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Synopsis, International digest of children’s palliative care research abstracts

Together for Short Lives is a UK wide charity that, together with our members, speaks out for all children and young people who are expected to have short lives. Together with everyone who provides care and support to these children and families we are here to help them have as fulfilling lives as possible and the very best care at the end of life. We can't change the diagnosis, but we can help children and families make the most of their time together.

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

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

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

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
## Contents

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Synopsis Editors’ picks</td>
<td>1</td>
</tr>
<tr>
<td>Ethical and Clinical Decision Making</td>
<td>6</td>
</tr>
<tr>
<td>Education, Professional and Research</td>
<td>19</td>
</tr>
<tr>
<td>Epidemiology and Pathology</td>
<td>47</td>
</tr>
<tr>
<td>Outcomes and Instruments</td>
<td>58</td>
</tr>
<tr>
<td>Pain and Symptom Control</td>
<td>86</td>
</tr>
<tr>
<td>Psychosocial and Family Issues</td>
<td>108</td>
</tr>
<tr>
<td>Services for children and families</td>
<td>134</td>
</tr>
</tbody>
</table>
Synopsis Editor’s Picks

We set out here 30 of the favourite articles from this edition of Synopsis as selected by our editorial team.


Editor’s comment: This is a practical article in response to the NICE guidelines on end of life care for children. It’s important as it helps to support putting this guidance into practice.


Editor’s comment: This article uses meta-ethnography (a systematic approach to synthesising qualitative research) to explore the understanding of children, young people with brain tumours, their parents and health professionals of the term ‘Quality of Life’. It proposes a conceptual model with ‘Normalcy’ (a new normal) as a key concept rather than the more common health-related quality of life.


Editor’s comment: This study analyses 5 commonly used activities in research with children to draw out their perspectives on their care and treatment, including the "Draw and Write" technique, a sticker activity, a paper-person exercise, informal interviews, and participant observation. Each of these methods was examined with regard to ease of use, data generation, and utility.


Editor’s comment: In this research, bereaved parents were asked to reflect on their experiences of research participation, with a focus on recruitment methods, timing of research contact, and the location of their interview. Despite being emotionally difficult, parents’ overall experiences of research participation were positive, with an overall preference for an opt-in approach with initial contact by letter within 12-24 months of their child’s death.


Editor’s comment: Interesting article which provides useful insight for care professionals on how social media is used, especially by children and young people, during their illness, after death, and in the bereavement process.


Section: Education, Professional Development and Research
Editor’s comment: This article explores the use of a simulation model in undergraduate paediatric nurse education to provide an experience of caring for a dying child and family including developing self-awareness of their own emotions and thoughts when faced with a pediatric death and how these might impact on a family.


Section: Outcomes and Instruments
Editor’s comment: This study shines a light on the outcome of advance care planning with children, adolescents, and young adults with complex chronic conditions. It evaluates whether advance care planning and assessment of specific family considerations were associated with differences in parent-reported end-of-life outcomes.


Section: Outcomes & instruments
Editor’s comment: This paper reviews relevant outcomes and quality measures in PPC, the current state of science on outcome measurement for children and young people with life-limiting and life-threatening conditions and the development of the African Children’s Palliative Outcome Scale which is a key milestone in the journey towards developing robust outcome measures for PPC.


Section: Psychosocial and Family Issues
Editor’s comment: This article seeks to understand the lived experience of parents who have lost their child to a chronic life-limiting condition. Thematic synthesis was used to categorise findings into 13 themes that were further organized into a four-phase trajectory of parental bereavement, with practical recommendations for enhancing parental bereavement support services.


Section: Psychosocial and Family Issues
Editor’s comment: This study synthesises published qualitative research to explore healthcare users’ experiences of communicating with healthcare professionals about children with life-limiting conditions. Findings were that 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.


Section: Services for Children and Families
Editor’s comment: This study reviews charts of children with life-limiting conditions who have attended A&E and as might be expected found that often paediatric palliative care patients often present to A&E acutely ill, often at their end of life, and that goals of care are not always discussed.

2

Section: Education, Professional Development and Research

Editor’s comment: The aim of this article is to understand the contribution of previous qualitative research on paediatric palliative care that includes the voices of children. This is an important issue and good to see a systematic review on it.


Section: Education, Professional Development and Research

Editor’s comment: Important and topical. This paper gives practical guidance about the importance of good advance care planning and includes the insight of two bereaved parents. My top pick!


Section: Pain and Symptoms

Editor’s comment: It’s important to know what is preventing the use of these preparations to inform better symptom control.


Section: Pain and Symptoms

Editor’s comment: The aim of this study was to describe how children with cancer, aged 4-7 years, express their symptoms through drawings and concludes that it can help initiate communication regarding how they feel, and develop rapport between the interviewer and children.


Section: Education, Professional Development and Research

Editor’s comment: This review seeks to clarify the role and scope of speech-language pathologists in children’s palliative care. Roles identified included management of communication, feeding, upper-airway and oral health as well as a role within a multidisciplinary PPC team. This is an issue we come across a lot, so good to see an article devoted to this.


Section: Education, Professional Development and Research

Editor’s comment: This study assessed 153 phone calls from next of kin notifying the treating medical team that their relative had died. They notified either directly (using specific words such as ‘died’) or indirectly (using words like ‘it’s over’). 30% of callers expressed doubt or uncertainty and 20% expressed emotions. This is an important subject that I’ve not seen much written about.


Section: Outcomes and Instruments
Editor’s comment: This systematic review prioritises issues that matter to children and their families to inform the development of a health outcomes framework. As well as identifying 5 priority themes, it also highlights the gaps in research - of the 81 studies included, most (n = 68) were from high-income countries and focussed on young people with cancer (n = 58). Young people did not contribute in 30% of studies.


Section: Services for Children and Families
Editor’s comment: An interesting study which explores the cultural and practical differences experienced by young people and parents when transitioning from children's to adult services. Six conflicting realities are identified.


Section: Pain and Symptoms
Editor’s comment: This study used a retrospective chart review to analyse 70 cases of acute attack of respiratory disease in children with life-limiting conditions in order to describe the circumstances of the use of intranasal fentanyl and to describe outcomes and adverse events after its use. Intranasal fentanyl may be a safe and effective medication for the treatment of acute attacks of respiratory distress in children with life-limiting conditions.


Section: Epidemiology and Pathology
Editor’s comment: This study looks at 2008-15 Child Death Overview Panel data to describe trends in place of death for children in South Yorkshire (748 deaths). Excluding neonatal deaths (which accounted for 46% of the total), 58% were ‘expected’ and of these 19% died in home, 19% died in hospice and 61% died in hospital. This was significantly different from comparable national data which showed considerably more hospital deaths. There was no significant change in POD over time.

https://www.ncbi.nlm.nih.gov/pubmed/30446488


Section: Psychosocial and Family issues
Editor’s comment: A nice way of exploring the reflections of young people with cancer about their experiences and preferences in relation to their involvement in decision-making about their care.


Section: Outcomes and Instruments
Editor’s comment: Dystonia is a difficult area of symptom management on which children’s palliative care doctors are often asked to advise. Practice in this area is changing and collaboration with paediatric neurologists is really important.


Section: Psychosocial and Family Issues
Editor’s comment: The purpose of this Delphi study was to gather opinions from parents and care providers about the processes essential to parental hope to increase understanding of support needs. Eight major themes were identified.

Section: Ethical and Clinical Decision Making
Editor’s comment: An interesting study to identify religious and spiritual factors affecting parental decision-making. The study concludes that parents consider religion & spirituality as fundamental to decision-making, but apply the concepts in vague ways, which suggests that it impacts more on how decisions are made than what decisions are made. The authors present three models of how this vagueness functions in parental decision-making and suggest clinical applications.

Section: Outcome and Instruments
Editor’s comment: This study used the computer-based (PediQUEST) system to administer a modified version of the Memorial Symptom Assessment Scale (PQ-MSAS) to look at the factors associated with fatigue and associated distress in children with advanced cancer. There was a close correlation of fatigue with particular symptoms (anorexia, nausea, sleep disturbance, sadness, and irritability), whilst fatigue distress was associated with distress from nausea, cough, and pain.

Section: Outcomes and Instruments
Editor’s comment: I selected this article as it’s focussed on end of life care for children with progressive and non-progressive severe physical disability. It concludes that there is an opportunity for greater consistency in offering advanced care planning and palliative care, especially to those with NPSPD. These children make up a big proportion of the children’s hospice population.
https://www.ncbi.nlm.nih.gov/pubmed/29943874

Section: Psychosocial and Family Issues
Editor’s comment: An interesting narrative on how an academic philosophical perspective helped a bereaved parent cope with their experience in NICU.

Section: Education, Professional Development and Research
Editor’s comment: This study evaluates the use of a new curriculum for paediatric residents on end of life care – an area we need to develop further in the UK.

Section: Outcomes and Instruments
Editor’s comment: This article argues for moving beyond the biological outcomes that can be achieved for babies through modern technology towards a "Slow Medicine" which adopts a more rounded view of the broader suffering of patients and their families outside the NICU.
Ethical and Clinical Decision Making


 Debates on morally acceptable and lawful end-of-life (EOL) practices in pediatrics were reignited by the recent amendment in Belgian law to allow euthanasia for minors of any age who meet the criteria for capacity. Euthanasia and its legalization in pediatrics are often opposed based on the availability of aggressive palliative sedation. For terminally ill patients, this type of sedation is often identified as continuous and deep sedation until death (CDS). We demonstrate that this reasoning is based on flawed assumptions: (1) CDS is a morally preferable alternative to euthanasia; (2) CDS can meet the same patient needs as euthanasia; (3) children lack the capacity and experience to make EOL decisions; (4) unlike euthanasia, CDS does not raise capacity issues. Our aim is not to reject CDS as a valid option at the EOL, nor to offer a clear-cut defense of euthanasia for minors, but to emphasize the ethical issues with both practices.


 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.


OBJECTIVE:: This study aims to assess the experiences and wishes of parents of children with severe spinal muscular atrophy regarding information and decision-making throughout the course of the illness. STUDY DESIGN:: A full population survey, conducted in 2015, among parents of children with severe spinal muscular atrophy who were born in Denmark between January 1, 2003, and December 31, 2013. We used a study-specific questionnaire with items about experiences and wishes concerning the provision of information about diagnosis, treatment, and end-of-life care. RESULTS:: Among the 47 parents that were identified, 34 parents of 21 children participated. Eleven of them were nonbereaved and 23 were bereaved parents. All parents stated that health care staff did not take any decisions without informing them. A proportion of parents indicated that they were not informed about what spinal muscular atrophy entails (32%), possible treatment options (18%), or the fact that their child would have a short life (26%) or that death was imminent (57%). Most of the bereaved parents who had wishes concerning how and where their child would pass away had their wishes fulfilled. CONCLUSIONS:: The study showed that health care staff did not take treatment decisions without parents being informed. However, there is room for improvement concerning information about what spinal muscular atrophy entails, treatment options, and prognosis. Possibilities of palliative care and advance care planning should be investigated for these parents, their child, and health care staff.


AIMS: To study parent’s levels of uncertainty related to the transfer from pediatric to adult care in adolescents with congenital heart disease (CHD) and to identify potentially correlating factors. BACKGROUND: Parents acknowledge that during transition they struggle with finding ways of feeling secure in handing over the responsibility and letting go of control. Well-prepared and informed parents who feel secure are most likely better skilled to support their adolescent and to hand over the responsibility. DESIGN: A cross-sectional study. METHODS: Overall, 351 parents were included (35% response rate). Parental uncertainty was assessed using a Linear Analogue Scale (0-100). Data were collected between January - August 2016. Potential correlates were assessed using the readiness for transition questionnaire and sociodemographic data. RESULTS: The mean parental uncertainty score was 42.5. Twenty-four percent of the parents had a very low level of uncertainty (score 0-10) and 7% had a very high level (score 91-100). Overall, 26% of the mothers and 36% of the fathers indicated that they had not started thinking of the transfer yet. The level of uncertainty was negatively associated with the level of perceived overall readiness. Adolescents' age, sex, CHD complexity, and parental age were not related to uncertainty. CONCLUSION: A wide range in the levels of uncertainty was found. Parents who were less involved in the care, or perceived their adolescent as readier for the transition, felt less uncertain. Still, thirty percent of the parents had not started to think about the transfer to adult care.
Health care in the United States is increasingly delivered in cross-cultural contexts. Empathy, mutual regard, respect, and compassionate communication are necessary to achieve the highest standard of care for each individual. Moral and ethical perspectives on life and death, health, and health care are not universal but rather have their origins within culture and societal norms. In a cross-cultural context, "the right decision" may be seen differently depending on an individual's cultural background, discipline, and type of education. This pediatric case study is intended to stimulate conversation on the need for culturally sensitive health care decision making and the shortcomings of a "one-size-fits-all" approach to bioethics in our increasingly interconnected world.


BACKGROUND: Despite advancements in treatment and survival, pediatric organ failure and transplant populations continue to face significant risks of morbidity and mortality. Little scientific attention has been given to addressing the end-of-life care needs of this growing population of young people. This study characterized current practices, beliefs, and challenges specific to the disclosure of prognosis and end-of-life care topics among providers caring for pediatric organ failure and transplant populations. METHODS: This cross-sectional study included 144 healthcare providers actively caring for children, adolescents, and young adults with organ failure or solid organ transplant history. Participants completed an electronic survey measuring frequency and comfort in discussing the following topics with patients and parents: prognosis/survival statistics, re-transplantation, advance care planning (ACP), and death/dying. Descriptive statistics, two-sample t tests, and analysis of variance were used. RESULTS: Fewer than half of respondents regularly discuss prognosis/survival statistics and potential need for re-transplantation with their pediatric and young adult patients. Less than 20% of providers engage their pediatric patients in ACP discussions, and approximately 30% facilitate such discussions with young adult patients. Pediatric organ failure and transplant providers endorse a number of barriers specific to discussing these topics. CONCLUSION: Pediatric organ failure and transplant providers do not regularly discuss prognosis or end-of-life care topics with this patient population. Communication-focused intervention research is needed to improve honest and compassionate discussion of these topics that is aligned with both patients' and parents' needs and preferences.

A recent case of conjoined twins required multiple hospitalizations in the pediatric intensive care unit and led to a difficult situation confronting staff regarding the potential separation where surgery would result in the death of one twin. The hospital ethics committee was consulted. A systematic approach was utilized to examine medical standards, historical precedents, and various ethical and legal frameworks. The ethics committee believed that either proceeding with or forgoing attempted separation surgery would be ethically acceptable. We share our reasoning and lessons learned for others facing this situation in the future.


Objective: Adolescent and young adults' (AYAs) involvement in advance care planning and end-of-life discussions may enhance the decision-making process, reduce stress and improve the patient's quality of life. Given the importance of establishing adequate communication and having culturally-appropriate tools to introduce advance care planning, our paper will describe the cross-cultural adaptation of the advance care planning guide, Voicing My CHOICES () in Australia and in Brazil. Methods: In Brazil, the process involved initially translating the document to Portuguese followed by evaluation by a group of providers and patients (aged 18-39) undergoing cancer treatment. The document was revised based on the feedback received, then back-translated to English and discussed with Voicing My CHOICES () authors to refine the final version in Portuguese. In Australia, a multi-perspective interview-based study was undertaken with AYA cancer patients/survivors (aged 15-25), siblings, parents, and a range of healthcare providers from the oncology setting, to determine the perceived acceptability of the tool within the Australian clinical context. Results: These interviews pointed to a variety of recommended adaptations ranging from the aesthetic and linguistic, through to the re-structuring of content within the tool. Adaptations for the Australian setting were then revised in an iterative capacity within several focus groups of AYA participants and healthcare providers. Conclusions: The processes used in both countries highlight ways to engage youth living with a life-limiting illness in conversations about advance care planning and how to develop culturally-appropriate clinical tools.


This article explores the ethical challenges of providing Medical Assistance in Dying (MAID) in a paediatric setting. More specifically, we focus on the theoretical questions that came to light when we were asked to develop a policy for responding to MAID requests at our tertiary paediatric institution. We illuminate a central point of conceptual confusion about the nature of MAID that emerges at the level of practice, and explore the various entailments for clinicians and patients that would flow from different understandings. Finally, we consider the ethical challenges of building policy on what is still an extremely controversial social practice. While MAID is currently available to capable patients in Canada who are 18 years or older—a small but important subsection of the population our hospital serves—we write our policy with an eye to the near future when capable young people may gain access to MAID. We propose that an opportunity exists for MAID-
providing institutions to reduce social stigma surrounding this practice, but not without potentially serious consequences for practitioners and institutions themselves. Thus, this paper is intended as a road map through the still-emerging legal and ethical landscape of paediatric MAID. We offer a view of the roads taken and considered along the way, and our justifications for travelling the paths we chose. By providing a record of our in-progress thinking, we hope to stimulate wider discussion about the issues and questions encountered in this work.


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.


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


Determination and declaration of death by neurologic criteria, brain death, is an established and legally accepted clinical practice with profound implications. Concerns regarding the accuracy of this diagnosis raise important clinical, ethical, and legal issues. A recent magazine article highlights these concerns by describing a poignant example of a patient meeting accepted clinical and ancillary testing criteria for brain death in the setting of post cardiac arrest hypoxic ischemic encephalopathy (CA-HIE). With continuation of ventilatory and nutritional support, this patient not only survived but over time demonstrated findings that were no longer consistent with brain death. Offered here is a review of the course of events described in the article, an overview of the variable clinical implications of CA-HIE and their relationship to the diagnosis of brain death, a proposed pathophysiologic correlation, and recommendations for palliative clinicians providing consultation with regard to goals of care and intervention options in cases of CA-HIE with severe neurologic injury.


Background: It is difficult to decide whether to inform the child of the incurable illness. We investigated attitudes of the general population and physicians toward prognosis disclosure to children and associated factors in Korea. Methods: Physicians working in one of 13 university hospitals or the National Cancer Center and members of the general public responded to the questionnaire. The questionnaire consisted of the age appropriate for informing children about the prognosis and the reason why children should not be informed. This survey was conducted as part of research to identify perceptions of physicians and general public on the end-of-life care in Korea. Results: A total of 928 physicians and 1,241 members of the general public in Korea completed the questionnaire. Whereas 92.7% of physicians said that children should be informed of their incurable illness, only 50.7% of the general population agreed. Physicians were also more likely to think that younger children should know about their poor prognosis compared with the general population. Physicians who opposed incurable illness disclosure suggested that children might not understand the situation, whereas the general public was primarily concerned that disclosure would exacerbate the disease. Physicians who were women or religious were more likely to want to inform children of their poor prognosis. In the general population, gender, education, comorbidity, and caregiver experience were related to attitude toward poor prognosis disclosure.
to children. Conclusion: Our findings indicate that physicians and the general public in Korea differ in their perceptions about informing children of poor prognosis.


AIM: The Charter of the Rights of the Dying Child was formulated as a professional guide for caring the child in the final stages. The study examines the nurses' degree of agreement with the Charter's principles and their perception of the implementation of those principles in hospital.

MATERIALS AND METHODS: A multicenter, cross-sectional study to observe the nurses' positions about the 10 rights outlined in the Charter, using an online questionnaire in 5 pediatric hospitals in northern Italy. RESULTS: A total of 119 nurses (44.9%) completed the questionnaire. The majority (range: 86.6-100%) expressed their agreement with the Charter's principles (Likert >/=4). Lower ratings were reported in Charter's principles implementation items (range: 42.9-89.1%). Being older and working in a smaller hospital lead the nurses to overlook the child's right to be informed and to be given the opportunity to make decisions about his/her own life and death (p = 0.02, p < 0.01). Postgraduate training induced greater awareness of the dying child (p = 0.01).

CONCLUSIONS: This study highlights the value of the Charter of the Rights of the Dying Child as a reference guideline for nurses working in pediatric hospitals. Better training is important to improve the nurse's communication skills and the pediatric palliative care should be offered to all families that have a child with incurable disease mostly in the end of life.


It is one of the primary goals of medical care to secure good quality of life (QoL) while prolonging survival. This is a major challenge in severe medical conditions with a prognosis such as amyotrophic lateral sclerosis (ALS). Further, the definition of QoL and the question whether survival in this severe condition is compatible with a good QoL is a matter of subjective and culture-specific debate. Some people without neurodegenerative conditions believe that physical decline is incompatible with satisfactory QoL. Current data provide extensive evidence that psychosocial adaptation in ALS is possible, indicated by a satisfactory QoL. Thus, there is no fatalistic link of loss of QoL when physical health declines. There are intrinsic and extrinsic factors that have been shown to successfully facilitate and secure QoL in ALS which will be reviewed in the following article following the four ethical principles (1) Beneficence, (2) Non-maleficence, (3) Autonomy and (4) Justice, which are regarded as key elements of patient centered medical care according to Beauchamp and Childress. This is a JPND-funded work to summarize findings of the project NEEDSinALS (www.NEEDSinALS.com) which highlights subjective perspectives and preferences in medical decision making in ALS.

An infant with complex congenital heart disease suffers a prolonged cardiac arrest with minutes of anoxia. He is left with severe brain damage and profound neurologic impairment. He no longer responds to caregivers. Much of the time, he cries and grimaces as if in pain. He has required increasing sedation to control these symptoms. His parents live hours from the hospital and seldom visit. When their infant's situation is explained to them over the telephone, they request that doctors "do everything to keep him alive." His bedside caregivers report high levels of moral and psychological distress and frequently discuss J.S.'s "suffering." An ethics consultation is requested, asking whether it is permissible to withdraw life support despite the parents' request that therapy continue.


OBJECTIVES: In many Euro-American societies, the ideal of patient and family involvement in clinical decision-making prevails. This ideal exists alongside the doctor's obligation and responsibility to make decisions and to be accountable for them. In this article, we explore how medical staff navigate the tension between autonomy and authority when engaging life-and-death decision-making in a Danish NICU. METHODS: The study rests on ethnographic fieldwork in a Danish NICU, involving participant observations in everyday care and decision-making work and semistructured interviews with staff and parents. All interviews were taped and transcribed. The empirical material was analyzed using thematic coding and validated in discussions with staff, parents, and social scientists. RESULTS: Decisions are relational. Multiple moves, spaces, temporalities, and actors are involved in life-and-death decisions in the NICU. Therefore, the concept of medical decision-making fails to do justice to the complex efforts of moving infants in or out of life. Yet, many of these decision-making moments are staged, timed, and coordinated by medical staff. Therefore, we introduce an alternative vocabulary for talking about life-and-death decision-making in neonatology to help us attend to the moral stakes, the emotional tenor, and the fine-grained mechanisms of authority implied in such decisions around tiny infants. CONCLUSIONS: We conceptualize decisions as an art of "careography." Careography is the work of aligning care for the infant, care for the parents, care for staff, care for other infants, and care for society at large, in the process of deciding whether it is best to continue or withdraw life support.


BACKGROUND:: Metaphors are often used within the context of ethics and healthcare but have hardly been explored in relation to moral reasoning. OBJECTIVE:: To describe a central set of metaphors in one case and to explore their contribution to moral reasoning. METHOD:: Semi-structured interviews were conducted with 16 parents of a child suffering from the neurodegenerative disease CLN3. The interviews were recorded, transcribed, and metaphors were analyzed. The researchers wrote memos and discussed about their analyses until they reached consensus. ETHICAL CONSIDERATIONS:: Participants gave oral and written consent and their confidentiality and anonymity were respected. FINDINGS:: A central set of metaphors referred to
the semantic field of the hands and arms and consisted of two central metaphors that existed in a dialectical relationship: grasping versus letting go. Participants used these metaphors to describe their child’s experiences, who had to “let go” of abilities, while “clinging” to structures and the relationship with their parent(s). They also used it to describe their own experiences: participants tried to “grab” the good moments with their child and had to “let go” of their child when (s)he approached death. Participants, in addition, “held” onto caring for their child while being confronted with the necessity to “let go” of this care, leaving it to professional caregivers.

DISCUSSION: The ethical analysis of the findings shows that thinking in terms of the dialectical relationship between “grasping” and “letting go” helps professional caregivers to critically think about images of good care for children with CLN3. It also helps them to bear witness to the vulnerable, dependent, and embodied nature of the moral self of children with CLN3 and their parents. CONCLUSION: Metaphorical reasoning may support the inclusion of marginalized perspectives in moral reasoning. Future studies should further explore the contribution of metaphorical reasoning to moral reasoning in other cases.


When a child needs surgery, both the surgeon and the anesthesiologist must obtain informed consent from the parents. In theory, each specialist obtains permission for their respective portion of the procedure, with the anesthesiologist only obtaining informed consent for the administration of anesthesia and management in the operating room and recovery room. However, he or she may occasionally realize that the parents have misunderstandings about what the surgery and perioperative course entail. In such cases, he or she must decide whether their role is only to discuss the issues related to anesthesia care or whether he or she should also clarify the range of expected outcomes and the postoperative course after surgery. We present a case in which such a dilemma arose and on which we sought experts in anesthesia and ethics to comment.


The diagnosis and treatment of cancer leads to short-term and long-term challenges for every patient. This is especially true for adolescents and young adults (AYAs) with cancer who strive to gain independence, autonomy, confidence, and social status while developing into adulthood. In this article, we review prominent ethical issues in AYA oncology that are related to autonomy, shared decision-making, care refusal or abandonment, end-of-life care, truth telling, and fertility preservation. Clinicians should recognize that AYA patients develop at their own pace; the onus lies with clinicians to determine the patient’s interests, values, maturity, and desire to participate in decision-making.


The question of whether children with cancer who enroll in clinical trials have superior outcomes compared with those who do not participate has been pursued for more than 4 decades, and recent studies have provided conflicting answers. Whether clinical trial participation influences outcome has important implications for how clinicians should present trial participation to patients and families. Methodological challenges limit generalizations about the impact of clinical trial participation on outcome compared with nonparticipation. Oncologists should inform patients and families that clinical trials are the engine for future progress because they identify more effective therapies and that clinical trial participation is a reasonable option to consider for children with cancer. However, as noted in by Truong and colleagues in this issue, the rationale for trial enrollment should not include an expectation of better outcomes compared with nonenrollment.


OBJECTIVE: To summarize and analyze the impact of specialized pediatric palliative care (SPPC) programs on communication and decision-making for children with life-threatening conditions. METHODS: Our search strategy covered MEDLINE, PsycINFO, Cochrane Central Register of Controlled Trials, Web of Science, CINAHL, Scopus, and Embase through September 2018. RESULTS: We reviewed 13 studies analyzing the impact of SPPC programs on communication and decision-making using a wide range of outcome indicators. Study quality was poor in 58% of included papers. SPPC programs improved communication and decision-making between families and healthcare professionals (HCPs), within and between families, and among HCPs. CONCLUSION: SPPC programs generally support and improve communication and decision-making for children with life-threatening conditions, their families and associated HCPs. Families referred to an SPPC program had more discussions with HCPs on a broad variety of topics. However, data on communication with children, siblings, and other family members was scarce and of poor quality.
PRACTICE IMPLICATIONS: More research on SPPC program efficacy is needed from the perspective of the ill child, as well as about barriers to end-of-life discussions and the specific aspects of SPPC programs responsible for improving outcomes.  


BACKGROUND: Medical advances have led to new challenges in decision-making for parents of seriously ill children. Many parents say religion and spirituality (R&S) influence their decisions, but the mechanism and outcomes of this influence are unknown. Health care providers (HCPs) often feel unprepared to discuss R&S with parents or address conflicts between R&S beliefs and clinical recommendations. Our study sought to illuminate the influence of R&S on parental decision-making and explore how HCPs interact with parents for whom R&S are important. METHODS: A longitudinal, qualitative, descriptive design was used to (1) identify R&S factors affecting parental decision-making, (2) observe changes in R&S themes over time, and (3) learn about HCP perspectives on parental R&S. The study sample included 16 cases featuring children with complex life-threatening conditions. The length of study for each case varied, ranging in duration from 8 to 531 days (median = 380, mean = 324, SD = 174). Data from each case included medical records and sets of interviews conducted at least monthly with mothers (n = 16), fathers (n = 12), and HCPs (n = 108). Thematic analysis was performed on 363 narrative interviews to identify R&S themes and content related to decision-making. RESULTS: Parents from 13 cases reported R&S directly influenced decision-making. Most HCPs were unaware of this influence. Fifteen R&S themes appeared in parent and HCP transcripts. Themes most often associated with decision-making were Hope & Faith, God is in Control, Miracles, and Prayer. Despite instability in the child's condition, these themes remained consistently relevant across the trajectory of illness. R&S influenced decisions about treatment initiation, procedures, and life-sustaining therapy, but the variance in effect of R&S on parents' choices ultimately depended upon other medical & non-medical factors. CONCLUSIONS: Parents consider R&S fundamental to decision-making, but apply R&S concepts in vague ways, suggesting R&S impact how decisions are made more than what decisions are made. Lack of clarity in parental expressions of R&S does not necessarily indicate insincerity or underestimation of the seriousness of the child's prognosis; R&S can be applied to decision-making in both functional and dysfunctional ways. We present three models of how religious and spiritual vagueness functions in parental decision-making and suggest clinical applications.  


Parents whose child is diagnosed with a serious disease such as trisomy 18 first rely on the medical community for an accurate description and prognosis. In the case of trisomy 18, however, many families are told the disease is "incompatible with life" even though some children with the condition live for several years. This paper considers parents' response to current medical
discourse concerning trisomy 18 by examining blogs written by the parents of those diagnosed. Using interpretive humanistic reading and foregrounding Cathryn Molloy’s recuperative ethos theory (2015), we find that parents demonstrate recuperative ethos in response to physicians’ descriptions of trisomy 18, particularly in rhetoric addressing survival, medicalized language, and religious and/or spiritual rhetoric. We argue that, by using language such as “incompatible with life,” physicians distance themselves from families, creating not care, but the very gulf that requires recuperation. We conclude that medical professionals would do well to engage with the trisomy 18 community-including learning from blogs and online forums- employ palliative care practices, and seek more accurate, descriptive language that is compatible with care.


In this article, I identify 3 ways of justifying neonatal policies of when to provide life-saving treatment to infants who were born extremely premature: by appealing to universal principles or rights, to considerations of the best interests of the children, or to considerations of the best interests of the families. I go on to show how each of these justifications can be used to characterize the discourse on neonatal policies in 1 of the Scandinavian countries.


OBJECTIVES: The role of parents in life-and-death decision-making for infants born at the border of viability is challenging. Some argue that parents should have the final say in decisions about life-sustaining treatment. Others disagree. In this article, we report views from health care personnel (HCP) on the appropriate parental role. METHODS: Focus group interviews with 5 different groups of HCP (neonatal nurses, midwives, obstetricians, mother-fetal specialists, and neonatologists) dealing with life-and-death decisions throughout pregnancy and birth were performed at the Norwegian University of Science and Technology and at St Olav's Hospital in Trondheim, Norway in 2014-2017. Interviews were taped and transcribed. Inductive analysis was performed for each group discussion for emergent ethical themes. A summary of the transcribed discussion was sent to the relevant focus group participants for comments. RESULTS: Our participants felt strongly that doctors, not parents, should have the final say. They did not think parents should have to live with the burden of the decision. The possible disagreement between parents, lack of necessary knowledge, experience, time, and emotional stability all point toward the neonatologist as the optimal decision-maker, within a model of “Patient Preference-Satisfaction Paternalism.” CONCLUSIONS: The general attitude of our groups was that parents should have a say and be included in a thorough information and decision-making process. The doctor, or a team of HCP, however, should make the final decision, being in the best position both epistemologically and normatively to promote the best interest of both parents and the child.

An examination of the policies regarding the care of extremely premature newborns reveals unexpected differences between Scandinavian countries and the Netherlands. Three topics related to decision-making at the beginning and at the end of life are identified and discussed. https://www.ncbi.nlm.nih.gov/pubmed/30171145
Education, Professional and Research


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.


PURPOSE: This study explores the experience of disclosing critical information in the care of children with palliative care needs, from the perspective of physicians, nurses, and mothers in Jordan. DESIGN AND METHODS: This study employed a qualitative case study approach. It was conducted in three paediatric units in a Jordanian hospital. Each case comprised a child aged 1-12 years with a condition eligible for palliative care who received health care in one of these units, and their most involved carers (e.g. mother, physician and nurse). Two data collection methods were employed: participant observation and semi-structured interviews with three categories of participants: mothers, physicians, and nurses. Ethical approval was obtained from the hospital ethical review board. Written consent was obtained from all participants. RESULTS: Qualitative case studies were developed around 15 children (aged 1-12 years, nine were boys and six were girls, with varying diagnoses: renal disease, neurological conditions, and congenital heart defects). A total of 197 observational hours and 60 interviews were completed (15 mothers, 12 physicians and 21 nurses). The findings demonstrate that the practice of 'mutual protection' dominated communication between children, parents and clinical staff. Parents protected their children by disclosing only partial information about their disease, and by avoiding any information they thought would cause the child distress or loss of hope. Similarly, children avoided expression to
their parents of their anxieties or fears, in order to protect them. In turn, nurses attempted to ensure observance of professional boundaries with children and mothers to avoid a sense of loss when a child died. CONCLUSION: The findings of the current study indicate that while open and honest communication between parents and children is generally recommended by literature, not all mothers agree with adopting open communication with their children concerning their illnesses. Therefore, any future intervention planned for them should respect parents' autonomy and decisions in addition to their cultural backgrounds. PRACTICAL IMPLICATIONS: The provision of ongoing education and specialised training for professionals to provide them with culturally sensitive skills in communication and provision of emotional support for children and parents is needed to improve clinical practice in healthcare settings with limited access to specialist palliative care such as Jordan.


OBJECTIVE: Engaging bereaved parents in the review process that examines their care before and after a perinatal death might help parents deal with their grief more effectively and drive improvements in patient safety. The objective of this study is to explore whether healthcare professionals would accept or support parent engagement in the perinatal mortality review process. DESIGN: Qualitative focus group interviews. Transcripts were analysed with an inductive thematic approach. SETTING: Two geographically distinct tertiary maternity hospitals in the UK. PARTICIPANTS: Five focus groups were conducted with clinical staff including midwives, obstetricians, neonatologists, nursing staff and chaplaincy services. RESULTS: Twenty-seven healthcare professionals unanimously agreed that parents' involvement in the perinatal mortality review process is useful and necessary. Six key themes emerged including: parental engagement; need for formal follow-up; critical structure of perinatal mortality review meeting; coordination and streamlining of care; advocacy for parents including role of the bereavement care lead; and requirement for training and support for staff to enable parental engagement. CONCLUSIONS: Healthcare professionals strongly advocated engaging bereaved parents in the perinatal mortality review: empowering parents to ask questions, providing feedback on care, helping generate lessons and providing them with the opportunity to discuss a summary of the review conclusions with their primary healthcare professional contact. The participants agreed it is time to move on from 'a group of doctors reviewing notes' to active learning and improvement together with parents, to enable better care and prevention of perinatal death.


BACKGROUND: In planning high-quality research in any aspect of care for children and young people with life-limiting conditions, it is important to prioritise resources in the most appropriate
areas. AIM: To map research priorities identified from existing research prioritisation exercises relevant to infants, children and young people with life-limiting conditions, in order to inform future research. DESIGN: We undertook a systematic scoping review to identify existing research prioritisation exercises; the protocol is publicly available on the project website. DATA SOURCES: The bibliographic databases ASSIA, CINAHL, MEDLINE/MEDLINE In Process and Embase were searched from 2000. Relevant reference lists and websites were hand searched. Included were any consultations aimed at identifying research for the benefit of neonates, infants, children and/or young people (birth to age 25 years) with life-limiting, life-threatening or life-shortening conditions; their family, parents, carers; and/or the professional staff caring for them. RESULTS: A total of 24 research prioritisation exercises met the inclusion criteria, from which 279 research questions or priority areas for health research were identified. The priorities were iteratively mapped onto an evolving framework, informed by World Health Organization classifications. This resulted in identification of 16 topic areas, 55 sub-topics and 12 sub-sub-topics. CONCLUSION: There are numerous similar and overlapping research prioritisation exercises related to children and young people with life-limiting conditions. By mapping existing research priorities in the context in which they were set, we highlight areas to focus research efforts on. Further priority setting is not required at this time unless devoted to ascertaining families’ perspectives. https://www.ncbi.nlm.nih.gov/pubmed/30404588


Increased emphasis on the child’s voice and point of view in care and treatment has led to an expansion in the development of methods to access and identify their perspectives. Drawing on our experiences in a study of children with leukemia in hospital, this article explains the challenges and opportunities that arise in the use of five commonly used methods in a study of hospitalized children’s experiences with health care professionals, including the “Draw and Write” technique, a sticker activity, a paper-person exercise, informal interviews, and participant observation. Each of these methods was examined with regard to ease of use, data generation, and utility of data for accessing children’s perspectives and development of initial clinical guidance. https://www.ncbi.nlm.nih.gov/pubmed/30270755


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


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.


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.


Background: For children with life-shortening illness, achieving a "good death" can be a tacit goal. There is little understanding of how different stakeholders perceive what a "good death" might be. Objective: To review empirical literature to construct an understanding of a "good death" for children with life-shortening conditions. Design: An integrative review approach was followed. This involved searching across Embase, Web of Science, Medline, CINAHL, and PsycINFO (no date limits set), as well as identifying eligible studies tracking reference lists. Appraisal of shortlisted articles in full text was performed, followed by data extraction, synthesis, and interpretation. Results: Analysis of articles (n = 24) yielded a dynamic and layered narrative about a good death that revolved around three themes. (1) Level of needs: includes both practical support and aspirational goals such as "do everything." (2) The composite experience: whether positive or negative adds to produce a sense of suffering. (3) Control (preservation and letting go): moving from maintaining status quo to acceptance of the child's death, the experience of which also contributes to suffering.
Framed using a health care system perspective, a concept map that interprets a good death in children with life-shortening conditions is represented. Conclusions: A single yet holistic understanding of a good death experienced in the “real world” is suggested. Pediatric health and social care providers, and even policy makers, can use this new understanding to conceive alternative approaches to enhance support to dying children and their families.


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

https://www.ncbi.nlm.nih.gov/pubmed/29552895


PURPOSE: Preparing a future nurse to respond to the complex and sensitive needs of a child and family during the end-of-life requires more than didactic content in a classroom. During clinical experiences, students may care for children diagnosed with a terminal illness however; it is less likely that a student will have a clinical opportunity to care for a child and their family at the end-of-life. Without having an experience, it is challenging to teach students how to care for the dying child and family including how to appreciate the emotions, thoughts, and expectations when faced with a pediatric death (Lindsay, 2010). DESIGN AND METHODS: The instructional model integrates an end-of-life simulation into an undergraduate pediatric nursing course allowing students to practice caring for a child and their family while developing an understanding of the unique needs of a dying pediatric patient. RESULTS: Post simulation, students participating in guided reflection, identified several themes impacting their experience with end-of-life care, including symptom management, emotional care and “what to say”. CONCLUSIONS: The structured simulated
experience provided knowledge, skill and awareness to the role of the nurse when providing care at the end-of-life. PRACTICE IMPLICATIONS: Nurses’ behaviors and responses when caring for a child can have a significant impact on the family’s experience and memory of their child’s death. Unintended actions may result in the family experiencing negative impressions, causing further distress to the grieving family (Butler, Hall, Willetts, & Copnell, 2015).


BACKGROUND: Paediatric palliative care (PPC) aim to ensure the control of symptoms and the best possible quality of life for patients whose underlying disease, characterized by an unstoppable evolution and negative prognosis, no longer responds to specific treatments. The scientific evidence in this context are very deficient and, in order to obtain welfare objectives consistent with the situation, in the overwhelming majority of cases the prescription of drugs is off-label for indication of use and/or for age and/or for way of administration and/or formulation. The Agenzia Italiana del Farmaco - AIFA and the Italian Society of Palliative Care (Societa Italiana di Cure Palliative - SICP), under a dedicated working group, wrote a document that collects the scientific evidence available to support the off-label use of medicines more frequently used in PPC. The goal is to certify the consolidated off-label use of these drugs and propose their use under the Law 648/96, in the absence of data from its pivotal clinical trials. Aim of the commentary is to report the conditions for this important work and to present the 10 drugs, usually used off-label in PPC and in pain therapy, now included in Law 648/96. CONCLUSION: This work is deemed essential to resolve, at least in part, the lack of availability of medicines researched and approved.


OBJECTIVE: About 14% of cancer patients live with dependent children. Healthcare professionals are well placed to help patients support their children as part of a patient-centred practice. Children tend to appreciate open communication during the course of illness, but patients often find this difficult. However, research is unclear about patients' preferences and their willingness to talk with healthcare professionals about their dependent children. METHODS: We conducted 15 in-depth interviews with patients from haematological (N = 11) and gynaecological oncology (N = 4). The interviews and subsequent analysis focused on patients' communicative preferences, taking the theoretical framework of "biographical disruption" as a starting point and using Jenkins' concept of identity as a social, relational and dynamic process. RESULTS: We identified two overall identities at stake for seriously ill patients with parental responsibility: "patient identity" and "parent identity." As "patients," patients were ambivalent about relating to their children, but as "parents" they wanted healthcare professionals to talk about their children. CONCLUSION: In order to be patient-centred, clinicians should, we suggest, acknowledge that patients have these conflicting perspectives and identities, which surface at various times and situations throughout
their illness trajectories. Research is needed to further explore these findings in different illness groups and cultures.


The International Children's Palliative Care Network (ICPCN) held its third international conference on children's palliative care in Durban, South Africa, from May 30 2018 to 2 June 2018. The conference—inspiration, innovation and integration—brought together 250 participants from 41 countries and was held in conjunction with local partners-Umduduzi Hospice Care for Children, Palliative Treatment for Children South Africa (Patch SA) and the Hospice and Palliative Care Association of South Africa. It built on national and global developments in palliative care such as its inclusion in Universal health coverage (UHC), the Lancet Commission report on pain and palliative care and the sustainable development goals (SDGs), and aimed to raise the profile of children’s palliative care in KwaZulu-Natal (KZN) and nationally. Seven pre-conference workshops were held prior to the conference on topics such as pain and symptom management, children’s palliative care within a humanitarian crisis, perinatal palliative care, research, developing programmes, ethical issues and difficult conversations in children’s palliative care. Delegates were welcomed in true Durban style at the welcome reception hosted by the City of Durban and uShaka Marine World. The opening plenary included entertainment from the Open Air School and Hillcrest Primary School, and inspirational talks from the Member of the Executive Council (MEC) for Health, a representative of the World Health Organization (WHO), the Chief Executive of ICPCN and the Noble Peace Prize Nominee Dr MR Rajagopal from Pallium India. Plenary sessions were interspersed throughout the conference with 56 oral concurrent presentations and workshops, six ‘Meet the expert sessions’ 100 poster presentations and the South African Premier of the film 'Hippocratic: 18 Experiments in gently shaking the world'. There was a great feeling of networking and learning throughout the conference, with the conference being well evaluated, and an increase in the level of presentations and research from previous conferences demonstrating the steps that are being taken in children’s palliative care globally.


The purpose of this paper is to present a conceptual-theoretical-empirical model addressing factors that influence the relation between parental uncertainty and health-related quality of life in children with cancer. The basic concepts identified and defined in the model include parental uncertainty, health-related quality of life, parental trait anxiety, parental depression, and perceived parental social support. The proposed relationships between the concepts are explained with explicit linkages to their empirical indicators. There is limited research in childhood cancer regarding the relation between parental uncertainty and health-related quality of life; therefore, the proposed model will help to better understand this relationship.

Paediatric palliative care has only been clearly referred to as such in France since 2008. It is defined as care provided from the antenatal period until the age of 19 to children suffering from a disease which limits or threatens their life expectancy. Many nurses will encounter patients such as these at some point during their career in a health or medical-social facility. They require multi-disciplinary care and coordination between all the health professionals involved in the treatment.


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.


BACKGROUND: The goal of the 4th edition of the National Consensus Project Clinical Practice Guidelines for Quality Palliative Care (NCP Guidelines) is to improve access to quality palliative care for all people with serious illness regardless of setting, diagnosis, prognosis, or age. OBJECTIVE: The NCP Guidelines are intended to encourage and guide healthcare organizations and clinicians (including nonpalliative care specialists) across the care continuum to integrate palliative care principles and best practices into their routine assessment and care of all seriously ill patients and their family caregivers. METHODS: The NCP Guidelines formalize and delineate evidence-based processes and practices for the provision of safe and reliable high-quality palliative care for adults, children, and families with serious illness in all care settings. RESULTS: This article presents the key domains and guidelines of the 4th edition.


OBJECTIVE: Qualitative research is pivotal in gaining understanding of individuals' experiences in pediatric palliative care. In the past few decades, the number of qualitative studies on pediatric palliative care has increased slightly, as has interest in qualitative research in this area. Nonetheless, a limited number of such studies have included the first-person perspective of children. The aim of this article is to understand the contribution of previous qualitative research on pediatric palliative care that included the voices of children. METHOD: A systematic review of qualitative studies and a meta-summary were conducted. MEDLINE, CINAHL, PsycINFO, PsycARTICLES, and ERIC were searched without limitations on publication date or language. Eligible articles were qualitative research articles in which the participants were children ranging in age from 3 to 18 years. Result We retrieved 16 qualitative research articles reporting on 12 unique studies, and we selected two mixed-method articles. The meta-summary shows eight themes: the relationship with professional caregivers, pain and its management, "living beyond pain," the relationship between pediatric patients and their families, children's view on their treatment and service provision, meanings children give to their end-of-life situation, consequences of clinical decisions, and the relationships among children in pediatric palliative care and their peers. Significance of results This meta-summary presents the "state of the art" of pediatric palliative care qualitative research on children and highlights additional research areas that warrant qualitative study.


Background: The "Children's Palliative Care Project" was initiated in October 2010 in the Indian state of Maharashtra with a view to improve the quality of life of children with life-limiting conditions. This study evaluates its education and training component through a questionnaire. Materials and Methods: A cross-sectional survey was carried out pre-/post-training among 258 doctors, nurses, social workers, and counselors at three sites in Maharashtra in March 2015. Descriptive statistics were used for data analysis. Results: Sixty-two participants responded. Posttraining, doctors and the nurses had a better level of knowledge, skill set, and attitude; whereas social workers and counselors fared better with prevailing care practices. Participants advocated using morphine only when other analgesics had failed and suggested ways for better service delivery of care. Conclusion: The study gives a rough idea of the prevailing practice of pediatric palliative care among the health-care workers (who participated in the survey) and suggests practical ways to improve it.


OBJECTIVES: To describe how parents of neonatal intensive care unit (NICU) graduates with cerebral palsy (CP) perceive both the accuracy of prognoses provided in the NICU and the timing of their child’s diagnosis of CP, and to assess the influence of functional outcome on these perceptions. STUDY DESIGN: We surveyed parents of NICU graduates with CP about timing and benefit of diagnosis, accuracy of prognosis, and functional abilities of their children. After piloting and validation, CP parent support groups circulated the survey on social media, websites, and email lists. Bivariate relationships between categorical responses to survey questions were assessed with the chi(2) test, and multivariable logistic regression was performed to identify independent factors associated with perceptions about the timing of diagnosis. RESULTS: Parents of 463 children were included. Two-thirds (67%) of the children were diagnosed with CP before age 2 years, yet 40% of the respondents felt that diagnosis was made late, and only 11% categorized diagnosis as early. More than one-half (59%) perceived a benefit to diagnosis. There was a significant association between earlier age at diagnosis and greater functional limitations; 24% of parents who recalled being given a prognosis reported that their child functioned as predicted, and 46% reported that their child exceeded expectations. Parents were more likely to believe that children with fewer functional limitations had exceeded expectations. CONCLUSION: Parents remember prognostic discussions about children who develop CP as underestimating functional outcome. Diagnosis is rarely seen as early and is associated with benefits. These observations suggest that clinicians should aim to diagnose CP early and to maintain guarded optimism about future outcomes. Tools for improved communication are urgently needed. 


AIM: We examined how physicians in different medical specialties would evaluate treatment decisions for vulnerable patients in need of resuscitation. METHODS: A survey depicting six acutely ill patients from newborn infant to aged, all in need of resuscitation with similar prognoses, was distributed (in 2009) to a representative sample of 1650 members of the Norwegian Medical Association and 676 members of the Norwegian Pediatric Association. RESULTS: There were 1335 respondents (57% participation rate). The majority of respondents across all specialties thought resuscitation was in the best interest of a 24 weeks' gestation preterm infant and would resuscitate the patient, but would also accept palliative care on the family's demand. Accepting a family's refusal of resuscitation was more common for the newborn infants. Specialists were overall similar in their answers, but specialty, age and gender were associated with different answers for the patients at both ends of the age spectrum. CONCLUSION: Resuscitation decisions for the very young do not always seem to follow the best interest principle. Specialty and personal characteristics still have an impact on how we consider important ethical issues. We must be cognisant of our own valuations and how they may influence care. 

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.


Given the limited number of pediatric-specific palliative care programs, palliative care providers of all disciplines may be called on to care for infants, children, and adolescents with serious illness. This article provides a review of the unique components of pediatric palliative care, including key roles within an interdisciplinary team, pediatric developmental considerations, use of medical technology and complexities of symptom management in children with serious illness, hospice utilization, as well as pointers for discussions with families regarding a patient’s quality of life and goals of care.


Historically, communication research in pediatric oncology has relied on surveys and interviews, resulting in cross-sectional and retrospective studies constrained by selection, recognition, and recall biases. This systematic review identifies and synthesizes the published literature analyzing primary data from recorded conversations between pediatric oncologists, patients with cancer, and their families, with the following objectives: (1) to identify the extent and content of the evidence base, (2) to describe methodological strategies utilized in the analysis of recorded medical dialogue, (3) to aggregate salient findings, and (4) to generate recommendations for future prospective research related to analysis of medical dialogue in pediatric oncology.


OBJECTIVE: This paper presents data on licensure/certification status, supervision of multidisciplinary pediatric psychosocial staff, and training opportunities in pediatric cancer.
programs in the United States, data that are critical to provide care aligned with the Standards of Psychosocial Care in Pediatric Cancer (Psychosocial Standards). METHODS: An online survey of psychosocial care consistent with the Psychosocial Standards was completed from a national sample of pediatric cancer programs (144/200). Licensure/certification status, availability and format of supervision for multidisciplinary staff (social workers, psychologists, psychiatrists, child life specialists/recreational therapists), and types and number of psychosocial trainees were reported. RESULTS: Nearly all pediatric psychosocial providers were licensed/certified. Peer consultation was the most frequently endorsed form of staff supervision although a sizeable group of centers reported no systematic ongoing supervision. Trainees in social work and child life were most common although the size of trainee cohorts is generally small. Psychosocial trainees are more prevalent in sites with pediatric hematology/oncology medical fellowship programs and in larger programs. CONCLUSIONS: A properly trained and supported psychosocial workforce is essential to providing evidence-based care consistent with the Psychosocial Standards. Psychosocial providers are appropriately licensed. However, supervision opportunities are variable and may be inadequate for the intensity of the work. It is important to address the limited opportunities for trainees in pediatric cancer programs, which may influence the pipeline for ongoing and future work in this area.


BACKGROUND: Given the dearth of literature and no clinical practice guidelines written for speech-language pathologists (SLPs) working in paediatric palliative care (PPC), a need has been identified to explore the scope of clinical practice and strategies used by SLPs. OBJECTIVE: This study aims to undertake an international investigation into the role and scope of practice of SLPs working in PPC to develop consensus-driven 'Recommendations for Speech-Language Pathologists in Paediatric Palliative Care Teams' (ReSP(3)CT). METHODS: A modified Delphi process will be used to synthesise consensus-based statements from SLPs in six different countries about their role and practice working in PPC. Initially, preliminary survey data will be collected from SLPs to obtain demographic and caseload information. Respondents will then be invited to participate in an in-depth interview to explore common and unique themes that emerge from the online survey. Participants from the interview will then 'opt-in' to become Delphi panel members and receive questionnaires comprising statements for agreement over multiple rounds. Statements will be based on common themes that arise from the literature review, survey and interview data. The Delphi process for each statement will stop if statements achieve >/= 70 % agreement and an IQR of </= 1 (maximum of five rounds). CONCLUSION: This is the first study to investigate the role and practice of SLPs in PPC across internationally accepted scope of practice areas. The study will use existing frameworks for statistical analysis and a mixed-methods approach to aid in the synthesis of statements/recommendations for international consensus.

PURPOSE: Attempts have been made within the literature to clarify the role and scope of speech-language pathologists (SLPs) within paediatric palliative care (PPC). As SLP literature regarding adult/geriatric populations is gaining traction, it is fitting to investigate the role of SLPs in the management of infants and children in end-of-life care. METHOD: Arksey and O’Malley’s (2005) scoping review method was utilised for searching multiple databases. Two database searches were undertaken. The first located literature in which SLP PPC intervention is specifically addressed. The second search utilised internationally recognised SLP scope of practice areas. Manual searching of reference lists was also utilised. RESULT: Themes identified included management of communication, feeding, upper-airway and oral health as well as the role of SLPs within a multidisciplinary PPC team. CONCLUSION: There is acknowledgement that SLPs have a role in PPC. However, there is little information identifying SLP involvement in the diagnosis and management of swallowing, cognition/communication, oral hygiene and upper airway issues. The available literature predominantly relies on limited adult palliative care research and does not address age-specific management approaches across the paediatric life-stage. Given an absence of SLP PPC guidelines, further research is warranted to explicitly define SLP scope of practice within this population.


PURPOSE: Low recruitment of adolescents and young adults in cancer clinical trials is widely reported and may be linked to limited improvements in survival. Research to date does not adequately explain all underlying reasons for poor trial accrual. This paper reports health professional perceptions of communicating with adolescents and young adults with bone sarcoma about clinical trial participation. METHODS: This study used narrative inquiry. Findings are reported from thematic analysis of in-depth interviews with 18 multidisciplinary health professionals working in a supra-regional bone and soft tissue sarcoma centre. RESULTS: Participants described professional expertise, the development of specialist knowledge and skills and strategies used to develop trusting relationships with adolescents and young adults with bone sarcoma. These factors were perceived to facilitate communication about clinical trial participation. Emergent themes were having credibility through expertise of the team, developing specialist communication skills through reflection on practice, having inclusive approaches to education and training about clinical trials, individual communication styles used to form trusting relationships, using a patient-centred approach to connect with adolescents and young adults, creating time needed to form trusting relationships and effective team working. CONCLUSIONS: We aligned findings of this study with characteristics of patient-physician trust and provide a basis for transferable recommendations. Our findings can be used to inform the development of age-specific, specialist communication skills and highlight health professional education needs about clinical trials. Additional research is needed to explore which elements of team working optimise improved clinical trial participation, in what contexts and why.


BACKGROUND: High-quality oncology care is marked by skillful communication, yet little is known about patient and family communication perceptions or content preferences. Our study sought to elicit pediatric oncology patient and parent perceptions of early cancer communication to establish whether informational needs were met and identify opportunities for enhanced communication throughout cancer care. METHOD: An original survey instrument was developed, pretested, and administered to 129 patients, age 10-18 years, and their parents at 3 cancer centers between 2011 and 2015. Statistical analysis of survey items about perceived communication, related associations, and patient/parent concordance was performed. RESULTS: A greater percentage of participants reported "a lot" of discussion about the physical impact of cancer (patients, 58.1% [n = 75]; parents, 69.8% [n = 90]) compared with impact on quality of life (QOL) (patients, 44.2% [n = 57]; parents, 55.8% [n = 72]) or emotional impact (patients, 31.8% [n = 41]; parents, 43.4% [n = 56]). One fifth of patients (20.9% [n = 27]) reported they had no up-front discussion about the emotional impact of cancer treatment. Parents indicated a desire for increased discussion regarding impact on family life (27.9% [n = 36]), long-term QOL (27.9% [n = 36]), and daily activities (20.2% [n = 26]). Patients more frequently than parents indicated a desire for increased physician/patient discussion around the impact on daily activities (patients, 40.3% [n = 52]; parents, 21.7% [n = 28]; P < .001), long-term QOL (patients, 34.9% [n = 45]; parents, 16.3% [n = 21]; P < .001), pain management (patients, 23.3% [n = 30]; parents, 7% [n = 9]; P < .001), physical symptom management (patients, 24% [n = 31]; parents, 7.8% [n = 10]; P < .001), short-term QOL (patients, 23.3% [n = 30]; parents, 9.3% [n = 12]; P < .001), and curative potential (patients, 21.7% [n = 28]; parents, 8.5% [n = 11]; P = .002, P values calculated using McNemar's test). CONCLUSION: Oncologists may not be meeting the informational needs of many patients and some parents/caregivers. Communication could be enhanced through increased direct physician-patient communication, as well as proactive discussion of emotional symptoms and impact of cancer on QOL. https://www.ncbi.nlm.nih.gov/pubmed/30602057


Adolescent and young adults diagnosed with cancer represent a vulnerable population needing careful collaborative care from interprofessional teams. Healthcare providers must understand and appreciate the respective scopes of practice of palliative care team members to maximize the quality of care provided to these patients. A team of graduate students engaged in a collaborative learning activity to explore professional roles and responsibilities of palliative care team members when caring for adolescent and young adult oncology patients. Following a literature review and community expert interviews, students identified shared responsibilities of all team members and unique contributions of various professions. Engaging in this process highlighted and clarified the full scope of practice for each specialized team member. Educators should consider utilizing a
similar collaborative learning activity to enhance students' understanding of the roles and responsibilities of each member of the interprofessional healthcare team. 


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 showdowns and discusses accepted moral practice in this difficult area. 


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. 

BACKGROUND: The principal aim of this study was to understand how communication between parents and health professionals concerning prematurity occurs, from delivery to admission to the neonatal Intensive Care Unit. METHODS: This is an exploratory, descriptive study with a qualitative methodology. Data were collected using tape-recorded and Focal Groups technique interview with mothers of premature newborns and health professionals involved in caring for preterm infants, at southeast Brazil. RESULTS: The word “premature” was not said or heard during prenatal care. From the narratives, it was observed that there was a lack of information available to pregnant women about preterm birth, failure in medical care regarding signs and symptoms reported by pregnant women, and lack of communication between the medical teams, mothers and family during delivery and Neonatal Intensive Care Unit (NICU) admission. CONCLUSION: There is a fine line between born too soon and die too soon, that increases stress, fear and distance impacting negatively over communication between mothers and health professionals during antenatal care, childbirth and NICU admission.


The experiences of end-of-life care by nurses in the pediatric intensive care unit are the subject of this systematic review. Six qualitative articles from three different countries were chosen for the review using methods from Joanna Briggs Institute. The themes discovered included the following: insufficient communication, emotional burden, moral distress from medical futility, strengthening resilience, and taking steps toward hospice. These themes are discussed in detail followed by recommendations for practice to assist nurses in their quest for a good death for their pediatric patients.


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.


In the recent decades, expressive arts (EXA) has been used in end-of-life care (EOLC) for facilitating the quality of life of the patients and the caregivers. However, it may not be practical for every EOLC service to dispense EXA activities solely by extensively trained art therapy specialists. There is currently a lack of brief training for nonart therapists, which may have stifled the application of the techniques in clinical settings. The current study therefore described and evaluated the effectiveness of a 2-day EXA training workshop in enhancing practice, knowledge, and self-competence among health and social care professionals working in EOLC using a mixed-method approach. The quantitative findings show significant improvement in perceived competence of providing services per holistic and person-centered EOLC objectives, nonpharmaceutical management of symptoms, and evidence-based psychosocial care as well as self-competence in death work (SCDW) after the workshop. The qualitative findings corroborated the quantitative results by suggesting that the improvement in competence could be associated with enhanced communication, meaning reconstruction, and therapeutic relationship with the clients as well as the improvement in mood, socialization, and self-esteem among the clients through the learned EXA activities. Our findings support the efficacy of a brief training of EXA activities for nonart therapists in enhancing multifaceted intervention competence. Further research on brief training will be needed to promote the use of EXA activities in the EOLC context.


Being able to communicate effectively is an essential skill for all nurses. Communication in paediatric end of life care can be challenging for both the student and lecturer as it is a rare experience and challenging to teach. Innovative approaches to teaching communication skills such as role play, simulation and drama have been used; however there is a dearth of literature examining the use of drama in this specialist context. The aim of this study was to explore the effectiveness of a novel workshop in teaching transferable knowledge and skills in palliative, end of life and bereavement care communication to a convenience sample of first year pre-registration nursing students undertaking clinical skills training at a UK university. Qualitative and quantitative data were obtained from pre and post intervention questionnaires exploring student’s perception of communication skills. Qualitative data were analysed thematically and quantitative data presented as standard descriptive statistics. The novel communication workshop facilitated students’ exploration of how good and poor communication looks and feels and introduced aids to inform communication in clinical practice. Exposure to different learning approaches provided opportunities to both gain confidence in engaging in new learning activities and develop knowledge and skills through purposeful engagement.


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.


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.


The portrait of a dying child is an homage to a child's journey from initial diagnosis to the terminal stages of illness in metastatic neuroblastoma, raising the critical question of the importance of defining a beautiful death - a concept I first came across as a literature student in Henrik Ibsen's renowned tragedy Hedda Gabler. In this article, we discuss a case study of a child named Peter (real names have been changed to maintain the confidentiality of the patient) and his family, whom I met during my oncology rotation as a junior pediatric resident, and various aspects of care - ranging from symptom management, pain control, the family as an emotional and spiritual unit and complications of metastatic disease. Interlaced amidst references of current practices related to pain control and palliation of symptoms are quotes from Dr Myra BluebondLangner's books and a personal encounter with the child to construct the child as a child and not another dying patient. The enigma of the definition of a beautiful death is also discussed from the patient and the family's point of view. An ideal medical death would be one without pain and with optimal symptom control; however, a beautiful death is so much more - encompassing a peaceful passing surrounded not by machines but by happiness around and at heart. We hope this article would
encourage pediatricians to continue to practice pediatric palliative care in the daily setting when dealing with critically ill patients or children in their final stages of life.


CONTEXT: In our increasingly multicultural society, providing sensitive and respectful pediatric palliative care is vital. OBJECTIVES: We held a one-day workshop conference with stakeholders and pediatric clinicians to identify suggestions for navigating conflict when cultural differences are present and for informing standard care delivery. METHODS: Participants explored cases in one of four workshops focused on differences based on race/ethnicity, economic disparity, religion/spirituality, or family values. Each workshop was facilitated by two authors; separate transcriptionists recorded workshop discussions in real time. We used content analyses to qualitatively evaluate the texts and generate recommendations. RESULTS: Participants included 142 individuals representing over six unique disciplines, 25 of the U.S., and three nations. Although the conference focused on pediatric palliative care, findings were broadly generalizable to most medical settings. Participants identified key reasons cultural differences may create tension and then provided frameworks for communication, training, and clinical care. Specifically, recommendations included phrases to navigate emotional conflict, broken trust, unfamiliar family values, and conflict. Suggested approaches to training and clinical care included the development of core competencies in communication, history taking, needs assessment, and emotional intelligence. Important opportunities for scholarship included qualitative studies exploring diverse patient and family experiences, quantitative studies examining health disparities, and randomized clinical trials testing interventions designed to improve community partnerships, communication, or child health outcomes. CONCLUSION: Taken together, findings provide a foundation for collaboration between patients, families, and clinicians of all cultures.


Purpose: Around 170 multidisciplinary staff of the Oncology Services Group at Queensland Children's Hospital, Brisbane, care for children with oncology, hematology, and palliative care needs from throughout Queensland and northern New South Wales. A series of challenges impacted staff resilience and retention, and strategies were needed to improve staff well-being and enable them to flourish despite the inherent work stressors. Methods: A needs analysis was conducted using themes from Discovery Interviews with 51 staff, surveys related to "The Work Stressors Scale - Pediatric Oncology" and "The Work Rewards Scale - Pediatric Oncology" completed by 59 staff, and an organizational staff survey responded to by 51 staff. Results: The needs analysis informed the development of a customized Oncology Staff Well-being Program with a range of strategies aligned to a PERMA framework for flourishing (positive emotion, engagement, relationships, meaning, and accomplishment). Positive emotion areas included education on topics such as well-being, resilience, responding to escalating behaviors, grief and
loss, and self-care. Staff attended the available mindfulness sessions, debriefing and counselors on site, developed self-care plans, and followed a well-being Facebook Group. Engagement was supported through exploring character strengths, improving communication, supporting innovation, and addressing frustrations and safety concerns. Relationships within the team were addressed through team building and social events. Meaning of the work was emphasized through sharing family updates and end of treatment celebrations. Accomplishments of staff were acknowledged in newsletters and meetings. Conclusion: The needs analysis drove a multifaceted approach to staff well-being with the development of strategies which aligned to a framework that would empower staff to flourish at work. Implementation and evaluation are ongoing and will be reported in a subsequent paper.

https://www.ncbi.nlm.nih.gov/pubmed/30532608


Purpose: Challenges experienced by staff in the Oncology Services Group at Queensland Children's Hospital led to issues with staff retention, well-being, and stress on team culture. Therefore, a customized program was developed through a needs analysis to improve the well-being and resilience of oncology staff, enabling them to cope with stressors and critical incidents inherent in their everyday work and to flourish. The program included education, on-site counselors, mindfulness sessions, debriefing, well-being resources, and improved engagement, support, and communication. Methods: Evaluation of the program in the first year examined program participation, staff feedback following education workshops and mindfulness sessions, staff retention rates, and the results of an annual organizational staff survey and a program outcome survey. Results: Approximately 76% of staff attended the Introduction to Well-being workshop, and 98% of responses to survey questions were positive. Staff also provided positive feedback on the other well-being workshops and sessions embedded within existing education programs. Employee Assistance Program counseling sessions had an 81% uptake, with a wide variety of presenting issues, 62% related to work. All participants in mindfulness sessions agreed that it was a valuable tool to improve clinical practice, 94% said it had an immediate positive impact on their well-being, and 70% agreed that they were applying mindfulness principles outside the sessions. Staff retention and turnover improved. Staff reported a positive effect on awareness of self-care, addressing risks to resilience, seeking support from trusted colleagues, coping with critical incidents, and the ability to interact positively with patients and families. Conclusion: The evaluation showed a positive impact on staff well-being. Although there was a wide variety of successful interventions reported in the literature, sustainability needs to be considered. Feedback on this program found that staff appreciated being listened to, valued, and supported through the strategies, and the ongoing program will continue to monitor staff needs and be responsive in building their resilience and well-being.


Purpose: The Quality of Care Collaborative Australia (QuoCCA) provided pediatric palliative care education across Australia with the aim of improving the quality of services. The education was delivered through a collaboration of six tertiary pediatric palliative care services, through funding for Nurse Educators, Medical Fellows, a National Allied Health Educator, and national project staff.

Methods: Pre- and post-education surveys were completed by participants immediately following the education, and confidence and knowledge were measured along nine domains related to the care of the child and family, including managing a new referral, symptom management, medications, preparing the family, and using local agencies. Results: Education was provided to over 5,500 health and human service professionals in 337 education sessions across Australia between May 2015 and June 2017. Paired pre- and post-surveys were completed by 969 participants and showed a significant improvement in all the domains measured. Those with no experience in caring for children receiving palliative care showed greater improvement following QuoCCA education compared to those with experience, although the latter had higher scores both before and after education. Similarly, those with no previous education showed greater improvement, but those with previous education showed higher scores overall. Participants in full-day and half-day sessions showed greater improvement than those in short day sessions. Thus, the dosage of education in the length of the sessions and prior attendance impacted knowledge and confidence. Topics requested by the participants were analyzed. Educator learnings were that education was more effective when tailored to the needs of the audience, was interactive, and included story-telling, case studies, and parent experiences. Conclusion: These results encouraged the continuation of the provision of education to novice and experienced professionals who care for children with a life-limiting condition, leading to higher levels of confidence and knowledge. The learnings from this evaluation will be transferred into the second round of funding for the national QuoCCA education project. The next stage will focus on developing simulation and interactive training, accessible training modules, and videos on a national website.


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.


BACKGROUND: In the UK, rates of neonatal post-mortem (PM) are low. Consent for PM is required, and all parents should have the opportunity to discuss whether to have a post-mortem examination of their baby. OBJECTIVES: We aimed to explore neonatal healthcare professionals' experiences, knowledge, and views regarding the consent process for post-mortem examination after neonatal death. METHOD: An online survey of neonatal healthcare providers in the UK was conducted. Responses from 103 healthcare professionals were analysed, 84 of whom were doctors. The response rate of the British Association of Perinatal Medicine (BAPM) members was 11.7%. RESULTS: Perceived barriers to PM included cultural and religious practices of parents as well as a lack of rapport between parents and professionals. Of the respondents, 69.4% had observed a PM; these professionals had improved satisfaction with their training and confidence in counselling (p < 0.001 and p < 0.001) but not knowledge of the procedure (p = 0.77). Healthcare professionals reported conservative estimates of the likelihood that a PM would identify significant information regarding the cause of death. CONCLUSIONS: Confidence of neonatal staff in counselling could be improved by observing a PM. Training for staff in developing a rapport with parents and addressing emotional distress may also overcome significant barriers to consent for PM.


The purpose of this study was to analyze the impact of interprofessional pediatric end-of-life simulations for health professions students. A quasiexperimental design was used with three TeamSTEPPS(R) tools. Forty-one students were enrolled (nursing = 20, medicine = 10, pharmacy = 10, public health = 1). TeamSTEPPS 2.0 Teamwork Attitudes Questionnaire and Teamwork Perceptions Questionnaire analysis indicated a significant difference in mean pretest and posttest scores (p = .015 and p = .028, respectively). The Team Performance Observation Tool indicated statistical significance between simulations (p < .001, df = 18, r = .8). Simulations were significantly related to an increase in faculty observation scores, TeamSTEPPS 2.0 Teamwork Attitudes Questionnaire pre-post scores, and TeamSTEPPS 2.0 Teamwork Perceptions Questionnaire pre-post scores.


AIM: Children and adolescents with end-stage renal disease face a high morbidity and mortality. Palliative care provides a multidisciplinary approach to reduce disease burden and improve quality of life. This study evaluated concepts and current structures of palliative care from the perspective of a multidisciplinary paediatric nephrology team including physicians, nurses and psychosocial health professionals. METHODS: Evaluation was done by an online survey sent to the members of the German Society of Nephrology and to the nurse managers of German paediatric dialysis centres between April 9, 2018 and May 31, 2018. RESULTS: Out of the 52 respondents, 54% were physicians, 21% nurses and 25% psychosocial health professionals. The quality of actual palliative care service was rated as moderate (3.3 on a scale from one to six). Specialised palliative care teams (54%) and the caring paediatric nephrologist (50%) were considered as primarily responsible for palliative care. Two thirds wished for training in palliative care. In only 15% of the respondents' centres, palliative care specialisation existed. CONCLUSION: Palliative care structures in paediatric nephrology were not sufficient in the view of the multidisciplinary healthcare team. Therefore, efforts should be taken to integrate palliative care into the routine treatment of children and adolescents with chronic kidney diseases.


BACKGROUND: In Pediatric Intensive Care Unit (PICU) two types of population require the intervention of neuropediatricians (NP): chronic brain diseases' patients who face repetitive and prolonged hospitalizations, and patients with acute brain failure facing the risk of potential neurologic sequelae, and both conditions may result in a limitation of life-sustaining treatments (LLST) decision. OBJECTIVE: To assess NP's involvement in LLST decisions within the PICU of a tertiary hospital. METHOD: Retrospective study of medical reports of patients hospitalized during 2014 in the Necker-Hospital PICU. Patients were selected using keywords ("cardiorespiratory arrest", "death", "withdrawal of treatment", "palliative care", "acute brain failure", or "chronic neurological disease"), and/or if they were assessed by a NP during the hospitalization. Demographic and medical data were analysed, including the NP's assessment and data about Collaborative Multidisciplinary Deliberation (CMD) to discuss potential LLST. RESULTS: Among 1160 children, 274 patients were included and 142 (56%) were assessed by a NP during their hospitalization for diagnosis (n = 55) and/or treatment (n = 95) management. NP was required for 59%-100% of patients with neurological acute failure, and for 14-44% of patients with extra neurological failure. A LLST decision was taken after a CMD for 27 (9.8%) of them, and a NP was involved in 19/27 (70%) of these decisions that occurred during the hospitalization (n = 19) or before (n = 8).12 patients died thereafter the LLST decision (40% of the 30 dead patients). CONCLUSION: NP are clearly involved in the decision-process of LLST for patients admitted in
PICU, claiming for close collaboration to improve current practices and the quality of the care provided to children.  


CONTEXT: Implementation of pediatric palliative care as a primary practice relevant for all pediatricians and pediatric subspecialists requires a grounding, shared knowledge. This study reports on the innovative application of a monthly Palliative Care E-Journal Club (Pal Care Club) to foster shared palliative care knowledge hospital wide. OBJECTIVES: To explore the impact of a monthly electronic journal club to increase the number of palliative care-relevant articles read and discussed and to enhance provider comfort with the integration and introduction of palliative care. METHODS: A single cohort, predesign-post-design was utilized to explore the impact of a monthly palliative care electronic journal club. RESULTS: Preintervention barriers to reading pediatric palliative care literature were primarily access and time. The mean of paired differences (post-pre) for the number of full-text articles read per month was 2.56 (SD = 1.25). The journal club intervention increased participant personal comfort with integrating palliative care principles at the bedside (p < 0.0001) and introducing pediatric palliative care to patients and families (p < 0.0001). CONCLUSION: An electronic journal club is a feasible and acceptable means of increasing number of palliative care articles read and discussed across an institution as well as enhancing pediatric palliative care knowledge across subspecialist and general pediatric services.  


Palliative care competencies at the pediatric resident training level expand learned knowledge into behavior. The objective of this study was to investigate mode of palliative care education delivery preferred by pediatric residents and to report on participatory approach to resident palliative care curriculum design. A one-hour monthly palliative care curriculum was designed and implemented in a participatory manner with 20 pediatric residents at a free-standing Midwestern children's hospital. Outcome measures included pediatric residents' personal attitude and perceived training environment receptivity before and after implementation of a palliative care competency-based curriculum. An 18-item survey utilizing Social Cognitive Theory Constructs was administered at baseline and after palliative care curriculum implementation (2017(-)2018 curricular year). Pediatric residents prioritized real case discussions in group format (16/20) over other learning formats. Topics of highest interest at baseline were: discussing prognosis and delivering bad news (weighted average 12.9), pain control (12.3), goals of care to include code status (11.1), and
integrative therapies (10.7). Summary of ordinal responses revealed improvement in self-assessment of personal attitude toward palliative care and training environment receptivity to palliative care domains after year-long curriculum implementation. Curricular approach which is attentive to pediatric residents' preferred learning format and self-assessment of their behaviors within their care setting environment may be beneficial in competency-based primary palliative training.


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.


INTRODUCTION: Surgical residents often need to break bad news (BBN) to patients and family members. While communication skills are a core competency in residency training, these specific skills are rarely formally taught. We piloted a simulation training to teach pediatric surgical residents how to compassionately BBN of an unexpected, traumatic pediatric death to surviving family members. This training was unique in that it was influenced by family systems theory and was a collaborative effort between our institution’s surgery residency and medical family therapy (MedFT) programs. METHOD: This study provides outcomes of surgery residents' communication
skills, attitudes, and self-perceptions after a BBN simulation activity with standardized family members at a major academic teaching hospital. Each resident participated in two 30-min simulations and received feedback from observers. Outcome data were collected through self-assessments completed before, immediately after, and 6 months after the simulation. Participants were 15 surgery residents, and MedFT students served as simulated family members and trainers.

RESULTS: A statistically significant change with medium to large effect sizes in participant self-reported perceptions of skill and confidence were documented and maintained over 6 months. Responses to open-ended questions supported practice changes in response to the training.

DISCUSSION: This collaborative training promoted significant improvement in resident compassionate communication skills. The curriculum was highly valued by the learners and resulted in sustained application of learned skills with patients and families. Our novel approach was feasible with promising results that warrant further investigation and could be reproduced in other institutions with similar programs. (PsycINFO Database Record (c) 2018 APA, all rights reserved).


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 pre-implementation 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.


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.


BACKGROUND:: Lack of education and training in palliative care has been described to be one of the most important barriers to pediatric palliative care implementation. OBJECTIVE:: To examine what factors determine the degree of knowledge and level of comfort Mexican pediatricians have providing pediatric palliative care. METHODS:: A questionnaire that assessed palliative care concepts was developed and applied online to Mexican pediatricians, both generalists and specialists. RESULTS:: A total of 242 pediatricians responded. The majority had not received palliative care education (92.6%) and felt uncomfortable discussing palliative needs with patients and families (92.1%). The mean score of the questionnaire was 6.8 (+/-1.4) of 10 correct answers. Knowledge in palliative care was associated with exposure to oncologic patients ( P = .01) and previous palliative care education ( P = .02) but inversely related to the pediatrician’s age ( P = .01). Comfort addressing patient’s palliative care needs was associated with knowledge in palliative care ( P < .01), exposure to oncologic patients ( P = .03), and previous education in palliative care ( P = .02). CONCLUSIONS:: Although Mexican pediatricians have basic knowledge of palliative care concepts, they do not feel comfortable addressing palliative care needs, suggesting that the main barrier for implementing palliative care is not the lack of knowledge but rather feeling uncomfortable when addressing these issues with patients and families. Educational programs should incorporate strategies that could help physicians develop comfort in approaching palliative care patients.

Epidemiology and Pathology


Multiple sclerosis (MS) is an inflammatory idiopathic autoimmune disease causing demyelination of central nervous system (CNS). The incidence of pediatric MS is relatively rare, affecting 0.2 to 0.64/100,000 subjects; cases with MS onset before age 10-12 years, account for less than 1% of all MS cases, while 2.7 to 10.5% of all MS cases worldwide are seen in children <18 years of age, with a strong female preponderance. The disease course of MS varies from a benign type with relatively low level of disability after a long duration (15 years) of the disease, to a malignant type of MS with severe disability or even death within few months following onset. Diagnostic criteria for pediatric MS include >/= 2 clinical events involving more areas of CNS inflammation in the absence of encephalopathy, separated by > 30 days, along with the involvement of brainstem. Pediatric MS generally presents relapsing-remitting form of MS, with majority of the patients recovering from the first attack. Major histocompatibility complex, more specifically, mutations in the human leukocyte antigen (HLA) DRB1*15 allele, are considered most important genetic factors that are contributory to the disease. Treatment choices for pediatric MS include many disease-modifying therapies (DMT) that are currently being used for adult MS and these are interferon-beta 1a/1b (IFN-beta1a/1b), glatiramer acetate, teriflunomide, dimethyl fumarate, alemtuzumab, etc. However, most of these have not gone through complete testing in randomized, placebo-controlled clinical trials for pediatric MS and are being prescribed off-label by clinicians. As these studies are progressing, it is important to address if these approaches of treating pediatric MS patients have any long-term impact on patients, in particular, physical, cognitive, developmental and social outcomes of the children.


INTRODUCTION: Advances in paediatric oncology led to the increase in long-term survival, revealing the burden of therapy-related long-term side effects. We evaluated overall and cause-specific mortality in a large cohort of Italian childhood cancer survivors (CCSs) and adolescent cancer survivors identified through the off-therapy registry. MATERIALS AND METHODS: CCSs alive 5 years after cancer diagnosis occurring between 1960 and 1999 were eligible; the last follow-up was between 2011 and 2014. Outcomes were reported as standardised mortality ratios (SMRs) and absolute excess risks (AERs). RESULTS: Among 12,214 CCSs, 1113 (9.1%) deaths occurred. Survival at 35 years since diagnosis was 87% (95% confidence interval [CI]: 86-88) and at 45 years was 81% (95% CI: 77-84). CCSs had an 11-fold increased risk of death (SMR 95% CI: 10.7-12),
corresponding to an AER of 48 (95% CI: 45-51). Mortality decreased by 60% for survivors treated most recently (1990-1999). The most frequent causes of death were recurrence of the original cancer (56%), a subsequent neoplasm (19%) and cardiovascular diseases (5.8%). Among those who survived at least 15 years after diagnosis, a secondary malignancy was the leading cause of death.

CONCLUSIONS: This study confirms the impact of recent advances in anticancer therapy in reducing mortality, mainly attributable to recurrence but also to other causes. However, overall mortality continues to be higher than in the general population. A long-term follow-up is needed to prevent late mortality due to secondary neoplasms and non-neoplastic causes in CCSs.


BACKGROUND: Krabbe disease is a rare neurological disorder caused by a deficiency in the lysosomal enzyme, beta-galactocerebrosidase, resulting in demyelination of the central and peripheral nervous systems. If left without treatment, Krabbe disease results in progressive neurodegeneration with reduced quality of life and early death. The purpose of this prospective study was to describe the natural progression of early onset Krabbe disease in a large cohort of patients.

METHODS: Patients with early onset Krabbe disease were prospectively evaluated between 1999 and 2018. Data sources included diagnostic testing, parent questionnaires, standardized multidisciplinary neurodevelopmental assessments, and neuroradiological and neurophysiological tests. RESULTS: We evaluated 88 children with onset between 0 and 5 months. Median age of symptom onset was 4 months; median time to diagnosis after onset was 3 months. The most common initial symptoms were irritability, feeding difficulties, appendicular spasticity, and developmental delay. Other prevalent symptoms included axial hypotonia, abnormal deep tendon reflexes, constipation, abnormal pupillary response, scoliosis, loss of head control, and dysautonomia. Results of nerve conduction studies showed that 100% of patients developed peripheral neuropathy by 6 months of age. Median galactocerebrosidase enzyme activity was 0.05 nmol/h/mg protein. The median survival was 2 years. CONCLUSIONS: This is the largest prospective natural history study of Krabbe disease. It provides a comprehensive description of the disease during the first 2 years of life. With recent inclusion of state mandated newborn screening programs and promising therapeutic interventions, enhancing our understanding of disease progression in early onset Krabbe disease will be critical for developing treatments, designing clinical trials, and evaluating outcomes.


OBJECTIVE: To describe the long-term prognosis of childhood epilepsy, with special emphasis on seizure remission, relapse, medication, associated neurologic impairment, mortality rate, and cause of death. METHODS: A prospective longitudinal study on a population-based total cohort of 195 children with epileptic seizures in 1962-1964. Data were collected from medical records and a questionnaire. RESULTS: Follow-up data from 94% of the initial cohort showed the best long-term prognosis for seizure freedom for children with no intellectual or neurologic impairment. These
children had later seizure onset, shorter total duration of epilepsy, and were more often medication free. Only a few of them had isolated relapses. Generalized, rather than focal, epilepsy was associated with fewer relapses and less ongoing medication. The “true incidence” group, with onsets during the inclusion period of 1962-1964, had the best long-term prognosis for seizure freedom, with 90% seizure-free after 50 years. Although only 10% of this group had ongoing seizures at follow-up, 22% still used anticonvulsant medication, often with old drugs, that is, phenobarbital or phenytoin, as one of the anticonvulsant drugs. The standardized mortality ratio (SMR) was 2.61 for the whole group, with no difference between those with or without other neurodeficits. Those who died young either had neurologic impairment or died from epilepsy-related conditions; later deaths often followed non-epilepsy-related conditions. No one in the incidence group died of SUDEP (sudden unexpected death in epilepsy). SIGNIFICANCE: This 50-year, long-term follow-up of a cohort of persons with childhood epilepsy in general demonstrates a better outcome for seizure freedom compared to our follow-up after 12 years and to previous reports. We also report a low incidence of seizure relapses. Remission of seizures does not automatically lead to termination of medication. The mortality rate associated with SUDEP was lower than previously reported.


Spinal muscular atrophy (SMA) is an autosomal recessive neuromuscular disorder. The infantile form is the most common genetic cause of infantile death due to respiratory insufficiency. The disorder is caused by the premature death of motor neurons of anterior horn, leading to progressive weakness and muscular atrophy. Longtime considered as untreatable, the pathology knew a real revolution during the last two years. Views on this terrible disease have completely changed, changing, therefore, the management of the patients and constituting new challenges. https://www.ncbi.nlm.nih.gov/pubmed/30793560


INTRODUCTION: Lafora disease is autosomal recessive progressive myoclonus epilepsy with late childhood-to teenage-onset caused by loss-of-function mutations in either EPM2A or EPM2B genes encoding laforin or malin, respectively. DEVELOPMENT: The main symptoms of Lafora disease, which worsen progressively, are: myoclonus, occipital seizures, generalized tonic-clonic seizures, cognitive decline, neuropsychiatric symptoms and ataxia with a fatal outcome. Pathologically, Lafora disease is characterized by the presence of polyglucosans deposits (named Lafora bodies), in the brain, liver, muscle and sweat glands. Diagnosis of Lafora disease is made through clinical, electrophysiological, histological and genetic findings. Currently, there is no treatment to cure or prevent the development of the disease. Traditionally, antiepileptic drugs are used for the management of myoclonus and seizures. However, patients become drug-resistant after the initial stage. CONCLUSIONS: Lafora disease is a rare pathology that has serious consequences for patients and their caregivers despite its low prevalence. Therefore, continuing research in order to clarify the underlying mechanisms and hopefully developing new palliative and curative treatments for the disease is necessary. https://www.ncbi.nlm.nih.gov/pubmed/30638256

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


AIM: To assess the long-term natural course of early-onset facioscapulohumeral dystrophy (FSHD), which is important for patient management and trial-readiness, and is currently lacking. METHODS: We had the unique opportunity to evaluate 10 patients with early-onset FSHD after 22 years follow-up. Patients underwent a semi-structured interview, physical examination and additional genotyping. RESULTS: Nine initial study participants (median age 37 years) were included, one patient died shortly after first publication. At first examination, one patient was wheelchair dependent, one patient walked aided, and eight patients walked unaided. After 22 years, four patients were wheelchair dependent, three walked aided, and two walked unaided. Systemic features, including hearing loss (56%), intellectual disability (44%), and a decreased respiratory function (56%), were frequent. Patients participated socially and economically with most patients living in a regular house (n = 6) and/or having a paid job (n = 4). DISCUSSION: Patients with early-onset FSHD generally had a severe phenotype compared to classical onset FSHD. However, after 22 years of follow up they showed a wide variation in severity and, despite these physical limitations, participated socially and economically. These observations are important for patient management and should be taken into account in clinical trials.


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% of 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.


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.


Importance: The United States has higher infant and youth mortality rates than other high-income countries, with striking disparities by racial/ethnic group. Understanding changing trends by age and race/ethnicity for leading causes of death is imperative for focused intervention. Objective: To estimate trends in US infant and youth mortality rates from 1999 to 2015 by age group and race/ethnicity, identify leading causes of death, and compare mortality rates with Canada and England/Wales. Design, Setting, and Participants: This descriptive study analyzed death certificate data from the US National Center for Health Statistics, Statistics Canada, and the UK Office of National Statistics for all deaths among individuals younger than 25 years. The study took place from January 1, 1999, to December 31, 2015, and analyses started in September 2017. Exposures: Race/ethnicity. Main Outcomes and Measures: Average annual percent changes in mortality rates from 1999 to 2015 and absolute rate change between 1999 to 2002 and 2012 to 2015 for each age group, race/ethnicity, and cause of death. Results: Among individuals from birth to age 24 years, 1169537 deaths occurred in the United States, 80540 in Canada, and 121183 in England/Wales from 1999 to 2015. In the United States, 64% of deaths occurred in male individuals and 52.6% occurred in white individuals (25.1% deaths occurred in black individuals and 17.9% in Latino individuals). All-cause mortality declined for all age groups (infants younger than 1 year [38.5% of deaths], children aged 1-9 years [10.6%], early adolescents aged 10-14 years [5%], late adolescents aged 15-19 years [17.7%], and young adults aged 20-24 years [28.1%]) in the United States, Canada, and England/Wales from 1999 to 2015. However, rates were highest in the United States. Within the United States, annual declines in all-cause mortality rates occurred among all age groups of black, Latino, and white individuals, except for white individuals aged 20 to 24 years, whose rates remained stable. Mortality rates declined across most major causes of death from 1999 to 2002 and 2012 to 2015, with notable declines observed for sudden infant death syndrome, unintentional injury death, and homicides. Among infants, unintentional suffocation and strangulation in bed increased (difference between 2012-2015 and 1999-2002 range, 6.11-29.03
Further, suicide rates among Latino and white individuals aged 10 to 24 years (range, 0.21-2.63 per 100000) and black individuals aged 10 to 19 years (range, 0.10-0.45 per 100000) increased, as did unintentional injury deaths in white young adults (0.79 per 100000). The rise in unintentional injury deaths is attributed to increases in drug poisonings and was also observed in black and Latino young adults. Conclusions and Relevance: Mortality rates in the United States have generally declined for infants and youths from 1999 to 2015 owing to reductions in sudden infant death syndrome, unintentional injury death, and homicides. However, US mortality rates remain higher than Canada and England/Wales, with particularly elevated rates among black and American Indian/Alaskan Native youth. Further, there is a concerning increase in suicide and drug poisoning death rates among US adolescents and young adults.


Whole or partial trisomy of the short arm of chromosome 9 (9p) is considered to be one of the more frequent chromosome abnormalities compatible with life. The duplication may affect various organs, however the most common symptoms are certain specific facial dysmorphisms and abnormalities of the fingers, toes and nails. A one month old boy presented with failure to thrive, jaundice, ventricular septal defect (VSD) and dysmorphic face. He displayed symptoms of heart failure. The cardiologic examination revealed a significant VSD, hypoplasia of the aortic arch, pulmonary hypertension, decompensated circulatory failure and moderate left ventricle dysfunction. Routine cytogenetic analysis revealed a supernumerary marker chromosome. Fluorescence in situ hybridization (FISH) identified this as the short arm of chromosome 9. The child’s karyotype was determined as 47,XY,+der(9)dup(9)(p10p24)dn. Due to his worsening condition and the high risk of the operation, it was decided to forego the procedure. After a short palliative care the child passed away. The child’s clinical presentation and the uncharacteristic severity of his condition show that chromosome abnormalities involving duplicated genetic material are extremely heterogeneous. Thus treatment of each child should be individualized and may also involve difficult ethical considerations. Orv Hetil. 2018; 159(47): 1994-2000.


BACKGROUND: Niemann-Pick disease Type C (NP-C) is a lysosomal lipid storage disorder characterized by progressive neurodegenerative symptomatology. The signs and symptoms of NP-C vary with age at disease onset, and available therapies are directed at alleviating symptoms and
stabilizing disease progression. We report the characteristics and factors related to disease progression, and analyze the effect of miglustat treatment on disease progression and patient survival using NP-C disability scales. METHODS: This retrospective, observational chart review included patients with NP-C from five expert NP-C centers. Patient disability scores were recorded using three published NP-C disability scales, and a unified disability scale was developed to allow comparison of data from each scale. Disease progression was represented by scores on the unified NP-C disability scale. Patients were stratified as infantile (< 4 years), juvenile (>/= 4 - < 16 years), and adult (>/= 16 years) based on age at diagnosis, and treated >/=1 year and non-treated/treated < 1 year based on the duration of miglustat treatment. RESULTS: The analysis included 63 patients; the majority (61.9%) were on miglustat therapy for >/=1 year. Ataxia and clumsiness/frequent fall were the most common neurologic symptoms across age groups, whereas, hypotonia and delayed developmental milestones were specific to infantile patients. In both infantile and juvenile patients, visceral signs preceded diagnosis and neurologic signs were noted at or shortly after diagnosis. Adult patients presented with a range of visceral, neurologic, and psychiatric signs in years preceding diagnosis. Patients on miglustat therapy for >/=1 year had a lower mean annual disease progression compared with those untreated/treated < 1 year (1.32 vs 3.54 points/year). A significant reduction in annual disease progression in infantile patients, and a trend towards reduced disease progression in juvenile patients after >/=1 year of miglustat treatment, translated into higher age at last contact or death in these groups. CONCLUSIONS: The type and onset of symptoms varied across age groups and were consistent with descriptions of NP-C within the literature. Miglustat treatment was associated with a reduced rate of disability score worsening in infantile and juvenile patients, both in agreement with increased age at last contact.


OBJECTIVES: Place of death (POD) is considered a key quality indicator for adult end of-life care, but paediatric evidence is limited. Data from Child Death Overview Panel (CDOP) databases provides an opportunity to describe trends in POD as regional paediatric palliative medicine (PPM) options have increased. Aims were to identify and describe trends in POD for children in South Yorkshire. METHODS: Retrospective cohort study. Anonymised data extracted from five CDOP databases 2008-2015. Data included age, gender, ethnicity, postcode (outward code only), POD, classification and category of death. Descriptive statistical analysis using chi(2) test was used to assess intergroup differences. RESULTS: 748 deaths were notified from 2008 to 2015. Neonatal deaths were excluded, 46% (n=345). Of non-neonatal deaths (n=403), 58% (n=232) were 'expected'. Of expected deaths (n=232), 19% (n=45) died in home, 19% (n=45) died in hospice and 61% (n=141) died in hospital. This was significantly different from comparable national data which showed considerably more hospital deaths. There was no significant change in POD over time. CONCLUSION: Hospital remains the POD for most children, whether deaths are 'expected' or not, suggesting specialised PPM should be expanded into the hospital setting. More research is needed regarding preference for POD. This study may help inform future service planning for PPM and hospice development.

https://www.ncbi.nlm.nih.gov/pubmed/30446488
BACKGROUND: Population trends of disease prevalence and incidence over time measure burden of disease and inform healthcare planning. Neuromuscular disorders (NMD) affect muscle and nerve function with varying degrees of severity and disease progression. OBJECTIVE: Using health administrative databases we described trends in incidence, prevalence, and mortality of adults and children with NMD. We also explored place of death and use of palliative care. METHODS: Population-based (Ontario, Canada) cohort study (2003 to 2014) of adults and children with NMD identified using International Classification of Disease and health insurance billing codes within administrative health databases. RESULTS: Adult disease prevalence increased on average per year by 8% (95% confidence interval (CI) 6% to 10%, P < .001), with the largest increase in adults 18-39 years. Childhood disease prevalence increased by 10% (95% CI 8% to 11%, P < .0001) per year, with the largest increase in children 0 to 5 years. Prevalence increased across all diagnoses except amyotrophic lateral sclerosis and spinal muscular atrophy for adults and all diagnoses for children. Adult incidence decreased by 3% (95% CI -4% to -2%, P < .0001) but incidence remained stable in children. Death occurred in 34,336 (18.5%) adults; 21,236 (61.8%) of whom received palliative care. Death occurred in 1,009 (5.6%) children; 507 (50.2%) of whom received palliative care. Mortality decreased over time in adults (odds ratio (OR) 0.86, 95% CI 0.86-0.87, P < .0001) and children (OR 0.79, 95% CI 0.76-0.82, P < .0001). Use of palliative care over time increased for adults (OR 1.18, 95% CI 1.09 to 1.28, P < .0001) and children (OR 1.22, 95% CI 1.20 to 1.23, P < .0001).

CONCLUSIONS: In both adults and children, NMD prevalence is rising and mortality rates are declining. In adults incidence is decreasing while in children it remains stable. This confirms on a population-based level the increased survival of children and adults with NMD.


Inborn errors of metabolism (IEMs) are particularly frequent as diseases of the nervous system. In the pediatric neurologic presentations of IEMs neurodevelopment is constantly disturbed and in fact, as far as biochemistry is involved, any kind of monogenic disease can become an IEM. Clinical features are very diverse and may present as a neurodevelopmental disorder (antenatal or late-onset), as well as an intermittent, a fixed chronic, or a progressive and late-onset neurodegenerative disorder. This also occurs within the same disorder in which a continuum spectrum of severity is frequently observed. In general, the small molecule defects have screening metabolic markers and many are treatable. By contrast only a few complex molecules defects have metabolic markers and most of them are not treatable so far. Recent molecular techniques have considerably contributed in the description of many new diseases and unexpected phenotypes. This paper provides a comprehensive list of IEMs that affect neurodevelopment and may also present with neurodegeneration.

AIM: to communicate HPN data obtained from the HPN registry of the NADYA-SENPE group (www.nadya-senpe.com) for the year 2017. MATERIAL AND METHODS: descriptive analysis of the data collected from adult and pediatric patients with HPN in the NADYA-SENPE group registry from January 1st, 2017 to December 31st, 2017. RESULTS: there were 308 patients from 45 Spanish hospitals (54.5% women), 38 children and 270 adults, with 3,012 episodes, which represent a prevalence rate of 6.61 patients/million inhabitants/year 2017. The most frequent diagnosis in adults was "palliative cancer" (25.6%), followed by "others". In children, it was Hirschsprung's disease with six cases (15.8%). The first indication was short bowel syndrome in both children (55.3%) and adults (33.7%). The most frequently used type of catheter was tunneled in both children (73.4%) and adults (38.2%). Ending 81 episodes, the most frequent cause was death (62.9%) and transition to oral feeding (34.7%). CONCLUSIONS: the progressive increase of collaborating centers and professionals in the registry of patients receiving NPD is maintained. The main indications of HPN and the motive for ending have remained stable.


Autophagy is a tightly modulated lysosomal degradation pathway. Genetic disorders of autophagy during nervous system development may lead to developmental delay, neurodegeneration, and other neurological signs in children. Here we aimed to summarize single gene disorders that perturb various steps of autophagy pathway and their roles in the causation of childhood neurological diseases. Numerous childhood-onset disorders are caused by mutations that impact the autophagy pathway. These can manifest with a range of features including ataxia, spastic paraplegia, and intellectual disability. Defective proteins causing such diseases can interfere with autophagy flux at different stages of the itinerary. Defective autophagy may be an important contributor to the pathological features of various childhood neurodegenerative diseases and lead to the accumulation of aberrant protein and dysfunctional organelles. Insights into the relevant cell biological processes may help understand pathophysiological mechanisms and inspire autophagy-restoring therapeutic approaches. WHAT THIS PAPER ADDS: Numerous childhood-onset disorders
are caused by mutations that impact the autophagy pathway. Defective autophagy is a feature of some mutations that cause ataxia, spastic paraplegia, and intellectual disability.

Outcomes and Instruments


PURPOSE: Enterococci are a common cause of bacteremia in immunocompromised patients. Although the increase of vancomycin-resistant enterococci (VRE) makes appropriate antibiotic therapy difficult, clinical characteristics of enterococcal bacteremia and the impact of VRE infection on outcomes have rarely been reported in immunocompromised children. METHODS: We enrolled children and adolescents (< 19 years of age) with underlying malignancies who were diagnosed with enterococcal bacteremia during febrile neutropenia between 2010 and 2017. Medical records of the enrolled children were retrospectively reviewed to evaluate the clinical characteristics of enterococcal bacteremia and impact of VRE infection on outcomes. RESULTS: Thirty-six episodes of enterococcal bacteremia were identified in 30 patients. VRE infection was identified in 11 episodes (30.6%); the 7- and 30-day mortalities were 27.8% and 44.4%, respectively. Acute lymphoblastic leukemia (50.0%) and acute myeloid leukemia (30.6%) were the most common underlying disorders. Three (8.3%) of the patients were in complete remission, and palliative and reinduction chemotherapies were administered in 47.2% and 36.1% of episodes, respectively. Empirical antibiotic therapy was appropriate in 64.0% of patients with vancomycin-susceptible enterococcal infection and in none of the VRE-infected patients (p = 0.001). However, the 30-day mortality was not significantly different between the two patient groups (44.0% vs. 45.5%, p = 1.000). CONCLUSIONS: Most episodes of enterococcal bacteremia occurred in advanced stages of underlying malignancies, and still showed high mortality. The prognosis seemed to be related to the underlying disease condition rather than vancomycin resistance of the isolated enterococci, although the number of enrolled patients was small.


Niemann-Pick disease, type C (NPC) is a neurodegenerative lysosomal storage disease affecting the visceral organs and the central nervous system. The age of initial presentation varies from fetal to adult onset, although childhood onset is most common. The life expectancy for the full spectrum of NPC patients is not well defined, and it is unknown if current supportive care impacts the natural history. In order to assess age of death for a large cohort of NPC patients, we "crowd-sourced" age and year of death from information posted on disease support group website.
memorial walls. We analyzed data from 338 individuals who died between 1968 and 2018. In addition to age of death, gender can be inferred from given names and photographs. The median age of death was 13 years with a range from 0.1-69 years. Although sex significantly affects survival of NPC1 mutant mice, we did not observe a gender dependent survival difference in NPC patients. Median age of survival across time increased between the earliest patients and the most recently deceased patient; however, we found no significant change in survival over the last 20 years. These data suggest that supportive medical care has not impacted survival in the recent past and provides support for the use of historic controls in evaluating therapeutic interventions.


In 2006, PAIN published a systematic review of the measurement properties of self-report pain intensity measures in children and adolescents (Stinson JN, Kavanagh T, Yamada J, Gill N, Stevens B. Systematic review of the psychometric properties, interpretability and feasibility of self-report pain intensity measures for use in clinical trials in children and adolescents. PAIN 2006;125:143-57). Key developments in pediatric pain necessitate an update of this work, most notably growing use of the 11-point numeric rating scale (NRS-11). Our aim was to review the measurement properties of single-item self-report pain intensity measures in children 3 to 18 years old. A secondary aim was to develop evidence-based recommendations for measurement of child and adolescent self-report of acute, postoperative, and chronic pain. Methodological quality and sufficiency of measurement properties for reliability, validity, responsiveness, and interpretability was assessed by at least 2 investigators using CONsensus based Standards for the selection of health Measurement INstruments (COSMIN). Searches identified 60 unique self-report measures, of which 8 (reported in 80 papers) met inclusion criteria. Well-established measures included the NRS-11, Color Analogue Scale (CAS), Faces Pain Scale-Revised (FPS-R; and original FPS), Pieces of Hurt, Oucher-Photographic and Numeric scales, Visual Analogue Scale, and Wong-Baker FACES Pain Rating Scale (FACES). Quality of studies ranged from poor to excellent and generally reported sufficient criterion and construct validity, and responsiveness, with variable reliability. Content and cross-cultural validity were minimally assessed. Based on available evidence, the NRS-11, FPS-R, and CAS were strongly recommended for self-report of acute pain. Only weak recommendations could be made for self-report measures for postoperative and chronic pain. No measures were recommended for children younger than 6 years, identifying a need for further measurement refinement in this age range. Clinical practice and future research implications are discussed.

https://www.ncbi.nlm.nih.gov/pubmed/30180088


OBJECTIVE: This study examines health care provider perspectives about barriers to pediatric palliative care for seriously ill children 15 years after an initial study within the same academic health system. METHODS: Anonymous validated surveys were sent electronically to inpatient
nursing unit distribution lists \(n = 1315\). Reminders were sent through e-mail twice over a two-month data collection period. RESULTS: Response rate was 20.9\% \(275/1315\) with 45.2\% of responses from critical care units and 21.6\% from hematology/oncology units. Of the participants, 58.2\% \(n = 160\) had \(\geq 10\) years nursing experience, 58.5\% \(n = 161\) had one to five patients die in the past 12 months, and 50.2\% \(n = 138\) had one to five patients receiving subspecialty pediatric palliative care in the past year. Approximately one-half of the participants reported 3 of 26 barriers listed on the study survey as frequently or almost always occurring, including (1) family preference for more life-sustaining treatment than staff \(n = 177, 64.8\%\), (2) family not ready to acknowledge incurable condition \(n = 175, 64.1\%\), and (3) parent discomfort with possibility of hastening death \(n = 146, 53.7\%\). Study findings were similar between 2002 and 2017, particularly in the extremes of the most and least commonly cited barriers. CONCLUSIONS: Barriers to palliative care for hospitalized children persist and commonly include perceptions that families deny, prefer, or have discomfort with forgoing life-sustaining treatments. Increasingly, studies have shown that families can be simultaneously hopeful and aware of their child’s worsening health. Further palliative care education and research about these barriers and their impacts are necessary to support seriously ill children and their families.


CONTEXT: Do-not-resuscitate (DNR) orders are common among children receiving palliative care, who may nevertheless benefit from surgery and other procedures. Although anesthesia, surgery, and pediatric guidelines recommend systematic reconsideration of DNR orders in the perioperative period, data regarding how clinicians evaluate and manage DNR orders in the perioperative period are limited. OBJECTIVES: To evaluate perioperative management of DNR orders at a tertiary care children's hospital. METHODS: We reviewed electronic medical records for all children with DNR orders in place within 30 days of surgery at a tertiary care pediatric hospital from February 1, 2016, to August 1, 2017. Using standardized case report forms, we abstracted the following from physician notes: 1) patient/family wishes with respect to the DNR, 2) whether preoperative DNR orders were continued, modified, or suspended during the perioperative period, and 3) whether life-threatening events occurred in the perioperative period. Based on data from these reports, we created a process flow diagram regarding DNR order decision-making in the perioperative period. RESULTS: Twenty-three patients aged six days to 17 years had a DNR order in place within 30 days of 29 procedures. No documented systematic reconsideration took place for 41\% of procedures. DNR orders were modified for two (7\%) procedures and suspended for 15 (51\%). Three children (13\%) suffered life-threatening events. We identified four time points in the perioperative period where systematic reconsideration should be documented in the medical record, and identified recommended personnel involved and important discussion points at each time point. CONCLUSION: Opportunities exist to improve how DNR orders are managed during the perioperative period.

Epilepsy is a common pediatric neurological condition, and approximately one-third of children with epilepsy are refractory to medical management. For these children, neurosurgery may be indicated, but operative success is dependent on complete delineation of the epileptogenic zone. In this review, surgical techniques for pediatric epilepsy are considered. First, potentially-curative operations are discussed and broadly divided into resections and disconnections. Then, two palliative approaches to seizure control are reviewed. Finally, future neurosurgical approaches to epilepsy are considered.


OBJECTIVE: In paediatric cardiopulmonary arrest, International Liaison Committee on Resuscitation (ILCOR) states, 'there are no simple guidelines to determine when resuscitative efforts become futile'. Considerations to assist this decision-making include cause of arrest, pre-existing medical conditions, age, site of arrest, duration of untreated cardiopulmonary arrest, witnessed arrest and presence of shockable rhythm. Outcomes are poor in out-of-hospital cardiac arrests (OHCA), particularly for infants. This single-centre observational study describes the characteristics and outcomes of the subgroup of children presenting to our hospital's ED following OHCA still receiving cardiac compressions, to assist development of guidelines for future resuscitation efforts in our ED, particularly for cessation of cardiopulmonary resuscitation (CPR). METHODS: The ED database was searched for children presenting in cardiopulmonary arrest receiving cardiac compressions. Data were reviewed on pre-hospital, ED and hospital management and outcome, particularly looking at considerations outlined by ILCOR. RESULTS: From January 2000 to December 2013, 60 children were identified: median age 1.71 years; 87% arresting at home; 68% with bystander CPR; median CPR duration pre-hospital 42 min, and in ED 19.5 min; total CPR median 61 min. Fifty patients (83%) died in ED, 10 (17%) were admitted to intensive care but all died within 4 days. CONCLUSION: Children presenting to ED still receiving cardiac compressions following OHCA had a universally poor outcome, regardless of age and underlying cause. This implies resuscitative efforts could be discontinued earlier in this subgroup. A national, multicentre study is needed to determine if this finding is reproducible with a larger population.


OBJECTIVES: For children, adolescents, and young adults with complex chronic conditions advance care planning may be a vital component of optimal care. Advance care planning outcomes research has previously focused on seriously ill adults and adolescents with cancer where it is correlated with high-quality end-of-life care. The impact of advance care planning on end-of-life...
outcomes for children, adolescents, and young adults with complex chronic conditions is unknown, thus we sought to evaluate parental preferences for advance care planning and to determine whether advance care planning and assessment of specific family considerations during advance care planning were associated with differences in parent-reported end-of-life outcomes. DESIGN: Cross-sectional survey. SETTING: Large, tertiary care children's hospital. SUBJECTS: Bereaved parents of children, adolescents, and young adults with complex chronic conditions who died between 2006 and 2015. INTERVENTIONS: None. MEASUREMENT AND MAIN RESULTS: One-hundred fourteen parents were enrolled (54% response rate) and all parents reported that advance care planning was important, with a majority (70%) endorsing that discussions should occur early in the illness course. Parents who reported advance care planning (65%) were more likely to be prepared for their child’s last days of life (adjusted odds ratio, 3.78; 95% CI, 1.33-10.77), to have the ability to plan their child's location of death (adjusted odds ratio, 2.93; 95% CI, 1.06-8.07), and to rate their child’s quality of life during end-of-life as good to excellent (adjusted odds ratio, 3.59; 95% CI, 1.23-10.37). Notably, advance care planning which included specific assessment of family goals was associated with a decrease in reported child suffering at end-of-life (adjusted odds ratio, 0.23; 95% CI, 0.06-0.86) and parental decisional regret (adjusted odds ratio, 0.42; 95% CI, 0.02-0.87). CONCLUSIONS: Parents of children, adolescents, and young adults with complex chronic conditions highly value advance care planning, early in the illness course. Importantly, advance care planning is associated with improved parent-reported end-of-life outcomes for this population including superior quality of life. Further studies should evaluate strategies to ensure high-quality advance care planning including specific assessment of family goals.


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.

reliability of the revised Face, Legs, Activity, Cry, and Consolability (r-FLACC) scale ratings."

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. Interrater 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 THIS 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.

Outcomes in Pediatric Palliative Care: A Systematic Review." Pediatrics 143(1).

CONTEXT: Pediatric palliative care (PPC) is intended to promote children's quality of life by using a
family-centered approach. However, the measurement of this multidimensional outcome remains
challenging. OBJECTIVE: To review the instruments used to assess the impact of PPC interventions. DATA SOURCES: Five databases (Embase, Scopus, The Cochrane Library, PsychInfo, Medline) were searched. STUDY SELECTION: Inclusion criteria were as follows: definition of PPC used; patients aged 0 to 18 years; diseases listed in the directory of life-limiting diseases; results based on empirical data; and combined descriptions of a PPC intervention, its outcomes, and a measurement instrument. DATA EXTRACTION: Full-text articles were assessed and data were extracted by 2 independent researchers, and each discrepancy was resolved through consensus. The quality of the studies was assessed by using the Standard Quality Assessment Criteria for Evaluating Primary Research Papers From a Variety of Fields checklist. RESULTS: Nineteen of 2150 articles met the eligibility criteria. Researchers in 15 used quantitative methods, and 9 were of moderate quality. Multidimensional outcomes included health-related quality of life, spiritual well-being, satisfaction with care and/or communication, perceived social support, and family involvement in treatment or place-of-care preferences. PPC interventions ranged from home-based to hospital and respite care. Only 15 instruments (of 23 reported) revealed some psychometric properties, and only 5 included patient-reported (child) outcome measures. LIMITATIONS: We had no access to the developmental process of the instruments used to present the underlying concepts that were underpinning the constructs. CONCLUSIONS: Data on the psychometric properties of instruments used to assess the impact of PPC interventions were scarce. Children are not systematically involved in reporting outcomes. https://www.ncbi.nlm.nih.gov/pubmed/30530504


BACKGROUND:: Few studies have analyzed the benefit of limb amputations in children with metastatic osteosarcoma and limited life span. OBJECTIVE:: We studied outcomes of limb amputations in children with metastatic osteosarcoma. DESIGN:: We performed a retrospective review of patients who underwent limb amputations (January 1995-June 2015) and died within 1 year of surgery. SETTING/PARTICIPANTS:: We studied 12 patients with osteosarcoma at a single institution. MEASUREMENTS:: Data on mobility, pain, and emotional and psychological well-being were retrieved from medical records from 1 month before surgery to 6 months after surgery. RESULTS:: Of the 12 patients (7 females and 5 males; median age at surgery 13 years [range, 7-20 years]) meeting study criteria, 3 patients and 9 patients had primary osteosarcoma in upper and lower limbs, respectively. Mobility improved postamputation in 8 bedridden/wheelchair-bound patients. Postamputation, emotional, and psychological well-being improved for 9 patients, 3 patients had persistent psychological and/or emotional symptoms, and no patient experienced signs of regret. Daily mean pain scores were significantly lower at 1 week (median 3 [range, 0-6]; P = .03) and 3 months (median 0 [range, 0-8]; P = .02) postsurgery than at 1 week presurgery (median 5.5 [range, 0-10]). Morphine consumption (mg/kg/d) showed a trend toward higher values at 1 week (median 0.2 [range, 0-7.6]; P = .6) and 3 months (median 0.2 [range, 0-0.5]; P = .3) postsurgery than at 1 week presurgery (median 0.1 [range, 0-0.5]). CONCLUSIONS:: Patients undergoing limb amputations had reduced pain and improved mobility and emotional and psychological well-being. Amputations are likely to benefit children with limited life expectancy. https://www.ncbi.nlm.nih.gov/pubmed/30058346
PURPOSE: Emergent palliative radiation therapy (PRT) of symptomatic metastases can significantly increase the quality of life of patients with cancer. In some contexts, this treatment may be underused, but in others PRT may represent an excessively aggressive intervention. The characterization of the current use of emergent PRT is warranted for optimized value and patient-centered care.

METHODS AND MATERIALS: This study is a cross-sectional retrospective analysis of all emergent PRT courses at a single academic tertiary institution across 1 year. RESULTS: A total of 214 patients received a total of 238 treatment courses. The most common indications were bone (39%) and brain (14%) metastases. Compared with outpatients, inpatients had lower mean survival rates (2 months vs 6 months; P < .001), higher rates of stopping treatment early (19.1% vs 9.0%; P = .034), and greater involvement of palliative care (44.8% vs 24.1%; P < .001), but the same mean planned fractions (9.10 vs 9.40 fractions; P = .669). In a multiple predictor survival analysis, palliative care involvement (P = .025), male sex (P = .001), ending treatment early (P = .011), and having 1 of 3 serious indications (airway compromise, leptomeningeal disease, and superior/inferior vena cava involvement; P = .007) were significantly associated with worse overall survival. CONCLUSIONS: Survival is particularly poor in patients who receive emergent PRT, and patient characteristics such as functional status and indication should be considered when determining fractionation schedule and dosing. A multi-institutional study of practice patterns and outcomes is warranted.


PURPOSE: Status Epilepticus can be a serious life threatening event in epileptic patients. The definition of refractory or super-refractory Status Epilepticus was based on the therapeutic response to anti-epileptic and anesthetic drugs. Vagal Nerve Stimulation showed efficacy in treating drug-resistant epilepsy but there are only few reports on emergent placement of Vagal Nerve Stimulator for refractory or super-refractory Status Epilepticus. METHODS: Among 49 children implanted at our Institution with Vagal Nerve Stimulation for drug-resistant epilepsy, the authors retrospectively identified those implanted for refractory or super-refractory Status Epilepticus, according with the current definitions. RESULTS: 4 patients were operated upon at ages ranging 7 to 17 months and reached the programmed output current of 1 mA over a time ranging from 24 to 36 h (fast ramping-up). In 3 out of 4 patient we observed the abrupt of Status Epilepticus; one patient was refractory both to drugs and Vagal Nerve Stimulation and later died, without recovering from SE. At follow up, ranging from 24 to 45 months, the remaining 3 patients showed a decrease of the seizures frequency >80% without relapse of Status Epilepticus; in all the patients, output current and/or Duty Cycle were increased later. CONCLUSION: VNS can be effective in treating refractory or super-refractory Status Epilepticus.

BACKGROUND: Aim of the study was to analyze the association of shock index (SI) from 0 to 6 hours with early mortality in severe sepsis/septic shock and to explore its age-specific cut-off values. To investigate association of change in SI over first 6 hours with early mortality. METHODS: A prospective cohort study of children (<14 years) admitted in emergency department, tertiary care hospital with severe sepsis or septic shock, divided into 3 groups: group 1: 1 month to <1 year; group 2: 1 to <6 years; group 3: 6 to 12 years. Shock index (SI = heart rate/systolic blood pressure) measured at admission (X0) and hourly till 6 hours (X1-6). Primary outcome was death within 48 hours of admission. Area under receiver operating characteristic curves were constructed for SI (0-6). Optimal cut-offs of SI 0 and SI 6, maximizing both sensitivity and specificity were determined and positive and negative predictive values (PPV, NPV) were calculated. RESULTS: From 2015 to 2016, 120 children were recruited. Septic shock was present at admission in 56.7% children. Early mortality was 50%. All hourly shock indices (SI 0-6) were higher among non survivors in group 2 (P <= .03) and group 3 (P < .001). In group 1, SI after 2 hours was higher in non survivors (P 2-6: <= .02). Area under receiver operating characteristic curves (95% CI) for SI at 0 hour was 0.72 (0.5-0.9), 0.66 (0.5-0.8), and 0.77 (0.6-0.9) and at 6 hours was 0.8 (0.6-1), 0.75 (0.6-0.9), and 0.8 (0.7-1) in 3 groups. The cut-off values of SI 0 (sensitivity; specificity; PPV; NPV) in 3 groups: 1.98 (77; 75; 67; 83), 1.50 (65; 65; 68; 63), and 1.25 (90; 67; 77; 83) and SI6: 1.66 (85; 80; 73; 89), 1.36 (73; 70; 73; 70), and 1.30 (74; 73; 78; 69). Improvement of SI over 6 hours was associated with better outcome. Children with higher SI at both time points had higher mortality than those with SI score below the cut-offs (P = .001). CONCLUSIONS: Age-specific SI cut-off values may identify children at high risk of early mortality in severe sepsis/septic shock and allow for better targeted management.


Mitochondrial neurogastrointestinal encephalomyopathy syndrome is a rare autosomal recessive multisystem disorder caused by nuclear TYMP gene mutations, which leads to deficiency in thymidine phosphorylase enzyme. This deficiency then leads to mitochondrial dysfunction, which causes the features characteristic of this syndrome, including severe muscle wasting, gastrointestinal dysmotility, leukoencephalopathy, peripheral neuropathy, and ophthalmoplegia. Here, we present a case series of 3 patients with mitochondrial neurogastrointestinal encephalomyopathy from Saudi Arabia who underwent allogeneic stem cell transplant at King Faisal Specialist Hospital (Riyadh, Saudi Arabia). Two patients died within the first year of transplant, and the third is still alive but without improvement in clinical features. Allogeneic hematopoietic stem cell transplant-related mortality appears to be high; this may at least be partially related to established end-organ effects with decreased performance status. Although allogeneic hematopoietic stem cell transplant clearly affects correction of genetic and biochemical defects in mitochondrial neurogastrointestinal encephalomyopathy, its ability to reverse or improve established clinical manifestations has not been proven.
BACKGROUND:: There is a lack of appropriate, validated person-centred outcome measures (PCOM) for paediatric palliative care in the scientific literature, and as a result there is not a tool to drive and evaluate care of children and young people. METHODS:: In line with COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN) guidance, an expert group was convened to elicit views on the domains/items to include in a PCOM, implementation challenges and requirements for use in routine care by practitioners. Data were content analysed. RESULTS:: 36 UK-wide clinicians, advocates, and researchers participated. 1) Items included were: specific symptoms, education, play and social interaction, parental time for partner and other children, sex and intimacy, and sibling wellbeing. 2) Implementation challenges: supporting children and young people to engage meaningfully, that the instrument could be seen as a 'test' of parents' care quality, raising unrealistic expectations, proxy validity. 3) There is a need for clear administration and interpretation guidance and for data ownership/access to be agreed. CONCLUSIONS:: This expert meeting addressed the initial step in COSMIN guidance, informing face validity and acceptability. It provides the information necessary for the first phase of tool development and informs potential use and implementation.


OBJECTIVE: Due to the unique importance of parental and sibling relationships and concurrently existing developmental challenges, the loss of a parent or sibling due to cancer is a highly stressful event for children and adolescents. This is the first systematic review that integrates findings on psychosocial outcomes after parental or sibling cancer bereavement. METHODS: A systematic search of Web of Science, PubMed, PsycINFO, and PubPsych was conducted, last in December 2017. Quantitative studies on psychosocial outcomes of children and adolescents who lost a parent or sibling due to cancer were included. RESULTS: Twenty-four studies (N = 10 parental and N = 14 sibling bereavement), based on 13 projects, were included. Ten projects had cross-sectional designs. Only 2 projects used large, population-based samples and nonbereaved comparison groups. Outcomes were partially measured by single-item questions. Bereaved children and adolescents showed similar levels of depression and anxiety compared with nonbereaved or norms. Severe behavioral problems were found rarely. However, in 2 large, population-based studies, about half of the bereaved individuals reported unresolved grief. Bereaved adolescents had a higher risk for self-injury compared with the general population in one large, population-based study. Communication with health-care professionals, family, and other people; social support; distress during illness; age; gender; and time because loss were associated with psychosocial bereavement outcomes. CONCLUSIONS: Results indicate a high level of adjustment in cancer-bereaved children and adolescents. A modifiable risk factor for adverse psychosocial consequences is poor communication. Prospective designs, representative samples, and validated instruments, eg, for prolonged grief, are suggested for future research.

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.


Introduction: Pediatric palliative care (PPC) seeks longitudinal relationships with patients facing life-threatening conditions. The majority of pediatric deaths occur within the first year of life, especially neonatal intensive care unit (NICU); however, the consultation by PPC in the NICU is not routine. This project sought to improve the PPC’s presence within 1 NICU for patients facing life-limiting conditions through quality improvement techniques. Methods: A trigger list of severe, life-threatening conditions impacting neonates was created and implemented to increase PPC consultation within the NICU. Interventions to improve compliance with the trigger list included

the collaborative creation of the trigger list, education, modification of PPC staff modeling, and expansion of the perinatal palliative care program. Results: Over the 2 years that the project occurred, 31 prenatal and postnatal patients were eligible for PPC consultation based on the trigger list. Of these, 24 received PPC consultation. The primary outcome measure of the project was to increase PPC consultations for those NICU infants identified on a severe diagnosis "trigger" list from 25% to 80% and to maintain this increase for 6 months. This project achieved 100% compliance within 12 months. Conclusions: Utilization of quality improvement methodology to address PPC underutilization within an NICU successfully led to the implementation of a trigger list for patients with severe diagnoses to receive PPC services. Such modeling could be used in other health systems to improve palliative care referrals. 


PURPOSE: Intensity of end-of-life care receives much attention in oncology because of concerns that high-intensity care is inconsistent with patient goals, leads to worse caregiver outcomes, and is expensive. Little is known about such care in those undergoing allogeneic hematopoietic cell transplantation (HCT), a population at high risk for morbidity and mortality. PATIENTS AND METHODS: We conducted a population-based analysis of patients who died between 2000 and 2013, within 1 year of undergoing an inpatient allogeneic HCT using California administrative data. Previously validated markers of intensity were examined and included: hospital death, intensive care unit (ICU) admission, and procedures such as intubation and cardiopulmonary resuscitation at end of life. Multivariable logistic regression models determined clinical and sociodemographic factors associated with: hospital death, a medically intense intervention (ICU admission, cardiopulmonary resuscitation, hemodialysis, intubation), and >/= two intensity markers. RESULTS: Of the 2,135 patients in the study population, 377 were pediatric patients (age </= 21 years), 461 were young adults (age 22 to 39 years), and 1,297 were adults (age >/= 40 years). The most common intensity markers were: hospital death (83%), ICU admission (49%), and intubation (45%). Medical intensity varied according to age, underlying diagnosis, and presence of comorbidities at time of HCT. Patients with higher-intensity end-of-life care included patients age 15 to 21 years and 30 to 59 years, patients with acute lymphoblastic leukemia, and those with comorbidities at time of HCT. CONCLUSION: Patients dying within 1 year of inpatient allogeneic HCT are receiving medically intense end-of-life care with variations related to age, underlying diagnosis, and presence of comorbidities at time of HCT. Future studies need to determine if these patterns are consistent with patient and family goals. 


Outcomes of neonatal intensive care unit (NICU) graduates have been categorized by rates of neurodevelopmental impairment at 2 years old. Although useful as metrics for research, these early childhood assessments may underestimate or overestimate later functional capabilities. Often overlooked are less severe but more prevalent neurobehavioral dysfunctions seen later in
childhood, and chronic health concerns that may impact the child's quality of life (QoL). Comprehensive NICU follow-up should include measures of less severe cognitive/learning delays, physical/mental well-being, and the promotion of resilience in children and families. Studies are needed to identify QoL measures that will optimize children’s assessments and outcomes.


BACKGROUND: End-of-life (EOL) care in neonatal intensive care units (NICUs) can vary depending on religious beliefs of health care providers and families as well as the sociocultural environment. Although guidelines exist for EOL care in NICUs, most are based on Western studies, and little is known about such care in Asian countries, which have different religious and social background. OBJECTIVE: This review synthesized empirical research to reveal the state of the science on infant EOL care in Asian countries. DESIGN: This was an integrative review. SETTING/SUBJECTS: Data were collected from studies identified in CINAHL, Embase, PsycINFO, and PubMed. The search was limited to current empirical studies involving infant EOL care in Asian countries and published in English between 2007 and 2016. RESULTS: Of 286 studies initially identified, 11 empirical studies conducted in Hong Kong, India, Israel, Japan, Mongolia, Taiwan, and Turkey were included in the review. Four themes were captured: factors influencing decision making, trends in decision making, practical aspects of EOL care, and health care providers' preparation. In most NICUs, health care providers controlled decisions regarding use of life-sustaining treatment, with parents participating in decision making no more than 60% of the time. Although care decisions were gradually changing from "do everything" for patient survival to a more palliative approach, comfort care at the EOL was chosen no more than 63% of the time. CONCLUSION: While infant EOL care practice and research vary by country, few articles address these matters in Asia. This integrative review characterizes infant EOL care in Asia and explores cultural influences on such care.


We analyzed treatment outcomes and prognostic factors in adult patients with therapy-related myeloid neoplasms (t-MNs) to select patients who would be benefited by active anticancer treatment. After excluding 18 patients who received palliative care only and 13 patients with acute promyelocytic leukemia, 72 t-MN patients (45 with acute myeloid leukemia and 27 with myelodysplastic syndrome) were retrospectively evaluated. Among them, 10 (13.9%), 32 (44.4%), and 30 patients (41.7%) had favorable, intermediate- and adverse-risk cytogenetics, respectively. Among patients with intermediate-risk cytogenetics, patients with a normal karyotype (NK; N = 20) showed superior allogeneic stem cell transplantation-censored overall survival (AC-OS) and OS compared to those with non-NK-intermediate-risk cytogenetics (P < 0.001). In the multivariate analysis, male sex, age >/= 70 years, and unfavorable cytogenetics (non-NK-intermediate plus adverse risk cytogenetics) were associated with inferior AC-OS. Those results suggest that a more-
refined subdivision of risk stratification would be necessary in patients with intermediate-risk cytogenetics.


Among 131 children admitted to our institution for early phase rehabilitation after freshwater near-drowning (ND) between the year 1986 and 2000, 87 were in unresponsive wakefulness syndrome (UWS) for at least 4 weeks after the accidents. An anonymous questionnaire was sent to the families after 0.5 to 15.0 years (median: 4.6) and 48 mothers and 51 fathers of 55 of these 87 children were interviewed after 6.6 to 23.8 years (median: 13.8) of ND. At the time of the interviews, 8/55 children were able to perform daily living activities independently, 36/55 children were not able to do so (many of them suffered from chronic medical conditions like spasticity or disorders of swallowing), and 11/55 children had died. Health-related quality of life (HRQoL) was, however, similar to the normal population for mothers, and even higher for fathers. Furthermore, the ND accident had apparently not lead to a higher rate of separations of parents but had increased their likelihood to have further children. Feelings of guilt were highly prevalent (23/47 mothers, 20/47 fathers), and correlated with lower HRQoL of the respective parent. We found correlations between duty of supervision and feelings of guilt and between outcome and HRQoL for only the fathers. In conclusion, we found that after 4 weeks in UWS, the long-term neurological outcome of pediatric ND victims is often but not always poor. Despite often severe disabilities or death of the child during long-term care, parents surprisingly report little impact on their HRQoL, on the stability of their partnership or on their wish to have further children. Our findings may help parents and physicians to choose the best treatment for a child in UWS due to different etiologies striking the balance between rehabilitation and palliative care.


OBJECTIVE: To assess the health-related quality of life of children with neurofibromatosis type 1-related plexiform neurofibromas (pNF) using a battery of patient-reported outcome measures selected based on a conceptual framework derived from input by patients, parents, and clinicians regarding the most important pNF symptoms and concerns. STUDY DESIGN: There were 140 children with pNF ages 8-17 years who completed the Patient-Reported Outcomes Measurement Information System (including domains anxiety, depressive symptom, psychosocial stress experiences, fatigue, pain interference, meaning and purpose, positive affect, peer relationships, physical function-mobility) and Quality of Life in Neurological Disorders measurement system (stigma) via an online platform. T-scores for each measure were compared with US population norms. RESULTS: Children with pNF reported significantly worse scores than the population norms on 8 of 10 domains. Children with at least 1 family member having a diagnosis of
neurofibromatosis type 1 and those having pain reported significantly worse symptoms and functioning on all domains. Boys reported significantly worse pain interference, stigma, meaning and purpose, mobility function, and upper extremity function than girls. CONCLUSIONS: Children with pNF experience significantly worse health-related quality of life on all but 1 domain, highlighting the importance of monitoring children’s quality of life over time in clinical research and practice. Future research should evaluate the replicability of these findings and evaluate the validity of the Patient-Reported Outcomes Measurement Information System and Quality of Life in Neurological Disorders measurement system in relation to clinical characteristics among children with pNF.


Childhood Lymphoblastic leukemia’s (ALL) early mortality (EM) is an undesirable treatment outcome for a disease for which >90% long term success is achievable. In the Western world EM constitutes no >3%; yet, in Chiapas, Mexico, remains around 15%. With the objective of improving on EM, we determined associated elements in 28 ALL who died within 60 days of arriving at Hospital de Especialidades Pediatricas in Chiapas (HEP), by comparing them to those in 84 controls who lived beyond the first 90 days. chi, t test, and binary logistic regression (BLR) were used to determine significant individual and multiple variables associated to outcome. On arrival, fever, liver and spleen enlargement, active bleeding, lower albumin, less platelets, higher creatinine, and uric acid, more diploid and less hyperdiploid cases were associated with EM cases. Time to diagnosis, nutritional status, risk group and leukocyte count were not related. Antileukemic treatment approach was similar in both groups. The BLR model including fever, active bleeding, liver enlargement, <10,000 platelets/microL, and >2X upper normal lactic dehydrogenase, determined outcome in 66.7% EM and 90.2% controls. To improve on EM in ALL, patients with characteristics defined here ought to be treated differently at HEP.

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BACKGROUND: Primary dystonia is a neurologic disease with characteristics of abnormal, involuntary twisting and turning movements, which greatly affect quality of life of patients. Treatments for dystonia consist of oral medications, botulinum neurotoxin injections, physical therapy, and surgery. For medication-refractory dystonia, surgery, especially deep brain stimulation (DBS), is the optimal option. CASE DESCRIPTION: The patient was a 13-year-old boy suffering from extremely severe primary dystonia, with a Burke-Fahn-Marsden Dystonia Rating Scale-motor score of 118 and a Toronto Western Spasmodic Torticollis Rating Scale-severity score of 29. The examination of 173 genes, including DYT, failed to identify any abnormality. He responded ineffectively to medications. After both bilateral subthalamic nucleus DBS and unilateral thalamic lesion in ventralis intermedius nucleus and ventralis oralis nucleus (Vim-Vo thalamotomy), his movement disorder improved dramatically. Four and 7 months after the operation, the scores of 2 rating scales sharply decreased. Potential brain structural changes were reflected in sensorimotor-
related cortical thickness, surface area, and gray matter volume from magnetic resonance imaging, which may reveal a valid method to evaluate surgical effect on the brain with enough patients.

CONCLUSIONS: DBS and thalamotomy is potentially an effective combination of treatments for severe medication-refractory dystonia.


BACKGROUND:: Although most children at end of life have commercial insurance, little is known about their demographic and clinical characteristics, what care they are receiving, and how much it costs. OBJECTIVES:: To describe commercially insured children who enrolled in hospice care during their last year of life and to examine differences across age-groups. METHODS:: A retrospective cohort study was conducted using 2005 to 2014 data from the MarketScan Commercial Claims and Encounters database from Truven Health Analytics. Variables were created for demographics, health, utilization, and spending. Analyses included chi(2) and analysis of variance tests of differences. RESULTS:: Among the 17 062 children who utilized hospice, 49% had a preferred provider organization (PPO). Hospice length of stay averaged less than 5 days. Over 80% of children visited their primary care physician. Eight percent had hospital readmissions, and 38% had emergency department (ED) visits. Average expenditures were US$3686 per month or US$44 232 annually. The most common condition for children less than 1 year was cardiovascular (21.96%). Neuromuscular conditions were the most frequent (7.89%) in children aged 1 to 5 years, while malignancies (10.53% and 11.32%, respectively) were prevalent in ages 6 to 14 and 15 to 17. Children less than 1 year had the highest frequency of hospital readmissions (16.25%) with the lowest ED visits (28.67%) while incurring the highest expenses (US$11 211/month).

CONCLUSIONS:: The findings suggest that commercially insured children, who enroll in hospice, have flexible coverage with a PPO. Hospital readmissions and ED visits were relatively low for a population who was seriously ill. There were significant age-group differences.


Neonatal Intensive Care Units (NICU) provide special equipment designed to give life support for the increasing number of prematurely born infants and assure their survival. More recently NICU's strive to include developmentally oriented care and modulate sensory input for preterm infants. Music, among other sensory stimuli, has been introduced into NICUs, but without knowledge on the basic music processing in the brain of preterm infants. In this study, we explored the cortico-subcortical music processing of different types of conditions (Original music, Tempo modification, Key transposition) in newborns shortly after birth to assess the effective connectivity of the primary auditory cortex with the entire newborn brain. Additionally, we investigated if early exposure during NICU stay modulates brain processing of music in preterm infants at term equivalent age. We approached these two questions using Psychophysiological Interaction (PPI) analyses. A group
of preterm infants listened to music (Original music) starting from 33 weeks postconceptional age until term equivalent age and were compared to two additional groups without music intervention; preterm infants and full-term newborns. Auditory cortex functional connectivity with cerebral regions known to be implicated in tempo and familiarity processing were identified only for preterm infants with music training in the NICU. Increased connectivity between auditory cortices and thalamus and dorsal striatum may not only reflect their sensitivity to the known music and the processing of its tempo as familiar, but these results are also compatible with the hypothesis that the previously listened music induces a more arousing and pleasant state. Our results suggest that music exposure in NICU's environment can induce brain functional connectivity changes that are associated with music processing.


Several children receiving palliative care experience dyspnea and pain. An order protocol for distress (OPD) is available at Sainte-Justine Hospital, aimed at alleviating respiratory distress, pain and anxiety in pediatric palliative care patients. This study evaluates the clinical use of the OPD at Sainte-Justine Hospital, through a retrospective chart review of all patients for whom the OPD was prescribed between September 2009 and September 2012. Effectiveness of the OPD was assessed using chart documentation of the patient’s symptoms, or the modified Borg scale. Safety of the OPD was evaluated by measuring the time between administration of the first medication and the patient’s death, and clinical evolution of the patient as recorded in the chart. One hundred and four (104) patients were included in the study. The OPD was administered at least once to 78 (75%) patients. A total of 350 episodes of administration occurred, mainly for respiratory distress (89%). Relief was provided in 90% of cases. The interval between administration of the first protocol and death was 17 h; the interval was longer in children with cancer compared to other illnesses (p = 0.02). Data from this study support the effectiveness and safety of using an OPD for children receiving palliative care.


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


BACKGROUND: Spinal muscular atrophy (SMA) is an autosomal-recessive neuromuscular disorder resulting in progressive muscle weakness. In December 2016, the U.S. Food and Drug Administration approved the first treatment for SMA, a drug named nusinersen (Spinraza) that is administered intrathecally. However many children with SMA have neuromuscular scoliosis or spinal instrumentation resulting in challenging intrathecal access. Therefore alternative routes must be considered in these complex patients. OBJECTIVE: To investigate routes of drug access, we reviewed our institutional experience of administering intrathecal nusinersen in all children with spinal muscular atrophy regardless of spinal anatomy or instrumentation. MATERIALS AND METHODS: We reviewed children with SMA who were referred for intrathecal nusinersen injections from March to December 2017 at our institution. In select children with spinal hardware, spinal imaging was requested to facilitate pre-procedure planning. Standard equipment for intrathecal injections was utilized. All children were followed up by their referring neurologist. RESULTS: A total of 104 intrathecal nusinersen injections were performed in 26 children with 100% technical success. Sixty procedures were performed without pre-procedural imaging and via standard interspinous technique. The remaining 44 procedures were performed in 11 complex (i.e. neuromuscular scoliosis or spinal instrumentation) patients requiring pre-procedural imaging for planning purposes. Nineteen of the 44 complex procedures were performed via standard interspinous technique from L2 to S1. Twenty-two of the 44 complex procedures were performed using a neural-foraminal approach from L3 to L5. Three of the 44 complex procedures were performed via cervical puncture technique. There were no immediate or long-term complications but there was one child with short-term complications of meningismus and back pain at the injection site. CONCLUSION: Although we achieved 100% technical success in intrathecal nusinersen administration, our practices evolved during the course of this study. As a result of our early experience we developed an algorithm to assist in promoting safe and effective nusinersen administration in children with spinal muscular atrophy regardless of SMA type, abnormal spinal anatomy and complex spinal instrumentation.


OBJECTIVE: To describe the perceptions of caregivers of children with medical complexity (CMC) about their decision to pursue tracheostomy for their children, in particular the satisfaction with their decision. STUDY DESIGN: In this qualitative study conducted in western North Carolina
between 2013 and 2014, we interviewed 56 caregivers of 41 CMC who had received tracheostomies in the past 5 years. Three of the CMC were deceased at the time of the interview; 8 were decannulated. In-depth interviews (35 English, 6 Spanish) were conducted, audio-recorded, and transcribed verbatim. We used ATLAS.ti software to manage data and identified themes related to caregiver perceptions about tracheostomy decision. RESULTS: We found that caregivers often chose tracheostomy because extending the lives of their children and being able to care for them at home were important. Caregivers reported the many benefits of tracheostomy including improvement in respiratory symptoms, physical and developmental health, quality of life, and means to provide medical care quickly when needed. There were negative effects of tracheostomy such as mucus plugs, excessive secretions, accidental decannulation necessitating emergency tracheostomy tube change, and the increased infection risk. Providing medical care for CMC with tracheostomy at home was difficult, but improved over time. Caregivers were generally satisfied with their decision to pursue tracheostomy for their CMC. CONCLUSIONS: Decisional satisfaction with tracheostomy for CMC is high. In counseling caregivers about tracheostomy, clinicians should present both the benefits and risks. Future studies should quantify the outcomes described in this study.


BACKGROUND: The design and provision of quality pediatric palliative care should prioritize issues that matter to children and their families for optimal outcomes. OBJECTIVE: This review aims to identify symptoms, concerns and outcomes that matter to children and young people (“young people”) with terminal illnesses and their families. Findings from the systematic review will inform the development of a relevant framework of health outcomes. METHOD: This is a systematic literature review across multiple databases for identification of eligible primary evidence. Data sources included PsycINFO, MEDLINE, Embase, CINAHL, OpenGrey, and Science Direct Journals, searched from 1 August 2016 to 30 July 2017. The study also incorporates consultations with experts in the field, citation searches via Scopus, and a hand search of reference lists of included studies. RESULTS: Of the 13,567 articles that were evaluated, 81 studies were included. Most of these studies (n = 68) were from high-income countries and focused on young people with cancer (n = 58). A total of 3236 young people, 2103 family carers, 108 families, and 901 healthcare providers were included in the studies. Young people did not contribute to data in 30% of studies. Themes on priority concerns are presented by the following domains and health outcomes: (1) physical (n = 62 studies), e.g., physical symptoms; (2) psychological (n = 65), e.g., worry; (3) psychosocial (n = 31), e.g., relationships; (4) existential (n = 37), e.g., existential loss; and (5) “other” (n = 39), e.g., information access. CONCLUSION: Burdensome symptoms and concerns affect young people with malignant and nonmalignant conditions and occur across the disease trajectory; pediatric palliative care should not be limited to the end-of-life phase. A child-family-centered framework of health outcomes, spanning the patient, family, and quality of service levels is proposed to inform service development. Future research should address gaps identified across the literature (i.e., the involvement of young people in research, evidence for developing countries, and a focus on nonmalignant conditions.

Objective: Identification of palliative care needs in patients with liver cirrhosis using the MELD/Na score and the Child-Pugh score. Materials and Methods: A retrospective study of hospitalized patients with hepatic cirrhosis between January 2015 and December 2016 using the Child-Pugh score and the MELD/Na score in January 2018. Results: Recognizing end-of-life patients (the past 12 months of life) is a challenge for health professionals, especially in diseases with poorly defined criteria, such as cirrhosis of the liver. The verification of rapid functional decline and health indicators can be verified using already defined scales such as the Child-Pugh score and the MELD/Na score. Patients were classified according to the Child-Pugh score in Class A (17%), Class B (48.9%), and Class C (34%). The corresponding survival rate was as follows: class A (87.5%), Class B (30.4%), and Class C (31.25%). The MELD/Na score intervals were >9 (2.15%), score 10-19 (46.8%), score 20-29 (27.7%), score 30-40 (19.1%), and score >40 (4.3%). Nearly 51.1% had a MELD/Na score >20 and 48.9% <20. The study revealed that 59.6% of patients died before 12 months. They were end-of-life patients who needed palliative care to reduce the impact of the disease. Conclusions: The Child-Pugh score and the MELD/Na score represent a viable and easy-to-use tool to identify patients in need of palliative care, among those with liver cirrhosis. Early identification, timely evaluation, and effective treatment of physical, spiritual, family, and social problems improve the quality of life of people with incurable diseases and their families.


OBJECTIVE: To investigate the effectiveness of vagus nerve stimulation (VNS) in combination with pharmacological therapy in a longitudinal retrospective study at a single center. MATERIALS AND METHODS: Data from 130 consecutive patients implanted with a VNS device between the years 2000 and 2013 was analyzed. Seizure frequency and pharmacological antiepileptic drug (AED) treatments were recorded prior to as well as at one, two, and five years after VNS implantation. RESULTS: Median age at epilepsy onset was five years and mean years from diagnosis to VNS implantation was 16.5 years. There was a significant seizure reduction overall (all p < 0.001). The responder (>/=50% seizure frequency reduction) rate increased from 22.1 to 43.8% between the first and fifth year for the cohort as a whole, with the largest increase between the first and second year (22.1-38.1%) and regardless of AED changes. VNS effectiveness did not differ between patients who altered or remained on the same AEDs. Patients were treated with a median of three AEDs throughout the study and the number of AEDs significantly increased after two (p = 0.007) and five (p = 0.001) years. CONCLUSIONS: VNS is a well-tolerated palliative neuromodulatory treatment for drug resistant epilepsy with a 43.8% seizure reduction after five years. Our data supports the idea that VNS effectiveness increases with time. Therefore we suggest that VNS should be evaluated for at least two years after implantation. AED changes should try to be kept to a minimum during evaluation in order to determine the effectiveness of VNS.
BACKGROUND: Pediatric palliative care programs aim to improve the quality of life of children with severe life-threatening illnesses, and that of their families. Although rehabilitation and physical therapy provides a valuable tool for the control of symptoms, it has been poorly researched to date. Since the family represents such a fundamental support in these cases, it is important to deepen our understanding regarding the value of implementing rehabilitation programs from the parents' perspective. AIM: The aim of this paper was to explore parents' experiences regarding the implementation of a physical rehabilitation program in pediatric palliative care. DESIGN: A qualitative methodology was chosen. SETTING: The unit of pediatric palliative care at the Hospital Nino Jesus (Madrid, Spain). POPULATION: The inclusion criteria were: a) parents of children, irrespective of their diagnosis, b) integrated within the program of palliative care at the time of study, c) aged between 0-18 years, c) must be receiving Home-Based Rehabilitation Program by the Pediatric Palliative Care team. Fourteen parents were included. METHODS: Purposeful sampling method was implemented. Data collection consisted of unstructured and semi-structured interviews. A thematic analysis was performed to interpret transcripts. Guidelines for conducting qualitative studies established by the Consolidated Criteria for Reporting Qualitative Research were followed. RESULTS: Three main themes were identified: a) The meaning of physical rehabilitation to parents, b) Physical rehabilitation as an opportunity for patients to stay in their home environment and c) Home-based physical rehabilitation as part of the families' social environment. CONCLUSIONS: The main needs of a home physical rehabilitation program are to decrease pain and suffering, together with improving family education and training. CLINICAL REHABILITATION IMPACT: The experience of rehabilitation programs at home is essential in order to improve both the quality of life and the quality of care of affected children and parents.


OBJECTIVE: To explore the relation between time to reintubation and death or bronchopulmonary dysplasia (BPD) in extremely preterm infants. STUDY DESIGN: This was a subanalysis from an ongoing multicenter observational study. Infants with birth weight \( \leq 1250 \) g, requiring mechanical ventilation, and undergoing their first elective extubation were prospectively followed throughout hospitalization. Time to reintubation was defined as the time interval between first elective extubation and reintubation. Univariate and multivariate logistic regression analyses were performed to evaluate associations between time to reintubation, using different observation windows after extubation (24-hour intervals), and death/BPD (primary outcome) or BPD among survivors (secondary outcome). AORs were computed with and without the confounding effects of cumulative mechanical ventilation duration. RESULTS: Of 216 infants included for analysis, 103...
(48%) were reintubated at least once after their first elective extubation. Reintubation was associated with lower gestational age/weight and greater morbidities compared with infants never reintubated. After adjusting for confounders, reintubation within observation windows ranging between 24 hours and 3 weeks postextubation was associated with increased odds of death/BPD (but not BPD among survivors), independent of the cumulative mechanical ventilation duration. Reintubation within 48 hours from extubation conferred higher risk-adjusted odds of death/BPD vs other observation windows. CONCLUSIONS: Although reintubation after elective extubation was independently associated with increased likelihood of death/BPD in extremely preterm infants, the greatest risk was attributable to reintubation within the first 48 hours postextubation. Prediction models capable of identifying the highest-risk infants may further improve outcomes.


Current research demonstrates that pediatric symptom management care is often initiated in the late stages of disease once clinicians are no longer able to meaningfully impact symptom burden. Given that physicians or nurse practitioners are responsible for initiating palliative care referrals, it is incumbent upon registered nurses to advocate when improved symptom management care is needed. The pediatric palliative care screening instrument pilot provides a centralized instrument to document and quantify a patient’s symptom profile, giving registered nurses the opportunity to objectively communicate and track a patient’s need for improved symptom management care within the areas of pain, secretions, dyspnea, intractable seizures, nausea, vomiting, constipation, diarrhea, anorexia, cachexia, sleep disturbance, lethargy, anxiety, depression, and/or agitation. The 4-week quality improvement project at an academic teaching hospital formally incorporated the bedside registered nurses' symptom assessment into a centralized document. Fifty-three patients were identified as having an uncontrolled symptom burden in at least one of the symptom domains, indicating that excessive and untreated symptom burden was present on the acute care floor. The pediatric palliative care screening instrument could act as a conduit between bedside registered nurses and the palliative care team, serving to reduce the time between onset of excessive symptom burden and initiation of symptom management services.


Compared with younger children and older adults, adolescent and young adult (AYA) patients with cancer receive more intensive end-of-life (EOL) care. We hypothesize that enhanced understanding of AYA preferences, increased engagement of these patients in decision-making, and improved communication of their preferences with family members and the medical team will lead to
increased provision of goal-concordant care and decreased intensity of EOL care. In this study, we describe the development of a novel tool that quantifies the relative importance of numerous factors considered by AYA patients with cancer, their parents, and health care providers when choosing between treatment options. 


Spinal muscular atrophy (SMA) is a neurodegenerative disorder that affects motor neurons, primarily in young children. SMA is caused by mutations in the Survival Motor Neuron 1 (SMN1) gene. SMN functions in the assembly of spliceosomal RNPs and is well conserved in many model systems including mouse, zebrafish, fruit fly, nematode, and fission yeast. Work in Drosophila has focused on the loss of SMN function during larval stages, primarily using null alleles or strong hypomorphs. A systematic analysis of SMA-related phenotypes in the context of moderate alleles that more closely mimic the genetics of SMA has not been performed in the fly, leading to debate over the validity and translational value of this model. We, therefore, examined 14 Drosophila lines expressing SMA patient-derived missense mutations in Smn, with a focus on neuromuscular phenotypes in the adult stage. Animals were evaluated on the basis of organismal viability and longevity, locomotor function, neuromuscular junction structure, and muscle health. In all cases, we observed phenotypes similar to those of SMA patients, including progressive loss of adult motor function. The severity of these defects is variable and forms a broad spectrum across the 14 lines examined, recapitulating the full range of phenotypic severity observed in human SMA. This includes late-onset models of SMA, which have been difficult to produce in other model systems. The results provide direct evidence that SMA-related locomotor decline can be reproduced in the fly and support the use of patient-derived SMN missense mutations as a comprehensive system for modeling SMA.


The care of individuals with Duchenne muscular dystrophy (DMD) now extends into adulthood. Childhood to adulthood transition planning is an important aspect of care, affecting health outcomes as well as other important aspects of adult life. In this article, we address transition planning as it relates to DMD health care, education, steps toward vocations, personal care, accessing the home and community, and the importance of relationships with others. Because of the complex, disabling, and progressive nature of DMD, coordinated, well-timed planning is critical to ensure that all components of transition are accomplished. In this article, we introduce the DMD Transition Toolkit. The toolkit is designed to help assess readiness for transition, track progress toward transition goals, and provide a template for documenting key elements of medical care, medical equipment, and services. The transition readiness assessment for young adults with DMD is used to gauge readiness for adult health care and living practices. Consistent with the 2018 DMD Care Considerations, the transition checklist for young adults with DMD is a comprehensive list to
be considered, discussed, and planned for during transition. The medical summary for young adults with DMD can be used by a provider or individuals with DMD to communicate details of their health plan, provider contacts, and medical equipment needs. It can be used in transition handoffs, when adding new providers, or when informing new nursing agencies or personal care attendants. It could also be useful in urgent care settings by providing baseline information about the individual with DMD.


: media-1vid110.1542/5804909711001PEDS-VA_2017-4182Video Abstract BACKGROUND: Knowledge about how children die in pediatric hospitals is limited, and this hinders improvement in hospital-based end-of-life care. METHODS: We conducted a retrospective chart review of all the patients who died in a children's hospital between July 2011 and June 2014, collecting demographic and diagnostic information, hospital length of stay, location of death, and palliative care consultation. A qualitative review of provider notes and resuscitation records was used to create 5 mutually exclusive modes of death, which were then assigned to each patient. Analysis included the calculation of descriptive statistics and multinomial logistic regression modeling. RESULTS: We identified 579 patients who were deceased; 61% were <1 year of age. The ICU was the most common location of death (NICU 29.7%; PICU 27.8%; cardiac ICU 16.6%). Among the 5 modes of death, the most common was the withdrawal of life-sustaining technology (40.2%), followed by nonescalation (25.6%), failed resuscitation (22.8%), code then withdrawal (6.0%), and death by neurologic criteria (5.3%). After adjustment, patients who received a palliative care consultation were less likely to experience a code death (odds ratio 0.31; 95% confidence interval 0.13-0.75), although African American patients were more likely than white patients to experience a code death (odds ratio 2.46; 95% confidence interval 1.05-5.73), mostly because of code events occurring in the first 24 hours of hospitalization. CONCLUSIONS: Most deaths in a children's hospital occur in ICUs after the withdrawal of life-sustaining technology. Race and palliative care involvement may influence the manner of a child's death.


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 Sisom 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.

heart disease: existing evidence, conflicts and concerns." J Matern Fetal Neonatal Med,

INTRODUCTION: Congenital heart disease is one of the most of the groups of congenital
anomalies with an incidence of about 1 per 100 live births. Almost one-third of these infants
require some type of intervention, usually in the first year of life and increasingly often in the
neonatal period. Innovative reparative and palliative surgical procedures and advanced medical
support in the Neonatal Intensive Care Unit have significantly reduced the mortality related to
congenital heart disease. Achieving survival is not the only target of clinicians for these patients.
Appropriate growth, development, and improved quality of life are also very important. Growth
failure is a very common problem of these children and nutritional support and management are a
challenge for health care providers. Early intervention and identification of at-risk patients have the
potential to decrease morbidity and mortality related to malnutrition. AIM/METHODS: The purpose
of this article is to analyze the existing evidence and common concerns about perioperative and
postdischarge nutritional management of neonates with congenital heart disease based on the
special issues or complications that may arise. Furthermore, we reviewed the recent literature
about current practices and proposed policies that could prevent malnutrition and improve the
outcomes of neonates with congenital heart disease. RESULTS/CONCLUSION: A standardized
institutional protocol and clear guidelines referring to feeding initiation, prompt estimation of
caloric needs and provision of adequate and appropriate nutrient intake is likely to benefit these
patients. Clear definitions for the nutritional approach in the setting of medical complications and
close assessment of growth by pediatricians and specialized nutritionists are crucial for the long-
term outlook and quality of life of these infants.

(2018). "Self-reported fatigue in children with advanced cancer: Results of the PediQUEST

BACKGROUND: Pediatric cancer-related fatigue is prevalent and significantly impairs health-related
quality of life, yet its patterns and correlates are poorly understood. The objectives of this study
were to describe fatigue as prospectively reported by children with advanced cancer and to
identify the factors associated with fatigue and associated distress. METHODS: Children (age >/=2
years) with advanced cancer (N = 104) or their parents at 3 academic hospitals reported symptoms
at most weekly over 9 months using the computer-based Pediatric Quality of Life Evaluation of
Symptoms Technology (PediQUEST) system. PediQUEST administered a modified version of the
Memorial Symptom Assessment Scale (PQ-MSAS) as part of a randomized controlled trial. Clinical
information was abstracted from medical records. Primary outcomes were: 1) fatigue prevalence
(yes/no response to PQ-MSAS fatigue item) and 2) fatigue distress (composite score of severity,
frequency, and bother). Multivariable models were constructed to identify factors independently

82
associated with fatigue prevalence and scores reflecting fatigue distress (ie, burden). RESULTS: Of 920 reports, 46% (n = 425) noted fatigue. When reported, fatigue was of high frequency in 41% of respondents (n = 174), severity in 25% of respondents (n = 107), and bother in 34% of respondents (n = 143). Most reports (84%; n = 358) were associated with scores indicating fatigue distress. In multivariable analyses, fatigue was associated with older age, lower hemoglobin, and distress from particular symptoms (anorexia, nausea, sleep disturbance, sadness, and irritability). In contrast, fatigue distress was associated with distress from nausea, cough, and pain. CONCLUSIONS: Fatigue is common among children with advanced cancer and is often highly distressing. Interventions focused on uncontrolled symptoms may ease fatigue distress in children with advanced cancer. https://www.ncbi.nlm.nih.gov/pubmed/30291811


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. https://www.ncbi.nlm.nih.gov/pubmed/29943874


BACKGROUND: In the last years, the structure of Pediatric Palliative Care in Germany has developed more and more. Since 2007, there is a legal claim for a specialized palliative care in German which also applies to children. Therefore, the need of an advance care planning for children is frequently discussed. In Germany, a written advance is judicially approved only when
the person concerned is of full age, intentions to change this legal ground are going on. Nevertheless in many institutions involved in pediatric palliative care standard forms similar to an advance patient directive are used, especially since 2009 when a Do-Not-Resuscitate-Order (DNR-Order) equivalent for children was published on which many German pediatric medical societies had agreed. METHODS: To get an overview which DNR-Order equivalents are actually used in pediatric palliative care in Germany we sent a questionnaire with 10 items to 174 institutions that are involved in pediatric oncological palliative care between August 2012 and October 2013. RESULTS: Only 46.9% of replying institutions used the DNR-Order equivalent for children approved by many German pediatric societies. When asked for optimizing such an advance patient directive for minors it was mostly suggested to include always a protocol of the consenting talk, an individualized treatment algorithm of all therapeutic options (not only emergency measures) in the palliative setting, and a more detailed information about the patient's current palliative situation. CONCLUSIONS: All collected data were summarized within a suggestion for a new advance pediatric oncological care planning standard form for minors in Germany.


PURPOSE OF REVIEW: People with epilepsy have an increased risk of mortality when compared to the general population. Sudden unexpected death in epilepsy (SUDEP) is the most common cause of epilepsy-related death in children and adults. The purpose of this review is to discuss SUDEP, with an emphasis on SUDEP risk factors, their mitigation and prevention. RECENT FINDINGS: SUDEP affects approximately 1 in 1000 people with epilepsy each year. Recent studies suggest that the incidence in children is similar to that of adults. The most important risk factor for SUDEP is the presence and frequency of generalized tonic-clonic seizures. The presence of nocturnal supervision may decrease risk along with the use of nocturnal listening devices. Underlying genetic influences, both cardiac and epilepsy-related may further alter risk. Risk mitigation strategies include reducing seizure frequency, optimizing therapy, and the use of nocturnal supervision/seizure detection devices. Risk factors for SUDEP are well established; however, pediatric specific risk factors have not been identified. Current prevention strategies are focused on reduction of risk factors and the possible role of seizure detection devices. More research is needed to better understand the varied underlying pathological mechanisms and develop targeted prevention strategies. Further understanding the genetic factors that influence SUDEP risk may potentially aid in understanding the underlying pathophysiology of SUDEP.


BACKGROUND: Specialized pediatric palliative care programs aim to improve quality of life and ease distress of patients and their families across the illness trajectory. These programs require further development, which should be based on how they improve outcomes for patients, families, health care professionals, and the health care system. OBJECTIVE: To identify and compare definitions of indicators used to assess the impact of specialized pediatric palliative care programs.
DESIGN: The scoping review protocol was prospectively registered on PROSPERO 2017 (CRD42017074090). DATE SOURCES: MEDLINE, PsycINFO, Cochrane Central Register of Controlled Trials, Web of Science, CINAHL, Scopus, and Embase databases were searched from January 2000 to September 2018. Eligible studies included randomized controlled trials, experimental studies, or observational studies that compared specialized programs with usual care. Studies were excluded if most care recipients were older than 19 years or the article was not available in English, French, German, or Spanish. RESULTS: Forty-six studies were included; one was a randomized controlled trial. We identified 82 different indicators grouped into 14 domains. The most common indicators included the following: location of death, length of stay in hospital, and number of hospital admissions. Only 22 indicators were defined identically in at least 2 studies. Only one study included children’s perspectives in assessing indicators. CONCLUSIONS: Many indicators were used to assess program outcomes with little definition consensus across studies. Development of a set of agreed-upon indicators to assess program impact concurrent with family and patient input is essential to advance research and practice in pediatric palliative care.


To date, outcomes in neonatology have focused mainly on the biological outcomes of the babies under our care. In this article, we argue that we must move beyond this proband and biological bias, towards a "Slow Medicine" that recognizes the distinction between the remarkable technical capabilities of the modern medical world and how those intersect with our society, and its values, more broadly. Practically speaking, this involves consideration of the impacts of neonatal intensive care and its sequelae on families, as well as non-biological outcomes such as finances and stress. Implementing this Slow Medicine does not mean that neonatologists must forego effective therapy or the improved mortality and morbidity it has brought, but rather that we adopt a committed and compassionate view of medicine, in which we engage outside the neonatal intensive care unit to address the nonbiological suffering of our patients and their equally vulnerable families.

BACKGROUND: Children with cerebral palsy (CP) face particular challenges, e.g. daily pain that threaten their participation in school activities. This study focuses on how teachers, personal assistants, and clinicians in two countries with different cultural prerequisites, Sweden and South Africa, manage the pain of children in school settings. METHOD: Participants’ statements collected in focus groups were analysed using a directed qualitative content analysis framed by a Frequency of attendance-Intensity of involvement model, which was modified into a Knowing-Doing model. RESULTS: Findings indicated that pain management focused more on children's attendance in the classroom than on their involvement, and a difference between countries in terms of action-versus-reaction approaches. Swedish participants reported action strategies to prevent pain whereas South African participants primarily discussed interventions when observing a child in pain. CONCLUSION: Differences might be due to school- and healthcare systems. To provide effective support when children with CP are in pain in school settings, an action-and-reaction approach would be optimal and the use of alternative and augmentative communication strategies would help to communicate children's pain. As prevention of pain is desired, structured surveillance and treatment programs are recommended along with trustful collaboration with parents and access to “hands-on” pain management when needed. Implications for rehabilitation * When providing support, hands-on interventions should be supplemented by structured preventive programs and routines for parent collaboration (action-and-reaction approach). * When regulating support, Sweden and South Africa can learn from each other; o In Sweden, the implementation of a prevention program has been successful. o In South Africa, the possibilities giving support directly when pain in children is observed have been beneficial. 


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.


Background: Venipuncture is described by children as one of the most painful and frightening medical procedures. Objective: To evaluate the effectiveness of Virtual Reality (VR) as a distraction technique to help control pain in children and adolescents undergoing venipuncture. Methods: Using a within-subjects design, fifteen patients (mean age 10.92, SD = 2.64) suffering from oncological or hematological diseases received one venipuncture with "No VR" and one venipuncture with "Yes VR" on two separate days (treatment order randomized). "Time spent thinking about pain", "Pain Unpleasantness", "Worst pain" the quality of VR experience, fun during the venipuncture and nausea were measured. Results: During VR, patients reported significant reductions in "Time spent thinking about pain," "Pain unpleasantness," and "Worst pain". Patients also reported significantly more fun during VR, and reported a "Strong sense of going inside the computer-generated world" during VR. No side effects were reported. Conclusion: VR can be considered an effective distraction technique for children and adolescents' pain management during venipuncture. Moreover, VR may elicit positive emotions, more than traditional distraction techniques. This could help patients cope with venipuncture in a non-stressful manner. Additional research and development is needed.


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 SDY, 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.


https://www.ncbi.nlm.nih.gov/pubmed/30537003


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: 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 analgesics. 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 (ps<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.


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. https://www.ncbi.nlm.nih.gov/pubmed/30181088


OBJECTIVES: Abscess incision and drainage (I&D) are painful and distressing procedures in children. Intranasal (IN) fentanyl is an effective analgesic for reducing symptomatic pain associated with fractures and burns but has not been studied for reducing procedural pain during abscess I&D. Our objective was to compare the analgesic efficacy of IN fentanyl with intravenous (IV) morphine for abscess I&D in children. METHODS: We performed a randomized noninferiority trial in children aged 4 to 18 years undergoing abscess I&D in a pediatric emergency department. Patients received IN fentanyl (2 mug/kg; maximum, 100 mug) or IV morphine (0.1 mg/kg; maximum, 8 mg). The primary outcome, determined independently by blinded assessors, was the Observational Scale of Behavioral Distress-Revised (OSBD-R). The prestated margin of noninferiority (Delta) was 1.80. Secondary outcomes included self-reported pain, treatment failure, and patient and parental satisfaction. RESULTS: We enrolled 20 children (median age, 15.4 years), 10 in each group. The difference between total OSBD-R scores was -13.45 (95% confidence interval, -24.24 to -2.67), favoring IN fentanyl. There was less self-reported pain in patients who received IN fentanyl immediately after the procedure. Four patients (40%) receiving IV morphine had treatment failures and required moderate sedation or had the procedure terminated. More patients who received IN fentanyl were satisfied with the analgesic administered compared with those who received IV morphine. CONCLUSIONS: In a small sample of children aged 4 to 18 years undergoing abscess I&D, IN fentanyl was noninferior, and potentially superior, to IV morphine for reducing procedural pain and distress. https://www.ncbi.nlm.nih.gov/pubmed/27387971
BACKGROUND: This is an update of the original Cochrane review first published in Issue 1, 2003, and previously updated in 2009, 2012 and 2014. Chronic pain, defined as pain that recurs or persists for more than three months, is common in childhood. Chronic pain can affect nearly every aspect of daily life and is associated with disability, anxiety, and depressive symptoms. OBJECTIVES: The aim of this review was to update the published evidence on the efficacy of psychological treatments for chronic and recurrent pain in children and adolescents. The primary objective of this updated review was to determine any effect of psychological therapy on the clinical outcomes of pain intensity and disability for chronic and recurrent pain in children and adolescents compared with active treatment, waiting-list, or treatment-as-usual care. The secondary objective was to examine the impact of psychological therapies on children's depressive symptoms and anxiety symptoms, and determine adverse events. SEARCH METHODS: Searches were undertaken of CENTRAL, MEDLINE, MEDLINE in Process, Embase, and PsycINFO databases. We searched for further RCTs in the references of all identified studies, meta-analyses, and reviews, and trial registry databases. The most recent search was conducted in May 2018. SELECTION CRITERIA: RCTs with at least 10 participants in each arm post-treatment comparing psychological therapies with active treatment, treatment-as-usual, or waiting-list control for children or adolescents with recurrent or chronic pain were eligible for inclusion. We excluded trials conducted remotely via the Internet. DATA COLLECTION AND ANALYSIS: We analysed included studies and we assessed quality of outcomes. We combined all treatments into one class named 'psychological treatments'. We separated the trials by the number of participants that were included in each arm; trials with > 20 participants per arm versus trials with < 20 participants per arm. We split pain conditions into headache and mixed chronic pain conditions. We assessed the impact of both conditions on four outcomes: pain, disability, depression, and anxiety. We extracted data at two time points; post-treatment (immediately or the earliest data available following end of treatment) and at follow-up (between three and 12 months post-treatment). MAINRESULTS: We identified 10 new studies (an additional 869 participants) in the updated search. The review thus included a total of 47 studies, with 2884 children and adolescents completing treatment (mean age 12.65 years, SD 2.21 years). Twenty-three studies addressed treatments for headache (including migraine); 10 for abdominal pain; two studies treated participants with either a primary diagnosis of abdominal pain or irritable bowel syndrome, two studies treated adolescents with fibromyalgia, two studies included adolescents with temporomandibular disorders, three were for the treatment of pain associated with sickle cell disease, and two studies treated adolescents with inflammatory bowel disease. Finally, three studies included adolescents with mixed pain conditions. Overall, we judged the included studies to be at unclear or high risk of bias. Children with headache pain We found that psychological therapies reduced pain frequency post-treatment for children and adolescents with headaches (risk ratio (RR) 2.35, 95% confidence interval (CI) 1.67 to 3.30, P < 0.01, number needed to treat for an additional beneficial outcome (NNTB) = 2.86), but these effects were not maintained at follow-up. We did not find a beneficial effect of psychological therapies on reducing disability in young people post-treatment (SMD -0.26, 95% CI -0.56 to 0.03), but we did find a beneficial effect in a small number of studies at follow-up (SMD -0.34, 95% CI -0.54 to -0.15). We found no beneficial effect of psychological interventions on depression or anxiety symptoms. Children with mixed pain conditions We found that psychological therapies reduced pain intensity post-
treatment for children and adolescents with mixed pain conditions (SMD -0.43, 95% CI -0.67 to -0.19, P < 0.01), but these effects were not maintained at follow-up. We did find beneficial effects of psychological therapies on reducing disability for young people with mixed pain conditions post-treatment (SMD -0.34, 95% CI -0.54 to -0.15) and at follow-up (SMD -0.27, 95% CI -0.49 to -0.06). We found no beneficial effect of psychological interventions on depression symptoms. In contrast, we found a beneficial effect on anxiety at post-treatment in children with mixed pain conditions (SMD -0.16, 95% CI -0.29 to -0.03), but this was not maintained at follow-up. Across all pain conditions, we found that adverse events were reported in seven trials, of which two studies reported adverse events that were study-related. Quality of evidence We found the quality of evidence for all outcomes to be low or very low, mostly downgraded for unexplained heterogeneity, limitations in study design, imprecise and sparse data, or suspicion of publication bias. This means our confidence in the effect estimate is limited: the true effect may be substantially different from the estimate of the effect, or we have very little confidence in the effect estimate; or the true effect is likely to be substantially different from the estimate of effect.

AUTHORS’ CONCLUSIONS: Psychological treatments delivered predominantly face-to-face might be effective for reducing pain outcomes for children and adolescents with headache or other chronic pain conditions post-treatment. However, there were no effects at follow-up. Psychological therapies were also beneficial for reducing disability in children with mixed chronic pain conditions at post-treatment and follow-up, and for children with headache at follow-up. We found no beneficial effect of therapies for improving depression or anxiety. The conclusions of this update replicate and add to those of a previous version of the review which found that psychological therapies were effective in reducing pain frequency/intensity for children with headache and mixed chronic pain conditions post-treatment.

https://www.ncbi.nlm.nih.gov/pubmed/30939227


Background: Chronic pain is a prevalent health condition associated with parenting difficulties. Pain-specific parenting, such as protectiveness and catastrophizing, may contribute to chronic pain in children. Additional work is needed to test predictors of pain-specific parenting. Aim: The current study tested parent mental health symptoms as predictors of protectiveness and catastrophizing about child pain and whether comorbid pain and mental health symptoms exacerbate risk for problematic responses to children’s pain. Methods: Parents with chronic pain (n = 62) and parents without chronic pain (n = 80) completed self-report questionnaires assessing pain characteristics, mental health symptoms, and pain-specific parenting responses. Results: Results indicated significantly higher rates of depression, anxiety, and somatization in parents with chronic pain. Depression predicted protectiveness and catastrophizing over and above chronic pain status. Chronic pain status moderated the association between increased anxiety and greater catastrophizing about child pain. Conclusions: Findings highlight the potential impact of mental health symptoms on pain-specific parenting even when accounting for chronic pain status.


Spinal muscular atrophy (SMA) is a recessive disorder caused by a mutation in the survival motor neuron 1 gene (SMN1); it affects 1 in 11,000 newborn infants. The most severe and most common form, type 1 SMA, is associated with early mortality in most cases and severe disability in survivors. Nusinersen, an antisense oligonucleotide, promotes production of full-length protein from the pseudogene SMN2. Nusinersen treatment prolongs survival of patients with type 1 SMA and allows motor milestone acquisition. Patients with type 2 SMA also show progress on different motor scales after nusinersen treatment. Nusinersen was recently approved by the European Medicines Agency and the US Food and Drug Administration; it is now reimbursed in several European countries and in the USA. In Australia, the transition from expanded access programme to commercial availability is coming soon. In New Zealand, an expanded access programme is opened, and in Canada price negotiation for the treatment is in progress. In this review we exemplify the clinical benefit of nusinersen in subgroups of patients with SMA. Nusinersen represents the first efficacious marked approved drug in type 1 and type 2 SMA. Different knowledge gaps, such as results in older patients, in patients with permanent ventilation, in patients with neonatal forms, or in patients after spinal fusion, still need to be addressed. WHAT THIS PAPER ADDS: Identifies gaps in knowledge about the efficacy of nusinersen in broader populations of patients with spinal muscular atrophy. Identifies open questions in populations of patients where proof of efficacy is available.


Methadone is a synthetic opioid with unique pharmacodynamic and pharmacokinetic properties. It is effective in treating both nociceptive and neuropathic pain, which commonly co-exist in children with cancer. Upon reviewing the literature describing the use of methadone in pediatric oncology patients, publications are limited in number and low in quality of evidence; nevertheless, there is support for the safety and efficacy of methadone in treating pain in children with cancer, particularly when pain is refractory to conventional treatment. Although the risk of life-threatening arrhythmia is commonly cited as an argument against the use of methadone, our review of the literature did not support this finding in children. Further evaluation with prospective studies is warranted to develop evidence-based recommendations for the use of methadone in pediatric oncology.


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. https://www.ncbi.nlm.nih.gov/pubmed/30322510


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). https://www.ncbi.nlm.nih.gov/pubmed/29893452

There is a scarcity of work examining the relationship between culture and pain-related caregiver behaviors. Moreover, no pediatric pain studies have examined the relationship between caregiver cultural values and pain-related caregiver behaviors nor discern if this process is mediated by caregiver parenting styles and moderated by ecosocial context. Based on cross-cultural developmental theories, this study hypothesized that ecosocial context would moderate the relationship between cultural values, parenting styles, and pain-related caregiver behaviors; and that parenting styles mediate the effect of cultural values on pain-related caregiver behaviors. A cross-cultural survey design was employed using a convenience sample of 547 caregivers of 6 to 12 year olds living in Canada (n = 183), Iceland (n = 184), and Thailand (n = 180). Multigroup structural equation modeling showed that ecosocial context did not affect which cultural model of parenting the caregiver adopted. Parenting styles mediated the relationship between cultural values and pain-related caregiver behavior. Vertical/horizontal individualism, collectivism, and authoritative- and authoritarian-parenting styles positively predicted solicitousness. Vertical individualism and authoritarian-parenting style positively predicted discouraging behavior, whereas other predictors did not. The findings support the sociocommunication model of children's pain by showing that cultural context does affect parents' behaviors. They also corroborate with others' claims of solicitousness universality in a pediatric pain context. However, solicitousness may have different cultural meanings among individuals and may be used in conjunction with discouraging behavior. The findings from this study have implications for the theory development about culture and pediatric pain, but do not provide specific clinical recommendations.


Nusinersen is the first approved drug to treat spinal muscular atrophy (SMA). Its periodic intrathecal delivery may cause psychological burden in infants and in their parents. We report our experience during expanded access program (EAP) for type 1 SMA in a single Italian center. Because of the occurrence of stress emotional states, anxious reactions and fear before, during, and after lumbar puncture (LP), a specific psychological intervention was implemented based on regulation of emotions, anticipatory expectations, and post-event attributions. Activities included the use of fairy tales, distraction, music play through listening preferred cartoon themes in the youngest children, and contextual games and solution of fun riddle quizzes in the oldest ones. State anxiety greatly reduced in children and their parents. Treatment of psychological aspects should therefore become an integral part of health care in SMA infants and children during Nusinersen treatment.


The diagnosis of cancer in a child leaves parents and families devastated and vulnerable. In an effort to do everything possible, families often choose an integrative medicine approach to their
Surveys have found that 31%-84% of children with cancer use complementary and alternative medicine and most often as supportive care agents. Several systematic reviews have demonstrated a clinical benefit for some select therapies; however, the safety and efficacy of the combination of biological therapies with conventional treatment remain largely unknown and garner concern due to the potential for interactions with conventional therapy. Given the sustained use and potential benefit of integrative medicine, additional research is warranted in pediatric oncology. Utilizing the available literature, clinical providers should aim to conduct open and non-judgmental discussions with families about the use of integrative medicine so as to guide the safe integration of the two modalities.


Nurses are often the first to recognize and respond to children's symptoms. This descriptive, exploratory study characterized how pediatric oncology health care providers characterize and assess children's cancer-related symptoms. The study also explored challenges associated with symptom assessment and information perceived as helpful in planning interventions. The setting was a Children's Oncology Group-affiliated hospital in the Intermountain West of the United States. Twenty-two pediatric oncology health care providers (95% female; 68% nurses) participated in one of four focus group sessions. Sessions were facilitated by two individuals and included six open-ended questions addressing participants' perspectives of cancer-related symptoms, approaches to symptom assessment, challenges and frustrations encountered when assessing symptoms, and information needed to plan interventions. Participants identified 75 physical and psychosocial responses that included both subjectively experienced symptoms and other consequences of the cancer experience. Qualitative content analysis procedures organized other responses into categories and subcategories. Participants most frequently reported using observational approaches including physical assessment findings and observation of the child's behavior to identify symptoms. Strategies that sought the child's input such as the use of a rating scale or seeking the child's verbal description were less frequently named. Participants related discerning and interpreting the child's behaviors as a challenge to symptom assessment. They also reported attention to symptom characteristics as important to planning interventions. Future directions include building capacity to support child-centric symptom assessment. Development of reliable and valid resources for use in clinical settings may support a more child-centric approach to symptom assessment.


AIM: Sensorial saturation (SS) is an analgesic approach to babies' pain that includes three types of stimulations: oral sugar, massage and caregivers' voice. The aim of this review is to assess its efficacy. METHODS: We performed an analysis of scientific literature from 2001 to 2017, retrieving those clinical trials where SS had been compared with other analgesic treatments during procedural pain in babies. RESULTS: We retrieved 14 studies. Pain sources were heel-prick in nine,
eye examination and intramuscular shots in two each, and endotracheal aspiration in one. SS was the most effective treatment in all cases, except in endotracheal suctioning. No drawbacks were reported in any study using SS. CONCLUSION: SS is a safe and effective approach to neonatal pain due to heel-prick, more effective than oral sucrose or glucose in both term and preterm babies; it seems also effective in other types of acute procedural pain like eye examination or intramuscular injections, but more studies are needed to confirm these preliminary data. More studies are also needed to test SS efficacy for other procedures, and for older infants.


CONTEXT: Methadone is a long-acting opioid known for its unique pharmacokinetic and pharmacodynamic properties. Most research on methadone in children is limited to its effect on the prolongation of the corrected QT (QTc) interval. OBJECTIVES: To better understand the attitudes, beliefs, and practices of pediatric palliative care physicians regarding the use of methadone in children with advanced cancer. METHODS: A survey was sent to the American Academy of Pediatrics Section of Hospice and Palliative Medicine LISTSERV. Information on demographics, dosing of methadone, and the use of electrocardiograms (ECGs) was collected. RESULTS: One-hundred and five respondents (91%) provide palliative care to children >/= 50% of the time, and a majority (81, 77%) prescribe methadone. Most (62, 77%) physicians were board certified in Hospice and Palliative Medicine, and most (39, 63%) certified via the direct pathway (“grandfathering”). Most physicians (57, 70%) do not use loading doses of methadone. Board-certified physicians trended toward decreasing methadone dose more (40% +/- 19%) than non-board-certified physicians (28%, +/-20%) when changing from the oral to intravenous route (P = 0.07). Respondents defined a QTc interval as "prolonged" (mean +/- SD) at 444 milliseconds (+/-68 milliseconds). The percentage of patients receiving a baseline ECG was 65% (+/-33%). The most common reason for not performing a baseline ECG was that the patient was on hospice (13, 36%). CONCLUSIONS: There are consistent practices, attitudes, and beliefs of pediatric palliative care providers with regard to methadone. More education is needed on the accurate value of a prolonged QTc interval.


BACKGROUND: Systematic symptom assessment is not a standard of care in children with cancer. Many well-known symptom assessment tools are lengthy or difficult to integrate into a daily pediatric palliative care practice. We created a series of brief and simple questions to be systematically given to children and their caregivers. OBJECTIVE: The primary objective was to determine the percentage of eligible children and caregivers exposed to the questions that were able to complete the assessment. Secondary objectives included documenting the symptom burden at the time of consultation, evaluating the level of agreement in symptom reporting between children and caregivers, as well as between children/caregivers and the referring medical
team. DESIGN: A series of systematic questions were presented to all caregivers (if present) and children who were seven years of age or older at the time of initial consultation with pediatric palliative care. RESULTS: One hundred twenty-two consecutive children/caregiver dyads were given the survey. One hundred seven of 108 (99%) eligible caregivers and 83 of 97 (86%) eligible children completed the survey. Lack of appetite (child-72/83, 87%; caregiver-89/107, 83%) and pain (child-71/83, 86%; caregiver-86/107, 80%) were the most commonly reported symptoms. Caregivers reported irritability (p = 0.005) and nervousness (p < 0.001) more frequently than children. Referring medical teams significantly underdiagnosed psychological and other less clinically evident symptoms such as lack of appetite, fatigue, and sleep disturbance (p < 0.001). CONCLUSIONS: Our series of questions is easy to complete by children and caregivers. Systematic symptom assessment of children with cancer referred to palliative care should become a true standard of care.


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.


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.


Pain is a major problem in sick newborn infants, especially for those needing intensive care. Pharmacological pain relief is the most commonly used, but might be ineffective and has side effects, including long-term neurodevelopmental sequelae. The effectiveness and safety of alternative analgesic methods are ambiguous. The objective was to review the effectiveness and safety of non-pharmacological methods of pain relief in newborn infants and to identify those that are the most effective. PubMed and Google Scholar were searched using the terms: "infant", "premature", "pain", "acupuncture", "skin-to-skin contact", "sucrose", "massage", "musical therapy" and 'breastfeeding'. We included 24 studies assessing different methods of non-pharmacological analgesic techniques. Most resulted in some degree of analgesia but many were ineffective and some were even detrimental. Sucrose, for example, was often ineffective but was more effective than music therapy, massage, breast milk (for extremely premature infants) or non-invasive electrical stimulation acupuncture. There were also conflicting results for acupuncture, skin-to-skin care and musical therapy. Most non-pharmacological methods of analgesia provide a modicum of relief for preterm infants, but none are completely effective and there is no clearly superior method. Study is also required to assess potential long-term consequences of any of these methods.


This case report discusses the problems encountered with dystonia in a teenage boy with end-stage calcifying leucodystrophy. It records the successful use of prolonged continuous subcutaneous clonidine and the change to thrice weekly transdermal clonidine.

OBJECTIVES: Benzodiazepine use may be associated with delirium in critically ill children. However, benzodiazepines remain the first-line sedative choice in PICUs. Objectives were to determine the temporal relationship between administration of benzodiazepines and delirium development, control for time-varying covariates such as mechanical ventilation and opiates, and evaluate the association between dosage of benzodiazepines and subsequent delirium. DESIGN: Retrospective observational study. SETTING: Academic tertiary care PICU. PATIENTS: All consecutive admissions from January 2015 to June 2015. INTERVENTIONS: Retrospective assessment of benzodiazepine exposure in a population that had been prospectively screened for delirium. MEASUREMENTS AND MAIN RESULTS: All subjects were prospectively screened for delirium throughout their stay, using the Cornell Assessment for Pediatric Delirium, with daily cognitive status assigned as follows: delirium, coma, or normal. Multivariable mixed effects modeling determined predictors of delirium overall, followed by subgroup analysis to assess effect of benzodiazepines on subsequent development of delirium. Marginal structural modeling was used to create a pseudorandomized sample and control for time-dependent variables, obtaining an unbiased estimate of the relationship between benzodiazepines and next day delirium. The cumulative daily dosage of benzodiazepines was calculated to test for a dose-response relationship. Benzodiazepines were strongly associated with transition from normal cognitive status to delirium, more than quadrupling delirium rates (odds ratio, 4.4; CI, 1.7-11.1; p < 0.002). Marginal structural modeling demonstrated odds ratio 3.3 (CI, 1.4-7.8), after controlling for time-dependent confounding of cognitive status, mechanical ventilation, and opiates. With every one log increase in benzodiazepine dosage administered, there was a 43% increase in risk for delirium development. CONCLUSIONS: Benzodiazepines are an independent and modifiable risk factor for development of delirium in critically ill children, even after carefully controlling for time-dependent covariates, with a dose-response effect. This temporal relationship suggests causality between benzodiazepine exposure and pediatric delirium and supports limiting the use of benzodiazepines in critically ill children.


This study explored the feasibility of generating reliable information on the frequency, nature and management of breakthrough pain (BTP) in children with life-limiting conditions and life-threatening illnesses (LTIs) from narrative clinical records. In the absence of standardized ways for documenting BTP, we conducted a consensus exercise to develop a glossary of terms that could denote BTP in the records. Thirteen clinicians who contributed to the records reached consensus on 45 terms which could denote BTP, while emphasizing the importance of contextual information. The results of this approach together with guidance for improving the reliability of retrospective reviews informed a data extraction instrument. A pilot test of this instrument showed poor agreement between raters. Given the challenges encountered, we do not recommend a
retrospective review of BTP using narrative records. This study highlighted challenges of data extraction for complex symptoms such as BTP from narrative clinical records. For both clinical and research purposes, the recording of complex symptoms such as BTP would benefit from clear criteria for applying definitions, a more structured format and the inclusion of validated assessment tools. This study also showed the value of consensus exercises in improving understanding and interpretation of clinical notes within a service.


Aim: The aim of this study is to describe the experiences of other countries regarding the status of pediatric palliative care in the field of symptom management and to compare it with the current status in Iran to achieve an appropriate level of symptom management for children with cancer.

Materials and Methods: This is a comparative study. The research population includes the palliative care systems of Jordan, England, Australia, and Canada, which were ultimately compared with Iran's palliative care system. Results: The results showed that in the leading countries in the field of palliative care, such as Australia and Canada, much effort has been made to improve palliative care and to expand its service coverage. In the UK, as a pioneer in the introduction of palliative care, a significant portion of clinical performance, education and research, is dedicated to childhood palliative care. Experts in this field and policymakers are also well aware of this fact. In developing countries, including Jordan, palliative care is considered a nascent specialty, facing many challenges. In Iran, there is still no plan for providing these services coherently even for adults.

Conclusion: Children with cancer experience irritating symptoms during their lives and while they are hospitalized. Regarding the fact that symptom management in developed countries is carried out based on specific and documented guidelines, using the experiences of these successful countries and applying them as an operational model can be useful for developing countries such as Iran.


BACKGROUND AND OBJECTIVE: The link between humour and sense of humour with pain has been a topic of research for decades. The purpose of the present article was to review the different studies that have been conducted to date on the association between humour and sense of humour with pain.

DATABASES AND DATA TREATMENT: The literature search was conducted using the PubMed, Science Direct and ProQuest databases. Forty-one studies were reviewed, and the results are summarized and structured into three sections: experimental pain, chronic pain and pain in children.

RESULTS: For experimental pain, the findings support the idea that humorous distractions, such as watching a comedy clip, increase pain tolerance, although most of the studies indicate that other non-humorous distractions produce similar effects. Regarding chronic pain, humour has been studied as a way of coping with pain and the emotional distress produced by chronic pain conditions. The results of correlational studies show significant associations between the use of humour and main variables such as anxiety and catastrophizing. Finally, concerning pain
in children, similar findings to those described for the previous sections have been reported, with a notable presence of studies on clinic clown interventions, which promote emotional well-being among children and their parents, although their effectiveness in pain reduction is controversial.

CONCLUSIONS: The study of the link between humour and pain is still on an early stage, and overcoming the limitations of previous studies is required to strengthen the promising results that have been observed up to date. SIGNIFICANCE: This review summarizes all main findings regarding humour, sense of humour and pain up until the first half of 2018 and offers a list of aspects to be considered in further studies regarding the link of humour and pain to contribute to a more systematic research.


BACKGROUND: Reducing acute pain in premature infants during neonatal care improves their neurophysiological development. The use of pharmacological and non-pharmacological analgesia, such as sucrose, is limited per day, particularly for very preterm infants. Thus, the usual practice of non-nutritive sucking is often used alone. Facilitated tucking could be an additional strategy to non-nutritive sucking for reducing pain. To the best of our knowledge, no randomized trial has compared the combination of facilitated tucking and non-nutritive sucking to non-nutritive sucking alone. OBJECTIVES: To compare the efficacy of facilitated tucking in combination with non-nutritive sucking (intervention group) to non-nutritive sucking alone (control group) in reducing pain during the heel-stick procedure in very preterm infants. DESIGN: Prospective, randomized controlled trial. SETTINGS: Level III and II neonatal care units, including the neurosensory care management program. METHODS: Very preterm infants (gestational age between 28 and 32 weeks) were randomly assigned by a computer programme to the intervention or control group during a heel-stick procedure within the first 48h of life. In both groups, infants were placed in an asymmetric position on a cushion; noise and light were limited following routine care. A heel-stick was performed first in the care sequence. In the intervention group, facilitated tucking was performed by a nurse or nursing assistant. The procedure was video recorded from 15s (T-15s) before the procedure until three minutes (T+3min) after the end of the procedure. Pain was blindly assessed by two independent specialist nurses. The primary outcome was the pain score evaluated 15s before the procedure and 30s immediately after by the premature infant pain profile (PIPP) scale. The secondary outcome was the pain score evaluated between T-15s and T+3min by the DAN scale (a French acronym for the acute pain of a newborn). RESULTS: Sixty infants were included (30 in each group). The PIPP pain scores did not differ between the intervention group (median: 8.0; interquartile range (IQR): 6.0-12.0) and the control group (median: 9.5; IQR: 7.0-13.0, p=0.32). Pain assessed by the DAN scale at T+3min was lower in the intervention group than in the control group (median: 0.3; IQR: 0.0-1.0 and 2.0; IQR: 0.5-3.0, respectively, p=0.001). CONCLUSIONS: The combined use of facilitated tucking and non-nutritive sucking did not significantly alleviate pain during the heel-stick procedure. However, the addition of facilitated tucking facilitated faster pain recovery following the heel-stick procedure.


BACKGROUND: Respiratory distress is one of the most common and frightening symptoms of children with life-limiting conditions. Because treatment of the underlying cause is frequently impossible or insufficient, in many children, symptomatic treatment is warranted. The purpose of this study was to describe the circumstances of the use of intranasal fentanyl in an acute attack of respiratory distress (AARD) in children receiving palliative care, as well as to describe outcomes and adverse events after its use. METHODS: Children and adolescents treated in a pediatric palliative unit or attended by a specialized home care team between 2010 and 2016 were included in this study. A retrospective chart review was conducted of those who were treated with intranasal fentanyl for an AARD. RESULTS: During the study period 16 children (0.5-18.6 years) with various life-limiting conditions were treated with intranasal fentanyl for AARD. In total, 70 AARDs were analyzed. In 74% of all AARDs, a single dose of intranasal fentanyl was used. Frequent causes for an AARD were excessive secretions and acute respiratory infection. The median starting dose of intranasal fentanyl was 1.5 mug/kg body weight. Labored breathing (96%), tachypnea (79%) and related suffering (97%) improved after treatment. An adverse event occurred in one child. CONCLUSIONS: Intranasal fentanyl may be a safe and effective medication for the treatment of acute attacks of respiratory distress in children with life-limiting conditions. However, prospective studies with larger sample sizes and a control group are needed to validate these findings.


Pain assessment is difficult in individuals with cerebral palsy (CP). This is of particular relevance in children with communication difficulties, when non-verbal pain behaviors could be essential for appropriate pain recognition. Parents are considered good proxies in the recognition of pain in their children; however, health professionals also need a good understanding of their patients' pain experience. This study aims at analyzing the agreement between parents' and physiotherapists' assessments of verbal and non-verbal pain behaviors in individuals with CP. A written survey about pain characteristics and non-verbal pain expression of 96 persons with CP (45 classified as communicative, and 51 as non-communicative individuals) was performed. Parents and physiotherapists displayed a high agreement in their estimations of the presence of chronic pain, healthcare seeking, pain intensity and pain interference, as well as in non-verbal pain behaviors. Physiotherapists and parents can recognize pain behaviors in individuals with CP regardless of communication disabilities.


PURPOSE OF REVIEW: Good pain management in children, especially those at end of life, is a crucial component of palliative medicine. The current review assesses some of the new and/or innovative ways to manage pain in children. The article focuses on some recent
medications/pharmaceutical options such as cannabinoids and also innovative ways to administer medication to children, such as intranasal and inhalation. RECENT FINDINGS: Current approaches to pain management now include (1) new uses of old drugs such as ketamine and lidocaine, (2) use of new drugs/medications such as cannabinoids, and (3) creative use of old technology such as atomizers, intranasal drops, and inhalation. Typically, novel approaches to care rarely start in pediatrics or palliative care. The current review has presented some new and old drugs being utilized in new and old ways. 


BACKGROUND: Almost 50,000 children and young people are affected by life-limiting conditions in the United Kingdom, around a third of which use children's hospices. Anecdotal evidence suggests that cannabinoid-based medicines (CBMs), specifically cannabis oil (CO), are being used by families with increasing frequency to manage distressing symptoms. The use of most non-prescription CBMs in the United Kingdom remains illegal. OBJECTIVE: The objective of the study was to identify the prevalence of CO use by families who use children's hospices in the United Kingdom, and the approaches taken by those services to manage it. DESIGN: An electronic survey was sent to each of the 54 children's hospices in the United Kingdom between May and July 2018, comprising 10 questions. RESULTS: Forty children's hospices from across the four countries of the United Kingdom responded to the survey, representing 74% of British children's hospices. About 87.5% of hospices knew of children who use CO therapeutically. Sixty-nine percent of those hospices have received requests to administer CO during an episode of care. Approaches by organizations around CO management varied across the sectors, including arrangements for storage, administration, and recording of its use. Hospices highlighted how the lack of available guidance made decision making more challenging. Only a third of responding organizations routinely questioned families about the use of cannabis when prescribing medicines. CONCLUSION: CO is used extensively by children who use children's hospices. Despite recognizing the use of CO, many hospices are unable to support it. There is a need for clear guidelines on how hospices should approach the care needs of children, allowing hospices to meet the needs of children who use CO, and families in a safe, consistent, and relevant way, safeguarding all children, families, and professionals within the organization. 


BACKGROUND: Preterm infants spend the early days of their lives in neonatal intensive care units, where they undergo many minor painful procedures. There are many nonpharmacologic methods that can effectively reduce the pain response of neonates who undergo routine procedures. AIMS: This study aimed to investigate whether oral glucose and listening to lullabies could bring pain relief during the removal and reinsertion of the tracheal tube and also oronasopharyngeal suctioning in premature infants to whom nasal continuous positive airway pressure was applied.
DESIGN: A double-blind, randomized controlled trial. SETTING: This study was conducted in the neonatal intensive care unit in the tertiary setting between November 2012 and September 2013. PARTICIPANTS/SUBJECTS: A total of 106 preterm infants were divided into three groups, including 37 infants in the control group, 35 infants in the lullaby group, and 34 infants in the glucose group. METHODS: All preterm infants were randomly assigned to either the intervention groups or the control group. Pain responses were assessed using the Neonatal Infant Pain Scale and the Premature Infant Pain Profile. RESULTS: An assessment of the pain severity of the preterm infants after the intervention indicated that the preterm infants in the lullaby and glucose groups had lower pain, whereas the preterm infants in the control group experienced more pain (p < .05). CONCLUSION: The findings suggest that pain could be reduced significantly in preterm infants after the suggested intervention, although further studies are required to identify the benefits of lullabies or glucose in infants during other painful procedures.

https://www.ncbi.nlm.nih.gov/pubmed/29776872


Widespread use of cannabis as a drug and passage of legislation on its use should lead to an increase in the number of scientific publications on cannabis. The aim of this study was to compare trends in scientific publication for papers on medical cannabis, papers on cannabis in general, and all papers between the years 2000 and 2017. A search of PubMed and Web of Science was conducted. The overall number of scientific publications in PubMed increased 2.5-fold. In contrast, the number of publications on cannabis increased 4.5-fold and the number of publications on medical cannabis increased almost 9-fold. The number of publications on medical cannabis in Web of science increased even more (10-fold). The most significant number of publications was in the field of psychiatry. In the fields of neurology and cancer treatment there was a significant increase in the years 2011-2013. There was a rise in the number of publications on children and the elderly after 2013. The specific indications with the largest number of publications were HIV (261), chronic pain (179), multiple sclerosis (118), nausea and vomiting (102), and epilepsy (88). More than half of the publications on medical cannabis originated from the United States, followed by Canada. More than 66% of the publications were original studies. The spike in the number of scientific publications on medical cannabis since 2013 is encouraging. In light of this trend the authors expect an even greater increase in the number of publications in this area in coming years.


Epilepsy affects millions of people worldwide. Approximately one-third have pharmacoresistant epilepsy, and of these, the majority are not candidates for epilepsy surgery. Vagus nerve stimulation (VNS) therapy has been an option to treat pharmacoresistant seizures for 30 years. In this update, we will review the clinical data that support the device’s efficacy in children, adolescents, and adults. We will also review its side-effect profile, quality of life and cost benefits, and the impact the device has on sudden unexpected death in epilepsy (SUDEP). We will then discuss candidate selection and provide guidance on dosing and future models. Vagus nerve stimulation therapy is an effective treatment for many seizure types and epilepsy syndromes with a
predictable and benign side-effect profile that supports its role as the most commonly prescribed device to treat pharmacoresistant epilepsy. "This article is part of the Supplement issue Neurostimulation for Epilepsy."


PURPOSE: This study was designed to report information regarding symptomology of incurable pediatric cancer to promote proactive medicine and support for children and their families in the palliative phase in Mainland China. METHOD: A multi-center retrospective cohort study including 205 children who died from incurable cancer between June 2008 and September 2013 were analyzed. RESULTS: An incurable diagnosis was confirmed between 0 and 1726 (median, 279) days from initial diagnosis with death occurring between 1 and 239 (median, 83) days. The most frequent symptoms were fatigue (93.7%), pain (87.3%), and poor appetite (76.1%). The earliest symptoms were pain and fatigue. Children with leukemia and lymphoma also complained early of nausea/vomiting, and children with solid tumors complained early of disturbed sleep. Later in the palliative phase, altered consciousness and seizures were found in children with central nervous system tumors and solid tumors, while children with leukemia and lymphoma were found to have fever, diarrhea, and bleeding. However, these symptoms only persisted for a short time. DNR discussions were held in 89 cases (43.4%) at a median of 37 (range, 4-178) days before death. A total of 154 patients (75.1%) died at home and 51 patients (24.9%) in the hospital. CONCLUSIONS: This study provides new knowledge about symptomology to health care professionals and parents of children in Mainland China. Given our results, an improved alternative care plan should be developed and implemented earlier to facilitate end-of-life planning.


Background: Medical cannabis has been available in the State of Minnesota since July 2015 through the Minnesota Medical Cannabis Program (MMCP). Objectives: Our study aimed to delineate oncology providers' views on medical cannabis, identify barriers to patient enrollment, and assess clinicians' interest in a clinical trial of medical cannabis in patients with stage IV cancer. Methods: From June to August 2017, we distributed a 14-question survey to Minnesota oncology physicians, advanced practice nurses, and physician assistants who care for adults and children with cancer. Descriptive analyses for each question were provided for all survey respondents. Results: Of the 529 eligible survey participants, 153 (29%) responded to our survey; 68 respondents were registered with the MMCP. Most identified themselves as a medical oncologist or medical oncology nurse practitioner/physician assistant (n=125, 82%), and most practiced in a community setting (n=102, 67%). Overall, 65% of respondents supported the use of medical cannabis. Perceived cost and inadequate research were the highest barriers to MMCP patient enrollment. The lowest barriers included lack of health group support for allowing certification of patients and risk of social stigma. Of all respondents, 36% lacked confidence in discussing the risks and benefits
of medical cannabis, and 85% wanted more education. Conclusions: Although support for cannabis use in the cancer setting is growing, significant barriers remain. This study illustrates a clear need to give clinicians both data and education to guide their discussions about the benefits, risks, and cost considerations of using medical cannabis for cancer-related symptoms.

Psychosocial and Family issues


Pena-Shokeir syndrome (PSS) type 1, also known as fetal akinesia deformation sequence, is a rare genetic syndrome that almost always results in intrauterine or early neonatal death. It is characterized by markedly decreased fetal movements, intrauterine growth restriction, joint contractures, short umbilical cord, and features of pulmonary hypoplasia. Antenatal diagnosis can be difficult. Ultrasound features are varied and may overlap with those of Trisomy 18. The poor prognosis of PSS is due to pulmonary hypoplasia, which is an important feature that distinguishes PSS from arthrogryposis multiplex congenital without pulmonary hypoplasia, which has a better prognosis. If diagnosed in the antenatal period, a late termination of pregnancy can be considered following ethical discussion (if the law allows). In most cases, a diagnosis is only made in the neonatal period. Parents of a baby affected with PSS require detailed counseling that includes information on the imprecise recurrence risks and a plan for subsequent pregnancies.


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 determine whether code status, advance directives, and decisions to limit life support were different for patients with limited English proficiency (LEP) in the intensive care unit (ICU) as compared with patients whose primary language was English. PATIENTS AND METHODS: We conducted a retrospective cohort study in adult patients admitted to 7 ICUs in a single tertiary academic medical center from May 31, 2011, through June 1, 2014. RESULTS: Of the 27,523 patients admitted to the ICU, 779 (2.8%) had LEP. When adjusted for severity of illness, sex, education level, and insurance status, patients with LEP were less likely to change their code status from full code to do not resuscitate during ICU admission (odds ratio [OR], 0.62; 95% CI, 0.46-0.82; P<.001) and took 3.8 days (95% CI, 1.9-5.6 days; P<.001) longer to change to do not resuscitate. Patients with LEP who died in the ICU were less likely to receive a comfort measures order set (OR, 0.38; 95% CI, 0.16-0.91; P=.03) and took 19.1 days (95% CI, 13.2-25.1 days; P<.001) longer to transition to comfort measures only. Patients with LEP were less likely to have an advance directive (OR, 0.23; 95% CI, 0.18-0.29; P<.001), more likely to receive mechanical ventilation (OR, 1.26; 95% CI, 1.07-1.48; P=.005), and more likely to have restraints used (OR, 1.36; 95% CI, 1.11-1.65; P=.003). The hospital length of stay was 2.7 days longer for patients with LEP. Additional adjustment for religion, race, and age yielded similar results. CONCLUSION: There are important differences in end-of-life care and decision making for patients with LEP.


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.


PROBLEM: Little is known about how parents perceive their role or the role of health care providers (HCPs) during end-of-life decision making (EOL DM) in the context of the pediatric intensive care unit (PICU). ELIGIBILITY CRITERIA: The authors searched CINAHL, PubMed, Ovid Medline, Web of Science, Social Science Database, PsycINFO, and Google scholar for English language studies performed in the United States related to parental perception of parental or HCP roles in EOL DM in the PICU since 2008. SAMPLE: Eleven studies of parents and health care providers (HCPs) of
critically ill children in the PICU and/or receiving inpatient pediatric palliative care, and bereaved parents of PICU patients. RESULTS: Most parents reported belief that EOL DM is within the domain of parental role, a minority felt it was a physician's responsibility. Parental EOL DM is rooted more firmly in emotion and perception and a desire to be a 'good parent' to a child at EOL in the way they see fit than HCP recommendations or 'medical facts'. Parents need HCPs to treat them as allies, communicate well, and be trustworthy. CONCLUSIONS: Role conflict may exist between parents and HCPs who are prioritizing different attributes of the parental role. The role of the nurse in support of parental role in the PICU is not well-elucidated in the extant literature.

IMPLICATIONS: Future research should focus on what parents need from HCPs, especially nurses, to support their parental role, and factors that facilitate the development of trust and good communication.


A pregnancy following a perinatal bereavement has a rather particular dimension to it. It requires attentive support and monitoring in order to enable the future baby to take its place within the family. An interview with Marie-Jose Soubieux, child psychiatrist and psychoanalyst.


BACKGROUND: As understandings of the impacts of end-of-life experiences on parents' grief and bereavement increase, so too does the inclusion of bereaved parents into research studies exploring these experiences. However, designing and obtaining approval for these studies can be difficult, as guidance derived from bereaved parents' experiences of the research process are limited within the current literature. METHODS: We aimed to explore bereaved parents' experiences of research participation in a larger grounded theory study exploring experiences of the death of a child in the paediatric intensive care unit. Data were obtained during follow-up phone calls made to 19 bereaved parents, five of whom provided data from their spouse, 1 week after their participation in the study. Participants were asked to reflect on their experiences of research participation, with a focus on recruitment methods, timing of research contact, and the location of their interview. Parents' responses were analysed using descriptive content analysis. RESULTS: Our findings demonstrate that despite being emotionally difficult, parents' overall experiences of research participation were positive. Parents preferred to be contacted initially via a letter, with an opt in approach viewed most favourably. Most commonly, participants preferred that research contact occurred within 12-24 months after their child's death, with some suggesting contact after 6 months was also appropriate. Parents also preferred research interviews conducted in their own homes, though flexibility and parental choice was crucial. CONCLUSIONS: Findings from this study offer further insight to researchers and research review committees, to help ensure that future studies are conducted in a way that best meets the unique needs of bereaved parents participating in research.


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.


This paper reports on a study that examined the grief and coping of 29 parents whose child has hypoplastic left heart syndrome using the Dual Process Model. The study employed a secondary thematic analysis of interviews at key times of treatment and recovery for the child. After the diagnosis, parents experienced intense loss (LO), but focused upon restoration-orientated tasks (RO) to support their child. Over time, most parents employed a healthy oscillation between LO coping and RO coping, with waves of grief and with some grieving suppressed. There are some specific grief and coping and gender patterns employed by parents.


BACKGROUND: There is few literature on the difficulties and different meanings of gastrostomy tubes (GST) for parents of children with palliative needs, and what specific palliative care teams contribute to this process. AIM: To explore the process of information in the decision of performing a gastrostomy and the meanings that parents of children with palliative needs build around them. DESIGN: Semi-structured interviews which were transcribed and analysed using Grounded Theory. SETTING/PARTICIPANTS: Parents and caretakers of children admitted in Paediatric Palliative Care Unit of Madrid Autonomous Community (Spain) whose children bore a gastrostomy device. RESULTS: Two core categories arise (‘Fight’ and ‘The child as a life-meaning generator’). In all the cases, the child supplied the meaning to go on, and the Palliative Care Unit (CPU) helped in the daily care of the child and solving problems derived from the handling of the GT. CONCLUSIONS: It is necessary to improve the process of giving bad news and to introduce models of health care that focus on parents and child as the center of palliative care. It is also necessary to develop educational programs that enable continuity of care at home for children with palliative needs.

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.


When the premature death of a baby occurs in a family, brothers and sisters need support. Their silence, their absence of questions or reactions must set off alarm bells. The parents, beyond their own bereavement, have a key role to play in enabling their children to integrate this loss into their lives.


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 life day 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.


PURPOSE: Family caregivers of children with cancer face emotional, psychological, and spiritual challenges coping with their child's illness. For ensuring comprehensive multidisciplinary pediatric care, there is a need to understand and define what spirituality means for them in relation to their
child's illness. The purpose of this study is to understand the meaning of spirituality for parents of cancer patients in Lebanon. METHODS: This qualitative study followed the Heideggerian interpretive phenomenological method. Through purposeful sampling, 11 parents (mother or father) of children with cancer receiving treatment at a tertiary care center in Beirut, Lebanon were interviewed. Data were analyzed following the hermeneutical process as described by Diekelmann and Ironside (1998). RESULTS: A constitutive pattern and overarching theme, "spirituality is a two-level relationship. It is a relation with God and with people. It is the act of receiving and giving back" and five major themes emerged from the data. These were "Being there for me; " "Connectedness with other parents is a blessing and a torment; " "The power of knowing; " "Communication with Unknown" and "Spirituality is not religiosity". CONCLUSION: Lebanese parents of children with cancer defined the elements of their own spirituality. Relational aspects dominated and communication was an important factor. IMPLICATIONS FOR PRACTICE: This is the first study in the Middle East to address the meaning of spirituality in this population, and would pave the way for a customized palliative care program and integrative approach to patient care. 


To understand the lived experience of parents who have lost their child to a chronic life-limiting condition, six major databases were searched by adhering to the Preferred Reporting Items for Systematic Review and Meta-Analyses guidelines. Articles were screened for appropriateness using the Sample, Phenomenon of Interest, Design, Evaluation, Research type tool, and relevant qualitative studies were selected for full-text data analysis using Thematic Synthesis. Findings were categorized into 13 themes that were further organized into a four-phase trajectory of parental bereavement experience of child loss, namely: Liminal Margin, Holding Space, Navigating Losses, and Reconstructing Lives. The findings are discussed in the light of existing literature with practical recommendations for enhancing parental bereavement support services. 


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. 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.


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.


Ethical dilemmas in critical care may cause healthcare practitioners to experience moral distress: incoherence between what one believes to be best and what occurs. Given that paediatric decision-making typically involves parents, we propose that parents can also experience moral distress when faced with making value-laden decisions in the neonatal intensive care unit. We propose a new concept—that parents may experience "moral schism"—a genuine uncertainty regarding a value-based decision that is accompanied by emotional distress. Schism, unlike moral distress, is not caused by barriers to making and executing a decision that is deemed to be best by the decision-makers but rather an encounter of significant internal struggle. We explore factors that appear to contribute to both moral distress and "moral schism" for parents: the degree of available support, a sense of coherence of the situation, and a sense of responsibility. We propose that moral schism is an underappreciated concept that needs to be explicated and may be more
prevalent than moral distress when exploring decision-making experiences for parents. We also suggest actions of healthcare providers that may help minimize parental "moral schism" and moral distress.


BACKGROUND: Identifying characteristics of individuals at greatest risk for prolonged grief disorder (PGD) can improve its detection and elucidate the etiology of the disorder. The Safe Passage Study, a study of women at high risk for sudden infant death syndrome (SIDS), prospectively examined the psychosocial functioning of women while monitoring their healthy pregnancies. Mothers whose infants died of SIDS were followed in bereavement. METHODS: Pre-loss data were collected from 12,000 pregnant mothers and analyzed for their associations with grief symptoms and PGD in 50 mothers whose infants died from SIDS, from 2 to 48 months after their infant's death, focusing on pre-loss risk factors of anxiety, depression, alcohol use, maternal age, the presence of other living children in the home, and previous child loss. RESULTS: The presence of any four risk factors significantly predicted PGD for 24 months post-loss (p < 0.003); 2-3 risk factors predicted PGD for 12 months (p = 0.02). PGD rates increased in the second post-loss year, converging in all groups to approximately 40% by 3 years. Pre-loss depressive symptoms were significantly associated with PGD. Higher alcohol intake and older maternal age were consistently positively associated with PGD. Predicted risk scores showed good discrimination between PGD and no PGD 6-24 months after loss (C-statistic = 0.83). CONCLUSIONS: A combination of personal risk factors predicted PGD in 2 years of bereavement. There is a convergence of risk groups to high rates at 2-3 years, marked by increased PGD rates in mothers at low risk. The risk factors showed different effects on PGD.


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.


There is a need for guiding theory to understand the experiences and outcomes of bereaved siblings, particularly from a family systems framework. The present study investigated the relevance of emotional security theory in a sample of 72 young adults who experienced sibling bereavement. We investigated (1) whether perceptions of prolonged parental grief predicted key aspects of emotional security (disengagement, preoccupation, and security), and (2) whether emotional security mediated a relation between perceptions of prolonged parental grief and young adult emotional functioning. Results supported the potential utility of emotional security theory as a theoretical framework for understanding sibling bereavement.


Young children construct a biological conception of death, recognizing that death terminates mental and bodily processes. Despite this recognition, many children are receptive to an alternative conception of death, which affirms that the deceased has an afterlife elsewhere. A plausible interpretation of children's receptivity to this alternative conception is that human beings, including young children, are naturally disposed to remember and keep in mind individuals to whom they are attached even when those individuals leave and are absent for extended periods. This disposition is reflected in the pervasive tendency to talk about death as a departure rather than a terminus. It also enables the living to sustain their ties to the dead, even if, in the case of death, the departure is permanent rather than temporary. Linguistic and developmental evidence for these claims is reviewed. Possible biological origins and implications for archaeological research are also discussed. This article is part of the theme issue 'Evolutionary thanatology: impacts of the dead on the living in humans and other animals'.


Perinatal loss is one of the most devastating events a family can experience. This practice point focuses on circumstances that are likely to involve paediatric health care professionals. Recommendations are provided for compassionate communication, bereavement, sibling care and counselling to support families.


BACKGROUND AND AIMS:: Children with severe spinal muscular atrophy have complex care needs due to progressive muscle weakness, eventually leading to respiratory failure. To design a care system adapted to families' needs, more knowledge about parents' experience of care and its coordination between settings is required. This study explores (1) whether parents felt that health professionals took every opportunity to help the child feel as good as possible, (2) parents'
satisfaction with various care settings, and (3) parents' satisfaction with coordination between settings. METHODS:: Data derive from nationwide Swedish and Danish surveys of bereaved and nonbereaved parents of children with severe spinal muscular atrophy born between 2000 and 2010 in Sweden and 2003 and 2013 in Denmark (N = 95, response rate = 84%). Descriptive statistics and content analysis were used. RESULTS:: Although most of the parents reported that care professionals had taken every opportunity to help the child feel as good as possible, one-third reported the opposite. Bereaved parents were significantly more satisfied with care than nonbereaved (81% vs 29%). The children received care at many different locations, for all of which parents rated high satisfaction. However, some were dissatisfied with care coordination, describing lack of knowledge and communication among staff, and how they as parents had to take the initiative in care management. CONCLUSIONS:: This study highlights the importance of improving disease-specific competence, communication and knowledge exchange among staff. For optimal care for these children and families, parents should be included in dialogues on care and staff should be more proactive and take care management initiatives.


Communication plays an essential role in social relationships. Yet it is unclear how young cancer patients and survivors communicate with peers, and whether this contributes to increased rates of social difficulties. We aimed to analyze how childhood cancer patients and survivors communicate about their cancer with family and peers. We systematically searched Medline, Embase and PsycINFO for peer-reviewed studies on cancer-related communication among patients and survivors (any cancer, <25 years at diagnosis). We screened 309 articles, and included 6 qualitative studies. Studies were assessed using a standardized quality assessment tool. Participants were adolescents and young adults, 16-34 years of age at the time of study. Included studies related to different forms of cancer-related communication, benefits, and challenges. We found that cancer-related communication was an individual, complex process, addressing medical, existential, and emotional aspects of cancer. Communication occurred on a spectrum with variation in who information was shared with, as well as differences in the frequency at which information was shared, and the amount and type of information shared. Communication often occurred at uncertain or significant times for participants, or was initiated by others. Communicating about cancer yielded benefits as a coping strategy, prompted social support, and appeared central to significant relationships. Barriers to communication, including fear of stigma and poor peer reactions, hindered willingness to disclose. The number of studies analyzing this topic was limited. Communicating about cancer is a significant yet complex process for young patients and survivors. Further research is needed to complement the existing literature and to establish the evidence base for the development of future effective interventions promoting social and communication skills.
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.

PURPOSE: Grandparents can be profoundly emotionally affected when a grandchild is diagnosed with cancer. They also often provide invaluable support for the family (e.g., caring for the sick child and/or siblings). Multigenerational family functioning may therefore change. Limited research has assessed grandparents' perspectives after their grandchild is diagnosed with cancer. In this study, we aimed to (1) assess differences in perceived family functioning among grandparents of a child with cancer and grandparents of healthy children and (2) assess the cancer-specific and demographic factors related to perceived family functioning in grandparents of a grandchild with cancer. PROCEDURE: Grandparents of a child with cancer (n = 89) and grandparents of healthy children (n = 133) completed the general functioning, communication, and problem-solving scales of the Family Assessment Device. We used multilevel models with a random intercept to detect (1) between-group differences and (2) identify factors related to perceived family functioning among grandparents with a grandchild with cancer. RESULTS: Grandparents with a grandchild with cancer reported poorer family functioning than grandparents with healthy grandchildren. Among the grandparents with a grandchild with cancer, impairments in family functioning were correlated with fewer years since diagnosis, providing care to their sick grandchild and/or siblings and living far away from the sick grandchild. CONCLUSIONS: The detrimental impact of childhood cancer likely extends beyond the immediate family members. Including grandparents in interventions—beginning at diagnosis—to reduce distress and increase cohesion for families of a child with cancer is warranted, particularly for grandparents who provide care to their sick grandchild or siblings.

Individuals with chronic illnesses must manage long-term uncertainty as they cope with the ways the illness influences their lives. In the context of pediatric illnesses, parents must manage uncertainty during the diagnosis and treatment of their child’s illness. It is common for children with complex chronic illnesses to see multiple specialists for the treatment of their condition. While
previous research has explored parents' uncertainty during a child's diagnosis and during end-of-life care, less is known about these experiences when the child is referred to a team of specialists for treatment. The aim of the current study was to explore how specialists, as credible authorities, influence parents’ uncertainty during parents’ first visit to a multidisciplinary clinic for the care of their child’s complex chronic illness. Data were collected through semi-structured interviews with 29 parents after their child's first visit to a vascular anomaly clinic at a large Midwestern children’s hospital. The results suggest parents’ communication with credible authorities facilitates effective uncertainty management primarily through the mechanism of uncertainty reappraisal. The results also suggest that specialists, as credible authorities, are a key mechanism in the appraisal of uncertainty for conditions that are often misdiagnosed and mismanaged.


Parents hit by perinatal bereavement are often confronted with misunderstanding and awkwardness on the part of those around them. However, they need to talk, to be listened to and respected in their grief from the moment the death is announced. Support from Agapa association enables them to talk about their child, to break the isolation in which they find themselves, and thereby move forward along the path of bereavement and reconstruction.


Advancing technology in reproductive medicine has led to more frequent perinatal diagnoses of fatal or life-limiting anomaly. For those parents who choose to continue pregnancies while facing such a diagnosis, compassionate and communal perinatal palliative care provides beneficial physical and psycho-socio-emotional support for these families, so that they may preserve and acknowledge the relationship with their child, no matter how brief his/her life.


This study aims to identify parenting experiences after the death of a child. Using interpretive phenomenological analysis, we mapped the experiences of 16 parents with school-aged surviving children after the death of their sibling to the 2014 Sewol ferry disaster in South Korea. Interviews illuminate five master themes of parenting surviving children following a child's death: (a) parental anxiety, (b) conflicts and obstacles in the parent-child relationship, (c) changes in parenting style, (d) striving to support children's grief, and (e) seeking outside help for parenting. Implications for supporting grieving parents and their children are discussed in light of the findings.
OBJECTIVE: To determine the preliminary feasibility, acceptability, and effects of Meaning-Centered Grief Therapy (MCGT) for parents who lost a child to cancer. METHOD: Parents who lost a child to cancer and who were between six months and six years after loss and reporting elevated levels of prolonged grief were enrolled in open trials of MCGT, a manualized, one-on-one cognitive-behavioral-existential intervention that used psychoeducation, experiential exercises, and structured discussion to explore themes related to meaning, identity, purpose, and legacy. Parents completed 16 weekly sessions, 60-90 minutes in length, either in person or through videoconferencing. Parents were administered measures of prolonged grief disorder symptoms, meaning in life, and other assessments of psychological adjustment preintervention, mid-intervention, postintervention, and at three months postintervention. Descriptive data from both the in-person and videoconferencing open trial were pooled. Result: Eight of 11 (72%) enrolled parents started the MCGT intervention, and six of eight (75%) participants completed all 16 sessions. Participants provided positive feedback about MCGT. Results showed postintervention longitudinal improvements in prolonged grief (d = 1.70), sense of meaning (d = 2.11), depression (d = 0.84), hopelessness (d = 1.01), continuing bonds with their child (d = 1.26), posttraumatic growth (ds = 0.29-1.33), positive affect (d = 0.99), and various health-related quality of life domains (d = 0.46-0.71). Most treatment gains were either maintained or increased at the three-month follow-up assessment. Significance of results: Overall, preliminary data suggest that this 16-session, manualized cognitive-behavioral-existential intervention is feasible, acceptable, and associated with transdiagnostic improvements in psychological functioning among parents who have lost a child to cancer. Future research should examine MCGT with a larger sample in a randomized controlled trial.


In the last century, decreases in infant and child mortality, urbanization and increases in healthcare efficacy have reduced children's personal exposure to death and dying. So how do children acquire accurate conceptions of death in this context? In this paper, we discuss three sources of children's learning about death and dying, namely, direct experience of death, parental communication about death and portrayals of death in the media and the arts. We conclude with recommendations about how best to teach modern children about this aspect of life. This article is part of the theme issue 'Evolutionary thanatology: impacts of the dead on the living in humans and other animals'.


A child’s death is a traumatic life experience for parents. Health-care professionals (HCPs) have sought guidance on how to intervene with grieving parents, particularly with fathers. Having therapeutic conversations is an effective way for HCPs to support grieving fathers. In our previous study, fathers identified core beliefs that influenced their experience of grief and coping. In this article, the Illness Beliefs Model was integrated with the findings to provide a framework for interventions to create open conversations, ease fathers’ suffering, and thereby help their spouse and family suffering as well. This article will guide HCPs to engage in therapeutic conversations to support bereaved fathers.


Background: When a mother loses a baby after the period of viability, there is no way to fathom her grief, neither any words, nor an explanation. It is an unexpected event. Stillbirth presents a situation where the early activation of the grief process primarily in mother is exacerbated by the circumstances surrounding the loss. It thus becomes imperative for the healthcare providers to evaluate the significance of parent’s perception on the loss and the factors contributing to it before the initiation of therapy. Objective: To evaluate the psychosocial impact of stillbirth among mothers and its contributing factors. Materials and Methods: A WHO-funded prospective study was conducted in VMMC and Safdarjung Hospital from September 2015 to August 2016 on all women who gave birth to a stillborn baby, using a questionnaire based on EPDS, after taking their written informed consent. Data were entered on the predesigned proforma and analyzed after applying Chi-square test, keeping a null hypothesis value of 15% for all the variables. Results: Out of the 709 women who delivered stillborn babies, 645 respondents, who willingly consented to participate, were included in the study. There was a significant relationship between psychosocial impact after perinatal loss and support from caregiver and family. Conclusion: Mothers with stillborn fetuses should be screened for psychosocial impact and offered support when needed. Appropriate counseling by healthcare providers and continued psychosocial and emotional support by family members must be provided.


Adolescents and young adults (AYAs) with cancer aged 15-39 years have unique medical, psychosocial, and informational needs. At the time of diagnosis, they are often going through important life milestones, such as establishing their independence, attending school or work, and maintaining romantic and/or family relationships. This article describes some of the critical time points for AYAs with cancer and the resources available to support the nursing profession in meeting the unique care needs of this population.

Research demonstrates that severe forms of grief and grief-related pathology exist in the general population. Less attention, however, has been paid to the grief of parents following the death of a young, dependent child. In this review, we summarize a search of Pubmed, PsycINFO and Web of Science from 1995 to 2017, using the terms ‘parental complicated grief’, ‘parental traumatic grief’, and ‘parent Prolonged Grief Disorder’, specifically addressing parental grief and identified risk factors for complicated or prolonged grief. Forty-two studies met criteria and indicate a significant burden of complicated or prolonged grief in parents of children dying from virtually any cause. It appears that the empiric literature is undermined by great variability, including the composition of samples, the causes of death studied, the psychometric measures used, and post-loss intervals. We conclude that the uniform severity of grief experiences following the death of a young child is potentially a distinct subtype of grief, deserving of attention in its own right in future research and diagnostic formulations.


Objective: Serious childhood illness is associated with significant parent psychological distress. This study aimed to (a) document acute and posttraumatic stress symptoms (PTSS) in parents of children with various life-threatening illnesses; (b) identify trajectory patterns of parental PTSS and recovery over 18 months; (c) determine psychosocial, demographic, and illness factors associated with trajectory group membership. Methods: In total, 159 parents (115 mothers, 44 fathers) from 122 families participated in a prospective, longitudinal study that assessed parent psychological responses across four time points—at diagnosis, and 3, 6, and 18 months later. Children were admitted to the Cardiology, Oncology, and Pediatric Intensive Care Departments in a tertiary pediatric hospital. The primary outcome was parent PTSS. Results: Three distinct parent recovery profiles were identified—“Resilient,” “Recovery,” and “Chronic.” The “Resilient” class (33%) showed low distress responses across the trajectory period, whereas the “Recovery” class (52%) showed significantly higher levels of distress at the time of diagnosis that gradually declined over the first months following their child’s illness. Both of these classes nevertheless remained within the normative range throughout. In contrast, the “Chronic” class (13%) was consistently high in severity, remaining within the clinical range across the entire period. Psychosocial factors such as mood, anxiety, and emotional responses predicted group membership, whereas demographic and illness factors did not. Conclusions: Parents show considerable resilience in the face of children’s life-threatening illnesses. Early assessment of parent psychosocial factors may aid identification of those who would benefit from early intervention.


The death of a significant person, especially when it comes tragically or prematurely, can shake the foundations of our assumptive and relational world and lead to anguished attempts to find meaning in the loss and in our lives in its aftermath. In this article, I review one program of research focused on this attempt at meaning reconstruction, describe recently developed measures of meaning in mourning, and discuss several therapeutic techniques for helping clients make sense of the death and rework their attachment relationship to the deceased. I conclude by illustrating some of this work in my therapy with a couple grieving the loss of not one but two children to tragic accidents, as they try to adapt to a compound traumatic bereavement.


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.


In recent years, clinical approaches to anticipatory grief and inclusivity amongst the medical team and family members have grown. In thinking about the end-of-life concerns within the pediatric care setting, practice concepts, and innovations inform how physicians and members of the interdisciplinary care team choose to approach conversations with parents and family members, as well as the particular level of involvement parents should have in decisions regarding the end of their child’s life.


PURPOSE: Young adults (YAs) aged 18-35 years with cancer often experience unmet psychosocial needs. We aimed to evaluate a conversation aid (“Snapshot”) that offered a framework for discussing YA-specific psychosocial concerns between patients and clinicians. METHODS: We developed and implemented Snapshot between 2014 and 2016 as part of a quality improvement initiative at Dana-Farber Cancer Institute. We extracted pre- and postimplementation data from chart documentation of psychosocial concerns. YAs and social workers provided qualitative feedback on the use of Snapshot in clinical care. RESULTS: Postintervention chart reviews revealed a significant increase in the median number of topics documented in charts after implementation of Snapshot (preintervention median = 9 [range: 1-15] vs. postintervention median = 11 [range 6-15]; p = 0.003). Overall, YAs and social workers reported that using Snapshot improved communication and consistency of psychosocial care, with documented improvement in the following domains: understanding illness (p < 0.001), sexuality and intimacy (p = 0.03), symptom burden (p = 0.003), care planning (p < 0.001), support for caregivers and children (p = 0.02), and social, work, and home changes (p = 0.05). CONCLUSION: Snapshot improved the quality of psychosocial needs assessment among YAs with cancer. Implementation was successful in reducing variability identified in the preintervention cohort and increasing the number of YA-specific psychosocial topics discussed. A standardized conversation aid has the potential to improve quality of care for YAs by enabling early identification and intervention of psychosocial issues for all patients.


OBJECTIVE: To classify NICU interventions for parental distress and quantify their effectiveness. STUDY DESIGN: We systematically reviewed controlled studies published before 2017 measuring NICU parental distress, defined broad intervention categories, and used random-effects meta-analysis to quantify treatment effectiveness. RESULTS: Among 1643 unique records, 58 eligible trials predominantly studied mothers of preterm infants. Interventions tested in 22 randomized trials decreased parental distress (p < 0.001) and demonstrated improvement beyond 6 months (p < 0.005). In subgroup analyses, complementary/alternative medicine and family-centered instruction interventions each decreased distress symptoms (p < 0.01), with fathers and mothers improving to similar extents. Most psychotherapy studies decreased distress individually but did not qualify for meta-analysis as a group. CONCLUSION: NICU interventions modestly reduced parental distress. We identified family-centered instruction as a target for implementation and complementary/alternative medicine as a target for further study. Investigators must develop psychosocial interventions that serve NICU parents at large, including fathers and parents of full-term infants.


Today every aspect of our life is published and shared online, including grief. The virtual cemeteries and social networks' use could be considered as a new modern mortuary ritual. Starting from the keyword stillbirth, 50 videos published on YouTube since 2008 have been analyzed qualitatively. The videos, 70% published by the mother, with an average length of 5.52 minutes, a mean of 2,429,576 views and 2,563 of comments, follow a sort of script: the second part with black and white photos, background music, and religious references. Could the continuous access to the child's technological grave encourage a complicated grief or be a support, given by the interaction with users, limiting the sense of isolation. The parent shows his or her own conceptions about death and, as a modern baptism, presents the child to the whole society. Videos keep child's memory alive and fuel a process of personalization and tenderness in the user.


This study aimed to provide a better understanding of the medical decision-making preferences and experiences of young adult survivors of pediatric, adolescent, and young adult cancers. We conducted key informant interviews and a cross-sectional mailed survey with young adult survivors (currently aged 18-39 years) of pediatric, adolescent, and young adult cancers in South Texas. Of the responding survivors, almost all wanted to be actively involved in medical decision-making, but preferences regarding family and doctor involvement varied. In open-ended responses, the most commonly reported concerns related to medical decision-making were feelings of uncertainty and fear of receiving bad news. Survivors reported that they desired more information in order to feel better about medical decision-making. Due to the variety of preferences regarding decision-making and who to include in the process, physicians should be prepared to ask and accommodate patients regarding their decision-making preferences.


PURPOSE: Childhood chronic conditions have a considerable effect on the quality of life (QoL) of pediatric patients and their caregivers. The purpose of this meta-analysis was to evaluate the effects of caregiver-involved interventions on the QoL of children and adolescents with chronic conditions and their caregivers. METHODS: The PubMed, EMBASE, Web of Science, Cumulative Index of Nursing and Allied Health Literature, Academic Search Complete, Education Resource Information Center, and PsycINFO databases were searched for published randomized controlled trials from inception to April 2016. Two reviewers (NS and JM) independently screened included studies and assessed study quality. The meta-analyses and meta-regressions using random-effects models were performed with the Comprehensive Meta-analysis software (version 3, Biostat, Englewood, NJ). RESULTS: Fifty-four studies involving 10075 pediatric patients diagnosed with asthma, diabetes, cancer, hypersensitivity, cerebral palsy, arthritis, or sickle cell diseases and 10015 caregivers were included in our analysis. The interventions mainly involved education about disease, skill training, environment change, psychological intervention, physical exercise,
experience sharing, monitoring, or social support. The results demonstrated that caregiver-involved interventions significantly improved the health-related QoL (HRQoL) of caregivers [standardized mean difference (SMD) = 0.26, 95% CI 0.14-0.38, p < 0.001], particularly those delivered through the face-to-face mode (SMD = 0.32, 95% CI 0.21-0.43, p < 0.001). However, no improvements in the QoL (SMD = 0.00, 95% CI -0.22 to 0.22, p = 1.00) and HRQoL (SMD = 0.06, 95% CI -0.02 to 0.14, p = 0.16) of children and both caregivers and children (SMD = 0.04, 95% CI -0.08 to 0.17, p = 0.52) were observed. CONCLUSIONS: This meta-analysis provides evidence on the positive effects of caregiver-involved interventions on the HRQoL of caregivers. Moreover, face-to-face mode is the delivery approach with a promising effect on the HRQoL of caregivers. Further research on conditions not found in this review is warranted.


BACKGROUND: Despite growing evidence and support for shared decision making, little is known about the experiences of parents who hold more active roles than they wish. METHODS: This was a prospective cohort study of 372 parents of children with cancer and their oncologists at 2 academic pediatric hospitals. Parents were surveyed within 12 weeks of the diagnosis, and they were assessed for associated factors and outcomes of holding a more active decision-making role than they preferred. Parents were asked about their preferred and actual roles in decision making. Oncologists were asked to estimate parental preferences. RESULTS: Most parents preferred to share decision making with the oncologist (64% [236 of 372]); however, 13% (49 of 372) preferred oncologist-led decision making. Most parents fulfilled their ideal decision-making role (66% [244 of 372]), but a notable minority were either more involved (14% [52 of 372]) or less involved than they preferred (20% [76 of 372]; P < .0001 [McNemar test]). Oncologists recognized parents’ preferred roles in 49% of cases (167 of 341); 24% (82 of 341) of parents preferred more active roles than the oncologist recognized, and 27% (92 of 341) preferred less active roles than recognized. No parent or communication characteristics were found that were associated with parents’ holding a more active role than desired in decision making. Parents who held more active roles in decision making than they wished had higher odds of decisional regret (odds ratio, 3.75; 95% confidence interval, 2.07-6.80; P < .0001). CONCLUSIONS: Although many parents fulfill their desired roles in decision making about their child’s cancer, some are asked to take on more active roles than they wish. Holding a more active role than desired may lead to increased decisional regret.


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.


BACKGROUND: The impact of a child's life-limiting or life-threatening illness is significant on parents who experience a great deal of emotional, physical, and spiritual upheaval. Hope has been identified as an important inner resource for parental caregivers. Specifically, parental hope has been described as having four subprocesses including Accepting Reality, Establishing Control, Restructuring Hope, and Purposive Positive Thinking. PURPOSE: The purpose of this Delphi study was to gather expert opinions from parents and formal care providers about the four subprocesses essential to parental hope, to increase understanding of parental caregivers current support needs. As Phase one of a three-phase study, the findings provided direction in the development of a theory-based hope intervention. DESIGN AND METHODS: A Delphi study consisting of three rounds of survey questions and controlled feedback to experts was employed. Experts suggested strategies for each subprocess and ranked them in order of highest to lowest according to feasibility and effectiveness. RESULTS: Sixty-eight experts consisting of parental caregivers of children diagnosed with life-limiting or life-threatening illnesses and those who care for them (community members, nurses, social workers, and physicians) were recruited to participate. Through three rounds of survey questions, response rates ranged from 92-97%. A consensus revealed eight major themes that support parental hope: Organize Basic Needs; Connect with Others; Prioritize Self-care; Obtain Meaningful Information; Take Things Day by Day; Advocate for Parental Participation; Manifest Positivity; and Celebrate Milestones. PRACTICE IMPLICATIONS: This study identified a wide variety of psychosocial needs for parental caregivers. Results also offered direction for a theory-based hope intervention while highlighting the need for additional research in this area. These results will provide the foundation for a booklet parents can work through in their journey of caring for a child with a life-limiting or life-threatening illness.


OBJECTIVE: Health-related quality of life (HRQoL) concerns of adolescents and young adults (AYAs) aged 14-25 years were compared with those of older adults (26-60 years) with cancer. METHODS: AYAs and older adults receiving curative intent treatment or supportive palliative care for cancer were recruited from eight research centres across Europe. Participants used a rating scale to score the relevance and importance of a list of 77 issues covering 10 areas of HRQoL concern: symptoms; activity restrictions; social; emotional; body image; self-appraisals; outlook on life; lifestyle; treatment-related and life beyond treatment. RESULTS: HRQoL issues were reviewed by 33 AYAs and 25 older adults. Several issues were recognised as relevant and important across all age groups: symptoms, emotional impact, outlook on life, lifestyle and treatment-related. A number of issues were more relevant or important to AYAs including interrupted education, greater motivation to achieve academic goals, increased maturity, boredom, fertility and change in living situation. CONCLUSION: While there is overlap in several of the HRQoL concerns across the age span, it is important that HRQoL measures used with AYAs capture the diverse and unique psychosocial aspects of this developmental stage.


BACKGROUND: Bereaved parents may be at higher risk to develop persistent, severe and disabling grief, termed prolonged grief. Grief rumination, repetitive thinking about the causes and consequences of the loss, is a malleable cognitive process that maintains prolonged grief. Grief rumination can be measured with the Utrecht Grief Rumination Scale (UGRS). The present study aimed to examine the psychometric properties of the new Swedish version of the UGRS in a sample of bereaved parents. METHODS: A Swedish nationwide postal survey including measures of demographic and loss-related variables, grief rumination (UGRS), and symptoms of prolonged grief, posttraumatic stress, anxiety, depression, and insomnia, was completed by 226 parents (133 mothers and 93 fathers) who lost a child to cancer in the past five years. Psychometric properties of the UGRS were examined through confirmatory factor analyses (CFA), reliability analyses, and assessment of UGRS score associations with symptoms of prolonged grief, posttraumatic stress, depression, anxiety, and insomnia. RESULTS: The internal consistency of the Swedish UGRS was good. The CFA yielded an acceptable fit for a two-factor hierarchical model with five sub-factors. Grief rumination was positively associated with all psychopathology symptom measures. Higher scores on UGRS were found in parents with possible prolonged grief disorder compared to those without (d = 1.47). Moreover, the Swedish UGRS was associated with prolonged grief symptoms over and above loss-related and demographic variables and other psychopathology symptoms. CONCLUSIONS: The Swedish UGRS demonstrated good psychometric properties, which supports its use as a measure to assess grief rumination in Swedish bereaved parents in research and practice.


Traditionally, family-focused care extends to parents and siblings of children with life-limiting conditions. Only a few studies have focused on the needs of grandparents, who play an important role in the families of children with illness and with life-limiting conditions, in particular. Interpretative phenomenological analysis was used as the methodological framework for the study. Seven bereaved grandparents participated in this study. Semistructured, individual, face-to-face interviews were conducted. A number of contextual factors affected the experience of bereaved grandparents, including intergenerational bonds and perceived changes in role following the death of their grandchild. The primary motivation of grandparents stemmed from their role as a parent, not a grandparent. The breadth of pain experienced by grandparents was complicated by the multigenerational positions grandparents occupy within the family. Transition from before to after the death of a grandchild exacerbated the experience of pain. These findings about the unique footprint of grandparent grief suggest the development of family nursing practice to better understand and support grandparents during the illness of a grandchild, in addition to bereavement support.


AIM: To increase understanding of grandparental grief following the death of a grandchild from a life-limiting condition. DESIGN: Meta-ethnography. DATA SOURCES: Academic Search Complete CINHAL, Embase, psycINFO, PubMed and Web of Science, supplemented by manual search strategies (in 2015, updated 2018). REVIEW METHODS: Studies were appraised and synthesized using the principles of meta-ethnography. FINDINGS: Three superordinate themes were identified: 'influence of the relationship with their grandchild', 'influence of the relationship with the grandchild's family' and 'pain'. The simultaneous, multigenerational position of grandparents meant individuals experience emotional pain from witnessing the experience of family members. CONCLUSION: Many factors that contribute to the bereavement experience of grandparents are outside of their control. The roles, positions, and support needs of grandparents need to be acknowledged to better meet their needs as parents, grandparents, and individuals who have experienced a child death.


BACKGROUND: Cystic fibrosis (CF) is a chronic disease that has an impact on Health-Related Quality of Life (HRQoL). OBJECTIVES: To identify demographic and clinical factors associated with HRQoL in adolescents and young adults with CF. METHODS: The sample comprised adolescent and young adult patients with CF. They completed the Cystic Fibrosis Quality of Life (CFQoL)

questionnaire, which includes Physical, Social, Treatment, Chest Symptoms, Emotional Functioning, Future Concerns, Relationships, Body Image, and Career dimensions. We examined the relationships between gender, age, body weight, FEV1, pain, sleep, anxiety, depression and HRQoL.

RESULTS: The sample comprised 95 patients (aged 14-25 years; female/male: 43.1/56.8%). The lowest CFQoL score was observed in Future Concerns. FEV1 and body weight were positively associated with Physical Functioning (ss = 0.21; P < 0.01) and Body Image (ss = 0.30; P< 0.01), respectively. Females perceived themselves more negatively in Future Concerns (ss = -0.26; P< 0.01), Relationships (ss = -0.17; P< 0.01) and Career Concerns (ss = -0.20; P < 0.01) than males. Pain intensity (ss = -0.37), anxiety (ss = -0.39) and poor sleep quality (ss = -0.21) were negatively associated with global CFQoL (P < 0.001). CONCLUSIONS: Pain intensity, anxiety and quality of sleep have the broadest impact on HRQoL. Regular assessment of psycho-emotional functioning, quality of sleep and pain intensity may improve a patient's well-being.


OBJECTIVES: Spinal muscular atrophy (SMA) is a rare, hereditary, autosomal recessive neuromuscular disorder that, in its most severe forms, impacts infants and children. Once symptomatic, it is characterized clinically by a distinct inability to achieve motor milestones, such as the ability to lift the head, sit, stand, or walk. Quality of life (QOL) measurement in very young infants presents a particular challenge. Therefore, this review aims to highlight commonly used measurement tools and identifies future research opportunities for QOL measurement in SMA.

METHODS: A systematic literature review was carried out focusing on the various tools used to measure QOL in children < 18 years of age with formally diagnosed SMA type I, II, or III. Although the disease area of interest was SMA, data on Duchenne's muscular dystrophy were also included because of the rare nature of SMA. RESULTS: The Pediatric Quality of Life Inventory was the most commonly utilized tool to measure QOL in children; this included the generic and neuromuscular modules. No disease-specific tool to capture QOL in children with SMA was identified. Additionally, no measurement tools exist for very young infants (i.e., under 12 months) with SMA Type 1.

CONCLUSIONS: Evolving standards of care will lead to increased interest by stakeholders, on the methods used to measure QOL in infants and children across all types of SMA. Generic tools may not adequately capture QOL changes in SMA, especially given the age group affected by the disease. Further research is required to explore the scope for a disease-focused approach.


INTRODUCTION: Grief among bereaved parents is known to cause psychological distress and physical illness, but knowledge concerning factors that can contribute to health promotion after bereavement is scarce. Childhood cancer remains the most common non-accidental cause of death among children in Norway. The aim of the present study was to explore if resilience factors among cancer-bereaved parents could predict whether they will be able to come to terms with their grief 2-8 years following the loss. METHODS: A Norwegian cross-sectional national survey
was conducted among 161 cancer-bereaved parents using a study-specific questionnaire. Logistic regression was used to explore whether resilience factors predicted parents’ grief outcome 2-8 years after their loss. RESULTS: On the Resilience Scale for Adults (RSA), three of the resilience factors contributed significantly in predicting whether the parents in the present study would come to terms with their grief 2-8 years after the loss their child: "Perception of self" (OR 2.08, p = .048), "Social resources" (OR 2.83, p = .008) and "Family cohesion" (OR .41, p = .025). The results showed a negative relationship between time since loss (2-6 years) and whether the parents answered that they had come to terms with their grief (p = < .05). The loss of a parent (OR .30, p = .030) combined with the loss of their child had a negative and significant effect on whether they indicated that they had processed their grief. CONCLUSION: The total score of RSA and three of the six resilient factors contributed significantly in predicting whether cancer-bereaved parents in the present study indicated that they had come to terms with their grief to a great extent. The present study supports hypotheses that regard resilience as an important contribution in predicting healthy outcomes in people exposed to adverse life events.


After the death of my daughter Zoe in neonatal intensive care unit (NICU), a colleague asked me whether my status as an academic philosopher changed my experience in the NICU. In this short narrative, I outline 5 ways in which philosophical perspective helped me understand and cope with our hospital experience.


Music therapy and attachment is an expanding field and the number of studies addressing the theoretical work is slowly growing. There are both qualitative and quantitative approaches to studying the effect of regular music therapy sessions on parent-child interactions and these cover a range of patient populations including: children at risk of neglect, parents with a trauma history, children coping with bereavement and a large number addressing the disability population, including autism spectrum disorder. These studies suggest that music therapy benefits the parent-child relationship through the improvement communication, especially non-verbal communication, and so increased the feeling of closeness and understanding. Following a review of the available literature, a pilot study is described using transcripts of video recordings of music therapy sessions, and subsequent colour coding and conversion of the data into pie charts provides a potential method of analysis that produces an “interaction profile” of each parent-child dyad. Preliminary results of this method of analysis suggest that music therapy sessions might be able to improve interactions through therapists addressing the power dynamics within a relationship. The new method developed in this pilot study to visualise and study the parent-child relationship in music therapy sessions was effective and could be used and developed by music therapy researchers in the future.

https://www.ncbi.nlm.nih.gov/pubmed/30439844
BACKGROUND: Although patients in palliative care commonly report high emotional and spiritual needs, effective psychosocial treatments based on high quality studies are rare. First research provides evidence for benefits of psychosocial interventions in advanced cancer care. To specifically address end-of-life care requirements, life review techniques and creative-arts based therapies offer a promising potential. Therefore, the present study protocol presents a randomized controlled trial on the effectiveness of a newly developed music therapy technique that is based on a biographically meaningful song ("Song of Life"; SOL). METHODS: In a design with two parallel arms, 104 patients at two palliative care units will be randomly assigned to three sessions of either SOL (experimental group) or relaxation exercises (control group). Improvements in the psychological domain of quality of life will be the primary endpoint, while secondary outcomes encompass spiritual well-being, ego-integrity, overall quality of life, and distress. Additionally, caregivers will be asked to provide feedback about the treatment. Assessment of biopsychological stress markers and qualitative analysis of perceived strengths and weaknesses will complement data collection. DISCUSSION: Based on the results of a previous pilot study, we dedicated considerable efforts to optimizing the intervention and selecting appropriate outcomes for the present trial. We are confident to have designed a methodologically rigorous study that will contribute to the evidence-base and help to develop the potential of psychosocial interventions in palliative care. TRIAL REGISTRATION: German Clinical Trials Register (DRKS) - DRKS00015308 (date of registration: September 07th 2018).


Bereaved families fear their child being forgotten by those who knew their loved child, including their child’s oncology team. Thoughtfully timed, family-centric condolences shared by pediatric oncology team members have the potential to extend our compassion and kindness toward a family during the darkness of grief. Well-intended medical teams sometimes feel "at a loss" in terms of what to say to a grieving family and how or when to say it. This paper provides a tangible overview of written or verbal condolence communication in a format that can be personalized to the provider and the patient’s family.


Parents, after learning of a life-limiting fetal condition (LLFC), experience emotional distress and must consider options that impact the remainder of the pregnancy, their future lives, and family
members. For those who continue, little is known about their long-term presence or absence of regret about their choice, the reasons for this feeling, or its impact on their life. The aim of this research was to examine the concept of decision regret in parents who opted to continue a pregnancy affected by an LLFC. The contextual factors, conditions, and consequences surrounding the presence or absence of regret were analyzed. Data were retrieved from a cross-sectional study using the Quality of Perinatal Palliative Care and Parental Satisfaction Instrument. Participants were parents (N = 405) who experienced a life-limiting prenatal diagnosis and opted to continue their pregnancy. Secondary data analysis examined qualitative responses (121/402) to an item addressing regret. Dimensional analysis was used to examine data, identifying context, conditions, and consequences associated with the presence or absence of regret. Absence of regret was articulated in 97.5 percent of participants. Parents valued the baby as a part of their family and had opportunities to love, hold, meet, and cherish their child. Participants treasured the time together before and after the birth. Although emotionally difficult, parents articulated an empowering, transformative experience that lingers over time.

Services for Children and Families


Children with central nervous system (CNS) malignancies often suffer from high symptom burden and risk of death. Pediatric palliative care is a medical specialty, provided by an interdisciplinary team, which focuses on enhancing quality of life and minimizing suffering for children with life-threatening or life-limiting disease, and their families. Primary palliative care skills, which include basic symptom management, facilitation of goals-of-care discussions, and transition to hospice, can and should be developed by all providers of neuro-oncology care. This chapter will review the fundamentals of providing primary pediatric palliative care.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6315897/


INTRODUCTION: Early palliative care is recommended for cancer patients. However, palliative care consults (PCC) are often delayed in Lebanon. The aim of this study was to identify the factors associated with timing of PCC and their impact on the place of death. METHODS: This is a retrospective, single institution, study conducted at Hotel Dieu de France University Hospital in Lebanon. The clinical and demographic characteristics of oncology patients who received PCC were obtained. Cox and logistic regression models were used to evaluate the factors determining the time to first PCC and location of death, respectively. RESULTS: Two hundred and ten patients were included in our analyses with a median age of 69 years (range 22-92 years). The median survival times were: overall survival 18.7 months, time to first PCC 17.9 months, and survival post-PCC 0.6 months. Among patients who were followed-up at home, the median time spent at home was 0.6 months. Late PCC were associated with a childless status (HR = 0.57, 95%CI = 0.37-0.86, p = 0.007), awareness of the diagnosis (HR = 0.64, 95%CI = 0.45-0.91, p = 0.013), and lack of palliative home care (HR = 0.42, 95%CI = 0.25-0.65, p < 0.001). Older patients (OR = 1.03, 95%CI = 1.01-1.05, p = 0.026) and those who had been followed up at home during the PCC (OR = 160.56, 95%CI = 21.39-1205.50, p < 0.001) were significantly more likely to have died at home as opposed to the hospital. DISCUSSION: Cancer patients often receive PCC only shortly before their death. PCC for Lebanese cancer patients were found to be significantly delayed in patients that are childless, knowledgeable of their diagnosis, and lack home palliative care.


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.


Treatment for cystic fibrosis (CF) remains arduous and time-consuming, with young people in particular struggling to balance these demands with living a 'normal' life. Transferring to adult services is an important milestone that should be preceded by a gradual process of empowerment. This service evaluation aimed to explore the views of young people with CF before their transfer to adult care and to co-produce revisions to the transition and transfer programme. A total of 37 participants, aged 11-17 years, completed questionnaires during routine clinic visits with 81% expressing good knowledge of CF and treatment, and 59% reporting that they undertook their own treatment. Only 40% had seen a doctor alone for part of their clinic visit, 64% supported recruitment of a youth worker and 48% viewed dedicated adolescent clinics as beneficial. Participants expressed overall satisfaction with their care, however, improvements were suggested. Based on these suggestions, funding was secured for a youth worker, ‘transition’ clinics were established with children’s and adult CF team members, and doctors started seeing young people on their own for part of the clinic visit from age 13 years.


Palliative care (PC) aims to improve quality of life for patients and their families. The World Health Organization and American Academy of Pediatrics recommend that PC starts at diagnosis for children with cancer. This systematic review describes studies that reported PC timing in the pediatric oncology population. The following databases were searched: PubMed, Web of Science, CINAHL, and PsycInfo databases. Studies that reported time of PC initiation were independently screened and reviewed by 2 researchers. Studies describing pilot initiatives, published prior to 1998, not written in English, or providing no empirical time information on PC were excluded. Extracted data included sample characteristics and timing of PC discussion and initiation. Of 1120
identified citations, 16 articles met the inclusion criteria and comprised the study cohort. Overall, 54.5% of pediatric oncology patients received any palliative service prior to death. Data revealed PC discussion does not occur until late in the illness trajectory, and PC does not begin until close to time of death. Despite efforts to spur earlier initiation, many pediatric oncology patients do not receive any palliative care service, and those who do, predominantly receive it near the time of death. Delays occur both at first PC discussion and at PC initiation. Efforts for early PC integration must recognize the complex determinants of PC utilization across the illness timeline.


Background: Children with complex chronic conditions (CCCs) are dying at home with increased frequency, yet the number of studies on the financial feasibility of community-based pediatric palliative care is limited. Objective: The objectives of this study were to (1) describe characteristics of patients who died in a community-based palliative care program and (2) evaluate cost differences associated with participant characteristics and location of death. Design: A retrospective cohort analysis of administrative and electronic medical record data was employed. Setting/Subjects: Children enrolled in the community-based pediatric palliative care program, CompassionNet, who died between 2008 and 2015 were included (N = 224). Measurements: Demographic data, program expense, and paid claims were extracted from an insurance provider database and clinical data from the electronic medical record. Results: Sixty-six (29%) of the children were <1 year old at death; 80 (36%) were 1-9 years old, and 78 (35%) were 10-22 years old. Malignancy was the most common primary CCC diagnosis for the 158 children/adolescents (n = 89, 56%), whereas neuromuscular conditions (n = 20, 30%) were most frequent for infants. Death at home occurred 21% of the time for infants, 48% for children of ages 1-9 years, and 46% for children of ages 10-22 years. The mean total cost in the final year of life for pediatric patients was significantly related to location of death, a malignancy diagnosis, and participation in Medicaid. The largest estimated difference was between costs of care associated with death at home ($121,111) versus death in the hospital ($200,050). Conclusions: Multidisciplinary community-based pediatric palliative care teams provide the opportunity for a home death to be realized as desired. Significant cost differences associated with location of death may support program replication and sustainability.


Practices around the care of stillborn babies have evolved considerably over the last 15 years. Perinatal bereavement care requires a team approach to support the parents experiencing this ordeal. The place of rituals is important as is the personalisation of the care. The humanity which surrounds such moments constitutes the foundation on which the future equilibrium of these bereaved families will be based.

Over recent years, palliative care in maternity units has developed considerably. This is due to the evolution of legislation, medicine and requests from many parents, faced with a fatal prenatal diagnosis, to continue with the pregnancy and support their baby at birth. In parallel, the neonatal intensive care of extremely premature babies has improved significantly. Different situations can be concerned by the setting up of palliative care in maternity units. This specific support comprises significant challenges.


OBJECTIVES: The objective of this study was to describe the characteristics of pediatric palliative care (PPC) patients presenting to a pediatric emergency department (ED) and these patients’ ED visits. METHODS: This retrospective chart review was conducted from April 1, 2007, to March 31, 2012, in a tertiary care pediatric university-affiliated hospital. Eligible patients had initial PPC consultations during the study period; all ED visits by these patients were included. Data were drawn from the ED’s electronic data system and patient’s medical chart. RESULTS: A total of 290 new patients were followed by the PPC team, and 94 (32.4%) consulted the ED. Pediatric palliative care patients who consulted the ED had a median age of 7 years and baseline diagnoses of cancer (39.4%) or encephalopathy (27.7%). No patients died in the ED, but 36 (38.3%) died in hospital after an ED visit and 18 (19.1%) within 72 hours of admission. Pediatric palliative care patients consulted 219 times, with a median number of visits per patient of 2 (range, 1-8). They presented acutely ill as per triage scales. Reasons for consultation included respiratory distress/dyspnea (30.6%), pain (12.8%), seizure (11.4%), and fever (9.1%). Patients were often admitted to wards (61.2%) and the pediatric intensive care unit (7.3%). Two thirds (65.7%) of patients had signed an advanced care directive at the time of their visit. Discussions about goals of care occurred in 37.4% of visits. CONCLUSIONS: Pediatric palliative care patients present to the ED acutely ill, often at their end of life, and goals of care are not always discussed. This is a first step toward understanding how to improve PPC patients’ ED care.


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 objective was to characterize the relation between different sources of school-based social support (friends, peers, and teachers) and bereaved siblings' grief and grief-related growth and to examine whether nonparental sources of social support buffer the effects of low parent support on bereaved siblings. Families (N = 85) were recruited from cancer registries at 3 pediatric institutions 3-12 months after a child's death. Bereaved siblings were 8-18 years old (M = 12.39, SD = 2.65) and majority female (58%) and White (74%). During home visits, siblings reported their perceptions of social support from parental and nonparental sources using the Social Support Scale for Children, as well as grief and grief-related growth using the Hogan Sibling Inventory of Bereavement. Parent, friend, and teacher support were positively correlated with grief-related growth, whereas parent and peer support were negatively correlated with grief for adolescents. Teacher and friend support significantly moderated the association between parent support and grief such that teacher and friend support accentuated the positive effects of parent support. Friend and peer support moderated associations between parent support and grief/growth for adolescents but not children. School-based social support, namely from friends, peers, and teachers, appears to facilitate the adjustment of bereaved siblings. Findings suggest that bereaved siblings may benefit from enhanced support from teachers and friends regardless of age, with middle/high school students particularly benefitting from increased support from close friends and peers. (PsycINFO Database Record


BACKGROUND: The transition to adulthood has been described as a difficult time in the lives of young people with intellectual disability. There has been little emphasis on young people with severe or profound intellectual disability specifically, even though their pathways may differ, due to greater support needs across the life course. METHODS: A systematic review was conducted utilising Bronfenbrenner’s ecological model to inform framework analysis to synthesise qualitative findings. RESULTS: Taking an ecological perspective proved valuable. The transition process was described as stressful and barriers were identified across the ecological levels. Parents accounted for the majority of participants in studies, and the needs of young people and their parents emerged as highly interdependent. CONCLUSION: Themes reflect the complex nature of the
question what adulthood should look like for individuals with severe or profound intellectual disability. There is a lack of involvement of multiple stakeholders and young people themselves within studies.


CONTEXT: Approximately 500,000 children in the United States suffer from life-limiting illnesses each year, many of whom are hospice eligible each year. Few hospice agencies, however, offer formal pediatric programs. OBJECTIVE: To determine the levels of experience and comfort of hospice nurses who provide care to children and families in the community. METHODS: A cross-sectional survey was developed to assess hospice nurse experience/comfort across the domains of symptom management, end-of-life care, goals of care, family-centered care, and bereavement. The survey was pilot tested and distributed to hospice nurses across a tristate region. RESULTS: A total of 551 respondents across 71 hospices completed surveys. The majority of nurses reported no training in pediatric palliative or hospice care (89.8%), with approximately half reporting <5 years of hospice experience (53.7%) and no pediatric hospice experience (49.4%). Those with pediatric hospice experience reported limited opportunities to maintain or build their skills, with the majority providing care to children several times a year or less (85.7%). Nurses reported feeling somewhat or very uncomfortable providing services to children during the illness trajectory and at the end of life across all domains. CONCLUSION: Children with serious illness who receive care from local hospices often interface with nurses who lack training, experience, and comfort in the provision of palliative and hospice care to pediatric patients. These findings should inform future development and investigation of educational resources, training programs, and child- and family-centered policies to improve the delivery of palliative and hospice care to children in the community.


OBJECTIVES: To understand the experiences of young adults with cancer for whom cure is not likely, in particular what may be specific for people aged 16-40 years and how this might affect care. DESIGN: We used data from multiple sources (semi-structured interviews with people with cancer, nominated family members and healthcare professionals, and workshops) informed by a preliminary programme theory: realist analysis of data within these themes enabled revision of our theory. A realist logic of analysis explored contexts and mechanisms affecting outcomes of care. SETTING: Three cancer centres and associated palliative care services across England. PARTICIPANTS: We aimed for a purposive sample of 45 people with cancer from two groups: those aged 16-24 years for whom there may be specialist cancer centres and those 16-40 years cared for through general adult services; each could nominate for interview one family member and one
healthcare professional. We interviewed three people aged 16-24 years and 30 people 25-40 years diagnosed with cancer (carcinomas; blood cancers; sarcoma; central nervous system tumours) with a clinician-estimated prognosis of <12 months along with nominated family carers and healthcare professionals. 19 bereaved family members and 47 healthcare professionals participated in workshops. RESULTS: Data were available from 69 interviews (33 people with cancer, 14 family carers, 22 healthcare professionals) and six workshops. Qualitative analysis revealed seven key themes: loss of control; maintenance of normal life; continuity of care; support for professionals; support for families; importance of language chosen by professionals; and financial concerns. CONCLUSIONS: Current care towards end of life for young adults with cancer and their families does not meet needs and expectations. We identified challenges specific to those aged 16-40 years. The burden that care delivery imposes on healthcare professionals must be recognised. These findings can inform recommendations for measures to be incorporated into services. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6352841/


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

Objective: The objective of this study was to estimate palliative care needs and to describe the cohort of children with life-limiting illnesses (LLI) dying in hospitals. Design: This study was a retrospective cohort study. The national hospital admissions database was reviewed and children who had died who had life-limiting illnesses were identified. Setting: This study was conducted at Ministry of Health hospitals, Malaysia. Patients: Children aged 18 years and below who had died between January 1, 2012 and December 31, 2014. Main Outcome Measures: Life-limiting diagnoses based on Hain et al.’s directory of LLI or the ACT/RCPCH categories of life-limiting disease trajectories. Results: There were 8907 deaths and 3958 (44.4%) were that of children with LLI. The majority, 2531 (63.9%) of children with LLI were neonates, and the most common diagnosis was extreme prematurity <28 weeks with 676 children (26.7%). For the nonneonatal age group, the median age at admission was 42 months (1-216 months). A majority, 456 (32.0%) had diagnoses from the ICD-10 chapter "Neoplasms" followed by 360 (25.3%) who had a diagnoses from "Congenital malformations, deformations, and chromosomal abnormalities" and 139 (9.7%) with diagnoses from "Disease of the nervous system." While a majority of the terminal admissions were to the general ward, there were children from the nonneonatal age group, 202 (14.2%) who died in nonpediatric wards. Conclusion: Understanding the characteristics of children with LLI who die in hospitals could contribute toward a more efficient pediatric palliative care (PPC) service development. PPC service should include perinatal and neonatal palliative care. Palliative care education needs to extend to nonpediatric healthcare providers who also have to manage children with LLI.


BACKGROUND: Researchers have shown that most youth with special health care needs (YSHCN) are not receiving guidance on planning for health care transition. This study examines current transition planning among US youth with and without special health care needs (SHCN). METHODS: The 2016 National Survey of Children’s Health is nationally representative and includes 20 708 youth (12-17 years old). Parents and/or caregivers were asked if transition planning occurred, based on the following elements: (1) doctor or other health care provider (HCP) discussed the eventual shift to an HCP who cares for adults, (2) an HCP actively worked with youth to gain self-care skills or understand changes in health care at age 18, and (3) youth had time alone with an HCP during the last preventive visit. Sociodemographic and health system characteristics were assessed for associations with transition planning. RESULTS: Nationally, 17% of YSHCN and 14% of youth without SHCN met the overall transition measure. Older age (15-17 years) was the only sociodemographic factor associated with meeting the overall transition
measure and individual elements for YSHCN and youth without SHCN. Other sociodemographic characteristics associated with transition planning differed among the 2 populations. Receipt of care coordination and a written plan was associated with transition planning for YSHCN.

CONCLUSIONS: This study reveals that few youth with and without SHCN receive transition planning support. It underscores the need for HCPs to work with youth independently and in collaboration with parents and/or caregivers throughout adolescence to gain self-care skills and prepare for adult-focused care.


BACKGROUND: Research indicates that informal caregiving can have intense physical and mental impact on the individual. Relative to caregivers of adults, pediatric palliative caregivers appear less in literature despite experiencing greater mental, physical, financial, and social strain. There is limited research on the creation and evaluation of interventions specifically for this population despite clear need. OBJECTIVE: This study aims to evaluate the feasibility and engagement of the Photographs of Meaning Program, a modified meaning-making intervention for pediatric palliative caregivers. DESIGN: Participants completed a pre-post intervention meaning-in-life measure. Over a 9-week period, participants followed a meaning-making curriculum whereby they created and shared photo narratives via social media. As part of the intervention, a community photo exhibition was held featuring these photo narratives. Exit interviews were also conducted at study close.

SETTING/PARTICIPANTS: Nine individuals providing informal care to children in a pediatric palliative care program participated in the intervention. All participants were female and are older than 18 years. Settings for research include participant homes and at The Center for Hospice and Palliative Care in Cheektowaga, New York. RESULTS: Participants posted 95 photographs and 96 narratives during the intervention, posting on average once each week. Statistical analysis within the small sample indicated an increased presence of meaning in the lives of participants (P = .022). Exit interviews conveyed satisfaction with the intervention. CONCLUSIONS: Findings suggest that the Photographs of Meaning Program is a practical intervention with life-enhancing potential for pediatric palliative. Future research should aim to collect additional evidence of the intervention’s effectiveness.


Background: Many young adults with cerebral palsy (CP) face limited participation in activities, including employment and independent living. Physical therapy during the transition period can help to support participation through promotion of self-care, ambulation, and functional mobility. Thus, ensuring appropriate access to physical therapy services for young people who can benefit from them before, during, and after transition is imperative. Objective: The objective of this study was to identify factors contributing to the utilization of physical therapy services for youth with CP.
both during and after secondary school. Design: The design was a deidentified secondary analysis of the National Longitudinal Transition Study 2 (NLTS2). Methods: Multivariate regression models were run to examine demographic and disability characteristics influencing utilization of physical therapy services for youth with CP both during and after secondary school. Results: The total weighted population sample included 35,290 young people with CP. When all youth were in secondary school, 59.4% of the youth utilized physical therapy services; however, once all youth were out of school, only 33.7% of them were reported to have utilized physical therapy since leaving secondary school. For young people with difficulties accessing general disability support services, demographic characteristics, including sex, race, income, and parent education status, influenced use of physical therapy services in addition to disability characteristics. Limitations: This population sample included only young people in special education with Individual Education Plans (IEPs) and may not generalize to young people with CP in general education settings. Conclusions: Frequency of physical therapy services decreases drastically once young adults with CP leave secondary school. Future work should examine this trend in more depth to identify therapy intervention strategies to optimize participation in young adult life for persons with CP.


BACKGROUND: Advance care planning (ACP) is recommended for people with cystic fibrosis (CF), yet guidance for optimal implementation is lacking. OBJECTIVE: To assess ACP-related thoughts, comfort level, and preferences among people with CF to guide evidence-based routine implementation of ACP in the CF clinic. DESIGN: A cross-sectional survey assessed ACP-related experiences and preferences. SUBJECTS: Thirty-eight adolescents and adults with CF from an urban CF center. RESULTS: Few subjects reported talking to their CF team about ACP care preferences (5%) or completing advance directives detailing desired medical treatments (11%). However, most participants worried about living with advanced disease (84%) and felt comfortable discussing ACP preferences with CF providers (92%). Subjects largely preferred that ACP conversations occur when they are generally healthy, in the outpatient setting, and with any familiar CF team member. Disease severity was not associated with frequency of worry about living with advanced disease, comfort level with ACP discussions, or ACP setting preferences. CONCLUSIONS: People with CF worry about advanced disease and feel comfortable discussing ACP, but need more guidance to understand and document ACP choices. CF patient experiences and preferences support implementation of an early, active approach to ACP for people with CF.


Mental health professionals can play a key role in helping pediatric patients and their families prepare for and endure the death of a child. Impactful interventions include assisting a family’s transition toward acceptance of a child’s pending death, using prognostication as a tool in
emotional preparedness, and education on expectant symptoms to optimize management and sense of caregiver efficacy.


It has long been recognized that patients with neurological conditions, and particularly pediatric neurology patients, are well suited for palliative care because they frequently have a high symptom burden and variable prognoses. In 1996, the American Academy of neurology formally recognized a need for neurologists to "understand and apply the principles of palliative medicine."

Subsequently, some reviews have proposed a simultaneous care model in which palliative care is integrated for all neurology patients from the time of diagnosis. This article will review the current status of palliative care in pediatric neurology and discuss barriers to its integration.


BACKGROUND: Due to medical advances, growing numbers of adolescents with congenital heart disease (CHD) survive into adulthood and transferring from paediatric to adult healthcare. This transfer is significant step in a young person's life, and this study examines the views of Irish healthcare professionals' on how best to manage this transition. METHODS: Purposeful sampling was used to invite participation by healthcare professionals (HCPs) from a variety of disciplines whose caseloads include adolescents and young adults with CHD. Fourteen professionals participated in semistructured interviews regarding their experiences of the transition process and their recommendations. Data were collected during Spring 2016 and analysed using thematic analysis. RESULTS: Results indicated that the current approach to transition and transfer could be improved. Professionals identified barriers hindering the transition process such as cultural and attitudinal differences between HCPs dealing with child and adult patients, inadequate preparation and education of patients about their condition, parental reluctance to transfer, and concern about parents' role in on-going treatment. Measures such as better support and education for both the patients and their parents were recommended, in order to facilitate a smoother transition process for all parties involved. Additionally, HCPs identified the need for better collaboration and communication, both between paediatric and adult healthcare professionals and between hospitals, to ensure greater continuity of care for patients. CONCLUSIONS: Action is required in order to improve the current transition process. Measures need to be taken to address the barriers that currently prevent a smooth transition process for young adult CHD patients. Professionals recommended the implementation of a structured transition clinic to deal with the wide variety of needs of transitioning adolescent patients and their families. Recommendations for future research are also made.

STUDY PURPOSE: The purpose of this qualitative study was to understand, from the parent perspective, the experience of the family whose child has Type 1 spinal muscular atrophy (Type 1 SMA), in the emergency center, hospital, and clinical care settings to identify opportunities for improved family-centered care (FCC). DESIGN AND METHODS: This study used a qualitative descriptive design with individual or small group interviews guided by a semi-structured questionnaire. Reviewers used framework analysis to identify gaps in the provision of FCC and opportunities for improvement with respect to services health professionals may provide families of children with Type 1 SMA. RESULTS: Nineteen families with 22 children with Type 1 SMA participated. Results are organized according to eight basic tenets of FCC. Family-to-family interactions strongly impacted participants’ decision-making and perceived level of support. Participants valued strong family/provider partnerships, feeling heard and respected by their providers, and receiving complete education regarding disease trajectory. CONCLUSIONS: Our analyses revealed both successful application of FCC and gaps in care where FCC could have been used to benefit families who have children with Type 1 SMA. As a pediatric chronic illness affects the whole family, FCC is important in maintaining the providers' focus on the family during the child’s care. PRACTICE IMPLICATIONS: There are opportunities for nursing, social work, care managers and others to engage as care coordinators to explain the family's goals and values to the medical team. Care coordinators help ensure understanding between families and providers, empowering the family to articulate their hopes and concerns.


OBJECTIVES: When active treatment is no longer in the best interests of the patient, redirection of care to palliation is an important transition. We review, within a tertiary neonatal intensive care unit (NICU), the journey leading to the decision to redirect care, the means of symptom control and the provision of psychosocial supports. METHODS: A retrospective review of all 166 deaths of NICU-affiliated patients during a 10-year epoch. Medical notes were reviewed, and the provision and type of, or barriers to, effective palliative care was defined. RESULTS: Extreme prematurity accounted for 71/145 (49%) of deaths with relatively high proportions of Maori 17/71 (25%) and Pacific Islanders 9/71 (13%). Almost all eligible infants received some form of palliation. Transition from curative to palliative care was refused by the family in a single case. Median time from decision to redirect care until first recorded action was 80 min, and median time from action until death was 60 min. The majority of infants received some form of comfort cares, (128/166) most commonly morphine (94/128, 73%). Three infants had documented seizure activity or respiratory distress but did not receive any pharmacological intervention. Psychosocial supports were offered in 98/145 (67%) of cases, but only 71/145 (49%) of families were formally offered an opportunity to discuss the infant's clinical course after their death. CONCLUSIONS: Clinical documentation of care plans was often incomplete, potentially leading to inconsistent delivery of care, increased risk of symptom breakthrough and/or inadequate psychosocial supports for family. Formal individualised
Palliative care plans are under development to standardise documentation and improve therapeutic and psychosocial interventions available to the infant and their family. [https://www.ncbi.nlm.nih.gov/pubmed/30470701](https://www.ncbi.nlm.nih.gov/pubmed/30470701)


Play is of vital importance for the healthy development of children. From a developmental perspective, play offers ample physical, emotional, cognitive, and social benefits. It allows children and adolescents to develop motor skills, experiment with their (social) behavioural repertoire, simulate alternative scenarios, and address the various positive and negative consequences of their behaviour in a safe and engaging context. Children with a chronic or life-threatening disease may face obstacles that negatively impact play and play development, possibly impeding developmental milestones, beyond the actual illness itself. Currently, there is limited understanding of the impact of (1) aberrant or suppressed play and (2) play-related interventions on the development of chronic diseased children. We argue that stimulating play behaviour enhances the adaptability of a child to a (chronic) stressful condition and promotes cognitive, social, emotional and psychomotor functioning, thereby strengthening the basis for their future health. Systematic play research will help to develop interventions for young patients, to better cope with the negative consequences of their illness and stimulate healthy development. [https://www.ncbi.nlm.nih.gov/pubmed/30273634](https://www.ncbi.nlm.nih.gov/pubmed/30273634)


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. [https://www.ncbi.nlm.nih.gov/pubmed/30047155](https://www.ncbi.nlm.nih.gov/pubmed/30047155)

**Pinkerton, R., Donovan, L. and Herbert, A. (2018). “Palliative Care in Adolescents and Young Adults With Cancer-Why Do Adolescents Need Special Attention?” Cancer J 24(6): 336-341.**

Meeting shortfalls in the provision of care to adolescents and young adults with cancer has focused largely on improving outcomes and psychosocial support. A significant percentage of adolescents and young adults with cancer will die of disease because of initial poor prognosis conditions or disease relapse. In adults, progress has been made in the concept of an integrated cancer/palliative care service. In pediatric oncology, the application of this philosophy of care has lagged behind somewhat. In the case of adolescents, particularly those with advanced cancer, the palliative care needs, in a broader sense than only end-of-life care, are often not adequately met, irrespective of whether treatment is delivered in a pediatric or adult cancer service. There are a number of age-specific aspects to palliative and supportive care for adolescents. Complex interactions between clinicians, parents, and patients potentially limit the young person’s ability to influence care planning. The wide variation in real or perceived competency at this age, the developmental challenges in relation to behavior, communication, and coping strategy all require particular professional expertise that is not always available. [https://www.ncbi.nlm.nih.gov/pubmed/30480579](https://www.ncbi.nlm.nih.gov/pubmed/30480579)


Limited studies exist regarding the timing, location, or physicians involved in do-not-resuscitate (DNR) order placement in pediatrics. Prior pediatric studies have noted great variations in practice during end-of-life (EOL) care. This study aims to analyze the timing, location, physician specialties, and demographic factors influencing EOL care in pediatrics. We examined the time preceding and following the implementation of a pediatric palliative care team (PCT) via a 5-year, retrospective chart review of all deceased patients previously admitted to inpatient services. Thirty-five percent (167/471) of the patients in our study died with a DNR order in place. Sixty-two percent of patients died in an ICU following DNR order placement. A difference was noted in DNR order timing between patients on general inpatient units and those discharged to home compared with those in the ICUs (p = 0.02). The overall DNR order rate increased following the initiation of the PCT from 30.8% to 39.2% (p = 0.05), but no change was noted in the rate of death in the ICUs. Our study demonstrates a variation in the timing of death following DNR order placement when comparing ICUs and general pediatric floors. Following the initiation of the PCT, we saw increased DNR frequency but no change in the interval between a DNR order and death. [https://www.ncbi.nlm.nih.gov/pubmed/30544741](https://www.ncbi.nlm.nih.gov/pubmed/30544741)

Background: Congenital syndrome of Zika virus (CSZV) is associated with neuromotor and cognitive developmental disorders, limiting the independence and autonomy of affected children and high susceptibility to complications, so palliative care needs to be discussed and applied. Aim: To identify factors associated with emergency visits and hospitalizations of patients with CSZV and clinical interventions performed from the perspective of palliative care. Design: This is a cross-sectional study with bidirectional longitudinal component. Data were collected between May and October 2017 through the review of medical records and interviews with relatives of patients hospitalized. Setting/Participants: The study was developed in a tertiary care hospital involving patients with confirmed CSZV born as of August 2015 and followed up until October 2017. Patients under investigation were excluded. Results: 145 patients were followed up at the specialized outpatient clinic, 92 (63.5%) were consulted at least once in the emergency room, and 49% had already been hospitalized, with the main reason being neurological causes, while 24.1% had never required any emergency visit or hospitalization. No risk factors were associated with the occurrence of consultations or hospitalizations. Such events happened at an early age and were accompanied by a high number of invasive procedures and interventions. An approach in palliative care was only identified in two hospitalized patients. Conclusions: For the patient with known severe malformations caused by congenital infection by the Zika virus with indication of palliative care, this approach could be used in order to allow life without suffering and disproportionate invasive method.


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.


This article reviews the state and practice of pediatric palliative care (PC) within the pediatric intensive care unit (PICU) with specific consideration of quality issues. This includes defining PC and end of life (EOL) care. We will also describe PC as it pertains to alleviating children’s suffering through the provision of "concurrent care" in the ICU environment. Modes of care, and attendant strengths, of both the consultant and integrated models will be presented. We will review salient issues related to the provision of PC in the PICU, barriers to optimal practice, parental, and staff perceptions. Opportunity areas for quality improvement and the role of initiatives and measures such as education, family-based initiatives, staff needs, symptom recognition, grief, and communication follow. To conclude, we will look to the literature for PC resources for pediatric intensivists and future directions of study.


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.


Background: The impact of pediatric palliative care (PPC) is well established for children with chronic complex diseases. However, PPC likely also benefits previously healthy children with acute life-threatening conditions. Objective: To determine the incidence and impact of PPC for previously healthy patients who died in a pediatric hospital. Design: Retrospective chart review of all pediatric deaths over four years. Setting/Subjects: Patients were 0 to 25 years old, died during an inpatient stay at an academic pediatric hospital >/=48 hours after admission, and had no complex chronic conditions (CCCs) before admission. Measurements: One hundred sixty-seven patients met the
eligibility criteria. Most died in intensive care settings (n = 149, 89%), and few (n = 34, 20%) received PPC consultations or services. Results: Patients who received PPC services were more likely to receive a multidisciplinary care conference than did patients without PPC support (70.5% vs. 39.9%; p = 0.001), which also occurred earlier for patients who received PPC services (seven days vs. two days before death; p = 0.04). Most patients had documented end-of-life planning in their medical records; however, this occurred earlier for patients who received PPC consultation (9.5 days before death) than for those who did not (two days before death; p < 0.0001). Patients receiving PPC support (67.7%) were also more likely to have a do-not-resuscitate/intubate order before death than those who did not (39.9%; p = 0.004). Conclusions: Pediatric patients without known CCCs who subsequently die as inpatients benefit from PPC in terms of goals of care discussions and documentation of end-of-life care preferences.


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 in 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.


Background: In pediatric health care, non-pharmacological interventions such as music therapy have promising potential to complement traditional medical treatment options in order to facilitate recovery and well-being. Music therapy and other music-based interventions are increasingly applied in the clinical treatment of children and adolescents in many countries worldwide. The purpose of this overview is to examine the evidence regarding the effectiveness of music therapy and other music-based interventions as applied in pediatric health care. Methods: Surveying recent literature and summarizing findings from systematic reviews, this overview covers selected fields of application in pediatric health care (autism spectrum disorder; disability; epilepsy; mental health; neonatal care; neurorehabilitation; pain, anxiety and stress in medical procedures; pediatric oncology and palliative care) and discusses the effectiveness of music interventions in these areas. Results: Findings show that there is a growing body of evidence regarding the beneficial effects of music therapy, music medicine, and other music-based interventions for children and adolescents, although more rigorous research is still needed. The highest quality of evidence for the positive effects of music therapy is available in the fields of autism spectrum
disorder and neonatal care. Conclusions: Music therapy can be considered a safe and generally well-accepted intervention in pediatric health care to alleviate symptoms and improve quality of life. As an individualized intervention that is typically provided in a person-centered way, music therapy is usually easy to implement into clinical practices. However, it is important to note that to exploit the potential of music therapy in an optimal way, specialized academic and clinical training and careful selection of intervention techniques to fit the needs of the client are essential.


OBJECTIVES: Following publication of detailed national neonatal palliative care guidance, practical regional guidance, in the form of multidisciplinary ‘checklists’, was implemented aiming to improve the quality of neonatal palliative care. METHODS: Case note audit was used to examine the quality of locally delivered neonatal palliative care before and after regional guidance implementation. RESULTS: 27 patients were allocated to the ‘before’ cohort and 10 to the ‘after’ cohort. Introduction of the checklists was apparently associated with improvements in domains of pain relief and comfort care, monitoring, fluids and nutrition, completion of diagnostics, treatment ceiling decisions, resuscitation status and discussion with parents. Other support for parents was poorly adhered to. CONCLUSION: Regional guidance improved some aspects of palliative care delivery though other areas remained suboptimal. Other strategies, for example, consultation with paediatric palliative care services, need to be considered to further improve the quality of palliative care delivered to babies with life-limiting illnesses.


For most families, the preferred location of death for their child is home, yet most children still die in the hospital. Many children with life-threatening and life-limiting illness are medically dependent on technology, and palliative transport can serve as a bridge from the intensive care unit to the family’s home to achieve family-centered goals of care. Palliative transport may also present an opportunity to prioritize cultural care and rituals at end of life which cannot be provided in the hospital. We describe a case series of pediatric patients from communities espousing markedly diverse cross-cultural values and limited financial resources. Specific cultural considerations at end of life for these children included optimizing the presence of the shared community or tribe, the centrality of healing rituals, and varied attitudes toward withdrawal of life-sustaining medical treatment. By addressing each of these components, we were able to coordinate palliative transport to enhance cross-cultural care and meaning at end of life for children with life-limiting illness.

ABSTRACT
Objective: The present study intended to evaluate the impact of a standardized format-called the “Music Givers,” based on a single session of music intervention followed by a buffet-on the psychological burden and well-being of hospitalized cancer patients. METHOD: The Distress Thermometer (DT), the Hospital Anxiety and Depression Scale (HADS), and self-reported visual analogue scales (score range = 1-10) to assess pain, fatigue, and five areas of well-being (i.e., physical, psychological, relational, spiritual, and overall well-being) were administered to 242 cancer patients upon admission to and at discharge from the hospital. Among them, 103 were hospitalized during which time a live concert took place (intervention group), whereas 139 patients were hospitalized when it did not (control group). RESULTS: Compared to the control group, patients in the intervention group demonstrated less distress at discharge according to the DT (adjusted estimate of difference = -0.8, p = 0.001), lower HADS-Anxiety (-1.7, p < 0.001) and HADS-Depression scores (-1.3, p = 0.001), and higher scores on all the well-being scales, with the exception of spiritual well-being. In addition, no between-group differences were found in terms of pain and fatigue scores at discharge. SIGNIFICANCE OF RESULTS: The one-session format of the Music Givers intervention is an effective, standardized, easy-to-replicate, and low-cost intervention that reduces psychological burden and improves the well-being of hospitalized cancer patients. Listening to live music and the opportunity to establish better relationships between patients and staff could explain these results.


CONTEXT: All inpatient children receiving pediatric palliative care consults at a free-standing children's hospital. OBJECTIVES: To explore the impact of massage therapy on pediatric palliative care patients’ symptom burden and medication use pattern, to describe the impact of massage therapy on family caregiver distress, and to report on bedside nursing staff perception of massage therapy for children and their families. METHODS: A 1-time point, single-center exploratory study offering 10-minute bedside massage to children receiving palliative care and 10-minute massage to their family caregivers. RESULTS: A total of 135 massages were provided to children and their caregivers. Difference in child Face, Legs, Activity, Cry, Consolability scale (FLACC) score was detectable (P < .0001) with the median (interquartile range [IQR]) before FLACC score being 2 (1-3) and after FLACC score being 0 (0-1). Difference in “as-needed” pain medication usage in the 24 hours before and after the massage was detectable (P = .0477). Median difference in family caregiver distress with massage was -3.0 (IQR = 2.0, P < .0001). Bedside nurses (100%) reported massage to be a meaningful way to care for their families and patients. CONCLUSION: Massage therapy is a potentially meaningful intervention for pediatric palliative care patients with noted impact on symptom burden, benefit to family caregivers, and acceptance by nursing staff.


OBJECTIVE: The death of a child has been associated with adverse parental outcomes, including a heightened risk for psychological distress, poor physical health, loss of employment income, and diminished psychosocial well-being. Psychosocial standards of care for centers serving pediatric cancer patients recommend maintaining at least one meaningful contact between the healthcare team and bereaved parents to identify families at risk for negative psychosocial sequelae and to provide resources for bereavement support. This study assessed how this standard is being implemented in current healthcare and palliative care practices, as well as barriers to its implementation. METHOD: Experts in the field of pediatric palliative care and oncology created a survey that was posted with review and permission on four listservs. The survey inquired about pediatric palliative and bereavement program characteristics, as well as challenges and barriers to implementation of the published standards of care. Result: The majority of participants (N = 100) self-reported as palliative care physicians (51%), followed by oncologists (19%). Although 59% of staff reported that their center often or always deliver bereavement care after a child's death, approximately two-thirds reported having no policy for the oncology team to routinely assess bereavement needs. Inconsistent types of bereavement services and varying duration of care was common. Twenty-eight percent of participants indicated that their center has no systematic contact with bereaved families after the child's death. Among centers where contacts are made, the person who calls the bereaved parent is unknown to the family in 30% of cases. Few centers (5%) use a bereavement screening or assessment tool. Significance of results: Lack of routine assessment of bereavement needs, inconsistent duration of bereavement care, and tremendous variability in bereavement services suggest more work is needed to promote standardized, policy-driven bereavement care. The data shed light on multiple areas and opportunities for improvement.


The aims of this article are twofold: (I) provide a general overview of perinatal bereavement services throughout the healthcare system and (II) identify future opportunities to improve bereavement services, including providing resources for the creation of standardized care guidelines, policies and educational opportunities across the healthcare system. Commentary is provided related to maternal child services, the neonatal intensive care unit (NICU), prenatal clinics, operating room (OR) and perioperative services, emergency department (ED), ethics, chaplaincy and palliative care services. An integrated system of care increases quality and safety and contributes to patient satisfaction. Physicians, nurses and administrators must encourage pregnancy loss support so that regardless of where in the facility the contact is made, when in the pregnancy the loss occurs, or whatever the conditions contributing to the pregnancy ending, trained caregivers are there to provide bereavement support for the family and palliative symptom management to the fetus born with a life limiting condition. The goal for respectful caregiving throughout an entire hospital system is achievable and critically important.


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.


