

# Synopsis

International digest of children's palliative  
care research abstracts

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Dr Linda Maynard, Sue Langley and Lizzie Chambers

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**Synopsis, International digest of children's palliative care research abstracts**

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**Together for Short Lives is the leading UK charity for all children with life-threatening and life-limiting conditions and all those who support, love and care for them. We support families, professionals and services, including children's hospices. Our work helps to ensure that children can get the best possible care, wherever and whenever they need it.**

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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](http://www.each.org.uk/library)

For information on membership, please contact: Sue Langley, Library and Information Services Manager [sue.langley@each.org.uk](mailto:sue.langley@each.org.uk); 01223 815103



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## Proceedings of the seventh northern region paediatric colloquium

Barkla, X. and C. Kaplan

*Med Sci Law* 2014; 54(1): 15-21

Ethical and legal dilemmas frequently arise in paediatric practice. Given the nature of the speciality, these issues are relevant to both the medical and legal professions. To this end, senior figures from the medical and legal professions in the Northern region have met on a regular basis in order to discuss anonymised case material. We report on the proceedings of the seventh such meeting. Six cases are described and key points arising from the subsequent discussion are presented.

<http://www.ncbi.nlm.nih.gov/pubmed/23864488>

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## Ethical aspects of clinical research with minors

Bos, W., et al

*Eur J Pediatr* 2013; 172(7): 859-866

Over the past decades, clinical research has increasingly been subjected to ethical requirements and legal regulation. The specific focus of ethical and legal frameworks on competent adults (which serve as the paradigmatic research subject), however, has created an ambivalent attitude towards paediatric clinical research. On one hand, minors are regarded as a vulnerable population that deserves additional protection against the risks and burdens involved in clinical research. On the other hand, the population of minors should not be denied (or not get timely) access to the benefits of clinical research. In this paper, we will explore the legal regulation and ethical guidance that currently governs paediatric clinical research in the European Union and discuss the future challenges in this field. In addition, we will discuss major ethical concerns in paediatric clinical research, with a focus on the acceptability of research risks and the informed consent process. In the discussion, we will address key concerns in both regulating paediatric clinical research and implementing ethical and legal requirement in the actual paediatric research conduct.

<http://www.ncbi.nlm.nih.gov/pubmed/23073901>

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## The 'window of opportunity' for death after severe brain injury: family experiences

Kitzinger, J. and Kitzinger, C.

*Health Illn* 2013; 35(7): 1095-1112.

This article builds on and develops the emerging bioethics literature on the 'window of opportunity' for allowing death by withholding or withdrawing treatment. Our findings are drawn from in-depth interviews with 26 people (from 14 different families) with severely brain injured relatives. These interviews were specifically selected from a larger study on the basis of interviewees' reports that their relatives would not have wanted to be kept alive in their current condition (e.g. in vegetative or minimally conscious states). Our analysis tracks the decision-making processes that have led to the situation in which life-sustaining treatments continue to be delivered to these patients--maintaining them in a state that some families describe as a 'fate worse than death'. We show how the medico-legal 'window of opportunity' for allowing the patient to die structures family experience and fails to deliver optimal outcomes for patients. We end with some suggestions for change.

<http://www.ncbi.nlm.nih.gov/pubmed/23278317>

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## Communication and trust in the care provided to a dying parent: A nationwide study of cancer-bereaved youths

Grenklo, T. B., et al.

*J Clin Oncol* 2013; 31(23): 2886-2894

**PURPOSE:** To assess children's trust in the care provided to a dying parent during the final week of life in relation to end-of-life medical information about disease, treatment, and death. **METHODS:** This nationwide population-based survey included 622 (73%) of 851 youths who, 6 to 9 years earlier, at age 13 to 16 years, lost a parent to cancer. We asked about the children's reception of end-of-life professional information and trust in the care provided. We also asked about depression and several potential risk factors of distrust in the care provided. **RESULTS:** A majority (82%) reported moderate/very much trust in the care provided. Compared with children who received end-of-life medical information before their loss, the risk of distrust in the care provided was higher in those who received no information (risk ratio [RR], 2.5; 95% CI, 1.5 to 4.1), in those who only received information afterward (RR, 3.2; 95% CI, 1.7 to 5.9), and in those who did not know or remember if end-of-life medical information was provided (RR, 1.7; 95% CI, 1.1 to 2.5). Those reporting distrust in the care provided had an RR of 2.3 (95% CI, 1.5 to 3.5) for depression. Furthermore, the risk of distrust in the care provided was higher among children reporting poor efforts to cure (RR, 5.1; 95% CI, 3.6 to 7.3), and/or a poor relationship with the surviving parent (RR, 2.9; 95% CI, 2.0 to 4.1). **CONCLUSION:** Our study suggests that children's trust in the care provided to a dying parent was highest when they received end-of-life medical information before their loss.

<http://www.ncbi.nlm.nih.gov/pubmed/23857973>

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## Association between religious and socio-economic background of parents of children with solid tumors and DNR orders

Hileli, I., et al

*Pediatr Blood Cancer* 2014; 61(2): 265-268

**BACKGROUND:** The influence of socio-economic and religious background on decisions made by parents of children with incurable cancer regarding DNR orders is not fully understood. **PROCEDURE:** A retrospective analysis of medical charts of patients who died between January 2000 and January 2011 was performed. The following data were sought: written evidence of DNR discussion with parents, religious background, educational level, monthly income. **RESULTS:** There was evidence of a discussion on DNR in 73/90 charts. DNR consent was obtained in 14/17 (82.4%) cases where at least one parent had >15 years of education versus in only 24/45 (53.3%) cases where both parents had ≤15 years education as determined by univariate analysis (P = 0.03). DNR consent was also more likely to be obtained among parents of children with income >10,000 NIS (24/30, 80.0% vs. 20/38, 52.6%, P = 0.013). Parents of Jewish (22/30, 73.3%), Islamic (16/26, 61.5%), and Christian (8/9, 88.9%) background were equally likely to provide DNR consent. However, Druze families were less likely to do so (2/8, 25.0%, P = 0.036). **CONCLUSIONS:** The process of decision-making to a DNR request was associated with parents' educational level and monthly family income, and not by religious background, with the exception of Druze families.

<http://www.ncbi.nlm.nih.gov/pubmed/23940107>

## Cancer patients and advance directives: A survey of patients in a hematology and oncology outpatient clinic

Hubert, E., et al.

*Onkologie* 2013; 36(7-8): 398-402

**BACKGROUND:** In 2009, Germany enacted a new law supporting advance directives that led to heated discussions in the media and the public. 3 years after the law passed, we surveyed patients with malignant diseases with regards to their views on advance directives. **PATIENTS AND METHODS:** Between September 2011 and July 2012 an anonymous survey on advance directives was conducted among 617 patients at the hematology and oncology outpatient department of the University Hospital Mannheim, using a standardized questionnaire developed for this investigation. **RESULTS:** Of the 503 patients who returned the questionnaire, 31% (n = 157) indicated having an advance directive. Of these 157, 54% (n = 85) completed the advance directive after 2009. 56% (282 out of 503) desired more information on advance directives. Of these, 71% (201 out of 282) wanted their general physician and 45% (128 out of 282) their specialist, to provide more information about this issue. Of the 339 patients without an advance directive, 47% (n = 158) stated that they had 'not worried about that yet'. **CONCLUSION:** Although the percentage of patients with advance directives has increased since the legislative amendment, more information is still required by patients. It is recommended that physicians should discuss advance directives more frequently with their patients.

<http://www.ncbi.nlm.nih.gov/pubmed/23921757>

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## Transition to non-curative end-of-life care in paediatric oncology: A nationwide follow-up in Sweden

Jalmsell, L., et al.

*Acta Paediatr* 2013; 102(7): 744-748

**AIM:** To estimate whether and when children dying from a malignancy are recognized as being beyond cure and to study patterns of care the last weeks of life. **METHODS:** A nationwide retrospective medical record review was conducted. Medical records of 95 children (60% of eligible children) who died from a malignancy 2007-2009 in Sweden were studied. **RESULTS:** Eighty-three children (87%) were treated without curative intent at the time of death. Children with haematological malignancies were less likely to be recognized as being beyond cure than children with brain tumours [relative risks (RR) 0.7; 95% confidence interval (CI) 0.6-0.9] or solid tumours (RR 0.8; 0.6-1.0). The transition to noncurative care varied from the last day of life to over four years prior to death (median 60 days). Children with haematological malignancies were treated with a curative intent closer to death and were also given chemotherapy (RR 5.5; 1.3-22.9), transfusions (RR 2.0; 1.0-4.0) and antibiotics (RR 5.3; 1.8-15.5) more frequently than children with brain tumours the last weeks of life. **CONCLUSION:** The majority of children dying from a malignancy were treated with noncurative intent at the time of death. The timing of a transition in care varied with the diagnoses, being closer to death in children with haematological malignancies.

<http://www.ncbi.nlm.nih.gov/pubmed/23557514>

## The duty of the physician to care for the family in pediatric palliative care: Context, communication, and caring

Jones, B. L., et al.

*Pediatrics* 2014; 133 Suppl 1: S8-15

Pediatric palliative care physicians have an ethical duty to care for the families of children with life-threatening conditions through their illness and bereavement. This duty is predicated on 2 important factors: (1) best interest of the child and (2) nonabandonment. Children exist in the context of a family and therefore excellent care for the child must include attention to the needs of the family, including siblings. The principle of nonabandonment is an important one in pediatric palliative care, as many families report being well cared for during their child's treatment, but feel as if the physicians and team members suddenly disappear after the death of the child. Family-centered care requires frequent, kind, and accurate communication with parents that leads to shared decision-making during treatment, care of parents and siblings during end-of-life, and assistance to the family in bereavement after death. Despite the challenges to this comprehensive care, physicians can support and be supported by their transdisciplinary palliative care team members in providing compassionate, ethical, and holistic care to the entire family when a child is ill.

<http://www.ncbi.nlm.nih.gov/pubmed/24488541>

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## Unilateral pediatric do not attempt resuscitation orders: The pros, the cons, and a proposed approach

Mercurio, M. R., et al.

*Pediatrics* 2014; 133 Suppl 1: S37-43

A unilateral do not attempt resuscitation (DNAR) order is written by a physician without permission or assent from the patient or the patient's surrogate decision-maker. Potential justifications for the use of DNAR orders in pediatrics include the belief that attempted resuscitation offers no benefit to the patient or that the burdens would far outweigh the potential benefits. Another consideration is the patient's right to mercy, not to be made to undergo potentially painful interventions very unlikely to benefit the patient, and the physician's parallel obligation not to perform such interventions. Unilateral DNAR orders might be motivated in part by the moral distress caregivers sometimes experience when feeling forced by parents to participate in interventions that they believe are useless or cruel. Furthermore, some physicians believe that making these decisions without parental approval could spare parents needless additional emotional pain or a sense of guilt from making such a decision, particularly when imminent death is unavoidable. There are, however, several risks inherent in unilateral DNAR orders, such as overestimating one's ability to prognosticate or giving undue weight to the physician's values over those of parents, particularly with regard to predicted disability and quality of life. The law on the question of unilateral DNAR varies among states, and readers are encouraged to learn the law where they practice. Arguments in favor of, and opposed to, the use of unilateral DNAR orders are presented. In some settings, particularly when death is imminent regardless of whether resuscitation is attempted, unilateral DNAR orders should be viewed as an ethically permissible approach.

<http://www.ncbi.nlm.nih.gov/pubmed/24488539>

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## Deceased donor liver transplantation in infants and small children: Are partial grafts riskier than whole organs?

Cauley, R. P., et al.

*Liver Transplant* 2013; 19(7): 721-729

Infants have the highest wait-list mortality of all liver transplant candidates. Although previous studies have demonstrated that young children may be at increased risk when they receive partial grafts from adult and adolescent deceased donors (DDs), with few size-matched organs available, these grafts have increasingly been used to expand the pediatric donor pool. We aimed to determine the current adjusted risks of graft failure and mortality in young pediatric recipients of partial DD livers and to determine whether these risks have changed over time. We analyzed 2683 first-time recipients of DD livers alone under the age of 24 months in the United Network for Organ Sharing database (1995-2010), which included 1118 partial DD livers and 1565 whole DD organs. Transplant factors associated with graft loss in bivariate analyses ( $P < 0.1$ ) were included in multivariate proportional hazards models of graft and patient survival. Interaction analysis was used to examine risks over time (1995-2000, 2001-2005, and 2006-2010). Although there were significant differences in crude graft survival by the graft type in 1995-2000 ( $P < 0.001$ ), graft survival rates with partial and whole grafts were comparable in 2001-2005 ( $P = 0.43$ ) and 2006-2010 ( $P = 0.36$ ). Furthermore, although the adjusted hazards for partial graft failure and mortality were 1.40 [95% confidence interval (CI) = 1.05-1.89] and 1.41 (95% CI = 0.95-2.09), respectively, in 1995-2000, the adjusted risks of graft failure and mortality were comparable for partial and whole organs in 2006-2010 [hazard ratio (HR) for graft failure = 0.81, 95% CI = 0.56-1.18; HR for mortality = 1.02, 95% CI = 0.66-1.71]. In conclusion, partial DD liver transplantation has become less risky over time and now has outcomes comparable to those of whole liver transplantation for infants and young children. This study supports the use of partial DD liver grafts in young children in an attempt to significantly increase the pediatric organ pool.

<http://www.ncbi.nlm.nih.gov/pubmed/23696310>

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## Pediatric palliative care and pediatric medical ethics: Opportunities and challenges

Feudtner, C. and Nathanson, P. G

*Pediatrics* 2014; 133 Suppl 1: S1-7

The fields of pediatric palliative care (PPC) and pediatric medical ethics (PME) overlap substantially, owing to a variety of historical, cultural, and social factors. This entwined relationship provides opportunities for leveraging the strong communication skills of both sets of providers, as well as the potential for resource sharing and research collaboration. At the same time, the personal and professional relationships between PPC and PME present challenges, including potential conflict with colleagues, perceived or actual bias toward a palliative care perspective in resolving ethical problems, potential delay or underuse of PME services, and a potential undervaluing of the medical expertise required for PPC consultation. We recommend that these challenges be managed by: (1) clearly defining and communicating clinical roles of PPC and PME staff, (2) developing questions that may prompt PPC and PME teams to request consultation from the other service, (3) developing explicit recusal criteria for PPC providers who also provide PME consultation, (4) ensuring that PPC and PME services remain organizationally distinct, and (5) developing well-defined and broad scopes of practice. Overall, the rich relationship between PPC and PME offers substantial opportunities to better serve patients and families facing difficult decisions.

<http://www.ncbi.nlm.nih.gov/pubmed/24488535>

## Waiver of informed consent in pediatric resuscitation research: A systematic review

Eltorki, M., et al.

*Acad Emerg Med* 2013; 20(8): 822-834

**BACKGROUND:** In critical care and emergency medicine research, obtaining consent can be problematic when patients present with life-threatening conditions. This issue is further complicated in children, as even while coherent, they are often incapable of making decisions regarding their own care. To enable the ethical conduct of research in such situations, the Food and Drug Administration (FDA) of the United States has set recommendations for the conduct of research employing a waiver of consent. These regulations have been termed exception from informed consent, or EFIC. As this is an evolving concept with limited pediatric experience, the authors conducted a review to examine the conduct of emergency research in the absence of prospectively obtained informed consent. Our review focused both on opinions and on the ability to conduct research without informed consent in life-threatening situations. **METHODS:** A systematic review of the literature was undertaken in accordance with the PRISMA guidelines. Medline, CINAHL, and EMBASE databases were searched on January 9, 2013. Eligibility criteria included: 1) examined a method of conducting research in a life-threatening situation, 2) involved a real or theoretical clinical situation, 3) involved patients less than 18 years of age or a substitute decision-maker, and 4) reported at least one quantifiable outcome. The findings were synthesized qualitatively with the pertinent results summarized and discussed. **RESULTS:** Eleven articles matched the eligibility criteria. Six focused on community consultation and public disclosure, three focused on the feasibility of employing a waiver of consent, and two examined attitudes toward emergency research. Of the studies focusing on community consultation, four defined the community as previous or current patients and health care providers and administrators in the study's home institution; the other two defined the community as the general population. Although there was heterogeneity in study designs, settings, and outcome measures, overall 68% (3,219 of 4,767) of subjects surveyed supported the use of EFIC under select circumstances (individual study range = 50% to 92%). Caregiver support increased among those in whom the situation was a more possible reality (e.g., critical care unit patients) and varied by the scenario and method of presentation (e.g., bulleted handout vs. preferred). Several studies revealed that patient accrual and time to intervention are impeded when prospective informed consent is required. Finally, deferred consent, although endorsed and used outside of the United States, continues to raise important ethical questions, particularly related to the need and timing of disclosure. **CONCLUSIONS:** Limited data exist evaluating ethical issues in pediatric acute care resuscitation research. This review highlighted the fact that every proposal is unique and the method of obtaining consent (or waiver) requires careful consideration by local ethics committees. Particular attention must be paid to use of the population selected for community consultation. Several studies highlighted the need to consider the use of alternatives to prospective informed consent to enable the conduct of research in emergency departments (EDs) in life-threatening situations. Future research should evaluate children's opinions on this topic.

<http://www.ncbi.nlm.nih.gov/pubmed/24033626>

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## When life-sustaining treatment is withdrawn and the patient doesn't die

Kutzsche, S., et al.

*Pediatrics* 2013; 132(5): 893-897

One of the most difficult decisions that doctors and parents must make is the decision to withdraw life-sustaining treatment. Doctors find it easier to withdraw treatments in situations where withdrawal will be rapidly fatal rather than in situations in which treatment withdrawal will lead to a prolonged dying process. Mechanical ventilation is usually such a treatment. Withdrawal of ventilation generally leads to the patient's rapid demise. Doctors may tell parents that death will occur quickly after a ventilator is withdrawn. But what happens when the doctors are wrong and a patient survives without life support? What should doctors do next? We present a case in which that happened and asked 3 experts to comment on the case. Stefan Kutzsche is a senior consultant in neonatology at Oslo University Hospital Ullevål in Norway. John Colin Partridge is a neonatologist and professor of pediatrics at University of California, San Francisco. Steven R. Leuthner is a neonatologist and professor of pediatrics and bioethics at the Medical College of Wisconsin. They each recommend slightly different approaches to this dilemma.

<http://www.ncbi.nlm.nih.gov/pubmed/24101767>

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## Tracheostomies and assisted ventilation in children with profound disabilities: Navigating family and professional values

Wilfond, B. S.

*Pediatrics* 2014; 133 Suppl 1: S44-49

Parental requests for gastrostomies, tracheostomies, or assisted ventilation in children with profound disabilities raise ethical concerns about children's interests, parental decision-making, and health care costs. The underlying concern for many relates to the perceived value of these children. Clinicians should make efforts to appreciate the family's perspective regarding children with profound disabilities who require respiratory and nutritional medical support. Finding opportunities to learn about the family members' lives outside of the health care setting may facilitate a deeper understanding of what it means to live with a child who has profound disabilities. In conversations with families, referring to interventions as futile and conditions as lethal will obscure the value-based nature of these decisions. Respiratory and nutritional interventions are not clearly against the interests of most children. Even for children with a limited life span, life-sustaining interventions may be important for the child and family. Health care costs are a serious societal issue; however, the costs associated with profound disabilities are not the most significant contributor. Societal decisions not to provide life-sustaining health care to children with profound disabilities would require a public process. Clinicians may have personal views regarding decisions for their own family or for their vision for society. However, clinicians have professional obligations to families who have different values. It is important to present balanced information and support parental decision-making so parents may decide to forgo or use life-sustaining interventions according to their values and goals.

<http://www.ncbi.nlm.nih.gov/pubmed/24488540>

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## Communicating about prognosis: Ethical responsibilities of pediatricians and parents

Mack, J. W. and S. Joffe

*Pediatrics* 2014; 133 Suppl 1: S24-30

Clinicians are sometimes reluctant to discuss prognosis with parents of children with life-threatening illness, usually because they worry about the emotional impact of this information. However, parents often want this prognostic information because it underpins informed decision-making, especially near the end of life. In addition, despite understandable clinician concerns about its emotional impact, prognostic disclosure can actually support hope and peace of mind among parents struggling to live with a child's illness. Children, too, may need to understand what is ahead to manage uncertainty and make plans for the ways their remaining life will be lived. In this article, we describe the ethical issues involved in disclosure of prognostic information to parents and children with life-threatening illness and offer practical guidance for these conversations.

<http://www.ncbi.nlm.nih.gov/pubmed/24488537>

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## Length of time from extubation to cardiorespiratory death in neonatal intensive care patients and assessment of suitability for organ donation

Saha, S. and A. L. Kent

*Arch Dis Child Fetal Neonatal Ed* 2014; 99(1): F59-63

**OBJECTIVE:** A common concern for parents when end of life decisions are made is the length of time their baby may take to die. Postcardiac death organ donation is now becoming more common, along with neonatal organ donation. The aim was to determine the length of time from extubation until cardiorespiratory death (CRD) in neonatal intensive care patients and consideration of potential organ donation. **DESIGN:** Retrospective review of medical records of neonates who died in a neonatal intensive care unit between 2000 and 2009. **PATIENTS:** Data collected included gestation at birth, age at death, birth weight, reason for cessation of intensive care, inotrope and ventilation requirements, sedation and muscle relaxation prior to death, time from extubation to documented CRD. An assessment was made for potential suitability for consideration of organ donation with a gestation at birth  $\geq$  34 weeks and birth weight  $>2.0$  kg. **RESULTS:** 117 neonates were included, median gestation 29 weeks and median birth weight 1220 grams. The median age at death was 4 days of age. The median time from discussing prognosis to death was 137 min. The median time from extubation to CRD was 30 min. Seven (6%) neonates were considered suitable for organ donation, and for these infants the median time from extubation to CRD was 120 min. Two neonates donated heart valves. **CONCLUSIONS:** This provides a guide for grieving parents on time frames for the interval between extubation and CRD. More accurate postextubation CRD times are required to determine likely potential for postcardiac death organ donation.

<http://www.ncbi.nlm.nih.gov/pubmed/24105623>

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## Which newborn infants are too expensive to treat? Camosy and rationing in intensive care

Wilkinson, D.

*J Med Ethics* 2013;39(8): 502-506

Are there some newborn infants whose short- and long-term care costs are so great that treatment should not be provided and they should be allowed to die? Public discourse and academic debate about the ethics of newborn intensive care has often shied away from this question. There has been enough ink spilt over whether or when for the infant's sake it might be better not to provide life-saving treatment. The further question of not saving infants because of inadequate resources has seemed too difficult, too controversial, or perhaps too outrageous to even consider. However, Roman Catholic ethicist Charles Camosy has recently challenged this, arguing that costs should be a primary consideration in decision-making in neonatal intensive care. In the first part of this paper I will outline and critique Camosy's central argument, which he calls the 'social quality of life (sQOL)' model. Although there are some conceptual problems with the way the argument is presented, even those who do not share Camosy's Catholic background have good reason to accept his key point that resources should be considered in intensive care treatment decisions for all patients. In the second part of the paper, I explore the ways in which we might identify which infants are too expensive to treat. I argue that both traditional personal 'quality of life' and Camosy's 'sQOL' should factor into these decisions, and I outline two practical proposals.

<http://www.ncbi.nlm.nih.gov/pubmed/23355229>

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## What parents want from doctors in end-of-life decision-making for children

Sullivan, J., et al.

*Arch Dis Child* 2014; 99(3): 216-220

OBJECTIVE: End-of-life decision-making is difficult for everyone involved, as many studies have shown. Within this complexity, there has been little information on how parents see the role of doctors in end-of-life decision-making for children. This study aimed to examine parents' views and experiences of end-of-life decision-making. DESIGN: A qualitative method with a semistructured interview design was used. SETTING: Parent participants were living in the community. PARTICIPANTS: Twenty-five bereaved parents. MAIN OUTCOMES: Parents reported varying roles taken by doctors: being the provider of information without opinion; giving information and advice as to the decision that should be taken; and seemingly being the decision maker for the child. The majority of parents found their child's doctor enabled them to be the ultimate decision maker for their child, which was what they very clearly wanted to be, and consequently enabled them to exercise their parental autonomy. Parents found it problematic when doctors took over decision-making. A less frequently reported, yet significant role for doctors was to affirm decisions after they had been made by parents. Other important aspects of the doctor's role were to provide follow-up support and referral. CONCLUSIONS: Understanding the role that doctors take in end-of-life decisions, and the subsequent impact of that role from the perspective of parents can form the basis of better informed clinical practice.

<http://www.ncbi.nlm.nih.gov/pubmed/24311188>

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## Transfusion in critically ill children: an ongoing dilemma

Secher, E. L., et al.

*Acta Anaesthesiol Scand* 2013; 57(6): 684-691

Transfusion of blood products is a cornerstone in managing many critically ill children. Major improvements in blood product safety have not diminished the need for caution in transfusion practice. In this review, we aim to discuss the interplay between benefits and potential adverse effects of transfusion in critically ill children by including 65 papers, which were evaluated based on previously agreed selection criteria. Current practice on transfusing critically ill children is mainly founded on the basis of adult studies, common practices with cut-off values, and expert opinions, rather than evidence-based medicine. Paediatric patients have explicit physiological challenges and requirements to be addressed. Critically ill children often suffer from anaemia, have substantial iatrogenic blood loss with subsequent transfusions, and are at a higher risk of complications, often due to human errors. Transfusion in children is associated with increased morbidity. A restrictive transfusion strategy is not associated with increased morbidity. Thus, transfusion in paediatrics should be considered a high-risk treatment and requires individual clinical assessment. Current level of evidence support the notion that in most stable cases, despite high severity of illness (cyanotic children and neonates excluded), a restrictive haemoglobin threshold of 70 g/l (4.3 mmol/l) is no more harmful than to transfuse at a liberal trigger, e.g. haemoglobin 95 g/l (5.9 mmol/l). Thus, balanced against potential benefits and often its necessity, a restrictive approach may be appropriate due to the associated risks of transfusion.

<http://www.ncbi.nlm.nih.gov/pubmed/23692309>

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## Judging the quality of mercy: Drawing a line between palliation and euthanasia

Morrison, W. and T. Kang

*Pediatrics* 2014; 133 Suppl 1: S31-36

Clinicians frequently worry that medications used to treat pain and suffering at the end of life might also hasten death. Intentionally hastening death, or euthanasia, is neither legal nor ethically appropriate in children. In this article, we explore some of the historical and legal background regarding appropriate end-of-life care and outline what distinguishes it from euthanasia. Good principles include clarity of goals and assessments, titration of medications to effect, and open communication. When used appropriately, medications to treat symptoms should rarely hasten death significantly. Medications and interventions that are not justifiable are also discussed, as are the implications of palliative sedation and withholding fluids or nutrition. It is imperative that clinicians know how to justify and use such medications to adequately treat suffering at the end of life within a relevant clinical and legal framework.

<http://www.ncbi.nlm.nih.gov/pubmed/24488538>

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## Cancer in adolescents and young adults in countries with limited resources

Magrath, I. and S. Epelman

*Curr Oncol 2013; Rep 15(4): 332-346*

Cancer in adolescents and young adults (AYA) represents a higher fraction of all cancer in countries that are still undergoing a demographic transition. Such countries tend to have much younger populations, and therefore unless they have a particularly low incidence of cancer in this age group, will have a higher burden of cancer (absolute number of cases with cancer) in AYA. Cancers in AYA are comprised of the tail end of the incidence curve of cancers that have their peak incidence, or occur almost exclusively in childhood, the beginning of the incidence curve of cancers that primarily affect the elderly, and a third set of cancers that have their peak incidence (or are at least common) in the AYA age group (e.g., testicular cancer, sarcomas, melanoma, thyroid cancer). Many, but not all, of these cancers require radiation or cancer surgery, but the poorest countries do not have a sufficient number of radiation therapy units and surgical oncologists, or indeed medical and pediatric oncologists, to deal with the burden of cancer they face. The AYA age group is particularly important, both with regard to their contribution to the economy now and in the future (the majority are in the “working” age-group defined as 15-64 years), as well as their important role in caring for their families. Moreover, some of these cancers are eminently curable with chemotherapy alone, and more could be cured by simply improving the efficiency of existing health services and providing education and training to both the public as well as oncologists and other specialists required for the care of AYA (although such individuals will not necessarily be exclusively concerned with this age group). Of particular importance is the detection and diagnosis of cancer patients at the earliest possible time in the course of their disease. Avoiding delays in initiating therapy, which are partly due to the poverty and lack of education of the public as well as to a failure on the part of primary health care providers to recognize the possibility of cancer, would lead not only to improved survival and less toxicity, but is likely to reduce the need for radiation as well as the cost of treatment. There are few good quality clinical trials that take place in the LMIC (in relationship to the extent of the existing cancer burden), and research training should be an integral component of capacity building. Research on the efficacy and toxicity of standardized treatment approaches that are either based on principles established in the HIC, or adapted from treatment protocols used in the HIC, would be a good place to begin, but health policy and multisectoral collaboration are essential if improved survival rates are to be achieved. Decisions will also need to be made regarding the treatment of diseases in which radiation or cancer surgery are important elements, when one or both of the latter are unavailable. Late effects are important in this young population in HIC, and protocol adaptations or design in LMIC should take into consideration the significant fraction of cured patients with late effects who were treated in HIC in an era where improving response and survival rates was the paramount consideration—the situation that applies today in less developed countries. Special adolescent units which better deal with psychological issues of young cancer patients are rare in LMIC and the psychosocial issues faced by adolescents are much less studied. Although survival is the first consideration, attention to psychosocial and financial issues may reduce existing delays in initiating therapy and also the fraction of patients that abandon therapy.

<http://www.ncbi.nlm.nih.gov/pubmed/23832788>

## The ethics consultation and the pediatric surgeon

Statter, M. B.

*Semin Pediatr Surg* 2013; 22(3): 149-153

The cultural, ethnic, religious, socioeconomic, and educational diversity of the patient population and the expanded surgical options provided by innovation and technology can pose significant ethical challenges. The questions confronting pediatric surgeons and their patients' families have greater complexity, and both the pediatric surgeon and the family perceive increasing vulnerability and uncertainty. The analysis and management of ethical issues in pediatric surgery cannot simply be extrapolated from the approach applied to adult cases. By reviewing the history of the events that contributed to the creation and utilization of hospital ethics committees and examining the role of the ethics consultant in the context of pediatric surgical care, practitioners and trainees will be better able to address these multifaceted situations.

<http://www.ncbi.nlm.nih.gov/pubmed/23870209>

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## Towards evidence-based dosing regimens in children on the basis of population pharmacokinetic pharmacodynamic modelling

Admiraal, R., et al.

*Arch Dis Child* 2014; 99(3): 267-272

When growing up, the pharmacokinetic (PK) and pharmacodynamic (PD) profiles of drugs change, which may alter the effect of drugs. To ensure optimal drug efficacy and safety in paediatric care, PK and PD relationships of drugs need to be explored in children. This article presents an outline on performing a population PK/PD study and translating these results into rational dosing regimens, with the development and prospective evaluation of PK/PD derived evidence-based dosing regimen being discussed. Examples on amikacin, morphine and busulfan are provided, showing how PK/PD modelling not only led to optimization and individualization in paediatric clinical care for the specific drugs but also to insight in maturation of organ systems involved. It is shown that the latter results can subsequently be used as a basis for dosing of other drugs eliminated through the same pathway. Ultimately, these efforts should lead to predictable drug efficacy and safety across all age groups.

<http://www.ncbi.nlm.nih.gov/pubmed/24356807>

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## Status dystonicus: A practice guide

Allen, N. M., et al.

*Dev Med Child Neurol* 2014; 56(2): 105-112

Status dystonicus is a rare, but life-threatening movement disorder emergency. Urgent assessment is required and management is tailored to patient characteristics and complications. The use of dystonia action plans and early recognition of worsening dystonia may potentially facilitate intervention or prevent progression to status dystonicus. However, for established status dystonicus, rapidly deployed temporizing measures and different depths of sedation in an intensive care unit or high dependency unit are the most immediate and effective modalities for abating life-threatening spasms, while dystonia-specific treatment takes effect. If refractory status dystonicus persists despite orally active anti-dystonia drugs and unsuccessful weaning from sedative or anaesthetic agents, early consideration of intrathecal baclofen or deep brain stimulation is required. During status dystonicus, precise documentation of dystonia sites and severity as well as the baseline clinical state, using rating scales and videos is recommended. Further published descriptions of the clinical nature, timing of evolution, resolution, and epidemiology of status dystonicus are essential for a better collective understanding of this poorly understood heterogeneous emergency. In this review, we provide an overview of the clinical presentation and suggest a management approach for status dystonicus.

<http://www.ncbi.nlm.nih.gov/pubmed/24304390>

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## Improving the practice of child death overview panels: A paediatric perspective

Allen, L., et al

*Arch Dis Child* 2014; 99(3): 193-196

**OBJECTIVE:** In England, every death in childhood is reviewed by a local multidisciplinary Child Death Overview Panel (CDOP) with the intention of understanding causation and implementing interventions to reduce future deaths. This study aimed to establish how well panels work from the perspective of the paediatricians involved and to ascertain whether they deliver good value and identify areas for improvement. **DESIGN:** A questionnaire was sent to every CDOP paediatrician in the country (n=93). Questions focused on the quality of CDOP case discussions as well as examples of effective and significant recommendations. Responses were analysed using simple quantitative and qualitative methods. **RESULTS:** 84/93 (90%) of the paediatricians responded. Among the respondents, 60 (71%) believe that investment in CDOPs is offering good value, 73 (87%) feel that case discussions are rigorous and consistent and over 90% believe that the correct issues are emerging from discussions. However, responders noted many areas for improvement: 40 (48%) suggested devolving the discussion of specialist deaths (eg, neonates) to hospital-based review meetings or holding themed meetings with invited specialists, 11 (13%) suggested filtering out cases where learning is unlikely before full CDOP meetings and 13 (15%) called for national integration and analysis of data. **CONCLUSIONS:** In this time of economic austerity it is vital that the CDOPs add value to the invested resources. Although CDOP paediatricians feel that panels are working well, there is scope for improvement through enhancing relationships with commissioning bodies, aggregate review and analysis of CDOP data at a national level and consideration of specialist and/or network review of certain categories of deaths such as cardiac surgery, oncology and neonates.

<http://www.ncbi.nlm.nih.gov/pubmed/24255566>

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## Advancing the management of childhood epilepsies

Cross, J. H., et al.

*Eur J Paediatr Neurol* 2013; 17(4): 334-347

Childhood epilepsies comprise a heterogeneous group of disorders and syndromes that vary in terms of severity, prognosis and treatment requirements. Effective management requires early, accurate recognition and diagnosis, and a holistic approach that addresses each individual's medical and psychosocial needs within the context of their overall health status and quality of life. With increasing understanding of underlying aetiologies, new approaches to management and treatment are emerging. For example, genetic testing is beginning to provide a tool to aid differential diagnosis and a means of predicting predisposition to particular types of epilepsy. Despite the availability of an increasing number of antiepileptic drugs (AEDs)--due not only to the development of new AEDs, but also to changes in regulatory requirements that have facilitated clinical development--seizure control and tolerability continue to be suboptimal in many patients, and there is therefore a continuing need for new treatment strategies. Surgery and other non-pharmacological treatments (e.g. vagus nerve stimulation, ketogenic diet) are already relatively well established in paediatric epilepsy. New pharmacological treatments include generational advances on existing AEDs and AEDs with novel modes of action, and non-AED pharmacological interventions, such as immunomodulation. Emerging technologies include novel approaches allowing the delivery of medicinal agents to specific areas of the brain, and 'closed-loop' experimental devices employing algorithms that allow treatment (e.g., electrical stimulation) to be targeted both spatially and temporally. Although in early stages of development, cell-based approaches (e.g., focal targeting of adenosine augmentation) and gene therapy may also provide new treatment choices in the future.

<http://www.ncbi.nlm.nih.gov/pubmed/23558251>

## Bronchopulmonary dysplasia: A review

Ali, Z., et al.

*Arch Gynecol Obstet* 2013; 288(2): 325-333

INTRODUCTION: The prevalence of bronchopulmonary dysplasia (BPD), one of the most frequently occurring complications following preterm birth, is increasing due to increased survival of preterm infants. METHODS: Systematic literature review. CONCLUSION: The etiology is multifactorial, with prematurity being a prerequisite for the development of BPD. Over time, there have been many different and new treatment modalities, some of them have reduced the severity of the disease, but none of them have been able to impact upon the increasing incidence of BPD.

<http://www.ncbi.nlm.nih.gov/pubmed/23420126>

## Effect of communication skills training for residents and nurse practitioners on quality of communication with patients with serious illness: A randomized trial

Curtis, J. R., et al.

*JAMA* 2013; 310(21): 2271-2281

IMPORTANCE: Communication about end-of-life care is a core clinical skill. Simulation-based training improves skill acquisition, but effects on patient-reported outcomes are unknown. OBJECTIVE: To assess the effects of a communication skills intervention for internal medicine and nurse practitioner trainees on patient- and family-reported outcomes. DESIGN, SETTING, AND PARTICIPANTS: Randomized trial conducted with 391 internal medicine and 81 nurse practitioner trainees between 2007 and 2013 at the University of Washington and Medical University of South Carolina. INTERVENTION: Participants were randomized to an 8-session, simulation-based, communication skills intervention (N = 232) or usual education (N = 240). MAIN OUTCOMES AND MEASURES: Primary outcome was patient-reported quality of communication (QOC; mean rating of 17 items rated from 0-10, with 0 = poor and 10 = perfect). Secondary outcomes were patient-reported quality of end-of-life care (QEOLC; mean rating of 26 items rated from 0-10) and depressive symptoms (assessed using the 8-item Personal Health Questionnaire [PHQ-8]; range, 0-24, higher scores worse) and family-reported QOC and QEOLC. Analyses were clustered by trainee. RESULTS: There were 1866 patient ratings (44% response) and 936 family ratings (68% response). The intervention was not associated with significant changes in QOC or QEOLC. Mean values for postintervention patient QOC and QEOLC were 6.5 (95% CI, 6.2 to 6.8) and 8.3 (95% CI, 8.1 to 8.5) respectively, compared with 6.3 (95% CI, 6.2 to 6.5) and 8.3 (95% CI, 8.1 to 8.4) for control conditions. After adjustment, comparing intervention with control, there was no significant difference in the QOC score for patients (difference, 0.4 points [95% CI, -0.1 to 0.9]; P = .15) or families (difference, 0.1 [95% CI, -0.8 to 1.0]; P = .81). There was no significant difference in QEOLC score for patients (difference, 0.3 points [95% CI, -0.3 to 0.8]; P = .34) or families (difference, 0.1 [95% CI, -0.7 to 0.8]; P = .88). The intervention was associated with significantly increased depression scores among patients of postintervention trainees (mean score, 10.0 [95% CI, 9.1 to 10.8], compared with 8.8 [95% CI, 8.4 to 9.2]) for control conditions; adjusted model showed an intervention effect of 2.2 (95% CI, 0.6 to 3.8; P = .006). CONCLUSIONS AND RELEVANCE: Among internal medicine and nurse practitioner trainees, simulation-based communication training compared with usual education did not improve quality of communication about end-of-life care or quality of end-of-life care but was associated with a small increase in patients' depressive symptoms. These findings raise questions about skills transfer from simulation training to actual patient care and the adequacy of communication skills assessment. TRIAL REGISTRATION: clinicaltrials.gov Identifier: NCT00687349.

<http://www.ncbi.nlm.nih.gov/pubmed/24302090>

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<http://www.ncbi.nlm.nih.gov/pubmed/24302090>

## Summary of the British Transplantation Society guidelines for transplantation from donors after deceased circulatory death

Andrews, P. A., et al.

*Transplantation* 2014; 97(3): 265-270

The second edition of the British Transplantation Society Guidelines for Transplantation from Donors after Deceased Circulatory Death was published in June 2013. The guideline has been extensively revised since the previous edition in 2004 and has used the GRADE system to rate the strength of evidence and recommendations. This article summarizes the Statements of Recommendation contained in the guideline, which provide a framework for transplantation after deceased circulatory death in the U.K. and may be of wide international interest. It is recommended that the full guideline document is consulted for details of the relevant references and evidence base.

[http://www.bts.org.uk/MBR/Clinical/Guidelines/Current/Member/Clinical/Current\\_Guidelines.aspx](http://www.bts.org.uk/MBR/Clinical/Guidelines/Current/Member/Clinical/Current_Guidelines.aspx).

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## Safety in pediatric imaging: An update

Arthurs, O. J. and A. A. Bjorkum

*Acta Radiol* 2013; 54(9): 983-990

Many assumptions are made when imaging children. In particular a judgement is made regarding how safe or unsafe each imaging modality is, using relatively arbitrary definitions and distinctions, due to the lack of robust scientific data. Here, the latest evidence is reviewed, particularly regarding the medical exposure to ionizing radiation (X-rays and CT) and MRI in childhood. The best evidence currently available suggests a small but convincing risk of cumulative low-dose ionizing radiation in children. Given our predictions for the children imaged today, it seems reasonable to pursue non-ionizing-based techniques wherever possible, although there is emerging evidence that MRI and ultrasound may have hitherto unknown effects. As our knowledge base expands, we must continually review our practice in light of the latest scientific data.

<http://www.ncbi.nlm.nih.gov/pubmed/23550188>

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## Educating children's nurses for communicating bad news

Crawford, D., et al.

*Nurs Child Young People* 2013; 25(8): 28-33; quiz 36

Some parents are unhappy with the way news is broken to them. This article seeks to educate and inform the reflective practitioner on a series of communication strategies to enhance their skills. This is important because the way news is disclosed can affect the way news is accepted and the level of support the family will require. The importance of clarity, honesty and empathy is emphasised.

<http://www.ncbi.nlm.nih.gov/pubmed/24112022>

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## Role of MRI in patient selection for surgical treatment of intractable epilepsy in infancy

Daghistani, R. and E. Widjaja

*Brain Dev* 2013; 35(8): 697-705

Epilepsy surgery is an effective treatment in selected patients with localization-related intractable epilepsy. The success of epilepsy surgery is in part dependent upon identification of a lesion on MRI. In infants, the surgical epileptogenic substrates include focal cortical dysplasia (FCD), hemimegalencephaly, tuberous sclerosis complex, Sturge Weber syndrome, hypoxic-ischemic or cerebrovascular injury and low-grade tumour. The sensitivity of MRI in identifying the epileptogenic substrate is influenced by the nature of the epileptogenic substrate, MRI technique and expertise of the interpreting physician. The MRI features of some lesions such as FCD may differ in infants compared to children and adults; the white matter adjacent to FCD may demonstrate lower T2 and higher T1 signal in some infants due to premature myelination, while in others, the white matter demonstrates higher T2 or lower T1 signal due to demyelination, dysmyelination or gliosis, similar to children and adults. The appearances of some lesions, such as FCD, may change with time, due to brain maturation or seizure related changes. MRI for patients with localization-related intractable epilepsy should have high-resolution, multiplanar and multisequence. In infants, volumetric T1 and high-resolution T2 imaging are recommended. FLAIR and proton density sequences are less helpful in infants due to lack of myelin in the white matter. The physician interpreting the scan should be familiar with the imaging appearances of epileptogenic substrates and may need to review the scan more than once if a lesion is not seen on initial inspection.

<http://www.ncbi.nlm.nih.gov/pubmed/23632127>

## Motor and cognitive assessment of infants and young boys with Duchenne Muscular Dystrophy: Results from the Muscular Dystrophy Association DMD Clinical Research Network

Connolly, A. M., et al.

*Neuromuscul Disord* 2013; 23(7): 529-539

Therapeutic trials in Duchenne Muscular Dystrophy (DMD) exclude young boys because traditional outcome measures rely on cooperation. The Bayley III Scales of Infant and Toddler Development (Bayley III) have been validated in developing children and those with developmental disorders but have not been studied in DMD. Expanded Hammersmith Functional Motor Scale (HFMSE) and North Star Ambulatory Assessment (NSAA) may also be useful in this young DMD population. Clinical evaluators from the MDA-DMD Clinical Research Network were trained in these assessment tools. Infants and boys with DMD (n = 24; 1.9 +/- 0.7 years) were assessed. The mean Bayley III motor composite score was low (82.8 +/- 8; p <= .0001) (normal = 100 +/- 15). Mean gross motor and fine motor function scaled scores were low (both p <= .0001). The mean cognitive comprehensive (p=.0002), receptive language (p <= .0001), and expressive language (p = .0001) were also low compared to normal children. Age was negatively associated with Bayley III gross motor (r = -0.44; p = .02) but not with fine motor, cognitive, or language scores. HFMSE (n=23) showed a mean score of 31 +/- 13. NSAA (n = 18 boys; 2.2 +/- 0.4 years) showed a mean score of 12 +/- 5. Outcome assessments of young boys with DMD are feasible and in this multicenter study were best demonstrated using the Bayley III.

## A comparison of burnout among oncology nurses working in adult and pediatric inpatient and outpatient settings

Davis, S., et al.

*Oncol Nurs Forum* 2013; 40(4): E303-311

PURPOSE/OBJECTIVES: To investigate differences in burnout among oncology nurses by type of work setting, coping strategies, and job satisfaction. DESIGN: Descriptive. SETTING: A metropolitan cancer center. SAMPLE: A convenience sample of 74 oncology nurses. METHODS: Participants completed a demographic data form, the Nursing Satisfaction and Retention Survey, and the Maslach Burnout Inventory. MAIN RESEARCH VARIABLES: Burnout, coping strategies, job satisfaction, and oncology work setting (inpatient versus outpatient and adult versus pediatric). FINDINGS: The participants most often used spirituality and co-worker support to cope. Emotional exhaustion was lowest for youngest nurses and highest for outpatient RNs. Personal accomplishment was highest in adult settings. Job satisfaction correlated inversely with emotional exhaustion and the desire to leave oncology nursing. CONCLUSIONS: The findings support that the social context within the work environment may impact emotional exhaustion and depersonalization, and that demographics may be more significant in determining burnout than setting. IMPLICATIONS FOR NURSING: The findings raise questions of whether demographics or setting plays a bigger role in burnout and supports organizational strategies that enhance co-worker camaraderie, encourage nurses to discuss high-stress situations, and share ways to manage their emotions in oncology settings. KNOWLEDGE TRANSLATION: Spirituality and co-worker relationships were positive coping strategies among oncology nurses to prevent emotional exhaustion. Nurses who rely on supportive social networks as a coping mechanism have lower levels of depersonalization. Age was inversely related to emotional exhaustion.

<http://www.ncbi.nlm.nih.gov/pubmed/23803274>

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## Physicians' conceptualization of closure as a benefit of physician-parent follow-up meetings after a child's death in the pediatric intensive care unit

Eggle, S., et al.

*J Palliat Care* 2013; 29(2): 69-75

We examined physicians' conceptualization of closure as a benefit of follow-up meetings with bereaved parents. The frequency of use and the meaning of the word "closure" were analyzed in transcripts of interviews with 67 critical care physicians affiliated with the Collaborative Pediatric Critical Care Research Network. In all, 38 physicians (57 percent) used the word "closure" at least once (median: 2; range: 1 to 7), for a total of 86 times. Physicians indicated that closure is a process or trajectory rather than an achievable goal. They also indicated that parents and physicians can move toward closure by gaining a better understanding of the causes and circumstances of the death and by reconnecting with, or resolving relationships between, parents and health professionals. Physicians suggested that a primary reason to conduct follow-up meetings is that such meetings offer parents and physicians an opportunity to move toward closure. Future research should attempt to determine whether followup meetings reduce the negative effects of bereavement for parents and physicians.

<http://www.ncbi.nlm.nih.gov/pubmed/23923469>

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## Pattern and predictors of complementary and alternative medicine (CAM) use among pediatric patients with epilepsy

Doering, J. H., et al.

*Epilepsy Behav* 2013; 29(1): 41-46

Parents of pediatric patients with chronic conditions such as epilepsy increasingly opt for complementary and alternative medicine (CAM). However, data on the pattern and reasons of CAM use in childhood epilepsy are scarce. The objectives of this study were as follows: first, to characterize CAM use among pediatric patients with epilepsy by assessing its spectrum, prevalence, costs, and frequency of use; second, to evaluate the influence of CAM use on compliance and satisfaction with conventional care as well as to explore parent-child neurologist communication concerning CAM; and third, to investigate predictors of CAM use. A postal survey was administered to all parents of pediatric outpatients with epilepsy aged 6 to 12, who have received treatment at the neuropaediatric outpatient clinic of the University Children's Hospital Heidelberg between 2007 and 2009. One hundred thirty-two of the 297 distributed questionnaires were suitable for inclusion in statistical analysis (44.7%). Forty-nine participants indicated that their children used CAM during the previous year (37.1%). Thirty different types of CAM were used, with homeopathy (55.1%), osteopathy (24.5%), and kinesiology (16.3%) being the most commonly named. A mean of 86euro (0euro-500euro) and 3h (1 h-30 h) per month was committed to CAM treatment. Only 53% of the users informed their child neurologist of the additional CAM treatment, while 85.6% of all parents wished to discuss CAM options with their child neurologist. Seventy-five percent of users considered the CAM treatment effective. Among the participants most likely to seek CAM treatment are parents whose children show a long duration of epileptic symptoms, parents who make use of CAM treatment themselves, and parents who value a holistic and natural treatment approach. A substantial portion of pediatric patients with epilepsy receive CAM treatment. The high prevalence of use and significant level of financial and time resources spent on CAM indicate the high importance of these treatment options for parents. On the other hand, communication concerning CAM with the child neurologist is largely insufficient despite the wish to speak about CAM. Complementary and alternative medicine users' high compliance with conventional treatment and high perceived effectiveness of CAM support an integrative approach to CAM for pediatric patients with epilepsy. Our study implies that in addition to open parent-child neurologist communication, active inquiry on CAM treatments is necessary to enable informed decision making by parents and to establish the suitability of CAM treatment for the patient. Reliable predictors for CAM use, which allow for improved identification of patients with a high likelihood to receive CAM treatment, are the duration of the illness, use of CAM by the parents themselves, and the desire of the parents to receive a holistic and natural treatment for their child.

<http://www.ncbi.nlm.nih.gov/pubmed/23933629>

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## The pediatric surgeon and palliative care

Feudtner, C. and T. A. Blinman

*Semin Pediatr Surg* 2013; 22(3): 154-160

Palliative care is now a core component of pediatric care for children and families who are confronting serious illness with a low likelihood of survival. Pediatric surgeons, in partnership with pediatric palliative care teams, can play a pivotal role in assuring that these patients receive the highest possible quality of care. This article outlines a variety of definitions and conceptual frameworks, describes decision-making strategies and communication techniques, addresses issues of interdisciplinary collaboration and personal self-awareness, and illustrates these points through a series of case vignettes, all of which can help the pediatric surgeon perform the core tasks of pediatric palliative care.

<http://www.ncbi.nlm.nih.gov/pubmed/23870210>



## End-of-life care in Toronto neonatal intensive care units: Challenges for physician trainees

El Sayed, M. F., et al.

*Arch Dis Child Fetal Neonatal Ed* 2013; 98(6): F528-533

**BACKGROUND:** Physician trainees in neonatology can find it extremely challenging to care for patients from diverse linguistic and multicultural backgrounds. This challenge is particularly highlighted when difficult and ethically challenging end-of-life (EOL) decision-making with parents is required. While these interactions are an opportunity for growth and learning, they also have the potential to lead to misunderstanding and uncertainty and can add to trainees' insecurity, unpreparedness and stress when participating in such interactions. **OBJECTIVES:** To explore the challenges for trainees when EOL decisions are undertaken and to encourage them to reflect on how they might influence such decision-making. **DESIGN AND INTERVIEW:** An in-depth, semi-structured interview guide was developed: the interview questions address trainees' beliefs, attitudes, preferences and expectations regarding discussions of EOL neonatal care. Twelve interviews were completed and the audio records transcribed verbatim, after removal of identifying personal information. **RESULTS:** Participants identified six domains of challenge in EOL care: withdrawal of life-sustaining treatment based on poor outcome, explaining 'no resuscitation options' to parents, clarifying 'do not resuscitate (DNR)' orders, empowering families with knowledge and shared decision-making, dealing with different cultures and managing personal internal conflict. Participants experienced the most difficulty during the initial stages of training and eventually reported good knowledge of the EOL care process. They had a sense of security and confidence working within a multidisciplinary care team, which includes experienced nursing staff as well as bereavement and palliative care coordinator within the neonatal intensive care unit. **CONCLUSIONS:** The challenges experienced by physician trainees when providing EOL care can serve as focal points for improving EOL educational programmes for neonatal fellowship training.

<http://www.ncbi.nlm.nih.gov/pubmed/23899436>

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## The adolescent and young adult with cancer: State of the art-brain tumor

Epelman, S.

*Curr Oncol Rep* 2013; 15(4): 308-316

The management of adolescents and young adults with brain tumors, which consist of many different histologic subtypes, continues to be a challenge. Better outcome with a decrease of the side effects of the disease and therapy and improvement of quality of life has been demonstrated in recent decades for some tumors. Significant differences in survival and cure are also observed between adult and pediatric tumors of the same histologic grade. Genetic, developmental, and environmental factors likely influence the type of tumor and response observed, even though no clear pathologic features differentiate these lesions among children, adolescents, and adults. Similarly, treatment strategies are not identical among these populations; most patients receive surgery, followed by radiation therapy and multiagent chemotherapy. Advances in understanding the biology underlying the distribution of tumors in adolescents and young adults may influence the development of prospective trials. A more individualized view of these tumors will likely influence stratification of patients in future studies as well as selection for targeted agents. Accordingly, outcomes may improve and long-term morbidities may decrease.

<http://www.ncbi.nlm.nih.gov/pubmed/23737251>

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## Surgical intervention for feeding and nutrition difficulties in cerebral palsy: A systematic review

Ferluga, E. D., et al.

*Dev Med Child Neurol* 2014; 56(1): 31-43.

AIM: The aim of the study was to systematically review surgical intervention for feeding difficulties in cerebral palsy. METHOD: We searched databases including MEDLINE from 1980 to July 2012. Two reviewers independently assessed studies and rated the overall quality and strength of the evidence. RESULTS: Thirteen publications (11 unique studies) met the inclusion criteria and addressed gastrostomy outcomes or treatment of reflux via fundoplication. In nine studies, gastrostomy-fed children gained weight. Relative to typically developing populations, baseline weight z-scores ranged from -3.56 to -0.39 and follow-up z-scores ranged from -2.63 to -0.33. Other growth measures were mixed. Two studies assessed fundoplication: in one, both Nissen fundoplication and vertical gastric plication reduced reflux (by 57% and 43% respectively), while in one case series, reflux recurred within 12 months in 30% of children. The highest rates of adverse events across studies were site infection (59%), granulation tissue (42%), and recurrent reflux (30%). Death rates ranged from 7 to 29%; however, the underlying cause was probably not surgery. INTERPRETATION: Evidence for the effectiveness of surgical interventions is insufficient to low. Studies of gastrostomy typically demonstrated significant weight gain. Results for other measures were mixed. Many children remained underweight, although, given a lack of appropriate reference standards, these results should be interpreted cautiously.

<http://www.ncbi.nlm.nih.gov/pubmed/23738903>

## Skeletal muscles of ambulant children with Duchenne muscular dystrophy: Validation of multicenter study of evaluation with MR imaging and MR spectroscopy

Forbes, S. C., et al.

*Radiology* 2013; 269 (1): 198-207.

PURPOSE: To validate a multicenter protocol that examines lower extremity skeletal muscles of children with Duchenne muscular dystrophy (DMD) by using magnetic resonance (MR) imaging and MR spectroscopy in terms of reproducibility of these measurements within and across centers. MATERIALS AND METHODS: This HIPAA-compliant study was approved by the institutional review boards of all participating centers, and informed consent was obtained from each participant or a guardian. Standardized procedures with MR operator training and quality assurance assessments were implemented, and data were acquired at three centers by using different 3-T MR imaging instruments. Measures of maximal cross-sectional area (CSA<sub>max</sub>), transverse relaxation time constant (T<sub>2</sub>), and lipid fraction were compared among centers in two-compartment coaxial phantoms and in two unaffected adult subjects who visited each center. Also, repeat MR measures were acquired twice on separate days in 30 boys with DMD (10 per center) and 10 unaffected boys. Coefficients of variation (CVs) were computed to examine the repeated-measure variabilities within and across centers. Results: CSA<sub>max</sub>, T<sub>2</sub> from MR imaging and MR spectroscopy, and lipid fraction were consistent across centers in the phantom (CV, <3%) and in the adult subjects who traveled to each site (CV, 2%-7%). High day-to-day reproducibility in MR measures was observed in boys with DMD (CSA<sub>max</sub>, CV = 3.7% [25th percentile, 1.3%; 75th percentile, 5.1%]; contractile area, CV = 4.2% [25th percentile, 0.8%; 75th percentile, 4.9%]; MR imaging T<sub>2</sub>, CV = 3.1% [25th percentile, 1.2%; 75th percentile, 4.7%]; MR spectroscopy T<sub>2</sub>, CV = 3.9% [25th percentile, 1.5%; 75th percentile, 5.1%]; and lipid fraction, CV = 4.7% [25th percentile, 1.0%; 75th percentile, 5.3%]). CONCLUSION: The MR protocol implemented in this multicenter study achieved highly reproducible measures of lower extremity muscles across centers and from day to day in ambulatory boys with DMD.

<http://www.ncbi.nlm.nih.gov/pubmed/23696684>

## Signs and symptoms of childhood cancer: A guide for early recognition

Fragkandrea, I., et al.

*Am Fam Physician* 2013; 88(3): 185-192

Although cancer in children is rare, it is the second most common cause of childhood mortality in developed countries. It often presents with nonspecific symptoms similar to those of benign conditions, leading to delays in the diagnosis and initiation of appropriate treatment. Primary care physicians should have a raised index of suspicion and explore the possibility of cancer in children who have worrisome or persisting signs and symptoms. Red flag signs for leukemia or lymphoma include unexplained and protracted pallor, malaise, fever, anorexia, weight loss, lymphadenopathy, hemorrhagic diathesis, and hepatosplenomegaly. New onset or persistent morning headaches associated with vomiting, neurologic symptoms, or back pain should raise concern for tumors of the central nervous system. Palpable masses in the abdomen or soft tissues, and persistent bone pain that awakens the child are red flags for abdominal, soft tissue, and bone tumors. Leukokoria is a red flag for retinoblastoma. Endocrine symptoms such as growth arrest, diabetes insipidus, and precocious or delayed puberty may be signs of endocranial or germ cell tumors. Paraneoplastic manifestations such as opsoclonus-myoclonus syndrome, rheumatic symptoms, or hypertension are rare and may be related to neuroblastoma, leukemia, or Wilms tumor, respectively. Increased suspicion is also warranted for conditions associated with a higher risk of childhood cancer, including immunodeficiency syndromes and previous malignancies, as well as with certain genetic conditions and familial cancer syndromes such as Down syndrome, Li-Fraumeni syndrome, hemihypertrophy, neurofibromatosis, and retinoblastoma.

<http://www.ncbi.nlm.nih.gov/pubmed/23939697>

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## Late effects care as an emerging clinical specialty in paediatric oncology: How to prepare the workforce?

Greenfield, D. and L. Hjorth

*Curr Opin Support Palliat Care* 2013; 7(3): 296-302

**PURPOSE OF REVIEW:** There is an increasing recognition of the healthcare needs of long-term childhood and adolescent cancer survivors, but less information is known about the education and training needs of healthcare professionals. **RECENT FINDINGS:** The need for the provision of late effects care for this cohort of patients is now universally accepted by the paediatric cancer community in the western world. As evidence of healthcare needs become known, internationally agreed evidence-based screening practices are emerging. **SUMMARY:** The next clear step is to ensure that clinical staff, medical and nursing alike, are sufficiently prepared to provide safe, compassionate, patient-centred, contemporaneous and clinically effective late effects care. This review considers evidence for existing educational approaches and considers how we may prepare the workforce.

<http://www.ncbi.nlm.nih.gov/pubmed/23912388>

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## Errors in filling WHO death certificate in children: Lessons from 1251 death certificates

Gupta, N., et al.

*J Trop Pediatr* 2014; 60(1): 74-78

Our objective was to identify the frequency and types of various pediatric death certification errors. All available death certificates (n = 1424, from January to December 2005 and from January to July 2007) were retrieved from medical record library in a tertiary pediatric hospital. These were analyzed retrospectively (66%) and prospectively (34%) for the year 2005 and 2007, respectively. Only 11% of death certificates were filled accurately. In the remaining 89%, the total number of errors ranged from 0-5 per death certificate. The most common major and minor errors were improper sequencing (50.3%) and "absence of time interval" (74.7%), respectively. The combination of major and minor errors was observed in 51.9% of the certificates. The frequency of errors was significantly less in prospective data as compared with retrospective data (p < 0.05). Given the high rate of errors, there is an urgent need to design relevant training programs to streamline this dismal situation.

<http://www.ncbi.nlm.nih.gov/pubmed/23902671>

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## The cooperative international neuromuscular research group Duchenne natural history study: Glucocorticoid treatment preserves clinically meaningful functional milestones and reduces rate of disease progression as measured by manual muscle testing and other commonly used clinical trial outcome measures

Henricson, E. K., et al.

*Muscle Nerve* 2013; 48(1): 55-67

INTRODUCTION: Glucocorticoid (GC) therapy in Duchenne muscular dystrophy (DMD) has altered disease progression, necessitating contemporary natural history studies. METHODS: The Cooperative Neuromuscular Research Group (CINRG) DMD Natural History Study (DMD-NHS) enrolled 340 DMD males, ages 2-28 years. A comprehensive battery of measures was obtained. RESULTS: A novel composite functional "milestone" scale showed clinically meaningful mobility and upper limb abilities were significantly preserved in GC-treated adolescents/young adults. Manual muscle test (MMT)-based calculations of global strength showed that those patients <10 years of age treated with steroids declined by 0.4 +/- 0.39 MMT unit/year, compared with -0.4 +/- 0.39 MMT unit/year in historical steroid-naive subjects. Pulmonary function tests (PFTs) were relatively preserved in steroid-treated adolescents. The linearity and magnitude of decline in measures were affected by maturational changes and functional status. CONCLUSIONS: In DMD, long-term use of GCs showed reduced strength loss and preserved functional capabilities and PFTs compared with previous natural history studies performed prior to the widespread use of GC therapy.

<http://www.ncbi.nlm.nih.gov/pubmed/23649481>

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## Spinal muscle atrophy (SMA) is autosomal recessive and one of the most common inherited lethal diseases in childhood

Islander, G.

*Paediatr Anaesth* 2013; 23(9): 804-816

The spectrum of symptoms of SMA is continuous and varies from neonatal death to progressive symmetrical muscle weakness first appearing in adulthood. The disease is produced by degeneration of spinal motor neurons and can be described in three or more categories: SMA I with onset of symptoms before 6 months of age; SMA II with onset between 6 and 18 months and SMA III, which presents later in childhood. Genetics: The disease is in more than 95% of cases caused by a homozygous deletion in survival motor neuron gene 1 (SMN1). Pathophysiology: The loss of full-length functioning SMN protein leads to a degeneration of anterior spinal motor neurons which causes muscle weakness. Anesthetic risks: Airway: Tracheal intubation can be difficult. Respiration: Infants with SMA I almost always need postoperative respiratory support. Patients with SMA II sometimes need support, while SMA III patients seldom need support. Circulation: Circulatory problems during anesthesia are rare. Anesthetic drugs: Neuromuscular blockers: Patients with SMA may display increased sensitivity to and prolonged effect of nondepolarizing neuromuscular blockers. Intubation without muscle relaxation should be considered. Succinylcholine should be avoided. Opioids: These should be titrated carefully. Anesthetic techniques: All types of anesthetic technique have been used. Although none is absolutely contraindicated, none is perfect: anesthesia must be individualized. Conclusion: The perioperative risks can be considerable and are mainly related to the respiratory system, from respiratory failure to difficult/impossible intubation.

<http://www.ncbi.nlm.nih.gov/pubmed/23601145>

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## Recruitment in pediatric clinical research was influenced by study characteristics and pediatricians' perceptions: A multicenter survey

Kaguelidou, F., et al.

*J Clin Epidemiol* 2013; 66(10): 1151-1157

OBJECTIVES: The aim of this survey was to quantify refusal rates and identify factors of refusal pertaining to studies and recruiting pediatricians in the research recruitment process. STUDY DESIGN AND SETTING: We performed a cross-sectional survey on all clinical studies conducted in six pediatric Clinical Investigation Centers in France over an 18-month period. Data were retrieved using a data collection form for the characteristics of each of the studies included in the survey and a questionnaire addressed to recruiting pediatricians. Multilevel models were used for the statistical analysis. RESULTS: Overall, 145 pediatricians approached the families of 999 children and adolescents for participation in 44 studies. In the 36 of the 44 studies that enrolled subjects, median refusal rate was 12.5% (Q1-Q3, 0-28%). Lower refusal rates were associated with therapeutic drug use as the focus of the study [odds ratio (OR), 0.51; 95% CI: 0.25, 1.05], additional hospital stays required for the study (OR, 0.53; 95% CI: 0.28, 0.99), longer duration of the inclusion visit (OR, 0.93/10 min; 95% CI: 0.87, 1), and recruitment by a pediatrician with university teaching responsibilities (OR, 0.26; 95% CI: 0.10, 0.68). Refusal rate was higher when the recruiting pediatrician perceived the study as generating heavy practical burden for the subject and/or its family (OR, 1.3; 95% CI: 1.17, 1.45). CONCLUSION: Refusal to participate in clinical research was low and was influenced by factors associated to the objectives and conduct of the studies and factors related to the characteristics and perceptions of the recruiting pediatricians.

<http://www.ncbi.nlm.nih.gov/pubmed/23856188>

## A simple method for percutaneous endoscopic gastrostomy tube removal: Tie and retrograde pull

Karakus, S. C., et al.

*J Pediatr Surg* 2013; 48(8): 1810-1812

**BACKGROUND/PURPOSE:** Various techniques have been presented to remove the percutaneous endoscopically placed gastrostomy tube in children, but tubes with semi-rigid internal retaining discs are difficult or impossible to remove by external traction. We describe a simple and effective endoscopic removal technique that should be applicable to any type of percutaneous endoscopic gastrostomy tube. **METHODS:** Percutaneous endoscopic gastrostomy tube removal was performed with the tie and retrograde pull technique. After a polypropylene suture was placed and tied 1 cm over the skin level, the percutaneous endoscopic gastrostomy tube was cut 0.5 cm over the knot. The suture was cut from the connection point between the needle and the suture. The distal end of the suture was pushed through the stoma into the stomach. Then a forceps was inserted through the gastroscope. The suture was caught, and the residual percutaneous endoscopic gastrostomy portion was retrieved via retrograde traction on the suture. **RESULTS:** The causes of exchange were determined to be planned tube replacement in 9, buried bumper syndrome in 1, and tube occlusion in 3 patients. The mean tube dwell time was 10.8 +/- 3.9 months. Esophageal mucosal tear developed in 1 patient with epidermolysis bullosa during removal. No other complications occurred during PEG tube exchanges. **CONCLUSION:** This is a rapid and useful technique that does not require any complex endoscopic devices.

<http://www.ncbi.nlm.nih.gov/pubmed/23932627>

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## Prognostic factors of infantile spasms: Role of treatment options including a ketogenic diet

Lee, J., et al.

*Brain Dev* 2013; 35(8): 821-826

**OBJECTIVES:** The aim of this study was to provide additional evidences on prognostic factors for infantile spasms and the possible role of a ketogenic diet. **METHODS:** A retrospective analysis was performed for patients with infantile spasms who had been followed up for more than 6 months between January 2000 and July 2012 at Samsung Medical Center (Seoul, Republic of Korea). We analyzed the association between possible prognostic factors and seizure/developmental outcomes. **RESULTS:** Sixty-nine patients were included in this study and their mean follow-up duration was 52.5 (9-147) months. In the patients who had been followed up for more than 2 years, 53.6% (n=30/57) remained seizure-free at the last visit. Sixty patients (86.9%) showed developmental delay at last follow-up. Forty-two patients (60.9%) became spasm-free with one or two antiepileptic drugs, one patient with epilepsy surgery for a tumor, and seven patients with a ketogenic diet after the failure of two or more antiepileptic drugs. The etiology and age of seizure onset were the significant prognostic factors. **CONCLUSIONS:** In this study, about 60% of the patients became spasm-free with vigabatrin and topiramate. Ketogenic diet increased the rate by 10% in the remaining antiepileptic drug resistant patients. However, 86.9% of the patients showed developmental delay, mostly a severe degree. Early diagnosis and prompt application of treatment options such as antiepileptic drugs, a ketogenic diet or epilepsy surgery can improve outcomes in patients with infantile spasms.

<http://www.ncbi.nlm.nih.gov/pubmed/23856457>

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## Skin-to-skin care for dying preterm newborns and their parents: A phenomenological study from the perspective of NICU nurses

Kymre, I. G. and T. Bondas

*Scand J Caring Sci* 2013; 27(3): 669-676

**BACKGROUND:** Consequences of separation between preterm newborns and their parents have been discussed in many aspects, thus skin-to-skin care (SSC) has become common practice in Scandinavian Neonatal Intensive Care Units (NICUs) since the 1980s. The International workshop on Kangaroo Mother Care (KMC), 2009, recommends implementation of continuous KMC as the gold standard pervading all medical and nursing care, based on empirical studies and clinical guidelines and they suggest that KMC may be used during terminal care in agreement with parents. Parents have a strong desire to be near their child and give support and emotional comfort when the condition of the child requires it, and it has been suggested that medical staff expect parents to be with the neonates, and therefore, encourages them to hold the neonate while it is dying. The practice of SSC at the end of life has been under-researched, however. **AIM:** The aim of this study, which is part of a larger study on neonatal nursing care, was to describe the phenomenon of how nurses enact SSC for dying preterm newborns and their parents. **DESIGN:** A phenomenological reflective life world design. **SETTING AND PARTICIPANTS:** A purposive sample of 18 nurses from three Scandinavian NICUs. **FINDINGS:** The essential meaning of the phenomenon was expressed as strong belief in the urgency of SSC in providing mutual proximity and comfort for dying preterm newborns and their parents. The nurses act upon this belief and upon an engagement in securing the best possible present and future experiences of being close, in which the SSC is understood as a necessary premise in achieving the intended optimal conditions. The findings are elaborated in relation to previous caring and nursing research and phenomenology. **CONCLUSIONS:** Skin-to-skin care for dying preterm newborns and their parents is the preferred caring practice among Scandinavian NICU nurses who consider it of major importance to facilitate proximity and comfort through SSC when the newborn is still alive. The authors suggest this practical knowledge from NICU nurses perspective to be acknowledged in discussions concerning end-of-life care for preterm newborns and their parents and we recommend more formal establishment of this practice. Further research is needed on parents' experiences of skin-to-skin caring in this vulnerable end of life situation of 'being with' their dying newborn.

<http://www.ncbi.nlm.nih.gov/pubmed/23016802>

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## Towards improved ways of knowing children with profound multiple disabilities: Introducing startle reflex modulation

Lyons, G. S., et al.

*Dev Neurorehabil* 2013; 6(5): 340-344

**OBJECTIVE:** To propose startle reflex modulation (SRM) as an objective measure of emotions of children with profound multiple disabilities (PMD). Knowledge about emotion states of children with PMD is crucial to their individualised care and support. Proxy reporting, observational and physiological measures of emotion are reported in the literature. Despite advances in this science, the rigour of the findings and methods are contested. In this article, we introduce SRM; a neurophysiological measure untried with children with PMD, despite its well-known sensitivity to even subtle changes in affective processing without depending on explicit responses. **RESULTS:** We propose a research agenda that aims to deliver a more comprehensive and accurate profile of the inner states of these children, based upon previous research undertaken using SRM. **CONCLUSION:** It is suggested that this objective measure has potential to provide useful information about the inner emotional states of children with PMD.

<http://www.ncbi.nlm.nih.gov/pubmed/24020879>

## A novel treatment regimen for Duchenne muscular dystrophy

Li, M., et al.

*Neuroreport* 2013; 24(16): 924-927

Duchenne muscular dystrophy (DMD) is the most common, X-linked genetic, skeletal muscle disease, with various regimens of treatment. The objective of this study was to determine the safety and efficacy of a novel treatment regimen for this disease. Thirty boys with DMD were administered prednisone according to the following regimen: in the first year, 1.5 mg/kg/day for the first 3 months, 1.0 mg/kg/day for the next 3 months, 0.75 mg/kg/day for the next 3 months, and 0.5 mg/kg/day for the last 3 months. In the second year, prednisone was administered 0.5 mg/kg on the alternate day for 12 months. The muscle strength (Medical Research Council sum score and Gower's sign), serum enzymes (creatine kinase, creatine kinase isoenzyme-2, and lactate dehydrogenase), pulmonary function (forced vital capacity, maximum voluntary ventilation), body weight, height, and BMI were determined before treatment and 3, 6, 9, 12, and 24 months after treatment. The results showed that the patients' mean Medical Research Council sum score increased from 46.1 at the baseline to 53.6 at 12 months and was maintained at 24 months. Gower's sign disappeared in 22 (73.3%) patients at 12 months and 21 (70.0%) at 24 months. The serum levels of creatine kinase, creatine kinase isoenzyme-2, and lactate dehydrogenase decreased and pulmonary function improved after 24 months of treatment. Significantly increased weight gain, osteoporosis, and cushingoid features were not observed. Our results suggested that this novel prednisone regimen for DMD has similar efficacy and safety as other regimens.

<http://www.ncbi.nlm.nih.gov/pubmed/24045777>

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## Seeing is believing: Reducing misconceptions about children's hospice care through effective teaching with undergraduate nursing students

Price, J., et al.

*Nurse Educ Pract* 2013; 13(5): 361-365

Children's palliative care has evolved in recent years and is now recognised as a distinct area of health and social care practice. Whilst children's hospices are viewed as central to quality care for these children and families, lack of knowledge regarding the exact nature of care they provide exists. Education can go part way to changing attitudes and knowledge about the key contribution of hospices, thus improving future care. Alternative and innovative strategies to stimulate meaningful learning are pivotal to children's nurse education and this paper examines one such innovation adopted with 2nd year children's nursing students. Aiming to help students explore the ethos of children's hospice an educational visit was arranged, followed by an on line discussion. Although some practical challenges were encountered, the visit heightened student awareness moving them from the readily held perception that children's hospices were exclusively for dying children and was viewed by students as more effective than a traditional classroom session.

<http://www.ncbi.nlm.nih.gov/pubmed/23111411>

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## Feasibility and perceived benefits of a framework for physician-parent follow-up meetings after a child's death in the PICU

Meert, K. L., et al.

*Crit Care Med* 2014; 42(1): 148-157

**OBJECTIVE:** To evaluate the feasibility and perceived benefits of conducting physician-parent follow-up meetings after a child's death in the PICU according to a framework developed by the Collaborative Pediatric Critical Care Research Network. **DESIGN:** Prospective observational study. **SETTING:** Seven Collaborative Pediatric Critical Care Research Network-affiliated children's hospitals. **SUBJECTS:** Critical care attending physicians, bereaved parents, and meeting guests (i.e., parent support persons, other health professionals). **INTERVENTIONS:** Physician-parent follow-up meetings using the Collaborative Pediatric Critical Care Research Network framework. **MEASUREMENTS AND MAIN RESULTS:** Forty-six critical care physicians were trained to conduct follow-up meetings using the framework. All meetings were video recorded. Videos were evaluated for the presence or absence of physician behaviors consistent with the framework. Present behaviors were evaluated for performance quality using a 5-point scale (1 = low, 5 = high). Participants completed meeting evaluation surveys. Parents of 194 deceased children were mailed an invitation to a follow-up meeting. Of these, one or both parents from 39 families (20%) agreed to participate, 80 (41%) refused, and 75 (39%) could not be contacted. Of 39 who initially agreed, three meetings were canceled due to conflicting schedules. Thirty-six meetings were conducted including 54 bereaved parents, 17 parent support persons, 23 critical care physicians, and 47 other health professionals. Physician adherence to the framework was high; 79% of behaviors consistent with the framework were rated as present with a quality score of 4.3 +/- 0.2. Of 50 evaluation surveys completed by parents, 46 (92%) agreed or strongly agreed the meeting was helpful to them and 40 (89%) to others they brought with them. Of 36 evaluation surveys completed by critical care physicians (i.e., one per meeting), 33 (92%) agreed or strongly agreed the meeting was beneficial to parents and 31 (89%) to them. **CONCLUSIONS:** Follow-up meetings using the Collaborative Pediatric Critical Care Research Network framework are feasible and viewed as beneficial by meeting participants. Future research should evaluate the effects of follow-up meetings on bereaved parents' health outcomes.

<http://www.ncbi.nlm.nih.gov/pubmed/24105453>

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## Diagnosis and management of life-threatening cardiac malformations in the newborn

Mellander, M.

*Semin Fetal Neonatal Med* 2013; 18(5): 302-310

Approximately 1-2 per 1000 newborn babies have a cardiac defect that is potentially life-threatening usually because either the systemic or the pulmonary blood flow is dependent on a patent ductus arteriosus. A significant proportion of newborns with such cardiac defects are being discharged from well-baby nurseries without a diagnosis and therefore risk circulatory collapse and death. This risk is greatest for defects with duct-dependent systemic circulation, notably aortic arch obstruction, but is also significant in transposition of the great arteries, for example. The solution to this problem, apart from improving prenatal detection rates, is to introduce effective neonatal screening including routine pulse oximetry.

## Tracheostomy in mucopolysaccharidosis type II (Hunter's Syndrome)

Malik, V., et al.

*Int J Pediatr Otorhinolaryngol* 2013; 77(7): 1204-1208

**OBJECTIVE:** Patients with mucopolysaccharidosis type II (MPS II) may develop progressive multi-level upper airway obstruction. Despite the unique challenges presented by these complex patients, tracheostomy remains an important intervention to safeguard the airway when other interventions have failed or when the airway obstruction involves multiple sites. Airway involvement is largely responsible for the significant anaesthetic risk seen in MPS II. We reviewed our tertiary unit's experience of tracheostomies in patients with MPS II. **STUDY DESIGN:** Retrospective study. **METHODS:** Case note review of MPS II patients requiring tracheostomy at our tertiary institution. The primary outcome measure used for this study was complications following tracheostomy. **RESULTS:** We identified 10 MPS II patients requiring tracheostomy to manage upper airway obstruction. Mean age at which tracheostomy was 11 years 2 months (range 4 years 6 months to 28 years 10 months). Tracheostomy insertion was indicated in 3 scenarios: (1) to safeguard an anticipated difficult airway prior to a planned non-ENT surgical procedure, (2) to treat refractory progressive upper airway obstruction and (3) emergency airway management. Complications recorded included infratip and suprastomal granulations, local wound infection and skin ulceration from mechanical trauma. There were no immediate postoperative complications. **CONCLUSIONS:** Progressive upper airway obstruction is common in children with MPS II. Tracheostomy is an effective way of managing airway obstruction when less invasive interventions are no longer adequate. Tracheostomy in these patients can be technically difficult and although the complications of tracheostomy in MPS II do not significantly differ from other patient groups, the implications and management complexity vary considerably. The impact of ERT on airway obstruction is not yet fully understood, with tracheostomies likely to remain an important airway adjunct in some patients who fail to respond to ERT, or in those patients surviving into adulthood. It is vital that a multidisciplinary team, comprising clinicians with experience in managing such patients, are involved in airway management of patients with MPS II to enable the best standard of care to be given. The significant additional implications of a tracheostomy in a patient with MPS II, in terms of safety, aftercare and potentially life-threatening complications must be discussed in detail with the patient's family and/or carers. **LEVEL OF EVIDENCE:** 2c.

<http://www.ncbi.nlm.nih.gov/pubmed/23726952>

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## Grief and the experiences of nurses providing palliative care to children and young people at home

Reid, F.

*Nurs Child Young People* 2013; 25(9): 31-36

**AIMS:** To elicit the views of children's nurses with regard to the personal, contextual and interprofessional challenges faced when delivering palliative and end of life care to children and young people in the community. **METHODS:** Semi-structured interviews were conducted with seven nurses who provided palliative care to one or more child or young person in the home. Data generated were analysed thematically to define topics. **FINDINGS:** Four themes emerged: service delivery, nurse-family relationships, nurses' grief, funeral rites and bereavement support. **CONCLUSIONS:** Nurses experienced considerable internal and external pressures. Some are inevitable but others, such as organisation of care provision to families and nurses' personal coping, could be improved by adequately resourced workforces, integrated service structures and guidance on reflective practice. Further research is needed.

<http://www.ncbi.nlm.nih.gov/pubmed/24200187>

## Donation after circulatory death: Current practices, ongoing challenges, and potential improvements

Morrissey, P. E. and A. P. Monaco

*Transplantation* 2014; 97(3): 258-264

Organ donation after circulatory death (DCD) has been endorsed by the World Health Organization and is practiced worldwide. This overview examines current DCD practices, identifies problems and challenges, and suggests clinical strategies for possible improvement. Although there is uniform agreement on DCD donor candidacy (ventilator-dependent individuals with nonrecoverable or irreversible neurologic injury not meeting brain death criteria), there are variations in all aspects of DCD practice. Utilization of DCD organs is limited by hypoxia, hypotension, reduced--then absent--organ perfusion, and ischemia/reperfusion syndrome. Nevertheless, DCD kidneys exhibit comparable function and survival to donors with brain death kidneys, although they have higher rates of primary graft nonfunction, delayed graft function, discard, and retrieval associated injury. Concern over ischemic organ injury underscores the reluctance to recover extrarenal DCD organs since lack of medical therapy to support inadequate allograft function limits their acceptability. Nevertheless, limited results with DCD pancreas, liver, and lung allografts (but not heart) are now approaching that of donors with brain death organs. Pretransplant machine perfusion of DCD kidneys (vs. static storage) may reduce delayed graft function but has no effect on long-term organ function and survival. Normothermic regional perfusion used during DCD abdominal organ retrieval may reduce ischemic organ injury and increase the number of usable organs, although critical confirmative studies have yet to be done. Minor increases in usable DCD kidneys could accrue from increased use of pediatric DCD kidneys and from selective use of DCD/ECD kidneys, whereas a modest increase could result through utilization of donors declared dead beyond 1 hr from withdrawal of life support therapy. A significant increase in transplantable kidneys could be achieved by extension of the concept of living kidney donation in relation to imminent death of potential DCD donors. Progress in research to identify, prevent, and repair DCD-associated organ retrieval injury should improve utilization of DCD organs. Recent results using ex situ pretransplant organ perfusion of DCD organs has been encouraging in this regard.

<http://www.ncbi.nlm.nih.gov/pubmed/24492420>

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## Lived experiences of adult community nurses delivering palliative care to children and young people in rural areas

Reid, F. C.

*Int J Palliat Nurs* 2013; 19(11): 541-547

The anticipated death of a child or young person is a relatively rare occurrence in the Western world. Many families receive support from children's health-care services until the late stages of palliation, with adult community nurses being involved in just an occasional end-of-life care episode in the home during their entire career. This creates challenges in nurses' experiential reflection, development of knowledge and skills, and building of nurse-family relationships. Individual semi-structured interviews were conducted with 10 adult community nurses from a rural part of Scotland to explore their experiences of providing palliative care to children. The material was analysed using a qualitative phenomenological thematic approach. Four key themes emerged: emotional preparedness, navigating the professional 'road', becoming part of the family, and it's everybody's business. Significant issues were highlighted in relation to nurses' coping, with implications for practice. Recommendations are made for further research into rural contextual dilemmas.

<http://www.ncbi.nlm.nih.gov/pubmed/24263898>

## To be a phenomenal doctor you have to be the whole package: physicians' interpersonal behaviors during difficult conversations in paediatrics

Orioles, A., et al.

*J Palliat Med* 2013; 16(8): 929-933

**BACKGROUND:** Delivery of bad news is a challenging task for physicians and other health care professionals. Several studies have assessed parental perceptions of the delivery of bad news, but none have focused on the role of physicians' interpersonal behaviors in the communication process. **OBJECTIVE:** The study's objective was to assess parental perceptions of physicians' interpersonal behaviors and their role in communication of bad news. **DESIGN:** The design was a cross-sectional qualitative interview study of 13 parents of patients hospitalized or previously hospitalized in the pediatric intensive care unit or oncology/bone marrow transplant unit at an academic children's hospital. **RESULTS:** Eleven interpersonal behaviors were identified as important by parents. The majority of parents identified empathy in physicians as critical. Availability, treating the child as an individual, and respecting the parent's knowledge of the child were mentioned by almost half of parents. Themes also considered important but by a smaller number of parents were allowing room for hope, the importance of body language, thoroughness, going beyond the call of duty, accountability, willingness to accept being questioned, and attention to the suffering of the child. **CONCLUSIONS:** To increase parental satisfaction and enhance the parent-physician therapeutic partnership, we recommend that physicians consider attending to the 11 interpersonal behaviors described in this manuscript, and that educational programs pay particular attention to these behaviors when training health care providers in the communication of bad news.

<http://www.ncbi.nlm.nih.gov/pubmed/23808643>

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## Bilateral retinoblastoma: Clinical presentation, management and treatment

Pichi, F., et al.

*Int Ophthalmol* 2013; 33(5): 589-593

Management of retinoblastoma (Rb), the most common intraocular malignant tumor in childhood, is tailored to each individual case and based on the overall situation. We present a case of bilateral Rbs in a 4-month-old girl, referred to our center for bilateral leukocoria. In the right eye, the optic disc was partially visible, and three large foci of retinoblastoma were noted adjective in the vitreous cavity with satellite retinal detachment. The macula was obscured by the tumors. The tumor was therefore classified as group D (International classification of retinoblastoma), and thus underwent intravenous chemotherapy with the standard three-agent protocol of ifosfamide, carboplatin and etoposide (ICE protocol) delivered monthly for six cycles, regressing with a type I calcified pattern. The left eye presented three multifocal yellow-white retinal masses, with a total retinal detachment, and secondary glaucoma. The lesions were classified as group E and, therefore, taking the results obtained by Shields et al. in group E tumors into consideration, underwent enucleation. This case clearly shows that this cancer is curable if detected at a stage in which it is still contained within the retina, subretinal space or vitreous, and that the management of Rb relies on an experienced team of ocular oncologists, pediatricians and pediatric ophthalmologists working together for the single goal of saving the child's life.

<http://www.ncbi.nlm.nih.gov/pubmed/23277210>

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## Fundoplication versus postoperative medication for gastro-oesophageal reflux in children with neurological impairment undergoing gastrostomy

Vernon-Roberts, A. and P. B. Sullivan

*Cochrane Database Syst Rev 2013; 8: CD006151*

**BACKGROUND:** Children with neurological impairments frequently experience feeding difficulties, which can lead to malnutrition and growth failure. Gastrostomy feeding is now the preferred method of providing nutritional support to children with neurological impairments who are unable to feed adequately by mouth. Complications may arise as a result of gastrostomy placement, and the development or worsening of gastro-oesophageal reflux (GOR) has been widely reported. This has led to the frequent use of surgical antireflux treatment in the form of a fundoplication, or other antireflux procedures. Fundoplication is associated with a high recurrence rate, surgical failure, and significant morbidity and mortality. Since proton pump inhibitors (PPIs) were introduced in the 1990s, they have come to play a larger part in the medical management of GOR in children with neurological impairments. Uncontrolled studies suggest that PPIs may be a safe, appropriate treatment for GOR. Other agents currently used include milk thickeners, acid suppression drugs, acid buffering agents, gut motility stimulants and sodium alginate preparations. There are risks and benefits associated with both surgical and medical interventions and further comparison is necessary to determine the optimal treatment choice. **OBJECTIVES:** To compare the effectiveness of antireflux surgery and antireflux medications for children with neurological impairments and GOR who are undergoing placement of a gastrostomy feeding tube. **SEARCH METHODS:** We searched the following databases on 23 March 2012: the Cochrane Central Register of Controlled Trials (CENTRAL), Ovid MEDLINE, EMBASE, CINAHL, LILACS and ISI Web of Science. Previously, we searched the Child Health Library in June 2009. We also performed online searches of trial registries, medical journals, conference proceedings, dissertations and theses. We contacted specialists in the medical and industry setting for knowledge of completed or ongoing trials. **SELECTION CRITERIA:** We sought to include randomised controlled trials that recruited children up to the age of 18 years with neurological impairments and GOR who were undergoing gastrostomy tube insertion. **DATA COLLECTION AND ANALYSIS:** The review authors worked independently to select trials; none were identified. **MAIN RESULTS:** We identified no trials that satisfied the criteria for this review. **AUTHORS' CONCLUSIONS:** There remains considerable uncertainty regarding the optimal treatment when faced with the decision of fundoplication surgery versus antireflux medications for children with GOR and neurological impairment who are undergoing gastrostomy insertion. There is a need for robust scientific evidence in order to provide data on the comparable risks or benefits of the two interventions.

<http://www.ncbi.nlm.nih.gov/pubmed/23986351>

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## After a baby has died

Moore, A.

*Nurs Stand 2013; 27(50): 20-21*

Princess Alexandra Hospital in Harlow has developed an innovative staff training course, 'Death of a Baby', which has improved the care provided to parents who have lost a baby. It is open to all hospital staff, who are taught how to communicate sensitively and how to answer parents' questions about practical issues. The course also equips staff to cope with their own feelings of distress and loss in such a situation, and to be able to support each other.

<http://www.ncbi.nlm.nih.gov/pubmed/23944798>

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## CT head in children

Rao, P., et al.

*Eur J Radiol* 2013; 82(7): 1050-1058

The advances in computerized technology (CT) technique over the last few decades have greatly modified imaging protocols in children. The range of pathologies that can now be demonstrated has broadened with the advent of newer techniques such as CT perfusion and the ability to perform complex reconstructions. Increasing speed of scanning and reduction in scan time have influenced the need for sedation and general anaesthetic as well as impacting on motion artefact. Additionally, concerns about radiation safety and avoidance of unnecessary radiation have further impacted on the inclusion of CT in the imaging armamentarium. Justification and image optimisation are essential. It is important to familiarize oneself with the appearances of normal variants or age related developmental changes. CT does however remain an appropriate investigation in a number of conditions.

<http://www.ncbi.nlm.nih.gov/pubmed/22209524>

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## Update on pediatric cancer predisposition syndromes

Schiffman, J. D., et al.

*Pediatr Blood Cancer* 2013; 60(8): 1247-1252

Hereditary cancer syndromes in children and adolescents are becoming more recognized in the field of pediatric hematology/oncology. A recent workshop held at the American Society of Pediatric Hematology/Oncology (ASPHO) 2012 Annual Meeting included several interactive sessions related to specific familial cancer syndromes, genetic testing and screening, and ethical issues in caring for families with inherited cancer risk. This review highlights the workshop presentations, including a brief background about pediatric cancer predisposition syndromes and the importance of learning about them for the practicing pediatric hematologists/oncologists. This is followed by a brief summary of the newly described cancer predisposition syndromes including Rhabdoid Tumor Predisposition Syndrome, Hereditary Paragangliomas and Pheochromocytoma Syndrome, and Familial Pleuropulmonaryblastoma Tumor Predisposition (DICER1) Syndrome. The next section covers genetic testing and screening for pediatric cancer predisposition syndromes. Ethical issues are also discussed including preimplantation genetic diagnosis or testing (PGD/PGT), suspicious lesions found on tumor screening, and incidental mutations discovered by whole genome sequencing. Finally, the perspective of a family with Li-Fraumeni Syndrome is shared.

<http://www.ncbi.nlm.nih.gov/pubmed/23625733>

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## Pediatric computed tomography imaging guideline

Young, C. and C. M. Owens

*Acta Radiol* 2013; 54(9): 998-1006

The use of computed tomography (CT) in pediatric diagnostic imaging is demanding generally, but when coupled with an awareness to limit the radiation dose associated with this imaging modality, the procedure becomes challenging. Although new techniques have been developed in line with the introduction of faster multidetector computed tomography (MDCT) scanners to aid radiation reduction, it still remains the responsibility of the clinical practitioner to ensure each examination request is justified and the scanning protocol and parameters selected are optimized to the individual patient's requirement. It is the purpose of this article to outline the basic principle of CT radiation dose optimization based on modification of scanning parameters and application of different imaging techniques.

<http://www.ncbi.nlm.nih.gov/pubmed/23436822>

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## What parents want from emails with their pediatrician: Implications for teaching communication skills

Schiller, J. H., et al.

*Patient Educ Couns* 2013; 92(1): 61-66

**OBJECTIVE:** Physician-patient email communication is increasing but trainees receive no education on this communication medium. Research eliciting patient preferences about email communication could inform training. Investigators elicited parents' perspectives on physician-parent email communication and compared parent and faculty assessments of medical students' emails. **METHODS:** This mixed methods study explored physician-parent email communication in 5 parent focus groups using qualitative analyses to identify themes. Differences between faculty and parent assessment scores for students' email responses were calculated using univariate general linear modeling. **RESULTS:** Themes that emerged were: (1) Building the Relationship, (2) Clarity of Communication and (3) Expectations. Parents criticized student's statements as condescending. The sum of assessment scores by parents and faculty were moderately correlated ( $r(44) = .407, P < .01$ ), but parents gave students lower scores on acknowledges validity/ expresses empathy ( $P = .01$ ) and higher scores on provides next steps ( $P < .01$ ) and identifies issues ( $P < .01$ ). **CONCLUSION:** Parents place value on students' abilities to communicate clearly and convey respect and empathy in email. Parent and faculty perspectives on email communication are similar but not the same. **PRACTICE IMPLICATIONS:** Differences between parental and faculty assessments of medical students' emails supports the need for the involvement of patients and families in email communication curriculum development.

<http://www.ncbi.nlm.nih.gov/pubmed/23510794>

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## Treating myoclonic epilepsy in children: State-of-the-art

Striano, P. and V. Belcastro

*Expert Opin Pharmacother* 2013; 14(10): 1355-1361

**INTRODUCTION:** Myoclonic seizures can be observed in various clinical settings and different epileptic conditions, including some forms of both idiopathic and symptomatic epilepsies. Relatively little has been written on treatment of myoclonic seizures. Some old antiepileptic drugs, such as valproate and some benzodiazepines, are widely used but more treatment options exist today for some newer antiepileptic drugs. Nevertheless, patients can be refractory to drug treatment and some drugs may exacerbate or even induce myoclonus. **AREAS COVERED:** Key safety, tolerability, and efficacy data are presented for different antiepileptic drugs with antimyoclonic effect, alone and/or in combination. **EXPERT OPINION:** Treatment of myoclonic seizures in children is mainly based on prospective and retrospective studies, with little evidence from randomized clinical trials. Valproate is commonly the first choice alone or in combination with some benzodiazepines or levetiracetam. There is still insufficient evidence for the use of topiramate and zonisamide as monotherapy. Of major importance remains avoidance of medication that may aggravate the seizures. Better understanding of pathophysiologic mechanisms of myoclonic seizures and myoclonic epilepsies could yield great improvement in the treatment and quality of life of patients.

<http://www.ncbi.nlm.nih.gov/pubmed/23668690>

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## Hospice nurses' perspectives of spirituality

Tiew, L. H., et al.

*J Clin Nurs* 2013; 22(19-20): 2923-2933

**AIMS AND OBJECTIVES:** To explore Singapore hospice nurses' perspectives of spirituality and spiritual care. **DESIGN:** A descriptive, cross-sectional design was used. **BACKGROUND:** Spiritual care is integral to providing quality end-of-life care. However, patients often report that this aspect of care is lacking. Previous studies suggest that nurses' neglect of this aspect of care could be attributed to poor understanding of what spirituality is and what such care entails. This study aimed to explore Singapore hospice nurses' perspectives about spirituality and spiritual care. **METHODS:** A convenience sample of hospice nurses was recruited from the eight hospices in Singapore. The survey comprised two parts: the participant demographic details and the Spirituality Care-Giving Scale. This 35-item validated instrument measures participants' perspectives about spirituality and spiritual care. **RESULTS:** Sixty-six nurses participated (response rate of 65%). Overall, participants agreed with items in the Spiritual Care-Giving Scale related to Attributes of Spiritual Care; Spiritual Perspectives; Spiritual Care Attitudes; and Spiritual Care Values. Results from general linear model analysis showed statistically significant main effects between race, spiritual affiliation and type of hospice setting, with the total Spiritual Care-Giving Scale score and four-factor scores. **CONCLUSIONS:** Spirituality was perceived to be universal, holistic and existential in nature. Spiritual care was perceived to be relational and centred on respecting patients' differing faiths and beliefs. Participants highly regarded the importance of spiritual care in the care of patients at end-of-life. Factors that significantly affected participants' perspectives of spirituality and spiritual care included race, spiritual affiliation and hospice type. **RELEVANCE TO CLINICAL PRACTICE:** Study can clarify values and importance of spirituality and care concepts in end-of-life care. Accordingly, spirituality and care issues can be incorporated in multi-disciplinary team discussions. Explicit guidelines regarding spiritual care and resources can be developed.

<http://www.ncbi.nlm.nih.gov/pubmed/23808790>

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## Imaging in childhood cancer: A Society for Pediatric Radiology and Children's Oncology Group Joint Task Force report

Weiser, D. A., et al.

*Pediatr Blood Cancer* 2013; 60(8): 1253-1260

Contemporary medical imaging is a cornerstone of care for children with cancer. As 5-year survival rates for children with cancer exceed 80%, imaging technologies have evolved in parallel to include a wide array of modalities. Here, we overview the risks and benefits associated with commonly used imaging modalities and survey the current landscape of medical imaging for children with cancer. We find evidence-based imaging guidelines to assist in protocol development and to guide decision-making for optimal patient care are often lacking. The substantial variation in protocol-based recommendations for imaging both during and following therapy may hinder optimal clinical research and clinical care for children with cancer.

<http://www.ncbi.nlm.nih.gov/pubmed/23572212>

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## Is communication guidance mistaken? Qualitative study of parent-oncologist communication in childhood cancer

Young, B., et al.

*Br J Cancer* 2013; 109(4): 836-843

**BACKGROUND:** Guidance encourages oncologists to engage patients and relatives in discussing the emotions that accompany cancer diagnosis and treatment. We investigated the perspectives of parents of children with leukaemia on the role of paediatric oncologists in such discussion. **METHODS:** Qualitative study comprising 33 audio-recorded parent-oncologist consultations and semi-structured interviews with 67 parents during the year following diagnosis. **RESULTS:** Consultations soon after the diagnosis were largely devoid of overt discussion of parental emotion. Interviewed parents did not describe a need for such discussion. They spoke of being comforted by oncologists' clinical focus, by the biomedical information they provided and by their calmness and constancy. When we explicitly asked parents 1 year later about the oncologists' role in emotional support, they overwhelmingly told us that they did not want to discuss their feelings with oncologists. They wanted to preserve the oncologists' focus on their child's clinical care, deprecated anything that diverted from this and spoke of the value of boundaries in the parent-oncologist relationship. **CONCLUSION:** Parents were usually comforted by oncologists, but this was not achieved in the way suggested by communication guidance. Communication guidance would benefit from an enhanced understanding of how emotional support is experienced by those who rely on it.

<http://www.ncbi.nlm.nih.gov/pubmed/23900218>

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## Electrographic seizures and status epilepticus in critically ill children and neonates with encephalopathy

Abend, N. S., et al.

*Lancet Neurol* 2013; 12(12): 1170-1179

Electrographic seizures are seizures that are evident on EEG monitoring. They are common in critically ill children and neonates with acute encephalopathy. Most electrographic seizures have no associated clinical changes, and continuous EEG monitoring is necessary for identification. The effect of electrographic seizures on outcome is the focus of active investigation. Studies have shown that a high burden of electrographic seizures is associated with worsened clinical outcome after adjustment for cause and severity of brain injury, suggesting that a high burden of such seizures might independently contribute to secondary brain injury. Further research is needed to determine whether identification and management of electrographic seizures reduces secondary brain injury and improves outcome in critically ill children and neonates.

<http://www.ncbi.nlm.nih.gov/pubmed/24229615>

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## Rare bleeding disorders in children: Identification and primary care management

Acharya, S. S.

*Pediatrics* 2013; 132(5): 882-892

Bleeding symptoms are common in healthy children but occasionally may indicate an underlying congenital or acquired bleeding diathesis. The rare bleeding disorders (RBDs) comprise inherited deficiencies of coagulation factors I (congenital fibrinogen deficiencies), II, V, VII, X, XI, and XIII and combined factor deficiencies, most notably of factors V and VIII and of vitamin K-dependent factors. These disorders often manifest during childhood and may present with recurrent or even serious or life-threatening bleeding episodes, particularly during the neonatal period. Accordingly, primary care and other nonhematologist pediatric providers should be familiar with the clinical presentation and initial evaluation of these rare disorders. Bleeding manifestations generally vary within the same RBD and may be indistinguishable from 1 RBD to another or from other more common bleeding disorders. Serious bleeding events such as intracranial hemorrhage may be heralded by less serious bleeding symptoms. The results of initial coagulation studies, especially prothrombin time and activated partial thromboplastin time, are often helpful in narrowing down the potential factor deficiency, with factor XIII deficiency being an exception. Consultation with a hematologist is advised to facilitate accurate diagnosis and to ensure proper management and follow-up. The approach to bleeding episodes and invasive procedures is individualized and depends on the severity, frequency, and, in the case of procedures, likelihood of bleeding. Prophylaxis may be appropriate in children with recurrent serious bleeding and specifically after life-threatening bleeding episodes. When available, specific purified plasma-derived or recombinant factor concentrates, rather than fresh frozen plasma or cryoprecipitate, are the treatment of choice.

<http://www.ncbi.nlm.nih.gov/pubmed/24127475>

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## Central nervous system tumors in the first year of life: A clinical and pathologic experience from a single cancer center

Al-Hussaini, M., et al.

*Childs Nerv Syst* 2013; 29(10): 1883-1891

**PURPOSE:** This study aims to review our experience with central nervous system (CNS) tumors occurring during the first year of life and to report differing features found in our series. **METHODS:** This is a retrospective study of infants with CNS tumors diagnosed at our institution from 2006 to 2011. **RESULTS:** A total of 19 cases were identified, with a median age of 232 days and predominance of male gender. Males were younger than females at the time of diagnosis ( $p$  value = 0.039). There were 13 low-grade tumors, glial tumors being the most common (11/13,  $p$  value = 0.003) and six high-grade tumors, atypical teratoid rhabdoid tumor being the most common (4/6). Low-grade tumors predominated in the supratentorial region, while high-grade tumors were seen in the infratentorial area ( $p$  value = 0.035). Males had a predilection to have more supratentorial tumors ( $p$  value = 0.058). Four patients underwent gross total resection, and eight received chemotherapy; none received radiotherapy. Two patients had spinal cord tumors; both were of pilomyxoid astrocytoma histology. Rare tumors included hemangiopericytoma ( $n = 1$ ) and atypical choroid plexus tumor ( $n = 1$ ), both occurring in the supratentorial area and affecting the youngest patients in this group; they were diagnosed prenatally and at 107 days, respectively. The median progression-free and overall survivals were 269 and 667 days, respectively. Among all tested parameters, only the grade of the tumor affected the outcome. **CONCLUSIONS:** Diagnosis and management of infant's CNS tumors remain challenging. Pathologists should be aware of the diversity of histological types. Assigning appropriate tumor grade is fundamental in predicting the outcome.

<http://www.ncbi.nlm.nih.gov/pubmed/23525892>

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## Predictors of survival and incidence of hepatoblastoma in the paediatric population.

Allan, B. J., et al.

*HPB (Oxford)* 2013; 15(10): 741-746

**OBJECTIVES:** This study evaluates current trends in incidence, clinical outcomes and factors predictive of survival in children with hepatoblastoma (HB). **METHODS:** The Surveillance, Epidemiology and End Results (SEER) database was queried for the period 1973-2009 for all patients aged <20 years with HB. **RESULTS:** A total of 606 patients were identified. The age-adjusted incidence was 0.13 patients per 100 000 in 2009. An annual percentage change of 2.18% (95% confidence interval (CI) 1.10-3.27;  $P < 0.05$ ) was seen over the study period. Overall survival rates at 5, 10 and 20 years were 63%, 61% and 59%, respectively. Ten-year survival rates significantly improved in patients with resectable disease who underwent operative treatment in comparison with those with non-resectable HB (86% versus 39%;  $P < 0.0001$ ). Multivariate analysis showed surgical treatment (hazard ratio (HR) = 0.23, 95% CI 0.17-0.31;  $P < 0.0001$ ), Hispanic ethnicity (HR = 0.61, 95% CI 0.43-0.89;  $P = 0.01$ ), local disease at presentation (HR = 0.43, 95% CI 0.29-0.63;  $P < 0.0001$ ) and age < 5 years (HR = 0.63, 95% CI 0.41-0.95;  $P < 0.03$ ) to be independent prognostic factors of survival. **CONCLUSIONS:** The incidence of paediatric HB has increased over time. Hepatoblastoma is almost exclusively seen in children aged < 5 years. When HB presents after the age of 5 years, the prognosis is most unfavourable. Tumour extirpation markedly improves survival in paediatric patients with local disease.

<http://www.ncbi.nlm.nih.gov/pubmed/23600968>

## An analysis of 73 cases of pediatric malignant tumors of the thymus

Allan, B. J., et al.

*J Surg Res* 2013; 184(1): 397-403

**BACKGROUND:** Tumors of the thymus are very rare in the pediatric population. This study examines the current trends and outcomes of children with thymus tumors. **METHODS:** The Surveillance, Epidemiology and End Results (SEER) registry was queried for all patients <20 y of age with primary thymic malignancies from 1973 to 2008. **RESULTS:** A total of 73 pediatric patients were identified with malignant thymic tumors. The median age at diagnosis was 13 y old. Among the 20 patients that presented with distant disease, 70% died. Conversely, among the 23 patients that presented with locoregional disease, 70% survived. Although the overall mean survival time was 89 +/- 116 mo, 45% of patients died over the study period. Patients with Hodgkin lymphomas and germ cell tumors exhibited the highest survival (76% and 60% at 10 y, respectively). Multivariate analysis was used to identify local or regional tumor stage (odds ratio = 4.5, 95% confidence interval = 1.4-14.5) and surgical resection (OR = 3.8, 95% confidence interval = 1.4-10.8) as independent predictors of survival. **CONCLUSIONS:** Malignant thymomas and lymphomas are the most common histological variants of pediatric thymus tumors, and patients with Hodgkin lymphomas exhibit the highest survival. Surgery is more commonly performed on malignant thymomas and is an independent prognostic indicator of survival.

<http://www.ncbi.nlm.nih.gov/pubmed/23570972>

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## A guide to diagnosis and treatment of Leigh syndrome

Baertling, F., et al.

*J Neurol Neurosurg Psychiatry* 2013; 85(3): 257-265

Leigh syndrome is a devastating neurodegenerative disease, typically manifesting in infancy or early childhood. However, also late-onset cases have been reported. Since its first description by Denis Archibald Leigh in 1951, it has evolved from a postmortem diagnosis, strictly defined by histopathological observations, to a clinical entity with indicative laboratory and radiological findings. Hallmarks of the disease are symmetrical lesions in the basal ganglia or brain stem on MRI, and a clinical course with rapid deterioration of cognitive and motor functions. Examinations of fresh muscle tissue or cultured fibroblasts are important tools to establish a biochemical and genetic diagnosis. Numerous causative mutations in mitochondrial and nuclear genes, encoding components of the oxidative phosphorylation system have been described in the past years. Moreover, dysfunctions in pyruvate dehydrogenase complex or coenzyme Q10 metabolism may be associated with Leigh syndrome. To date, there is no cure for affected patients, and treatment options are mostly unsatisfactory. Here, we review the most important clinical aspects of Leigh syndrome, and discuss diagnostic steps as well as treatment options.

<http://www.ncbi.nlm.nih.gov/pubmed/23772060>

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## Neurobehavioral features and natural history of juvenile neuronal ceroid lipofuscinosis (Batten disease)

Adams, H. R. and J. W. Mink

*J Child Neurol* 2013; 28(9): 1128-1136

Juvenile neuronal ceroid lipofuscinosis is a childhood-onset neurodegenerative disease with prominent symptoms comprising a pediatric dementia syndrome: intellectual decline, mood and behavioral impairments, and loss of adaptive skills. We review the history of neurobehavioral features in juvenile neuronal ceroid lipofuscinosis and the work of the University of Rochester Batten Center to characterize the extent and progression of neurobehavioral symptoms over the disease course, and discuss the relevance of neurobehavioral studies as an aid to understanding the clinical phenotype of juvenile Batten disease and potential targets for intervention.

<http://www.ncbi.nlm.nih.gov/pubmed/24014508>

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## Inflammatory myofibroblastic tumors following the treatment of malignancy in childhood: case reports

Adamski, J. K., et al.

*J Pediatr Hematol Oncol* 2014; 36(2): 159-162

Inflammatory myofibroblastic tumors (IMT) are rare, mostly benign soft tissue tumors. Occurring mainly in children the presentation, clinical features, and diagnostic dilemmas raised are well reported. Here we describe 4 patients diagnosed with IMT after the treatment of childhood cancer, and review the literature regarding IMT and malignancy. Discussing them in this context raises clinical questions; Are these tumors incidental or a consequence of treatment? Are they more common than we think and are any miss-diagnosed as tumor recurrence? This paper aims to raise awareness of IMT as diagnostic possibilities after treatment for childhood malignancies.

<http://www.ncbi.nlm.nih.gov/pubmed/23274383>

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## The adolescent and young adult with cancer: State of the art- acute leukemias

Gramatges, M. M. and K. R. Rabin

*Curr Oncol Rep* 2013; 15(4): 317-324

Despite survival gains over the past several decades, adolescent and young adult (AYA) patients with both acute lymphoblastic leukemia (ALL) and acute myeloid leukemia (AML) demonstrate a consistent survival disadvantage. The AYA population exhibits unique disease and host characteristics, and further study is needed to improve their outcomes. This review will highlight distinctive aspects of disease biology in this population, as well as salient treatment-related toxicities including osteonecrosis, pancreatitis, thromboembolism, hyperglycemia, and infections. The impact of obesity and differences in drug metabolism and chemotherapy resistance will also be discussed, as well as optimal treatment considerations for the AYA population.

<http://www.ncbi.nlm.nih.gov/pubmed/23757222>

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## Histologically proven, low-grade brainstem gliomas in children: 30-year experience with long-term follow-up at Mayo Clinic

Ahmed, K. A., et al.

*Am J Clin Oncol* 2014; 37(1): 51-56

**INTRODUCTION:** To evaluate long-term overall survival (OS), progression-free survival (PFS), and outcomes in pathologically proven brainstem low-grade gliomas (BS-LGG) in children. **METHODS:** The Mayo Clinic tumor registry identified 48 consecutive children ( $\leq 20$  y, 52% female) with biopsy-proven BS-LGG treated at Mayo Clinic between January 1971 and December 2004. Medical records were retrospectively reviewed. For analysis, patients were censored at the time of recurrence, death, or last follow-up. **RESULTS:** The median age at diagnosis was 12 years with a median follow-up of 6.0 years. The majority of tumors were grade I (69%) and pathology was consistent with an astrocytoma in the majority of patients (98%). Gross total resection was obtained in 4, subtotal in 17, and 27 patients were biopsied only. Postoperative radiotherapy (RT) was used in 29 patients. Median OS for the entire group was 14.8 years with a 1-, 5-, and 10-year OS of 85%, 67% and 59%, respectively. Median PFS for the entire group was 7.3 years. Improved survival was associated with undergoing resection versus biopsy-only with 5-year OS rates of 85% and 50% ( $P=0.002$ ), respectively. A high proportion of patients (42%) had diffuse tumors and 13 patients (27%) had diffuse pontine gliomas (DPGs). DPGs had an OS of 1.8 years with a worse median PFS than non-DPGs (1.8 vs. 11.1 y;  $P=0.009$ ). RT was used preferentially in patients with poor prognosis such as those who had a biopsy-only procedure (19/27) and DPGs (9/13). **CONCLUSIONS:** OS in this single institution retrospective study in pathologically proven BS-LGG with extensive follow-up displayed favorable long-term outcomes. Improved outcomes were associated with nondiffuse classification.

<http://www.ncbi.nlm.nih.gov/pubmed/23357966>

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## Oral corticosteroids and onset of cardiomyopathy in Duchenne muscular dystrophy

Barber, B. J., et al.

*J Pediatr* 2013; 163(4): 1080-1084 e1081

**OBJECTIVE:** To estimate the age when cardiomyopathy develops in boys with Duchenne muscular dystrophy (DMD) and to analyze the effect of corticosteroid treatment on the age of cardiomyopathy onset. **STUDY DESIGN:** We identified a population-based sample of 462 boys with DMD, born between 1982 and 2005, in 5 surveillance sites in the US. Echocardiographic and corticosteroid treatment data were collected. Cardiomyopathy was defined by a reduced fractional shortening ( $<28\%$ ) or ejection fraction ( $<55\%$ ). The age of cardiomyopathy onset was determined. Survival analysis was performed to determine the effects of corticosteroid treatment on cardiomyopathy onset. **RESULTS:** The mean (SD) age of cardiomyopathy onset was 14.3 (4.2) years for the entire population and 15.2 (3.4) years in corticosteroid-treated vs 13.1 (4.8) in non-treated boys. Survival analysis described a significant delay of cardiomyopathy onset for boys treated with corticosteroids ( $P < .02$ ). By 14.3 years of age, 63% of non-treated boys had developed cardiomyopathy vs only 36% of those treated. Among boys treated with corticosteroids, there is a significant positive effect of duration of corticosteroid treatment on cardiomyopathy onset ( $P < .0001$ ). For every year of corticosteroid treatment, the probability of developing cardiomyopathy decreased by 4%. **CONCLUSIONS:** Oral corticosteroid treatment was associated with delayed cardiomyopathy onset. The duration of corticosteroid treatment also correlated positively with delayed cardiomyopathy onset. Our analysis suggests that a boy with DMD treated for 5 years with corticosteroids might experience a 20% decrease in the likelihood of developing cardiomyopathy compared with untreated boys.

<http://www.ncbi.nlm.nih.gov/pubmed/23866715>

## Diffuse intrinsic pontine glioma treated with prolonged temozolomide and radiotherapy: Results of a United Kingdom phase II trial (CNS 2007 04)

Bailey, S., et al.

*Eur J Cancer* 2013; 49(18): 3856-3862

Diffuse intrinsic pontine glioma (DIPG) has a dismal prognosis with no chemotherapy regimen so far resulting in any significant improvement over standard radiotherapy. In this trial, a prolonged regimen (21/28d) of temozolomide was studied with the aim of overcoming O(6)-methylguanine methyltransferase (MGMT) mediated resistance. Forty-three patients with a defined clinico-radiological diagnosis of DIPG received radiotherapy and concomitant temozolomide (75 mg/m<sup>2</sup>) after which up to 12 courses of 21d of adjuvant temozolomide (75-100mg/m<sup>2</sup>) were given 4 weekly. The trial used a 2-stage design and passed interim analysis. At diagnosis median age was 8 years (2-20 years), 81% had cranial nerve abnormalities, 76% ataxia and 57% long tract signs. Median Karnofsky/Lansky score was 80 (10-100). Patients received a median of three courses of adjuvant temozolomide, five received all 12 courses and seven did not start adjuvant treatment. Three patients were withdrawn from study treatment due to haematological toxicity and 10 had a dose reduction. No other significant toxicity related to temozolomide was noted. Overall survival (OS) (95% confidence interval (CI)) was 56% (40%, 69%) at 9 months, 35% (21%, 49%) at 1 year and 17% (7%, 30%) at 2 years. Median survival was 9.5 months (range 7.5-11.4 months). There were five 2-year survivors with a median age of 13.6 years at diagnosis. This trial demonstrated no survival benefit of the addition of dose dense temozolomide, to standard radiotherapy in children with classical DIPG. However, a subgroup of adolescent DIPG patients did have a prolonged survival, which needs further exploration.

<http://www.ncbi.nlm.nih.gov/pubmed/24011536>

## Adverse effects of antiepileptic drugs in North Indian pediatric outpatients

Bansal, D., et al.

*Clin Neuropharmacol* 2013; 36(4): 107-113

**PURPOSE:** The present study investigates the pattern and predictors of treatment-emergent adverse drug reactions (ADRs) in children diagnosed with epilepsy. **METHODS:** We conducted prospective observational study in a tertiary care teaching hospital on 277 epileptic children. Antiepileptic drug (AED)-associated ADRs, demographic and clinical characteristics, AED regimen, and so on were recorded. Causality, severity, and preventability were performed by World Health Organization-Uppsala Monitoring Center scale, Hartwig's severity scale, and Schumock and Thornton questionnaire, respectively. **RESULTS:** Of the enrolled population, 53% children had symptomatic epilepsy, and 51% were in 5- to 10-year age group. More than two-thirds of children were on monotherapy, with phenytoin (n = 176, 63.5%) being the most common AED. Three hundred fifty-three AED-related ADRs were recorded in 175 children (63.2%). Poor scholastic performance (19%) was the most common ADR, followed by gum hypertrophy (13.3%), headache (10.2%), behavioral problems (5.7%), drowsiness (5.7%), and others. Two hundred sixteen ADRs were probable, and 126 ADRs were possible. Severe ADRs were noted in 6 children. Girls (odds ratio [OR], 1.93; 95% confidence interval [95% CI], 1.07-3.45; P = 0.03), children with secondary epilepsy (OR, 3.31; 95% CI, 1.76-6.23; P <= 0.001), children older than 5 years (5-10 years; OR, 6.28; 95% CI, 2.79-14.12; P <= 0.001), and those older than 10 years (OR, 9.04; 95% CI, 3.69-22.17; P <= 0.001) were found to be at higher risk of experiencing ADRs. **CONCLUSIONS:** Monotherapy was the preferred treatment. Phenytoin was the most common ADR causative agent. Female sex, symptomatic epilepsy, and older age (> 5 years) were found to be associated with higher probability of ADR development.

<http://www.ncbi.nlm.nih.gov/pubmed/23860344>

## Effectiveness of fundoplication at the time of gastrostomy in infants with neurological impairment

Barnhart, D. C., et al.

*JAMA Pediatr* 2013; 167(10): 911-918

**IMPORTANCE:** Gastrostomy tube (GT) placement is the most common gastrointestinal operation performed on neonates. Concomitant fundoplication is used variably to prevent complications of gastroesophageal reflux, but its effectiveness is unproven. **OBJECTIVE:** To compare the effect of fundoplication at the time of GT placement vs GT placement alone on subsequent reflux-related hospitalizations in infants with neurological impairment. **DESIGN, SETTING, AND PARTICIPANTS:** Retrospective, observational cohort study, defined by birth between January 1, 2005, and December 31, 2010, at 42 children's hospitals in the United States, with a 1-year follow-up period among 4163 infants with neurological impairment who underwent GT placement with or without fundoplication during their neonatal intensive care unit stay. **INTERVENTION:** Fundoplication and GT placement vs. GT placement alone. **MAIN OUTCOMES AND MEASURES:** One-year postprocedural reflux-related hospitalization rates, defined as hospitalization for asthma, mechanical ventilation, gastroesophageal reflux disease, and aspiration or other types of pneumonia. Propensity to undergo concomitant fundoplication was modeled using demographics, prior procedures (tracheostomy and mechanical ventilation), and prior diagnoses (eg, pneumonia, gastroesophageal reflux disease, and other comorbidities). **RESULTS:** Overall, 4163 of 42,796 infants (9.7%) with neurological impairment admitted to the neonatal intensive care unit underwent GT placement alone or with fundoplication. Infants who concomitantly underwent fundoplication had more reflux-related hospitalizations during the first year than those who underwent GT placement alone (mean, 1.02; 95% CI, 0.93-1.10 vs mean, 0.92; 95% CI, 0.91-1.00). Of 1404 infants who underwent fundoplication, 1027 (73.1%) were matched based on propensity scores. The mean difference of the matched cohort for any reflux-related hospitalizations was -0.05 (95% CI, -0.20 to 0.15) per year. **CONCLUSIONS AND RELEVANCE:** Infants with neurological impairment who underwent fundoplication at the time of GT placement did not have a reduced rate of reflux-related hospitalizations during the first year compared with those who underwent GT placement alone, despite propensity score matching. This may be due to a lack of effectiveness of fundoplication in preventing these complications or due to differences in the patient groups that were inadequately accounted for in the matching.

<http://www.ncbi.nlm.nih.gov/pubmed/23921627>

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## Why short stature is beneficial in Duchenne muscular dystrophy

Bodor, M. and C. M. McDonald

*Muscle Nerve* 2013; 48(3): 336-342

**INTRODUCTION:** Duchenne muscular dystrophy (DMD) is caused by a genetic defect resulting in absent dystrophin, yet children are able to walk when small and young but lose this ability as they grow. The mdx mouse has absent dystrophin yet does not exhibit significant disability. **METHODS:** Allometric modeling of linearly increasing load per muscle fiber and stress on the sarcolemma with growth and exponential decline associated with loss of muscle fibers correlated with case studies and animal models of DMD. **RESULTS:** Smaller species or breeds are predictably less affected than large as follows: mdx mice < small golden retriever muscular dystrophy (GRMD) dogs < large GRMD dogs < humans. Case reports of combined growth hormone and dystrophin deficiency show a relatively benign course of disease. **CONCLUSIONS:** Future therapeutic trials in DMD might include specific growth inhibitors in combination with standard of care treatments to delay the clinical onset and reduce the severity of disease and disability.

<http://www.ncbi.nlm.nih.gov/pubmed/23893308>



## Cognitive decline in Dravet syndrome: Is there a cerebellar role?

Battaglia, D., et al.

*Epilepsy Res* 2013; 106(1-2): 211-221

**PURPOSE:** The aim of the study was to perform a detailed assessment of cognitive abilities and behaviour in a series of epileptic patients with Dravet syndrome (DS) in order to establish a possible cerebellar-like pattern. **METHODS:** Nine children with DS without major behavioural disturbances and with cognitive abilities compatible with the assessment of specific cognitive skills (IQ>45) were enrolled in the study, in parallel with another group of nine epileptic patients (cryptogenic or symptomatic with minor brain injuries) consecutively admitted into the hospital matched for chronological age and IQ. All cases underwent neurological examination, long term EEG monitoring, neuroimaging and genetic analysis as well as a neuropsychological assessment including specific cognitive skills. **RESULTS:** On neurological examination 8 of the 9 DS patients had cerebellar signs, which were mild in six and more severe in the other two cases. DS patients had a constant discrepancy between verbal and performance items scales (verbal better than visual-spatial) that was not found in the control group. As to specific cognitive competence, the DS patients differ from the control group in the pattern of cognitive defects involving four main areas of cognitive abilities (a) expressive language with relatively spared comprehension, (b) visual-spatial organization, (c) executive function defects, (d) behavioural disorders. **CONCLUSIONS:** The pattern of cognitive difficulties found in DS patients is consistent with what is reported in literature as cerebellar cognitive syndrome and may account for a possible cerebellar origin (at least as co-factor) of the cognitive decline observed in DS patients, as suggested by other clinical and experimental studies.

<http://www.ncbi.nlm.nih.gov/pubmed/23642572>

## Antiepileptic drugs and high prevalence of low bone mineral density in a group of inpatients with chronic epilepsy

Beerhorst, K., et al.

*Acta Neurol Scand* 2013; 128(4): 273-280

**PURPOSE:** Long-term antiepileptic drug use is associated with low bone mineral density (BMD), fractures and abnormalities in bone metabolism. We aimed at determining the prevalence of bone mineral disorders in patients with refractory epilepsy treated with antiepileptic drugs. **METHODS:** A cross-sectional survey was conducted in adult patients (n = 205) from a residential unit of a tertiary epilepsy centre. Screening for bone mineral disorders was performed with dual-energy X-ray absorptiometry (DXA) scan of spine and hip (including bone mineral density and vertebral fracture assessment) and laboratory measurements. Patient information regarding demography, epilepsy characteristics and medication use was recorded. Based on DXA T-scores, prevalence of bone mineral disorders (osteopenia and osteoporosis) was calculated. Correlations between DXA T-scores and epilepsy parameters were explored. **RESULTS:** Of the 205 patients, there were 10 dropouts. 80% (n = 156/195) of the patients had low BMD: 48.2% had osteopenia and 31.8% had osteoporosis. Of those having low BMD, 51.9% (n = 81/195) was between 18 and 50 years. The T-score of the femoral neck correlated significantly with total duration of epilepsy, cumulative drug load and history of fractures. Linear regression analysis showed that of the epilepsy-related parameters, only cumulative drug load significantly predicted low femoral neck T-score (P = 0.001). **CONCLUSION:** In this high-risk population, we obtained a very high prevalence of 80% of low BMD. Both men and women were affected as well as patients <50 years of age. This study illustrates the magnitude of the problem of bone mineral disorders in chronic epilepsy.

<http://www.ncbi.nlm.nih.gov/pubmed/23461582>

## Mortality risks in new-onset childhood epilepsy

Berg, A. T., et al.

*Pediatrics* 2013; 132(1): 124-131

**OBJECTIVES:** Estimate the causes and risk of death, specifically seizure related, in children followed from onset of epilepsy and to contrast the risk of seizure-related death with other common causes of death in the population. **METHODS:** Mortality experiences from 4 pediatric cohorts of newly diagnosed patients were combined. Causes of death were classified as seizure related (including sudden unexpected death [SUDEP]), natural causes, nonnatural causes, and unknown. **RESULTS:** Of 2239 subjects followed up for >30 000 person-years, 79 died. Ten subjects with lethal neurometabolic conditions were ultimately excluded. The overall death rate (per 100 000 person-years) was 228; 743 in complicated epilepsy (with associated neurodisability or underlying brain condition) and 36 in uncomplicated epilepsy. Thirteen deaths were seizure-related (10 SUDEP, 3 other), accounting for 19% of all deaths. Seizure-related death rates were 43 overall, 122 for complicated epilepsy, and 14 for uncomplicated epilepsy. Death rates from other natural causes were 159, 561, and 9, respectively. Of 48 deaths from other natural causes, 37 were due to pneumonia or other respiratory complications. **CONCLUSIONS:** Most excess death in young people with epilepsy is not seizure-related. Mortality is significantly higher compared with the general population in children with complicated epilepsy but not uncomplicated epilepsy. The SUDEP rate was similar to or higher than sudden infant death syndrome rates. In uncomplicated epilepsy, sudden and seizure-related death rates were similar to or higher than rates for other common causes of death in young people (eg, accidents, suicides, homicides). Relating the risk of death in epilepsy to familiar risks may facilitate discussions of seizure-related mortality with patients and families.

<http://www.ncbi.nlm.nih.gov/pubmed/23753097>

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## Acute lymphoblastic leukemia in early childhood as the presenting sign of ataxia-telangiectasia variant

Bielorai, B., et al.

*Pediatr Hematol Oncol* 2013; 30(6): 574-582

Ataxia-telangiectasia (A-T), an autosomal recessive disorder is characterized by progressive neurodegeneration, immunodeficiency, sensitivity to ionizing radiation, and predisposition to cancer, especially to lymphoid malignancies. A-T variant is characterized by a milder clinical phenotype and is caused by missense or leaky splice site mutations that produce residual ataxia telangiectasia mutated (ATM) kinase activity. Lymphoid malignancy can precede the diagnosis of A-T, particularly in young children with mild neurological symptoms. We studied a consanguineous family with four A-T variant patients, three of them developed T-ALL at a young age before the diagnosis of A-T was established. ATM mutation analysis detected two new missense mutations both within exon 12: c.1514T>C and c.1547T>C. All four patients are homozygous for the two mutations, while their parents are heterozygous for the mutations. ATM protein level was low in all patients and the response to the radiomimetic agent, neocarzinostatin, was reduced. Leukemic presentation in a young age in three members of consanguineous family led to the identification of a new missense mutation in the ATM gene. The diagnosis of A-T or A-T variant should be considered in children with neurological abnormalities who develop T-ALL at a young age.

<http://www.ncbi.nlm.nih.gov/pubmed/23509889>

## Bilateral hypertrophic olivary nucleus degeneration on magnetic resonance imaging in children with Leigh and Leigh-like syndrome

Bindu, P. S., et al.

*Br J Radiol* 2014; 87(1034): 20130478

**OBJECTIVE:** Bilateral hypertrophic olivary degeneration on brain MRI has been reported in a few metabolic, genetic and neurodegenerative disorders, including mitochondrial disorders. In this report, we sought to analyse whether bilateral symmetrical inferior olivary nucleus hypertrophy is specifically associated with mitochondrial disorders in children. **METHODS:** This retrospective study included 125 children (mean age, 7.6 +/- 5 years; male: female, 2.6:1) diagnosed with various metabolic and genetic disorders during 2005-2012. The routine MRI sequences (T1 weighted, T2 weighted and fluid-attenuated inversion-recovery sequences) were analysed for the presence of bilateral symmetrical olivary hypertrophy and central tegmental tract or dentate nuclei signal changes. The other imaging findings and the final diagnoses were noted. **RESULTS:** The cohort included patients with Leigh and Leigh-like syndrome (n = 25), other mitochondrial diseases (n = 25), Wilson disease (n = 40), Type 1 glutaric aciduria (n = 14), maple syrup urine disease (n = 13), giant axonal neuropathy (n = 5) and L-2 hydroxy glutaric aciduria (n = 3). Bilateral inferior olivary nucleus hypertrophy was noted in 10 patients, all of whom belonged to the Leigh and Leigh-like syndrome group. **CONCLUSION:** Bilateral hypertrophic olivary degeneration on MRI is relatively often, but not routinely, seen in children with Leigh and Leigh-like syndrome. Early detection of this finding by radiologists and physicians may facilitate targeted metabolic testing in these children. **Advances in knowledge:** This article highlights the occurrence of bilateral hypertrophic olivary nucleus degeneration on MRI in children with Leigh and Leigh-like syndrome, compared with other metabolic disorders.

<http://www.ncbi.nlm.nih.gov/pubmed/24470583>

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## Mortality and morbidity of VLBW infants with trisomy 13 or trisomy 18

Boghossian, N. S., et al.

*Pediatrics* 2014; 133(2): 226-235

**OBJECTIVE:** Little is known about how very low birth weight (VLBW) affects survival and morbidities among infants with trisomy 13 (T13) or trisomy 18 (T18). We examined the care plans for VLBW infants with T13 or T18 and compared their risks of mortality and neonatal morbidities with VLBW infants with trisomy 21 and VLBW infants without birth defects. **METHODS:** Infants with birth weight 401 to 1500 g born or cared for at a participating center of the Eunice Kennedy Shriver National Institute of Child Health and Human Development Neonatal Research Network during the period 1994-2009 were studied. Poisson regression models were used to examine risk of death and neonatal morbidities among infants with T13 or T18. **RESULTS:** Of 52,262 VLBW infants, 38 (0.07%) had T13 and 128 (0.24%) had T18. Intensity of care in the delivery room varied depending on whether the trisomy was diagnosed before or after birth. The plan for subsequent care for the majority of the infants was to withdraw care or to provide comfort care. Eleven percent of infants with T13 and 9% of infants with T18 survived to hospital discharge. Survivors with T13 or T18 had significantly increased risk of patent ductus arteriosus and respiratory distress syndrome compared with infants without birth defects. No infant with T13 or T18 developed necrotizing enterocolitis. **CONCLUSIONS:** In this cohort of liveborn VLBW infants with T13 or T18, the timing of trisomy diagnosis affected the plan for care, survival was poor, and death usually occurred early.

<http://www.ncbi.nlm.nih.gov/pubmed/24446439>

## Rare malignant pediatric tumors registered in the German Childhood Cancer Registry 2001 -2010

Brecht, I. B., et al.

*Pediatr Blood Cancer* 2014; 61(7): 1202-1209

**BACKGROUND:** The German Childhood Cancer Registry (GCCR) annually registers approximately 2,000 children diagnosed with a malignant disease (completeness of registration >95%). While most pediatric cancer patients are diagnosed and treated according to standardized cooperative protocols of the German Society for Pediatric Oncology and Hematology (GPOH), patients with rare tumors are at risk of not being integrated in the network including trials and reference centers. **PROCEDURE:** A retrospective analysis of all rare extracranial solid tumors reported to the GCCR 2001-2010 (age <18 years) was undertaken using a combination of the International Classification of Childhood Cancer (ICCC-3) and the International Classification of Diseases-Oncology (ICD-O-3). Tumors accounting for <0.3% of all malignancies were defined as rare (approx. 6 cases/year and registered malignancy). **RESULTS:** According to this definition 1,189 rare extracranial solid tumors (18.2% of all malignant extracranial solid tumors) were registered, among these 232 patients (19.5% of rare tumor cases), were not included in preexisting GPOH studies/registries. Within 10 years, the number of registered non-GPOH-trial patients with a rare tumor increased. **CONCLUSIONS:** Though most of the GCCR-registered patients with rare malignant tumors are treated within GPOH trials, there is a considerable number of patients that have been diagnosed and treated outside the structures of the GPOH. These patients should be reported to the recently founded German Pediatric Rare Tumor Registry (STEP). Active data accrual and the development of appropriate structures will allow for better registration and improvement of medical care in these patients.

<http://www.ncbi.nlm.nih.gov/pubmed/24585499>

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## Inpatient characteristics of the child admitted with chronic pain

Coffelt, T. A., et al.

*Pediatrics* 2013; 132(2): e422-429

**OBJECTIVE:** To define the demographic, diagnostic, procedural, and episode of care characteristics for children admitted with chronic pain. **METHODS:** We used the Pediatric Health Information System database to obtain data on demographic characteristics, length of stay, readmission rates, diagnoses, and procedures for children admitted with chronic pain. Patients with sickle cell disease, cancer, burns, cerebral palsy, transplants, and ventilator-dependent children were excluded. **RESULTS:** A total of 3752 patients with chronic pain were identified from 2004 through 2010. Admissions increased by 831% over this time period. The mean age of these patients was 13.5 years, the most common race was white (79%), and female subjects outnumbered male subjects by 2.41 to 1. The most common admission and principal discharge diagnosis was abdominal pain; comorbid diagnoses were common, with a mean of 10 diagnoses per patient. In total, 65% of patients had a comorbid gastrointestinal diagnosis and 44% had a psychiatric diagnosis. The mean length of stay was 7.32 days, with an expected length of stay of 4.24 days; 12.5% were readmitted at least once within 1 year. They underwent a mean of 3.18 procedures per patient. **CONCLUSIONS:** The average child admitted with chronic pain is a teenaged female with a wide variety of comorbid conditions, many of which are gastrointestinal and psychiatric in nature. Admissions for chronic pain are rising and account for substantial resource utilization. Future studies should further characterize this population, with the overall objective of improving outcomes and optimizing cost-effective care.

<http://www.ncbi.nlm.nih.gov/pubmed/23821701>

## Congenital myotonic dystrophy: Canadian population-based surveillance study

Campbell, C., et al.

*J Pediatr* 2013; 163(1): 120-125 e121-123

**OBJECTIVES:** To determine the incidence and neonatal morbidity and mortality of congenital myotonic dystrophy (CDM) in Canada. **STUDY DESIGN:** The study has 2 phases. A 5-year prospective monthly surveillance of incident cases of CDM conducted via the Canadian Pediatric Surveillance Program, from March 1, 2005-February 28, 2010, and a 5-year cohort study of eligible incident cases, which is ongoing and not the subject of this report. **RESULTS:** A total of 121 cases were reported, with 38 confirmed as CDM. The incidence of CDM in Canada is 2.1/100,000 (1/47,619) live births. The cases were reported from 8 provinces and 1 territory. The highest reported incidence was Ontario with 15, followed by British Columbia with 7, and Quebec with 6. External validation of cases was performed. The trinucleotide repeat level varied from 550-3100. Twenty-two (58%) of the children were the index cases for their families. Seventeen children are currently enrolled in the ongoing cohort study. **CONCLUSION:** Surveillance and prospective examination of CDM at a population level is important, as the impact of this rare disease is systemic, chronic, and associated with significant morbidity and mortality throughout childhood.

<http://www.ncbi.nlm.nih.gov/pubmed/23415617>

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## Pediatric demyelinating diseases

Chitnis, T.

*Continuum (Minneapolis)* 2013; 19(4 Multiple Sclerosis): 1023-1045

**PURPOSE OF REVIEW:** In the past decade, the number of studies related to demyelinating diseases in children has exponentially increased. Demyelinating disease in children may be monophasic or chronic. Typical monophasic disorders in children are acute disseminated encephalomyelitis and clinically isolated syndromes, including optic neuritis and transverse myelitis. However, some cases of acute disseminated encephalomyelitis or clinically isolated syndrome progress to become chronic disorders, including multiple sclerosis and neuromyelitis optica. This review summarizes the current knowledge on monophasic and chronic demyelinating disorders in children, focusing on an approach to diagnosis and management. **RECENT FINDINGS:** Improved diagnostic definitions for pediatric demyelinating diseases have led to enhanced recognition of these disorders. Additionally, increased awareness and focused national and international efforts continue to inform about the clinical course, response to treatment, and disease pathogenesis. **SUMMARY:** Significant advances have been made in the recognition, diagnosis, and management of pediatric demyelinating disorders over the past 10 years. This review summarizes these advances and provides an updated approach to the diagnosis and management of pediatric demyelinating disorders.

<http://www.ncbi.nlm.nih.gov/pubmed/23917099>

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## Highlights of the third International Conference on Immunotherapy in Pediatric Oncology.

Brehm, C., et al.

*Pediatr Hematol Oncol* 2013; 30(5): 349-366

The third International Conference on Immunotherapy in Pediatric Oncology was held in Frankfurt/Main, Germany; October 1-2, 2012. Major topics of the conference included (i) cellular therapies using antigen-specific and gene-modified T cells for targeting leukemia and pediatric solid tumors; (ii) overcoming hurdles and barriers with regard to immunogenicity, immune escape, and the role of tumor microenvironment; (iii) vaccine strategies and antigen presentation; (iv) haploidentical transplantation and innate immunity; (v) the role of immune cells in allogeneic transplantation; and (vi) current antibody/immunoconjugate approaches for the treatment of pediatric malignancies. During the past decade, major advances have been made in improving the efficacy of these modalities and regulatory hurdles have been taken. Nevertheless, there is still a long way to go to fully exploit the potential of immunotherapeutic strategies to improve the cure of children and adolescents with malignancies. This and future meetings will support new collaborations and insights for further translational and clinical immunotherapy studies.

<http://www.ncbi.nlm.nih.gov/pubmed/23758210>

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## Muscular dystrophies share pathogenetic mechanisms with muscle sarcomas

Fanzani, A., et al.

*Trends Mol Med* 2013; 19(9): 546-554

Several lines of recent evidence have opened a new debate on the mechanisms underlying the genesis of rhabdomyosarcoma, a pediatric soft tissue tumor with a widespread expression of muscle-specific markers. In particular, it is increasingly evident that the loss of skeletal muscle integrity observed in some mouse models of muscular dystrophy can favor rhabdomyosarcoma formation. This is especially true in old age. Here, we review these experimental findings and focus on the main molecular and cellular events that can dictate the tumorigenic process in dystrophic muscle, such as the loss of structural or regulatory proteins with tumor suppressor activity, the impaired DNA damage response due to oxidative stress, the chronic inflammation and the conflicting signals arising within the degenerated muscle niche.

<http://www.ncbi.nlm.nih.gov/pubmed/23890422>

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## The adolescent and young adult with cancer: State of the art-epithelial cancer

Ferreira, C. G., et al.

*Curr Oncol Rep* 2013; 15(4): 287-295

The adolescent and young adult (AYA) is defined as a patient of 15 to 39 years of age at initial cancer diagnosis, and this group has particular medical needs and age-related issues. Excluding violent deaths, cancer is the leading cause of death among the AYA population. Lymphomas, melanoma, testicular cancer, female genital tract malignancies, thyroid cancer, bone and soft tissue sarcomas, leukemias, central nervous system tumors, breast cancer, and nongonadal germ cell tumors account for 95 % of the cancers in this group. Among those, the epithelial cancer of AYA comprehends the minimum amount and its incidence rates tend to increase with age. This review presents information about epidemiology, biologic peculiarities, as well as standard treatment strategies for epithelial cancers in AYA.

<http://www.ncbi.nlm.nih.gov/pubmed/23754487>

## Chronic residual lesions in metastatic medulloblastoma patients

Fried, I., et al.

*J Pediatr Hematol Oncol* 2014; 36(1): 71-75

In a retrospective review of 24 metastatic medulloblastoma patients whose treatment included craniospinal irradiation, 5 patients presented with gross residual abnormalities at completion of therapy. This report describes 2 medulloblastoma patients with persistent residual abnormalities on serial follow-up imaging studies. The patients aged 2 and 2.5 years old at the time of diagnosis underwent surgery followed by multiagent chemotherapy. One patient progressed on therapy and underwent salvage craniospinal radiation. The second showed residual tumor on end of treatment imaging and received low-dose craniospinal irradiation. Despite persistent magnetic resonance imaging findings, the patients are alive and well 13 and 7 years after diagnosis with no further treatment applied. The nature of these residual abnormalities is discussed.

<http://www.ncbi.nlm.nih.gov/pubmed/23528902>

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## Brain stem tumors in children and adolescents: Single institutional experience

Garzon, M., et al.

*Childs Nerv Syst* 2013; 29(8): 1321-1331

**PURPOSE:** Pediatric brain stem tumors (BsT) are a heterogeneous group of diseases. Our aim was to analyze our experience to find out prognostic factors. **METHOD:** A retrospective study with BsT patients was performed. Imaging characteristics, extension of surgery, pathology, and adjuvant therapy were analyzed and correlated with overall survival (OS) and progression-free survival (PFS) as outcome measures. **RESULT:** Since 1980 to 2010, we analyzed 65 BsT patients, 41 of them girls (63%), median age of 8 years (range 13.9 months to 17.6 years). Twenty-two patients (33.8%) had diffuse intrinsic pontine gliomas (DIPG) and 43 (66.2%) presented with focal BsT. Histology was available in 42 patients; the most frequent is low-grade glioma in 24/42 patients (57%). DIPG's histology (obtained usually at necropsy) confirmed five high-grade gliomas. After median follow-up of 49.3 months (0.5-175 months), 20/22 DIPG patients have died (90.9%), while 27/43 with focal tumors were alive (62.8%). Variables related to outcome were histology (better for low-grade glioma (LGG) OS  $p < 0.001$ ), surgery (better if operated OS  $p < 0.001$ ), and adjuvant therapy (worse if given, PFS  $p = 0.001$ , OS  $p = 0.024$ ). The outcome for DIPG was dismal, median OS/EFS of 14.2/9.4 months, significantly worse than focal BsT ( $p = 0.000$ ), while OS/EFS was 122.8/87.2 months for focal intrinsic, 88.2/47.1 months for exophytic, and 124.4/54 months for cervico-medullary tumors: no differences were found among them, except the histology (OS  $p < 0.001$  for low-grade vs high-grade tumors). **CONCLUSION:** BsT in children comprised two different groups: diffuse (DIPG) and focal gliomas. The DIPGs continue having a dismal prognosis, needing new approaches, while focal tumors including LGG have better prognosis.

<http://www.ncbi.nlm.nih.gov/pubmed/23666431>

## Ambulatory capacity and disease progression as measured by the 6-minute-walk-distance in Duchenne muscular dystrophy subjects on daily corticosteroids

Goemans, N., et al.

*Neuromuscul Disord* 2013; 23(8): 618-623

In order to understand contemporary natural history of Duchenne muscular dystrophy (DMD), we report 6-minute walk distance (6MWD) and its change over time from a large single centre population of corticosteroid treated DMD boys. Sixty-five boys on daily corticosteroid treatment were identified with a mean (SD) age of 9.5 (2.3) years at first observation. 6MWD was described for 1 year age groupings. In addition, changes in 6MWD at 1, 1.5 and 2 years (+/- 12 weeks) of follow-up were evaluated. The same evaluations were applied to 6MWD data converted to percent predicted values based on the Geiger equation. 6MWD showed an increase from age group 4.5-5.5 years to age group 6.5-7.5 years, followed by a decline, which became precipitous from 12.5 years onwards. From 15.5 years, all boys were unable to perform the 6-min test. Changes in 6MWD demonstrated a mean (median, SD) decline of -43 (-14, 90) m at 1 year (N=25, mean baseline age 9.5 years), -64 (-56, 99) m at 1.5 years (N=18, mean baseline age 9.6 years), -125 (-106, 139) m at 2 years (N=14, mean baseline age 10.0 years). Conversion to percent predicted values showed the same pattern of evolution. This study provides data on the ambulatory capacity and its changes over time in a homogenous cohort of 65 DMD boys on daily corticosteroids. The variability, the age-related aspects and the slope of decline of the 6MWD should be considered in the design and interpretation of therapeutic trials in ambulant DMD patients.

<http://www.ncbi.nlm.nih.gov/pubmed/23770101>

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## Survival among adolescents and young adults with cancer in Germany and the United States: An international comparison

Gondos, A., et al.

*Int J Cancer* 2013; 133(9): 2207-2215

Serious concern arose in the scientific literature about the state of and progress in cancer survival among adolescent and young adult (AYA) patients in the recent years. We provide an up-to-date international comparison of survival among AYA patients. Using population-based cancer data from 11 German cancer registries and the SEER Program of the United States (covering populations of 39 and 33 million people, respectively), standardized tumor group classifications, period analysis and modeling, we compared the 5-year relative survival of AYA patients in the age groups 15-29 and 30-39 to survival seen among adults aged 40-49 for the 2002-2006 period. Additionally, we also provide an age-specific survival comparison between the two countries. In 2002-2006, for the overwhelming majority of the more than 30 types of cancer examined, AYA patients aged both 15-29 and 30-39 years had higher or similar survival than patients in the age group 40-49 in both countries. A numerically large and statistically significant survival deficit among AYA patients was only found for breast carcinomas in both populations, and colorectal and stomach carcinoma in the United States for the age group 15-29. Overall, results of the country-specific comparisons did not indicate systematic differences. With very few exceptions, no survival deficit between AYA patients and adults aged 40-49 years was found in either of the examined countries in the first decade of the 21st century.

<http://www.ncbi.nlm.nih.gov/pubmed/23616284>



## Child deaths due to injury in the four UK countries: A time trends study from 1980 to 2010

Hardelid, P., et al.

*PLoS One* 2013; 8(7): e68323

**BACKGROUND:** Injuries are an increasingly important cause of death in children worldwide, yet injury mortality is highly preventable. Determining patterns and trends in child injury mortality can identify groups at particularly high risk. We compare trends in child deaths due to injury in four UK countries, between 1980 and 2010. **METHODS:** We obtained information from death certificates on all deaths occurring between 1980 and 2010 in children aged 28 days to 18 years and resident in England, Scotland, Wales or Northern Ireland. Injury deaths were defined by an external cause code recorded as the underlying cause of death. Injury mortality rates were analysed by type of injury, country of residence, age group, sex, and time period. **RESULTS:** Child mortality due to injury has declined in all countries of the UK. England consistently experienced the lowest mortality rate throughout the study period. For children aged 10 to 18 years, differences between countries in mortality rates increased during the study period. Inter-country differences were largest for boys aged 10 to 18 years with mortality rate ratios of 1.38 (95% confidence interval 1.16, 1.64) for Wales, 1.68 (1.48, 1.91) for Scotland and 1.81 (1.50, 2.18) for Northern Ireland compared with England (the baseline) in 2006-10. The decline in mortality due to injury was accounted for by a decline in unintentional injuries. For older children, no declines were observed for deaths caused by self-harm, by assault or from undetermined intent in any UK country. **CONCLUSION:** Whilst child deaths from injury have declined in all four UK countries, substantial differences in mortality rates remain between countries, particularly for older boys. This group stands to gain most from policy interventions to reduce deaths from injury in children.

<http://www.ncbi.nlm.nih.gov/pubmed/23874585>

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## Psychiatric symptoms causing delay in diagnosing childhood cancer: Two case reports and literature review

Hensgens, T. B., et al.

*Eur Child Adolesc Psychiatry* 2013; 222(7): 443-450

**INTRODUCTION:** A somatic disorder may initially be overlooked when a child presents with psychiatric symptoms. We report two children with anorexia nervosa as initial diagnosis and in whom there was a delay in the final diagnosis of the underlying malignancy. A literature survey was performed including patients under 18 years of age with psychiatric symptoms in whom later on an oncological diagnosis became evident as an explanation. **RESULTS:** We have found 30 additional cases, with a median delay of 12 months until the diagnosis of the tumour. Overall, 16 boys and 16 girls had a solid tumour: 26 central nervous system tumours, 3 tumours of the gastrointestinal tract and 3 others. In 25 out of 32 patients anorexia nervosa was assumed, although it always appeared to be atypical. Patients younger than 7 years had a significantly longer delay until final diagnosis, while no other patient characteristics correlated with such delay. **DISCUSSION:** In addition to careful physical (including full neurological) examination, we advise additional neuroimaging especially in each case of atypical presentation of anorexia nervosa, in order to avoid a delay in diagnosis of a possible malignancy. Furthermore, it is desirable to perform a re-examination when a psychiatric disorder does not respond to therapy, in order not to overlook an underlying oncological disease.

<http://www.ncbi.nlm.nih.gov/pubmed/23296472>

## Epilepsy and risk of death and sudden unexpected death in the young: A nationwide study

Holst, A. G., et al.

*Epilepsia* 2013; 54(9): 1613-1620

**PURPOSE:** Patients with epilepsy are at increased risk of premature death from all causes and likely also from sudden unexplained death (SUD). Many patients with epilepsy have significant comorbidity, and it is unclear how much of the increased risk can be explained by epilepsy itself. We aimed to chart the incidence of sudden unexpected death in epilepsy (SUDEP) and estimate the risk of death from all causes and SUD conferred by epilepsy independently. **METHODS:** We conducted a historical cohort study using data from Danish registries and a complete manual review of all death certificates. The population studied consisted of all Danish residents in the age group 1-35 years, in the period 2000-2006 (inclusive), and the main outcome measures were risk of death and SUD. **KEY FINDINGS:** We identified 33,022 subjects with epilepsy (median follow-up 3.7 years) and 3,001,952 subjects without (median follow-up 7.0 years). Among 685 deaths in the population with epilepsy, we identified 50 cases of definite and probable SUDEP corresponding to an incidence rate of 41.1 (95% confidence interval [CI] 31.6-54.9) per 100,000 person-years. Incidence rates increased with age from 17.6 (95% CI 9.5-32.8) in the age group 1-18 years to 73.8 (95% CI 52.5-103.8) for the age group 24-35 years. Having epilepsy increased the crude risk of death with a hazard ratio (HR) of 11.9 (95% CI 11.0-12.9). When adjusting for sex and comorbidities often encountered in patients with epilepsy (neurologic disease including cerebral palsy, psychiatric disease including mental retardation, and congenital disorders), as well as the Charlson comorbidity score, the HR fell to 5.4 (95% CI 4.9-6.0). The crude HR for SUD was 27.5 (95% CI 18.1-41.8) and fell to 16.3 (95% CI 9.8-26.9) when adjusted for the same covariates as above. **SIGNIFICANCE:** Epilepsy in and of itself carries a significant risk of premature death and SUD. These findings highlight the potential gains of risk factor modification for the prevention of premature death and SUDEP in patients with epilepsy.

<http://www.ncbi.nlm.nih.gov/pubmed/23895621>

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## The relationship between spasticity and gross motor capability in nonambulatory children with spastic cerebral palsy

Katusic, A. and S. Alimovic

*Int J Rehabil Res* 2013; 36(3): 205-210

Spasticity has been considered as a major impairment in cerebral palsy (CP), but the relationship between this impairment and motor functions is still unclear, especially in the same group of patients with CP. The aim of this investigation is to determine the relationship between spasticity and gross motor capability in nonambulatory children with spastic CP. Seventy-one children (30 boys, 41 girls) with bilateral spastic cerebral palsy and with Gross Motor Function Classification System (GMFCS) levels IV (n=34) and V (n=37) were included in the study. The spasticity level in lower limbs was evaluated using the Modified Ashworth Scale and the gross motor function with the Gross Motor Function Measure (GMFM-88). Spearman's correlation analysis was used to determine the nature and the strength of the relationship. The results showed a moderate correlation between spasticity and gross motor skills ( $\rho=0.52$  for the GMFCS level;  $\rho=0.57$  for the GMFM-88), accounting for less than 30% of the explained variance. It seems that spasticity is just one factor among many others that could interfere with gross motor skills, even in children with severe forms of spastic CP. Knowledge of the impact of spasticity on motor skills may be useful in the setting of adequate rehabilitation strategies for nonambulatory children with spastic CP.

<http://www.ncbi.nlm.nih.gov/pubmed/23895621>

## Characteristics of dysphagia in children with cerebral palsy, related to gross motor function

Kim, J. S., et al.

*Am J Phys Med Rehabil* 2013; 92(10): 912-919

**OBJECTIVE:** The aim of this study was to report the characteristics of dysphagia in children with cerebral palsy (CP), related to gross motor function. **DESIGN:** Videofluoroscopic swallow study was performed in 29 children with CP, according to the manual of Logemann. Five questions about oromotor dysfunction were answered. Gross motor function level was classified by the Gross Motor Function Classification System Expanded and Revised. **RESULTS:** The results of the videofluoroscopic swallowing studies showed that reduced lip closure, inadequate bolus formation, residue in the oral cavity, delayed triggering of pharyngeal swallow, reduced larynx elevation, coating on the pharyngeal wall, delayed pharyngeal transit time, multiple swallow, and aspiration were significantly more common in the severe group (Gross Motor Function Classification System Expanded and Revised IV or V). As for aspiration, 50% of the children with severe CP had problems, but only 14.3% of them with moderate (Gross Motor Function Classification System Expanded and Revised III) CP and none of them with mild CP had abnormalities. In addition, five of the seven aspiration cases occurred silently. **CONCLUSIONS:** This study shows that dysphagia is closely related to gross motor function in children with CP. Silent aspiration was observed in the moderate to severe CP groups. Aspiration is an important cause of medical problems such as acute and chronic lung disease, and associated respiratory complications contribute significantly in increasing morbidity and mortality in these patient groups. Therefore, the authors suggest that early dysphagia evaluation including videofluoroscopic swallow study is necessary in managing feeding problems and may prevent chronic aspiration, malnutrition, and infections.

<http://www.ncbi.nlm.nih.gov/pubmed/23739271>

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## Witnessed sleep-related seizure and sudden unexpected death in infancy: A case report

Kinney, H. C., et al.

*Forensic Sci Med Pathol* 2013; 9(3): 418-421

Witnessed reports of sudden death are rare, but critical to deciphering its mechanism(s). We report such a death in a seemingly healthy 8-month-old boy in whom seizures and respiratory distress in the prone position were witnessed upon discovery during a sleep period. Following cardiopulmonary resuscitation, anoxic encephalopathy resulted in brain death and withdrawal of life support after 2 days. The autopsy did not reveal a primary anatomic cause of death. Metabolic evaluation failed to uncover an inborn error of ammonia, amino, organic, or fatty acid metabolism. Seizures in this case may have been secondary to cerebral hypoxia-ischemia complicating cardiorespiratory arrest of unknown etiology. Yet, they may represent the first manifestation of idiopathic epilepsy, triggering cardiopulmonary arrest, analogous to the terminal events postulated in sudden and unexplained death in epilepsy. This report alerts the forensic community to the possibility that sudden and unexplained death in infants may be due to seizures.

<http://www.ncbi.nlm.nih.gov/pubmed/23852931>

## Neuroblastoma in older children, adolescents and young adults: A report from the International Neuroblastoma Risk Group project

Mosse, Y. P., et al.

*Pediatr Blood Cancer* 2014; 61(4): 627-635

**BACKGROUND:** Neuroblastoma in older children and adolescents has a distinctive, indolent phenotype, but little is known about the clinical and biological characteristics that distinguish this rare subgroup. Our goal was to determine if an optimal age cut-off exists that defines indolent disease and if accepted prognostic factors and treatment approaches are applicable to older children. **PROCEDURE:** Using data from the International Neuroblastoma Risk Group, among patients  $\geq 18$  months old ( $n = 4,027$ ), monthly age cut-offs were tested to determine the effect of age on survival. The prognostic effect of baseline characteristics and autologous hematopoietic cell transplant (AHCT) for advanced disease was assessed within two age cohorts;  $\geq 5$  to  $< 10$  years ( $n = 730$ ) and  $\geq 10$  years ( $n = 200$ ). **RESULTS:** Older age was prognostic of poor survival, with outcome gradually worsening with increasing age at diagnosis, without statistical evidence for an optimal age cut-off beyond 18 months. Among patients  $\geq 5$  years, factors significantly prognostic of lower event-free survival (EFS) and overall survival (OS) in multivariable analyses were INSS stage 4, MYCN amplification and unfavorable INPC histology classification. Among stage 4 patients, AHCT provided a significant EFS and OS benefit. Following relapse, patients in both older cohorts had prolonged OS compared to those  $\geq 18$  months to  $< 5$  years ( $P < 0.0001$ ). **CONCLUSIONS:** Despite indolent disease and infrequent MYCN amplification, older children with advanced disease have poor survival, without evidence for a specific age cut-off. Our data suggest that AHCT may provide a survival benefit in older children with advanced disease. Novel therapeutic approaches are required to more effectively treat these patients.

<http://www.ncbi.nlm.nih.gov/pubmed/24038992>

## Prominent fatigue in spinal muscular atrophy and spinal and bulbar muscular atrophy: Evidence of activity-dependent conduction block.

Noto, Y., et al.

*Clin Neurophysiol* 2013; 124(9): 1893-1898

**OBJECTIVES:** To clarify whether patients with spinal muscular atrophy (SMA) or spinal and bulbar muscular atrophy (SBMA) suffer disabling muscle fatigue, and whether activity-dependent conduction block (ADCB) contributes to their fatigue. ADCB is usually caused by reduced safety factor for impulse transmission in demyelinating diseases, whereas markedly increased axonal branching associated with collateral sprouting may reduce the safety factor in chronic lower motor neuron disorders. **METHODS:** We assessed the fatigue severity scale (FSS) in 22 patients with SMA/SBMA, and in 100 disease controls (multiple sclerosis, myasthenia gravis, chronic inflammatory demyelinating polyneuropathy (CIDP), and axonal neuropathy). We then performed stimulated-single fibre electromyography (s-SFEMG) in the extensor digitorum communis (EDC) muscle of 21 SMA/SBMA patients, 6 CIDP patients, and 10 normal subjects. **RESULTS:** The FSS score was the highest in SMA/SBMA patients [ $4.9 \pm 1.1$  (mean  $\pm$  SD)], with 81% of them complaining of disabling fatigue, compared with normal controls ( $3.5 \pm 1.0$ ), whereas patients with multiple sclerosis ( $4.3 \pm 1.6$ ), myasthenia gravis ( $4.0 \pm 1.6$ ) or CIDP ( $4.3 \pm 1.4$ ) also showed higher FSS score. When 2000 stimuli were delivered at 20 Hz in s-SFEMG, conduction block of single motor axons developed in 46% of patients with SMA/SBMA, and 40% of CIDP patients, but in none of the normal controls. **CONCLUSION:** SMA/SBMA patients frequently suffer from disabling fatigue presumably caused by ADCB induced by voluntary activity. **SIGNIFICANCE:** ADCB could be the mechanism for muscle fatigue in chronic lower motor neuron diseases.

<http://www.ncbi.nlm.nih.gov/pubmed/23643309w>

## Etiology and prognostic factors of acute liver failure in children

Kaur, S., et al.

*Indian Pediatr* 2013; 50 (7): 677-679

Acute liver failure (ALF) is a life-threatening condition characterized by jaundice, encephalopathy and coagulopathy leading to multiorgan failure in a patient with no prior history of liver disease. Forty three consecutive patients of ALF admitted in Pediatric ICU were studied for etiology and prognostic factors. Etiology was established in 91% cases. Viral infections were the most common cause. Mortality rate was 44%. Increasing grade of encephalopathy, >7 days interval between the onset of prodromal symptoms and encephalopathy, blood glucose <45mg/dL, serum bilirubin > 10mg/dL and pH <7.35 or >7.45 on admission were found to be associated with increased risk of mortality.

<http://www.ncbi.nlm.nih.gov/pubmed/23255683>

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## Diagnosis of mitochondrial neurogastrointestinal encephalopathy disease in gastrointestinal biopsies

Perez-Atayde, A. R.

*Hum Pathol* 2013; 44(7): 1440-1446

A 14-year-old boy with mitochondrial neurogastrointestinal encephalopathy (MNGIE) disease had a lifelong history of failure to thrive and gastrointestinal symptoms including vomiting, pain, and diarrhea, leading to progressive cachexia. At the age of 9 years, after an extensive workup, the diagnosis of Crohn disease was strongly suspected, and he underwent colonoscopy with multiple biopsies. At 11 years of age, vision change and poor balance lead to a diagnosis of leukodystrophy by magnetic resonance imaging. Investigations for metachromatic leukodystrophy, adrenal leukodystrophy, and globoid cell leukodystrophy were all negative. A diagnosis of MNGIE disease was suspected when he continued deteriorating with gastrointestinal symptoms, multiple neurologic deficits, and encephalopathy. Markedly diminished thymidine phosphorylase activity and increased thymidine plasma levels confirmed the diagnosis of MNGIE. At autopsy, megamitochondria were observed by light microscopy in submucosal and myenteric ganglion cells and in smooth muscle cells of muscularis mucosae and muscularis propria, along the entire gastrointestinal tract from the esophagus to the rectum. Megamitochondria in ganglion cells were also observed in a retrospective review of the endoscopic intestinal biopsies taken at age 9 and 13 years and in the appendectomy specimen obtained 1 month before his demise. This study corroborates the presence of megamitochondria in gastrointestinal ganglion cells in MNGIE disease, better illustrates their detailed morphology, and describes for the first time similar structures in the cytoplasm of gastrointestinal smooth muscle cells. Pathologists should be able to recognize these structures by light microscopy and be aware of their association with primary mitochondrialopathies.

<http://www.ncbi.nlm.nih.gov/pubmed/23453626>

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## Neurodegenerative disorders and metabolic disease

Pierre, G.

*Arch Dis Child* 2013; 98(8): 618-624

Most genetic causes of neurodegenerative disorders in childhood are due to neurometabolic disease. There are over 200 disorders, including aminoacidopathies, creatine disorders, mitochondrial cytopathies, peroxisomal disorders and lysosomal storage disorders. However, diagnosis can pose a challenge to the clinician when patients present with non-specific problems like epilepsy, developmental delay, autism, dystonia and ataxia. The variety of specialist tests involved can also be daunting. This review aims to give a practical approach to the investigation and diagnosis of neurometabolic disease from the neonatal period to late childhood while prioritising disorders where there are therapeutic options. In particular, patients who have a complex clinical picture of several neurological and non-neurological features should be investigated.

<http://www.ncbi.nlm.nih.gov/pubmed/23698595>

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## Successful heart transplantation from a donor with Ullrich congenital muscular dystrophy

Plonka, C., et al.

*Am J Transplant* 2013; 13(7): 1915-1917

Heart transplantation is the most effective therapy for children with end-stage heart disease; however, its use is limited by the number of donor organs available. This shortage may be further compounded by concerns about organ quality, leading to refusal of potential donor organ offers. We report on the successful transplantation and 5-year follow-up of a heart from a donor with Ullrich congenital muscular dystrophy (UCMD). The candidate was critically ill at the time of the transplant and the donor organ was declined repeatedly on the match run list due to concerns about organ quality, despite having normal cardiac function by echocardiography on minimal inotropic support. We believe the diagnosis of muscular dystrophy in the donor combined with a lack of understanding about the specifics of the diagnosis of UCMD enabled our candidate to receive a primary offer for this organ. We are unaware of any previous reports of the use of a heart from a donor with UCMD for orthotopic heart transplantation in adults or children.

<http://www.ncbi.nlm.nih.gov/pubmed/23668812>

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## Musicogenic seizures in Dravet syndrome

Sanchez-Carpintero, R., et al.

*Dev Med Child Neurol* 2013; 55(7): 668-670

Dravet syndrome is an epileptic encephalopathy characterized by multiple types of seizures. We report the first case of musicogenic reflex seizures in a 7-year-old male with a mutation in the SCN1A gene causing Dravet syndrome. Reflex seizures have been reported in patients with Dravet syndrome provoked by body temperature elevation, looking at visual patterns, or under intermittent photic stimulation. The case we report widens the spectrum of reflex seizures recorded in patients with Dravet syndrome. Cortical hyperexcitability of genetic origin could explain the tendency of these patients to experience reflex seizures.

<http://www.ncbi.nlm.nih.gov/pubmed/23517304>

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## Muscular dystrophy with large mitochondria associated with mutations in the CHKB gene in three British patients: Extending the clinical and pathological phenotype

Quinlivan, R., et al.

*Neuromuscul Disord* 2013; 23(7): 549-556

Three patients with CHKB deficient muscular dystrophy are described which broadens the previously described phenotype. Blood smear in one patient showed Jordan's anomaly (vacuolated leukocytes). Gastrointestinal features occurred in two patients and there appeared to be acute deterioration with infection/general anaesthesia. Brain imaging showed no structural changes but brain magnetic resonance proton spectroscopy (MRS) demonstrated significant reduction in choline:N-acetyl aspartate and choline:creatine ratios in keeping with a general decrease in the amount of choline and phosphocholine-based substrate. Muscle pathology showed either myopathic or dystrophic features, uneven oxidative enzyme staining, COX deficient fibres and peripherally located large mitochondria. CHKB activity was reduced in all three patients and complex 1 activity was significantly reduced in one patient.

<http://www.ncbi.nlm.nih.gov/pubmed/23692895>

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## Late effects of childhood cancer treatment: Severe hypertriglyceridaemia, central obesity, non alcoholic fatty liver disease and diabetes as complications of childhood total body irradiation

Rajendran, R., et al.

*Diabet Med* 2013; 30(8): e239-242

BACKGROUND: Childhood cancer survivors may develop a number of endocrine complications linked to organ failure, such as hypogonadism, diabetes and growth hormone deficiency. However, increasing evidence now suggests that total body irradiation treatment, specifically, is linked with future risk of insulin resistance, hepatic steatosis and dyslipidaemia, possibly because total body irradiation affects adipocyte differentiation and impairs subcutaneous adipose tissue depot expansion during times of positive energy balance. CASE REPORT: We describe a 20-year-old woman who developed pancreatitis with severe hypertriglyceridaemia (serum triglycerides > 300 mmol/l) that required plasmapheresis. She had received total body irradiation prior to her bone marrow transplant at age 6 years for relapsed acute lymphoblastic leukaemia. She developed ovarian failure at age 12 years. At age 15 years she was noted to have hyperglycaemia, increased blood pressure, hepatic steatosis and mild hypertriglyceridaemia. She presented with severe hypertriglyceridaemia and eruptive xanthoma, and developed pancreatitis 12 h after admission. She was treated with plasmapheresis and intravenous insulin and made an excellent recovery. We implicate and discuss total body irradiation as the major contributing factor to her severe hypertriglyceridaemia, compounded by worsening glycaemic control, oestrogen deficiency and a changing adult lifestyle. CONCLUSION: Children who have received total body irradiation are at risk of diabetes and an exaggerated form of the metabolic syndrome with hypertriglyceridaemia, which can be life-threatening. We suggest that survivors of total body irradiation treatment require careful lifelong monitoring of their metabolic status.

<http://www.ncbi.nlm.nih.gov/pubmed/23692373>

## The adolescent and young adult with cancer: State of the art: Bone tumors

Rainusso, N., et al.

*Curr Oncol Rep* 2013; 15(4): 296-307

Primary malignant bone tumors in the pediatric to young adult populations are relatively uncommon and account for about 6 % of all cancers in those less than 20 years old [1] and 3 % of all cancers in adolescents and young adults (AYA) within the age range of 15 to 29 years [2]. Osteosarcoma (OS) and Ewing's sarcoma (ES) comprise the majority of malignant bone tumors. The approach to treatment for both tumors consists of local control measures (surgery or radiation) as well as systemic therapy with high-dose chemotherapy. Despite earlier advances, there have been no substantial improvements in outcomes over the past several decades, particularly for patients with metastatic disease. This review summarizes the major advances in the treatment of OS and ES and the standard therapies available today, current active clinical trials, and areas of investigation into molecularly targeted therapies.

<http://www.ncbi.nlm.nih.gov/pubmed/23690089>

## Thyroid cancers in children, adolescents, and young adults with and without a history of childhood exposure to therapeutic radiation for other cancers

Sassolas, G., et al.

*Thyroid* 2013; 23(7): 805-810

**BACKGROUND:** The thyroid is highly sensitive to the carcinogenic effect of radiation in children. We compared, in patients with and without earlier childhood radiation, the features of papillary thyroid cancer (PTC) diagnosed in later childhood through young adulthood. **METHODS:** Patients were from the Rhone-Alpes Thyroid Cancer Registry. Twenty-four patients (RAD group) had been treated by radiation therapy for nonthyroid neoplasms at the age of 8.0+/-6.0 years (mean+/-SD) and by surgery for PTC at the age of 17+/-6.4 years. They were compared with 413 patients with PTC but no radiation exposure (sPTC group, age 23+/-4.8 years). The two groups were subdivided into three subgroups, ages 8-14 (children), 15-20 (adolescents), and 21-29 years (adults) at time of PTC diagnosis, and compared to matched subgroups from 80 patients in the sPTC group (M-sPTC). Age in years at PTC diagnosis (RAD vs. M-sPTC) was 12+/-2 compared with 12+/-2 for children, 17+/-1 compared with 19+/-1 for adolescents, and 25+/-3.2 compared with 25+/-2.5 for adults. The matched subgroups had comparable pTNM, treatments, and follow-up. We compared the histopathological characteristics of the initial specimens and the outcome events. **RESULTS:** The RAD group and the sPTC group were similar in terms of age when PTC was diagnosed. RAD tumors had significantly more lymph node metastases ( $p=0.007$ ) and a higher proportion of invasive pTN3 stage tumors ( $p=0.01$ ). The adult RAD subgroup ( $n=8$ ) was more likely to have lymph node metastases ( $p=0.004$ ) and a higher proportion of invasive pT3N+ stage tumors ( $p=0.01$ ) than the adult sPTC subgroup ( $n=316$ ). During the 6.5 years of follow-up, there was no difference in the risk of cervical recurrence between the RAD group and the M-sPTC groups. Risk of cervical recurrence was also similar for tumors that were high risk (pT3N+). **CONCLUSION:** Young adults with PTC associated with radiation therapy for nonthyroid neoplasms in childhood have a more aggressive initial presentation than young adults with sporadic PTC. The risk of recurrent disease in patients who received radiation in early childhood through adolescence and who developed PTC in late childhood through early adulthood is similar to those who did not receive radiation.

<http://www.ncbi.nlm.nih.gov/pubmed/23286372>



## Considerations for the treatment of infantile neuronal ceroid lipofuscinosis (infantile Batten disease)

Sands, M. S.

*J Child Neurol* 2013; 28(9): 1151-1158

The infantile form of neuronal ceroid lipofuscinosis (ie, infantile Batten disease) is the most rapidly progressing type and is caused by an inherited deficiency in the lysosomal enzyme palmitoyl protein thioesterase 1. The absence of enzyme activity leads to progressive accumulation of autofluorescent material in many cell types, particularly neurons of the central nervous system. Clinical signs of infantile neuronal ceroid lipofuscinosis appear between 6 months and 1 year of age and include vision loss, cognitive decline, motor deficits, seizures, and premature death, typically by 3 to 5 years of age. There is currently no effective treatment. However, preclinical experiments in the murine model of infantile neuronal ceroid lipofuscinosis have shown that gene therapy, enzyme replacement, stem cell transplantation, and small-molecule drugs, alone or in combination, can significantly slow disease progression. A more thorough understanding of the underlying pathogenesis of infantile neuronal ceroid lipofuscinosis will identify new therapeutic targets.

<http://www.ncbi.nlm.nih.gov/pubmed/23517304>

## Second malignant neoplasms after treatment of childhood acute lymphoblastic leukemia

Schmidt, E. and B. Burkhardt

*J Clin Oncol* 2013; 31(19): 2469-2476

**PURPOSE:** Second malignant neoplasms (SMNs) after diagnosis of childhood acute lymphoblastic leukemia (ALL) are rare events. **PATIENTS AND METHODS:** We analyzed data on risk factors and outcomes of 642 children with SMNs occurring after treatment for ALL from 18 collaborative study groups between 1980 and 2007. **RESULTS:** Acute myeloid leukemia (AML; n = 186), myelodysplastic syndrome (MDS; n = 69), and nonmeningioma brain tumor (n = 116) were the most common types of SMNs and had the poorest outcome (5-year survival rate, 18.1% +/- 2.9%, 31.1% +/- 6.2%, and 18.3% +/- 3.8%, respectively). Five-year survival estimates for AML were 11.2% +/- 2.9% for 125 patients diagnosed before 2000 and 34.1% +/- 6.3% for 61 patients diagnosed after 2000 (P < .001); 5-year survival estimates for MDS were 17.1% +/- 6.4% (n = 36) and 48.2% +/- 10.6% (n = 33; P = .005). Allogeneic stem-cell transplantation failed to improve outcome of secondary myeloid malignancies after adjusting for waiting time to transplantation. Five-year survival rates were above 90% for patients with meningioma, Hodgkin lymphoma, thyroid carcinoma, basal cell carcinoma, and parotid gland tumor, and 68.5% +/- 6.4% for those with non-Hodgkin lymphoma. Eighty-nine percent of patients with brain tumors had received cranial irradiation. Solid tumors were associated with cyclophosphamide exposure, and myeloid malignancy was associated with topoisomerase II inhibitors and starting doses of methotrexate of at least 25 mg/m<sup>2</sup> per week and mercaptopurine of at least 75 mg/m<sup>2</sup> per day. Myeloid malignancies with monosomy 7/5q- were associated with high hyperdiploid ALL karyotypes, whereas 11q23/MLL-rearranged AML or MDS was associated with ALL harboring translocations of t(9;22), t(4;11), t(1;19), and t(12;21) (P = .03). **CONCLUSION:** SMNs, except for brain tumors, AML, and MDS, have outcomes similar to their primary counterparts.

<http://www.ncbi.nlm.nih.gov/pubmed/23690411>

## Lymphoblastic lymphoma in childhood and adolescence

Schmidt, E. and B. Burkhardt

*Pediatr Hematol Oncol* 2013; 30(6): 484-508

Lymphoblastic lymphoma (LBL) are thought to derive from immature precursor T-cells or B-cells. LBL are the second most common subtype of Non-Hodgkin Lymphoma (NHL) in children and adolescents. LBL are closely related to acute lymphoblastic leukemia (ALL), the most common type of cancer in children. Using ALL-type treatment regimen to treat children with LBL was an important development in the treatment of LBL. During the last decades, several systematic clinical trials contributed to the controlled optimization of treatment. Today event-free survival (EFS) can be achieved for 75-90% of patients. However, acute and long-term toxicity, the lack of prognostic parameters and the poor outcome for patients who suffer from refractory or relapsed LBL remain highly relevant subjects for improvement. To date, the pathogenesis of LBL is poorly understood. Learning more about the biology and pathogenesis of LBL might pave the way for targeted treatment to improve survival especially in relapsed and refractory patients.

<http://www.ncbi.nlm.nih.gov/pubmed/23621872>

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## Management of children with spinal muscular atrophy type 1 in Australia

Tassie, B., et al.

*J Paediatr Child Health* 2013; 49(10): 815-819

AIMS: The study aims to: (i) estimate the prevalence of spinal muscular atrophy type 1 (SMA 1); (ii) describe what practices characterise end-of-life care of patients with SMA 1; and (iii) ascertain whether a consistent approach to the management of these patients exists in Australia. METHODS: An audit of the Australasian pathology laboratories offering the diagnostic SMN1 deletion test was conducted for patients diagnosed with SMA in Australia for 2010 and 2011. In addition, a retrospective clinical audit was conducted in eight major Australian paediatric hospitals of the end-of-life care provided to children with confirmed SMA 1 from 2005 to 2010. RESULTS: Thirty-five children were included in the clinical audit, accounting for an estimated 61% of children diagnosed with SMA 1 from 2005 to 2010. Twenty-six per cent were ventilated invasively, only two of whom were intubated after the diagnosis was confirmed. No children were ventilated long term (>90 days) or had a tracheostomy performed. Nasogastric tube feeding was a common measure to support adequate nutritional intake. Total parenteral nutrition, gastrostomy and fundoplication were not provided for any children. Conflict over end-of-life care decisions was documented in one instance, without the involvement of a guardianship tribunal. CONCLUSION: There appears to be a consistent approach in the management of children with SMA 1 in Australia, which can be characterised as 'actively managed dying'. This study could contribute to the development of Australian consensus guidelines for the management of these children. These results also highlight a number of ethical issues related to the management of children with SMA 1.

<http://www.ncbi.nlm.nih.gov/pubmed/23834358>

## SUDEP and other causes of mortality in childhood-onset epilepsy

Sillanpaa, M. and S. Shinnar

*Epilepsy Behav* 2013; 28(2): 249-255

**BACKGROUND:** There are few prospective studies on the causes of mortality in well-characterized cohorts with epilepsy and even fewer that have autopsy data that allow for reliable determination of SUDEP. We report causes of mortality and mortality rates in the Finnish cohort with childhood-onset epilepsy. **METHODS:** A population-based cohort of 245 children with epilepsy in 1964 has been prospectively followed for almost 40 years. Seizure outcomes and mortality were assessed. Autopsy data were available in 70% of the cases. Sudden unexpected death in epilepsy (SUDEP) rates were assessed, and SUDEP was confirmed by autopsy. **RESULTS:** During the follow-up, 60 subjects died. The major risk factor for mortality was lack of terminal remission ( $p < 0.0001$ ). Remote symptomatic etiology also increased the risk for death ( $p < 0.0001$ ) but did not remain significant on multivariate analysis after adjusting for effect of remission. Of the deaths, 33/60 (55%) were epilepsy-related including SUDEP in 23/60 (38%) using the Nashef criteria, status epilepticus in 4/60 (7%), and accidental drowning in 6/60 (10%). The nonepilepsy-related deaths occurred primarily in the remote symptomatic group and were often related to the underlying disorder or to medical comorbidities that developed after the onset of the epilepsy. Risk factors for SUDEP on multivariable analysis included lack of 5-year terminal remission and not having a localization-related epilepsy. In cryptogenic/idiopathic cases, SUDEP did not occur in childhood but begins only in adolescence. **CONCLUSION:** Childhood-onset epilepsy is associated with a substantial risk of epilepsy-related mortality, primarily SUDEP. In otherwise neurologically normal individuals, the increased SUDEP risk begins in adolescence. The higher mortality rates reported in this cohort are related to duration of follow-up as most of the mortality occurs many years after the onset of the epilepsy.

<http://www.ncbi.nlm.nih.gov/pubmed/23746924>

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## Second primary brain tumors following cranial irradiation for pediatric solid brain tumors

You, S. H., et al.

*Childs Nerv Syst* 2013; 29(10): 1865-1870

**PURPOSE:** We describe our institution's experience with seven patients who developed second brain tumors following cranial irradiation. **METHODS:** The median age at first irradiation was 8 years (range, 3-20 years). Initial diagnoses were two cases of germinoma, one non-germinomatous germ cell tumor (NGGCT), three cases of medulloblastoma, and one pineal gland tumor (pathology undetermined). All patients received craniospinal irradiation followed by local boost and the median dose to the initial tumor area was 54.0 Gy (range, 49.8-60.6 Gy). Four patients (two medulloblastomas, one germinoma, and one NGGCT) received chemotherapy. **RESULTS:** Second brain tumors were diagnosed a median of 114 months (range, 64-203) after initial radiation. Pathologic diagnoses were one glioblastoma, two cases of anaplastic astrocytoma, one medulloblastoma, one low-grade glioma, one high-grade glial tumor, and one atypical meningioma. Five patients underwent surgical resection with subsequent radiotherapy. One anaplastic astrocytoma patient received chemotherapy only following stereotactic biopsy. The meningioma patient was alive 32 months after total resection and radiosurgery for subsequent recurrences. Six patients died within 18 months and most deaths were due to disease progression. **CONCLUSIONS:** Most patients diagnosed with second brain tumors had received high-dose, large-volume radiotherapy with chemotherapy at a young age. Further studies are required to determine the relationship between radiotherapy/chemotherapy and the development of secondary brain tumors.

<http://www.ncbi.nlm.nih.gov/pubmed/23571774>

## Gastrostomy in children with cystic fibrosis and portal hypertension

Vandeleur, M., et al.

*J Pediatr Gastroenterol Nutr* 2013; 57(2): 245-247

Children with cystic fibrosis-associated liver disease (CFALD) and portal hypertension may require supplemental feeding with gastrostomy; however, this could lead to the development of stomal varices. We assessed this risk and nutritional and pulmonary outcome in a series of 7 children with CFALD and portal hypertension. In 35.1 patient-years of follow-up, none developed stomal varices or had a gastrointestinal bleed attributable to a varix. There was significant improvement in nutrition and lung function 2 years postinsertion. We conclude that gastrostomy placement for poor nutrition in children with CFALD and portal hypertension is safe and contributes to improved nutritional and pulmonary outcome.

<http://www.ncbi.nlm.nih.gov/pubmed/23575297>

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## Treatment of electrographic seizures and status epilepticus in critically ill children: A single center experience

Abend, N. S., et al.

*Seizure* 2013; 22(6): 467-471

**PURPOSE:** Electrographic seizures (ES) and electrographic status epilepticus (ESE) are common in encephalopathic children in the pediatric intensive care unit (PICU) and associated with worse short-term outcome. Survey data indicate most physicians treat ES and ESE with antiepileptic drugs (AEDs), but few data are available regarding AED usage patterns. We aimed to describe AED usage for ES and ESE in critically ill children. **METHODS:** We performed an observational study of patients who underwent continuous electroencephalographic (cEEG) monitoring in the PICU of a single quaternary care children's hospital. We collected data regarding age, clinical diagnoses, ES and ESE occurrence, and AEDs utilized. **RESULTS:** 200 subjects underwent cEEG. ES occurred in 21% (41/200) and ESE occurred in 22% (43/200). Of the 84 patients with ES or ESE, 80 received non-benzodiazepine AEDs including 48% (38 of 80) with ES and 52% (42 of 80) with ESE. The most commonly administered first AEDs were levetiracetam in 38% (30/80), phenobarbital in 31% (25/80), phenytoin-fosphenytoin in 28% (22/80), and valproate in 4% (3/80). Seizures terminated after administration of the first AED in 74% (28/38) with ES and 22% (9/41) with ESE. **CONCLUSIONS:** Levetiracetam, phenobarbital, and phenytoin-fosphenytoin are commonly used to manage ES and ESE at our center. Over half of subjects received multiple AEDs.

<http://www.ncbi.nlm.nih.gov/pubmed/23601851>

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## Does school attendance during initial cancer treatment in childhood increase the risk of infection?

Sandeberg, M., et al.

*Pediatr Blood Cancer* 2013; 60(8): 1307-1312

**BACKGROUND:** The present study aimed to investigate the relationship between school attendance and infection requiring antimicrobial treatment in children undergoing treatment for cancer. **PROCEDURE:** A national cohort of children aged 7-16 years undergoing cancer treatment was assessed during two observation periods of 19 days each, 1 month (n = 89) and 2.5 months (n = 89) poststart of treatment. Children free from infection at start of each observation period were included. Multivariable logistic regression analyses were performed including factors potentially associated with start of antimicrobial treatment. **RESULTS:** Twenty-seven (30%) children started antimicrobial treatment during the first observation period. Factors associated with an increased risk of starting antimicrobial treatment were diagnosed with sarcoma (OR = 24.37, P = 0.002) or non-Hodgkin lymphoma (OR = 17.57, P = 0.025), having neutropenia (OR = 5.92, P = 0.020) and age less than 13 years (OR = 8.54, P = 0.014). During the second observation period, when 20 (22%) children started antimicrobial treatment, the probability of starting treatment was increased in children with neutropenia (OR = 4.25, P = 0.007). There was no statistically significant association between starting treatment for infection and school attendance. **CONCLUSIONS:** In this study, children attending school while undergoing cancer treatment did not run a higher risk of starting antimicrobial treatment than children absent from school. However, there is a need for further studies evaluating risk of infections in children with ongoing cancer treatment.

<http://www.ncbi.nlm.nih.gov/pubmed/23596132>

## Hunter syndrome (Mucopolysaccharidosis type II), severe phenotype: Long term follow-up on patients undergone to hematopoietic stem cell transplantation

Annibali, R., et al.

*Minerva Pediatr* 2013; 65(5): 487-496

AIM: Our study aim is the evaluation of long-term effects of hematopoietic stem cell transplantation on Italian patients with severe Hunter syndrome. METHODS: Four boys, suffering from Hunter syndrome, severe phenotype, received hematopoietic stem cell transplantation between 2 years 6 months and 2 years 11 months of age, from 1992 to 2001. A complete multidisciplinary evaluation of hematopoietic stem cell transplantation long-term effects was performed periodically. RESULTS: All patients achieved successful engraftment. Urine glycosaminoglycans excretion was reduced or normalized, and the activity of leukocyte iduronate-2-sulphatase enzyme, absent before hematopoietic stem cell transplantation, remained constant, in all patients. Dysostosis multiplex progressed over time, according to the natural evolution of the disease. Joint stiffness improved in all affected districts. Hepatosplenomegaly decreased until it disappeared. The cardiovascular involvement stayed unchanged, as well as hearing loss. Skin became hyperelastical; face features seemed less coarse if compared to the natural evolution of the disease. Cerebral white matter alterations were constant in time. On the contrary, the hematopoietic stem cell transplantation did not prove to have long-term effectiveness on neurological symptoms of Hunter syndrome. CONCLUSION: The hematopoietic stem cell transplantation was successful in slowing the progression of Hunter syndrome, and even the evolution of neurological feature of the disease was slower in the first years after this treatment.

<http://www.ncbi.nlm.nih.gov/pubmed/24056375>

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## Treatment with topotecan plus cyclophosphamide in children with first relapse of neuroblastoma

Ashraf, K., et al.

*Pediatr Blood Cancer* 2013; 60(10): 1636-1641

BACKGROUND: Reports of responses and toxicities of salvage therapies for relapsed neuroblastoma are rare and often confounded by effects of additional treatments. Our objective was to describe the outcomes and toxicities for a topotecan and cyclophosphamide (TOPO/CTX) regimen for first relapse or progression of high-risk neuroblastoma. METHODS: We retrospectively reviewed charts of relapsed or refractory neuroblastoma patients treated between 1999 and 2009 with our standard-of-care outpatient TOPO/CTX (0.75 and 250 mg/m<sup>2</sup> /day x 5 days q3-4 weeks). RESULTS: Twenty-seven patients received 343 cycles of TOPO/CTX (median 10 cycles per patient, range 1-32). Most patients (N = 25) had undergone autologous stem cell transplantation. Seventeen (63%) patients had an objective response (CR + PR + MR). The 3-year progression-free survival (PFS) after relapse was 11 +/- 6% and 3-year overall survival (OS) after relapse was 33 +/- 9%. The median PFS was 1.2 years and the median OS was 2.3 years. Five patients are alive with follow-up of 3.1-5.5 years. Shorter time from diagnosis to relapse (6-18 months) was associated with shorter OS. The majority of patients experienced chemotherapy delays, transfusions, and febrile neutropenia, including eight bacterial infections. The mean number of hospitalized days was less than one per cycle. CONCLUSIONS: TOPO/CTX was well tolerated and resulted in response rates and PFS similar to those reported for patients treated on COG 9462. Our study provides additional toxicity, historical endpoints, and time-to-progression data against which new agents and combination therapies using TOPO/CTX as a backbone can be measured.

<http://www.ncbi.nlm.nih.gov/pubmed/23650219>

## Urological manifestations of Duchenne muscular dystrophy

Askeland, E. J., et al.

*J Urol* 2013; 190(4 Suppl): 1523-1528

**PURPOSE:** Duchenne muscular dystrophy is a dystrophinopathy affecting males that is associated with multiple organ system complications. To our knowledge urological complications of Duchenne muscular dystrophy have been described only anecdotally to date. **MATERIALS AND METHODS:** We reviewed the medical charts of 135 patients with Duchenne or Duchenne-Becker muscular dystrophy for demographics and disease progression, urological diagnoses, intervention and followup. **RESULTS:** Of 135 patients 67 (50%) had at least 1 documented urological diagnosis and 38 (28%) had multiple manifestations. Lower urinary tract symptoms were the most common urological diagnosis (32% of patients). Survival analysis revealed a median age at onset of lower urinary tract symptoms of 23 years (95% CI 17.7-23.9). Intervention was required in 12 patients (9%), most commonly due to nephrolithiasis. Urological morbidity increased with Duchenne muscular dystrophy progression when stratified by clinical progression. Lower urinary tract symptoms were more common in nonambulatory patients (40.7% vs 19%,  $p = 0.007$ ), those with a diagnosis of scoliosis (44% vs 19.7%,  $p = 0.003$ ) and/or scoliosis spine surgery (60% vs 22%,  $p < 0.001$ ), and those on invasive respiratory support (53% vs 29%,  $p = 0.046$ ). Likewise, nephrolithiasis was more common in nonambulatory patients (10% vs 0%,  $p = 0.017$ ), those with scoliosis (12% vs 0%,  $p = 0.004$ ) and/or scoliosis spine surgery (20% vs 1%,  $p < 0.001$ ), and those on invasive respiratory support (29% vs 3%,  $p < 0.001$ ). Only 28% of patients with a urological manifestation were referred to urology. **CONCLUSIONS:** As these patients transition into adolescence and adulthood, the increased prevalence of urological manifestations warrants increased awareness and referral to urologists.

<http://www.ncbi.nlm.nih.gov/pubmed/23357214>

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## External validity and reliability of the Psychosocial Assessment Tool (PAT) among Canadian parents of children newly diagnosed with cancer

Barrera, M., et al.

*Pediatr Blood Cancer* 2014; 61(1): 165-170

**BACKGROUND:** The Psychosocial Screening Tool (PAT) was developed and validated with a sample of caregivers of children newly diagnosed with cancer in the United States. This study aimed to assess cultural adaptation (Phase 1) and validity and reliability of the revised PAT (PATrev) with a Canadian sample (Phase 2). **PROCEDURE:** In Phase 1, a convenience sample of seven parents of children who were treated for cancer and six pediatric oncology healthcare experts participated. In Phase 2, 67 parents of children newly diagnosed with cancer from 4 Canadian pediatric cancer centers participated. To assess reliability and validity of the PATrev, parents completed behavioral (BASC-2) and quality of life (PedsQL) instruments about the child and an anxiety inventory (STAI) about themselves. **RESULTS:** The PAT required minor changes to be culturally adapted for the Canadian population. The PATrev had strong inter-rater (0.77) test-retest (0.75), and internal consistency reliability (0.85), as well as moderate to strong validity comparing PATrev child's problems and PedsQL total (-0.49), PedsQL anxiety (-0.47), BASC-2 internalizing (0.64), behavioral (0.63), and adaptive scores (-0.56). PATrev discriminative validity was confirmed with BASC-2 scores (AUR scores of 0.70-0.74). PATrev parental stressors were strongly correlated to STAI scores (0.53). Finally, agreement between PATrev child's problems and parental anxiety scores was moderate (0.47). **CONCLUSION:** This study supports the original PAT, demonstrates PATrev is a reliable and valid psychosocial screening tool, and provides unique evidence regarding early psychosocial risk in the family, which have important implications for guiding psychosocial practice.

<http://www.ncbi.nlm.nih.gov/pubmed/24106172>

## The relationship between the site of metastases and outcome in children with stage IV Wilms Tumor: Data from 3 European Pediatric Cancer Institutions

Berger, M., et al.

*Pediatr Hematol Oncol* 2013; 35(7): 518-524

The aim of this study was to analyze in detail the site of metastasis of stage 4 Wilms tumor (WT) and its correlation with outcome. The databases from 3 major European pediatric cancer institutions were screened for children with WT between 1994 and 2011. Of 208 children identified, 31 (14.9%) had metastases at diagnosis. The lung was affected in 29 children (93.5%) and the liver in 6 children (19.4%). Twenty-seven children (87.1%) had metastases isolated to 1 organ, with the lung being the most common site (80.7%). Five-year overall survival was significantly better in those children with distant disease in either lung or liver (95.8%) compared with those affected in both lung and liver (57.1%,  $P=0.028$ ). Further, prognostic markers were the response of metastases to preoperative chemotherapy ( $P=0.0138$ ), high-risk histology ( $P=0.024$ ), and local stage ( $P=0.026$ ). Five-year overall survival was 82.1% and 5-year event-free survival was 67.9%. The overall follow-up time was 74.1 and 87.2 (2 to 151) months among survivors, and the treatment-related complication rate was 16.7%. In conclusion, in our series of stage 4 WT, prognosis was excellent if histology was favorable, metastatic disease was isolated to either lungs or liver, and if metastases responded to preoperative chemotherapy.

<http://www.ncbi.nlm.nih.gov/pubmed/23588334>

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## Subsequent neoplasms of the CNS among survivors of childhood cancer: A systematic review

Bowers, D. C., et al.

*Lancet Oncol* 2013; 14(8): e321-328

Childhood cancer survivors are at risk for development of subsequent neoplasms of the CNS. Better understanding of the rates, risk factors, and outcomes of subsequent neoplasms of the CNS among survivors of childhood cancer could lead to more informed screening guidelines. Two investigators independently did a systematic search of Medline and Embase (from January, 1966, through March, 2012) for studies examining subsequent neoplasms of the CNS among survivors of childhood cancer. Articles were selected to answer three questions: what is the risk of CNS tumours after radiation to the cranium for a paediatric cancer, compared with the risk in the general population; what are the outcomes in children with subsequent neoplasms of the CNS who received CNS-directed radiation for a paediatric cancer; and, are outcomes of subsequent neoplasms different from primary neoplasms of the same histology? Our search identified 72 reports, of which 18 were included in this Review. These studies reported that childhood cancer survivors have an 8.1-52.3-times higher incidence of subsequent CNS neoplasms compared with the general population. Nearly all cancer survivors who developed a CNS neoplasm had been exposed to cranial radiation, and some studies showed a correlation between radiation dose and risk of subsequent CNS tumours. 5-year survival ranged from 0-19.5% for subsequent high-grade gliomas and 57.3-100% for meningiomas, which are similar rates to those observed in patients with primary gliomas or meningiomas. The quality of evidence was limited by variation in study design, heterogeneity of details regarding treatment and outcomes, limited follow-up, and small sample sizes. We conclude that survivors of childhood cancer who received cranial radiation therapy have an increased risk for subsequent CNS neoplasms. The current literature is insufficient to comment about the potential harms and benefits of routine screening for subsequent CNS neoplasms.

<http://www.ncbi.nlm.nih.gov/pubmed/23816298>



## Does goal setting in activity-focused interventions for children with cerebral palsy influence treatment outcome?

Brogren Carlberg, E. and K. Lowing

*Dev Med Child Neurol* 2013; 55 Suppl 4: 47-54

Today, treatment for children with cerebral palsy predominantly aims at improving the children's possibilities to perform everyday activities in their natural environment. The activities in focus for intervention are often expressed as specific goals, frequently defined in a collaborative goal-setting process between professionals and parents. The role of goal setting to improve the outcome of the intervention has not been shown in the literature so far. Thus, the aim of this systematic review was to explore if goal setting has an impact on treatment outcome assessed by standardized measures. CINAHL and MEDLINE were searched from January 2000 to October 2012, resulting in a final selection of 13 articles, six of which were randomized controlled trials. Methodological quality was assessed and study characteristics were analysed descriptively. Subject characteristics, type of intervention/s, frequency, and intensity of therapy varied largely. Outcome was assessed by standardized outcome measures as well as evaluated through aspects of goal attainment. Most studies showed robust within-group changes according to study-appropriate standardized measures, whereas the between-group comparisons exhibited less consistent differences in outcome. The review does not provide support for a positive effect of goal setting per se on treatment outcome. Studies that specifically measure the effect of goal setting on treatment outcome are needed.

<http://www.ncbi.nlm.nih.gov/pubmed/24237280>

## Invasive bacterial and fungal infections in paediatric patients with cancer: Incidence, risk factors, aetiology and outcomes in a UK regional cohort 2009-2011

Calton, E. A., et al.

*Pediatr Blood Cancer* 2014; 61(7): 1239-1245

**BACKGROUND:** Cancer is the second most common cause of childhood deaths in the United Kingdom and infection contributes to a quarter of all cancer-related deaths. This study aimed to estimate the risk, aetiology and outcome of bloodstream bacterial and fungal infections in children with cancer within a geographically defined region in South-West London over a 3-year period. **METHODS:** Web-based questionnaires were completed using case records of children with positive blood cultures admitted to five London hospitals during 2009-2011. **RESULTS:** A total of 112 children with a median age of 5.4 (IQR 3.6-11.2) years had 266 significant blood cultures during 149 infection episodes. Haematological malignancy affected 68 patients (60.7%) and solid tumours 44 (39.3%). The overall bloodstream infection rate was 1.5 episodes per 1,000 days-at-risk (95% CI, 1.2-1.8) and was similar for those with haematological malignancies and solid tumours. Most episodes were attributed to central venous catheter infection (120/149, 80.5%). Coagulase-negative staphylococci were isolated in almost half the bloodstream infections (127/266; 47.7%), while Gram-negative organisms accounted for a further quarter (64/266; 24.1%). Fungal isolates from blood were uncommon (8/112 children, 7.1%) but significantly associated with neutropenia (18/149 [12.1%] vs. 1/114 [0.9%],  $P = 0.0004$ ). Six children (5.4%) died, including three (2.7%; 95% CI, 0.6-7.6%) whose deaths were infection-related. **CONCLUSIONS:** This study provides an updated risk estimate for bloodstream infections in children with cancer and adds to the framework for developing evidence-based guidance for management of suspected infections in this highly vulnerable group.

<http://www.ncbi.nlm.nih.gov/pubmed/24615980>

## Screening for pain in pediatric brain tumor survivors using the pain thermometer

Chordas, C., et al.

*J Pediatr Oncol Nurs* 2013; 30(5): 249-259

Numerous instruments have been developed to measure pain within various populations; however, there remains limited understanding of how these tools are applicable to childhood cancer survivors. This study compared a single-item screening measure, the Pain Thermometer (PT), with a more in-depth measure, the Brief Pain Survey (BPS), in a cohort of childhood brain tumor survivors. Ninety-nine survivors (aged 13-32 years) with a median time from diagnosis of 9.9 years (range = 2-18 years) completed the 2 instruments. Thirty-seven survivors (37.4%) were identified on the BPS as having clinically significant pain, but the PT was not found to be an accurate tool for identifying these pain cases. Application of receiver operating characteristic curve analysis of PT ratings against BPS criterion indicated overall concordance between measures. No cutoff score on the PT were identified that resulted in acceptable sensitivity, meaning pain cases identified on the BPS would be missed on the PT. Findings suggest that a multi-item screening measure may better identify clinically significant pain in childhood brain tumor survivors compared with a 1-item screening measure alone.

<http://www.ncbi.nlm.nih.gov/pubmed/23867966>

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## Encephalopathy with status epilepticus during sleep or continuous spikes and waves during slow sleep syndrome: A multicenter, long-term follow-up study of 117 patients

Caraballo, R. H., et al.

*Epilepsy Res* 2013; 105(1-2): 164-173

**PURPOSE:** To retrospectively analyze the electroclinical features, etiology, treatment and prognosis of 117 patients with encephalopathy with status epilepticus during sleep (ESES) or continuous spike and waves slow sleep (CSWSS) syndrome with a long-term follow-up. **METHODS:** Charts of 117 patients with ESES/CSWSS syndrome followed between 1990 and 2012 were analyzed. Inclusion criteria were: (1) focal seizures or apparently generalized seizures and focal EEG epileptiform discharges; (2) further occurrence of atypical absences, and myoclonic, atonic, and/or generalized seizures; (3) cognitive impairment and/or behavior disturbances; (4) continuous spike-and-wave discharges during slow sleep in more than 85% of non-REM sleep. Patients with spike-and-wave discharges in less than 85% of slow sleep were also analyzed. **KEY FINDINGS:** 'Mean follow-up from onset of ESES/CSWSS was 13 years (range, 2-22 years) in the symptomatic/structural and non-idiopathic group consisting of 79 children and 10.5 years (range, 2-21 years) in the idiopathic group consisting of 38 children. The comparison of clinical findings and localization of paroxysmal EEG abnormalities (focal, multifocal, or generalized) at the different stages (before, during, and after ESES/CSWSS) and the percentage of spike-wave index during ESES/CSWSS between the symptomatic/structural and non-idiopathic and the idiopathic group was not statistically significant. **SIGNIFICANCE:** ESES/CSWSS syndrome is an epileptic encephalopathy with similar electroclinical findings in children with a >85% spike-wave index and those with a <85% spike-wave index. In this series of patients, the most commonly used treatments were clobazam, ethosuximide, sulthiame, alone or in combination. In refractory cases, high-dose steroids were administered. Among the AED responders, the idiopathic cases returned to normality and the structural cases returned to baseline cognitive development.

## Noninvasive ventilation for acute respiratory distress in children with central nervous system disorders

Falsaperla, R., et al.

*Respir Med* 2013; 107(9): 1370-1375

**BACKGROUND:** Acute respiratory distress (ARD) is a relatively frequent occurrence in patients suffering from central nervous system disorders (CNSD) and moderate to severe mental retardation. Whenever conventional therapy is little effective, noninvasive mechanical ventilation (NIV) is the additional treatment in patients with diseases of the peripheral nervous system. However, NIV is traditionally little employed in the acute phase in patients suffering from CNSD. In the latter, either conventional therapy is maintained or invasive mechanical ventilation is instituted if the patient's condition worsens severely. To challenge the traditional view, we conducted the study to prove that NIV is both applicable and effective in the treatment of ARD also in children with moderate to severe mental retardation. **METHODS:** We studied 44 children with ARD secondary to pneumonia and CNSD causing moderate to severe mental retardation. The children were divided in two groups. One group received conventional therapy and NIV, the other conventional therapy only, before being advanced to invasive ventilator support when nonresponding. On admission to hospital and one hour following admission we registered pH, PaCO<sub>2</sub>, PaO<sub>2</sub>, A - a DO<sub>2</sub> and the PaO<sub>2</sub>/FIO<sub>2</sub> ratio. The mean hospital stay was also recorded. **RESULTS:** After one hour on NIV PaO<sub>2</sub> and pH increased, PaCO<sub>2</sub> decreased, A - a DO<sub>2</sub> and PaO<sub>2</sub>/FIO<sub>2</sub> ratio improved. No changes in the above parameters were observed in children on conventional therapy only. Hospital stay was shorter when NIV was instituted. **CONCLUSIONS:** NIV is both applicable and beneficial in stabilizing blood gases, respiratory and cardiovascular parameters also in children with CNSD. Moreover its use shortens the hospital stay.

<http://www.ncbi.nlm.nih.gov/pubmed/23906815>

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## Prompt administration of antibiotics is associated with improved outcomes in febrile neutropenia in children with cancer

Fletcher, M., et al.

*Pediatr Blood Cancer* 2013; 60(8): 1299-1306

**BACKGROUND:** Time-to-antibiotic (TTA) administration is a widely used quality-of-care measure for children with cancer and febrile neutropenia (FN). We sought to determine whether TTA is associated with outcomes of FN. **PROCEDURE:** A single-center, retrospective cohort study was conducted of 1,628 FN admissions from 653 patients from 2001 to 2009. Outcome variables included (1) an adverse event (AE) composite of in-hospital mortality, pediatric intensive care unit (PICU) admission within 24 hours of presentation, and/or fluid resuscitation  $\geq 40$  ml/kg within 24 hours of presentation and (2) length of stay (LOS). TTA was measured as a continuous variable and in 60-minute intervals. Mixed regression models were constructed to evaluate associations of TTA with the outcome variables after adjusting for relevant covariates including cancer diagnosis, degree of myelosuppression, and presence of bacteremia. **RESULTS:** The composite AE outcome occurred in 11.1% of admissions including 0.7% in-hospital mortality, 4.7% PICU admission, and 10.1% fluid resuscitation. In univariate analysis, TTA was associated with the composite AE outcome (Odds Ratio [OR] 1.29, 95% CI 1.02-1.64) but not LOS. In multivariate analysis, after adjustment for relevant covariates, 60-minute TTA intervals were associated with the composite AE outcome (61-120 minutes vs.  $\leq 60$  minutes, OR 1.81, 95% CI 1.01-3.26). Unexpectedly, admission from the emergency department (ED) was also independently associated with the composite AE outcome (ED vs. clinic, OR 3.15, 95% CI 1.95-5.09). **CONCLUSIONS:** TTA and presentation to the ED are independently associated with poor outcomes of FN.

## Gastrostomy feeding versus oral feeding alone for children with cerebral palsy

Gantasala, S., et al.

*Cochrane Database Syst Rev* 2013; 7: CD003943

**BACKGROUND:** Children with cerebral palsy can be significantly disabled in terms of their ability to suck, chew and swallow. This can lead to significant impairment in feeding and, eventually, to undernutrition. It can also result in aspiration of food into the lungs. Length of feeding time may be considerably increased and, instead of being an enjoyable experience, mealtimes may be distressing for both child and carer. For children unable to maintain a normal nutritional state feeding by mouth, gastrostomy or jejunostomy tubes are increasingly being used to provide the digestive system with nutrients. A gastrostomy tube is a feeding tube inserted surgically through the abdominal wall directly into the stomach. A jejunostomy feeding tube is inserted into the jejunum, part of the small intestine, either directly or via a previous gastrostomy. Although gastrostomy or jejunostomy placement may greatly facilitate the feeding of children with cerebral palsy, many carers find it very emotionally difficult to accept this intervention. Moreover, the intervention is costly and there is the possibility of complications. The effectiveness and safety of the treatment requires further assessment. This review is an update of one previously published in 2004. **OBJECTIVES:** To assess the effects of nutritional supplementation given via gastrostomy or jejunostomy to children with feeding difficulties due to cerebral palsy. **SEARCH METHODS:** For this update, we searched the following databases in July 2012: CENTRAL, MEDLINE, Embase, CINAHL, Science Citation Index, Conference Proceedings Citation Index, LILACS and Zetoc. We searched for trials in ICTRP and Clinicaltrials.gov, and for theses in WorldCat and Proquest Index to Theses. We also contacted other researchers and experts in this field. **SELECTION CRITERIA:** We looked for randomised controlled trials that compared delivery of nutrition via a gastrostomy or jejunostomy tube compared with oral feeding alone for children up to the age of 16 years. **DATA COLLECTION AND ANALYSIS:** Screening of search results was undertaken independently by two review authors. No data extraction was possible as there were no included studies. **MAIN RESULTS:** No trials were identified that met the inclusion criteria for this review. **AUTHORS' CONCLUSIONS:** Considerable uncertainty about the effects of gastrostomy for children with cerebral palsy remains. A well designed and conducted randomised controlled trial should be undertaken to resolve the current uncertainties about medical management for children with cerebral palsy and physical difficulties in eating.

<http://www.ncbi.nlm.nih.gov/pubmed/23900969>

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## Cost-effectiveness analysis of antiepileptic drugs in the treatment of Lennox-Gastaut syndrome

Clements, K. M., et al.

*Epilepsy Behav* 2013; 29(1): 184-189

An economic model evaluated the costs and outcomes of adjunctive clobazam therapy for Lennox-Gastaut syndrome (LGS) compared with adjunctive lamotrigine, rufinamide, and topiramate. Clinical data were used to estimate baseline frequency and the percentage of drop-seizure reductions over 3 months (all comparators) and 2 years (rufinamide). Claims data from a large US health care plan were employed to estimate costs. After 3 months, 21.5% of those receiving clobazam were drop-seizure-free. Over a 3-month horizon, clobazam was more effective and less expensive than comparators, with the assumption that >0.77% of drop seizures required medical care. Below this threshold, topiramate was less costly than clobazam. With the base-case assumption that 2.3% of drop seizures were medically attended, costs for patients receiving clobazam totaled \$30,147 versus \$34,223-\$35,378 for comparators. Clobazam was more efficacious and less costly than rufinamide over a 2-year horizon. The percentage of medically attended drop seizures was a driver of results. Clobazam treatment may be cost-saving.

<http://www.ncbi.nlm.nih.gov/pubmed/23973644>

## Central venous catheters and catheter locks in children with cancer: A prospective randomized trial of taurolidine versus heparin

Handrup, M. M., et al.

*Pediatr Blood Cancer* 2013; 60(8): 1292-1298

**BACKGROUND:** To determine if the catheter lock taurolidine can reduce the number of catheter-related bloodstream infections (CRBSI) in pediatric cancer patients with tunneled central venous catheters (CVC). **PROCEDURE:** During a study period of 34 months, 129 newly placed tunneled CVCs in 112 patients were randomly assigned to standard lock with heparin solution or experimental lock with a taurolidine solution (ClinicalTrials.gov Identifier NCT00735813). **RESULTS:** Sixty-five CVCs were included in the standard group and 64 CVCs in the experimental group. The groups were comparable regarding patients' characteristics. A total number of 72 bloodstream infections of which 33 were CRBSIs were observed during 39,127 CVC-days. A lower rate of CRBSI (0.4 per 1,000 CVC-days) was observed in the experimental arm compared with the standard arm (1.4 per 1,000 CVC-days, incidence rate ratio (IRR) = 0.26; 95% confidence interval (CI) 0.09-0.61; P = 0.001). A lower rate of total bloodstream infections (1.2 per 1,000 CVC-days) was also observed in the experimental arm compared with the standard arm (2.5 per 1,000 CVC-days, IRR = 0.49; 95% CI 0.29-0.82; P = 0.004). Median interval from catheter insertion until first CRBSI was significantly lower in the standard group (156 days, range 12-602) compared with the experimental group (300 days, range 12-1,176; P = 0.02). Premature removal of the CVC due to infection and overall CVC survival were similar in the two study groups. **CONCLUSION:** Locking of long-term tunneled CVC with taurolidine significantly reduces catheter-related bloodstream infections in children with cancer.

<http://www.ncbi.nlm.nih.gov/pubmed/23417891>

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## Motor function measure: Validation of a short form for young children with neuromuscular diseases

de Lattre, C., et al.

*Arch Phys Med Rehabil* 94(11): 2218-2226

**OBJECTIVE:** To validate a useful version of the Motor Function Measure (MFM) in children with neuromuscular diseases aged <7 years old. **DESIGN:** Two prospective cohort studies that documented the MFM completion of children aged between 2 and 7 years old. **SETTING:** French-speaking rehabilitation departments from France, Belgium, and Switzerland. **PARTICIPANTS:** Healthy children (n=194) and children with a neuromuscular disease (n=88). **INTERVENTIONS:** Patients were rated by the MFM either once or twice by trained medical professionals, with a delay between the 2 MFMs ranging between 8 and 30 days. **MAIN OUTCOME MEASURE:** Intra- and interrater reliability of the MFM. **RESULTS:** The subtests making up the MFM-32, a scale monitoring severity and progression of motor function in patients with a neuromuscular disease in 3 functional domains, were carried out in healthy children aged 2 to 7 years. Twenty items of the MFM-32 were successfully completed by these children and were used to constitute the MFM-20. Principal component analysis of the MFM-20 confirmed the 3 functional domains. Inter- and intrarater reliability of the 3 subscores and total score were high (intraclass correlation coefficient >.90), and discriminant validity was good. **CONCLUSIONS:** The MFM-20 can be used as an outcome measure for assessment of motor function in young children with neuromuscular disease.

## Music to reduce pain and distress in the pediatric emergency department: A randomized clinical trial

Hartling, L., et al.

*JAMA Pediatr* 2013; 167(9): 826-835

**IMPORTANCE:** Many medical procedures aimed at helping children cause them pain and distress, which can have long-lasting negative effects. Music is a form of distraction that may alleviate some of the pain and distress experienced by children while undergoing medical procedures. **OBJECTIVE:** To compare music with standard care to manage pain and distress. **DESIGN, SETTING, AND PARTICIPANTS:** Randomized clinical trial conducted in a pediatric emergency department with appropriate sequence generation and adequate allocation concealment from January 1, 2009, to March 31, 2010. Individuals assessing the primary outcome were blind to treatment allocation. A total of 42 children aged 3 to 11 years undergoing intravenous placement were included. **INTERVENTIONS:** Music (recordings selected by a music therapist via ambient speakers) vs standard care. **MAIN OUTCOMES AND MEASURES:** The primary outcome was behavioral distress assessed blinded using the Observational Scale of Behavioral Distress-Revised. The secondary outcomes included child-reported pain, heart rate, parent and health care provider satisfaction, ease of performing the procedure, and parental anxiety. **RESULTS:** With or without controlling for potential confounders, we found no significant difference in the change in behavioral distress from before the procedure to immediately after the procedure. When children who had no distress during the procedure were removed from the analysis, there was a significantly less increase in distress for the music group (standard care group = 2.2 vs music group = 1.1,  $P < .05$ ). Pain scores among children in the standard care group increased by 2 points, while they remained the same in the music group ( $P = .04$ ); the difference was considered clinically important. The pattern of parent satisfaction with the management of children's pain was different between groups, although not statistically significant ( $P = .07$ ). Health care providers reported that it was easier to perform the procedure for children in the music group (76% very easy) vs the standard care group (38% very easy) ( $P = .03$ ). Health care providers were more satisfied with the intravenous placement in the music group (86% very satisfied) compared with the standard care group (48%) ( $P = .02$ ). **CONCLUSIONS AND RELEVANCE:** Music may have a positive impact on pain and distress for children undergoing intravenous placement. Benefits were also observed for the parents and health care providers. **TRIAL REGISTRATION:** clinicaltrials.gov Identifier: NCT00761033.

<http://www.ncbi.nlm.nih.gov/pubmed/23857075>

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## Outcome of children and adolescents with a second or third relapse of acute lymphoblastic leukemia (ALL): A population-based analysis of the Austrian ALL-BFM (Berlin-Frankfurt-Munster) study group

Reismuller, B., et al.

*J Pediatr Hematol Oncol* 2013; 35(5): e200-204

We analyzed outcome of a population-based cohort of 74 children with second and third acute lymphoblastic leukemia (ALL) relapse and aimed to identify prognostic factors. Duration of previous remission and site of relapse appeared of prognostic relevance as patients with a second remission duration >1.5 years and isolated extramedullary relapse did better. Neither patient with a second bone marrow relapse who underwent previous allogeneic transplantation nor patients with T-cell ALL survived. Overall, 7 of 74 (9%) patients are in long-term remission. Stem cell transplantation seemed to be the only curative option for systemic relapse of B-cell precursor ALL as all 4 surviving patients with a second/third relapse involving the bone marrow received a transplant. Conclusively, patients with a second ALL relapse are ideal candidates for phase I/II trials exploring new innovative drugs.

<http://www.ncbi.nlm.nih.gov/pubmed/23652878>

## High-risk childhood acute lymphoblastic leukemia in first remission treated with novel intensive chemotherapy and allogeneic transplantation

Marshall, G. M., et al.

*Leukemia* 2013; 27(7): 1497-1503

Children with acute lymphoblastic leukemia (ALL) and high minimal residual disease (MRD) levels after initial chemotherapy have a poor clinical outcome. In this prospective, single arm, Phase 2 trial, 111 Dutch and Australian children aged 1-18 years with newly diagnosed, t(9;22)-negative ALL, were identified among 1041 consecutively enrolled patients as high risk (HR) based on clinical features or high MRD. The HR cohort received the AIEOP-BFM (Associazione Italiana di Ematologia ed Oncologia Pediatrica (Italy)-Berlin-Frankfurt-Munster ALL Study Group) 2000 ALL Protocol I, then three novel HR chemotherapy blocks, followed by allogeneic transplant or chemotherapy. Of the 111 HR patients, 91 began HR treatment blocks, while 79 completed the protocol. There were 3 remission failures, 12 relapses, 7 toxic deaths in remission and 10 patients who changed protocol due to toxicity or clinician/parent preference. For the 111 HR patients, 5-year event-free survival (EFS) was 66.8% (+/-5.5) and overall survival (OS) was 75.6% (+/-4.3). The 30 patients treated as HR solely on the basis of high MRD levels had a 5-year EFS of 63% (+/-9.4%). All patients experienced grade 3 or 4 toxicities during HR block therapy. Although cure rates were improved compared with previous studies, high treatment toxicity suggested that novel agents are needed to achieve further improvement.

<http://www.ncbi.nlm.nih.gov/pubmed/23407458>

## Efficacy of vagus nerve stimulation in brain tumor-associated intractable epilepsy and the importance of tumor stability

Patel, K. S., et al.

*J Neurosurg* 2013; 119(2): 520-525

OBJECT: Vagus nerve stimulation (VNS) is a viable option for patients with medically intractable epilepsy. However, there are no studies examining its effect on individuals with brain tumor-associated intractable epilepsy. This study aims to evaluate the efficacy of VNS in patients with brain tumor-associated medically intractable epilepsy. METHODS: Epilepsy surgery databases at 2 separate epilepsy centers were reviewed to identify patients in whom a VNS device was placed for tumor-related intractable epilepsy between January 1999 and December 2011. Preoperative and postoperative seizure frequency and type as well as antiepileptic drug (AED) regimens and degree of tumor progression were evaluated. Statistical analysis was performed using odds ratios and t-tests to examine efficacy. RESULTS: Sixteen patients were included in the study. Eight patients (50%) had an improved outcome (Engel Class I, II, or III) with an average follow-up of 39.6 months. The mean reduction in seizure frequency was 41.7% ( $p = 0.002$ ). There was no significant change in AED regimens. Seizure frequency decreased by 10.9% in patients with progressing tumors and by 65.6% in patients with stable tumors ( $p = 0.008$ ). CONCLUSIONS: Vagus nerve stimulation therapy in individuals with brain tumor-associated medically intractable epilepsy was shown to be comparably effective in regard to seizure reduction and response rates to the general population of VNS therapy patients. Outcomes were better in patients with stable as opposed to progressing tumors. The authors' findings support the recommendation of VNS therapy in patients with brain tumor-associated intractable epilepsy, especially in cases in which imminent tumor progression is not expected. Vagus nerve stimulation may not be indicated in more malignant tumors.

<http://www.ncbi.nlm.nih.gov/pubmed/23600931>

## Development and validation of the distress thermometer for parents of a chronically ill child

Haverman, L., et al.

*J Pediatr* 2013; 163(4): 1140-1146 e1142

**OBJECTIVE:** To develop and validate a Distress Thermometer for Parents (DT-P) for chronically ill children and to determine a cutoff score for clinical distress. **STUDY DESIGN:** Parents of a chronically ill child (0-18 years) were recruited via announcements or were actively approached at the outpatient clinics of the Emma Children's Hospital/Academic Medical Center and Vrije Universiteit Medical Center. We modeled the development of the DT-P on the Distress Thermometer used in oncology medical care. The DT-P consists of a thermometer score from 0 (no distress) to 10 (extreme distress) and a problem list (practical, social, emotional, physical, cognitive, and parenting domains). The DT-P was validated with the Hospital Anxiety and Depression Scale (HADS) and the Parenting Stress Index. **RESULTS:** The mean thermometer score of the 706 participating parents was 3.7 (SD 3.0). The thermometer score and the scores in the practical, emotional, physical, and cognitive problem domains were strongly related to anxiety, depression, and the total score of the HADS ( $0.55 \leq r \leq 0.72$ ). The thermometer score and all problem domain scores were moderately-to-strongly related to the Parenting Stress Index ( $0.38 \leq r \leq 0.63$ ). A cutoff-score of 4 correctly identified 86% of clinical HADS cases" (sensitivity) and 67% of "nonclinical HADS cases" (specificity). **CONCLUSIONS:** We developed the DT-P and examined its diagnostic utility in a large sample. The DT-P appeared to be a valid and useful short screening-tool for identifying parental distress.

<http://www.ncbi.nlm.nih.gov/pubmed/23910979>

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## Deficits in physical function among young childhood cancer survivors

Hoffman, M. C., et al.

*J Clin Oncol* 2013; 31(22): 2799-2805

**PURPOSE:** Childhood cancer survivors (CCSs) are at risk for physical disability. The aim of this investigation was to characterize and compare physical performance among CCSs and a group of siblings age < 18 years and determine if diagnosis, treatment, and physical activity levels were associated with lower performance scores. **METHODS:** CCSs  $\geq 5$  years from diagnosis and a sibling comparison group were recruited and evaluated for strength, mobility, and fitness. Physical performance measures were compared in regression models between survivors and siblings by diagnosis and among survivors by treatment exposures and physical activity levels. **RESULTS:** CCSs (n = 183; mean age +/- standard deviation [SD], 13.5 +/- 2.5 years; 53% male) scored lower than siblings (n = 147; mean age +/- SD, 13.4 +/- 2.4 years; 50% male) on lower-extremity strength testing, the timed up-and-go (TUG) test, and the 6-minute walk (6MW) test, despite reporting similar levels and types of habitual physical activity. The lowest scores were prevalent among survivors of CNS tumors and bone and soft tissue sarcomas on strength testing (score +/- SD: CNS tumors, 76.5 +/- 4.7; sarcoma 67.1 +/- 7.2 v siblings, 87.3 +/- 2.4 Newton-meters quadriceps strength at 90 degrees per second; P = .04 and .01, respectively) and among CNS tumor survivors on the TUG (score +/- SD: 5.1 +/- 0.1 v siblings, 4.4 +/- 0.1 seconds; P < .001) and 6MW tests (score +/- SD: 533.3 +/- 15.6 v siblings, 594.1 +/- 8.3 m; P < .001). **CONCLUSION:** CCSs may have underlying physiologic deficits that interfere with function that cannot be completely overcome by participation in regular physical activity. These survivors may need referral for specialized exercise interventions in addition to usual counseling to remain physically active.

<http://www.ncbi.nlm.nih.gov/pubmed/23796992>



## Outcome of heart transplantation in pediatric cancer survivors

Shah, N., et al.

*Pediatr Transplant* 2013; 17(5): 423-428

The aim of this study is to evaluate the outcome of heart transplantation in children surviving malignancies. Pediatric heart transplant recipients were identified using the UNOS database. Follow-up data including survival and rate of malignancy were analyzed. A total of 7169 children received heart transplants between 1987 and 2011. Of these, 107 (1.5%) survived previous malignancy treatment (group I) and 7062 (98.5%) did not have prior malignancy (group II). Survival after transplant was 92.5%, 90.6%, 80.3%, and 65% at three months, one, five, and 10 yr in group I, similar to the rate in group II (90.1%, 84.4%, 73.8%, and 57.7%). Survival after transplantation was similar between group I and children who underwent OHT secondary to cardiomyopathy in group II. The rate of post-OHT malignancy in group I was higher than that in group II (14/107(13%) vs. 386/7062 (5.4%),  $p = 0.001$ ). Children who developed malignancy in group I had similar survival as children who developed malignancy in group II. Post-transplant survival is similar in children with and without pretransplant malignancy in spite of higher rate of malignancy in children with pretransplant malignancy. OHT appears to be a reasonable treatment option in children who develop end-stage heart disease after malignancy treatment.

<http://www.ncbi.nlm.nih.gov/pubmed/23527878>

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## Home monitoring program reduces interstage mortality after the modified Norwood procedure

Siehr, S. L., et al.

*J Thorac Cardiovasc Surg* 2014; 147(2): 718-723 e711

BACKGROUND: From 2002 to 2005, the interstage mortality after a modified Norwood procedure was 7% in our program. An interstage home monitoring program (HMP) was established to identify Norwood procedure patients at increased risk of decompensation and to reduce interstage mortality. METHODS: Results of the first 5 years of the Norwood HMP were reviewed retrospectively. Interstage was defined as the time between Norwood hospital discharge and admission for second stage surgical palliation. In the HMP, families documented oxygen saturation, heart rate, weight, and feedings daily. Nurse practitioners called each family at least weekly, and when issues arose, action plans were determined based on symptom severity. RESULTS: Between October 2005 and October 2010 there were 46 Norwood procedure patients who survived to hospital discharge. All were enrolled in the HMP. Forty-five patients had a Norwood procedure with right ventricle to pulmonary artery conduit, and 1 patient had a modified Blalock-Taussig shunt. Interstage survival was 100%. Nineteen patients (41%) were admitted interstage; 5 patients were admitted twice, 1 patient was admitted 4 times. Seventeen patients (37%) required interstage interventions. Eight patients (17%) required major interventions: conduit stenting, aortic arch balloon angioplasty, emergent shunt, or early Glenn surgery. Minor interventions included supplemental oxygen, blood transfusion, intravenous hydration, diuresis, anti-arrhythmic therapy, or feeding adjustments. CONCLUSIONS: In the first 5 years of the HMP, all infants discharged after a modified Norwood procedure survived the interstage period. The HMP altered clinical management in 37% of patients. Home monitoring of oxygen saturation, heart rate, weight, and feedings, along with comprehensive care coordination, allowed timely interventions and reduced interstage mortality from 7% to 0%.

<http://www.ncbi.nlm.nih.gov/pubmed/23663957>

## A decision analysis tool for the assessment of posterior fossa tumour surgery outcomes in children: The Liverpool Neurosurgical Complication Causality Assessment Tool

Zakaria, R., et al.

*Childs Nerv Syst* 2013; 29(8): 1277-1283

**INTRODUCTION:** Complications may occur following posterior fossa tumour surgery in children. Such complications are subjectively and inconsistently reported even though they may have significant long-term behavioural and cognitive consequences for the child. This makes comparison of surgeons, programmes and treatments problematic. **MATERIALS AND METHODS:** We have devised a causality tool for assessing if an adverse event after surgery can be classified as a surgical complication using a series of simple questions, based on a tool used in assessing adverse drug reactions. This tool, which we have called the "Liverpool Neurosurgical Complication Causality Assessment Tool", was developed by reviewing a series of ten posterior fossa tumour cases with a panel of neurosurgery, neurology, oncology and neuropsychology specialists working in a multidisciplinary paediatric tumour treatment programme. **DISCUSSION AND CONCLUSION:** We have demonstrated its use and hope that it may improve reliability between different assessors both in evaluating the outcomes of existing programmes and treatments as well as aiding in trials which may directly compare the effects of surgical and medical treatments.

<http://www.ncbi.nlm.nih.gov/pubmed/23494657>

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## Late morbidity leading to hospitalization among 5-year survivors of young adult cancer: A report of the childhood, adolescent and young adult cancer survivors research program

Zhang, Y., et al.

*Int J Cancer* 2014; 134(5): 1174-1182

To estimate the risk of late morbidity leading to hospitalization among young adult cancer 5-year survivors compared to the general population and to examine the long-term effects of demographic and disease-related factors on late morbidity, a retrospective cohort of 902 five-year survivors of young adult cancer diagnosed between 1981 and 1999 was identified from British Columbia (BC) Cancer Registry. A matched comparison group (N = 9020) was randomly selected from the provincial health insurance plan. All hospitalizations until the end of 2006 were determined from the BC health insurance plan hospitalization records. The Poisson regression model was used to estimate the rate ratios for late morbidity leading to hospitalization except pregnancy after adjusting for sociodemographic and clinical risk factors. Overall, 455 (50.4%) survivors and 3,419 (37.9%) individuals in the comparison group had at least one type of late morbidity leading to hospitalization. The adjusted risk of this morbidity for survivors was 1.4 times higher than for the comparison group (95% CI = 1.22-1.54). The highest risks were found for hospitalization due to blood disease (RR = 4.2; 95% CI = 1.98-8.78) and neoplasm (RR = 4.3; 95% CI = 3.41-5.33). Survivors with three treatment modalities had three-fold higher risk of having any type of late morbidity (RR = 3.22; 95% CI = 2.09-4.94) than the comparators. These findings emphasize that young adult cancer survivors still have high risks of a wide range of late morbidities.

<http://www.ncbi.nlm.nih.gov/pubmed/24037993>

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## Initial development of the Symptom Screening in Pediatrics Tool (SSPedi)

Tomlinson, D., et al.

*Support Care Cancer* 2014; 22(1): 71-75

**BACKGROUND:** We previously identified published scales for symptom assessment in pediatric cancer patients. The objectives of this study were to identify if any of these scales were suitable for use or adaptation as a self-report symptom screening tool, and if not, to begin the process of creating a new tool. **METHODS:** A focus group of ten healthcare professionals with expertise in pediatric cancer symptom management and a patient advocate were convened. First, the group identified the optimal properties of a symptom screening tool for pediatric cancer patients. Next, the previously identified symptom assessment scales were evaluated against these properties. As none of the existing scales were adequate for symptom screening, a nominal group technique was used to identify the most important symptoms for inclusion in a new symptom screening tool. **RESULTS:** Optimal properties of a symptom screening tool included minimal respondent burden, inclusion of 15 items or less, and inclusion of the most burdensome symptoms. None of the previously identified scales were adequate because they lacked content validity and were too long or would be too hard for children to understand. Nominal group technique identified 15 items to be included; an initial draft was developed and named the Symptom Screening in Pediatrics (SSPedi) Tool. **CONCLUSIONS:** This study identified the lack of an appropriate symptom screening tool for use by pediatric cancer patients. A preliminary version of SSPedi was developed. Subsequent work will ensure that it is understandable by children and evaluate its psychometric properties.

<http://www.ncbi.nlm.nih.gov/pubmed/23995756>

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## Outcome following neonatal seizures

Uria-Avellanal, C., et al.

*Semin Fetal Neonatal Med* 2013; 18(4): 224-232

Neonatal seizures are the most common manifestation of neurological disorders in the newborn period and an important determinant of outcome. Overall, for babies born at full term, mortality following seizures has improved in the last decade, typical current mortality rates being 10% (range: 7-16%), down from 33% in reports from the 1990s. By contrast, the prevalence of adverse neurodevelopmental sequelae remains relatively stable, typically 46% (range: 27-55%). The strongest predictors of outcome are the underlying cause, together with the background electroencephalographic activity. In preterm babies, for whom the outlook tends to be worse as background mortality and disability are high, seizures are frequently associated with serious underlying brain injury and therefore subsequent impairments. When attempting to define the prognosis for a baby with neonatal seizures, we propose a pathway involving history, examination, and careful consideration of all available results (ideally including brain magnetic resonance imaging) and the response to treatment before synthesizing the best estimate of risk to be conveyed to the family.

<http://www.ncbi.nlm.nih.gov/pubmed/23466296>

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## Midazolam oral transmucosal route: An alternative to rectal diazepam for some children

*Prescrire Int 2013; 22(140): 173-177*

In children, convulsive seizures lasting more than 5 minutes constitute a life-threatening condition. Outside of the hospital setting, the treatment of choice is rectal diazepam. Midazolam, a fast-acting benzodiazepine, is now authorised for use in this setting, in the form of a solution for oral transmucosal route. The results of five trials conducted in specialised centres suggest that midazolam oromucosal solution is at least as effective as rectal diazepam in reducing the duration of seizures and preventing early seizure recurrence. However, the evidence provided by these studies is weak. The main adverse effects of midazolam oromucosal solution are the same as those of rectal diazepam; in particular, respiratory depression occurs in 1% to 5% of patients and can necessitate intubation. Some anticonvulsants, such as carbamazepine, phenytoin and phenobarbital, may reduce the effectiveness of midazolam and diazepam. At the recommended doses, midazolam overdose may occur when the oromucosal solution is used to treat infants less than 6 months old. Midazolam oromucosal solution is sold in single-dose ready-to-use oral syringes. In France, diazepam doses for rectal administration must be prepared from a vial of solution. In early 2013, the usability of oral transmucosal versus rectal administration has not been compared prospectively in children with convulsive seizures. In practice, rectal diazepam is the drug of choice for paediatric convulsive seizures occurring outside the hospital setting. Midazolam oromucosal solution is an alternative for children over 6 months of age, especially when preparation or administration of the rectal diazepam dose poses a problem.

<http://www.ncbi.nlm.nih.gov/pubmed/23951591>

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## When opioids fail in chronic pain management: The role for buprenorphine and hospitalization

Berland, D. W., et al.

*Am J Ther 2013; 20(4): 316-321*

Clinicians are increasingly being challenged by patients who are treated for chronic pain with high-dose opioids that can cause medical, social, and societal harm. These patients may best be improved by psychological approaches, adjuvant medications, and opioid reduction or removal, rather than ever-escalating dosing that has become common. Opioid reduction or removal can be a difficult process that, when done incorrectly, may cause patient dissatisfaction or severe discomfort. Buprenorphine, a partial opioid agonist, is slowly becoming recognized as an effective pain treatment, possessing a wide safety margin while offering the opportunity for stabilization of opioid dosing or even removal. We have developed a protocol for hospitalization of the most fragile or toxic patients detailed herein that can permit a comfortable conversion to buprenorphine from prior high-dose full agonist opioid therapy. Seventy-six consecutive patients with serious medical, psychological, or addiction comorbidities, treated with morphine equivalent doses exceeding hundreds of milligrams per day, were followed after conversion for up to 25 months. Two-thirds reported moderate to dramatic improvements of pain and functional status with an increase seen in employment. Median length of hospital stay was 2 days, and the median daily buprenorphine discharge dose was 8 mg. No adverse reactions or outcomes were observed. A brief hospitalization for conversion from high-dose opioid therapy to a safer, more effective buprenorphine regimen can produce life-altering improvement.

<http://www.ncbi.nlm.nih.gov/pubmed/23584313>

## Neuropathic pain in patients with sickle cell disease

Brandow, A. M., et al.

*Pediatr Blood Cancer* 2014; 61(3): 512-517

**BACKGROUND:** Despite the suggestion of a neuropathic component to sickle cell disease (SCD) pain, there are minimal data on the systematic assessment of neuropathic pain in patients with SCD. Neuropathic pain is defined as pain primarily initiated by dysfunction of the peripheral or central nervous system. **PROCEDURE:** In a cross-sectional study, we used the painDETECT questionnaire, a one-page validated neuropathic pain screening tool, to determine the presence of neuropathic pain in patients with SCD and to evaluate the relationship between neuropathic pain, age, and gender. We hypothesized that 20% of patients with SCD will experience neuropathic pain and that neuropathic pain will be associated with older age and female gender. The completed painDETECT questionnaire yields a total score between 0 and 38 ( $\geq 19$  = definite neuropathic pain, 13-18 = probable neuropathic pain,  $\leq 12$  = no neuropathic pain). Scores  $\geq 13$  were designated as having evidence of neuropathic pain. **RESULTS:** A total of 56 patients participated. Median age was 20.3 years and 77% were female. We found 37% of patients had evidence of neuropathic pain. Age was positively correlated with total score ( $r = 0.43$ ;  $P = 0.001$ ) suggesting older patients experience more neuropathic pain. Females had higher mean total scores (13 vs. 8.4;  $P = 0.04$ ). Significantly more patients with neuropathic pain were taking hydroxyurea (90% vs. 59%;  $P = 0.015$ ). Despite 37% of patients experiencing neuropathic pain, only 5% were taking a neuropathic pain drug. **CONCLUSIONS:** Neuropathic pain exists in SCD. Valid screening tools can identify patients that would benefit from existing and future neuropathic pain therapies and could determine the impact of these therapies.

<http://www.ncbi.nlm.nih.gov/pubmed/24167104>

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## Inpatient characteristics of the child admitted with chronic pain

Coffelt, T. A., et al.

*Pediatrics* 2013; 132(2): e422-429

**OBJECTIVE:** To define the demographic, diagnostic, procedural, and episode of care characteristics for children admitted with chronic pain. **METHODS:** We used the Pediatric Health Information System database to obtain data on demographic characteristics, length of stay, readmission rates, diagnoses, and procedures for children admitted with chronic pain. Patients with sickle cell disease, cancer, burns, cerebral palsy, transplants, and ventilator-dependent children were excluded. **RESULTS:** A total of 3752 patients with chronic pain were identified from 2004 through 2010. Admissions increased by 831% over this time period. The mean age of these patients was 13.5 years, the most common race was white (79%), and female subjects outnumbered male subjects by 2.41 to 1. The most common admission and principal discharge diagnosis was abdominal pain; comorbid diagnoses were common, with a mean of 10 diagnoses per patient. In total, 65% of patients had a comorbid gastrointestinal diagnosis and 44% had a psychiatric diagnosis. The mean length of stay was 7.32 days, with an expected length of stay of 4.24 days; 12.5% were readmitted at least once within 1 year. They underwent a mean of 3.18 procedures per patient. **CONCLUSIONS:** The average child admitted with chronic pain is a teenaged female with a wide variety of comorbid conditions, many of which are gastrointestinal and psychiatric in nature. Admissions for chronic pain are rising and account for substantial resource utilization. Future studies should further characterize this population, with the overall objective of improving outcomes and optimizing cost-effective care.

<http://www.ncbi.nlm.nih.gov/pubmed/23821701>

## Are we failing to provide adequate rescue medication to children at risk of prolonged convulsive seizures in schools?

Cross, J. H., et al.

*Arch Dis Child* 2013; 98(10): 777-780

**OBJECTIVE:** This paper explores the issues that arise from the discussion of administering rescue medication to children who experience prolonged convulsive seizures in mainstream schools in the UK. **SITUATION ANALYSIS:** Current guidelines recommend immediate treatment of children with such seizures (defined as seizures lasting more than 5 min) to prevent progression to status epilepticus and neurological morbidity. As children are unconscious during prolonged convulsive seizures, whether or not they receive their treatment in time depends on the presence of a teacher or other member of staff trained and able to administer rescue medication. However, it is thought that the situation varies between schools and depends mainly on the goodwill and resources available locally. **RECOMMENDATIONS:** A more systematic response is needed to ensure that children receive rescue medication regardless of where their seizure occurs. Possible ways forward include: greater use of training resources for schools available from epilepsy voluntary sector organisations; consistent, practical information to schools; transparent guidance outlining a clear care pathway from the hospital to the school; and implementation and adherence to each child's individual healthcare plan. **IMPLICATIONS:** Children requiring emergency treatment for prolonged convulsive seizures during school hours test the goals of integrated, person-centred care as well as joined-up working to which the National Health Service (NHS) aspires. As changes to the NHS come into play and local services become reconfigured, every effort should be made to take account of the particular needs of this vulnerable group of children within broader efforts to improve the quality of paediatric epilepsy services overall.

<http://www.ncbi.nlm.nih.gov/pubmed/23899921>

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## Best BETs from the Manchester Royal Infirmary: BET 1: intranasal lorazepam is an acceptable alternative to intravenous lorazepam in the control of acute seizures in children

Allan, A. and J. Cullen

*Emerg Med* 2013; J 30(9): 768-769

A short-cut review was carried out to determine whether intranasal lorazepam was as effective as intravenous lorazepam in the control of seizures in children. Eighteen papers were found using the reported search, of which one was directly relevant and another compared intranasal lorazepam with intramuscular paraldehyde. The author, date and country of publication, patient group studied, study type, relevant outcomes, results and study weaknesses are shown in table 1. It is concluded that intranasal lorazepam appears to be a safe and effective treatment for this condition.

## Pediatric seizures

Agarwal, M. and S. M. Fox

*Emerg Med Clin North Am* 2013; 31(3): 733-754

Seizures are a commonly encountered condition within the emergency department and, because of this, can engender complacency on the part of the physicians and staff. Unfortunately, there is significant associated morbidity and mortality with seizures, and they should never be regarded as routine. This point is particularly important with respect to seizures in pediatric patients. The aim of this review is to provide a current view of the various issues that make pediatric seizures unique and to help elucidate emergent evaluation and management strategies.

<http://www.ncbi.nlm.nih.gov/pubmed/23915601>

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## Intravenous levetiracetam for treatment of neonatal seizures

Rakshasbhuvankar, A., et al.

*J Clin Neurosci* 2013; 20(8): 1165-1167

In this case series we report on eight neonates with refractory seizures who received intravenous levetiracetam when seizures did not respond to two or more conventional anticonvulsants. Six of the eight neonates had an excellent response with either cessation, or reduction in seizures by at least 80%. One neonate showed a partial response while one did not have any reduction in seizure frequency. We did not encounter any adverse effects that could be attributable to levetiracetam.

<http://www.ncbi.nlm.nih.gov/pubmed/23664133>

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## Intranasal fentanyl in the palliative care of newborns and infants

Harlos, M. S., et al.

*J Pain Symptom Manage* 2013; 46(2): 265-274

CONTEXT: Perinatal palliative care is an area of increasing focus among clinicians supporting newborns and their families. Although not every newborn will survive the neonatal period, assuring their comfort and quality of life remains an imperative for their care providers. It can be challenging to administer medications such as opioids in a minimally invasive yet effective manner. OBJECTIVES: To describe the experience using intranasal (IN) fentanyl in the management of distress in a case series of 11 dying neonates. METHODS: A retrospective chart review was undertaken of 58 consecutive referrals of newborns and infants aged six months or younger between November 2006 and July 2010 to the Winnipeg Regional Health Authority Pediatric Palliative Care Service to determine how often IN fentanyl was used and review documented responses after the medication. RESULTS: Of 58 referrals, IN fentanyl was used in 11 patients, in all cases for concerns regarding respiratory distress. Chart documentation indicated that fentanyl was tolerated well, with no circumstances of drug-related apnea and no occurrences of chest wall rigidity. In most cases, labored breathing and restlessness settled after medication administration. The average time from administration of the last dose of fentanyl until death was 61 minutes. CONCLUSION: We found IN fentanyl, which can be administered in a variety of care settings, to be a minimally invasive means of palliating distress in dying newborns and infants. No adverse events related to its use were noted.

<http://www.ncbi.nlm.nih.gov/pubmed/23017621>

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## Successful implementation of a neonatal pain and sedation protocol at 2 NICUs

Deindl, P., et al.

*Pediatrics* 2013; 132(1): e211-218

**OBJECTIVE:** To evaluate the implementation of a neonatal pain and sedation protocol at 2 ICUs. **METHODS:** The intervention started with the evaluation of local practice, problems, and staff satisfaction. We then developed and implemented the Vienna Protocol for Neonatal Pain and Sedation. The protocol included well-defined strategies for both nonpharmacologic and pharmacologic interventions based on regular assessment of a translated version of the Neonatal Pain Agitation and Sedation Scale and titration of analgesic and sedative therapy according to aim scores. Health care staff was trained in the assessment by using a video-based tutorial and bedside teaching. In addition, we performed reevaluation, retraining, and random quality checks. Frequency and quality of assessments, pharmacologic therapy, duration of mechanical ventilation, and outcome were compared between baseline (12 months before implementation) and 12 months after implementation. **RESULTS:** Cumulative median (interquartile range) opiate dose (baseline dose of 1.4 [0.5-5.9] mg/kg versus intervention group dose of 2.7 [0.4-57] mg/kg morphine equivalents;  $P = .002$ ), pharmacologic interventions per episode of continuous sedation/analgesia (4 [2-10] vs 6 [2-13];  $P = .005$ ), and overall staff satisfaction (physicians: 31% vs 89%;  $P < .001$ ; nurses: 17% vs 55%;  $P < .001$ ) increased after implementation. Time on mechanical ventilation, length of stay at the ICU, and adverse outcomes were similar before and after implementation. **CONCLUSIONS:** Implementation of a neonatal pain and sedation protocol at 2 ICUs resulted in an increase in opiate prescription, pharmacologic interventions, and staff satisfaction without affecting time on mechanical ventilation, length of intensive care stay, and adverse outcomes.

<http://www.ncbi.nlm.nih.gov/pubmed/23733799>

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## Methotrimeprazine for the management of end-of-life symptoms in infants and children

Hohl, C. M., et al.

*J Palliat Care* 2013; 29(3): 178-185

**OBJECTIVE:** This retrospective chart review assessed the efficacy, dose, and safety of methotrimeprazine in palliating end-of-life symptoms in children and infants. **METHODS:** A retrospective chart review was conducted of 18 hospitalized pediatric patients who were treated with methotrimeprazine in their last two weeks of life. Data collected included age, diagnosis, symptoms, methotrimeprazine dose, route, efficacy, and any documented adverse effects. **RESULTS:** Patients' ages ranged from 16 days to 17 years. Underlying conditions included malignancies, trauma, and various neurodegenerative and congenital diseases. All patients ( $n = 18$ ) were treated for symptoms of agitation, delirium, or restlessness. Most patients also experienced respiratory secretions/congestion ( $n = 15$ ), pain ( $n = 13$ ), and/or dyspnea ( $n = 9$ ). Less common symptoms included nausea/emesis ( $n = 5$ ) and spasticity ( $n = 1$ ). Methotrimeprazine dosages ranged from 0.02 mg/kg/dose to 0.5 mg/kg/dose. Routes of administration included intravenous ( $n = 13$ ), oral/gastrostomy tube ( $n = 6$ ), or subcutaneous ( $n = 4$ ). Sedation ( $n = 6$ ) was the only documented adverse effect, although when agitation was present, this was potentially an intended and perceived-to-be-beneficial effect. **CONCLUSION:** Methotrimeprazine, an old drug with diverse receptor activity and multiple routes of administration, appears to be an effective tool in treating complicated end-of-life symptoms in children and infants. This study provides a foundation for analysis with prospective and comparative trials, which may further quantify its benefit.

<http://www.ncbi.nlm.nih.gov/pubmed/24380217>



## Bioequivalent antiepileptic drug switching and the risk of seizure-related events

Hansen, R. N., et al.

*Epilepsy Res* 2013; 106(1-2): 237-243

**BACKGROUND:** Older antiepileptic drugs (AEDs) are known to have a narrow therapeutic index. As a consequence, switching between bioequivalent AEDs remains controversial in the management of epilepsy. We investigated the association between A-rated switching of each class of currently available AED and emergent treatment for a seizure-related event. **METHODS:** We used a case-control method and claims data from the 2010 to 2011 Truven Health MarketScan ((R)) Commercial Claims Database to estimate the risk of seizure following a medication switch. Cases and controls with an epilepsy diagnosis were identified by emergency/inpatient or outpatient visit claims, respectively. Cases and controls (N=9110) were matched 1:1 by age, epilepsy diagnosis category and seizure medication. The exposure was defined as a switch between A-rated AEDs during the 90 days prior to index date. Conditional logistic regression was used to estimate the association, adjusting for gender, baseline Deyo-Charlson Comorbidity Index (0, 1, 2, or 3+), region (Northeast, Central, South, and West), and total AED medications. **RESULTS:** A switch between A-rated AEDs occurred in 1053 (23.2%) cases and 827 (18.1%) matched controls. The unadjusted and adjusted odds ratios of a seizure-related event for switching were 1.38 (95% CI: 1.25-1.52) and 1.27 (95% CI: 1.14-1.41), respectively. The independent risk of an event also increased with each category increase in the Charlson score (CCI=1: 1.17, 95% CI: 1.02-1.33; CCI=2: 1.33, 95% CI: 1.09-1.62; CCI=3+: 1.99, 95% CI: 1.64-2.41). Older AEDs had infrequent switches compared to newer agents and were not associated with events. **DISCUSSION:** We found a modest association between AED switching and seizure-related events. Our analysis suggests that the behavior of switching alone may lead to seizure-related events regardless of the medication or type of switch. Other disease or environmental characteristics may contribute to this association. Based on these and other findings, health care professionals and patients should be cautious about switching bioequivalent AEDs.

<http://www.ncbi.nlm.nih.gov/pubmed/23726541>

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## Neuropathic pain in children

Howard, R. F., et al.

*Arch Dis Child* 2014; 99(1): 84-89

Neuropathic pain (NP), due to a lesion or disease of the somatosensory nervous system, is not well documented or researched in children. NP is a clinical diagnosis that can be difficult, especially in younger children. Nevertheless, it is important to recognise NP, as pain mechanisms and consequently management and prognosis differ from other types of long-term pain. NP is common in adult pain clinics but many of the underlying disease states in which it occurs are infrequently or never encountered in paediatric practice. However, NP in childhood has been reported, even in the very young in certain clinical situations. Causes of NP include traumatic injury, complex regional pain syndrome type II, cancer and chemotherapy, chronic infection, neurological and metabolic disease, and inherited sensory nerve dysfunction. The clinical and laboratory study of traumatic peripheral nerve injury has revealed important age-related differences in clinical presentation and prognosis. It is clear that mechanisms operating during development can profoundly modify the consequences of nerve damage and NP. Clinically, diagnosis, assessment and treatment of NP are based on methods and evidence derived from data in adults. Improvements in the understanding and management of NP are likely to come from developmentally appropriate improvements in the clarity and consistency of diagnosis and systematic, well-researched approaches to treatment.

<http://www.ncbi.nlm.nih.gov/pubmed/24114955>

## Phenobarbitone versus phenytoin for treatment of neonatal seizures: An open-label randomized controlled trial

Pathak, G., et al.

*Indian Pediatr* 2013; 50(8): 753-757

**OBJECTIVE:** To compare the efficacy of phenobarbitone and phenytoin for treatment of neonatal seizures in term and near-term neonates. **DESIGN:** Open labeled randomized controlled trial. **SETTING:** Neonatal intensive care unit of a level II unit from India, from November 2008 to September 2009. **PARTICIPANTS:** All term and late pre-term neonates admitted with clinically apparent seizures and not having any transient metabolic disorders (hypoglycemia or hypocalcemia) were randomly assigned. **INTERVENTION:** Phenobarbitone (n=54) or phenytoin (n=55) intravenously 20 mg/kg/dose over 20-30 min. Neonates whose seizures were not controlled by the assigned drug were then crossed over to be treated with other drug in same dose. **PRIMARY OUTCOME VARIABLE:** Clinical control of seizures (seizure free period of 24 hours after giving anticonvulsant). **RESULTS:** Baseline characteristics including mean birthweight, gestation age and sex were comparable in both groups. Seizures were controlled in 8 of the 55 (14.5%) neonates who received phenytoin, as compared to 39 of 54 (72.2%) neonates who received phenobarbitone (P <0.001). In babies not responding to assigned drugs, after cross-over to the other drug, seizure control was achieved in 44/55 (80%) of the neonates assigned to receive phenytoin first as compared to 49/54 (91%) of those assigned to receive phenobarbitone first (P=0.014). After maximum dose of phenobarbitone seizures were controlled in 49/55(89%) in phenytoin group and 52/54 (96%) in phenobarbitone group (P<0.05). **CONCLUSIONS:** Phenobarbitone is more efficacious than phenytoin in control of clinical seizures in term or near-term neonates, irrespective of etiology. To evaluate serum vascular endothelial growth factor (VEGF) levels in children with acute lymphoblastic leukemia (ALL) during the induction phase of chemotherapy.

<http://www.ncbi.nlm.nih.gov/pubmed/23502660>

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## Characteristics of pain in children and youth with cerebral palsy

Penner, M., et al.

*Pediatrics* 2013; 132(2): e407-413

**OBJECTIVES:** Pain in children with cerebral palsy (CP) is underrecognized, undertreated, and negatively affects quality of life. Communication challenges and multiple pain etiologies complicate diagnosis and treatment. The primary objectives of this study were to determine the impact of pain on activities and to identify the common physician-identified causes of pain in children and youth ages 3 to 19 years across all levels of severity of CP. **METHODS:** The study design was cross-sectional, whereby children/youth aged 3 to 19 years and their families were consecutively recruited. The primary caregivers were asked to complete a one-time questionnaire, including the Health Utilities Index 3 pain subset, about the presence and characteristics of pain. The treating physician was asked to identify the presence of pain and provide a clinical diagnosis for the pain, if applicable. **RESULTS:** The response rate was 92%. Of 252 participants, 54.8% reported some pain on the Health Utilities Index 3, with 24.4% of the caregivers reporting that their child experienced pain that affected some level of activities in the preceding 2 weeks. Physicians reported pain in 38.7% and identified hip dislocation/subluxation, dystonia, and constipation as the most frequent causes of pain. **CONCLUSIONS:** One-quarter of our sample experienced pain that limited activities and participation. Clinicians should be aware that hip subluxation/dislocation and dystonia were the most common causes of pain in children/youth with CP in this study. Potential causes of pain should be identified and addressed early to mitigate the negative impact of pain on quality of life.

<http://www.ncbi.nlm.nih.gov/pubmed/23858420>

## Uncontrolled epilepsy is not necessarily the same as drug-resistant epilepsy: Differences between populations with newly diagnosed epilepsy and chronic epilepsy

Hao, X., et al.

*Epilepsy Behav* 2013; 29(1): 4-6

**BACKGROUND:** A proportion of patients with seemingly “uncontrolled” epilepsy could still control their epilepsy with further pharmacological manipulations. It is possible that their epilepsy might not be truly “drug-resistant”. We audited the patients with “uncontrolled epilepsy” using the recent ILAE definition of drug-resistant epilepsy. **METHODS:** Patients with newly diagnosed epilepsy at Glasgow and patients with chronic epilepsy treated in Hong Kong were independently assessed at their last clinic visit. If the patient was not seizure-free, the epilepsy was considered “uncontrolled”. In this latter situation, if the patient had adequate trials of two or more tolerated, appropriately chosen, and appropriately used AED schedules, the epilepsy was classified as “drug-resistant” in accordance with the ILAE definition. If not, the outcome was classified as “undefined”, and the reason(s) for this was documented. **RESULTS:** In the newly diagnosed cohort with uncontrolled epilepsy (n=311), outcome was “undefined” in 175 (56%). The most common reasons were trying just one AED usually at the patient’s behest (n=68; 39%); intermittent compliance (60; 34%); adverse effects at low dosage (51; 29%); inadequate dosing (49; 28%); social issues such as imprisonment, alcohol, and recreational drug use (34; 19%); psychiatric problems affecting documentation, attendance, etc. (32; 18%); patient choice accepting less than optimal control (14; 8%); and seizure freedom of less than 12 months (12.7%). In the chronic cohort of 194 patients with uncontrolled epilepsy, drug responsiveness was “undefined” in just 79 (41%). The most common reasons were inadequate use of the AED(s) (35; 44%), followed by a lack of information on treatment response in the medical records (18; 23%) and failure of only one adequately used AED (11; 14%). **CONCLUSION:** Uncontrolled epilepsy is not necessarily the same as drug-resistant epilepsy. Efforts should be made to understand why a patient is not seizure-free so that appropriate adjustment in AED regimen can be taken to enable the patient to attain long-term seizure freedom.

<http://www.ncbi.nlm.nih.gov/pubmed/23911352>

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## Presentation and management of chronic pain

Rajapakse, D., et al.

*Arch Dis Child* 2014; 99(5): 474-480

Chronic pain is an important clinical problem affecting significant numbers of children and their families. The severity and impact of chronic pain on everyday function is shaped by the complex interaction of biological, psychological and social factors that determine the experience of pain for each individual, rather than a straightforward reflection of the severity of disease or extent of tissue damage. In this article we present the research findings that strongly support a biopsychosocial concept of chronic pain, describe the current best evidence for management strategies and suggest a common general pathway for all types of chronic pain. The principles of management of some of the most important or frequently encountered chronic pain problems in paediatric practice; neuropathic pain, complex regional pain syndrome (CRPS), musculoskeletal pain, abdominal pain and headache are also described.

<http://www.ncbi.nlm.nih.gov/pubmed/24554056>

## Pediatric clinical practice guidelines for acute procedural pain: a systematic review

Lee, G. Y., et al.

*Pediatrics* 2014; 133(3): 500-515

**BACKGROUND:** Procedural pain assessment and management have been extensively studied through multiple research studies over the past decade. Results of this research have been included in numerous pediatric pain practice guidelines. **OBJECTIVE:** To systematically review the quality of existing practice guidelines for acute procedural pain in children and provide recommendations for their use. **METHODS:** A systematic search was conducted on Medline, Embase, CINAHL, PsycINFO, and Scopus from 2000 to July 2013. A grey literature search was also conducted through the Translating Research Into Practice database, Guidelines International Network database, and National Guideline Clearinghouse. Four reviewers rated relevant guidelines using the Appraisal of Guidelines for Research and Evaluation (AGREE) II Instrument. Screening of guidelines, assessment of methodological quality, and data abstraction were conducted by 2 pairs of raters. Disagreements in overall assessments were resolved through consensus. **RESULTS:** Eighteen guidelines from 4930 retrieved abstracts were included in this study. Based on the AGREE II domains, the guidelines generally scored high in the scope and purpose and clarity of presentation areas. Information on the rigor of guideline development, applicability, and editorial independence were specified infrequently. Four of the 18 guidelines provided tools to help clinicians apply the recommendations in practice settings; 5 were recommended for use in clinical settings, and the remaining 13 were recommended for use with modification. **CONCLUSIONS:** Despite the increasing availability of clinical practice guidelines for procedural pain in children, the majority are of average quality. More transparency and comprehensive reporting are needed for the guideline development process.

<http://www.ncbi.nlm.nih.gov/pubmed/24488733>

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## Methylnaltrexone for opioid-induced constipation in pediatric oncology patients

Rodrigues, A., et al.

*Pediatr Blood Cancer* 2013; 60(10): 1667-1670

**BACKGROUND:** Pediatric oncology patients can experience opioid-induced constipation, which may not respond to laxative treatment. Methylnaltrexone is an opioid receptor antagonist that can reverse opioid-induced constipation without affecting analgesia. Published literature on the use of methylnaltrexone in children is very limited. This retrospective review describes the effectiveness and safety of methylnaltrexone for opioid-induced constipation in pediatric oncology patients. **PROCEDURE:** A retrospective review of health records was conducted for pediatric oncology in-patients who were prescribed methylnaltrexone between May 2008 and September 2012 at The Hospital for Sick Children. Demographic, clinical, efficacy, and safety data were collected, including; opioid, laxative, and methylnaltrexone dosing and frequency. **RESULTS:** Fifteen patients (median age: 14 years, range: 4-17 years) received methylnaltrexone; 12 received a single dose while three received multiple doses. At the time of methylnaltrexone administration, patients were receiving a median oral morphine dose equivalent of 5.7 mg/kg/day (range: 1.5-29.2 mg/kg/day) and had not had any bowel movements for several days despite treatment with multiple laxatives. Methylnaltrexone was given at a mean dose of 0.15 +/- 0.02 mg/kg/dose (range: 3-12 mg/dose) as a subcutaneous injection. After 14 of 19 doses administered, patients had a bowel movement within 4 hours. Three patients had documented mild gastrointestinal upset following methylnaltrexone administration. None reported a reduction of pain control or opioid withdrawal symptoms. **CONCLUSION:** This case series suggests that methylnaltrexone is safe and may be effective when given subcutaneously as a 0.15 mg/kg single dose to pediatric oncology patients with opioid-induced constipation.

<http://www.ncbi.nlm.nih.gov/pubmed/23766091>

## Oral morphine for cancer pain

Wiffen, P. J., et al.

*Cochrane Database Syst 2013; Rev 7: CD003868*

**BACKGROUND:** This is the second updated version of a Cochrane review first published in Issue 4, 2003 of The Cochrane Library and first updated in 2007. Morphine has been used for many years to relieve pain. Oral morphine in either immediate release or modified release form remains the analgesic of choice for moderate or severe cancer pain. **OBJECTIVES:** To determine the efficacy of oral morphine in relieving cancer pain, and assess the incidence and severity of adverse effects. **SEARCH METHODS:** We searched the following databases: Cochrane Pain, Palliative and Supportive Care Group Trials Register (June 2013); Cochrane Central Register of Controlled Trials (CENTRAL) (The Cochrane Library 2013, Issue 5, May); MEDLINE (1966 to June 2013); and EMBASE (1974 to June 2013). **SELECTION CRITERIA:** Published randomised controlled trials (RCTs) using placebo or active comparators reporting on the analgesic effect of oral morphine in adults and children with cancer pain. Trials with fewer than ten participants were excluded. **DATA COLLECTION AND ANALYSIS:** One review author extracted data, which were checked by another review author. There were insufficient comparable data for meta-analysis to be undertaken or to produce numbers needed to treat (NNTs) for the analgesic effect. We extracted any available data on the number or proportion of participants with 'no worse than mild pain' or treatment success (very satisfied, or very good or excellent on patient global impression scales). **MAIN RESULTS:** Ten new studies (638 participants) were identified for this update, bringing the total of included studies to 62, with 4241 participants. Thirty-six studies used a cross-over design ranging from one to 15 days, with the greatest number (11) for seven days for each arm of the trial. Fifteen studies compared oral morphine modified release (Mm/r) preparations with morphine immediate release (MIR). Fourteen studies compared Mm/r in different strengths; six of these included 24-hour modified release products. Fifteen studies compared Mm/r with other opioids. Six studies compared MIR with other opioids. Two studies compared oral Mm/r with rectal Mm/r. Three studies compared MIR with MIR by a different route of administration. Two studies compared Mm/r with Mm/r at different times and two compared MIR with MIR given at a different time. One study was found comparing each of the following: Mm/r tablet with Mm/r suspension; Mm/r with non-opioids; MIR with non-opioids; and oral morphine with epidural morphine. In this update a standard of 'no worse than mild pain' was set equivalent to a score of 30/100 mm or less on a visual analogue pain intensity scale (VAS), or the equivalent in other pain scales. Eighteen studies achieved this level of pain relief on average, and no study reported that good levels of pain relief were not attained. Where results were reported for individual participants in 17 studies, 'no worse than mild pain' was achieved by 96% of participants (362/377), and an outcome equivalent to treatment success in 63% (400/638). Morphine is an effective analgesic for cancer pain. Pain relief did not differ between Mm/r and MIR. Modified release versions of morphine were effective for 12- or 24-hour dosing depending on the formulation. Daily doses in studies ranged from 25 mg to 2000 mg with an average of between 100 mg and 250 mg. Dose titration was undertaken with both instant release and modified release products. A small number of participants did not achieve adequate analgesia with morphine. Adverse effects were common and approximately 6% of participants discontinued treatment because of intolerable adverse effects. **AUTHORS' CONCLUSIONS:** The effectiveness of oral morphine has stood the test of time, but the randomised trial literature for morphine is small given the importance of this medicine. Most trials recruited fewer than 100 participants and did not provide appropriate data for meta-analysis. Only a few reported how many people had good pain relief, but where it was reported, over 90% had no worse than mild pain within a reasonably short time period. The review demonstrates the wide dose range of morphine used in studies, and that a small percentage of participants are unable to tolerate oral morphine. The review also shows the wide range of study designs, and inconsistency in cross-over designs. Trial design was frequently based on titration of morphine or comparator to achieve adequate analgesia, then crossing participants over in cross-over design studies. It was not clear if these trials are sufficiently powered to detect any clinical differences between formulations or comparator drugs. New studies added to the review reinforce the view that it is possible to use modified release morphine to titrate to analgesic effect. There is qualitative evidence that oral morphine has much the same efficacy as other available opioids.

## Parent-reported pain in Rett syndrome

Symons, F. J., et al.

*Clin J Pain* 2013; 29(8): 744-746.

**OBJECTIVES:** Clinical reports suggest that patients with Rett syndrome (RTT) live with significant chronic health issues as well as severe motor and communication impairments. Consequently, patients with RTT may be at risk for living with pain but not having it recognized. The purpose of this preliminary study was to document parent reported estimates of pain frequency, pain communication, and pain source. **METHODS:** Caregivers of 44 patients with clinically diagnosed RTT (mean RTT age = 21.5, SD = 13.5) completed a health survey about their daughter that contained a number of items specific to pain from the Non-Communicating Children's Pain Checklist - Revised **SURVEY RESULTS:** Among survey responders, 24% reported that their child had experienced pain on 8 or more days (> 1 week) in the previous 30 days. The most frequent form of pain communication was facial expression (85%) and vocalization (82%, eg, moan, cry). The most commonly reported pain source was gastro-intestinal (66%). Pain frequency was significantly ( $P<0.05$ ) correlated with age (0.41), number of pain sources (0.72), and number of health problems (0.45); and the number pain sources was significantly ( $P<0.05$ ) correlated with number of health problems (0.67). **DISCUSSION:** These preliminary results suggest that pain is a problem for a significant subgroup of patients with RTT. Almost one quarter of respondents indicated their daughters experience over a week of pain per month. The frequent health and communication issues associated with RTT suggest an increased risk that pain may be overlooked or discounted in this vulnerable population.

<http://www.ncbi.nlm.nih.gov/pubmed/23835769>

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## Validation of self-report pain scales in children

Tsze, D. S., et al.

*Pediatrics* 2013; 132(4): e971-979

**BACKGROUND AND OBJECTIVES:** The Faces Pain Scale-Revised (FPS-R) and Color Analog Scale (CAS) are self-report pain scales commonly used in children but insufficiently validated in the emergency department setting. Our objectives were to determine the psychometric properties (convergent validity, discriminative validity, responsivity, and reliability) of the FPS-R and CAS, and to determine whether degree of validity varied based on age, sex, and ethnicity. **METHODS:** We conducted a prospective, observational study of English- and Spanish-speaking children ages 4 to 17 years. Children with painful conditions indicated their pain severity on the FPS-R and CAS before and 30 minutes after analgesia. We assessed convergent validity (Pearson correlations, Bland-Altman method), discriminative validity (comparing pain scores in children with pain against those without pain), responsivity (comparing pain scores pre- and postanalgesia), and reliability (Pearson correlations, repeatability coefficient). **RESULTS:** Of 620 patients analyzed, mean age was 9.2 +/- 3.8 years, 291(46.8%) children were girls, 341(55%) were Hispanic, and 313(50.5%) were in the younger age group (<8 years). Pearson correlation was 0.85, with higher correlation in older children and girls. Lower convergent validity was noted in children <7 years of age. All subgroups based on age, sex, and ethnicity demonstrated discriminative validity and responsivity for both scales. Reliability was acceptable for both the FPS-R and CAS. **CONCLUSIONS:** The FPS-R and CAS overall demonstrate strong psychometric properties in children ages 4 to 17 years, and between subgroups based on age, sex, and ethnicity. Convergent validity was questionable in children <7 years old.

<http://www.ncbi.nlm.nih.gov/pubmed/23999954>

## Managing children with raised intracranial pressure: Part one (introduction and meningitis)

Paul, S., et al.

*Nurs Child Young People* 2013; 25(10): 31-36

Intracranial pathologies in children need urgent identification and management. This article is presented in two parts, with part one describing intracranial pressure and outlining the features and management of meningitis. Part two, to be published in February 2014, outlines the features and management of brain tumours and intracranial bleeds. Each condition is accompanied by an illustrative case study to give an idea of what nurses might encounter in a child presenting with raised intracranial pressure.

<http://www.ncbi.nlm.nih.gov/pubmed/24308484>

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## Managing children with raised intracranial pressure: Part two (brain tumours and intracranial bleeds)

Paul, S., et al.

*Nurs Child Young People* 2014; 26(1): 30-37

This article is the second of two examining the causes of increased intracranial pressure in children. Key features and management associated with brain tumours and intracranial bleeds are highlighted. The conditions are accompanied by illustrative case studies to give an idea of what children's nurses may encounter in a patient presenting with raised intracranial pressure. Part one, published in December 2013, focused on the signs and symptoms of raised intracranial pressure and meningitis.

<http://www.ncbi.nlm.nih.gov/pubmed/24517622>

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## Pallidal deep brain stimulation for dystonia: A case series

Petrossian, M. T., et al.

*J Neurosurg Pediatr* 2013; 12(6): 582-587

**OBJECT:** Pallidal deep brain stimulation (DBS) is a treatment option for those with early-onset dystonia. However, there are limited data on long-term outcome and treatment complications. The authors report on the short- and long-term effects of pallidal DBS in a cohort of patients with early-onset dystonia. **METHODS:** Fourteen consecutive pediatric patients with early-onset dystonia were systematically evaluated and treated. The duration of follow-up ranged from 16 to 84 months. **RESULTS:** There were no immediate postoperative complications. At last follow-up, 12 of the 14 patients displayed a significant decline in the Burke-Fahn-Marsden Dystonia Rating Scale motor subscale score, with an average decrease of 62% +/- 8.4%. The most common hardware complication was lead fracture (14.3%). **CONCLUSIONS:** These data provide further evidence that DBS is a safe and effective treatment for those with early onset dystonia.

<http://www.ncbi.nlm.nih.gov/pubmed/24093589>

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## Pain assessment during a vaso-occlusive crisis in the pediatric and adolescent patient: Rethinking practice

Schiavenato, M. and O. Alvarez

*J Pediatr Oncol Nurs* 2013; 30(5): 242-248

Pain assessment of the child and adolescent with sickle cell disease is complex and challenging. We present a paradigm of pain assessment during a vaso-occlusive crisis in children and adolescents based on the Pain Assessment as a Social Transaction model. Using this model, the assessment of pain severity in sickle cell disease is uniquely highlighted as comprising at least 4 key factors: the limitations of current pain assessment tools, the existence of acute pain of various origins and the emergence and coexistence of chronic pain, the prevalence of cognitive deficits, and the sociocultural dynamics in America. Improved tools for pain assessment and targeted practitioner education are warranted.

<http://www.ncbi.nlm.nih.gov/pubmed/23850944>

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## Integrative care for pediatric patients with pain

Young, L. and K. J. Kemper

*J Altern Complement Med* 2013; 19(7): 627-632

**OBJECTIVES:** Although pediatric patients with chronic pain often turn to complementary therapies, little is known about patients who seek academic integrative pediatric care. **DESIGN:** The study design comprised abstraction of intake forms and physician records from new patients whose primary concern was pain. **SETTING/LOCATION:** The study setting was an academic pediatric clinic between January 2010 and December 2011. **Subjects:** Of the 110 new patients, 49 (45%) had a primary concern about headache (20), abdominal pain (18), or musculoskeletal pain (11). **RESULTS:** The average age was 13+/-4 years, and 37% were male. Patients reported an average pain level of 6+/-3 on a 10-point scale, and most reported more than one kind of pain; parents had an average of 7+/-3 health concerns per child, including fatigue (47%), mood or anxiety (45%), constipation/diarrhea (41%), and/or sleep problems (35%). Most patients (57%) were referred by specialists; 71% were taking prescription medications; and 53% were taking one or more dietary supplements at intake. Of those tested, most (61%) had suboptimal vitamin D levels. All families wanted additional counseling about diet (76%), exercise (66%), sleep (58%), and/or stress management (81%). In addition to encouraging continued medical care (100%) and referral to other medical specialists (16%), frequent advice included continuing or initiating dietary supplements such as vitamins/minerals (80%), omega-3 fatty acids (67%), and probiotics (31%). Stress-reducing recommendations included biofeedback (33%), gratitude journals (16%), and yoga/t'ai chi (8%). Other referrals included acupuncture (24%) and massage (20%). **Discussion:** Patients who have chronic pain and who present to an integrative clinic frequently have complex conditions and care. They are interested in promoting a healthy lifestyle, reducing stress, and using selected complementary therapies. **CONCLUSION:** Patients with chronic pain who seek integrative care may benefit from the kind of coordinated, integrated, comprehensive care provided in a medical home.

<http://www.ncbi.nlm.nih.gov/pubmed/23448120>

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## The role of cognitive behavioral therapy for chronic pain in adolescents

Zagustin, T. K.

*PM R* 2013; 5(8): 697-704

Chronic pain is frequently experienced in adolescents; it affects functionality and requires interventions to decrease the impairments caused by pain. Cognitive behavioral therapy (CBT) has been analyzed in numerous studies that evaluated its effects on reducing the different types of chronic pain in children and adolescents. Interestingly, the outcome of CBT was initially focused on pain intensity, but, because there is no correspondence between children's pain intensity and level of disability, the ability to participate in school and social and recreational activities have been the primary focus of recent studies. There are innovative methods of CBT (such as the third generation of CBT) with and without the use of technology that facilitates the availability of this psychological treatment to adolescents with chronic pain, optimizing its accessibility and comprehensiveness, and maintaining its effectiveness. In the future, specific types of CBT could be specific to the diagnosis of chronic pain in the adolescent, sociodemographics, and other unique features. Parents of children with chronic pain are usually included in these programs, either as coaches in the intervention or as recipients of psychological therapies (including CBT) to optimize benefits. CBT has no adverse effect on chronic pain in adolescents, and there is no literature that makes reference to the effectiveness of CBT in preventing chronic pain in adolescents. A review of the role of CBT in chronic pain in adolescents via a PubMed database search was performed to identify the role of CBT in the management of chronic pain in adolescents.

<http://www.ncbi.nlm.nih.gov/pubmed/23953015>

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## Percutaneous endoscopic gastrostomy tube placement in children with neurodevelopmental disabilities: Parents' perspectives

Alsaggaf, A. H., et al.

*Saudi Med* 2013; *J* 34(7): 695-700

**OBJECTIVE:** To study the attitudes of parents toward percutaneous endoscopic gastrostomy (PEG) tube placement and identify contributing factors to their negative attitudes. **METHODS:** Thirty consecutive parents were included retrospectively through a single endoscopy unit at the King Abdulaziz University Hospital, Jeddah, Kingdom of Saudi Arabia from January to July 2012. A structured 25-item questionnaire was designed to examine their demographics, attitudes, and experience with the PEG procedure. **RESULTS:** Patients' ages were 3-19 years (mean: 10.2), mostly with severe cerebral palsy (77%). Their PEG tubes were inserted 2-144 months (mean: 39) prior to the encounter. Only 43% of the parents felt informed and most (73%) had negative attitudes toward the procedure, which was associated with significant delays ( $p=0.016$ ). After the procedure, most parents (67%) reported a better-than-expected experience, which was associated with their information levels ( $p=0.03$ ). Most parents (80%) regretted not having the PEG tube placed earlier. This depended on their information level, as those who were not informed were more likely to have strong regrets when compared to those informed (82% versus 42%,  $p=0.008$ ). **CONCLUSION:** Most parents are not well-informed regarding the PEG procedure, which affects their expectations and experiences. Most parents found the experience better than what they expected and regretted not having carried it out earlier.

<http://www.ncbi.nlm.nih.gov/pubmed/23860888>

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## Parents' perspectives on the deaths of their children in two Brazilian paediatric intensive care units

Abib El Halal, G. M., et al.

*Int J Palliat Nurs* 2013; *19*(10): 495-502

**OBJECTIVES:** To evaluate the quality of care offered to terminally ill children and their families in the last days of life in two Brazilian Paediatric Intensive Care Units (PICUs) from the parents' perspectives. **METHODS:** This was a qualitative, exploratory study. Parents of a child who had died in one of the PICUs 6-12 months previously were invited to take part in two interviews: a private meeting with the PICU assistant physician who cared for their child, to discuss and review any outstanding issues related to the diagnosis, treatment, and prognosis, and a recorded interview with a researcher who was not involved in the child's treatment. Data from the interviews with the researcher were posteriorly grouped in categories according to recurrent terms. **RESULTS:** Six categories emerged, three of which are reported here. The quality of communication was low; the medical staff frequently used technical terms, limiting understanding. Parental participation in the decision-making process was scarce; decisions were based on the medical perspective. Finally, families reported uncompassionate attitudes from the medical staff and excessive technology in the final moments surrounding the child's death, although nurses were highly involved with palliative care measures and demonstrated sympathetic and supportive postures. **CONCLUSION:** The interviews uncovered deficiencies in the care provided to parents in the PICUs, indicating a need for changes in practice.

## Measuring exertion during caregiving of children and young adults with cerebral palsy who require assistance for mobility and self-care

Andrews, M., et al.

*Phys Occup Ther Pediatr* 2013; 33(3): 300-312

Our purpose was to compare objective and subjective measures of energy exertion during caregiving tasks. Participants were primary caregivers (N = 19) of children and young adults (aged 3 -22 years) with cerebral palsy (CP) who require assistance for mobility and self-care (67% classified in level V on the Gross Motor Function Classification System). Measures of exertion were collected during two caregiving tasks: (1) transfers and (2) dressing. Objective measures included volume of oxygen (VO<sub>2</sub>), heart rate (HR), and the subjective measure was a rating of perceived exertion (Borg RPE). Controlling for baseline status, perceived exertion correlated with VO<sub>2</sub> (0.43,  $p < .01$ ) and HR (0.29,  $p < .01$ ) during the tasks. Caregivers with high baseline HR and VO<sub>2</sub>, had high Borg RPE scores following a task. Correlations were found between HR and VO<sub>2</sub> during caregiving tasks (0.63,  $p < .01$ ). Patterns of association with caregiver and child characteristics were similar for VO<sub>2</sub> and Borg RPE. Subjective measures of exertion appear to capture the strain of caregiving. Understanding a caregiver's perception of exertion can guide therapists in assessing the need for equipment, pharmacological, or respite interventions.

<http://www.ncbi.nlm.nih.gov/pubmed/23356332>

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## Bereaved parents' intentions and suggestions about research autopsies in children with lethal brain tumors

Baker, J. N., et al.

*J Pediatr* 2013; 163(2): 581-586

**OBJECTIVE:** To determine bereaved parents' perceptions about participating in autopsy-related research and to elucidate their suggestions about how to improve the process. **STUDY DESIGN:** A prospective multicenter study was conducted to collect tumor tissue by autopsy of children with diffuse intrinsic pontine glioma. In the study, parents completed a questionnaire after their child's death to describe the purpose for, hopes (ie, desired outcomes of), and regrets about their participation in autopsy-related research. Parents also suggested ways to improve autopsy-related discussions. A semantic content analytic method was used to analyze responses and identify themes within and across parent responses. **RESULTS:** Responses from 33 parents indicated that the main reasons for participating in this study were to advance medical knowledge or find a cure, a desire to help others, and choosing as their child would want. Parents hoped that participation would help others or help find a cure as well as provide closure. Providing education/ anticipatory guidance and having a trusted professional sensitively broach the topic of autopsy were suggestions to improve autopsy discussions. All parents felt that study participation was the right decision, and none regretted it; 91% agreed that they would make the choice again. **CONCLUSION:** Because autopsy can help advance scientific understanding of the disease itself and because parents reported having no regret and even cited benefits, researchers should be encouraged to continue autopsy-related research. Parental perceptions about such studies should be evaluated in other types of pediatric diseases.

<http://www.ncbi.nlm.nih.gov/pubmed/23433673>

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## Symptoms of post-traumatic stress disorder in bereaved children and adolescents: Factor structure and correlates

Boelen, P. A. and M. Spuij

*J Abnorm Child Psychol* 41(7): 1097-1108

This study investigated the factor structure and correlates of posttraumatic stress-disorder (PTSD) symptoms among children and adolescents confronted with the death of a loved one. Three hundred thirty-two bereaved children and adolescents (aged 8-18; 56.9 % girls) who all received some form of psychosocial support after their loss, completed self-report measures of PTSD, together with measures tapping demographic and loss-related variables, depression, prolonged grief, and functional impairment. Parent-rated indices of impairment were also collected. We first evaluated the fit of six alternative models of the factor structure of PTSD symptoms, using confirmatory factor analyses. Outcomes showed that the 4-factor numbing model from King et al. (*Psychological Assessment* 10, 90-96, 1998), with distinct factors of reexperiencing, avoidance, emotional numbing, and hyperarousal fit the data best. Of all participants, 51.5 % met DSM-IV criteria for PTSD. PTSD-status and scores on the PTSD factors varied as a function of age and gender, but were unrelated to other demographic and loss-related variables. PTSD-status and scores on the PTSD factors were significantly associated symptom-levels of depression, prolonged grief, and functional impairment. Findings complement prior evidence that the DSM-IV model of the factor structure of PTSD symptoms may not represent the best conceptualization of these symptoms and highlight the importance of addressing PTSD symptoms in children and adolescents seeking help after bereavement.

<http://www.ncbi.nlm.nih.gov/pubmed/23612882>

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## Birth of a child with congenital heart disease: Emotional reactions of mothers and fathers according to time of diagnosis

Bevilacqua, F., et al.

*J Matern Fetal Neonatal Med* 2013; 26(12): 1249-1253

OBJECTIVE: To evaluate emotional distress, depression and quality of life in parents of infants with severe congenital heart disease (CHD) during their first hospitalization. METHODS: A pilot study for 38 parental couples of infants with CHD hospitalized within the 3 months of life. Parents filled up three self-administered questionnaires. We compared differences in the variables measuring emotional distress, depression and quality of life between mothers and fathers, and between prenatal and postnatal diagnosis. RESULTS: Stress and depression levels were significantly higher in mothers than in fathers (stress: 81.8% mothers versus 60.6% fathers; depression: 45.7% mothers versus 20.0% fathers). No difference were found between prenatal and postnatal groups in any field tested but, in percentage, mothers receiving prenatal diagnosis were more depressed, whereas those receiving postnatal diagnosis were more stressed. Fathers showed same tendency. CONCLUSIONS: Parents of newborns with severe CHD, especially mothers, need psychological support during their child's hospitalization. Parents of children diagnosed prenatally may need counseling throughout pregnancy to help them recover from the loss of the imagined healthy child.

<http://www.ncbi.nlm.nih.gov/pubmed/23419128>

## Variation in reported experience of involvement in cancer treatment decision making: Evidence from the National Cancer Patient Experience Survey

El Turabi, A., et al.

*Br J Cancer* 2013; 109(3): 780-787

**BACKGROUND:** Exploring variation in patients' experiences of involvement in treatment decision making can identify groups needing extra support, such as additional consultation time, when considering treatment options. **METHODS:** We analysed data from the 2010 English National Cancer Patient Experience Survey, a national survey of all patients attending hospitals in England for cancer treatment over a 3-month period, to examine how experience of involvement in decisions about treatment varied between patients with 38 different primary cancers using logistic regression. We analysed responses from 41 411 patients to a single question examining patient experience of involvement in treatment decision making. We calculated unadjusted odds ratios of reporting the most positive experience between patients of different sociodemographic and tumour characteristics and explored the effects of adjusting for age, gender, ethnicity, deprivation, cancer type and hospital of treatment. **RESULTS:** Of the 41 441 respondents, 29 776 (72%) reported positive experiences of decision-making involvement. Younger patients reported substantially less positive experiences of involvement in decision making (adjusted OR=0.49 16-24 vs 65-74;  $P<0.001$ ), as did ethnic minorities (adjusted ORs=0.52, 0.62 and 0.73 for Black, Chinese and Asian vs White patients, respectively;  $P<0.001$ ). Experience varied considerably between patients with different cancers (e.g., OR=0.52 for anal and 1.37 for melanoma vs colon cancer;  $P<0.001$ ), with ovarian, myeloma, bladder and rectal cancer patients reporting substantially worse experiences compared with other patients with gynaecological, haematological, urological and colorectal cancers, respectively. Clustering of different patient groups within hospitals with outlying performance report scores could not account for observed differences. **CONCLUSION:** Efforts to improve involvement in treatment decision making can focus on those who report the worst experience, in particular younger patients, ethnic minorities and patients with rectal, ovarian, multiple myeloma and bladder cancer.

<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3738115/>

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## Parenting stress evaluation and behavioral syndromes in a group of pediatric patients with epilepsy

Farrace, D., et al.

*Epilepsy Behav* 2013; 29(1): 222-227

The aim of the present work was to measure the amount of stress in parents of children with epilepsy and to determine whether and how parenting stress is linked to behavioral symptoms of the children. Parenting stress was measured with the Parenting Stress Index (PSI) and behavioral symptoms with the Child Behavior Checklist (CBCL). Data obtained from 26 parents of children with epilepsy were compared with those obtained from 31 parents of healthy children. Children with epilepsy obtained higher scores in all the subscales of PSI and in almost all the subscales of CBCL compared with healthy children. Epilepsy caused a high level of parenting stress and of problematic behaviors since the behavioral symptoms predicting the degree of parenting stress significantly differed between healthy children and children with epilepsy. Therefore, parents of children with epilepsy should be offered psychological support to cope with parenting stress and to improve the relationship with their children.

<http://www.ncbi.nlm.nih.gov/pubmed/24034672>

## Suicide ideation in pediatric and adult survivors of childhood brain tumors

Brinkman, T. M., et al.

*J Neurooncol* 2013; 113(3): 425-432

Survivors of pediatric brain tumors are at risk for long-term psychological morbidities. The current study investigated the prevalence and predictors of suicide ideation (SI) in a clinical sample of youth and adult survivors. Retrospective chart reviews were completed for 319 survivors of pediatric brain tumors who were assessed via clinical interview during routine neuro-oncology clinic visits between 2003 and 2007. Survivors were, on average, 18.0 years of age (SD = 4.9) and 10 years from diagnosis (SD = 5.0) at their most recent follow-up. The most common diagnosis was low-grade glioma (n = 162) followed by embryonal tumors (PNET/medulloblastoma; n = 64). Multivariable logistic regression was used to calculate odds ratios (OR) and 95 % confidence intervals (CI) for SI. Nearly 12 % of survivors (11.7 %, n = 37) reported SI. Five survivors (1.5 %) had documented suicide attempts, though none were fatal. In a multivariable model, adjusting for sex and age, history of depression (OR = 20.6, 95 % CI = 4.2-101.1), psychoactive medication treatment (OR = 4.5, 95 % CI = 1.8-11.2), observation or surgery only treatment (OR = 3.7, 95 % CI = 1.5-9.1), and seizures (OR = 3.6, 95 % CI = 1.1-11.1) were significantly associated with SI in survivors. Survivors of pediatric brain tumors appear to be at risk for experiencing SI. Our results underscore the importance of a multidisciplinary approach to providing follow-up care for childhood brain tumor survivors, including routine psychological screenings.

<http://www.ncbi.nlm.nih.gov/pubmed/23624716>

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## Health-related quality of life in childhood cancer

Fakhry, H., et al.

*J Dev Behav Pediatr* 2013; 34(6): 419-440

OBJECTIVE: Health-related quality of life (HRQoL) has become an increasingly important measure of research and treatment outcomes across all medical specialties. However, to date, there has not been an in-depth review of research relevant specifically to HRQoL in the populations of children and adolescents with cancer. In this review, the authors examine the effects of cancer on HRQoL from diagnosis to remission/survivorship and the end of life. DESIGN: A literature search was conducted using Medline and PsycINFO for articles published from 2002 to 2011. Studies included patients from diagnosis to remission and also the terminally ill. Twenty-nine studies specifically addressing HRQoL were selected after reaching consensus and study quality check. RESULTS: Children who are newly diagnosed with cancer and are undergoing treatment or are terminally ill have impaired HRQoL. Survivors of childhood cancer have high HRQoL (with the exception of those who experienced medical comorbidity or PTSD). The authors found that demographic differences, cancer types, and treatment regimens, all significantly influence the negative impact of cancer on patients' HRQoL. CONCLUSIONS: There are specific and identifiable impacts of childhood cancer on patients' HRQoL that are significant and complex across the span of the illness. There is a need for continued research in many areas related to this population, especially related to those with terminal illness in order to improve patient care.

<http://www.ncbi.nlm.nih.gov/pubmed/23838588>

## The adolescent and young adult with cancer: State of the art: Psychosocial aspects

Epelman, C. L. (2013)

*Curr Oncol Rep* 2013; 15(4): 325-331

Adolescents and young adults with cancer are a distinct subgroup of patients within oncology. From the onset of symptoms until the completion of therapy and beyond, they face physical, psychological, and social challenges that are significantly different from those of adults and children. Survival rates and quality of life outcomes for this population have not improved to the same extent that have for younger and older patients. Improvements in quality of care, overall survival and quality of life for these patients require access to specialized care and participation in clinical trials; assistance with management of disease and treatment effects (especially fertility and body image issues); assessment of psychosocial needs; facilitated transition to off-treatment care; and referral to age-appropriated information and support services. Staff team caring for young patients must be dedicated to working with this age group and should have specialist knowledge and training to support their specific needs.

<http://www.ncbi.nlm.nih.gov/pubmed/23700032>

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## Adolescent survivors of childhood cancer: Are they vulnerable for psychological distress?

Gianinazzi, M. E., et al.

*Psychooncology* 2013; 22(9): 2051-2058

**OBJECTIVES:** We aimed to (i) evaluate psychological distress in adolescent survivors of childhood cancer and compare them to siblings and a norm population; (ii) compare the severity of distress of distressed survivors and siblings with that of psychotherapy patients; and (iii) determine risk factors for psychological distress in survivors. **METHODS:** We sent a questionnaire to all childhood cancer survivors aged <16 years when diagnosed, who had survived  $\geq 5$  years and were aged 16-19 years at the time of study. Our control groups were same-aged siblings, a norm population, and psychotherapy patients. Psychological distress was measured with the Brief Symptom Inventory-18 (BSI-18) assessing somatization, depression, anxiety, and a global severity index (GSI). Participants with a T-score  $\geq 57$  were defined as distressed. We used logistic regression to determine risk factors. **RESULTS:** We evaluated the BSI-18 in 407 survivors and 102 siblings. Fifty-two survivors (13%) and 11 siblings (11%) had scores above the distress threshold (T  $\geq 57$ ). Distressed survivors scored significantly higher in somatization ( $p=0.027$ ) and GSI ( $p=0.016$ ) than distressed siblings, and also scored higher in somatization ( $p \leq 0.001$ ) and anxiety ( $p=0.002$ ) than psychotherapy patients. In the multivariable regression, psychological distress was associated with female sex, self-reported late effects, and low perceived parental support. **CONCLUSIONS:** The majority of survivors did not report psychological distress. However, the severity of distress of distressed survivors exceeded that of distressed siblings and psychotherapy patients. Systematic psychological follow-up can help to identify survivors at risk and support them during the challenging period of adolescence.

<http://www.ncbi.nlm.nih.gov/pubmed/23401292>

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## Young people describe their pre-diagnosis cancer experience

Gibson, F., et al.

*Psychooncology* 2013; 22(11): 2585-2592

**OBJECTIVE:** Young people often report a protracted journey to diagnosis and frequently report perceived delays. This study was undertaken to increase understanding of the self-reported pre-diagnosis experiences in young people with a non-haematological cancer, as close as possible to the time of diagnosis.

**METHODS:** Narrative interviews were conducted with 24 young people aged 16-24, 2-4 months from the diagnosis of a solid tumour. Data were analysed to identify whether prediagnosis narratives could be classified according to shared characteristics (typologies) to identify broader contextual issues concerning cancer, and cancer risk perceptions, in this age group. Case notes were also accessed to contextualize and confirm accounts. **RESULTS:** The main themes, which included a group narrative concerning perspectives of delay, included the impact on an individual's everyday life by symptoms; the role that significant others in young peoples' lives played in the interpretation of symptom significance; the negotiation of entry into, and experiences of, generalist health care; entry into specialist care; and the threshold points that exemplified when events shifted and a diagnosis was eventually obtained. **CONCLUSIONS:** The narratives reveal complex, and multidimensional explanations for delay with individual and contextual factors contributing. Insights were gained into preventable diagnostic delay; including investigations having been instigated, but not followed up. Each narrative also offered significant insights into how cancer symptoms should be considered within the context of young peoples' lives. This would help prevent signs and symptoms in this age group failing to trigger suspicion and not being treated seriously.

<http://www.ncbi.nlm.nih.gov/pubmed/23784978>

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## Illness uncertainty and quality of life in children with cancer

Fortier, M. A., et al.

*J Pediatr Hematol Oncol* 2013; 35(5): 366-370

**BACKGROUND:** Illness uncertainty is prevalent in children with cancer and has been associated with increased psychological distress. The relationship between illness uncertainty and quality of life in pediatric cancer patients remains unclear. The aim of the present study was to examine illness uncertainty as a predictor of health-related quality of life in children diagnosed with cancer. It was hypothesized that child-reported illness uncertainty would be negatively associated with child health-related quality of life.

**PROCEDURE:** Children aged 8 to 18 years old and receiving treatment for cancer were recruited to participate in this study. One hundred twenty children and their parent(s) completed measures of illness uncertainty, pain, anxiety, and quality of life during a routine visit to the Cancer Center at Children's Hospital of Orange County. **RESULTS:** Illness uncertainty was significantly associated with child age ( $P=0.02$ ), overall health-related ( $P<0.001$ ) and cancer-related ( $P<0.001$ ) quality of life, but not with treatment status (on/off chemotherapy) or demographic variables including sex and household income. Regression analyses statistically controlling for age, anxiety, and pain revealed that illness uncertainty significantly predicted child-reported cancer-related and health-related quality of life ( $P<0.01$ ) as well as parent-reported cancer-specific quality of life ( $P<0.01$ ). **CONCLUSIONS:** Illness uncertainty is prevalent and associated with lower quality of life in children diagnosed with cancer. Improved communication with children regarding disease state, treatment expectations, and prognosis may alleviate uncertainty and improve functioning in this vulnerable patient population.

<http://www.ncbi.nlm.nih.gov/pubmed/23669725>

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## Caregivers' estimations of their children's perceptions of death as a biological concept

Gaab, E. M., et al.

*Death Stud* 2013; 37(8): 693-703

Communication about death is often a sensitive topic in families with children. The present study compared answers of 141 school children aged 5-7 to questions about death, and their caregivers' predictions. Children were interviewed, and caregivers answered on paper, questions on inevitability, applicability, irreversibility, cessation, causation, and personal mortality. For causation, cessation, and irreversibility, children were significantly more correct than caregivers expected, and girls were more correct on applicability. Communicating with children about death may not always be as caregivers expect.

<http://www.ncbi.nlm.nih.gov/pubmed/24521028>

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## The voices of young New Zealanders involved in pediatric palliative care

Gaab, E. M., et al.

*J Palliat Care* 2013; 29(3): 186-192.

The perspectives of young New Zealanders receiving pediatric palliative care (PPC) are not well understood. A qualitative study of the perceptions of 16 PPC patients and their siblings, aged 9 to 18, was conducted through audio and written diary accounts. Inductive thematic analysis revealed several concerns of participants, including special treatment that patients had received, spending time with their families, their feelings of being judged or discriminated against, their sense of being understood themselves and of understanding others, and mortality. A nonjudgemental, open approach is recommended when consulting with patients and their siblings in order to determine their needs.

<http://www.ncbi.nlm.nih.gov/pubmed/24380218>

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## Type of continuing bonds expression and its comforting versus distressing nature: Implications for adjustment among bereaved mothers

Field, N. P., et al.

*Death Stud* 2013; 37(10): 889-912

This study investigated type of continuing bonds (CB) expression and its comforting versus distressing nature in relation to psychosocial adjustment among bereaved mothers. Twenty-eight mothers whose child had died within the previous five years participated in a CB interview in which they rated the extent they used each of 11 different types of CB expression during the past month and the degree to which they experienced each of the CB expressions as comforting and distressing. CB expressions involving illusions and hallucinations of the deceased child were predictive of greater distress whereas those involving belief that the deceased child was aware of the mother or communicating with her through dreams were not associated with symptoms, but instead linked to greater spirituality. Furthermore, mothers who reported CB as more comforting than distressing had lower symptom ratings. The implications of the findings for the attachment theory perspective on unresolved loss are discussed.

<http://www.ncbi.nlm.nih.gov/pubmed/24517520>

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## Uncovering an invisible network of direct caregivers at the end of life: A population study

Burns, C. M., et al.

*Palliat Med* 2013; 27(7): 608-615

**BACKGROUND:** Most palliative care research about caregivers relies on reports from spouses or adult children. Some recent clinical reports have noted the assistance provided by other family members and friends. **AIM:** This population study aims to define the people who actually provide care at the end of life. **SETTING/PARTICIPANTS:** A South Australian study conducted an annual randomized health population survey (n=23,706) over a 7 year period. A sample was obtained of self-identifying people who had someone close to them die and 'expected' death in the last 5 years (n=7915). Data were standardised to population norms for gender, 10-year age group, socioeconomic status, and region of residence. **RESULTS:** People of all ages indicated they provided 'hands on' care at the end of life. Extended family members (not first degree relatives) and friends accounted for more than half (n=1133/2028; 55.9%) of identified hands-on caregivers. These people came from the entire age range of the adult community. The period of time for which care was provided was shorter for this group of caregivers. People with extended family or friends providing care, were much more likely to be supported to die at home compared to having a spousal carer. **CONCLUSION:** This substantial network of caregivers who are mainly invisible to the health team provide the majority of care. Hospice and palliative care services need to create specific ways of identifying and engaging this cohort in order to ensure they are receiving adequate support in the role. Relying on 'next-of-kin' status in research will not identify them.

<http://www.ncbi.nlm.nih.gov/pubmed/23587738>

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## Assessment of family psychosocial functioning in survivors of pediatric cancer using the PAT2.0

Gilleland, J., et al.

*Psycho Oncology* 2013; 22(9): 2133-2139

**BACKGROUND:** This study aimed to examine clinical validity and utility of a screening measure for familial psychosocial risk, the Psychosocial Assessment Tool 2.0 (PAT2.0), among pediatric cancer survivors participating in long-term survivorship care. **METHODS:** Caregivers (N=79) completed the PAT2.0 during their child's survivorship appointment. Caregivers also reported on family engagement in outpatient mental health treatment. Medical records were reviewed for treatment history and oncology provider initiated psychology consults. **RESULTS:** The internal consistency of the PAT2.0 total score in this survivorship sample was strong. Psychology was consulted by the oncology provider to see 53% of participant families, and families seen by psychology had significantly higher PAT2.0 total scores than families without psychology consults. PAT2.0 total scores and corresponding subscales were higher for patients, parents, and siblings enrolled in outpatient mental health services since treatment completion. Results were consistent with psychosocial risk categories presented within the Pediatric Psychosocial Preventative Health Model. Fifty-one percent of families presenting for survivorship care scored in the "universal" category, 34% scored in the "targeted" category, and 15% scored in the "clinical" category. **CONCLUSIONS:** Data indicate that the overall proportions of families experiencing "universal", "targeted", and "clinical" levels of familial distress may be constant from the time of diagnosis into survivorship care. Overall, the PAT2.0 demonstrated strong psychometric properties among survivors of pediatric cancer and shows promise as a psychosocial screening measure to facilitate more effective family support in survivorship care.

<http://www.ncbi.nlm.nih.gov/pubmed/23529924>

## Trajectory of parental hope when a child has difficult-to-treat cancer: A prospective qualitative study

Granek, L., et al.

*Psycho Oncology* 2013; 22(11): 2436-2444

**OBJECTIVE:** This prospective and longitudinal study was designed to further our understanding of parental hope when a child is being treated for a malignancy resistant to treatment over three time points during the first year after diagnosis using a qualitative approach to inquiry. **METHODS:** We prospectively recruited parents of pediatric cancer patients with a poor prognosis who were treated in the Hematology/Oncology Program at a large children's hospital for this longitudinal grounded theory study. Parents were interviewed at three time points: within 3 months of the initial diagnosis, at 6 months, and at 9 months. Data collection and analysis took place concurrently using line-by-line coding. Constant comparison was used to examine relationships within and across codes and categories. **RESULTS:** Two overarching categories defining hope as a positive inner source were found across time, but their frequency varied depending on how well the child was doing and disease progression: future-oriented hope and present-oriented hope. Under future-oriented hope, we identified the following: hope for a cure and treatment success, hope for the child's future, hope for a miracle, and hope for more quality time with child. Under present-oriented hope, we identified hope for day-to-day/moment-to-moment, hope for no pain and suffering, and hope for no complications. **CONCLUSIONS:** For parents of children with a diagnosis of cancer with a poor prognosis, hope is an internal resource that can be present and future focused. These views fluctuated over time in response to changes in the child's well-being and disease progression.

<http://www.ncbi.nlm.nih.gov/pubmed/23737306>

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## Parents' perceptions of their child's symptom burden during and after cancer treatment

Heden, L., et al.

*J Pain Symptom Manage* 2013; 46(3): 366-375

**CONTEXT:** Previously reported studies of children with cancer mostly provide cross-sectional knowledge of the prevalence of symptoms but do not show when during the disease trajectory and after the end of successful treatment certain symptoms are most prevalent and/or distressing. **OBJECTIVES:** The aim was to describe parents' perceptions of their child's symptom burden longitudinally during and after cancer treatment and to investigate whether parents' perceptions vary with child characteristics and parent gender. **METHODS:** One hundred sixty parents (49% fathers) of 89 children answered a modified version of the Memorial Symptom Assessment Scale (MSAS) 10-18 at six different time points from one week after the child's diagnosis (T1) to 12-18 months after the end of successful treatment (T6). **RESULTS:** Feeling drowsy, pain, and lack of energy are initially the most prevalent symptoms. During treatment, the most prevalent symptom is less hair than usual. Pain, feeling sad, and nausea are initially the most distressing symptoms. Pain is both prevalent and distressing throughout the treatment. The child's symptom burden decreases over time. There is no difference regarding the reported symptom burden between the parents of a daughter or a son, or parents of a child older or younger than seven years of age. Mothers' and fathers' assessments of the symptom number, total MSAS and the subscales, are associated, but mothers' assessments are often higher than fathers' assessments. **CONCLUSION:** The prevalence and distress of symptoms and symptom burden decrease over time. However, even though the cancer is cured, feeling sad is reported as being prevalent and psychological distress is an issue. A dialogue between staff and the family about distressing symptoms and when they can be expected may increase acceptance and adaptation in children and parents during the disease trajectory.

<http://www.ncbi.nlm.nih.gov/pubmed/23498966>

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## Spinal muscular atrophy type I: Do the benefits of ventilation compensate for its burdens?

Gray, K., et al.

*J Paediatr Child Health* 2013; 49(10): 807-812

We report the progress of an 8-year-old child with spinal muscular atrophy (SMA) type 1. The parents elected in infancy that the child should be on long-term ventilation, but all attempts to establish this care at home have failed, so the child remains ventilated in the hospital. The leader of the long-term ventilation team reports on the child's progress and describes a week in the child's life. Two paediatricians argue that the benefits of long-term ventilation have not and do not compensate the child for the burdens imposed on her by this treatment and explain why they would not support the withdrawal of long-term ventilation now. They argue that long-term ventilation might have been avoided by applying to a court of law when the child was an infant. An ethicist discusses ethical aspects of decision-making in SMA type 1.

<http://www.ncbi.nlm.nih.gov/pubmed/24131113>

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## Correlation between religious coping and depression in cancer patients

Haghighi, F.

*Psychiatr Danub* 2013; 25(3): 236-240

**BACKGROUND:** Cancer often progresses very rapidly and either leads to various complications or patients eventually die of the disease. One of important consequences of cancer is depression which can increase the morbidity and mortality in non-treated cases. Religious coping is the use of religious beliefs or practices to reduce distress and deal with problems in life. This study aimed to determine the relationship between religious coping and depression in cancer patients. **SUBJECTS AND METHODS:** A descriptive-correlational study was conducted on 150 consequent cancer patients in three centers: Imam-Reza Hospital in Birjand, Qaem and Omid hospitals in Mashhad. Two questionnaires including Pargament's questionnaire for evaluation of religious coping and the Beck depression inventory were used. Data analysis was performed using multiple regression and correlation. **RESULTS:** There was no significant difference between men and women in the mean score of avoidant relationship with God and alternate fearfulness and hopefulness (ambivalence coping style). But the mean score of relationship with God in women was higher than men. The rate of depression was higher among patients who had an avoidant strategy. The religious coping method of relationship with God was effective in reducing depression. The rate of depression was lower among patients whose families had a better attitude to religion. **CONCLUSIONS:** Psychotherapy, individual/familial counseling, and especially increasing of religious beliefs such as praying and trust in God, as well as increasing the knowledge of patient and his/her family cause better acceptance of the disease and better confrontation of psychological problems.

<http://www.ncbi.nlm.nih.gov/pubmed/24048390>

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## Longitudinal study of parent caregiving self-efficacy and parent stress reactions with pediatric cancer treatment procedures

Harper, F. W., et al.

*Psycho Oncology* 2013; 22(7): 1658-1664

**BACKGROUND:** Pain/distress during pediatric cancer treatments has substantial psychosocial consequences for children and families. We examined relationships between parents' caregiving self-efficacy, parents' affect in response to their children's cancer-related treatment procedures, and parents' symptoms of post-traumatic stress at follow-up. **METHODS:** Participants were 75 pediatric cancer patients and parents. On the day of each of three procedures (i.e., port-start, lumbar puncture, or bone marrow aspiration), parents rated their self-efficacy for six caregiving goals. Parents also self-reported their negative affect (i.e., state anxiety, negative mood, and distress) in response to each procedure. Three months after the last procedure, parents reported their level of post-traumatic stress symptoms (PTSS). **RESULTS:** Higher parent self-efficacy about keeping children calm before treatment and/or keeping children calm during the procedure was associated with lower state anxiety. Self-efficacy for keeping the child calm during procedures was significantly correlated with distress in parents at the time of procedures, and self-efficacy for keeping the child calm before procedures was significantly correlated with PTSS. All three negative affect measures significantly mediated the effects of parents' caregiving self-efficacy for both goals on parents' PTSS 3 months later. **CONCLUSIONS:** Parents' caregiving self-efficacy influences their immediate and longer-term distress reactions to their children's treatment procedures. These findings provide a more nuanced understanding of how parents' cognitions contribute to their ability to cope with their children's treatment and suggest the benefit of an intervention that targets parents' procedure-specific caregiver self-efficacy.

<http://www.ncbi.nlm.nih.gov/pubmed/23034930>

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## Factors affecting parents' presence with their extremely preterm infants in a neonatal intensive care room

Heinemann, A. B., et al.

*Acta Paediatr* 2013; 102(7): 695-702

**AIM:** To describe parents' experiences of factors that influenced their stay with their extremely preterm infants in a neonatal intensive care unit (NICU). **METHODS:** This study has a qualitative descriptive design based on semistructured interviews conducted with seven mothers and six fathers. **RESULTS:** Opportunities to stay overnight together with their infant facilitated parental presence, and opportunities for taking over their infant's care empowered the parents in their parental role and increased their motivation to stay. Kangaroo mother care helped them to feel in control and feel needed, which increased their presence. High levels of illumination and noise rendered it difficult for parents to sleep and stay overnight with the infant. Low staffing levels limited their use of kangaroo mother care when they had to wait for assistance to transfer the infant from the incubator. Several participants perceived the performance of painful procedures on their child as stressful and as an obstacle to their presence. **CONCLUSION:** Kangaroo mother care and active involvement in the infant's care gave parents a sense of control and strengthened their motivation to be with their infant. High levels of noise and illumination and a dismissive staff attitude were obstacles to parents' presence.

<http://www.ncbi.nlm.nih.gov/pubmed/23590800>

## End-of-life caregivers' perception of medical and psychological support during the final weeks of glioma patients: A questionnaire-based survey

Heese, O., et al.

*Neuro Oncol* 2013; 15(9): 1251-1256

**BACKGROUND:** The prognosis for glioma remains dismal, and little is known about the final disease phase. To obtain information about this period, we surveyed caregivers of patients who were registered in the German Glioma Network and who died from the disease. **METHODS:** A questionnaire with 15 items, focusing on medical, logistic, and mental health support and symptom control during the final 4 weeks, was sent to caregivers. For some of the questions, a scale from 1 (inadequate) to 10 (excellent) was used. **RESULTS:** Of 1655 questionnaires, 605 were returned (36.6%) and evaluated. We found that 67.9% of the patients were taken care of at home for the last 4 weeks; 47.7% died at home, 22.6% died in hospitals, and 19.3% died in hospice facilities. Medical support was provided by general practitioners in 72.3% of cases, by physicians affiliated with a nursing home or hospice in 29.9%, and by general oncologists in 17%. Specialized neuro-oncologists were involved in 6%. The caregivers ranked the medical support with a mean of 7.2 (using a 10-point scale), nursing service with 8.1, and mental health support with 5.5. In 22.9% of cases, no support for the caregivers themselves was offered by medical institutions. **CONCLUSIONS:** Although these data reflect the caregivers' subjective views, they are useful in understanding and improving current patterns of care. While patients and their caregivers are supported mainly by neuro-oncologists for most of the disease phase, the end-of-life phase is managed predominantly by general practitioners and specialists in palliative care. Close cooperation between these specialties is necessary to meet the specific needs of glioma patients.

<http://www.ncbi.nlm.nih.gov/pubmed/23814266>

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## De-escalation of therapy for pediatric medulloblastoma: Trade-offs between quality of life and survival

Henrich, N., et al.

*Pediatr Blood Cancer* 2014; 61(7): 1300-1304

**BACKGROUND:** Treatment intensity for pediatric medulloblastoma may vary depending on the type of medulloblastoma. In some cases, the dose of radiation may be reduced or eliminated. Correspondingly, there may be trade-offs between quality of life and survival. In this study, focus groups were conducted with parents and clinicians to explore their opinions about these trade-offs as well as the alignment/misalignment between parents and clinicians regarding the trade-offs. **METHODS:** One hour semi-structured focus groups were conducted with parents of children with medulloblastoma and health care providers who were involved in the care of these children. **RESULTS:** Parents and providers showed differences in which factors they believe have the greatest impact on quality of life for children with medulloblastoma and their families. For parents, the most important factor is social functioning and their child's ability to make friends and have a social life. In contrast, providers thought that parents cared most about their child's cognitive functioning and ability to attend and perform in school. **CONCLUSION:** Understanding parents' perspectives on quality of life is important in terms of providing support services that target the areas that the parents prioritize. The types of functioning that are most strongly correlated with quality of life from the parents' perspective may be the ones that should be targeted to protect during treatment. *Pediatr Blood Cancer* 2014;61: 1300-1304. (c) 2014 Wiley Periodicals, Inc.

<http://www.ncbi.nlm.nih.gov/pubmed/24616367>

## Child-rearing in the context of childhood cancer: Perspectives of parents and professionals

Long, K. A., et al.

*Pediatr Blood Cancer* (2014); 61(2): 326-332

**BACKGROUND:** Elevated distress has been well documented among parents of children with cancer. Family systems theories suggest that cancer-related stressors and parental distress have the potential to affect child-rearing practices, but this topic has received limited empirical attention. The present work examined self-reported child-rearing practices among mothers and fathers of children with cancer and matched comparisons. **PROCEDURE:** Medical and psychosocial professionals with expertise in pediatric oncology selected items from the Child-Rearing Practices Report (CRPR) likely to differentiate parents of children with cancer from matched comparison parents. Then, responses on these targeted items were compared between parents of children with cancer (94 mothers, 67 fathers) and matched comparisons (98 mothers, 75 fathers). Effect sizes of between-group differences were compared for mothers versus fathers. **RESULTS:** Pediatric oncology healthcare providers predicted that 14 items would differentiate child-rearing practices of parents of children with cancer from parents of typically developing children. Differences emerged on six of the 14 CRPR items. Parents of children with cancer reported higher levels of spoiling and concern about their child's health and development than comparison parents. Items assessing overprotection and emotional responsiveness did not distinguish the two groups of parents. The effect size for the group difference between mothers in the cancer versus comparison groups was significantly greater than that for fathers on one item related to worry about the child's health. **CONCLUSION:** Parents of children with cancer report differences in some, but not all, domains of child-rearing, as predicted by healthcare professionals.

<http://www.ncbi.nlm.nih.gov/pubmed/24376229>

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## Breaking bad news: Patients' preferences and health locus of control

Martins, R. G. and I. P. Carvalho

*Patient Educ Couns* 2013; 92(1): 67-73

**OBJECTIVE:** To identify patients' preferences for models of communicating bad news and to explore how such preferences, and the reasons for the preferences, relate with personality characteristics, specifically patients' health locus of control (HLC): internal/external and 'powerful others' (PO). **METHODS:** Seventy-two patients from an oncology clinic watched videotaped scenarios of a breaking bad news moment, selected the model they preferred, filled an HLC scale and were interviewed about their choices. Data were analyzed with Chi-square, Kruskal-Wallis and Mann-Whitney tests. Interviews were content-analyzed. **RESULTS:** 77.8% preferred an empathic professional, 12.5% a distanced expert and 9.7% an emotionally burdened expert". Preferences varied significantly with HLC scores (patients with higher internal locus of control (ILC) and lower PO preferred the empathic model), presence of cancer, age and education. Patients explained their preferences through aspects of Caring, Professionalism, Wording, Time and Hope. ILC registered significant differences in regards to Wording and Time, whereas PO was associated with Hope and Time. **CONCLUSIONS:** HLC is an important dimension that can help doctors to better know their patients. **PRACTICE IMPLICATIONS:** Knowing whether patients attribute their health to their own behaviors or to chance/others can help tailor the disclosure of bad news to their specific preferences.

<http://www.ncbi.nlm.nih.gov/pubmed/23523195>

## Women's decision making and experience of subsequent pregnancy following stillbirth

Lee, L., et al.

*J Midwifery Women's Health* 2013; 58(4): 431-439

INTRODUCTION: This study sought to increase understanding of women's thoughts and feelings about decision making and the experience of subsequent pregnancy following stillbirth (intrauterine death after 24 weeks' gestation). METHODS: Eleven women were interviewed, 8 of whom were pregnant at the time of the interview. Modified grounded theory was used to guide the research methodology and to analyze the data. RESULTS: A model was developed to illustrate women's experiences of decision making in relation to subsequent pregnancy and of subsequent pregnancy itself. DISCUSSION: The results of the current study have significant implications for women who have experienced stillbirth and the health professionals who work with them. Based on the model, women may find it helpful to discuss their beliefs in relation to healing and health professionals to provide support with this in mind. Women and their partners may also benefit from explanations and support about the potentially conflicting emotions they may experience during this time.

<http://www.ncbi.nlm.nih.gov/pubmed/23855388>

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## Medication adherence and health care utilization in pediatric chronic illness: A systematic review

McGrady, M. E. and K. A. Hommel

*Pediatrics* 2013; 132(4): 730-740

BACKGROUND AND OBJECTIVE: Advanced understanding of modifiable predictors of health care use in pediatric chronic illness is critical to reducing health care costs. We examined the relationship between medication non-adherence and health care use in children and adolescents who have a chronic medical condition. METHODS: A systematic review of articles by using PubMed, PsycINFO, and CINAHL was conducted. Additional studies were identified by searching reference sections of relevant manuscripts. Studies that tested the relationship between medication non-adherence and health care use (ie, hospitalizations, emergency department visits, outpatient visits) or cost in children and adolescents (mean age  $\leq 18$  years) who have a chronic medical condition were included. Extraction of articles was completed by using predefined data fields. RESULTS: Ten studies met our inclusion criteria. Nine of the 10 studies reviewed (90%) demonstrated a relationship between medication non-adherence and increased health care use. The directionality of this relationship varied depending on the outcome variable of interest. CONCLUSIONS: Medication non-adherence is related to increased health care use in children and adolescents who have a chronic medical condition and should be addressed in clinical care. Future studies should include randomized controlled trials examining the impact of adherence promotion efforts on health care use and costs.

<http://www.ncbi.nlm.nih.gov/pubmed/23999953>

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## Cognitive and emotional aspects of fear of recurrence: Predictors and relations with adjustment in young to middle-aged cancer survivors

Park, C. L., et al.

*Psycho Oncology* 2013; 22(7): 1630-1638

**OBJECTIVE:** We investigated predictors of emotional (worry) and cognitive (perceived risk) dimensions of fear of recurrence (FOR) and their relationships with psychological well-being in a sample of young and middle-aged adult cancer survivors. **METHODS:** Eligible participants were survivors between 18 and 55 years old and diagnosed from 1 to 3 years prior. A total of 250 participants were recruited, and 167 responded to a 1-year follow-up. Demographic and psychosocial variables were assessed at baseline, and FOR and psychological well-being were assessed at follow-up. **RESULTS:** Race was associated with the cognitive dimension of FOR (such that minority race perceived less risk of recurrence), but no demographics were associated with the emotional dimension. Hierarchical regression analyses showed that spirituality was the only predictor of perceived risk independent of the effect of race, even when worry about general health was controlled. For the emotional dimension of FOR, avoidance coping predicted higher worry, but when controlling for a general tendency to worry about one's health, none of the psychosocial variables predicted worries about cancer's return. In addition, only worry about cancer's return predicted negative affect and intrusive thoughts. **CONCLUSIONS:** These results suggest that FOR comprises distinct dimensions, each of which has different implications for adjustment. These findings may have important clinical implications in developing interventions to deal with both FOR and more general health anxieties in cancer survivors.

<http://www.ncbi.nlm.nih.gov/pubmed/23060271>

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## Parent health and functioning 13 months after infant or child NICU/PICU death

Youngblut, J. M., et al.

*Pediatrics* 2013; 132(5): e1295-1301

**BACKGROUND:** After a child's death, parents may experience depression, posttraumatic stress disorder (PTSD), and increased risk for cancers, diabetes, psychiatric hospitalization, and suicide. Racial/ethnic differences are unknown. This longitudinal study investigated health and functioning of Hispanic, black, and white parents through 13 months after NICU/PICU death. **METHODS:** Parents (176 mothers, 73 fathers) of 188 deceased infants/children were recruited from 4 NICUs, 4 PICUs, and state death certificates 2 to 3 weeks after death. Deaths occurred after limiting treatment/withdrawing life support (57%), unsuccessful resuscitation (32%), or brain death (11%). Data on parent physical health (hospitalizations, chronic illness), mental health (depression, PTSD, alcohol use), and functioning (partner status, employment) were collected in the home at 1, 3, 6, and 13 months after death. **RESULTS:** Mean age for mothers was 32 +/- 8, fathers 37 +/- 9; 79% were Hispanic or black. Thirteen months after infant/child death, 72% of parents remained partnered, 2 mothers had newly diagnosed cancer, alcohol consumption was below problem drinking levels, parents had 98 hospitalizations (29% stress related) and 132 newly diagnosed chronic health conditions, 35% of mothers and 24% of fathers had clinical depression, and 35% of mothers and 30% of fathers had clinical PTSD. At 6 months after infant/child death, 1 mother attempted suicide. Week 1 after infant/child death, 9% of mothers and 32% of fathers returned to employment; 7 parents took no time off. More Hispanic and black mothers than white mothers had moderate/severe depression at 6 months after infant/child death and PTSD at every time point. **CONCLUSIONS:** Parents, especially minority mothers, have negative physical and mental health outcomes during the first year after NICU/PICU death.

<http://www.ncbi.nlm.nih.gov/pubmed/24101760>

## Family participation during intensive care unit rounds: Attitudes and experiences of parents and healthcare providers in a tertiary pediatric intensive care unit

Stickney, C. A., et al.

*J Pediatr* 2014; 164(2): 402-406 e401-404

**OBJECTIVE:** To compare the experiences and attitudes of healthcare providers and parents regarding parental participation in morning rounds, in particular to evaluate for differences in perception of parental comprehension of rounds content and parental comfort with attendance, and to identify subgroups of parents who are more likely to report comfort with attending rounds. **STUDY DESIGN:** Cross-sectional survey of 100 parents and 131 healthcare providers in a tertiary care pediatric medical/surgical intensive care unit. Descriptive statistics were used to analyze survey responses; univariate and multivariate analyses were performed to compare parent and healthcare provider responses. **RESULTS:** Of parents, 92% reported a desire to attend rounds, and 54% of healthcare providers reported a preference for parental presence. There were significant discrepancies in perception of understanding between the 2 groups, with healthcare providers much less likely to perceive that parents understood both the format (30% vs 73%,  $P < .001$ ) and content (21% vs 84%,  $P < .001$ ) of rounds compared with parents. Analysis of parent surveys did not reveal characteristics correlated with increased comfort or desire to attend rounds. **CONCLUSIONS:** A majority of parents wish to participate in morning rounds, whereas healthcare provider opinions are mixed. Important discrepancies exist between parent and healthcare provider perceptions of parental comfort and comprehension on rounds, which may be important in facilitating parental presence.

<http://www.ncbi.nlm.nih.gov/pubmed/24215826>

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## Daily life physical activity in long-term survivors of nephroblastoma and neuroblastoma

van Waas, M., et al.

*J Pediatr Hematol Oncol* 2013; 35(5): 361-365

The risk of metabolic late effects after childhood cancer, such as obesity, hypertension, and diabetes, can be positively influenced by a healthy lifestyle with sufficient physical activity. Nevertheless, studies on physical activity in adult survivors of childhood cancer are scarce and involve different and often nonvalidated questionnaires. We used the Short Questionnaire to Assess Health-enhancing physical activity (SQUASH), which was developed and validated to assess daily life physical activity in the Dutch adult population. The aim of the study was to assess daily life physical activity in Dutch adult long-term nephroblastoma and neuroblastoma survivors. Sixty-seven nephroblastoma and 36 neuroblastoma survivors (median age, 30 y; range, 18 to 51 y) and 60 sociodemographically similar healthy control subjects (median age, 32 y; range, 18 to 62 y) were asked to complete the SQUASH during their regular follow-up visit. The adjusted mean physical activity score in male neuroblastoma survivors (mean, 7155;  $P=0.004$ ) was significantly lower than in male controls (mean, 10,574), whereas it was not significantly lower in male nephroblastoma survivors (mean, 9122;  $P=0.108$ ). Adjusted means for physical activity scores in females were not different from their controls. In conclusions, male neuroblastoma survivors were identified as performing less daily physical activity.

<http://www.ncbi.nlm.nih.gov/pubmed/23389502>

## Tough courage: Oncology Nursing Forum addresses childhood cancer then and now

Rishel, C. J.

*Oncol Nurs Forum* 2013; 40(4): 308-310

Cancer is a devastating diagnosis for anyone, but none more so than for children and their parents-- so many questions to be asked, so much information to sift through and absorb, and so many difficult decisions to be made. It is no wonder that a diagnosis of childhood cancer is often met with fear, anger, guilt, and feelings of being overwhelmed, yet also a determined resilience on the part of families to do whatever it takes to help their child get well again (Rishel, 2010).

<http://www.ncbi.nlm.nih.gov/pubmed/23803262>

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## It is life threatening but I don't mind: A qualitative study using photo elicitation interviews to explore adolescents' experiences of renal replacement therapies

Wells, F., et al.

*Child Care Health Dev* 2013; 39(4): 602-612

BACKGROUND: Renal replacement therapy (RRT) transforms the life prospects of young people with established renal failure. However, these treatments can have significant physiological and psychological implications for adolescents as they prepare to transition into adulthood. Health policies increasingly emphasize children and youth's active participation and consultation as users of health services, yet studies infrequently seek their experiences directly. METHODS: Adolescents receiving RRT in a large UK teaching hospital took photographs illustrating the impact of their condition and treatment on their lives. Qualitative photo elicitation interviews were conducted to explore the significance of the images and the young person's experiences. Interviews were analysed using descriptive thematic analysis. RESULTS: Ten young people aged 13-17 years participated. Themes identified were: (1) understanding and acceptance of treatment; (2) living in a non-functioning body; (3) impact upon daily life; (4) sources of support. Young people found treatments challenging and experienced significant impact on relationships and daily routines. Yet, health was prioritized over body image and participants demonstrated great emotional resilience. Young people valued support from family and friends, although were wary of disclosing their condition in case it resulted in being highlighted as different. Young people reported hospital staff as being caring and professional, but their biggest virtue appeared to be their willingness to treat the young people as 'normal'. CONCLUSIONS: Young people engaged readily with the research, and frankly described the impact of RRT on their everyday lives. Service providers must ensure that adolescents' developmental needs are met as traditional tasks of adolescence may lose priority. However, it is also clear that young people's ability to cope with treatments should not be underestimated.

<http://www.ncbi.nlm.nih.gov/pubmed/22676493>

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## Quality of palliative care in children with cancer in Lebanon

Abu-Saad Huijer, H., et al.

*J Med Liban* 2013; 61(4): 228-236

**OBJECTIVE:** There is a growing research interest in pediatric palliative care in Lebanon. To date the existing studies have focused on the perspective of parents of children with cancer. The purpose of this study was to evaluate the quality of life (QoL), symptom prevalence and management, functional ability, and the quality of care among children with cancer at the Children's Cancer Center of Lebanon. **METHODS:** A cross-sectional survey design was used. A convenience sample of 85 patients on therapy aged 7 to 18 years participated in the study between 2010 and 2011. Using face to face interviews, a combination of four instruments were administered in Arabic. **RESULTS:** The mean age of the participants was 12.5 years, with the majority having leukemia. Overall, the children had satisfactory health-related QoL, with the exception of the nausea and worry subscales, had no limitations in functional abilities, and were satisfied with the care that they received. In children between 7 to 12 years, the most common symptoms were lack of appetite, pain, and nausea, while adolescents between 13 to 18 years experienced lack of energy, irritability, and pain. Pain and nausea were the most frequently treated symptoms. **CONCLUSION:** Although the participants reported satisfactory QoL, yet symptom management was inadequate and mainly focused on treating the physical symptoms. It is recommended to provide both pharmacological and psychological interventions in order to alleviate symptom burden and hence improve QoL in children with cancer.

<http://www.ncbi.nlm.nih.gov/pubmed/24428078>

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## National variation in costs and mortality for leukodystrophy patients in US children's hospitals

Brimley, C. J., et al.

*Pediatr Neurol* 2013; 49(3): 156-162 e151

**BACKGROUND:** Inherited leukodystrophies are progressive, debilitating neurological disorders with few treatment options and high mortality rates. Our objective was to determine national variation in the costs for leukodystrophy patients and to evaluate differences in their care. **METHODS:** We developed an algorithm to identify inherited leukodystrophy patients in deidentified data sets using a recursive tree model based on International Classification of Disease, 9th Edition, Clinical Modification, diagnosis and procedure charge codes. Validation of the algorithm was performed independently at two institutions, and with data from the Pediatric Health Information System (PHIS) of 43 US children's hospitals, for a 7-year period between 2004 and 2010. **RESULTS:** A recursive algorithm was developed and validated, based on six International Classification of Disease, 9th Edition, Clinical Modification, codes and one procedure code that had a sensitivity up to 90% (range 61-90%) and a specificity up to 99% (range 53-99%) for identifying inherited leukodystrophy patients. Inherited leukodystrophy patients comprise 0.4% of admissions to children's hospitals and 0.7% of costs. During 7 years, these patients required \$411 million of hospital care, or \$131,000/patient. Hospital costs for leukodystrophy patients varied at different institutions, ranging from two to 15 times more than the average pediatric patient. There was a statistically significant correlation between higher volume and increased cost efficiency. Increased mortality rates had an inverse relationship with increased patient volume that was not statistically significant. **CONCLUSIONS:** We developed and validated a code-based algorithm for identifying leukodystrophy patients in deidentified national datasets. Leukodystrophy patients account for \$59 million of costs yearly at children's hospitals. Our data highlight potential to reduce unwarranted variability and improve patient care.

<http://www.ncbi.nlm.nih.gov/pubmed/23953952>

## Ventilatory function in children with severe motor disorders using night-time postural equipment

Dawson, N. C., et al.

*Dev Med Child Neurol* 2013; 55(8): 751-757

AIM: Night-time postural equipment (NTPE) can prevent hip subluxation in children with severe motor disorders (SMDs). However, it is unclear how it affects ventilatory function. The aims of the study were to determine how NTPE use affects ventilatory function and to compare night-to-night variability of ventilatory function in children with SMDs and typically developing healthy children. METHOD: Fifteen NTPE users (six males, nine females), aged 1 to 19 years (mean age 8y 7mo) alternated sleep condition between NTPE and sleeping unsupported for 14 nights. In all but two participants, gross motor function was classified as Gross Motor Function Classification System (GMFCS) level V; in the other two it was level IV. Oxyhaemoglobin saturation (SpO<sub>2</sub>) was monitored each night and transcutaneous CO<sub>2</sub> (PtcCO<sub>2</sub>) for one night in each sleep condition. In 17 healthy children of similar age, home SpO<sub>2</sub> only was monitored for seven nights. RESULTS: In 13 of 15 NTPE users and 12 of the 17 typically developing children, SpO<sub>2</sub> monitoring was satisfactorily completed. Of the children with SMDs, two had mean SpO<sub>2</sub> levels below the treatment threshold for supplemental oxygen, which was uniquely associated with use of NTPE in only one participant, and three had nocturnal hypoventilation, which was uniquely associated with NTPE use in only one case. Night-to-night SpO<sub>2</sub> variability was higher in children with SMDs than in typically developing children. INTERPRETATION: NTPE may impair or enhance ventilatory function in a minority of children. Owing to night-to-night variability in SpO<sub>2</sub>, at least three nights of monitoring are recommended to determine optimal positioning for effective ventilation before and after NTPE introduction.

<http://www.ncbi.nlm.nih.gov/pubmed/23582011>

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## Pediatric palliative care programs in children's hospitals: A cross-sectional national survey

Feudtner, C., et al.

*Pediatrics* 2013; 132(6): 1063-1070

BACKGROUND: Pediatric palliative care (PPC) programs facilitate the provision of comprehensive care to seriously ill children. Over the past 10 years many such programs have been initiated by children's hospitals, but little is known about their number, staff composition, services offered, sources of support, or national distribution. METHODS: In the summer of 2012, we surveyed 226 hospitals as identified by the National Association of Children's Hospitals and Related Institutions. The survey instrument gathered data about whether their institution had a PPC program, and for hospitals with programs, it asked for a wide range of information including staffing, patient age range, services provided, and financial support. RESULTS: Of the 162 hospitals that provided data (71.7% response rate), 69% reported having a PPC program. The rate of new program creation peaked in 2008, with 12 new programs created that year, and 10 new programs in 2011. Most programs offer only inpatient services, and most only during the work week. The number of consults per year varied substantially across programs, and was positively associated with hospital bed size and number of funded staff members. PPC programs report a high level of dependence on hospital funding. CONCLUSIONS: PPC programs are becoming common in children's hospitals throughout the United States yet with marked variation in how these programs are staffed, the level of funding for staff effort to provide PPC, and the number of consultations performed annually. Guidelines for PPC team composition, funding, and consultation standards may be warranted to ensure the highest quality of PPC.

<http://www.ncbi.nlm.nih.gov/pubmed/24190689>

## Correlates of care for young men with Duchenne and Becker muscular dystrophy

Andrews, J. G., et al.

*Muscle Nerve* 2014; 49(1): 21-25

INTRODUCTION: In progressive conditions, such as Duchenne and Becker muscular dystrophy (DBMD), the need for care may outpace care use. We examined correlates that contribute to utilization of needed care. METHODS: Structured interviews were conducted on use of care among 34 young men with DBMD who were born before 1982. RESULTS: Disease severity, per capita income, and presence of other relatives with DBMD predicted greater use of services. Race/ethnicity, acculturation, and level of caregiver education did not significantly predict service utilization. CONCLUSIONS: We identified disparities in receipt of healthcare and related services in adult men with DBMD that can affect quality of life. Despite the high disease severity identified in this population, these men utilized only half of the services available to individuals with significant progressive conditions. Providers should be aware of low service utilization and focus on awareness and assistance to ensure access to available care.

<http://www.ncbi.nlm.nih.gov/pubmed/23558904>

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## Recipients of electric-powered indoor/outdoor wheelchairs provided by a national health service: A cross-sectional study

Frank, A. O. and L. H. De Souza

*Arch Phys Med Rehabil* 2013; 94(12): 2403-2409

OBJECTIVE: To describe the characteristics across all ages of powered wheelchair users and the assistive technology prescribed by a regional specialist wheelchair service. DESIGN: Cross-sectional study. SETTING: Regional wheelchair service. PARTICIPANTS: Electric-powered indoor/outdoor wheelchair (EPIOC) users (N=544) with 262 boys and men (mean age +/- SD, 41.7 +/- 20.7y; range, 8-82y) and 282 girls and women (mean age +/- SD, 47.2 +/- 19.7y; range, 7-92y). INTERVENTIONS: Not applicable. MAIN OUTCOME MEASURES: Demographic, clinical/diagnostic details of EPIOC recipients, including pain, (kypho) scoliosis, and ventilators. Technical features, including specialized (adaptive) seating, tilt in space, and modified control systems. Factors were related to age groups: 1 (0-15y), 2 (16-24y), 3 (25-54y), 4 (55-74y), and 5 (>=75y). RESULTS: Neurologic/neuromuscular conditions predominated (81%) with cerebral palsy (18.9%) and multiple sclerosis (16.4%). Conditions presenting at birth or during childhood constituted 39%. Of the participants, 99 had problematic pain, 83 had (kypho) scoliosis, and 11 used ventilators. Specialized (adaptive) seating was provided to 169 users (31%); most had cerebral palsy or muscular dystrophy. Tilt in space was used by 258 (53%) participants. Younger people were more likely to receive tilt in space than older ones. Only 92 had specialized (adaptive) seating and tilt in space (mean age +/- SD, 29 +/- 17.8y; range, 8-72y). Of the participants, 52 used modified control systems. CONCLUSIONS: The diversity of EPIOC users across age and diagnostic groups is shown. Their complex interrelations with these technical features of EPIOC prescriptions are explored. Younger users were more complex because of age-related changes. This study provides outcomes of the EPIOC prescription for this heterogeneous group of very severely disabled people.

<http://www.ncbi.nlm.nih.gov/pubmed/23891668>

## Transitional services for adolescents with epilepsy in the U.K.: A survey

Iyer, A. and R. Appleton

*Seizure* 2013; 22(6): 433-437

**PURPOSE:** To survey the current transitional epilepsy services in tertiary paediatric neurology centres in the UK within the principles of transitional care for young people with epilepsy. **METHODS:** An online web-based questionnaire was sent to the lead epilepsy clinicians in tertiary paediatric neurology centres on behalf of the British Paediatric Epilepsy Group, the specialist epilepsy group of the British Paediatric Neurology Association (BPNA). A transition clinic was defined as a 'clinic or service that provided joint paediatric and adult supervision of care from paediatric to adult services'. **RESULTS:** Twenty-three centres were approached of which 18 responded and 15 of which provided auditable data. The clinics were held between three and 12 times per year, mostly in the afternoon and sited equally between the paediatric and adult centre. Approximately three to five new, and three to eight follow up patients were seen in each clinic. Most clinics accepted new referrals with a minimum age of 14 and a maximum of 20 years. Most young people were seen only once in a transition clinic before then being promoted into the adult epilepsy service. Very few clinics accepted direct referrals from the GP. Adult, slightly more than the paediatric team provided out-of-hospital advice after the young person was seen in the transition clinic. **CONCLUSIONS:** Young people with epilepsy are a challenging, but interesting group and their care at this time may have a potentially irreversible impact on their life. Their progress from paediatric to adult services should be a dynamic, gradual and smoothly transitioned process to optimise their care. Although recommended by the National Institute for Health and Clinical Excellence (NICE) and the National Services Framework (NSF), the findings of this survey would suggest an un-met need of this population.

<http://www.ncbi.nlm.nih.gov/pubmed/23498777>

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## Facilitating pediatric patient-provider communications using wireless technology in children and adolescents with sickle cell disease

Jacob, E., et al.

*J Pediatr Health Care* 2013; 27(4): 284-292

**INTRODUCTION:** Use of wireless devices has the potential to transform delivery of primary care services for persons with sickle cell disease (SCD). The study examined text message communications between patients and an advanced practice registered nurse (APRN) and the different primary care activities that emerged with use of wireless technology. **METHODS:** Patients (N = 37; mean age 13.9 +/- 1.8 years; 45.9% male and 54.1% female) engaged in intermittent text conversations with the APRN as part of the Wireless Pain Intervention Program. Content analyses were used to analyze the content of text message exchanges between patients and the APRN. **RESULTS:** The primary care needs that emerged were related to pain and symptom management and sickle cell crisis prevention. Two primary care categories (collaborating and coaching), four primary care subcategories (screening, referring, informing, and supporting), and 16 primary care activities were evident in text conversations. **DISCUSSION:** The use of wireless technology may facilitate screening, prompt management of pain and symptoms, prevention or reduction of SCD-related complications, more efficient referral for treatments, timely patient education, and psychosocial support in children and adolescents with SCD.

<http://www.ncbi.nlm.nih.gov/pubmed/22446036>

## Life care planning for the child with cerebral palsy

Katz, R. T. and C. B. Johnson

*Phys Med Rehabil Clin N Am* 2013; 24(3): 491-505

A life care plan may be useful to plan the needs of the disabled child with cerebral palsy. A cost analysis for a life care plan depends on the life expectancy of the child, and careful review of the needs of the child. A wide variety of support services may be available in the public sector. Key physical disabilities are associated with diminished life span, as are diminished cognitive abilities, even in the absence of physical impairment. The life care plan must follow the generally accepted and peer-reviewed methodology, with an appropriate foundation for each item recommended.

<http://www.ncbi.nlm.nih.gov/pubmed/23910487>

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## Developing a palliative care service for children in the Queen Elizabeth Central Hospital, Blantyre, Malawi

Molyneux, E. M., et al.

*Arch Dis Child* 2013; 98(9): 698-701

There are too few palliative care services for children in resource poor countries. Health carers are overwhelmed with cases of acute illness that need their urgent attention, and chronically ill children with life-limiting diseases have been sidelined. The HIV epidemic in southern Africa revealed the huge needs in our own hospital, and in 2002, we started a hospital-based paediatric palliative care service. It was the first in Africa. We describe here how it developed and expanded in the ensuing years and how it has affected our staff, the children and their families in our care.

<http://www.ncbi.nlm.nih.gov/pubmed/23898158>

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## Lost in transition: Child to adult cancer services for young people

McInally, W.

*Br J Nurs* 2013; 22(22): 1314-1318

Cancer nursing care across the UK has dramatically improved for children and young people with cancer over the past 20 years (Department of Health, 2007). Around 70% of young people diagnosed with cancer survive into adulthood, albeit with long-term health complications (Scottish Government, 2012). This raises the contemporary concern of how best to transition these patients to an adult-focused care regime (National Cancer Survivorship Initiative, 2012). With support from a Florence Nightingale Foundation Travel Scholarship in 2012, this study compared the various transition models currently in use across the UK, Finland and the USA with a clear focus on individual patient choice, staff education and preparation to care for this group of patients and their families. The findings revealed wide discrepancy in current nursing practices across the globe. This article presents a series of findings and recommendations to improve further the overall cancer experience for young people living with and beyond a cancer diagnosis.

<http://www.ncbi.nlm.nih.gov/pubmed/24335869>

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## Differences in characteristics of dying children who receive and do not receive palliative care

Keele, L., et al.

*Pediatrics* 2013; 132(1): 72-78

OBJECTIVE: Comparing demographic and clinical characteristics associated with receipt of palliative care (PC) among children who died in children's hospitals to those who did not receive PC and understanding the trends in PC use. METHODS: This retrospective cohort study used the Pediatric Health Information System database. Children <18 years of age who died  $\geq 5$  days after admission to a Pediatric Health Information System hospital between January 1, 2001, and December 31, 2011 were included. Receipt of PC services was identified by the International Classification of Diseases, Ninth Revision code for PC. Diagnoses were grouped using major diagnostic codes. International Classification of Diseases codes and clinical transaction codes were used to evaluate all interventions. RESULTS: This study evaluated 24 342 children. Overall, 4% had coding for PC services. This increased from 1% to 8% over the study years. Increasing age was associated with greater receipt of PC. Children with the PC code had fewer median days in the hospital (17 vs 21), received fewer invasive interventions, and fewer died in the ICU (60% vs 80%). Receipt of PC also varied by major diagnostic codes, with the highest proportion found among children with neurologic disease. CONCLUSIONS: Most pediatric patients who died in a hospital did not have documented receipt of PC. Children receiving PC are different from those who do not in many ways, including receipt of fewer procedures. Receipt of PC has increased over time; however, it remains low, particularly among neonates and those with circulatory diseases.

<http://www.ncbi.nlm.nih.gov/pubmed/23753086>

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## Best practices for pediatric palliative cancer care: A primer for clinical providers

Levine, D., et al.

*J Support Oncol* 2013; 11(3): 114-125

Cancer is the leading cause of disease-related death in children and adolescents. Pediatric patients with cancer suffer greatly at the end of life. However, palliative care interventions can reduce suffering and significantly improve the care of these patients and their families. A large percentage of pediatric deaths occur outside of the hospital setting where pediatric palliative resources may not be readily available. Patients in the home setting may be cared for by community hospice programs, which are typically staffed for adult populations. Increasingly, nonpediatric providers are asked to provide palliative care for children and adolescents at the end of life, yet they receive little formal training in this area. This review focuses on the principles of best practice in the provision of palliative care for children and adolescents with cancer. Our intent is to aid clinical providers in delivering optimal care to this patient population. Topics unique to pediatric palliative care that are addressed include: providing pain and symptom management in the broad pediatric range from neonate to adolescent; caring for and interacting with developmentally distinct groups; engaging in shared decision making with parents and adolescents; providing accommodations for prognoses that are often more uncertain than in adult patients; and delivering concurrent disease-directed therapy with palliative care.

<http://www.ncbi.nlm.nih.gov/pubmed/24400391>

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## Who receives home-based perinatal palliative care: Experience from Poland

Korzeniewska-Eksterowicz, A., et al.

*Biomed Res Int* 2013; 652321

CONTEXT: The current literature suggests that perinatal palliative care (PPC) programs should be comprehensive, initiated early, and integrative. So far there have been very few publications on the subject of home-based PC of newborns and neonates. Most publications focus on hospital-based care, mainly in the neonatal intensive care units. OBJECTIVE: To describe the neonates and infants who received home-based palliative care in Lodz Region between 2005 and 2011. METHODS: A retrospective review of medical records. RESULTS: 53 neonates and infants were admitted to a home hospice in Lodz Region between 2005 and 2011. In general, they are a growing group of patients referred to palliative care. Congenital diseases (41%) were the primary diagnoses; out of 53 patients 16 died, 20 were discharged home, and 17 stayed under hospice care until 2011. The most common cause of death (56%) was cardiac insufficiency. Neurological symptoms (72%) and dysphagia (58%) were the most common clinical problems. The majority of children (45%) had a feeding tube inserted and were oxygen dependent (45%); 39 families received psychological care and 31 social supports. CONCLUSIONS: For terminally ill neonates and infants, perinatal palliative care is an option which improves the quality of their lives and provides the family with an opportunity to say goodbye.

<http://www.ncbi.nlm.nih.gov/pubmed/24083234>

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## Scaling up paediatric HIV care with an integrated, family-centred approach: An observational case study from Uganda

Luyirika, E., et al.

*PLoS One* 2013; 8(8): e69548

Family-centred HIV care models have emerged as an approach to better target children and their caregivers for HIV testing and care, and further provide integrated health services for the family unit's range of care needs. While there is significant international interest in family-centred approaches, there is a dearth of research on operational experiences in implementation and scale-up. Our retrospective case study examined best practices and enabling factors during scale-up of family-centred care in ten health facilities and ten community clinics supported by a non-governmental organization, Mildmay, in Central Uganda. Methods included key informant interviews with programme management and families, and a desk review of hospital management information systems (HMIS) uptake data. In the 84 months following the scale-up of the family-centred approach in HIV care, Mildmay experienced a 50-fold increase of family units registered in HIV care, a 40-fold increase of children enrolled in HIV care, and nearly universal coverage of paediatric cotrimoxazole prophylaxis. The Mildmay experience emphasizes the importance of streamlining care to maximize paediatric capture. This includes integrated service provision, incentivizing care-seeking as a family, creating child-friendly service environments, and minimizing missed paediatric testing opportunities by institutionalizing early infant diagnosis and provider-initiated testing and counselling. Task-shifting towards nurse-led clinics with community outreach support enabled rapid scale-up, as did an active management structure that allowed for real-time review and corrective action. The Mildmay experience suggests that family-centred approaches are operationally feasible, produce strong coverage outcomes, and can be well-managed during rapid scale-up.

<http://www.ncbi.nlm.nih.gov/pubmed/23936337>

## Long-term ventilation in children: Longitudinal trends and outcomes

McDougall, C. M., et al.

*Arch Dis Child* 2013; 98(9): 660-665

**BACKGROUND:** Cross-sectional studies have suggested a rapid expansion in paediatric long-term ventilation (LTV) over the last 20 years but information on longitudinal trends is limited. **METHODS:** Data were collected prospectively on all patients receiving LTV over a 15-year period (1.1.95-31.12.09) in a single regional referral centre. **RESULTS:** 144 children commenced LTV during the 15-year period. The incidence of LTV increased significantly over time, with an accompanying 10-fold increase in prevalence due to a significant increase in institution of non-invasive ventilation (NIV). There was no significant increase in invasive ventilation. 5-year survival was 94% overall and was significantly higher for patients on NIV (97%) than invasively ventilated patients (84%). 10-year survival was 91% overall. Although some children were able to discontinue respiratory support (21% at 5 years and 42% at 10 years), the number of patients transitioned to adult services increased significantly over time (26% of total cohort). Patients with neuromuscular disease were less likely to discontinue support than other patients. **CONCLUSIONS:** The paediatric LTV population has expanded significantly over 15 years. Future planning of paediatric hospital and community services, as well as adult services, must take into account the needs of this growing population.

<http://www.ncbi.nlm.nih.gov/pubmed/23838128>

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## Bereavement photography for children: Program development and health care professionals' response

Michelson, K. N., et al.

*Death Stud* 2013; 37(6): 513-528

Reports of in-hospital bereavement photography focus largely on stillborns and neonates. Empiric data regarding the implementation of bereavement photography in pediatrics beyond the neonatal period and the impact of such programs on healthcare professionals (HCPs) is lacking. The authors describe the implementation of a pediatric intensive care unit (PICU) bereavement photography program and use questionnaire data from HCPs to describe HCPs' reflections on the program and to identify program barriers. From July 2007 through April 2010, families of 59 (36%) of the 164 patients who died in the PICU participated in our bereavement photography program. Forty questionnaires from 29 HCPs caring for 39 participating patients/families indicated that families seemed grateful for the service (n = 34; 85%) and that the program helped HCPs feel better about their role (n = 30; 70%). Many HCPs disagreed that the program consumed too much of his/her time (n = 34; 85%) and that the photographer made his/her job difficult (n = 37; 92.5%). Qualitative analysis of responses to open-ended questions revealed 4 categories: the program's general value; positive aspects of the program; negative aspects of the program; and suggestions for improvements. Implementing bereavement photography in the PICU is feasible though some barriers exist. HCPs may benefit from such programs.

<http://www.ncbi.nlm.nih.gov/pubmed/24520925>

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## Racial and ethnic differences in hospice enrolment among children with cancer

Thienprayoon, R., et al.

*Pediatr Blood Cancer* 2013; 60(10): 1662-1666

**BACKGROUND:** Hospice is an important provider of end of life care. Adult minorities are less likely to enrol on hospice; little is known regarding the prevalence of pediatric hospice use or the characteristics of its users. Our primary objective was to determine whether race/ethnicity was associated with hospice enrolment in children with cancer. We hypothesized that minority (Latino) race/ethnicity is negatively associated with hospice enrolment in children with cancer. **PROCEDURE:** In this single-center retrospective cohort study, inclusion criteria were patients who died of cancer or stem cell transplant between January 1, 2006 and December 31, 2010. The primary outcome variable was hospice enrolment and primary predictor was race/ethnicity. **RESULTS:** Of the 202 patients initially identified, 114 met inclusion criteria, of whom 95 were enrolled on hospice. Patient race/ethnicity was significantly associated with hospice enrolment ( $P = 0.02$ ), the association remained significant ( $P = 0.024$ ) after controlling for payor status ( $P = 0.995$ ), patient diagnosis ( $P = 0.007$ ), or religion ( $P = 0.921$ ). Latinos enrolled on hospice significantly more often than patients of other races. Despite initial enrollment on hospice however, 34% of Latinos and 50% of non-Latinos had withdrawn from hospice at the time of death ( $P = 0.10$ ). Race/ethnicity was not significantly associated with dying on hospice. **CONCLUSIONS:** These results indicate that race/ethnicity and diagnosis are likely to play a role in hospice enrollment during childhood. A striking number of patients of all race/ethnicities left hospice prior to death. More studies describing the impact of culture on end of life decision-making and the hospice experience in childhood are warranted.

<http://www.ncbi.nlm.nih.gov/pubmed/23733549>

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