Staff Publications 2013

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This list is updated regularly.

Articles are listed alphabetically by first author.

Ski-interacting protein (SKIP) interacts with androgen receptor in the nucleus and modulates androgen-dependent transcription.
BACKGROUND: The androgen receptor (AR) is a member of the nuclear receptor (NR) superfamily of ligand-inducible DNA transcription factors, and is the major mediator of male sexual development, prostate growth and the pathogenesis of prostate cancer. Cell and gene specific regulation by the AR is determined by availability of and interaction with sets of key accessory cofactors. Ski-interacting protein (SKIP; SNW1, NCOA62) is a cofactor shown to interact with several NRs and a diverse range of other transcription factors. Interestingly, SKIP as part of the spliceosome is thought to link mRNA splicing with transcription. SKIP has not been previously shown to interact with the AR. RESULTS: The aim of this study was to investigate whether SKIP interacts with the AR and modulates AR-dependent transcription. Here, we show by co-immunoprecipitation experiments that SKIP is in a complex with the AR. Moreover, SKIP increased 5alpha-dihydrotestosterone (DHT) induced N-terminal/C-terminal AR interaction from 12-fold to almost 300-fold in a two-hybrid assay, and enhanced AR ligand-independent AF-1 transactivation. SKIP augmented ligand- and AR-dependent transactivation in PC3 prostate cancer cells. Live-cell imaging revealed a fast (half-time=129 s) translocation of AR from the cytoplasm to the nucleus upon DHT-stimulation. Forster resonance energy transfer (FRET) experiments suggest a direct AR-SKIP interaction in the nucleus upon translocation. CONCLUSIONS: Our results suggest that SKIP interacts with AR in the nucleus and enhances AR-dependent transactivation and N/C-interaction supporting a role for SKIP as an AR co-factor.

Adams C-l and Gill FJ
Co-bedding of multiples in the neonatal unit: Assessing nurses and midwives attitude and level of understanding.

Addo NK, Javadpour S, Kandasamy J, Sillifant P, May P and Sinha A
Central sleep apnea and associated Chiari malformation in children with syndromic craniosynostosis: treatment and outcome data from a supraregional national craniofacial center: Clinical article.

Agarwal A and Shipman PJ
Gallbladder polyposis in metachromatic leukodystrophy.
Gallbladder polyposis is a rare entity that can be associated with conditions such as metachromatic leukodystrophy (MLD), but the literature is sparse. We present a child with gallbladder polyposis who was diagnosed with MLD 15 months later despite normal neuroimaging and clinical examination initially.

Alblooshi AS, Simpson SJ, Stick SM and Hall GL
The safety and feasibility of the inhaled mannitol challenge test in young children.
Young children can complete a mannitol challenge and this may lead to improvements in our ability to diagnose exercise related asthma in this age group.

BACKGROUND: Existing studies of childhood dilated cardiomyopathy deal mainly with early survival. This population-based study examines long-term outcomes for children with dilated cardiomyopathy. METHODS AND RESULTS: The diagnosis of dilated cardiomyopathy was based on clinical, echocardiographic, and pathological findings. The primary study end point included time to the combined outcome of death or cardiac transplantation. There were 175 patients 0 to <10 years of age at the time of diagnosis. Survival free from death or transplantation was 74% (95% confidence interval, 67-80) 1 year after diagnosis, 62% (95% confidence interval, 55-69) at 10 years, and 56% (95% confidence interval, 46-65) at 20 years. In multivariable analysis, age at diagnosis <4 weeks or ≥5 years, familial cardiomyopathy, and lower baseline left ventricular fractional shortening Z score were associated with increased risk of death or transplantation, as well as lower left ventricular fractional shortening Z score during follow-up. At 15 years after diagnosis, echocardiographic normalization had occurred in 69% of surviving study subjects. Normalization was related to higher baseline left ventricular fractional shortening Z score, higher left ventricular fractional shortening Z score during follow-up, and greater improvement in left ventricular fractional shortening Z score. Children with lymphocytic myocarditis had better survival and a higher rate of echocardiographic normalization. At the latest follow-up, 100 of 104 of survivors (96%) were free of cardiac symptoms, and 83 (80%) were no longer receiving pharmacotherapy. CONCLUSIONS: Death or transplantation occurred in 26% of patients with childhood dilated cardiomyopathy within 1 year of diagnosis and ~1% per year thereafter. Risk factors for death or transplantation include age at diagnosis, familial cardiomyopathy, and severity of left ventricular dysfunction. The majority of surviving subjects are well and free of cardiac medication.


OBJECTIVES:: Through evidence review and the consensus of an expert panel, we developed recommendations for the clinical management of gastroesophageal reflux disease, constipation and abdominal bloating in Rett syndrome. METHODS:: Based upon review of the literature and family concerns expressed on RettNet, initial draft recommendations were created. Where the literature was lacking twenty five open-ended questions were included. Input from an international, multi-disciplinary panel of clinicians was sought using a 2-stage modified Delphi process to reach consensus agreement. Items related to the clinical assessment and management of gastroesophageal reflux disease, constipation and abdominal bloating. RESULTS:: Consensus was achieved on 78/85 statements. A comprehensive approach to the assessment of gastroesophageal reflux and reflux disease, constipation and abdominal bloating was recommended taking into account impairment of communication skills in Rett syndrome. A stepwise approach to management was identified with initial use of conservative strategies escalating to pharmacological measures and surgery if necessary. CONCLUSIONS:: Gastrointestinal dysmotility occurs commonly in Rett syndrome. These evidence- and consensus-based recommendations have the potential to improve care of dysmotility issues in a rare condition and stimulate research to improve the current limited evidence base.


AIM: This study aims to analyse the continuous relationship of each cardiometabolic risk factor with body mass index (BMI) and waist circumference percentiles in a population-based sample of children. METHODS: A cross-sectional sample of 996 school children aged 6-16.9 years in Busselton, Western Australia, (2005-2007) had
anthropometry and fasting blood tests for total cholesterol, high density lipoprotein, low density lipoprotein, triglycerides, glucose, insulin, high-sensitive C-reactive protein, liver function tests and adiponectin. Age- and menarche (for girls)-adjusted means of each risk factor were related to BMI and waist circumference centiles across the full normal-overweight-obese range. RESULTS: The correlations between BMI and waist circumference (boys 0.91 and girls 0.91) and between BMI z-score and waist z-score (boys 0.80 and girls 0.82) were high. An increase in insulin across all centile groups (for BMI and waist circumference) was found in both sexes. An increase was found for diastolic blood pressure and systolic blood pressure z-score, high density lipoprotein, high-sensitive C-reactive protein, alanine transaminase and gamma-glutamyltransferase in only the centile groups >85% for BMI and waist circumference for both sexes. Mixed and sex-discordant results were found for triglycerides, adiponectin and glucose. CONCLUSION: There are important differences in the relationships between increasing BMI/adiposity, and each comorbidity and these relationships can differ between boys and girls. This information has implications for screening and management of adiposity-related cardiometabolic risk factors in children and for public health initiatives to reduce future burden of cardiovascular disease.

Brazzale DJ, Hall GL and Pretto JJ. 
Effects of adopting the new Global Lung Function Initiative 2012 reference equations on the interpretation of spirometry. 

Brennan C, Newton M, Wood F, Schug SA, Allsop S and Browne AL. 
Training general practitioners in remote Western Australia in a method of screening and brief intervention for harmful alcohol use: A pilot study. 

OBJECTIVE: High levels of alcohol-related harm are a salient feature of many rural communities in Australia. General practitioners (GPs) are uniquely placed to identify and treat patients with harmful alcohol use in remote settings, yet corresponding opportunities for education in effective brief psychological interventions for harmful alcohol use are limited. This study piloted a training model for alcohol screening and brief intervention for GPs working in Kalgoorlie-Boulder, a remote Western Australian community facing significant alcohol-related problems. DESIGN: Observational pilot study. SETTING: Primary care. MAIN OUTCOME MEASURE(S): Perceived role in responding to harmful alcohol use, and confidence and knowledge of alcohol screening and brief intervention; satisfaction with a short training session focused on alcohol screening and brief intervention; and impact of training on implementation of screening and brief intervention for harmful alcohol use. RESULTS: Fifty per cent of GPs took up the training opportunity. GPs recognised their professional responsibility for conducting brief intervention but reported comparatively lower confidence and skills in implementing screening and intervention prior to training. The training improved knowledge and confidence in conducting alcohol screening and brief intervention. All GPs increased their frequency of alcohol screening, and 88% of GPs reported increasing the frequency of brief intervention at 6 months. CONCLUSIONS: Preliminary findings suggest that among participating GPs, subsequent compliance with identification and management of harmful alcohol use was improved. Further work examining methods to improve rural and remote GP participation in alcohol-related harm prevention training is required, as the potential impact on communities with disproportionately high alcohol-related difficulties is significant.

Respiratory impedance and bronchodilator responsiveness in healthy children aged 2–13 years. 

Cameron F, Cotterill A, Couper J, Craig M, Davis E, Donaghue K, Jones T, King B and Shell B. 
Short report: Care for children and adolescents with diabetes in Australia and New Zealand: Have we achieved the defined goals? 

Carapetis JR. 
Commentary on Remond et al. 

Carcione D, Blyth CC, Mak DB and Effler PV. 
User satisfaction with the Western Australian Vaccine Safety Surveillance (WAVSS) System. 

Chambers NA and Hullett B. Direct laryngoscopy after potential difficult intubation in children only predicts standard Cormack and Lehane view to within one grade. Paediatr Anaesth. 2013; 23(11): 1002-1005. BACKGROUND: Some techniques used to achieve intubation in children predicted to have a difficult airway do not involve direct laryngoscopy or assessment of the laryngeal grade. Direct laryngoscopy may therefore be performed immediately after intubation to provide a record for future anesthesists. It is unknown whether this postintubation grade accurately reflects the standard laryngeal grade in this group. AIM: The aim of the study was to identify those children who were predicted to be a difficult intubation and to perform direct laryngoscopy before and after intubation. We set out to ascertain if direct laryngoscopy performed after intubation could accurately predict the standard un-intubated laryngeal grade in this group. METHODS: All children presenting for general anesthesia who were clinically predicted to be a difficult intubation were considered for this study and prospectively recruited. After induction of anesthesia, one study anesthetist performed direct laryngoscopy before and another study anesthetist then performed direct laryngoscopy after intubation. These laryngeal grades were then compared. RESULTS: A total of 21 children were successfully recruited and studied, and all patients were successfully intubated. Overall, the postintubation grade did not reliably reflect the standard grade, but did not differ by more than one grade in any patient. In one-third of subjects, the postintubation grade was equal to the standard grade, in one-third it was a grade ‘easier’ and in one-third a grade ‘harder’. CONCLUSION: Assessment and documentation of a postintubation laryngeal grade does not appear to provide reliable information for future anesthesists and may even have the potential to be misleading. Any such documentation should always refer to the presence of an endotracheal tube and be interpreted with caution.


Cooper MN, O’Connell SM, Davis EA and Jones TW. A population-based study of risk factors for severe hypoglycaemia in a contemporary cohort of childhood-onset type 1 diabetes. Diabetologia. 2013; 56(10): 2164-2170. AIMS/HYPOTHESIS: Severe hypoglycaemia is a major barrier to optimising glycaemic control. Recent changes in therapy, however, may have altered the epidemiology of severe hypoglycaemia and its associated risk factors. The aim of this study was to examine the incidence rates and risk factors associated with severe hypoglycaemia in a contemporary cohort of children and adolescents with type 1 diabetes. METHODS: Subjects were identified from a population-based register containing data on >99% of patients (<16 years of age) who were being treated for type 1 diabetes in Western Australia. Patients attend the clinic approximately every 3 months, where data pertaining to diabetes management, demographics and complications including hypoglycaemia are prospectively recorded. A severe hypoglycaemic event was defined as an episode of coma or convulsion associated with hypoglycaemia. Risk factors assessed included age, duration of diabetes, glycaemic control, sex, insulin therapy, socioeconomic status and calendar year. RESULTS: Clinical visit data from 1,770 patients, providing 8,214 patient-years of data between 2000 and 2011 were analysed. During follow-up, 841 episodes of severe hypoglycaemia were observed. No difference in risk of severe hypoglycaemia was observed between age groups. Good glycaemic control (HbA1c <7% [53 mmol/mol]) compared with the
cohort average (HbA1c 8-9% [64-75 mmol/mol]) was not associated with an increased risk of severe hypoglycaemia. When compared with patients on injection regimens, subjects aged 12-18 years on pump therapy were at reduced risk of severe hypoglycaemia (incidence risk ratio 0.6; 95% CI 0.4, 0.9).

CONCLUSIONS/INTERPRETATION: In this population-based sample of children and adolescents with type 1 diabetes, contemporary therapy is associated with a changed pattern and incidence of severe hypoglycaemia.

Corscadden KJ, Kirkham L-AS, Thornton RB, Vijayasekaran S, Coates HL, Richmond PC and Wiertsema SP.

High pneumococcal serotype specific IgG, IgG1 and IgG2 levels in serum and the middle ear of children with recurrent acute otitis media receiving ventilation tubes.


Human rhinovirus species C infection in young children with acute wheeze is associated with increased acute respiratory hospital admissions.


RATIONALE: Human rhinovirus species C (HRV-C) is the most common cause of acute wheezing exacerbations in young children presenting to hospital, but its impact on subsequent respiratory illnesses has not been defined. OBJECTIVES: To determine whether acute wheezing exacerbations due to HRV-C are associated with increased hospital attendances due to acute respiratory illnesses (ARI). METHODS: Clinical information and nasal samples were collected prospectively from 197 children less than 5 years of age, presenting to hospital with an acute wheezing episode. Information on hospital attendances with an ARI before and after recruitment was subsequently obtained. MEASUREMENTS AND MAIN RESULTS: HRV was the most common virus identified at recruitment (n = 135 [68.5%]). From the 120 (88.9%) samples that underwent typing, HRV-C was the most common HRV species identified, present in 81 (67.5%) samples. Children with an HRV-related wheezing illness had an increased risk of readmission with an ARI (relative risk, 3.44; 95% confidence interval, 1.17-10.17; P = 0.03) compared with those infected with any other virus. HRV-C, compared with any other virus, was associated with an increased risk of a respiratory hospital admission before (49.4% vs. 27.3%, respectively; P = 0.004) and within 12 months (34.6% vs. 17.0%; P = 0.01) of recruitment. Risk for subsequent ARI admissions was further increased in atopic subjects (relative risk, 6.82; 95% confidence interval, 2.16-21.55; P = 0.001). Admission risks were not increased for other HRV species. CONCLUSIONS: HRV-C-related wheezing illnesses were associated with an increased risk of prior and subsequent hospital respiratory admissions. These associations are consistent with HRV-C causing recurrent severe wheezing illnesses in children who are more susceptible to ARIs.

Davey RJ, Bussau VA, Paramalingam N, Ferreira LD, Lim EM, Davis EA, Jones TW and Fournier PA.

A 10-s Sprint Performed After Moderate-Intensity Exercise Neither Increases nor Decreases the Glucose Requirement to Prevent Late-Onset Hypoglycemia in Individuals With Type 1 Diabetes.


Davey RJ, Howe W, Paramalingam N, Ferreira LD, Davis EA, Fournier PA and Jones TW.

The Effect of Midday Moderate-Intensity Exercise on Postexercise Hypoglycemia Risk in Individuals With Type 1 Diabetes.

J Clin Endocrinol Metab. 2013. Context:Exercise increases the risk of hypoglycemia in type 1 diabetes. Objective:Recently we reported a biphasic increase in glucose requirements to maintain euglycemia after late-afternoon exercise, suggesting a unique pattern of delayed risk for nocturnal hypoglycemia. This study examined whether this pattern of glucose requirements occurs if exercise is performed earlier in the day. Design, Participants, and Intervention: Ten adolescents with type 1 diabetes underwent a hyperinsulinemic euglycemic glucose clamp on 2 different occasions during which they either rested or performed 45 minutes of moderate-intensity exercise at midday. Glucose was infused to maintain euglycemia for 17 hours after exercise. Main Outcome Measures: The glucose infusion rate (GIR) to maintain euglycemia, glucose rates of appearance and disappearance, and levels of counterregulatory hormones were compared between conditions. Results: GIRs to maintain euglycemia were not significantly different between groups at baseline (9.8 +/- 1.4 and 9.5 +/- 1.6 g/h before the exercise and rest conditions, respectively) and did not change in the rest condition throughout the study. In contrast, GIR increased more than 3-fold during exercise (from 9.8 +/- 1.4 to 30.6 +/- 4.7 g/h), fell within the first hour of recovery, but remained elevated until 11 hours after exercise before returning to baseline levels. Conclusions: The pattern of glucose requirements to maintain euglycemia in response to moderate-intensity exercise performed at midday suggests that the risk of exercise-mediated hypoglycemia increases...
during and for several hours after moderate-intensity exercise, with no evidence of a biphasic pattern of postexercise risk of hypoglycemia.

Dawkins H, Watson HJ, Egan SJ and Kane RT.
Weight suppression in bulimia nervosa: Relationship with cognitive behavioral therapy outcome.

Duke J, Wood F, Semmens J, Edgar DW and Rea S.
Trends in hospital admissions for sunburn in Western Australia, 1988 to 2008.

Duncan CN, Riley TV, Carson KC, Budgeon CA and Siffleet J.
The effect of an acidic cleanser versus soap on the skin pH and micro-flora of adult patients: A non-randomised two group crossover study in an intensive care unit.

Egan SJ, Watson HJ, Kane RT, McEvoy P, Fursland A and Nathan PR.
Anxiety as a Mediator Between Perfectionism and Eating Disorders.

Evidence-based paramedic models of care to reduce unnecessary emergency department attendance--feasibility and safety.

Forbes D.
Mewling and puking: Infantile gastroesophageal reflux in the 21st century.
Infantile gastroesophageal reflux is challenging because of the difficulties in differentiating normal developmental physiology from disease, the imprecision of diagnostic tests and the paucity of evidence-based therapies for clinicians to implement. Careful clinical assessment is central to avoidance of inappropriate diagnosis and utilisation of ineffective or dangerous therapies, and at the same time, identification of those infants who warrant intervention.

Forbes D, Lim A and Ravikumara M.
Gastroesophageal reflux in the 21st century.
PURPOSE OF REVIEW: Gastroesophageal reflux (GER) remains a common, challenging problem for clinicians, with differentiation of normal development from disease a particular issue. This review updates clinicians on advances in diagnosis of GER, relationship to other problems, and current practice in management. RECENT FINDINGS: Development and understanding of multichannel intraluminal impedance-pH monitoring has given insights into the relationship of GER to symptoms. Medical treatment has changed little. Avoidance of overmedicalizing normal development is the major issue for clinicians. Laparoscopic fundoplication is established as equivalent to open fundoplication. Newer endoscopic techniques have only limited use in children to date. SUMMARY: Major changes in pediatric GER relate to understanding of physiology and relationship of GER to symptoms. The major challenge for clinicians involve differentiation of normal from abnormal GER, and applying the most relevant management.

Francis J, Anders M, Lobgeieir P and Nourse C.
Fatal Haemophilus influenzae type a sepsis in an infant.
Haemophilus influenzae type a can cause severe sepsis, as demonstrated by the case described. Epidemiology of sepsis in childhood is changing. Regardless of the pathogen involved, management of children with septic shock involves resuscitative measures and empiric antibiotics. The following case of H. influenzae type a sepsis proved fatal in spite of appropriate therapy.

Francis JR, Cherian S and Forbes D.
Seeking asylum: health and human rights in Australia.


Gibson N, Johnston K, Bear N, Stick S, Logie K and Hall G. Expiratory flow limitation and breathing strategies in overweight adolescents during sub maximal exercise. Int J Obes (Lond). 2013. ObjectiveTo investigate if ventilatory factors limit exercise in overweight and obese children during a six minute step test and to compare ventilatory responses during this test with those of healthy weight children. DesignCross sectional, prospective comparative study. Subjects26 overweight/obese subjects and 25 healthy weight subjects with no known respiratory illness. MeasurementsVarious fatness and fat distribution parameters are more likely to experience expiratory flow limitation during sub maximal exercise than their healthy weight peers [OR 7.2 (1.4,37.3), P<0.019]. Subjects who had lower lung volumes at rest, were even more likely to experience exercise induced expiratory flow limitations [OR 8.35 (1.4,49.3). Both groups displayed similar breathing strategies during submaximal exercise. ConclusionYoung people who are overweight/obese are more likely to display expiratory flow limitation during submaximal exercise than children of healthy weight. Use of compensatory breathing strategies appeared to enable overweight children to avoid the experience of breathlessness at this intensity of exercise. International Journal of Obesity accepted article preview online, 30 July 2013. doi:10.1038/ijo.2013.137.

Gill FJ, Leslie GD, Grech C and Latour JM. Health consumers' experiences in Australian critical care units: postgraduate nurse education implications. Nurs Crit Care. 2013; 18(2): 93-102. AIM: To explore critical care patients and families experiences and seek their input into nurses' postgraduate educational preparation and practice. BACKGROUND: There is an inconsistency in the expected standard of practice to 'qualify' Australian critical care nurses. There has also been a lack of health consumer input in the development of postgraduate course curriculum and content. METHOD: Following institutional ethics committee approval, purposive sampling was used to select participants for focus groups and individual interviews who had experienced intensive care or coronary care. FINDINGS: Seventeen participants provided data which created two main thematic categories; the role of the critical care nurse and; minimum practice standards for postgraduate critical care course graduates. Both physical patient care and socio-emotional support of patients and family were identified as important for the critical care nurse role. The level of socio-emotional support provided by nurses was reported to be inconsistent. Components of socio-emotional support included communication, people skills, facilitating family presence and advocacy. These components were reflected in participants' concepts of minimum practice standards for postgraduate critical care course graduates; talking and listening skills, relating to and dealing with stressed people, individualizing care and patient and family advocacy. CONCLUSION: Health consumers' views emphasize that socio-emotional skills and behaviours need to be explicitly described in postgraduate critical care nursing course curricula and instruments developed to consistently assess these core competencies.

Gill FJ, Leslie GD, Grech C and Latour JM. Using a web-based survey tool to undertake a Delphi study: Application for nurse education research. Nurse Educ Today. 2013; 33(11): 1322-1328. BACKGROUND: The Internet is increasingly being used as a data collection medium to access research participants. This paper reports on the experience and value of using web-survey software to conduct an eDelphi study to develop Australian critical care course graduate practice standards. METHODS: The eDelphi technique used involved the iterative process of administering three rounds of surveys to a national expert panel. The survey was developed online using SurveyMonkey. Panel members responded to statements using one rating scale for round one and two scales for rounds two and three. Text boxes for panel comments were provided. COLLECTING DATA AND PROVIDING FEEDBACK: For each round, the SurveyMonkey's email tool was used to distribute an individualized email invitation containing the survey web link. The distribution of panel responses, individual responses and a summary of comments were emailed to panel members. Stacked bar charts representing the distribution of responses were generated using the SurveyMonkey software. Panel response rates remained greater than 85% over all rounds. DISCUSSION: An online survey provided numerous
advantages over traditional survey approaches including high quality data collection, ease and speed of survey administration, direct communication with the panel and rapid collation of feedback allowing data collection to be undertaken in 12 weeks. Only minor challenges were experienced using the technology. Ethical issues, specific to using the Internet to conduct research and external hosting of web-based software, lacked formal guidance. CONCLUSIONS: High response rates and an increased level of data quality were achieved in this study using web-survey software and the process was efficient and user-friendly. However, when considering online survey software, it is important to match the research design with the computer capabilities of participants and recognize that ethical review guidelines and processes have not yet kept pace with online research practices.

Immunohistochemical, Ultrastructural and Functional Analysis of Axonal Regeneration through Peripheral Nerve Grafts Containing Schwann Cells Expressing BDNF, CNTF or NT3.  

Parent skills training treatment for parents of children and adolescents with eating disorders: A qualitative study.  

Medulloblastoma Down Under 2013: a report from the third annual meeting of the International Medulloblastoma Working Group.  
Medulloblastoma is curable in approximately 70 % of patients. Over the past decade, progress in improving survival using conventional therapies has stalled, resulting in reduced quality of life due to treatment-related side effects, which are a major concern in survivors. The vast amount of genomic and molecular data generated over the last 5-10 years encourages optimism that improved risk stratification and new molecular targets will improve outcomes. It is now clear that medulloblastoma is not a single-disease entity, but instead consists of at least four distinct molecular subgroups: WNT/Wingless, Sonic Hedgehog, Group 3, and Group 4. The Medulloblastoma Down Under 2013 meeting, which convened at Bunker Bay, Australia, brought together 50 leading clinicians and scientists. The 2-day agenda included focused sessions on pathology and molecular stratification, genomics and mouse models, high-throughput drug screening, and clinical trial design. The meeting established a global action plan to translate novel biologic insights and drug targeting into treatment regimens to improve outcomes. A consensus was reached in several key areas, with the most important being that a novel classification scheme for medulloblastoma based on the four molecular subgroups, as well as histopathologic features, should be presented for consideration in the upcoming fifth edition of the World Health Organization's classification of tumours of the central nervous system. Three other notable areas of agreement were as follows: (1) to establish a central repository of annotated mouse models that are readily accessible and freely available to the international research community; (2) to institute common eligibility criteria between the Children's Oncology Group and the International Society of Paediatric Oncology Europe and initiate joint or parallel clinical trials; (3) to share preliminary high-throughput screening data across discovery labs to hasten the development of novel therapeutics. Medulloblastoma Down Under 2013 was an effective forum for meaningful discussion, which resulted in enhancing international collaborative clinical and translational research of this rare disease. This template could be applied to other fields to devise global action plans addressing all aspects of a disease, from improved disease classification, treatment stratification, and drug targeting to superior treatment regimens to be assessed in cooperative international clinical trials.

Ham S, Meachem SJ, Choong CS, Charles AK, Baynam GS, Jones TW, Samarajeewa NU, Simpson ER and Brown KA.  

Hansford JR, Kotecha RS, Jevon G, Cole CH and Gottardo NG.
Efficacy of acute myeloid leukemia therapy without stem-cell transplantation in a child with blastic plasmacytoid dendritic cell neoplasm.

Hansford JR, Phillips M, Cole C, Francis J, Blyth CC and Gottardo NG.
Bacillus Cereus Bacteremia and Multiple Brain Abscesses During Acute Lymphoblastic Leukemia Induction Therapy.
Bacillus cereus can cause serious infections in immunosuppressed patients. This population may be susceptible to B. cereus pneumonia, bacteremia, cellulitis, and rarely cerebral abscess. Here we report an 8-year-old boy undergoing induction therapy for acute lymphoblastic leukemia who developed multifocal B. cereus cerebral abscesses, highlighting the propensity for B. cereus to develop cerebral abscesses. A review of the literature over the past 25 years identified another 11 cases (3 children and 8 adults) of B. cereus cerebral abscess in patients undergoing cancer therapy. B. cereus cerebral abscesses were associated with a high mortality rate (42%) and significant morbidity. Notably, B. cereus bacteremia with concomitant cerebral abscess was associated with induction chemotherapy for acute leukemia in both children and adults (10 of 12 case reports). Our case report and review of the literature highlights the propensity for B. cereus to develop cerebral abscess(es). Therefore, early consideration for neuroimaging should be given for any neutropenic cancer patient identified with B. cereus bacteremia, in particular those with acute leukemia during induction therapy.

Hawkrigg S, Johnson A, Flynn J, Thom G and Wright H.
Acute haemorrhagic oedema of infancy in a 5-week-old boy referred to the Child Protection Unit.
We describe the case of a 5-week-old infant boy presenting with purpura and oedema to both hands and torso. He was otherwise well, with no antecedent history of illness or trauma. Laboratory investigations were within normal limits. A review by the Child Protection Unit was organised during his admission for consideration of inflicted trauma as a cause of the lesions; this was felt most unlikely. A clinical diagnosis, following a dermatology consultation, of acute haemorrhagic oedema of infancy (AHO) was made.

Hawkrigg SL and Winterton PM.
Children and clandestine drug laboratories: the unseen victims.

Haynes A, Cooper MN, Bower C, Jones TW and Davis EA.
Maternal smoking during pregnancy and the risk of childhood type 1 diabetes in Western Australia.
Diabetologia. 2013.
AIMS/HYPOTHESIS: The aim of this study was to investigate the association between maternal smoking during pregnancy and type 1 diabetes in the offspring, using complete population data sources available in Western Australia. METHODS: A prospective cohort study was undertaken with cases defined as children born in Western Australia between 1998 and 2008 who were diagnosed with type 1 diabetes at <15 years of age up to 31 December 2010. Eligible cases were identified from the prospective, population-based Western Australian Children’s Diabetes Database. Record linkage was performed to identify perinatal records of cases from the Western Australian Midwives’ Notification System, which contains data on >99% of all births in Western Australia. Cox regression was used to analyse the data and adjust for recognised risk factors such as birthweight, gestational age, maternal age and socioeconomic status. RESULTS: The unadjusted HR for babies born to mothers who smoked during pregnancy being diagnosed with childhood type 1 diabetes was 0.70 (95% CI: 0.50, 0.97). After adjustment, the confidence interval widened but the point estimate remained relatively unchanged at 0.76 (95% CI: 0.54, 1.08). CONCLUSIONS/INTERPRETATION: Analyses of data from this population-based study indicate that maternal smoking during pregnancy may be associated with a reduced risk of childhood type 1 diabetes. Further investigation in larger populations with more detailed smoking data could lead to novel hypotheses regarding mechanisms that influence the immunopathogenesis of type 1 diabetes in early life.

Hegarty M, Calder A, Davies K, Shave M, Christiansen E, Meyer T and von Ungern-Sternberg BS.
BACKGROUND: More children are undergoing same-day surgery. While advances have been made in pediatric pain management, there have been few studies addressing pain management in the home (Br J Anaesth, 82, 1999 and 319). We wished to investigate whether issuing parents with take-home analgesia would improve postoperative pain scores and/or parental satisfaction following hospital discharge. METHODS: Two hundred
children, and their parents, attending for day case surgery at our institution were randomized into two groups. One group received advice regarding the management of postoperative pain and were given a pack containing discharge medications: group 'dispensed'. The other group received the same advice, but did not receive any medication: group 'advised'. Telephone interviews were conducted to assess pain scores, PONV, functional activity, analgesia requirements, and satisfaction rates. RESULTS: Data were available for 181 patients (median age, 4 years; range, 0-12 years): 89 children in group 'dispensed' and 92 children in group 'advised'. Postoperative instructions were followed by 86% in group 'advised' and 89% in group 'dispensed' (P = 0.68). Although all parents received analgesia advice, only 85/181 (48%) recalled the information. Rates for no/mild pain and moderate/severe pain were similar between the two groups: 59% (group 'advised') vs 62% (group 'dispensed') and 41% (group 'advised') vs 38% (group 'dispensed') (P = 0.78). DISCUSSION: Our study did not show any differences in the incidence of pain/parental satisfaction between the two groups. Analgesia advice given to parents was poorly retained, suggesting that other methods for disseminating information should be considered.


Ireland A, Gollow I and Gera P. Low risk, but not no risk, of umbilical hernia complications requiring acute surgery in childhood. Journal of paediatrics and child health. 2013. AIMS: Umbilical hernias are a common finding in the paediatric community, with a preponderance to affect Afro-Caribbean and premature children. The rate of incarceration varies greatly between populations. Therefore, it is valuable to obtain some Australian data on this topic. METHODS: We undertook a retrospective study of the records of all patients who underwent umbilical hernia repair over a 12-year period of between October 1999 and May 2012 at Princess Margaret Hospital. From this group, all patients that had an umbilical hernia repair for reason of acute complication were identified and analysed for age, ethnicity and co-morbidities. RESULTS: Between October 1999 and May 2012, 433 umbilical hernias were repaired at Princess Margaret Hospital, five of which were as the direct result of an acutely complicated umbilical hernia. The mean age of hernia repair was 5 years old, and the mean age of acute complication was 5 years old. Out of the patients with acutely complicated umbilical hernia, there were no Afro-Caribbean patients, and one was premature complicated by hyaline membrane disease and broncho-pulmonary dysplasia. CONCLUSIONS: Western Australia has an incidence of acutely complicated umbilical hernia requiring operative intervention of 1:3000 to 1:11 000. On an international scale, this is low, and studies with similar incidence do not advocate for immediate repair of all identified umbilical hernias. The authors believe repair should be guided by patient and guardian, but if there is an episode of incarceration, acute repair is advised.


Johnson SR, Cooper MN, Davis EA and Jones TW. Hypoglycaemia, fear of hypoglycaemia and quality of life in children with Type 1 diabetes and their parents.
AIM: To evaluate the association between fear of hypoglycaemia, episodes of hypoglycaemia and quality of life in children with Type 1 diabetes and their parents. METHODS: This was a cross-sectional, population-based study of 325 children with Type 1 diabetes and their parents. The children were aged 2-18 years. A total of 325 parents of the patients aged 2-18 years and 196 of the patients themselves (aged 8-18 years) completed questionnaires including the PedSQL Diabetes Module, the Hypoglycaemia Fear Survey and Clarke's hypoglycaemia awareness questionnaire. Data were compared with HbA1c results and the history of severe hypoglycaemia episodes. RESULTS: Parents with the highest levels of fear of hypoglycaemia reported that their children had a reduced quality of life (P < 0.001). Similarly children with the greatest fear also reported a reduced quality of life (P < 0.001); however a history of severe hypoglycaemia was not associated with the child's quality of life as perceived by the child or parent. Episodes of severe hypoglycaemia were associated with an increased fear of hypoglycaemia for the parents (P = 0.004) but not the children. Children in the highest fear quartile also had a higher HbA1c concentration compared with those in the lowest fear quartile [increase in HbA1c 7 mmol/mol (0.6%), P < 0.01]. CONCLUSIONS: Fear of hypoglycaemia and not episodes of hypoglycaemia per se is associated with increased psychological burden for children with Type 1 diabetes. Interventions to reduce fear of hypoglycaemia in these families may improve their quality of life.

Johnson SR, Cooper MN, Jones TW and Davis EA.
Long-term outcome of insulin pump therapy in children with type 1 diabetes assessed in a large population-based case-control study.
AIMS/HYPOTHESIS: We determined the impact of insulin pump therapy on long-term glycaemic control, BMI, rate of severe hypoglycaemia and diabetic ketoacidosis (DKA) in children. METHODS: Patients on pump therapy at a single paediatric tertiary hospital were matched to patients treated by injections on the basis of age, duration of diabetes and HbA1c at the time of pump start. HbA1c, anthropometric data, episodes of severe hypoglycaemia and rates of hospitalisation for DKA were collected prospectively. RESULTS: A total of 345 patients on pump therapy were matched to controls on injections. The mean age, duration of diabetes at pump start and length of follow-up were 11.4 (+/- 3.5), 4.1 (+/- 3.0) and 3.5 (+/- 2.5) years, respectively. The mean HbA1c reduction in the pump cohort was 0.6% (6.6 mmol/mol). This improved HbA1c remained significant throughout the 7 years of follow-up. Pump therapy reduced severe hypoglycaemia from 14.7 to 7.2 events per 100 patient-years (p < 0.001). In contrast, severe hypoglycaemia increased in the non-pump cohort over the same period from 6.8 to 10.2 events per 100 patient-years. The rate of hospitalisation for DKA was lower in the pump cohort (2.3 vs 4.7 per 100 patient-years, p = 0.003) over the 1,160 patient-years of follow-up.
CONCLUSIONS/INTERPRETATION: This is the longest and largest study of insulin pump use in children and demonstrates that pump therapy provides a sustained improvement in glycaemic control, and reductions of severe hypoglycaemia and hospitalisation for DKA compared with a matched cohort using injections.

Jones M and Khosa J.
Presacral tumours: a rare case of a dermoid cyst in a paediatric patient.

Judkins A, Pascoe E and Payne D.
Management of urinary tract infection in a tertiary children's hospital before and after publication of the NICE guidelines.
INTRODUCTION: The UK National Institute for Health and Clinical Excellence (NICE) introduced guidelines for the diagnosis, treatment and management of urinary tract infection (UTI) in children and adolescents in August 2007. AIM: The primary aim was to determine whether publication of NICE guidelines was associated with a change in the use of diagnostic imaging investigations in patients with a documented first UTI in a tertiary children's hospital. Secondary aims were to describe the epidemiology, microbiology, prescription of prophylactic antibiotics and follow-up for these children, and the incidence of structural renal tract abnormalities, vesicoureteric reflux and renal uptake defects identified. METHODS: Retrospective review of the case notes of patients presenting to Princess Margaret Hospital, Perth, Western Australia with a first UTI over a 4-year period (August 2005-2009). Details of demographics, radiological investigations, microbiology and follow-up were obtained. Data for subjects presenting before and after 31 August 2007 were compared. RESULTS: Data from 659 subjects, median age 6 (range 0-186) months were analysed. Compared with the pre-NICE period, there was no change in the proportion of patients undergoing renal USS in the 2 years following publication of the guidelines. There was a decrease in the proportion undergoing MCUG (p<0.0001) and receiving antibiotic prophylaxis (p<0.0001) and an increase in the proportion undergoing DMSA (p<0.001). CONCLUSIONS: Practice changed following publication of the NICE guidelines. While the reduction in MCUG requests and
prescription of antibiotic prophylaxis is in line with NICE guidelines, the increase in DMSA requests is contrary to the recommendations.

Khanna M, Shackleton C, Verheggen M, Sharp M, Wilson AC and Hall GL.
Evaluating hypoxia during air travel in healthy infants.

Kotecha RS, Jacoby P, Cole CH and Gottardo NG.
Morbidity in survivors of child and adolescent meningioma.
BACKGROUND: The extent of initial surgical resection has been identified as the strongest prognostic indicator for survival in child and adolescent meningioma. Given the paucity of data concerning long-term outcome, the authors undertook a meta-analysis to analyze morbidity in survivors of this disease. METHODS: Individual patient data were obtained from 19 case series published over the last 23 years through direct communication with the authors. Ordinal logistic regression models were used to assess the influence of risk factors on morbidity. RESULTS: Of 261 patients, 48% reported a completely normal life with no morbidity, and 25% had moderate/severe meningioma-associated morbidity at last follow-up. Multivariate analysis identified relapse as the only independent variable associated with an increased risk of morbidity (odds ratio, 4.02; 95% confidence interval, 2.11-7.65; P = .001). Univariate analysis also revealed an increased risk for patients with neurofibromatosis (odds ratio, 1.90; 95% confidence interval, 1.04-3.48; P = .04). Subgroup analysis identified a higher incidence of morbidity among patients who had intracranial tumors with a skull base location compared with a nonskull base location (P = .001). Timing at which morbidity occurred was available for 70 patients, with persistence of preoperative tumor-related symptoms in 67% and as a result of therapy in 20%. CONCLUSIONS: The majority of survivors of child and adolescent meningioma had no or only mild long-term morbidity, whereas 25% had moderate/severe morbidity, with a significantly increased risk in patients with relapsed disease. In the majority, morbidity occurred as a consequence of the tumor itself, justifying aggressive surgery to achieve gross total resection. However, for patients with neurofibromatosis and skull base meningioma, a more cautious surgical approach should be reserved.

Different Guidelines for Imaging After First UTI in Febrile Infants: Yield, Cost, and Radiation.

Larkins N and Murray KJ.
Major cluster of chilblain cases in a cold dry Western Australian winter.
AIM: Primary chilblains are an idiopathic cold-induced vasculopathy affecting the soft tissues of the hands and feet. Secondary chilblains occur in different forms of vasculitis and chronic autoimmune connective tissue disorders. Idiopathic chilblains are rarely reported in children and may generate significant anxiety to doctors and patients. We describe a cluster of idiopathic chilblains encountered over the winter of 2010 in Perth, Western Australia. METHODS: This is a retrospective review of patients identified from a prospectively compiled database of all new cases seen in our department. Data on history, examination, investigations, prescribed treatments and outcomes were collected. RESULTS: Thirty-two patients with isolated idiopathic chilblains were included, including 20 females and 12 males with a median age at onset of 13.5 years. Lesions were popular with signs of peripheral vasoconstriction causing acrocyanosis, and uncomfortable due to pain and/or pruritis in most. Thickening of the small joints was common where lesions involved these areas. Ulceration of lesions also occurred in some. One patient required hospitalisation for secondary bacterial infection. Most received some form of treatment including non-steroidal anti-inflammatory drugs, prednisolone or nifedipine. Most patients improved spontaneously with warmer weather or responded to cold protection advice. All had resolved completely by late spring (November). CONCLUSION: Our cluster of chilblains was associated with an unusually cold winter in Perth 2010. It is the largest series reported in the literature, suggesting that chilblains may be more common than previously thought. Chilblains are almost always benign in nature and patients are systemically well and usually need no further investigation and only symptomatic treatment. Prompt recognition can avoid excessive investigation and anxiety, allowing appropriate simple advice and treatment.

Lawrence JG, Carapetis JR, Griffiths K, Edwards K and Condon JR.
Acute Rheumatic Fever and Rheumatic Heart Disease: Incidence and Progression in the Northern Territory of Australia 1997-2010.
Circulation. 2013.
Leonard H, Ravikumara M, Baikie G, Naseem N, Ellaway C, Percy A, Abraham S, Geerts S, Lane J, Jones M, Bathgate K and Downs J. Assessment and management of nutrition and growth in Rett syndrome. J Pediatr Gastroenterol Nutr. 2013; 57(4): 451-460. OBJECTIVES: We developed recommendations for the clinical management of poor growth and weight gain in Rett syndrome through evidence review and the consensus of an expert panel of clinicians. METHODS: Initial draft recommendations were created based upon literature review and 34 open-ended questions in which the literature was lacking. Statements and questions were made available to an international, multidisciplinary panel of clinicians in an online format and a Microsoft Word-formatted version of the draft via e-mail. Input was sought using a 2-stage modified Delphi process to reach consensus. Items included clinical assessment of growth, anthropometry, feeding difficulties and management to increase energy intake, decrease feeding difficulties, and consideration of gastrostomy. RESULTS: Agreement was achieved on 101 of 112 statements. A comprehensive approach to the management of poor growth in Rett syndrome is recommended that takes into account factors such as feeding difficulties and nutritional needs. A body mass index of approximately the 25th centile can be considered as a reasonable target in clinical practice. Gastrostomy is indicated for extremely poor growth, if there is risk of aspiration and if feeding times are prolonged. CONCLUSIONS: These evidence- and consensus-based recommendations have the potential to improve care of nutrition and growth in a rare condition and stimulate research to improve the present limited evidence base.


Loh RK, Vale S and McLean-Tooke A. Quantitative serum immunoglobulin tests. Australian family physician. 2013; 42(4): 195-198. What is the test? Immunoglobulins are protein molecules. They contain antibody activity and are produced by the terminal cells of B-cell differentiation known as ‘plasma cells’. There are five classes of immunoglobulin (Ig): IgG, IgM, IgA, IgD and IgE. In normal serum, about 80% is IgG, 15% is IgA, 5% is IgM, 0.2% is IgD and a trace is IgE. Quantitative serum immunoglobulin tests are used to detect abnormal levels of the three major classes (IgG, IgA and IgM). Testing is used to help diagnose various conditions and diseases that affect the levels of one or more of these immunoglobulin classes. Some conditions cause excess levels, some cause deficiencies, and others cause a combination of increased and decreased levels. IgD and IgE will not be discussed in this article.

Ly TT, Nicholas JA, Retterath A, Lim EM, Davis EA and Jones TW. Effect of sensor-augmented insulin pump therapy and automated insulin suspension vs standard insulin pump therapy on hypoglycemia in patients with type 1 diabetes: a randomized clinical trial. JAMA : the journal of the American Medical Association. 2013; 310(12): 1240-1247. IMPORTANCE: Hypoglycemia is a critical obstacle to the care of patients with type 1 diabetes. Sensor-augmented insulin pump with automated low-glucose insulin suspension has the potential to reduce the incidence of major hypoglycemic events. OBJECTIVE: To determine the incidence of severe and moderate hypoglycemia with sensor-augmented pump with low-glucose suspension compared with standard insulin pump therapy. DESIGN, SETTING, AND PARTICIPANTS: A randomized clinical trial involving 95 patients with type 1 diabetes, recruited from December 2009 to January 2012 in Australia. INTERVENTIONS: Patients were randomized to insulin pump only or automated insulin suspension for 6 months. MAIN OUTCOMES AND MEASURES: The primary outcome was the combined incidence of severe (hypoglycemic seizure or coma) and moderate hypoglycemia (an event requiring assistance for treatment). In a subgroup, counterregulatory hormone responses to hypoglycemia were assessed using the hypoglycemic clamp technique. RESULTS: Of the 95 patients randomized, 49 were assigned to the standard-pump (pump-only) therapy and 46 to the low-glucose suspension group. The mean (SD) age was 18.6 (11.8) years; duration of diabetes, 11.0 (8.9) years; and duration of pump therapy, 4.1 (3.4) years. The baseline rate of severe and moderate hypoglycemic events in the pump-only group was 20.7 vs 129.6 events per 100 patient months in the low-glucose suspension group. After 6 months of treatment, the event rates decreased from 28 to 16 in the pump-only group vs 175 to 35 in the low-glucose suspension group. The adjusted incidence rate per 100 patient-months was 34.2 (95% CI, 22.0-53.3) for the pump-only group vs 9.5 (95% CI, 5.2-17.4) for the low-glucose suspension group. The incidence rate ratio was 3.6 (95% CI, 1.7-7.5; P <.001). There was no change in glycazed hemoglobin in either group: mean, 7.4 (95% CI, 7.2-7.6) to 7.4 (95% CI, 7.2-7.7) in the pump-only group vs mean, 7.6 (95%, CI, 7.4-7.9) to 7.5 (95% CI, 7.3-7.7) in the low-glucose suspension group. Counterregulatory hormone responses to hypoglycemia were not changed. There were no episodes of diabetic ketoacidosis or hyperglycemia with


Mariyappa B, Barker A, Samnakay N and Khosa J. Management of duplex-system ureterocele. Journal of paediatrics and child health. 2013. AIM: To analyse different treatment modalities, functional outcome and continence in children treated for duplex-system ureterocele and to review the relevant literature. METHODS: The medical records of patients with duplex-system ureterocele treated between 2001 and 2011 were reviewed retrospectively. RESULTS: Twenty-two cases were identified. Five patients underwent incision of the ureterocele as initial procedure. It was curative in only one patient. Seven patients underwent upper-pole nephroureterectomy. It was curative in 4 cases. Five patients underwent excision of ureterocele and common-sheath reimplant, and the remaining 5 patients had upper-pole nephroureterectomy and simultaneous excision of ureterocele with lower-moiet group for targeted prevention.


Martin AC, Coakley J, Forbes DA, Sullivan DR and Watts GF. Familial hypercholesterolaemia in children and adolescents: A new paediatric model of care. Journal of paediatrics and child health. 2013; 49(4): E263-272. Familial hypercholesterolaemia is a marked genetic disorder affecting more than 8000 children and adolescents throughout Australia. It results in marked elevation in plasma low-density lipoprotein cholesterol levels from birth that predisposes individuals to premature coronary heart disease in adult life. The majority of children and adolescents with FH are undiagnosed, as symptoms and signs only develop after decades of hypercholesterolaemia. Cascade screening of family members after detecting FH in an index case is an effective approach that allows the diagnosis of FH to be made in the young, before significant atherosclerosis develops. With the availability of effective therapies, mainly statins, paediatricians are ideally placed to improve the outcomes of this disorder by detecting and managing hypercholesterolaemia in childhood, thereby preventing premature coronary artery disease. We describe a new paediatric model of care for FH.

Martin L, Rea S, McWilliams T and Wood F. Hot ash burns in the children of Western Australia: How and why they happen. Burns : journal of the International Society for Burn Injuries. 2013. INTRODUCTION: Burns from hot ash are common in the paediatric population in Western Australia. Fifty children were admitted to the paediatric burn centre with hot ash contact burns to the feet in 2011 and 2012. It is important to examine the extent of the problem, seasonal variations, and identify those at risk to determine strategies for prevention campaigns. METHOD: Retrospective review of medical notes for all admissions to the paediatric burns unit was undertaken for 2011 and 2012. Data were collected for patient demographics, time, circumstance of injury, burn severity and treatment. RESULTS: Hot ash burns accounted for 8.6% of admissions but 16.1% of burns sustained in non-metro areas. Median age was just under 3 years, male or female. Median burn TBSA was 2%, and 44% of children required surgery. The burns were less common in summer, more common on non-school days and in children who were on camping trips away from home. DISCUSSION: Previous work has shown the value of targeted campaigns. The group for targeted prevention...
campaigns are the carers of very young children who go camping. Information distributed at camping shows and stores about the principles of campfire safety would reach the people at risk.


OBJECTIVE: To report the experience of implementing a 4-hour rule program. DESIGN, SETTING AND PARTICIPANTS: A 3-2013 whole-of-hospital clinical service redesign program in a tertiary paediatric hospital in Western Australia, involving all patients presenting to the emergency department (ED) from 1 January 2009 to 31 December 2011. MAIN OUTCOME MEASURES: Percentage of patients admitted, discharged or transferred from the ED within 4 hours of arrival at triage, and percentage of patients discharged from inpatient wards before 10 am. RESULTS: The percentage of patients admitted, discharged or transferred within 4 hours of arrival at the ED increased from 87% in 2009 to 95% in 2011. Safety and quality measures, including the admission rate from the ED, unplanned reattendances at the ED within 48 hours of discharge, patient complaints and inhospital mortality, remained unchanged. The percentage of patients discharged from inpatient wards before 10 am increased from 18% in 2009 to 30% in 2011. CONCLUSIONS: The introduction of a 4-hour rule program has resulted in improved timeliness of care for patients throughout the hospital, both in the ED and inpatient wards, with no adverse impact on the quality and safety of clinical care.


OBJECTIVE: To determine whether an outreach community-based training program on eating disorders enhances perceived capacity of rural health and education professionals to respond to and manage eating disorders. DESIGN: Survey conducted upon completion of outreach training. SETTING: Rural Western Australia. PARTICIPANTS: Health and education professionals working in rural Western Australia. MAIN OUTCOME MEASURES: Questionnaire responses analysed via descriptive statistics and inferential tests. RESULTS: There was a significant increase in perceived ability to identify, support and/or treat people with eating disorders among health and education professionals. CONCLUSIONS: Outreach training up-skilled rural gatekeepers and introduced systemic health system benefits of increased consultation and liaison, a fine-tuning of referral processes, a reduction in hospital admissions and better uptake of local services by patients discharged from hospital.


INTRODUCTION: Despite burns being common in children, research into the psychological experience and trauma remains limited. Improvements in the professional understanding of children's experiences will assist in improving holistic care. PURPOSE: This study uses phenomenology, a qualitative methodology to explore the psychological experiences following a burn injury in children. METHODS: In-depth interviews were conducted six months after burn with 12 (six girls and six boys) children who underwent surgery for a burn. The children were aged eight to 15 years. The interview examined the overall experience of children and included probing questions exploring participants' perceptions, thoughts and feelings. Transcripts were analysed according to the seven-step Coliazzi method. Relationships between themes were explored to identify core concepts. RESULTS: The findings demonstrated that trauma was central to the burn experience and comprised two phases: the burn trauma and the recovery trauma. Six themes emerged as a result of this experience: ongoing recurrent trauma; returning to normal activities; behavioural changes; scarring-the permanent reminder; family and adaptation. CONCLUSION: This research has clinical implications as its findings can be used to inform clinical care at all stages of the burn journey. These research conclusions could be used to develop comprehensive information and support management plans for children. This would complement and support the surgical and medical treatment plan, providing direction for comprehensive service delivery and improved psychosocial outcomes in children.

AIM: To investigate the impact of regular exposure to paediatric medical trauma on multidisciplinary teams in a paediatric hospital and the relationships between psychological distress, resilience and coping skills. METHOD: Symptoms of post-traumatic stress disorder, secondary traumatic stress, depression, anxiety, stress, burnout, compassion satisfaction, resilience and coping skills were measured in 54 health professionals and compared with published norms. RESULTS: Participants experienced more symptoms of secondary traumatic stress (P < 0.01), showed less resilience (P = 0.05) and compassion satisfaction (r = 0.01), more use of optimism and sharing as coping strategies, and less use of dealing with the problem and non-productive coping strategies than comparative groups. Non-productive coping was associated with more secondary traumatic stress (r = 0.50, P = 0.05), burnout (r = 0.45, P = 0.01), post-traumatic stress disorder (r = 0.41, P = 0.05), anxiety (r = 0.42, P = 0.05), depression (r = 0.54, P = 0.01) and stress (r = 0.52, P = 0.01) and resilience was positively associated with optimism (r = 0.48, P = 0.01). Health professionals <25 years old used more non-productive coping strategies (P = 0.05), less sharing as a coping strategy (P = 0.05) and tended to have more symptoms of depression (P = 0.06). CONCLUSION: Paediatric medical trauma can adversely affect a health professional's well-being, particularly those <25 years of age who make less use of positive coping strategies and more use of non-productive coping. These findings will assist the development of effective and meaningful interventions for health professionals working in paediatric hospitals.

McGarry S, Girdler S, McDonald A, Valentine J, Wood F and Elliott C. Paediatric medical trauma: The impact on parents of burn survivors. Burns : journal of the International Society for Burn Injuries. 2013; 39(6): 1114-1121. In order to identify parents at risk of developing ongoing psychological distress after their child has sustained a burn a greater understanding of paediatric medical trauma is required. AIM: To investigate the impact of exposure to paediatric trauma on parents of children with a burn and to identify risk factors and relationships between psychological distress and resilience. METHODS: Sixty-three parents were recruited. Parents completed standardised assessments measuring symptoms of posttraumatic stress disorder (PTSD), depression, anxiety, stress, and resilience within one week of the burn occurring. Statistical analysis included t-tests, Kruskal-Wallis one way ANOVA and Spearman's Roe. RESULTS: Parents experienced significantly more symptoms of PTSD (p=0.001) than a comparative community population. Factors including having a daughter, witnessing the event, feeling helpless or having past traumatic experiences significantly influenced symptoms of psychological distress and resilience (p=0.05). CONCLUSION: Parents of burn survivors experience significant psychological distress with low levels of resilience. As part of standard routine care health professionals should screen parents to identify those at greatest risk and provide effective evidence based interventions aimed at improving resilience and reducing stress.

McLean-Tooke A, O'Sullivan M, Easter T and Loh R. Differences between total IgG and sum of the IgG subclasses in clinical samples. Pathology. 2013; 45(7): 675-677. AIMS: IgG subclasses measurement is used in the investigation of patients with immunodeficiency and autoimmune diseases. In some patients a significant discrepancy between the sum of IgG subclasses (IgGsum) and total IgG may be seen. This study aimed to assess frequency and degree of such discrepancies in routine samples. METHODS: Data were collected retrospectively from 571 consecutive IgG subclass samples performed by an nephelometric/turbidimetric assay. Total IgG measurement was performed by nephelometry or turbidimetry. Fifty prospective samples with a difference between the IgGsum and total IgG >15% were re-run at dilution. RESULTS: IgGsum was a mean of 3.7% higher than the total IgG. Sixty-two samples (10.9%) had a difference between IgGsum and total IgG of >15%. Difference between IgGsum and total IgG correlated with the proportion but not level of IgG1. Repeat testing at dilution of samples with differences >15% did not significantly reduce the difference between results. CONCLUSIONS: Differences of >15% between IgGsum and total IgG are common. Using an adjusted range based on our data would reduce the number of samples requiring additional testing. Samples falling outside this range should be reviewed.


Minutillo C, Rao SC, Pirie S, McMichael J and Dickinson JE. Growth and developmental outcomes of infants with gastroschisis at one year of age: A retrospective study. J Pediatr Surg. 2013; 48(8): 1688-1696. BACKGROUND: The aim of the study was to describe the physical growth and developmental outcomes of babies born with gastroschisis. METHODS: We retrospectively reviewed all cases of gastroschisis in Western Australia born between 1997 and 2010. RESULTS: In the 128 pregnancies with fetal gastroschisis, 117 babies were live born. 112 (95.7%) survived to one year. 19% had z scores of<1.28 for weight at birth (<10th centiles)
compared with 30% at one year. Neurodevelopmental data were available in 88/112 (79%) of survivors (Griffiths scores in 67; reports of ages and stages questionnaire (ASQ) in 21). The mean GQ at 12months was 99 (SD 9.8). Suboptimal neurodevelopmental outcomes were noted in eight. Complex gastroschisis (present at birth) and acquired gut related complications were associated with adverse long term outcomes. The incidence of acquired gut complications was least (5%) in those who underwent silo reduction as the primary management. However, on univariate and multivariate analysis, the type of primary reduction did not significantly influence the outcome. CONCLUSIONS: A large proportion of infants with gastroschisis exhibit suboptimal weight gain during the first year. The incidence of adverse developmental outcomes appears to be low.

Outcomes of haematopoietic stem cell transplantation for inherited metabolic disorders: A report from the Australian and New Zealand Children’s Haematology Oncology Group and the Australasian Bone Marrow Transplant Recipient Registry.

Moore HC, Lehmann D, de Klerk N, Smith DW, Richmond PC, Keil AD and Blyth CC.
How Accurate Are International Classification of Diseases-10 Diagnosis Codes in Detecting Influenza and Pertussis Hospitalizations in Children?

Association of inhibitors of gastric Acid secretion and higher incidence of necrotizing enterocolitis in preterm very low-birth-weight infants.
Background Inhibitors of gastric acid (IGA) are used for upper gastrointestinal bleeding or gastroesophageal reflux in preterm infants. The resultant increase in gastric pH may enhance the growth of pathogens and increase the risk of necrotizing enterocolitis (NEC). Our systematic review examined the association between IGA and NEC in preterm infants.Methods Standard methodology of systematic reviews was followed. PubMed, Embase, Cochrane, and Cumulative Index to Nursing and Allied Health Literature (CINAHL) databases were searched in August 2012.Results One case-control and one prospective cohort study (n = 11,346), both evaluating H2-blockers as IGA, were included. Meta-analysis showed a significant association between NEC and IGA (odds ratio [OR]: 1.78, 95% confidence interval [CI]: 1.4, 2.27, p < 0.00001). The prospective cohort study found higher incidence of infection (sepsis, pneumonia, urinary tract infection) with IGA (37.4% versus 9.8%, OR: 5.5, 95% CI: 2.9 to 10.4, p < 0.001).Conclusions Exposure to H2 receptor antagonists may be associated with increased risk of NEC and infections in preterm infants.

More K, Athalye-Jape G, Rao SC and Patole SK.
Endothelin receptor antagonists for persistent pulmonary hypertension in term and late preterm infants.
The Cochrane Library. 2013; (5).

Mott LS, Graniel KG, Park J, de Klerk NH, Sly PD, Murray CP, Tiddens HA and Stick SM.
Assessment of early bronchiectasis in young children with cystic fibrosis is dependent on lung volume.

Mott LS, Park J, Gangel CL, de Klerk NH, Sly PD, Murray CP and Stick SM.
Distribution of early structural lung changes due to cystic fibrosis detected with chest computed tomography.

Mukkur T and Richmond P.
Vaccines and Vaccinations. 2013; 4(2).

Naylor LH, Yusof NM, Paramalingam N, Jones TW, Davis EA and Green DJ.
Acute hyperglycaemia does not alter nitric oxide-mediated microvascular function in the skin of adolescents with type 1 diabetes.

Serum ferritin and nutritional status: insights from an eating disorders clinic population.
Delivering a Healthy WA

Arch Dis Child. 2013. OBJECTIVE: To determine the relationship between serum ferritin and malnutrition in newly assessed patients at a paediatric eating disorders clinic. DESIGN: This was a prospectively assessed clinical cohort study. SETTING: Intake assessment clinic of a tertiary eating disorders service for children and adolescents. METHODS: Clinical, anthropometric and laboratory features of children and adolescents were systematically measured. The relationship of serum ferritin to other clinical, anthropometric and laboratory measures was determined using linear regression. RESULTS: A total of 121 female patients aged 9.5-17.6 years were included, with body mass index (BMI) z score -5.7 to 1.9 (median -1.3). Using multiple regression, serum ferritin was inversely associated with BMI z score (regression coefficient (beta)= -0.234, 95% CI -0.413 to -0.055) and serum insulin-like growth factor 1 (IGF-1) (beta= -0.476, 95% CI -0.884 to -0.068) and positively associated with alanine aminotransferase (beta=0.357, 95% CI 0.055 to 0.659, controlling for age, pubertal stage and serum iron). CONCLUSIONS: In malnourished adolescents with eating disorders increased serum ferritin is associated with lower BMI z score and serum IGF-1.


O'Grady MJ, Delaney J, Jones TW and Davis EA. Standardised mortality is increased three-fold in a population-based sample of children and adolescents with type 1 diabetes. Pediatr Diabetes. 2013; 14(1): 13-17. There are no type 1 diabetes (T1DM) mortality data on Australian children and limited contemporary data on their international counterparts. Fatalities in children and adolescents (1-17 yr) with T1DM were identified from the Western Australia Children's Diabetes Database between 1987-2011. Seventeen thousand four hundred and fifty-three patient-years of diabetes data were analysed and 13 deaths were confirmed (six male). The overall standardised mortality ratio was 3.1 [95% confidence interval (CI), 1.7-5.3] and was highest in the 10-14 yr age group, at 4.6 (95% CI, 1.5-10.8). Median age at death was 16.4 yr (range 5 to 17.8 yr), and median haemoglobin A1c at death was 10.5% (range 6.7 to >14). Cause of death was attributed to diabetes in 10 (77%) cases. Two patients were found 'dead-in-bed'. All diabetes-related deaths in subjects with known T1DM occurred outside the hospital setting.


O'Sullivan M, McLean-Tooke A and Loh RK. Antinuclear antibody test. Australian family physician. 2013; 42(10): 718-721. The antinuclear antibody (ANA) test is widely used as a serological marker of autoimmune disease. Antinuclear antibodies are immunoglobulins or antibodies that bind to one or more antigens expressed within the nucleus of human cells. Used selectively, the ANA test can be a useful laboratory tool to help confirm or exclude the diagnosis of systemic rheumatic disease. However, the relatively high prevalence of ANAs in other inflammatory conditions, as well as healthy individuals, can make a positive result difficult to interpret.

Oakley E, Borland M, Neutz J, Acworth J, Krieser D, Dalziel S, Davidson A, Donath S, Jachno K, South M, Theophilos T and Babi FE. Nasogastric hydration versus intravenous hydration for infants with bronchiolitis: a randomised trial. Lancet Respir Med. 2013; 1(2): 113-120. BACKGROUND: Bronchiolitis is the most common lower respiratory tract infection in infants and the leading cause of hospital admission. Hydration is a mainstay of treatment, but insufficient evidence exists to guide clinical practice. We aimed to assess whether intravenous hydration or nasogastric hydration is better for treatment of infants. METHODS: In this multicentre, open, randomised trial, we enrolled infants aged 2-12 months admitted to hospitals in Australia and New Zealand with a clinical diagnosis of bronchiolitis during three bronchiolitis seasons (April 1-Oct 31, in 2009, 2010, and 2011). We randomly allocated infants to nasogastric hydration or intravenous hydration by use of a computer-generated sequence and opaque sealed envelopes, with three randomly assigned block sizes and stratified by hospital site and age group (2-<6 months vs 6-12 months). The primary outcome was length of hospital stay, assessed in all randomly assigned infants. Secondary outcomes included rates of intensive-care unit admission, adverse events, and success of insertion. This trial is registered with the Australian and New Zealand clinical trials registry, ACTRN1260500033640.
FINDINGS: Mean length of stay for 381 infants assigned nasogastric hydration was 86.6 h (SD 58.9) compared with 82.2 h (58.8) for 378 infants assigned intravenous hydration (absolute difference 4.5 h [95% CI -3.9 to 12.9]; p=0.30). Rates of admission to intensive-care units, need for ventilatory support, and adverse events did not differ between groups. At randomisation, seven infants assigned nasogastric hydration were switched to intravenous hydration and 56 infants assigned intravenous hydration were switched to nasogastric hydration because the study-assigned method was unable to be inserted. For those infants who had data available for successful insertion, 275 (85%) of 323 infants in the nasogastric hydration group and 165 (56%) of 294 infants in the intravenous hydration group required only one attempt for successful insertion. INTERPRETATION: Intravenous hydration and nasogastric hydration are appropriate means to hydrate infants with bronchiolitis. Nasogastric insertion might require fewer attempts and have a higher success rate of insertion than intravenous hydration. FUNDING: Australian National Health and Medical Research Council, Samuel Nissen Charitable Foundation (Perpetual), Murdoch Children's Research Institute, Victorian Government.


Paul S, Rao S, Kohan R, McMichael J, French N, Zhang G and Simmer K. Poractant alfa versus beractant for respiratory distress syndrome in preterm infants: A retrospective cohort study. Journal of paediatrics and child health. 2013; 49(10): 839-844. AIM: Poractant alfa and beractant are the commonly used animal derived surfactants in preterm infants with respiratory distress syndrome. Between 2005 and 2007, poractant alfa and beractant were alternated every month in our neonatal intensive care unit for 27 months. The aim of this study was to compare the outcomes of preterm infants who received poractant alfa versus beractant. METHOD: Single-centre, retrospective cohort study of inborn preterm infants <32 weeks gestation (23-31(+6)). RESULTS: Six hundred sixty-four preterm infants (<32 weeks) were born during the study period, of which 415 received surfactant (poractant alfa: 214; beractant: 201). Infants in the poractant alfa group were 2.8 days younger than beractant (27.0 +/- 2.3 vs. 27.4 +/- 2.3 weeks; P = 0.03). All other baseline characters including Clinical Risk Index for Babies II scores were similar for both groups. No significant differences were found for the following outcomes: death or chronic lung disease (78/212 vs. 59/200; P = 0.28); death (24/214 vs. 15/201, P = 0.24); moderate to severe chronic lung disease (63/212 vs. 46/200; P = 0.45) and moderate to severe disability (20/163 vs. 19/151, P = 0.98) between poractant alfa and beractant, respectively. CONCLUSIONS: The results of our study do not support the need for preferential use of poractant alfa or beractant.


Priddis LE, Landy S, Moroney D and Kane R.

Rakshashbuvankar A, Rao S, Kohan R, Simmer K and Nagarajan L. Intravenous levetiracetam for treatment of neonatal seizures. J Clin Neurosci. 2013. 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.

Rath SR, Lee S, Kotecha RS, Taylor M, Junckerstorff RC and Choong CS. Childhood craniopharyngioma: 20-year institutional experience in Western Australia. Journal of paediatrics and child health. 2013; 49(5): 403-408. AIM: A retrospective audit was undertaken to evaluate modes of presentation and treatment outcomes for craniopharyngioma in a single paediatric institution over a 20-year period. METHODS: A search of the neurosurgical and histopathological databases for patients under 21 years of age treated for craniopharyngioma between 1990 and 2010 was performed at our institution. The clinical records of eligible patients were reviewed and information regarding presentation, medical and surgical management and post-treatment outcome were extracted and collated. RESULTS: Of 10 evaluable patients, the commonest presenting symptoms were headache and visual impairment. Clinical and biochemical evaluation undertaken prior to surgery revealed visual dysfunction in 70% and pituitary deficit in 30%. Gross total resection was achieved in 40% but was curative in only 20%. The remaining 80% required further surgical and/or radiotherapeutic intervention. Seven patients had radiation therapy with stabilisation in 70%. Multiple pituitary hormone deficiency evolved in all patients over time, while visual impairment worsened in 30% post-operatively and improved in 20%. Obesity was present in 50% after a mean follow-up interval of 5.6 years and was apparent within 1 year of initial surgery in 30%. Although neurocognitive, psychological and behavioural problems were noted for some patients during medical review, only 20% of patients were formally assessed. CONCLUSIONS: Craniopharyngioma is associated with significant long-term morbidity. Attention to an integrated care pathway that includes standardised neurocognitive and psychological and behavioural assessment would facilitate early appropriate intervention and support leading to an improved quality of life for children with craniopharyngioma.


Reynolds V, Meldrum S, Simmer K, Vijayasekaran S and French NP. Dysphonia in preterm children: Assessing incidence and response to treatment. Contemporary clinical trials. 2013. BACKGROUND: Mild dysphonia in childhood is surprisingly common, yet moderate to severe dysphonia is rare. The latter has been associated with complex medical conditions and congenital abnormalities. Intubation injury has also been documented as a cause of childhood dysphonia. Children born very preterm may be intubated as part of the intensive care administered in the perinatal and neonatal period, yet there are few studies investigating dysphonia in this population. This study will be the first to: use an objective acoustic voice assessment in a paediatric study, document the incidence of dysphonia in very preterm children at school age, and conduct a controlled trial of behavioural voice therapy in this population. DESIGN: This study will consist of three phases: assessment of voice quality and its impact on quality of life in up to 200 children born at less than 32 weeks gestation: assessment of the nature and extent of laryngeal pathology in children with moderate to severe dysphonia; and a non-blinded, randomised controlled trial of behavioural voice therapy in children with moderate to severe dysphonia. DISCUSSION: This study will be the first to use clinical assessment to examine the voice quality of very preterm children, and to use fibreoptic endoscopic evaluation of laryngeal function to determine the nature and extent of any laryngeal pathology in such children. Those participants with significant voice difficulties will be randomised to receive treatment immediately or after the eight week assessment. TRIAL REGISTRATION: This study is registered on the Australian New Zealand Clinical Trials Registry (ACTRN12613001015730/ACTRN12613001012763).


Russell P, von Ungern-Sternberg BS and Schug SA. Perioperative analgesia in pediatric surgery. Curr Opin Anaesthesiol. 2013; 26(4): 420-427. PURPOSE OF REVIEW: Poor pain management has continued to be a problem after pediatric surgery. This review examines the current situation and recent advances in the area. RECENT FINDINGS: Nonopioids such as paracetamol and NSAIDs play an increasing role as components of multimodal analgesia in children. However, studies on the safety and efficacy of many adjuvants in pediatrics are still lacking. The use of opioids is influenced understandably by safety concerns about respiratory depression, but data on its incidence are poor. The role of regional techniques in the treatment of pain after pediatric surgery is growing in line with the developments in adults; the emphasis here is more on peripheral techniques too. SUMMARY: The concept of multimodal analgesia including the use of regional analgesic techniques is governing the current developments and discussions in the area of pediatric postoperative analgesia.


Schultz A and Martin AC. Outpatient management of asthma in children. Clin Med Insights Pediatr. 2013; 7: 13-24. The principal aims of asthma management in childhood are to obtain symptom control that allows individuals to engage in unrestricted physical activities and to normalize lung function. These aims should be achieved using the fewest possible medications. Ensuring a correct diagnosis is the first priority. The mainstay of asthma management remains pharmacotherapy. Various treatment options are discussed. Asthma monitoring includes the regular assessment of asthma severity and asthma control, which then informs decisions regarding the stepping up or stepping down of therapy. Delivery systems and devices for inhaled therapy are discussed, as are the factors influencing adherence to prescribed treatment. The role of the pediatric health care provider is to establish a functional partnership with the child and their family in order to minimize the impact of asthma symptoms and exacerbations during childhood.

Shah S, Ghosh S and Nagarajan L.
Shah S, Keil A, Gara K and Nagarajan L.
Neurologic Complications of Influenza.

Sharma A, Mews C, Jevon G and Ravikumara M.
Duodenal bulb biopsy in children for the diagnosis of coeliac disease: experience from Perth, Australia.
AIM: The study aims to assess the usefulness of duodenal bulb biopsy in the diagnosis of coeliac disease (CD) in a paediatric population. METHODS: Since February 2009, in our institution, we have routinely included duodenal bulb biopsy in addition to distal duodenal biopsies in children undergoing diagnostic upper gastrointestinal endoscopy. All children diagnosed with CD between February 2009 and May 2011 were identified, and those children who had biopsy finding of CD limited to duodenal bulb were reviewed with regard to clinical, serological and histopathological parameters. Duodenal bulb biopsy reports of those children who did not have CD were also reviewed as control group. RESULTS: A total of 101 children were diagnosed with CD during the study period. The mean age was 8.21 years (±3.83), 33 males and 68 females. There were 8 out of 101 (7.92%) who had histological changes consistent with CD exclusively in the duodenal bulb, with normal histology in the distal duodenum. None of duodenal bulb biopsy was abnormal in the control group. CONCLUSIONS: In some children, diagnostic CD changes may be limited to the duodenal bulb only and hence we recommend that duodenal bulb biopsies be included routinely in children suspected with CD to improve the diagnostic yield.

Shetty VB, Kiraly-Borri C, Lamont P, Bikker H and Choong CS.
NKX2-1 mutations in brain-lung-thyroid syndrome: a case series of four patients.

Scoping review of the literature about family-centred care with caregivers of children with cystic fibrosis.

Siddiqui J, Brizard CP, Galati JC, Iyengar AJ, Hutchinson D, Konstantinov IE, Wheaton GR, Ramsay JM and d'Udekem Y.
Surgical valvotomy and repair for neonatal and infant congenital aortic stenosis achieves better results than interventional catheterization.

Sim G and Lannigan F.
Lateral sinus thrombosis following myringoplasty: a rare complication.
OBJECTIVE: We report lateral sinus thrombosis occurring as a rare complication following a routine and uneventful otological procedure. CASE REPORT: Lateral sinus thrombosis is a rare but known complication of otitis media. It has not been documented as a complication of routine otological surgery. We present a case of this rare complication following a myringoplasty. We also discuss the presentation, investigation and treatment of lateral sinus thrombosis. It is essential to be able to recognise and treat this rare complication early, due to its high mortality rate. CONCLUSION: Lateral sinus thrombosis is a rare but potentially life-threatening complication. It is therefore essential for clinicians to be able to recognise and treat this condition early.

Sim G and Vijayasekaran S.
Novel use of Coblation technology in an unusual congenital tracheal stenosis.
Background: We report the case of an unusual late presentation of congenital tracheal stenosis in a 13-year-old boy. He was treated with minimally invasive Coblation resection of the stenotic segment, avoiding a major open tracheal resection and reconstruction. This case report is the first to document the use of an ultra-fine Coblation wand in the treatment of congenital tracheal stenosis. Results: The case proceeded well, without any complications. The patient had a fully healed and patent trachea at 12-week post-operative review. Conclusion: Complex cases of congenital stenosis should be managed with a multidisciplinary approach. Different and novel treatment options should be explored to find one that suits the individual patient. Minimally invasive Coblation technology can offer less invasive treatment with quicker recovery and shorter hospitalisation.
Simmer K, Rakshasbhuvankar A and Deshpande G. Standardised parenteral nutrition. Nutrients. 2013; 5(4): 1058-1070. Parenteral nutrition (PN) has become an integral part of clinical management of very low birth weight premature neonates. Traditionally different components of PN are prescribed individually considering requirements of an individual neonate (IPN). More recently, standardised PN formulations (SPN) for preterm neonates have been assessed and may have advantages including better provision of nutrients, less prescription and administration errors, decreased risk of infection, and cost savings. The recent introduction of triple-chamber bag that provides total nutrient admixture for neonates may have additional advantage of decreased risk of contamination and ease of administration.

Sims C. A suction catheter is a simple technique to aid nasal intubation with the GlideScope. Paediatr Anaesth. 2013; 23(9): 874.


Sly PD, Gangell CL, Chen L, Ware RS, Ranganathan S, Mott LS, Murray CP and Stick SM. Risk factors for bronchiectasis in children with cystic fibrosis. N Engl J Med. 2013; 368(21): 1963-1970. BACKGROUND: Bronchiectasis develops early in the course of cystic fibrosis, being detectable in infants as young as 10 weeks of age, and is persistent and progressive. We sought to determine risk factors for the onset of bronchiectasis, using data collected by the Australian Respiratory Early Surveillance Team for Cystic Fibrosis (AREST CF) intensive surveillance program. METHODS: We examined data from 127 consecutive infants who received a diagnosis of cystic fibrosis after newborn screening. Chest computed tomography (CT) and bronchoalveolar lavage (BAL) were performed, while the children were in stable clinical condition, at 3 months and 1, 2, and 3 years of age. Longitudinal data were used to determine risk factors associated with the detection of bronchiectasis from 3 months to 3 years of age. RESULTS: The point prevalence of bronchiectasis at each visit increased from 29.3% at 3 months of age to 61.5% at 3 years of age. In multivariate analyses, risk factors for bronchiectasis were presented with meconium ileus (odds ratio, 3.17; 95% confidence interval [CI], 1.51 to 6.66; P=0.002), respiratory symptoms at the time of CT and BAL (odds ratio, 2.27; 95% CI, 1.24 to 4.14; P=0.008), free neutrophil elastase activity in BAL fluid (odds ratio, 3.02; 95% CI, 1.70 to 5.35; P<0.001), and gas trapping on expiratory CT (odds ratio, 2.05; 95% CI, 1.17 to 3.59; P=0.01). Free neutrophil elastase activity in BAL fluid at 3 months of age was associated with persistent bronchiectasis (present on two or more sequential scans), with the odds seven times as high at 12 months of age and four times as high at 3 years of age. CONCLUSIONS: Neutrophil elastase activity in BAL fluid in early life was associated with early bronchiectasis in children with cystic fibrosis. (Funded by the National Health and Medical Research Council of Australia and Cystic Fibrosis Foundation Therapeutics.)


Srinivasjois R, Rao S and Patole S. Prebiotic supplementation in preterm neonates: Updated systematic review and meta-analysis of randomised controlled trials. Clin Nutr. 2013; 32(6): 958-965. BACKGROUND & AIMS: Regular administration of prebiotic oligosaccharides promote beneficial gut flora in infants. We aimed to systematically review randomised controlled trials evaluating the safety and efficacy of prebiotic oligosaccharide supplementation in preterm infants <37 weeks of gestation. METHODS: Available studies from Medline, Embase, comparing formula milk supplemented with or without prebiotics, reporting on safety and the incidence of necrotising enterocolitis (NEC), late onset sepsis, feed tolerance, physical growth and various stool characteristics were eligible. RESULTS: 7 trials (n = 417) were included. Five trials (n = 345)
reported on the incidence of NEC, 3 trials (n = 295) reported on the incidence of late onset sepsis. Meta-analysis revealed a pooled RR (95% CI) of 1.24 (0.56-2.72) for NEC, 0.81 (0.57-1.15), p 0.23 for the risk of late onset sepsis. 3 individual trials (n = 295) did not observe any improvement in time to enteral feeds post intervention. Meta-analysis indicated a statistically significant difference in the growth of bifidobacteria in the oligosaccharide group with a weighted mean difference of 0.53 (95% CI: 0.33, 0.73) *10(6) colonies/g, p < 0.00001. A reduction in stool viscosity and pH was also observed. None of the trials reported life threatening adverse effects. CONCLUSIONS: Supplementation with prebiotic oligosaccharides was safe and did not result in decreased incidence of NEC, late onset sepsis and time to full enteral feeds but resulted in a significantly higher growth of beneficial microbes.

Stanko D, Bergesio R, Davies K, Hegarty M and von Ungern-Sternberg BS.
Postoperative pain, nausea and vomiting following adeno-tonsillectomy - a long-term follow-up.
BACKGROUND: Adenotonsillectomy is a common pediatric surgical procedure. Our knowledge of the recovery profile, parental understanding, and expectations is limited. We aimed to assess the incidence of pain, nausea, and vomiting in children undergoing adenotonsillectomy on postoperative day 3 and 7. We also wished to evaluate parental understanding regarding discharge instructions as well as parental expectations and experience of their child’s recovery. METHODS: We enrolled 100 children (0-16 years) undergoing elective adenotonsillectomy. On day 3 and 7, parents were questioned about their child's level of pain, nausea/vomiting and their understanding regarding postoperative instructions. RESULTS: Hundred children (median, 6.68 years) were recruited. 52% of parents rated their child's pain as VAS >= 5 on day 3, dropping to 30% by day 7. Almost 33% of patients experienced nausea on day 3, dropping to 11.6% by day 7. A similar trend was observed for postoperative vomiting. Most parents, 89%, agreed that postoperative instructions were clear. However, knowledge regarding when to seek emergency medical advice was found to be lacking. On day 7, only 44% of parents reported that their child's recovery met their expectations. CONCLUSION: Adenotonsillectomy is associated with significant pain and PONV, persisting into the seventh postoperative day. Parental education and information seems inadequate and needs to be improved.

Steer AC, Dale JB and Carapetis JR.
Progress toward a global group a streptococcal vaccine.

Stick S, Tiddens H, Aurora P, Gustafsson P, Ranganathan S, Robinson P, Rosenfeld M, Sly P and Ratjen F.
Early intervention studies in infants and preschool children with cystic fibrosis: are we ready?
Cystic fibrosis (CF) lung disease starts early in life and progresses even in the absence of clinical symptoms. Therefore, sensitive outcome measures to quantify and track these early abnormalities in infants and young children are needed; both for clinical care and interventional trials. Currently, the efficacy of most therapeutic interventions in CF has not been tested in children under the age of 6 years and drug development programmes have focused on assessing safety rather than efficacy in this age group. This article summarises the current status for outcome measures that can be utilised in clinical trials in infants and children with CF. Two methodologies are specifically highlighted in this review; chest computed tomography to assess structural damage of the lung and multiple breath washout as a technique to quantify ventilation inhomogeneity. While not all questions regarding the utility of these outcome measures in infants and young children have been resolved, significant advances have been made and it now appears feasible to design and conduct adequately powered efficacy studies in this age group. This could be a crucial step to further improve outcomes in CF patients as initiating effective treatment early is considered essential to prevent permanent lung damage.

Tanny SPT, Yong MS, d’Udekem Y, Kowalski R, Wheaton G, D’Orsogna L, Galati JC, Brizard CP and Konstantinov IE.

Taylor LJ, Maybery MT, Wray J, Ravine D, Hunt A and Whitehouse AJ.
Brief Report: Do the Nature of Communication Impairments in Autism Spectrum Disorders Relate to the Broader Autism Phenotype in Parents?

Thom GA and Cheah KC.
Perforating Foreign Body Reaction to Unheated Liquid Contents of Lava Lamp. Pediatric dermatology. 2013. A 21-month-old girl developed a local skin reaction after the unheated liquid contents of a broken lava lamp were in contact with her skin overnight. Several weeks later, small umbilicated erythematous papules containing central keratotic spines developed within the affected areas. Biopsy showed a granulomatous foreign body reaction with focal transepidermal elimination. Electron microscopy and energy-dispersive X-ray spectroscopy analysis of the tissue revealed carbon-based material, consistent with substances reported to be present in lava lamp liquid.


Tu J, Foster RS, Bint LJ and Halbert AR. Topical rapamycin for angiofibromas in paediatric patients with tuberous sclerosis: Follow up of a pilot study and promising future directions. The Australasian journal of dermatology. 2013. One of the most visible and potentially disfiguring cutaneous manifestations of tuberous sclerosis complex is the development of multiple facial angiofibromas, present in over 80% of patients. Topical rapamycin has been shown in many reports to be a safe and effective treatment for facial angiofibromas. In February 2012 we reported the results of a pilot study of four patients undertaken at a paediatric tertiary hospital in Australia. Since then, we have continued to refine the optimal formulation and concentration of topical rapamycin and expanded our selection of patients. We present an update on our current cohort of treated patients, discuss the optimal formulation of topical rapamycin and include a literature review on all published cases to date. Although topical rapamycin is not a curative treatment, we have demonstrated that its early institution significantly reduces both the vascularity and palpability of angiofibromas and prevents their progression with age. It is well tolerated and now a cost effective option.


von Ungern-Sternberg BS, Davies K, Hegarty M, Erb TO and Habre W. The effect of deep vs. awake extubation on respiratory complications in high-risk children undergoing adenotonsillectomy: a randomised controlled trial. Eur J Anaesthesiol. 2013; 30(9): 529-536. CONTEXT: There is ongoing debate regarding the optimal timing for tracheal extubation in children at increased risk of perioperative respiratory adverse events, particularly following adenotonsillectomy. OBJECTIVE: To assess the occurrence of perioperative respiratory adverse events in children undergoing elective adenotonsillectomy extubated under deep anaesthesia or when fully awake. DESIGN: Prospective, randomised controlled trial. SETTING: Tertiary paediatric hospital. PATIENTS: One hundred children (<16 years), with at least one risk factor for perioperative respiratory adverse events (current or recent upper respiratory tract infection in the past 2 weeks, eczema, wheezing in the past 12 months, dry nocturnal cough, wheezing on exercise, family history of asthma, eczema or hay fever as well as passive smoking). INTERVENTION: Deep or awake extubation. MAIN OUTCOME MEASURE: The occurrence of perioperative respiratory adverse events (laryngospasm, bronchospasm, persistent coughing, airway obstruction, desaturation <95%). RESULTS: There were no differences between the two groups with regard to age, medical and surgical parameters. The overall
incidence of complications did not differ between the two groups; tracheal extubation in fully awake children was associated with a greater incidence of persistent coughing (60 vs. 35%, \( P = 0.028 \)), whereas the incidence of airway obstruction relieved by simple airway manoeuvres in children extubated while deeply anaesthetised was greater (26 vs. 8%, \( P = 0.03 \)). There was no difference in the incidence of oxygen desaturation lasting more than 10 s. CONCLUSION: There was no difference in the overall incidence of perioperative respiratory adverse events. Both extubation techniques may be used in high-risk children undergoing adenotonsillectomy provided that the child is monitored closely in the postoperative period. TRIAL REGISTRATION: Australian New Zealand Clinical Trials Registry: ACTRN12609000387224.

Walters M, Claes P, Kakulas E and Clement JG.
Robust and regional 3D facial asymmetry assessment in hemimandibular hyperplasia and hemimandibular elongation anomalies.
Hemimandibular hyperplasia (HH) and hemimandibular elongation (HE) anomalies present with facial asymmetry and deranged occlusion. Currently, diagnosis and assessment of the facial dysmorphology is based on subjective clinical evaluation, supported by radiological scans. Advancements in objective assessments of facial asymmetry from three-dimensional (3D) facial scans facilitate a re-evaluation of the patterns of facial dysmorphology. Automated, robust and localised asymmetry assessments were obtained by comparing a 3D facial scan with its reflected image using a weighted least-squares superimposition. This robust superimposition is insensitive to severe asymmetries. This provides an estimation of the anatomical midline and a spatially dense vector map visualising localised directional differences between the left and right hemifaces. Analysis was conducted on three condylar hyperplasia phenotypes confirmed by clinical and CT evaluation: HH; HE; and hybrid phenotype. The midline extraction revealed chin point displacements in all cases. The upper lip philtrum and nose tip deviation to the affected side and a marked asymmetry of the mid face was noted in cases involving HE. Downward and medial rotation of the mandible with minor involvement of the midface was seen in the HH associated deformity. The hybrid phenotype case exhibited asymmetry features of both HH and HE cases.

Transcription Factor p63 Regulates Key Genes and Wound Repair in Human Airway Epithelial Basal Cells.

Watson HJ, Fursland A, Bulik CM and Nathan P.
Subjective binge eating with compensatory behaviors: A variant presentation of bulimia nervosa.
OBJECTIVES: To determine whether a variant bulimic-type presentation, whereby one meets criteria for bulimia nervosa (BN) except that binge eating episodes are not objectively large (i.e., “subjective bulimia nervosa,” SBN), has comparable clinical severity to established eating disorders, particularly BN. METHOD: Treatment-seeking adults with BN (N = 112), SBN (N = 28), anorexia nervosa restricting type (AN-R) (N = 45), and AN-binge/purge type (AN-B/P) (N = 24) were compared. RESULTS: Overall, SBN could not be meaningfully distinguished from BN. SBN and BN had equivalent eating pathology, depression and anxiety symptoms, low quality of life, impulsivity, Axis I comorbidity, and lifetime psychiatric history, and comparable clinical severity to AN-R and AN-B/P. DISCUSSION: Individuals with SBN, differing from BN only by the smaller size of their binge eating episodes, had a form of eating disorder comparable in clinical severity to threshold AN and BN and warranting clinical attention. Health professionals and the community require greater awareness of this variant to optimize detection, treatment-seeking, and outcomes. (c) 2012 by Wiley Periodicals, Inc. (Int J Eat Disord 2013).

Watson HJ, Fursland A and Byrne S.
Treatment engagement in eating disorders: who exits before treatment?
OBJECTIVE: Traditionally, drop-out has been investigated subsequent to treatment entry; yet some individuals "exit early," attending assessment but failing to commence the treatment offered. Early exit burdens administrative and clinical resources and means that individuals may not receive the care needed for recovery. This study aimed to describe the prevalence and characteristics associated with early exit at a statewide, outpatient eating disorder service. METHOD: From a pool of 972 consecutive referrals of adults and youth (16+ years), two groups were formed; an "early exit" group of individuals who attended assessment but chose to exit the service prior to treatment entry and a "non-early exit" group that attended assessment and entered treatment. The groups were compared on sociodemographic, clinical, and administrative features. RESULTS: The prevalence of early exit was 18.7%. The early exit group exhibited less pathology compared with the non-early exit group; specifically, a lower presence of Axis I comorbidity (p = .04) and self-induced vomiting (p =
DISCUSSION: The findings, considered in the context of previous research, suggest that there are no measured features to date that are robustly associated with early exit from outpatient eating disorder services. Future research should investigate decision-making processes at assessment, to inform patient-centered approaches that optimize transition to treatment.


Wilson S, Bremner AP, Mathews J and Pearson D. The Use of Oral Sucrose for Procedural Pain Relief in Infants Up to Six Months of Age: A Randomized Controlled Trial. Pain Manag Nurs. 2013; 14(4): e95-e105. The aim of this study was to evaluate the effectiveness of oral sucrose in decreasing pain during minor procedures in infants of 1-6 months corrected age. A blinded randomized controlled trial with infants aged 4-26 weeks who underwent venipuncture, heel lance or intravenous cannulation were stratified by correction age into >4-12 weeks and >12-26 weeks. They received 2 mL of either 25% sucrose or sterile water orally 2 minutes before the painful procedure. Nonnutritional sucking and parental comfort, provided in adherence to hospital guidelines, were recorded. Pain behavior was recorded using a validated 10 point scale at baseline, during and following the procedure. Data collectors were blinded to the intervention. A total of 21 and 20 infants received sucrose and water, respectively, in the >4-12-week age group, and 21 and 22, respectively, in the >12-26-week age group. No statistical differences were found in pain scores between treatment and control groups at any data collection points in either age group. Infants aged >4-12 weeks who did nonnutritional sucking showed statistically significantly lower median pain scores at 1, 2, and 3 minutes after the procedure than those who did not suck. Infants aged >4-26 weeks exhibited pain behavior scores that indicated moderate to large pain during painful procedures; however, there was insufficient evidence to show that 2 mL 25% sucrose had a statistically significant effect in decreasing pain. Infants should be offered nonnutritional sucking in compliance with the Baby Friendly Health Initiative during painful procedures.


androgen receptor gene (AR) mutation. DESIGN: Exons 2-7 of NR5A1 were PCR amplified and sequenced. Gene expression and cellular localization studies were performed on a novel NR5A1 variant c.744A>G (p.Y25C) identified in this study. RESULTS: We identified one novel mutation, c.744A>G (p.Y25C) in a patient characterized by penoscrotal hypospadias with bifid scrotum. He had elevated testosterone and gonadotropins in early infancy. Functional analysis of p.Y25C in vitro demonstrated reduced transcriptional activation by SF-1 and partially impaired nuclear localization in a proportion of transfected human adrenal NCI-H295R cells. CONCLUSION: This is the first reported case of a DSD patient with a NR5A1 mutation and elevated testosterone levels. Our finding supports evaluation of NR5A1 mutations in 46,XY DSD patients with a range of testosterone levels.

Wu M, Sabbaghian N, Xu B, Addidou-Kalucki S, Bernard C, Zou D, Reeve A, Eccles M, Cole C, Choong C, Charles A, Tan T, Iglesias D, Gooyer P and Foulkes W. Bi allelic DICER1 mutations occur in Wilms tumours. J Pathol. 2013. DICER1 is an endoribonuclease central to the generation of microRNAs (miRNAs) and short interfering RNAs (siRNAs). Germ-line mutations in DICER1 have been associated with a pleiotropic tumour predisposition syndrome and Wilms tumour (WT) is a rare manifestation of this syndrome. Three WTs, each in a child with a deleterious germ-line DICER1 mutation, were screened for somatic DICER1 mutations and were found to bear specific mutations in either the RNase IIIa (n = 1) or RNase IIIb domain (n = 2). In the two latter cases, we demonstrate that the germ-line and somatic DICER1 mutations were in trans, suggesting that the two-hit hypothesis of tumour formation applies for these examples of WT. Among 191 apparently sporadic WTs, we identified 5 different missense or deletion somatic DICER1 mutations (2.6%) in 4 individual WTs; one tumour has two very likely deleterious somatic mutations in trans in the RNase IIIb domain (c.5438A>G and c.5452G>A). In vitro studies of two somatic single base substitutions (c.5429A>G and c.5438A>G) demonstrate exon 25 skipping from the transcript, a phenomenon not previously reported in DICER1. Further we show that DICER1 transcripts lacking exon 25 can be translated in vitro. This study has demonstrated that a subset of WTs exhibit two "hits" in DICER1, suggesting that these mutations could be key events in the pathogenesis of these tumours.


Zappia T, Peter S, Hall G, Vine J, Martin A, Munns A, Shields L and Verheggenn M. Home oxygen therapy for infants and young children with acute bronchiolitis and other lower respiratory tract infections: the HI ThOx program. Issues Compr Pediatr Nurs. 2013; 36(4): 309-318. Background: Acute lower respiratory tract infection (LRTI) including bronchiolitis, is one of the leading causes of pediatric hospital admissions worldwide. Recent studies have demonstrated that some children with acute bronchiolitis can be successfully managed using home oxygen therapy. Aim: To report the impact of a Hospital in The Home oxygen therapy program (HI ThOx) for selected infants and young children with acute bronchiolitis and other LRTI. Findings: The HI ThOx program appears to be a safe model of care for carefully selected infants and young children with acute bronchiolitis and LRTI that reduces the hospital length of stay. Conclusions: The HI ThOx program provides an alternative model of care for infants and young children with acute LRTI. Implementation of models of care similar to that of the HI ThOx program in other pediatric health services may have the potential to create additional bed capacity, at the time of year when it is most needed.