUs can benefit from knowing these risks to best support bereaved parents, both during their child’s hospitalization and in the early postdeath period. The bereavement experience of parents is an adaptive process, and ongoing professional support may be required for vulnerable families. After the child’s hospitalization and death, a bereavement follow-up meeting with PICU physician(s) and staff may allow parents to gain additional information, emotional support, and provide an opportunity for parents to give feedback on their experiences.


This article examines the women-led natural deathcare movement in the early 21(st) century U.S., focusing upon the movement’s non-coincidental epistemological and gender-political similarities to the natural childbirth movement. Adopting an interdisciplinary approach and drawing upon the author’s intensive interviews with pioneers and leaders of the U.S. natural deathcare movement, as well as from the author’s own participation in the movement, this article argues that the political similarities between the countercultural natural childbirth and natural deathcare movements reveal a common cultural provocation-one that spans the natal transition and the fatal transition.


This article describes the preparation, rationale, and benefits of talking with adolescents who have life-threatening or life-limiting illness about advance care planning (ACP) and end-of-life concerns in a developmentally sensitive manner. The first step is to ensure that a health care provider is ready to work with adolescents in ACP discussions by taking a self-inventory, learning communication skills, and understanding individual barriers. The authors then outline how to assess patient and family readiness, including developmental, cultural, personal, and psychosocial considerations. Evidence-based techniques for respectfully and productively engaging adolescents in ACP conversations are discussed.


BACKGROUND/PURPOSE: Experiences of premature birth and neonatal intensive care unit (NICU) hospitalizations result in stress and family separation that have far-reaching implications. Prior studies of neonatal neurodevelopmental care show improved infant outcomes. Previous studies of mindfulness show improved stress and health outcomes in varied disease processes. No neonatal studies of parent training in mindfulness-based neurodevelopmental care exist. This study examines the impact of parent education and participation in mindfulness-based neurodevelopmental care on parent outcomes (stress, bonding, and satisfaction) and infant length of stay (LOS).

METHODS: This randomized controlled trial pilot study utilized a convenience sample of 55 parent-infant dyads. Parametric and nonparametric statistical tests examined differences in and between study groups in demographics and dependent study variables (stress, bonding, satisfaction, and LOS).

RESULTS: No statistically significant differences in parent outcomes were seen between groups. However, experimental group (EG) parents showed a significant reduction in stress scores from enrollment to discharge (P = .012) and EG infants had significantly shorter LOS (P = .026-.047) than control.

IMPLICATIONS FOR PRACTICE AND RESEARCH: While further research to confirm study results is warranted, changes in current NICU practices to incorporate additional parent education in mindfulness-based neurodevelopmental care may help alleviate parent stress and decrease LOS that impact financial, physical, and psychosocial outcomes for patients, families, healthcare systems, and society.

This paper seeks to examine orphaned children's experiences on grief and loss in Botswana, and its impact on their well-being and make policy recommendations. A cross sectional design which utilized survey questionnaires was employed. Data were collected from 11 districts (3 urban and 8 rural) among orphan children aged 10-18 years. Chi-squared test was used to identify variables believed to be associated with loss and grief. Unadjusted (simple) and adjusted multiple logistic regression was used to investigate factors associated with loss. Of the 732 participants (53.1%) were females and mean age was 13.5 years (SD = 2.7); and 44.6% of these children had experienced death of a close family member in the past year which had been communicated. Children had access to education, lower primary (19.5%), upper primary (39.1%), junior secondary (32.5%), senior secondary school (6.6%), and (0.3%) in tertiary institutions. Most children (98.6%) had not experienced stigma and discrimination at school; 55.2% lived with grandparents, aunts (23.4%), siblings (11.8%), uncles (4.0%), other relatives (3.5%) and non-relatives (0.1%). Unadjusted logistic regression indicated that loss was significantly associated with having someone to talk to (OR = 0.72, 95% CI, 0.53-0.98, p = 0.03), change of residence (OR = 3.08, 95% CI, 1.94-4.90, p < 0.01), having siblings (OR = 2.06, 95% CI, 1.38-3.07, p < 0.01) and being from urban areas (OR = 0.56, 95% CI, 0.41-0.78, p < 0.01). In the adjusted model, loss was significantly associated with change of residence (OR = 2.72, 95% CI, 1.69-4.35, p < 0.01), having siblings (OR = 1.98, 95% CI, 1.30-3.01, p < 0.01) and being from urban areas (OR = 0.65, 95% CI, 0.46-0.93, p = 0.02). Age-specific interventions aimed at addressing the emotional, psychosocial and economic impacts of grief and loss are critical in preventing negative coping behaviors and improving the quality of life of orphans.


BACKGROUND: Admission to the neonatal intensive care unit (NICU) is stressful for parents. Nurses often focus on maternal well-being and fail to acknowledge the stress of fathers. Research on fathers' psychological stress is limited.

PURPOSE: A systematic review of the literature was completed to examine the extent of psychological stress and types of stressors in fathers with infants admitted to the NICU.

METHODS/SEARCH STRATEGY: A search of Ovid MEDLINE, Cochrane Library, PsycINFO, CINAHL, and EMBASE was conducted to identify descriptive and observational studies reporting father-specific stress in the NICU. Studies using observational and descriptive designs, published in English, and reporting father-specific stress outcomes during a NICU admission were eligible for inclusion. Strengthening the Reporting of Observational Studies in Epidemiology guidelines were used for quality assessment.

RESULTS: Fifteen studies met inclusion criteria. Fathers find the NICU environment stressful and are more stressed than fathers of full-term, healthy infants. Parental role alteration, infant appearance, NICU environment, and staff communication are stressors.

IMPLICATIONS FOR PRACTICE/RESEARCH: By recognizing the extent and types of psychological stress in fathers, nurses can provide better support for fathers in their new role. Younger fathers and those with very low birth-weight premature infants may need additional support and resources. Future research on fathers' stress should include larger sample sizes, diverse populations, and tool development and evaluation.


BACKGROUND: Dignity therapy is becoming established in adult settings, with research supporting its effectiveness.

AIMS: This article aims to summarise and synthesise the research that has explored dignity therapy and related meaning-making interventions in palliative care with young people.

METHODS: A rapid structured review was undertaken. Quality appraisal was based on the randomised control trial or cohort study Critical Appraisals Skills Programme (CASP) tool.
RESULTS: Four studies met the inclusion criteria; one focused on young people (7-17 years), the other three included young people but mean ages were 50-70 years. Dignity therapy was found to improve aspects of wellbeing for the patient and was perceived as helpful for the family.

CONCLUSIONS: Dignity therapy is well received, with improvements in measures of wellbeing. However, few studies have included young people (24 years and below). This highlights a clear gap in the literature, suggesting the need to develop and evaluate a dignity therapy or related meaning-making intervention to support young people.


BACKGROUND: Caring for advanced cancer patients affects carers' psychological and physical health. Resilience has been defined as "the process of adapting well in the face of adversity, trauma, tragedy, threats or even significant sources of threat." AIM: The aim of this study was to explore factors promoting carer resilience, based on carers' experiences with and preferences for health care provider support.

DESIGN: Qualitative, semi-structured, individual interviews with family carers of advanced cancer patients were performed until data saturation. The interviews were recorded, transcribed, and analyzed using systematic text condensation.

SETTING/PARTICIPANTS: Carers (n = 14) of advanced cancer patients, not receiving curative treatment, admitted to an integrated curative and palliative care cancer outpatient clinic or to a university hospital cancer clinic, were included.

RESULTS: 14 carers of advanced cancer patients were included; 7 men, 7 women, and mean age of 59 years; 3 were bereaved; 12 were partners; 5 had young and teenage children. Four main resilience factors were identified: (1) being seen and known by health care providers-a personal relation; (2) availability of palliative care; (3) information and communication about illness, prognosis, and death; and (4) facilitating a good carer-patient relation.

CONCLUSION: Health care providers may enhance carers' resilience by a series of simple interventions. Education should address carers' support needs and resilience. Systematic assessment of carers’ support needs is recommended. Further investigation is needed into how health care providers can help carers and patients communicate about death.


Lived experiences of childhood cancer patients and their families have been described as interrupted and as a loss of normal life. Apart from symptoms due to the cancer disease, families continuously experience burden of treatment. Since coping capacities are unique to each individual, we captured variables that offer objective measures of treatment burden, with a particular focus on the disruptive effects of treatment on families’ lives. Our sample was comprised by 193 children that died of cancer. Medical records were extracted retrospectively. Quantitative data were statistically analysed with respect to variables related to treatment burden. Deceased children with cancer and their families faced a significant burden of treatment. Results revealed that deceased leukaemia patients had a higher number of inpatient stays, spent more time in the hospital both during their illness and during the last month of their life, and were more likely to die in the hospital when compared to deceased patients with CNS neoplasms and with other diagnoses. Our findings highlight the disruptive effects of treatment that are likely to have a great impact on families’ daily life, that go beyond exclusively focusing on side effects, and that needs to be taken into account by the treating staff.


BACKGROUND: Several studies have found that the loss of a child is associated with psychiatric health problems, yet few studies examined whether child loss influences psychotropic medication use. This study examined short- and long-term use of psychotropic medication, both before and after the death of a child, and its potential effect modifiers.
METHODOLOGY/PRINCIPAL FINDINGS: A random sample of 205,456 parents, including 902 bereaved parents, were selected from a Finnish total population registry. The analyses were based on linear regressions using generalised estimation equations (GEE) and adjusted for sociodemographic factors. Annual psychotropic use was defined as having purchased prescribed psychotropic medication between 1996 and 2012. Bereaved parents were followed for four years prior to and up to four years after the death of their child. An increase in the use of antidepressants and anxiolytics was found in parents following their loss. The highest percentage of use was found around one year after bereavement, followed by a steady decrease although this remained higher than the level of use among non-bereaved four years after the death. Between 20-25% of bereaved mothers and 10-15% of bereaved fathers used antidepressants or anxiolytics one year after bereavement while the corresponding number in non-bereaved was 5-10%. An increase in psychotropic medication was also found several years before the disease-related loss of a child.

CONCLUSIONS/SIGNIFICANCE: The use of psychotropic medication is markedly higher among parents after losing a child. Patterns of use leading up to and following the death of a child should be further examined in relation to clinical risk factors so as to identify at risk populations.


As physicians, we have lost many children who were under our care despite our best efforts. For most of us, after breaking the news to the family, we move on to treat our next patient who needs help. However, the family and most importantly the siblings have a harder time. The aim of this report is to try to understand how they cope with the loss of a sibling who was previously in intensive care unit.


Children who live with a complex chronic or life-threatening illness face extraordinary challenges. Whether they are receiving disease-oriented treatment (aimed at potential cure or prolongation of life) or palliative treatment—or both concurrently—our challenge is to enhance their comfort and minimize their distress. Symptom management is thus a critical component of pediatric palliative care. Symptoms may be either physical or psychological in nature (or a confluence of both) and their effective management has a direct impact on the child’s quality of life. This article provides an integrative overview of children’s experience of selected physical and psychological symptoms, as expressed through their words and images. Understanding their perspectives is an essential component in the design and provision of optimal symptom management. Included, as well, are examples from siblings—a reminder of the profound impact of illness on these children who also "live" the experience, albeit in a different way. The symptoms that are described are pain, nausea and vomiting, fatigue, weakness, seizures, hair loss, depression, and anxiety. Although psychological symptoms are often inextricable from the physical, they may also present independently as part of the overall illness experience.


BACKGROUND: A considerable number of terminally-ill adult children are outlived by at least one parent and receive palliative care prior to their death. At the same time, adult children continue to be confronted with their parents' terminal illnesses and end-of-life situations. The current study explores the specifics of dyadic interaction at the end of life between a) adult children suffering from a life-threatening disease and their parents, and b) terminally ill parents and their adult children.

METHODS: This prospective observational study aims at filling the existing gap on adult child-parent interaction specifics at the end of life using an exploratory mixed-methods framework. The mixed-methods framework combines a qualitative face-to-face interview and quantitative self-report questionnaires to study the topic at hand. The qualitative interview will focus on experiences, expectations, and wishes with regard to dyadic communication, information about illness and prognosis, expressed and perceived burden and support as well as caregiving role at the end of life. The questionnaires will cover socio-demographics, loneliness, attachment style, social support, and emotional closeness.

DISCUSSION: The research group is currently adjusting a semi-structured interview guide and questionnaire instructions based on the results of a multiprofessional scientific advisory board meeting (Jan. 2018). In a next step, and prior to qualitative and quantitative data collection, the questionnaires will be piloted on patients and their family members in a palliative care setting. The main expected results are i) a description of the specifics of the interaction within and between both dyads, ii) the development of hypotheses and a theoretical framework on the specifics, similarities, and differences for both study groups, and iii) clinical conclusions on specific psychosocial care needs of both groups.

TRIAL REGISTRATION: The study was registered prospectively in the Health Services Research Germany register (Versorgungsforschung Deutschland - Datenbank) (Registration N degrees VID_Dy@EoL_17_003897; date of registration: November 22, 2017) and in the German Clinical Trials Register (Deutsches Register Klinischer Studien) (Registration N degrees DRKS00013206 ; date of registration: October 27, 2017). The study is visible in the International Clinical Trials Registry Platform Search Portal of the World Health Organization under the German Clinical Trials Register number.


This article investigates children's views on providing peer support to bereaved children. The data (pre- and postinterviews and written documents) come from an action research study of a teacher-researcher and her 16 children aged 10-11 years old. Analysis of the data shows children's ideas on supporting a bereaved child and how this support should be provided, taking into consideration various factors such as the relationship with the bereaved and the role of memories. The paper emphasizes that children should have structured opportunities across the whole-school curriculum to learn how loss affects people's lives to support themselves and others.


People’s experiences of living with cancer suggest that spirituality creates purpose and meaning in life for patients. Strengthening spirituality has positive effects on coping, mental health, and symptoms of the disease. This study examines the effect of spiritual care on adolescents coping with cancer. This research is a single-group, quasi-experimental, pre-/poststudy conducted on 32 adolescents. The spiritual care program was presented individually and face to face over six 45-minute sessions held on the admission days. Data were collected using the Ways of Coping Questionnaire by Lazarus and Folkman. A significant difference was observed between the pretest and posttest mean values in all the coping subscales, which suggests the effect of spiritual care on adolescents’ coping with cancer, but no significant differences were observed between the posttest and follow-up mean values, which shows the importance of the continuity of spiritual care in adolescent patients. According to the results, spirituality-based care programs offered by nurses can positively affect adolescents’ coping with cancer and improve their overall coping.


AIMS AND OBJECTIVES: To explore the meaning of rituals that women and their families perform after a stillbirth.

BACKGROUND: A cultural taboo in Taiwan prohibits discussing death; thus, parents of stillborn babies have no established public mourning or burial ceremonies to perform for their stillborn children. Stillbirths are often treated as if they had never happened.

DESIGN: Qualitative descriptive study.

METHODS: In-depth interviews, which were transcribed and content analysed, were conducted with a purposive sample of 16 women discharged from two teaching hospitals in Taiwan after they had a stillbirth.

RESULTS: Families engaged in rituals for two underlying reasons: to benefit the deceased child and the immediate family. The meanings of the rituals for the child are presented through three themes: (i) sending the baby's spirit to a safe place, (ii) protecting it from suffering and (iii) preparing it for a better reincarnation. The meanings of rituals for the families are presented through four themes: (i) releasing parental guilt by doing their best for the deceased child, (ii) cutting bonds with the child, (iii) avoiding additional misfortune should they mishandle the funeral and (iv) praying for a successful subsequent pregnancy.

CONCLUSIONS: Death-related rituals are highly culturally diverse. This study fills a gap about Asian cultures. Participating in rituals permits a mother to do something for her deceased child, helps relieve her guilt and lets her cope with the stillbirth. Rituals after a stillbirth can help a woman recover from grieving and allow her to hope for a successful subsequent pregnancy.

RELEVANCE TO CLINICAL PRACTICE: Health professionals should discuss with bereaved parents what rituals they would like to perform and then respect their decisions. A continuum of care and support that exists from the prenatal diagnosis through the stillbirth and beyond is recommended for parents and families during this difficult time.


The issue of quality of life, particularly of patients affected by cancer, is very controversial, especially with adolescent patients who have no prospects of recovery. This article aims to describe the psychological mechanisms, which
affect teenage patients with terminal cancer, in order to allow the best quality of life possible. The adaptation of teenager patients suffering from terminal illnesses is also related to other non-medical issues such as psychological, legal and ethical considerations. A correct and balanced communication with such patients is needed, in order of maintaining a positive mental approach (“hope”) so that the teens are able to come to terms with the hard reality. Several case studies are presented, that according to the authors show the importance of developing an area of illusion, which allows patients to face up to the extreme distress and anxiety of their reality, without completely denying the reality itself. These hypotheses at the moment need more empirical evidences but they demonstrate the complex mental process involved with terminally ill adolescent patients coming to terms with their situations and show the importance of considering such process in all aspects of clinical care and treatment, in order to best address their physical and psychological well-being.


People seek to spend time in positive experiences, enjoying and savoring. Yet there is no escaping negative experiences, from the mundane (e.g. arguing) to the massive (e.g. death of a child). Might negative experiences confer a hidden benefit to well-being? We propose that they do, in the form of enhanced meaning in life. Research suggests that negative experiences can serve to boost meaning because they stimulate comprehension (understanding how the event fits into a broader narrative of the self, relationships, and the world), a known pillar of meaning in life. Findings on counterfactual thinking, reflecting on events’ implications, and encompassing experiences into broad-based accounts of one’s identity support the role of comprehension in contributing to life’s meaning from unwanted, unwelcome experiences.


OBJECTIVE: The objective was to compare mental illness diagnoses and treatment use among mothers who lost custody of their child through involvement with child protection services and those seen in mothers dealing with the death of a child.

METHODS: We studied mental health outcomes of a cohort of women whose first child was born in Manitoba, Canada between 1 April 1997 and 31 March 2015. Of these women, 5,792 had a child taken into care, and 1,143 mothers experienced the death of a child (<18 y old) before 31 March 2015. Adjusted relative rates (ARR) of 3 mental health diagnoses and 3 mental health treatment use outcomes between these 2 groups were examined.

RESULTS: Mothers with a child taken into care had significantly greater ARR of depression (ARR = 1.90; 95% CI, 1.82 to 1.98), anxiety (ARR = 2.51; 95% CI, 2.40 to 2.63), substance use (ARR = 8.54; 95% CI, 7.49 to 9.74), physician visits for mental illness (ARR = 3.01; 95% CI, 2.91 to 3.12), and psychotropic medication use (ARR = 4.95; 95% CI, 4.85 to 5.06) in the years after custody loss compared with mothers who experienced the death of a child.

CONCLUSION: Losing custody of a child to child protection services is associated with significantly worse maternal mental health than experiencing the death of a child. Greater acknowledgement and supportive services should be provided to mothers experiencing the loss of a child through the involvement of child protection services.


BACKGROUND: Losing a child is devastating for parents and grandparents. Family and friends generally focus on comforting and supporting the bereaved parents, unintentionally ignoring the bereaved grandparents. Grandmothers and grandfathers often struggle with wanting to help their adult children (deceased child’s parents) without usurping the parents’ responsibilities and decisions regarding the deceased child. Research on mothers’ and grandmothers’ health at about the same time after the same child’s death in the neonatal or pediatric intensive care unit is lacking. The aim of this study was to compare mothers and grandmothers on physical health, mental health, and functioning in the first 1-6 months after the same child’s death in a neonatal or pediatric intensive care unit.
METHODS: This cross-sectional secondary analysis compared 32 mothers with 32 grandmothers of the same 32 deceased children (newborn-6 years). Grandmothers were recruited through these 32 mothers. Most grandmothers and mothers were Hispanic (25%, 34%) or Black (44%, 41%), respectively. Mothers and grandmothers separately completed questions about their Physical Health, Mental Health [depression (Beck Depression Inventory), Post-Traumatic Stress Disorder (PTSD, Impact of Events-R), grief (Hogan Grief Reaction Checklist)], and Functioning (social support [MSPSS] and Employment) since the child’s/grandchild’s death. Paired t-tests and Chi Square tests were used to compare grandmothers with mothers of the same deceased infant/child on their private and separate responses to study measures.

RESULTS: Mothers had significantly more acute illnesses than grandmothers. More mothers (63%) than grandmothers (37%) were categorized as clinically depressed. More mothers (69%) than grandmothers (44%) had clinical PTSD. Mothers reported significantly higher levels of despair and detachment than grandmothers. Only 4 mothers and 2 grandmothers were in therapy at the time of interview. Grandmothers and mothers rated their ability to concentrate on their work and their level of social support similarly.

CONCLUSIONS: Mothers had more acute illnesses, more severe depression, and a higher level of grief than grandmothers. However, few received therapy despite their high levels of depressive and PTSD symptoms.


Shidu is the Chinese transliteration for 'losing an only child,' which indicates the death of the only child in the family. Shiduers are parents who have lost their only child. The grief research scholar Neimeyer (2012) argued that grief research should consider the role of different cultures in the grieving process. Familism culture is a collectivist culture that has a profound effect on Chinese society and is likely to produce a significant effect on the grieving process of shiduers; however, this effect has not yet received systematic attention in research. To explore the effect of familism culture on the grief of shiduers, we conducted semi-structured personal interviews in Beijing, China, with seven shiduers. The study results show that the effect of familism culture on the grief of shiduers includes three levels: cognition, emotion, and behavior. These levels are reflected in a variety of relationships, including relationships with ancestors, the deceased child, the spouse, relatives, Tong Ming Ren (the Chinese transliteration of 'people who share the same fate'), and the country. The first four types of relationships are reflections of 'direct familism culture,' and the latter two types of relationships are reflections of 'extended familism culture'. The relationships with the deceased child, relatives, and Tong Ming Ren are mainly supportive; the relationship with ancestors is mainly stressful; the relationship with the spouse has a dual nature; and the relationship with the country is contradictory.

Over time, shiduers have abandoned the concept of familism culture and have moved toward reducing stress and increasing supportiveness. Psychological professionals, social workers, and government staff may refer to the results of this study to help shiduers obtain support and reduce stress from the described relationships. Specific suggestions are provided in the text.


Bereavement may trigger different psychological outcomes, such as prolonged grief disorder or post-traumatic growth. The relationship between these two outcomes and potential precipitators remain unknown. The current study aimed to identify classes of Chinese bereaved individuals based on prolonged grief symptoms and post-traumatic growth and to examine predictors for these classes. We used data from 273 Chinese individuals who lost a relative due to disease (92.3%), accident (4.4%) and other reasons (1.8%). Latent class analysis revealed three classes: a resilient class, a growth class, and a combined grief/growth class. A higher level of functional impairment was found for the combined grief/growth class than for the other two classes. Membership in the combined grief/growth class was significantly predicted by the younger age of the deceased and the death of a parent, child or spouse. Subjective closeness with the deceased and gender were marginally significant predictors. When the four variables were included in the multinomial regression analysis, death of a parent, child or spouse significantly predicted the membership to the combined grief/growth class. These findings provide valuable information for the development of tailored interventions that may build on the bereaved individuals' personal strengths.

Providing Services for Children and Families


Paediatric Palliative Care: What is different in children compared to adults? Abstract. The number of children and adolescents (0 - 18 years) with life-limiting conditions and needs for paediatric palliative care (PPC) is rising. In Switzerland, the awareness for these needs lags largely behind other developed countries. In the United Kingdom, the prevalence for children with life-limiting conditions and PPC needs was estimated at 32 children per 10'000 population (0 - 19 years). In Switzerland, this would correspond to an absolute number of 5'000 children living with a life-limiting condition and potentially in need of PPC. In contrast, the number of deaths accounts for around 500 children (0 - 18 years) every year. Most common causes of death are perinatal conditions, contributing to nearly 50 % of all deaths in childhood, followed by accidents and complex chronic conditions such as genetic / congenital disorders, neurological and cardiac conditions and cancer. Compared to adults with palliative care needs, the group of children is significantly smaller but at the same time highly heterogenic. Heterogeneity relates to: the whole age continuum from neonates, infants and children to adolescents; a broad spectrum of diseases including rare diseases; a variety of needs due to age, development and the illness, e. g. needs for specialist care or technical support; various in- and outpatient settings. Paediatric care always encompasses the whole family and their particular needs. Internationally, hospital-based programmes have been developed and implemented to meet these particular needs of children and their families.


BACKGROUND/OBJECTIVE: Our home-care unit (HCU) is specialized for pediatric cancer patients and has a strong palliative care activity. We believe that the introduction of home-care services can influence the place of palliative care and of death as well as the length of hospitalization. We aimed at describing characteristics and care course of patients treated in our HCU, and tried to identify some factors contributing to home care at the end of life.

DESIGN/METHODS: We conducted a retrospective, observational, monocentric study about patients in pediatric onco-hematology, treated at least one day in our home-care unit, who died between July 1st 2013 and December 31st 2015. Statistical analysis was descriptive and analytic.

RESULTS: A total of 74 patients known by our HCU died during study period. Eight were excluded. Forty-three out of 66 patients died at home. During the last 3 months of life, oncology patients have significantly less classical hospitalization, when compared to hematology patients. The implication of general physicians (GP) and nurses and information given to the family increase the possibility for home death. No significant association was found between ages at death, distance between home and hospital, other life conditions and place of death.

CONCLUSIONS: Our HCU has a strong palliative care activity and a high rate of children dying at home. Good collaborations between our pediatric onco-hematology team and our HCU as well as between our HCU and caregivers optimize palliative care.


OBJECTIVES: To discuss the role of investigations after death in children as part of a supplement on "Death and Dying in the PICU."

DATA SOURCES: Literature review, personal experience, and expert opinion.

DATA SELECTION: Not applicable. DATA EXTRACTION: Moderated by three experts on investigations after death in children.

DATA SYNTHESIS: Not relevant.

CONCLUSIONS: A multidisciplinary cliniciopathologic conference is important after the death of a child in order to help bring closure to the family and to attempt to address any concerns they may have about the care. It is also an important part of the quality of care process for a tertiary care institution and provides an unique opportunity for
ongoing medical education. The model of a multidisciplinary clinicopathologic conference used by the Ontario Coroner’s Office to investigate sudden and unexpected deaths in children under 5 years old, which has been functioning for over 30 years, is described. Reports from this Pediatric Death Review Committee have been influential in improving the care of children in the province of Ontario.


Despite vast improvements in disease-based treatments, many children live with life-threatening disorders that cause distressing symptoms. These symptoms can be difficult to comprehensively assess and manage. Yet, frequent and accurate symptom reporting and expert treatment is critical to preserving a patient’s physical, psychological, emotional, social, and existential heath. We describe emerging methods of symptom and health-related quality-of-life (HRQOL) assessment through patient-reported outcomes (PROs) tools now used in clinical practice and novel research studies. Computer-based and mobile apps can facilitate assessment of symptoms and HRQOL. These technologies can be used alone or combined with therapeutic strategies to improve symptoms and coping skills. We review technological advancements, including mobile apps and toys, that allow improved symptom reporting and management. Lastly, we explore the value of a pediatric palliative care interdisciplinary team and their role in assessing and managing distressing symptoms and minimizing suffering in both the child and family. These methods and tools highlight the way that novel, new, and innovative approaches to symptom assessment and management are changing the way that pediatrics and pediatric palliative care will be practiced in the future.


Background: Medical advances continue to improve morbidity and mortality of serious pediatric diseases, including cancer, driving research addressing diminished physical and psychological quality of life in children with these chronic conditions. Empowerment enhances resilience and positively influences health, disease, and therapy understanding. We describe the development and usability assessment of a prototype Empower Stars! mobile video game grounded in behavioral and exercise theories with the purpose of coupling physical exercise with empowerment over disease in children with cancer.

Methods: Academic faculty, health-care providers, and community video game developers collaborated in this project. The iPadAir was selected as a delivery platform for its accelerometer and gyroscope features facilitating exercise design. Unity multiplatform technology provided animation and audiovisual features for immediate player feedback. Javascript, C#, Photoshop, Flash, and SketchUp were used for coding, creating graphical assets, Sprite sheets, and printing files, respectively. 3D-printed handles and case backing were used to adapt the iPad for physical exercise. Game usability, engagement, and enjoyment were assessed via a multilevel study of children undergoing cancer chemotherapy, their parents, and pediatric cancer health-care providers. Feedback crucial for ongoing game development was analyzed.

Results: A prototype Empower Stars! mobile video game was developed for children 7-14 years old with cancer. Active, sedentary, educational, and empowerment-centered elements intermix for 20 min of exercise within a 30 min "one-day treatment" gameplay session involving superheroes, space exploration, metaphorical cancer challenges, life restoration on a barren planet, and innumerable star rewards. No player "dies."* Usability assessment data analyses showed widespread enthusiasm for integrating exercise with empowerment over cancer and the game itself. Favorite elements included collecting star rewards and planet terraforming. Traveling in space and the Healthy Food Choice game were least liked. The need for improved gameplay instructions was expressed by all groups. The usability study provided essential feedback for converting the prototype into alpha version of Empower Stars!

Conclusion: Adapting exercise empowerment-promoting video game technology to mobile platforms facilitates usability and widespread dissemination for children with cancer. We discuss broader therapeutic applicability in diverse chronic pediatric diseases, including obesity, asthma, cystic fibrosis, diabetes, and juvenile idiopathic arthritis.

Social media is an important access point for engagement of children and adolescents. For individuals with a life-limiting illness or serving as the caregiver for an ill child, social media can be a helpful outlet for support and information gathering. It has democratized the process of being remembered through providing an ongoing account of thoughts, pictures, and videos that theoretically live on forever via a digital legacy. Providers should be familiar with how this new generation uses social media during their illness, after death, and in the bereavement process.


OBJECTIVES: To describe pediatric palliative care (PPC) in pediatric oncology, the importance of PPC for pediatric oncology patients, disparities within pediatric palliative oncology, innovative strategies for improving PPC access in underserved populations, and implications for oncology nursing practice.

DATA SOURCES: Published pediatric oncology and palliative peer-reviewed articles and guidance documents.

CONCLUSION: Disparities exist within pediatric palliative oncology. There is much work needed to improve the reach and quality of PPC for pediatric oncology patients, especially those from underserved populations.

IMPLICATIONS FOR NURSING PRACTICE: Nurses serve a critical role in advocating for PPC for seriously ill pediatric cancer patients and their 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.


Background: Survival of infants with complex care has led to a growing population of technology-dependent children. Medical technology introduces additional complexity to patient care. Outcomes after NICU discharge comparing Usual Care (UC) with Comprehensive Care (CC) remain elusive.
Objective: To compare the outcomes of technology-dependent infants discharged from NICU with tracheostomy following UC versus CC.

Methods: A single site retrospective study evaluated forty-three (N=43) technology-dependent infants discharged from NICU with tracheostomy over 5(1/2) years (2011-2017). CC provided 24-hour accessible healthcare-providers using an enhanced medical home. Mortality, total hospital admissions, 30-days readmission rate, time-to-mechanical ventilation liberation, and time-to-decannulation were compared between groups.

Results: CC group showed significantly lower mortality (3.4%) versus UC (35.7%), RR, 0.09 [95%CI, 0.12-0.75], P=0.025. CC reduced total hospital admissions to 78 per 100 child-years versus 162 for UC; RR, 0.48 [95% CI, 0.25-0.93], P=0.03. The 30-day readmission rate was 21% compared to 36% in UC; RR, 0.58 [95% CI, 0.21-1.58], P=0.29. In competing-risk regression analysis (treating death as a competing-risk), hazard of having mechanical ventilation removal in CC was two times higher than UC; SHR, 2.19 [95% CI, 0.70-6.84]. There was no difference in time-to-decannulation between groups; SHR, 1.09 [95% CI, 0.37-3.15].

Conclusion: CC significantly decreased mortality, total number of hospital admissions and length of time-to-mechanical ventilation liberation.


Background: Palliative care is recognized as an important component of care for children with cancer and other life-limiting conditions. In resource limited settings, palliative care is a key component of care for children with cancer and other life-limiting conditions. Globally, 98% of children who need palliative care live in low- or middle-income countries, where there are very few palliative care services available. There is limited evidence describing the practical considerations for the development and implementation of sustainable and cost-effective palliative care services in developing countries.

Objectives: Our aim is to describe the key considerations and initiatives that were successful in planning and implementing a hospital-based pediatric palliative care service specifically designed for a resource-limited setting.

Setting: Bangabandhu Sheikh Mujib Medical University (BSMMU) is a tertiary referral hospital in Bangladesh. Local palliative care services are very limited and focused on adult patients. In partnership with World Child Cancer, a project establishing a pediatric palliative care service was developed for children with cancer at BSMMU.

Results: We describe four key elements which were crucial for the success of this program: (1) raising awareness and sensitizing hospital administrators and clinical staff about pediatric palliative care; (2) providing education and training on pediatric palliative care for clinical staff; (3) forming a pediatric palliative care team; and (4) collecting data to characterize the need for pediatric palliative care.

Conclusion: This model of a hospital-based pediatric palliative care service can be replicated in other resource-limited settings and can be expanded to include children with other life-limiting conditions. The development of pilot programs can generate interest among local physicians to become trained in pediatric palliative care and can be used to advocate for the palliative care needs of children.


OBJECTIVES: To describe practical considerations and approaches to best practices for end-of-life care for critically ill children and families in the PICU.

DATA SOURCES: Literature review, personal experience, and expert opinion. STUDY SELECTION: A sampling of the foundational and current evidence related to the withdrawal of life-sustaining therapies in the context of childhood critical illness and injury was accessed.

DATA EXTRACTION: Moderated by the authors and supported by lived experience.

DATA SYNTHESIS: Narrative review and experiential reflection.
CONCLUSIONS: Consequences of childhood death in the PICU extend beyond the events of dying and death. In the context of withdrawal of life-sustaining therapies, achieving a quality death is impactful both in the immediate and in the longer term for family and for the team. An individualized approach to withdrawal of life-sustaining therapies that is informed by empiric and practical knowledge will ensure best care of the child and support the emotional well-being of child, family, and the team. Adherence to the principles of holistic and compassionate end-of-life care and an ongoing commitment to provide the best possible experience for withdrawal of life-sustaining therapies can achieve optimal end-of-life care in the most challenging of circumstances.


Purpose Palliative care remains an urgent, neglected need in the developing world. Global disparities in end-of-life care for children, such as those with advanced cancers, result from barriers that are complex and largely unstudied. This study describes these barriers at Bugando Medical Center, one of three consultant hospitals in Tanzania, to identify areas for palliative care development suitable to this context. Methods In-depth interviews were conducted with 20 caregivers of pediatric patients with cancer and 14 hospital staff involved in pediatric end-of-life care. This was combined with 1 month of participant observation through direct clinical care of terminally ill pediatric patients. Results Data from interviews as well as participant observation revealed several barriers to palliative care: financial, infrastructure, knowledge and cultural (including perceptions of pediatric pain), and communication challenges. Although this study focused on barriers, what also emerged were the unique advantages of end-of-life care in this setting, including community cohesiveness and strong faith background. Conclusion This study provides a unique but focused description of barriers to palliative care common in a low-resource setting, extending beyond resource needs. This multidisciplinary qualitative approach combined interviews with participant observation, providing a deeper understanding of the logistical and cultural challenges in this setting. This new understanding will inform the design of more effective-and more appropriate-palliative care policies for young patients with cancer in the developing world.


OBJECTIVE: To understand how family members view the ways Emergency Medical Services (EMS) and other first responders interact with distressed family members during an intervention involving a recent or impending pediatric death. METHODS: In depth interviews with 11 grieving parents of young children and survey results from 4 additional grieving parents of adult children were conducted as part of a larger study on effective ways for EMS providers to interact with distressed family members during a pediatric death in the field. The responses were analyzed using qualitative content analyses.

RESULTS: Family reactions to the crisis and the professional response by first responders were critical to family coping and getting necessary support. There were several critical competencies identified to help the family cope including: (1) that first responders provide excellent and expeditious care with seamless coordination, (2) allowing family to witness the resuscitation including the attempts to save the child’s life, and (3) providing ongoing communication. Whether the child is removed from the scene or not, keeping the family apprised of what is happening and why is critical. Giving tangible forms of support by calling friends, family, and clergy, along with allowing the family time with the child after death, giving emotional support, and follow-up gestures all help families cope.

CONCLUSION: The study generated hypothetical ways for first responders to interact with distressed family members during an OOH pediatric death.

Fardell, J. E., P. Patterson, C. E. Wakefield, C. Signorelli, R. J. Cohn, A. Anazodo, B. Zebrack and U. M. Sansom-Daly (2018). "A Narrative Review of Models of Care for Adolescents and Young Adults with Cancer: Barriers and Recommendations." J Adolesc Young Adult Oncol 7(2): 148-152.

Adolescents and young adults (AYAs) with cancer have unique cancer care needs that differ significantly from younger and older cancer patients. There has been an increasing appreciation for this unique group of patients internationally, with a rise in research and establishment of innovative models of care dedicated to addressing their
needs. This narrative review identifies common elements and barriers to care for AYA cancer patients and survivors, and offers recommendations for developing clinical care models for this age-defined population.


BACKGROUND: The multidisciplinary standardized interventions to educate and support patients with congenital heart disease (CHD) are described as “Transition Clinic” (TC). TC represents a key element to deliver care for patients during the transition from childhood to adulthood. So far, there is a lack of empirical evidence regarding the impact of TC models on the improvement of health perception in adolescent patients with CHD (CHD-specific TC model). For this reason, the aim of this study is assess the impact of the TC model on CHD adolescent patients’ health perception outcomes.

METHODS: This study has a quasi-experimental design. Quality of life, satisfaction and health perception were assessed in T0 and after 1 year from enrollment (T1). During the follow-up period, the patients enrolled (aged 14 to 21 years) were involved in the CHD-specific TC model.

RESULTS: The results are referred to the first 100 patients enrolled (mean age 14.79 +/- 1.85 years; 60% male), as they have already completed the follow-up. The overall study is currently ongoing. According to Warnes' classification, 29% of patients had simple heart defects, 46% showed moderate complexity, and 25% showed severe complexity. The comparison between T0 and T1 showed statistically significant improvement in T1 regarding pain/discomfort, anxiety/depression and perception of health status (EQ-5D), general satisfaction and quality of life (LAS QoL).

CONCLUSIONS: The preliminary results showed in this study are encouraging, and confirm the need to create a multidisciplinary standardized intervention of education and support to deliver care for adolescent patients with CHD.


Among the over 21 million children with life-limiting conditions worldwide that would benefit annually from a pediatric palliative care (PPC) approach, more than eight million would need specialized PPC services. In the United States alone, more than 42,000 children die every year, half of them infants younger than one year. Advanced interdisciplinary pediatric palliative care for children with serious illnesses is now an expected standard of pediatric medicine. Unfortunately, in many institutions there remain significant barriers to achieving optimal care related to lack of formal education, reimbursement issues, the emotional impact of caring for a dying child, and most importantly, the lack of interdisciplinary PPC teams with sufficient staffing and funding. Data reveals the majority of distressing symptoms in children with serious illness (such as pain, dyspnea and nausea/vomiting) were not addressed during their end-of-life period, and when treated, therapy was commonly ineffective. Whenever possible, treatment should focus on continued efforts to control the underlying illness. At the same time, children and their families should have access to interdisciplinary care aimed at promoting optimal physical, psychological and spiritual wellbeing. Persistent myths and misconceptions have led to inadequate symptom control in children with life-limiting diseases. Pediatric Palliative Care advocates the provision of comfort care, pain, and symptom management concurrently with disease-directed treatments. Families no longer have to opt for one over the other. They can pursue both, and include integrative care to maximize the child’s quality of life. Since most of the sickest children with serious illness are being taken care of in a hospital, every children’s hospital is now expected to offer an interdisciplinary palliative care service as the standard of care. This article addresses common myths and misconceptions which may pose clinical obstacles to effective PPC delivery and discusses the four typical stages of pediatric palliative care program implementation.


OBJECTIVE: Mature B-cell non-Hodgkin lymphoma (B-NHL) comprises more than 50% of all non-Hodgkin lymphoma (NHL) in children and adolescents. An official report published by the Mexican National Center for the Control and Prevention of Cancer in the Pediatric and Adolescent Populations, reported a lymphoma OS of 71% (including all Hodgkin and NHL). The Mexican Association of Pediatric Oncology and Hematology conducted a retrospective study to analyze the clinical characteristics and outcomes of children with diagnosis of B-NHL in Mexico, in order to perceive the main areas of improvement in the health care. METHODS: From 1 January 2000 to 31 December 2016, 166 pediatric patients were diagnosed with B-cell NHL at the participant institutions. RESULTS: According to histology the outcomes were 5-year EFS 63%, for BL/BLL, and 80% DLBCL, (P = .051), 5-year PFS 81%, for BL/BLL, and 91% for DLBCL, (P = .126), and 5-year OS 71%, for BL/BLL, and 83% for DLBCL, (P = .095). DISCUSSION: Overall, 18 patients died due to acute treatment toxicity, resulting in a cumulative incidence of toxic death of 10.84% and an early death rate of 7.23%, defined as <30 days after initial treatment. In conclusion, there is an urgent need to establish an academic collaboration to create strategies to improve pediatric cancer care according to our resources, especially in diseases with expected excellent prognosis as B-NHL. These strategies must include comprehensive supportive care, early referral, and the creation of easy communication between pediatric and adults centers as well as late-effects clinics.


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.


While bereavement camps serve as a support for children, this study examines a therapeutic recreation-based camp for families who have lost a child. The study triangulated documents, researcher reflection, and staff interviews to highlight the themes of Searching & Finding, Getting to Know, Finding the Balance, and Joining. Developing opportunistically through internal and external factors, the camp’s evolution represents a closing of the loop, from supporting families of living children to also supporting the families of children who have died. Understanding the camp’s evolution may facilitate other programs by highlighting the challenges in developing the program and the lessons learned.


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.

PURPOSE: Children with advanced cancer are often not referred to palliative or hospice care before they die or are only referred close to the child’s death. The goals of the current project were to learn about pediatric oncology team members’ perspectives on palliative care, to collaborate with team members to modify and tailor three separate interdisciplinary team-based interventions regarding initiating palliative care, and to assess the feasibility of this collaborative approach.

METHODS: We used a modified version of experience-based codesign (EBCD) involving members of the pediatric palliative care team and three interdisciplinary pediatric oncology teams (Bone Marrow Transplant, Neuro-Oncology, and Solid Tumor) to review and tailor materials for three team-based interventions. Eleven pediatric oncology team members participated in four codesign sessions to discuss their experiences with initiating palliative care and to review the proposed intervention including patient case studies, techniques for managing uncertainty and negative emotions, role ambiguity, system-level barriers, and team communication and collaboration.

RESULTS: The codesign process showed that the participants were strong supporters of palliative care, members of different teams had preferences for different materials that would be appropriate for their teams, and that while participants reported frustration with timing of palliative care, they had difficulty suggesting how to change current practices.

CONCLUSIONS: The current project demonstrated the feasibility of collaborating with pediatric oncology clinicians to develop interventions about introducing palliative care. The procedures and results of this project will be posted online so that other institutions can use them as a model for developing similar interventions appropriate for their needs.


BACKGROUND: When young people with life-limiting diagnoses become too old for children’s hospice services, they often experience challenges transitioning into adult services. A two-year pilot project was developed to try to aid transitioning, which involved a day service with occasional overnight trips.

AIM: To evaluate the pilot project.

METHOD: Three focus groups made up of key stakeholders (young people, their parents and staff) were set up and analysed using an adopted thematic analysis framework.

RESULTS: The participants consisted of three young people, seven parents and six staff members. Participants described the transition period as a difficult time for both young people and their families, with a perceived lack of adult services available. All groups agreed that the pilot had a positive impact on young people and their families and all were keen for the project to continue.

CONCLUSION: This pilot models a service that could be adopted by other organisations in order to aid the transition between child and adult hospice services, with further potential for application in mental health and special needs services.


OBJECTIVES: This article focuses on compassionate discharge from an ICU setting for pediatric patients. DATA SOURCES: Not Applicable.

STUDY SELECTION: Not Applicable.

DATA EXTRACTION: Not Applicable.
DATA SYNTHESIS: The rationale for compassionate discharge is described, along with suggestions for assessing feasibility. A patient case highlights the potential benefits of and provides specific examples of steps involved in the process. A general framework for consideration of compassionate discharge, along with a checklist, is provided to highlight the importance of detailed planning and communication.

CONCLUSIONS: Although many children die in an ICU setting, some families desire end-of-life care in a nonhospital setting, often at home. For children dependent on technology, there are considerable logistical challenges to overcome, and it may not always be possible. However, with meticulous planning and close collaboration between intensive care staff, palliative care staff, and other community services, compassionate discharge can be done successfully and provide the child and family the opportunity for end-of-life care in the place most meaningful to them.


BACKGROUND: In the U.S., more children die from cancer than from any other disease, and more than one third die in the hospital setting. These data have been replicated even in subpopulations of children with cancer enrolled on a palliative care service. Children with cancer who die in high-acuity inpatient settings often experience suffering at the end of life, with increased psychosocial morbidities seen in their bereaved parents. Strategies to preemptively identify children with cancer who are more likely to die in high-acuity inpatient settings have not been explored.

MATERIALS AND METHODS: A standardized tool was used to gather demographic, disease, treatment, and end-of-life variables for 321 pediatric palliative oncology (PPO) patients treated at an academic pediatric cancer center who died between 2011 and 2015. Multinomial logistic regression was used to predict patient subgroups at increased risk for pediatric intensive care unit (PICU) death.

RESULTS: Higher odds of dying in the PICU were found in patients with Hispanic ethnicity (odds ratio [OR], 4.02; p = .002), hematologic malignancy (OR, 7.42; p < .0001), history of hematopoietic stem cell transplant (OR, 4.52; p < .0001), total number of PICU hospitalizations (OR, 1.98; p < .0001), receipt of cancer-directed therapy during the last month of life (OR, 2.96; p = .002), and palliative care involvement occurring less than 30 days before death (OR, 4.7; p < .0001). Conversely, lower odds of dying in the PICU were found in patients with hospice involvement (OR, 0.02; p < .0001) and documentation of advance directives at the time of death (OR, 0.37; p = .033).

CONCLUSION: Certain variables may predict PICU death for PPO patients, including delayed palliative care involvement. Preemptive identification of patients at risk for PICU death affords opportunities to study the effects of earlier palliative care integration and increased discussions around preferred location of death on end-of-life outcomes for children with cancer and their families.

IMPLICATIONS FOR PRACTICE: Children with cancer who die in high-acuity inpatient settings often experience a high burden of intensive therapy at the end of life. Strategies to identify patients at higher risk of dying in the pediatric intensive care unit (PICU) have not been explored previously. This study finds that certain variables may predict PICU death for pediatric palliative oncology patients, including delayed palliative care involvement. Preemptive identification of patients at risk for PICU death affords opportunities to study the effects of earlier palliative care integration and increased discussions around preferred location of death on end-of-life outcomes for children with cancer and their families.


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.


Purpose: The purpose of this study was to generate baseline data on the health characteristics, health care utilization, and health care spending among privately insured adolescents and young adults (AYA), who were enrolled in hospice care during their last year of life.

Methods: A retrospective, nonexperimental design was used to collect and analyze longitudinal claims data from the Truven Health MarketScan database. The sample included AYA (aged 15-24 years) who utilized hospice during their last year of life.

Results: Totally, 17,408 AYA were included in this analysis. Mean hospice length of stay (LOS) was low overall, but there was a statistically significant difference in hospice LOS in ages 15-19 years (mean 3.56, SD 15.17 days) compared with those aged 20-24 years (mean 2.26, SD 8.24; P<0.001 days). More than a third (37%) of the AYAs used the emergency department during the last year of life and 83% sought care from a primary care visit. However, only 6% of the sample who were hospice enrollees used frequent inpatient hospital services.

Conclusions: This study provides preliminary data for private insurance expenditures and clinical utilization for AYA who were enrolled in hospice. This analysis also provides initial evidence to suggest extremely short hospice LOS for AYAs prior to the end of life and represents an area of future research need.

BACKGROUND: Improvements in care and treatment have led to more young adults with life-limiting conditions living beyond childhood, necessitating a transition from children’s to adult services. Given the lack of evidence on interventions to promote transition, it is important that those creating and evaluating interventions develop a theoretical understanding of how such complex interventions may work.

OBJECTIVES: To develop theory about the interventions, and organisational and human factors that help or hinder a successful transition from children’s to adult services, drawing on the experience, knowledge, and insights of young adults with life-limiting conditions, their parents/carers, and service providers.

DESIGN: A realist evaluation using mixed methods with four phases of data collection in the island of Ireland. Phase one: a questionnaire survey of statutory and non-statutory organisations providing health, social and educational services to young adults making the transition from children’s to adult services in Northern Ireland and one Health Services Executive area in the Republic of Ireland. Phase two: interviews with eight young adults. Phase three: two focus groups with a total of ten parents/carers. Phase four: interviews with 17 service providers. Data were analysed seeking to explain the impact of services and interventions, and to identify organisational and human factors thought to influence the quality, safety and continuity of care.

RESULTS: Eight interventions were identified as facilitating transition from children’s to adult services. The inter-relationships between these interventions supported two complementary models for successful transition. One focused on fostering a sense of confidence among adult service providers to manage the complex care of the young adult, and empowering providers to make the necessary preparations in terms of facilities and staff training. The other focused on the young adults, with service providers collaborating to develop an autonomous young adult, whilst actively involving parents/carers. These models interact in that a knowledgeable, confident young adult who is growing in decision-making abilities is best placed to take advantage of services - but only if those services are properly resourced and run by staff with appropriate skills. No single intervention or stakeholder group can guarantee a successful transition. Rather, service providers could work with young adults and their parents/carers to consider desired outcomes, and the range of interventions, in light of the organisational and human resources available in their context. This would allow them to supplement the organisational context where necessary and select interventions that are more likely to deliver outcomes in that context.


AIM: The aim of this study was to seek views of UK children’s and adult hospices on the availability and challenges of providing services for young adults with life-limiting conditions. BACKGROUND: Internationally, there are a growing number of young adults with life-limiting conditions and/or complex needs which are degenerative, progressive and diverse and involve complex life-long symptom, medication management as well as palliative care. There are 55,721 young adults, aged 18-40 in England, which continues to increase. The hospice sector is experiencing demands to extend services for this population despite concerns about the appropriateness of adult hospices and their nursing staff to provide care for the complex and unfamiliar conditions of this patient group. Evidence is needed of hospices’ views and the main challenges faced providing services for young adults. DESIGN: Descriptive cross-sectional survey. METHODS: xChildren and adult hospices completed an online survey exploring service provision and their views of respite care for young adults with life-limiting conditions from 18 years old and onward. Data were collected between October 2015 - February 2016. FINDINGS: Respondents (N = 76 hospices) reported that children’s hospices predominantly provided short breaks and end-of-life care; adult hospices provided mainly symptom management, end-of-life care and day services. Main challenges were lack of existing adult respite services; lack of funding and capacity; lack of a skilled workforce in adult hospices; and the need for better integrated service provision. CONCLUSION: Examples of good collaborative working were reported. With an increasing population of young adults and pressure on families, it is vital that services work together to find sustainable solutions to the challenges.

End-of-life care for many infants involves the withdrawal of mechanical ventilation. Usually this takes place in the hospital environment, but sometimes parents request that their infant dies at home. Facilitating this has significant practical and resource implications and raises both logistical and ethical questions. In this article, we report a neonatal case involving home extubation, explaining the processes involved as well as providing an ethical context.


PURPOSE: Although the UK has pioneered the development of specialist adolescent cancer units, the majority of teenagers and young adults (TYAs) continue to be treated at their local hospital or at a cancer centre alongside adults of all ages. This study aimed to elicit young people’s views on this experience of having cancer treatment in an adult setting. METHODS: Seventeen participants who had been treated for cancer in an adult hospital between the ages of 15 and 24 were recruited via cancer charities and social media. Telephone interviews were conducted with the participants and the resulting data were analysed using thematic analysis. RESULTS: Already feeling out of sync as a TYA with cancer, participants felt out of place in the adult setting. Four factors contributed to this negative experience: a lack of affinity with older patients; the challenging issues in the adult setting; the absence of empathy towards TYAs by staff; and the unsuitability of the environment for adolescents. CONCLUSION: Staff working with TYAs with cancer in the adult setting should be aware of the potentially detrimental impact of this environment on this cohort of patients, and consider ways of adapting and modifying their approach.


The purpose of this review is to explore the literature on the experience of adolescents and young adults (AYAs) having cancer treatment in an adult setting, rather than on a specialist adolescent cancer unit. The integrative review method was used to explore the current literature. Primary research on the topic was located systematically and then synthesized into a thematic narrative. The experience of AYAs undergoing treatment in an adult setting was generally negative. This can be attributed to three themes: feeling isolated in the adult setting; the lack of empathy from staff working in the adult setting; and the inappropriateness of the adult environment for this age group. As many AYAs with cancer will continue to have treatment in adult settings, staff working in this environment should be aware of the negative experience of this cohort and the impact this can have on a vulnerable group of patients. Staff could consider simple ways of improving the AYA experience, such as connecting AYA patients with their peers to reduce isolation; adapting their approach to take account of the unique emotional needs of this age group; and considering ways of making the environment more welcoming and age-appropriate.


Numerous policy directives highlight the need for planned and well-coordinated support to enable young people with long-term conditions and disabilities to negotiate the transition to adulthood, including making the leap from children-oriented to adult-centered health services. The journey is complex and multi-dimensional. For young people with a disability, long-term condition, or mental health problem there are additional challenges when transitioning between services with differences in expectations, delivery, and culture. This article explores findings from 6 case studies of young people who have recently experienced transition to adult health and care services, triangulating inter-related perspectives: those of young people, parents, and carers, and where possible the professionals involved. One of the case studies illustrates how the challenges are actually experienced. Analysis of emerging themes across the case studies leads to key messages from families to inform strategic development of services and practice.
improving the confidence of local care providers to deliver ongoing care. This paper looks at some of the factors related to care transfer for pediatric palliative patients from one care facility to another, home and the impact of this on the family and medical care.


OBJECTIVES: To describe important considerations during the process of caring for critically ill children who may be potential organ donors and supporting the family during the death of their child. DESIGN: Literature review and expert commentary.

MEASUREMENT AND MAIN RESULTS: Medical literature focusing on pediatric donation, best pediatric donation practices, donor management, and factors influencing donation were reviewed. Additional pediatric data were obtained and reviewed from the U.S. Organ Procurement and Transplantation Network. Achieving successful organ donation requires the coordinated efforts of the critical care team, organ donation organization, and transplant team to effectively manage a potential donor and recover suitable organs for transplantation. Collaboration between these teams is essential to ensure that all potential organs are recovered in optimal condition, to reduce death and morbidity in children on transplantation waiting lists as well as fulfilling the family’s wishes for their dying child to become a donor.

CONCLUSIONS: Organ donation is an important component of end-of-life care and can help the healing process for families and medical staff following the death of a child. The process of pediatric organ donation requires healthcare providers to actively work to preserve the option of donation before the death of the child and ensure donation occurs after consent/authorization has been obtained from the family. Medical management of the pediatric organ donor requires the expertise of a multidisciplinary medical team skilled in the unique needs of caring for children after neurologic determination of death and those who become donors following circulatory death after withdrawal of life-sustaining medical therapies.


AIMS: The aim of this study was to report a secondary qualitative analysis exploring the cultural and practical differences that young people and parents experience when transitioning from children’s to adult services.

BACKGROUND: Despite two decades of research and quality improvement initiatives, young people with life-limiting and life-threatening conditions still find transition unsatisfactory.

DESIGN: Secondary analysis: 77 qualitative interviews with children and young people (20), parents (35), siblings (1), professionals (21). METHODS: Qualitative framework analysis completed 2017.

FINDINGS: Six conflicting realities were identified: Planning to live and planning to die with different illness trajectories that misaligned with adult service models; being treated as an adult and the oldest "patient" in children's services compared with being treated as a child and the youngest "patient" in adult services; being a "child" in a child’s body in children’s services compared with being a "child" in an adult's body in adult services for those with learning impairments; being treated by experienced children's professionals within specialist children's services compared with being treated by relatively inexperienced professionals within generalist adult services; being relatively one of many with the condition in children’s services to being one of very few with the condition in adult services; meeting the same eligibility criteria in children’s services but not adult services.

CONCLUSION: Inequity and skills deficits can be addressed through targeted interventions. Expanding age-specific transition services, use of peer-to-peer social media, and greater joint facilitation of social support groups between health services and not-for-profit organizations may help mitigate age dilution and social isolation in adult services.


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.


BACKGROUND: Idiopathic pulmonary fibrosis has an uncertain and rapid trajectory after diagnosis. Palliative care is rarely utilized, although both patients and caregivers experience a distressingly high symptom burden. Most patients die in hospital. AIM: The purpose of this study was to explore bereaved caregivers’ experiences and perceptions of an early integrated palliative approach implemented at a Multidisciplinary Interstitial Lung Disease Clinic.

DESIGN: A narrative approach was used, with thematic and content analysis of open-ended interviews.

SETTING/PARTICIPANTS: The clinic is located in a large western Canadian city. Caregivers of deceased patients were recruited through purposive sampling. The eight participants were either spouses or adult children.

RESULTS: Five major themes were identified: Having a Terminal Disease; Planning Goals and Wishes for Care; Living Life and Creating Memories; Feeling Strain and Responsibility; and Nearing the End. Caregivers had little understanding of prognosis prior to advance care planning conversations at the clinic. Advance care planning conversations enabled caregivers to know and support patients’ goals and wishes. Caregivers described feeling informed, prepared, and supported when death was near. They expressed neither distress nor anxiety related to patients’ symptoms or strain of relationships.

CONCLUSION: Collaboration and close communication among caregivers, respirologists, and home care enabled effective symptom management and out of hospital deaths. Patients and caregivers had opportunities to enjoy events, create memories, determine preferences, and make plans. Further research on an early integrated palliative approach in Idiopathic Pulmonary Fibrosis is warranted related to quality of life, experience with death and dying, and caregiver bereavement.


BACKGROUND: Only limited data are available concerning the diseases managed before death and hospital palliative care (HPC) use according to place of death in France. We therefore conducted an observational study based on administrative health data in a large population to identify the diseases treated one year before death in 2013, the place of stay with or without hospital palliative care, and the place of death.

METHODS: French health insurance general scheme beneficiaries were identified in the National Health data Information System (Snds) with a selection of information. Diseases were identified by algorithms from reimbursement data recorded in the Snds database.

RESULTS: 347,253 people were included in this study (61% of all people who died in France). Place of death was short stay hospital for 51%, Rehab (7%), hospital at home (3%), skilled nursing home (13%) and other (26%). Chronic diseases managed in 2013 before death were cardiovascular/neurovascular diseases (56%), cancers (42%), and neurological and degenerative diseases (25%). During the year before death, 84% of people were hospitalized at least once, and 29% had received HPC. HPC was used by 52% of cancer patients (lung cancer: 62%; prostate cancer: 41%). In the absence of cancer, the use of HPC varied according to the disease: acute stroke: 24%, heart failure: 17%, dementia: 17%, multiple sclerosis: 23%.

CONCLUSIONS: Health administrative data can refine the knowledge of the care pathway prior to death and the HPC utilisation and can be useful to evaluate health policies and improve monitoring and assessment of HPC use.

BACKGROUND: Parents of children admitted to neonatal and pediatric intensive care units (ICUs) are at increased risk of experiencing acute and post-traumatic stress disorder. The integration of palliative care may improve child and family outcomes, yet there remains a lack of information about indicators for specialty-level palliative care involvement in this setting.

OBJECTIVE: To describe neonatal and pediatric critical care physician perspectives on indicators for when and why to involve palliative care consultants.

METHODS: Semistructured interviews were conducted with 22 attending physicians from neonatal, pediatric, and cardiothoracic ICUs in a single quaternary care pediatric hospital. Transcribed interviews were analyzed using content and thematic analyses.

RESULTS: We identified 2 themes related to the indicators for involving palliative care consultants: (1) palliative care expertise including support and bridging communication and (2) organizational factors influencing communication including competing priorities and fragmentation of care.

CONCLUSIONS: Palliative care was most beneficial for families at risk of experiencing communication problems that resulted from organizational factors, including those with long lengths of stay and medical complexity. The ability of palliative care consultants to bridge communication was limited by some of these same organizational factors. Physicians valued the involvement of palliative care consultants when they improved efficiency and promoted harmony. Given the increasing number of children with complex chronic conditions, it is important to support the capacity of ICU clinical teams to provide primary palliative care. We suggest comprehensive system changes and critical care physician training to include topics related to chronic illness and disability.


BACKGROUND: Literature in adult palliative care (PC) boasts fewer invasive procedures, shorter lengths of stay, and decreased cost of care. Benefits of pediatric PC are under-researched and are important to identify to optimize care.

OBJECTIVE: Our aim was to estimate the influence and utilization of PC on pediatric patient care.

DESIGN: We evaluated the electronic medical record of 43 patients at Cook Children’s Medical Center (CCMC) with complex chronic conditions, who died between January 1, 2013, and December 31, 2014, comparing the length and frequency of hospitalizations, number of medications administered and procedures performed, and established limits of resuscitation between patients who received PC and those who did not.

MEASUREMENTS: Data analyses were performed using SAS Enterprise (version 6.1; SAS Institute, Inc., Cary, NC). Continuous variables were described as medians and ranges and analyzed with Wilcoxon rank-sum test for ordinal data. Categorical variables were described as percentages and analyzed with chi-square test of independence. Repeated-measures analyses were performed utilizing multilevel linear modeling, which examined the data at the level of the 236 visits rather than the 43 patients.

RESULTS: Twelve (28%) eligible patients were seen by PC. PC patients had more hospitalizations, longer lengths of stay, and fewer medications and procedures than those patients without PC services. PC patients were also more likely to have a medical orders for scope of treatment in place.

CONCLUSION: These data demonstrate that PC services at CCMC are underutilized and support the need for PC services by decreased medications and procedures and identified family wishes for medical treatment.


Palliation in pediatric otorhinolaryngology is a rarely discussed but important aspect of care. This review encapsulates current thinking on pediatric palliative care (PC) and demonstrates, through one case, the impact of integrating PC into clinical care. We encourage early consideration of pediatric palliative care approaches for children with complex otorhinolaryngologic disorders.


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 continuation of needs of young people with cerebral palsy argue for close liaison between adult services and child services and creation of models of practice to improve coordination.


Pediatric palliative care is a comprehensive treatment approach (physical, psychological, social, spiritual) for children living with life-threatening conditions. These patients and siblings, as well as children of ill parents, face extraordinary psychological challenges. Structured art techniques incorporated into psychotherapy can be powerful for children dealing with life-and-death realities. This article provides the rationale, instructions, and examples for 3 techniques that the author has adapted for children facing illness and bereavement. Although these art techniques are simple to administer, they frequently evoke complex and powerful responses and thus are intended for use by or in consultation with mental health professionals.


Pediatric cancer has experienced significant improvement in overall survival rates over the past several decades. Despite this progress, however, it remains the leading cause of death from disease beyond infancy in children. Among the children and adolescents that survive their cancer diagnosis, significant symptom burden and toxicities of therapy are often experienced. The evidence presented affords great insight to the current empirical support for pediatric palliative care involvement, current utilization of palliative care services in the care of children with cancer and their families, and barriers that have been identified to date. Positive trends toward increased, appropriate integration of palliative care services in the care of children with cancer and their families have been observed. Continued research, advocacy, and education are necessary to optimize the care of this vulnerable population of patients and their families.

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: 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.


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

Vadeboncoeur, C. and M. McHardy (2018). "Benefits of Early Referral to Pediatric Palliative Care for a Child With a Rare Disease." Pediatrics 141(6).


BACKGROUND: Little is known about the composition, availability, integration, communication, perceived barriers, and work load of pediatric palliative care (PPC) providers serving children and adolescents with cancer.

 OBJECTIVE: To summarize the structure and services of programs to better understand successes and gaps in implementing palliative care as a standard of care.

 METHODS: Cross-sectional online survey about the palliative care domains determined by the Psychosocial Care of Children with Cancer and Their Families Workgroup.

 SUBJECTS: A total of 142 surveys were completed with representation from 18 countries and 39 states.

 RESULTS: Three-fourths of sites reported having a PPC program available for the pediatric cancer population at their center. Over one-fourth (28%) have been in existence less than five years. Fewer than half of sites (44%) offered 24/7 access to palliative care consultations. Neither hospital-based nor local community hospice services were available for pediatric patients at 24% of responding sites. A specific inpatient PPC unit was available at 8% of sites. Criteria for automatic palliative referrals ("trigger" diagnoses) were reported by 44% respondents. The presence of such "triggers" increased the likelihood of palliative principle introduction 3.41 times (p < 0.003). Six percent of respondents perceived pediatric oncology patients and their families "always" were introduced to palliative care concepts and 17% reported children and families "always" received communication about palliative principles. The most prevalent barriers to palliative care were at the provider level.

 DISCUSSION: Children and adolescents with cancer do not yet receive concurrent palliative care as a universal standard.


The national nursing shortage translates into a gap in home nursing care available to children with complex, chronic medical conditions and their family caregivers receiving palliative care consultations. A total of 38 home health nursing surveys were completed by families receiving pediatric palliative care consultation services at a freestanding children’s hospital in the Midwest. The gap in the average number of nursing hours allotted versus received was 40 h/wk per family, primarily during evening hours. Parents missed an average of 23 hours of employment per week to provide hands-on nursing care at home, ranking stress regarding personal employment due to nursing shortage at 6.2/10. Families invested an average of 10 h/mo searching for additional nursing coverage and often resorted to utilizing more than 6 different home nurse coverage personnel per month. Families reported multiple delays to hospital discharges (mean, 15 days per delay) due to inability to find home nursing coverage. Respiratory technology and lack of Medicaid coverage (P < .02) correlated with the gap in home nursing access. This study examines how the pediatric home nursing shortage translates into a lived experience for families with children with complex medical conditions receiving palliative care.


Purpose: The impact of specialized pediatric palliative care (SPPC) teams on patterns of end-of-life care is unknown. We sought to determine (1) which children with cancer access SPPC and (2) the impact of accessing SPPC on the risk of experiencing high-intensity end-of-life care (intensive care unit admission, mechanical ventilation, or in-hospital death). Methods: Using a provincial childhood cancer registry, we assembled a retrospective cohort of Ontario children with cancer who died between 2000 and 2012 and received care through pediatric institutions with an SPPC team. Patients were linked to population-based administrative data capturing inpatient, outpatient, and emergency visits. Children were classified as having SPPC, general palliative care, or no palliative care on the basis of SPPC clinical databases, physician billing codes, or inpatient diagnosis codes. Results: Of the 572 children, 166 (29%) received care from an SPPC team for at least 30 days before death, and 100 (17.5%) received general palliative care. SPPC involvement was significantly less likely for children with hematologic cancers (OR, 0.3; 95% CI, 0.3 to 0.4), living in the lowest income areas (OR, 0.4; 95% CI, 0.2 to 0.8), and living further from the treatment center (OR, 0.5; 95% CI, 0.4 to 0.5). SPPC was associated with a five-fold decrease in odds of intensive care unit admission (OR, 0.2; 95% CI, 0.1 to 0.4), whereas general palliative care had no impact. Similar associations were seen with all secondary indicators. Conclusion: When available, SPPC, but not general palliative care, is associated with lower intensity care at the end of life for children with cancer. However, access remains uneven. These results provide the strongest evidence to date supporting the creation of SPPC teams.


OBJECTIVE: To determine whether changes in emergency admission rates during transition from paediatric to adult hospital services differed in children and young people (CYP) with and without underlying long-term conditions (LTCs).

DESIGN: Cross-sectional study.


PARTICIPANTS: 763 199 CYP aged 10-24 years with and without underlying LTCs (LTCs were defined using the International Classification of Diseases, 10th Revision codes recorded in the past 5 years).

PRIMARY AND SECONDARY OUTCOME MEASURES: We calculated emergency admission rates before (10-15 years) and after transition (19-24 years), stratified by gender, LTC and primary diagnosis. We used negative binomial regression to estimate adjusted incidence rate ratios (IRRsr).

RESULTS: We included 1 109 978 emergency admissions, of which 63.2% were in children with LTCs. The emergency admission rate increased across the age of transition for all CYP, more so for those with LTCs (IRLTC: 1.55, 99% CI 1.47 to 1.63), compared with those without (IRPnolTC: 1.21, 99% CI 1.18 to 1.23). The rates increased most rapidly for CYP with mental health problems, MEDReG (metabolic, endocrine, digestive, renal, genitourinary) disorders, and multiple LTCs (both genders) and respiratory disorders (female only). Small or no increased rates were found for CYP without LTCs and for those with cancer or cardiovascular disease. Increases in length of stay were driven by long admissions (10+ days) for a minority (1%) of CYP with mental health problems and potentially psychosomatic symptoms. Non-specific symptoms related to abdominal pain (girls only), gastrointestinal and respiratory problems were the most frequent primary diagnoses.

CONCLUSIONS: The increased rates and duration of emergency admissions and predominance of non-specific admission diagnoses during transition in CYP with underlying LTCs may reflect unmet physical or mental health needs.

CONTEXT: According to the International Observatory on End of Life Care, the level of pediatric palliative care in Japan is Level 2 (capacity building) and the current status of palliative care for children in Japan has not been clarified.

OBJECTIVES: The objective of the study was to clarify the availability and utilization of specialist palliative care services among children with life-threatening conditions in Japan.

METHODS: A questionnaire was administered to assess the availability of specialist palliative care services among children with life-threatening conditions. All 427 certified regional cancer centers having hospital-based adult palliative care teams, 15 certified children’s cancer centers having pediatric palliative care teams, and 368 medical institutions having a certified palliative care unit were surveyed.

RESULTS: Fifteen to twenty-one percent of adult palliative care teams and more than 90% of pediatric palliative care teams had experience providing palliative care to children with cancer. By contrast, only 2%-3% of adult palliative care teams and 15% of pediatric palliative care teams had experience providing care for the noncancer population. An estimated 12% of children with cancer in Japan used hospital-based palliative care teams in 2015. Eight children used a palliative care unit in 2015, and of those, seven (88%) had a solid tumor. An estimated 1.3% of children with cancer who died in Japan used a palliative care unit.

CONCLUSION: An estimated 12% of children with cancer in Japan used hospital-based palliative care teams and an estimated 1.3% of children with cancer who died in Japan used a palliative care unit in 2015.
