1938 2013
The Royal Australasian College of Physicians
75 years SERVING FOR EXCELLENCE

VOLUME 50 SUPPLEMENT 2 MAY 2014

ISSN 1034-4810

Journal of Paediatrics and Child Health

Abstracts for the RACP Future Directions in Health Congress 2014
6–9 May 2014
Brisbane Convention & Exhibition Centre
Brisbane, Queensland, Australia

WILEY Blackwell

VOLUME 50 – SUPPLEMENT 2 – MAY 2014

Journal of Paediatrics and Child Health

Abstracts for the RACP Future Directions in Health Congress 2014
6–9 May 2014
Brisbane Convention & Exhibition Centre
Brisbane, Queensland, Australia

Plenary Speaker Abstracts

SUNDAY 18 MAY 2014

SCIENCE AND PUBLIC POLICY – RECONCILING TWO CULTURES

Peter Gluckman

Chief Science Advisor to the Prime Minister of New Zealand and Liggins Institute, University of Auckland, New Zealand

Science and technology are central to addressing many of the major issues confronting governments – from issues such as climate change, food, energy and water security to those emerging from major demographic and sociological changes. Yet whereas science is often perceived of as an endeavour creating certainty and some scientists have a tendency to minimize the limits of knowledge, what is sometimes called "post-modern science" is dealing with increasingly complex areas where policy decisions are urgent, facts are uncertain, high public interest and there are important values components. Some scientists and professionals are confused by the apparent lack of trust of, and consideration, of the evidence within the policy process, but policy is rarely determined by evidence alone. The confusion of scientific roles between advocacy and knowledge brokerage can create further tension and engender mistrust. Conversely science can be used as a proxy for debates that are really about values – for example in the climate change debate. There is an emerging understanding that science is a critical and potentially privileged input into the policy process but that requires an understanding of presenting objective knowledge in an as "values-free" way as possible and acknowledging the inferential gap between what is known and what is not. The role of the knowledge broker is to assist both the public and policy maker to better understandings of what we know and what we do not know. It is for the policy maker and politician to place values ranging from public opinion to fiscal and diplomatic considerations upon that knowledge – that is their role within a participatory democracy. Public policy will better served by greater scientific literacy on one hand and a greater understanding of the dangers of scientific hubris on the other. Only then will science be better employed to address the challenges that our societies face.

MONDAY 19 MAY 2014

Priscilla-Kincaid Smith Oration

EVIDENCE BASED INTRODUCTION OF HEALTH TECHNOLOGY AND DISINVESTMENT – TWO SIDES OF THE SAME COIN

Richard King AM

Over the past decade there has been an increase in health technology spending that has been estimated to be 25% of the increase in health costs,which cannot continued to be funded. In addition there has been an increased focus on patient safety. This talk will discuss our role, as a profession, in the safe, effective and affordable introduction of health technology and clinical procedures, and provide a framework as to how this might be done.

There is a groundswell of activity at international, national and institutional levels to do what is called

Disinvestment. There is a need to identify those procedures and technologies for which there is little or no evidence of efficacy or safety. There also a need to consider the concepts of Return on lnvestment(ROI) and Substitutional Reinvestment, as ways of not only funding new technology, but choosing something that is not only is more effective, but costs less. The ability to do this at an institutional level is one of the challenges. The value of "Low Value" lists and their place will be explored. Finally, a series of examples of success and failure and the reasons for both will be discussed, as will the potential role of the College going into the future.

TUESDAY 20 MAY 2014

Howard Williams Oration

THE LIFE-LONG LEGACIES OF PERINATAL MANAGEMENT

Jane Harding

Liggins Institute, University of Auckland, Auckland, New Zealand

Liggins and Liley, both working in Auckland in the 1960s, first developed new ways to treat babies before birth and markedly improved their chances of survival. We now know that fetal adaptations to the intrauterine environment have life-long implications for future health, but the longer-term health effects of specific therapeutic interventions have been more difficult to determine. New evidence about the long-term consequences of antenatal corticosteroid treatment and intrauterine transfusion for Rhesus haemolytic disease has important implications for how we treat babies today, and for their life-long health. Our research aims to incorporate these understandings into developing the best treatments for vulnerable babies both before and after birth.

FERGUSON-GLASS ORATION

Heron, Richard JL – 823

ABSTRACT NOT AVAILABLE AT THIS TIME

WEDNESDAY 21 MAY 2014

Redfern Oration

DECOLONISING MEDICAL EDUCATION AND PRACTICE TO ADVANCE INDIGENOUS HEALTH

Rhys Jones

University of Auckland

Health professionals are faced with an uncomfortable truth: our work predominantly serves to perpetuate social and ethnic inequalities in health. This tends to happen despite our best intentions and efforts, and is the result of a complex interplay of factors at different levels. In settler societies such as Australia and New Zealand, health and education systems are founded on colonial values, ideologies and practices; as a result health care serves to reinforce European privilege and Indigenous disadvantage.

In response to these disparities, elements of cultural competence and Indigenous health have been incorporated into educational curricula and health care environments. However these approaches will be ineffective unless professional and organisational cultures are also addressed. There is a need to fundamentally transform the way we educate and train doctors, the way we practice, and the systems in which we provide health care. Suggested approaches to 'decolonise' medical education and practice will be discussed with a view to improving Indigenous health and advancing equity.

THE DEATH OF DIAGNOSIS?

Rod Jackson

University of Auckland

Diagnosis is generally practiced as a binary process; patients either have a condition/disease or not. However most clinical conditions are neither completely present nor completely absent so binary diagnoses seldom reflect biology. Aside from death, certain infections and some injuries, almost all diseases, including most cancers are 'measured' on a continuous scale. Moreover the diagnoses of diseases like diabetes, atherosclerosis, mental disorders and most cancers are not only based on somewhat arbitrary thresholds levels of a measurement but there are often significant errors in the measurements.

These fundamental flaws in diagnostic practice are further compounded by the many other factors influencing a person's risk of disease. For example, the risk of symptomatic cardiovascular (CVD) events in people diagnosed with

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

binary conditions like diabetes, hypertension, or hyperlipidaemia, can vary more than ten or twenty-fold depending on age, gender and the presence of other CVD risk factors.

The alternative approach to diagnosis is individual risk prediction. Firstly, continuous risk better reflects the biology of most diseases and secondly, it reduces the need for choosing arbitrary thresholds because the patient, advised by their health practitioners, would ideally make the decision on whether the risk-benefit-harm balance justifies the type and intensity of treatment proposed. For example at what level of CVD risk would you agree to takes statins every day for the rest of your life, or what level of symptomatic prostate cancer risk justifies a prostatectomy? In fact it is impossible for a patient and a practitioner to have an informed discussion about the implications of a diagnosis without information on the individual's pre-treatment risk in addition to the benefits and harms of interventions.

There are however numerous challenges to using a risk prediction alternative to binary diagnoses. The two major ones are generating the necessary data for developing accurate patient-specific risk prediction and then communicating this information to patients. Addressing the data problem is becoming easier in the era of big data with the increasing ability to individually link health data on millions of people. The second problem of communicating risk to patients will be a greater challenge because most health professionals don't understand enough about risk themselves, let alone have the confidence or competence to communicate this with patients.

References:

1. Vickers A, Basch E, Kattan M. Against Diagnosis. *Ann Intern Med.* 2008;**149**:200–3.
2. Moynihan R. A new deal on disease definition. *BMJ.* 2011;**342**:d2548 doi: 10.1136/bmj.d2548.

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

Physicians as Medical Experts – P&CHD
Oral Abstracts

PAEDIATRIC HOSPITAL PRACTICE – FREE PAPERS

THE TRAJECTORY TOWARDS CHRONIC KIDNEY DISEASE AMONG ABORIGINAL YOUNG PEOPLE: THE ARDAC STUDY

Kim S[1,2], Hodson EM[1,2], Daylight J[1], Williams R[1], Vukasin N[1], Kearns R[1], Lyle DM[3] , Macaskill P[2], Craig JC[1,2]

[1]Centre for Kidney Research, The Children's Hospital at Westmead, Australia
[2]Sydney School of Public Health University of Sydney, Australia
[3]Department of Rural Health, University of Sydney, Australia

Background: The gap in chronic disease between Aboriginal and non-Aboriginal people remains substantial. We aimed to determine whether the increased prevalence of chronic kidney disease (CKD) in Aboriginal adults becomes evident in adolescence through examining early markers of chronic disease.

Methods: A prospective cohort study of Aboriginal and non-Aboriginal school children commenced in 2002 across 15 different screening centres involving 38 primary schools and 213 high schools across urban, regional and remote NSW. We have collected data on haematuria, albuminuria, blood pressure and BMI every 2 years.

Results: 3418 (1949 Aboriginal) participants were screened with 11,387 patient years of follow up; 67% of participants attended follow up. The average age at enrolment was 10 years. At baseline, 31% of the cohort was either overweight or obese; a significantly greater proportion of Aboriginal participants were overweight or obese (33% versus 29%, P = 0.02). At baseline Aboriginal participants were more likely to have albuminuria (12.6% versus 10.1%, P = 0.03) and haematuria (6.9% versus 3.5%, P < 0.01). Overall risk factors for albuminuria were increasing age (adjusted odds ratio [AOR] increase by each year over 10 years: 1.17, 95% confidence intervals [CI] 1.14–1.19, P < 0.01) and female gender (AOR 1.73 95% CI 1.49–2.01, P < 0.001). Aboriginal participants were more likely to have albuminuria when overweight or obese compared with non-Aboriginal participants. Among Aboriginal and non-Aboriginal participants, being underweight presented a greater risk of developing either transient (AOR: 0.88, 95% CI 0.80–0.96) or persistent albuminuria (AOR 0.75, 95% CI 0.64 to 0.88).

Conclusion: Weight gain increases the risk of albuminuria for Aboriginal compared with non-Aboriginal participants. Under nutrition increases the risk of albuminuria in all participants. Community based screening of participants in the ARDAC study will continue to assess whether the risk for albuminuria changes during early adulthood.

PREDICTORS OF OUTCOME IN CHILDREN HOSPITALISED WITH PANDEMIC INFLUENZA IN 2009: A PROSPECTIVE NATIONAL STUDY

Gulam Khandaker[1,2,12], Yvonne Zurynski[1,4], Greta Ridley[1,4], Jim Buttery[5], Helen Marshall,[6] Peter C Richmond FRACP,[7] Jenny Royle MD,[8] Michael Gold MD,[9] Tony Walls FRACP,[10] Bruce Whitehead,[11] Peter McIntyre,[2,3,12] Nicholas Wood,[2,3] Robert Booy[2,3,12] and Elizabeth J Elliott[1,2,3,4,12]

[1]The Discipline of Paediatrics and Child Health, The University of Sydney, Sydney, NSW, Australia,
[2]National Centre for Immunisation Research and Surveillance of Vaccine Preventable Diseases, The Children's Hospital at Westmead, Sydney, New South Wales, Australia
[3]The Sydney Children's Hospitals Network (Westmead), Sydney, NSW, Australia
[4]The Australian Paediatric Surveillance Unit, Sydney, NSW, Australia
[5]Murdoch Children's Research Institute and Monash Children's Hospital, Department of Paediatrics, Monash University, Melbourne, Victoria, Australia,
[6]Vaccinology and Immunology Research Trials Unit, Women's and Children's Hospital, Adelaide, South Australia, Australia,
[7]School of Paediatrics and Child Health, University of Western Australia, Perth, Western Australia,
[8]Immunisation Service, Department of General Medicine, Royal Children's Hospital, Melbourne, Victoria, Australia,
[9]School of Paediatrics and Reproductive Health, University of Adelaide, South Australia,
[10]Department of Paediatrics, University of Otago, Christchurch, New Zealand,
[11]Respiratory Medicine, John Hunter Children's Hospital, NSW, Australia,
[12]The Marie Bashir Institute for Infectious Diseases and Biosecurity (MBI), Sydney Medical School, the University of Sydney, Sydney, NSW, Australia

Background: Seasonal influenza is an important cause of hospital admission in children. The impact of the 2009 H1N1 pandemic posed enormous burden on paediatric hospital services.

Aims/Objectives: We aimed to describe the clinical epidemiology and examine predictors for adverse outcomes in children hospitalised in Australia with pandemic influenza.

Methods: Active hospital surveillance in 6 tertiary paediatric referral centres (June-September, 2009) for all children aged <15 years admitted with laboratory-confirmed pandemic influenza.

Findings: Of 601 children admitted with laboratory proven influenza, 506 (84.2%) had influenza A(H1N1)pdm09. Based on annual admissions to surveillance hospitals, the hospital admission rate for influenza was 33.9 per 1,000 admissions in 2009. Half (51%) the children with influenza A(H1N1)pdm09 were previously healthy. The mean length of hospital stay was 5.9 days. Rapid diagnosis decreased antibiotic use and length of hospital stay. Fifty (9.9%) children were admitted to paediatric intensive care (PICU), 30 (5.9%) required mechanical ventilation and 5 (0.9%) died. Laboratory proven bacterial co-infection and chronic lung disease were significant independent predictors of PICU admission (OR 6.89, 95% CI 3.15, 15.06 and OR 3.58, 95% CI: 1.41, 9.07 respectively) and requirement for ventilation (OR 5.61, 95% CI: 2.2, 14.28 and OR 5.18, 95% CI: 1.8, 14.86 respectively). Chronic neurological disease was a predictor of admission to PICU (OR 2.30, 95% CI: 1.14, 4.61).

Conclusions: During the 2009 pandemic, influenza was a major cause of hospitalisation in tertiary paediatric hospitals. Co-infection and underlying chronic disease increased risk of PICU admission and/or ventilation. Half the children admitted were previously healthy, supporting a role for universal seasonal influenza vaccination in children.

CAN CHILDREN WITH MODERATE TO SEVERE CELLULITIS BE EFFECTIVELY TREATED AT HOME WITH ONCE DAILY INTRAVENOUS ANTIBIOTICS?

Vicki Burneikis, Robert Parry

Central Coast Local Health District, New South Wales, Australia

Background: Although previous papers have shown that ambulatory management of moderate to severe cellulitis is possible with once daily intravenous (IV) antibiotics, failure rates were 9–21%, and there is no concensus on the most appropriate antibiotic regime[1,2].

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

Aim: To document rates of complications and success or failure of ambulatory management, in all children (0–17 years) with cellulitis (excluding periorbital cellulitis) managed with IV antibiotics through ambulatory care at Gosford and Wyong hospitals between 1st January and 31st December 2013.

Method: Details of cases were collected prospectively during the study period, and electronic records were used to gather information regarding presentation, management and outcome. Failure of ambulatory management was defined as the need for hospital admission, and was the primary outcome measure. Secondary outcomes included need for change in antibiotic regime, complications, unplanned emergency attendance during treatment, and representation with a related diagnosis within one month.

Results: 30 cases were found, and of these 29 (97%) were successfully managed in the ambulatory care units, without admission. There were no unplanned attendances, and no patient represented within a month of diagnosis.

24 patients received daily IV ceftriaxone and flucloxacillin, plus oral flucloxacillin 6 hourly, 6 received daily IV ceftriaxone only, and 2 received daily IV cefazolin and 6 hourly oral probenecid. 3 patients (10%) required a change in antibiotic due to poor response after 48 hours, one (3%) required incision and drainage of an abscess, which was done under local anaesthesia in the ambulatory care unit.

Of the 3 regimes, patients treated with cefazolin and probenecid were significantly more likely to require a change in antibiotic regime (OR 8.0, 95% CI 5.4–9.6, $p < 0.01$), compared with those treated with ceftriaxone with or without flucloxacillin. Ceftriaxone alone appeared less successful than the combination of ceftriaxone and flucloxacillin, although results were not statistically significant.

Age was a significant risk factor for poor response at 48 hours, with 4 of 7 patients aged 12 or over needing to change antibiotic, compared with none of 23 younger patients ($p < 0.05$).

Conclusion: 97% of our moderate to severe cellulitis patients were successfully managed through ambulatory care without admission or recurrence.

References:

1. Kam AJ et al, "Pediatric Cellulitis, Success of Emergency Department Short-Course Intravenous Antibiotics", *Ped Em Care* 2010
2. Gouin S et al, "Prospective Evaluation of the Management of Moderate to Severe Cellulitis with Parenteral Antibiotics in a Paediatric Day Treatment Centre", *J Paed &Ch Health* 2008

PAEDIATRIC DEPARTMENT NIGHT SHIFT AUDIT, CAMPBELLTOWN HOSPITAL, SW SYDNEY

Passarello L[1], Vosu J[1], Datta R[2], Chin R[2], Edwards M[1], Marsh D[2], Whitehall J[1]

[1]University of Western Sydney, New South Wales, Australia
[2]Campbelltown Hospital, New South Wales, Australia

Time and motion studies of paediatric junior staff are rare. Campbelltown Hospital is a busy peripheral hospital in Sydney with 24 paediatric and 16 special care neonatal beds (SCN), serving nearly 5000 paediatric hospitalisations, 3000 deliveries, and 5000 nocturnal ED attendances, annually. One paediatric registrar is rostered from 11.00 pm to 9.00 am to cover the wards and consult in ED to trainees of that specialty. Because of concerns of over-work it was decided to closely examine the registrar's activities.

Methods: For six weeks, the registrar was shadowed by one of two medical students who recorded each task (according to 14 definitions), its duration (five minute intervals) and location. Data was recorded by hand and entered in a spread sheet in the morning.

Results: 1,195 activities by five registrars were recorded over 264 hours. Percentage of time in each task is reported in Figure 1. The percentage of time spent in various locations is reported in Figure 2.

Discussion: The time and motion study reveals total commitment by the registrar to patient care in three competing sites in different parts of the hospital at the same time. There were no rostered breaks. <5 min/shift was taken rest and ablutions. All registrars returned home fatigued in traffic.

Bedside Patient Review	25.9%
Handover	25.4%
Reading /Writing Notes	16.2%
Phone Consultations	7.3%
Procedures	7.3%
Prescribing Medications	5.2%
Talking with Relatives	4.3%
Reviewing Test Results	2.1%
Ordering Tests	1.8%
Private Study	1.4%
Communications with consultants	1.2%
Consulting Guidelines	0.8%
Rest/Break	0.7%
Non patient Administration	0.3%

Figure 1. Percentage of Time per Activity.

Ward	46.3%
ED	39.7%
SCN	9.9%
Birthing Unit	2.1%
Theatres	1.9%

Figure 2. Percentage of Time per Location.

HOW SAFE ARE OUR KIDS? IMPROVING THE SAFETY AND QUALITY OF PAEDIATRIC CARE IN AN AUSTRALIAN ADULT ACADEMIC HEALTH SERVICE

Lynch C[1,2], Panozzo T[1], Reeves J[1]

[1]Alfred Health, Victoria, Australia
[2]Monash Childrens Hospital, Victoria, Australia

Introduction: Care of children occurs in many adult health services. Alfred Health is a large adult health network with a surprisingly large number of paediatric presentations. It was apparent that there was no transparent organization approach to ensuring safe effective and age appropriate care was provided.

Method: Royal Australasian College of Physicians (RACP) and National Safety and Quality Health Service Standards (NSQHS) have documented standards of care for children in hospitals Paediatric specific data was collected to provide an overview of paediatric care. RACP Audit tools revealed areas where standards were not met. An organization wide Paediatric Governance Committee oversaw implementation of a series of recommendations. Improvement was needed in facilities, resources and specific paediatric education for staff. These included introduction of age specific Paediatric Graphic observation charts, paediatric medication chart, monthly clinical data sets to allow for service delivery audits

Results: RACP and NSQHS audit tools were used pre and post implementation. Significant improvements included purpose built paediatric amenities to minimize colocation of children with adults .A new paediatric model of care in the emergency department includes the use of age specific observation charts and clinical guidelines. Regular audits with specific paediatric focus now occur. There has been an increased reporting of actual or near miss

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

clinical incidents. Graphic observation charts improved communication and allowed escalation of the deteriorating patient.

Conclusion: Paediatric patients cared for in predominantly adult health services are at risk without specialized and age specific systems of care. Accurate and meaningful data allows better understanding of service delivery. Executive sponsorship, multi service and multi disciplinary stakeholders are essential for change.

BIRTH THROUGH MECONIUM STAINED AMNIOTIC FLUID: DO ALL INFANTS REQUIRE 24 HOUR OBSERVATION IN HOSPITAL?

Gupta S, Birch P

Gold Coast University Hospital, Southport, Queensland, Australia

Background: Birth through meconium stained amniotic fluid (MSAF) can be associated with adverse neonatal outcomes leading many institutions to admit infants for a period of at least 24 hours of observation following birth.

Objective: To identify infants following birth through MSAF who would be at very low risk of requiring admission to the Newborn Care Unit (NCU) and would therefore be safe for early discharge at 6 hours of age.

Methods: This was a retrospective audit of term infants born through MSAF over an 11 month period from December2011 to November 2012

Results: 348 infants birthed through MSAF (111 per 1,000 live births). 2.6% of term infants born through MSAF developed meconium aspiration syndrome (MAS). 92.3% of term admissions following birth through MSAF occurred before 6 hours of age and no infant developed MAS beyond 6 hours of age. The development of MAS was associated with male gender (OR 8, 95% CI 66.7–1, $p = 0.05$), 5 minute Apgar score (5AS) < 9 (89% with MAS vs. 5% without MAS, $p < 0.0001$) and emergency caesarean section (40% with MAS vs. 11% without MAS, $p < 0.0001$). The 5AS was significantly lower in infants who developed MAS (6.3 vs. 8.9, $p = 0.006$). Having a 5AS ≥ 9 was strongly associated with not developing MAS (OR 0.007, CI 0.001–0.057, $p < 0.0001$) with 99.7% of 318 term infants who birthed through MSAF with a 5AS ≥ 9 not developing MAS.

Conclusion: Infants with 5AS ≥ 9 are unlikely to develop MAS and all infants with MAS developed symptoms within 6 hours of age. Rates of admission beyond 6 hours of age are low, therefore we conclude that for infants born through MSAF with a 5AS ≥ 9 it is safe for early discharge at 6 hours of age.

RUE WRIGHT MEMORIAL AWARD

PILOT STUDY OF eADVICE (ELECTRONIC ADVICE AND DIAGNOSIS VIA THE INTERNET FOLLOWING COMPUTERISED EVALUATION)

Caldwell PHY[1,2], Sureshkumar P[2], Hamilton S[2], Kerr M[2], Lau A[3], Craig JC[1,2]

[1]University of Sydney, New South Wales, Australia
[2]The Children's Hospital at Westmead (CHW), New South Wales, Australia
[3]University of NSW, New South Wales, Australia

Background: Waiting times for paediatric outpatient services are usually long, and is 24 months for the CHW continence service. Our research team have built a prototype interactive eHealth program/app (eADVICE) for managing paediatric incontinence, which can be accessed on the Internet and downloaded to a mobile device. This program follows an evidence-based algorithm for managing incontinence and transfers the exchange of information between the parents and specialists. It mimics multiple visits to a specialist paediatric continence service and combines assessment, diagnosis, tailored treatment advice, monitoring and feedback and well as education.

Aims: To assess the effects of an interactive eHealth for managing urinary incontinence in children awaiting a specialist appointment.

Methods: We conducted a pilot study of 10 children with urinary incontinence who used the eADVICE program for 4 months, supervised by their GP. We assessed whether the program provided the correct diagnosis and treatment advice, change in wetting from baseline, and adhered to the treatment advice given.

Results: The program was found to be accurate in assessing and providing appropriate treatment advice. Families visited the site an average of 3.2 times (range 2–6 visits) during the 4 months. 2 patients became completely dry, and another 2 improved using the program. However, up to 50% of treatment advice was not followed by families.

Conclusion: An interactive eHealth program is effective in providing tailored treatment advice for children. However, the poor adherence to eHealth advice needs to be addressed to improve the applicability of this methodology in clinical practice.

SPECIALIST HOME-BASED NURSING SERVICES FOR CHILDREN WITH ACUTE AND CHRONIC ILLNESSES (COCHRANE REVIEW)

Parab CS[1], Cooper C[2], Woolfenden S[3], Piper SM[4]

[1,4]Illawarra Shoalhaven Local Health District, Wollongong, Australia
[2]Royal North Shore Hospital, St Leonards, Australia
[3]Sydney Children's Hospitals Network, Randwick, Australia

Introduction: This review aimed to evaluate specialist home based nursing services as these were proposed as a cost-effective means of reducing distress resulting from hospital admissions, while enhancing primary care and reducing length of hospital stay.

Methods: The databases were searched electronically for initial screening of study titles and abstracts. Full text articles of the selected studies were then reviewed. The search was carried out independently by study authors and disagreements were resolved by consensus.

Results: Screening of 4226 titles yielded seven RCTs with a total of 840 participants. Participants, interventions and outcomes were diverse, thus metanalysis was not conducted. No significant differences were reported in health outcomes and hospital re-admission rates. Two studies reported a reduction in the hospital stay (2.37 vs 1.37 days, $p < 0.001$ and 96.9 hours vs 55.2 hours, $p = 0.001$). Two studies identified greater parent and child satisfaction. One study reported better parental coping and functioning ($p < 0.001$). Two studies respectively revealed no difference in the impact of illness on the family or parental burden of care. In terms of costing, one study reported parental cost savings of CAD 188 per child ($p < 0.001$) and increased cost to the hospital of CAD 87 ($p < 0.001$) with another study reporting similar findings qualitatively. One study did not report any significant cost difference.

Conclusion: Limited evidence base for home care programs; however suggestive evidence for greater parental satisfaction, improved quality of life and a reduction in the length of hospital stay. The cost-effectiveness of these programs is still to be determined. Further trials with adequate sample sizes, standardised clinical outcome measures and comprehensive costing analysis are required.

WILEY NEW INVESTIGATOR AWARD

PAEDIATRIC PRESENTATIONS TO A NON-TERTIARY PAEDIATRIC HOSPITAL OVER A 12 MONTH PERIOD: IMPLICATIONS FOR WORKFORCE PLANNING

Hardy A,[1] Fuller DG,[1,2,5] Forrester M,[1,2,5] Anderson PK,[1,2,5] Cooper C,[1,2,5] Jenner B,[1,2,5] Marshall I,[1,2,5] McCloskey K,[1,2,5] Sanderson C,[1,2,5] Standish J,[1,2,5] Worth J[1,2,5] and Vuillermin P (senior author)[1,2,3,4,5]

[1]Children's Services, Barwon Health, Geelong, Victoria, Australia
[2]Deakin University, Geelong, Victoria, Australia
[3]Murdoch Children's Research Unit, Victoria, Australia
[4]Child Health Research Unit, Barwon Health, Victoria, Australia
[5]St John of God Hospital, Geelong, Victoria, Australia

Introduction: Paediatricians working at non-tertiary hospitals have a substantial developmental and behavioural workload.[1] Data regarding the burden and scope of acute care is limited.

The present study aims to determine, for paediatricians working in non-tertiary hospitals:
(1) The volume and case mix of acute care paediatrics practised, and
(2) Whether the volume and case mix has changed since the 1990s.

Methods: Over a 12 month period (December 2012 to December 2013), acute paediatric inpatient, emergency department (ED) and neonatal case

© 2014 The Authors

mix was determined in the Barwon region, Victoria, Australia, by reviewing hospital database information. These data were compared to case mix data collected during 1996/1997.[2]

Results: Since 1996/1997, paediatric hospital admissions increased by 45% (95% confidence interval (CI) 38% to 52%; $p < 0.0001$) and presentations to the ED increased by 186% (95% CI 181% to 191%; $p < 0.0001$). A wide variety of problems are managed by paediatricians working in non-tertiary hospitals.

Conclusions: Paediatricians working in non-tertiary hospitals manage a diverse range of inpatients. General paediatric training and consultant paediatrician Continuing Medical Education programs should be designed to ensure the acquisition and maintenance of the knowledge and skills required to manage such patients. The significant increases in paediatric presentations to ED and paediatric admissions is relevant to workforce planning. If these trends continue, a greater number of general paediatricians with adequate acute care skills will be required to meet these needs.

References:

1. Hewson P. A 12-month profile of community paediatric consultations in the Barwon region. *Journal of Paediatrics and Child Health*. 1999; **35**: 16–22
2. Hewson P *et al*. The evolving role of community-based general paediatricians: The Barwon experience. *Journal of Paediatrics and Child Health*. 1999; **35**: 23–27

PROCEDURAL AND RESUSCITATION REQUIREMENTS FOR PAEDIATRICIANS WORKING IN A NON-TERTIARY CENTRE: IMPLICATIONS FOR TRAINING

Hardy A,[1] Fuller DG,[1,2,5] Forrester M,[1,2,5] Anderson PK,[1,2,5] Cooper C,[1,2,5] Jenner B,[1,2,5] Marshall I,[1,2,5] McCloskey K,[1,2,5] Sanderson C,[1,2,5] Standish J,[1,2,5] Worth J[1,2,5] and Vuillermin P (Senior Author)[1,2,3,4,5]

[1]Children's Services, Barwon Health, Geelong, Victoria, Australia
[2]Deakin University, Geelong, Victoria, Australia
[3]Murdoch Children's Research Unit, Victoria, Australia
[4]Child Health Research Unit, Barwon Health, Victoria, Australia
[5]St John of God Hospital, Geelong, Victoria, Australia

Introduction: Paediatricians working at non-tertiary hospitals are required to perform a variety of procedures and to lead paediatric resuscitations. Data regarding the scope of procedural skills required and the frequency with which procedural and resuscitation skills are utilised is required.

The present study aims to determine the procedural and resuscitation skills required by paediatricians working at non-tertiary hospitals and the frequency with which these skills are utilised.

Methods: Over a 12 month period (December 2012 to December 2013), each of the 11 paediatricians involved in acute inpatient care at the Geelong Hospital completed a weekly on-line survey regarding their inpatient clinical experience. This included procedures performed or directly supervised as well as their resuscitation involvement.

Results: Each of the 11 paediatricians who managed inpatients on a regular or semi-regular basis during the study period agreed to participate and each completed all of the weekly surveys. There were 7 Geelong Hospital paediatricians with an inpatient appointment (each with a 0.27 Full Time Equivalent (FTE) paediatrician workload) and 4 paediatricians providing inpatient cover on a locum basis. Apart from intravenous cannulation, paediatricians working in non-tertiary hospitals utilise procedural skills infrequently. Each 0.27 FTE paediatrician performed 0.86 intubations and was involved in 11.3 neonatal, 1.7 infant and 2.4 child resuscitations.

Conclusions: Paediatricians working at non-tertiary hospitals are required to perform and supervise critical procedural and resuscitation skills, but have limited opportunities to maintain proficiency in such skills. General paediatric training and consultant paediatrician Continuing Medical Education programs should be designed to ensure the acquisition and maintenance of the procedural and resuscitation skills required for the practice of non-tertiary acute care paediatrics.

MENSTRUAL MANAGEMENT FOR DEVELOPMENTALLY DELAYED GIRLS AT THE CHILDREN'S HOSPITAL WESTMEAD PAEDIATRIC AND ADOLESCENT GYNAECOLOGY OUTPATIENT CLINIC

Chuah I[1], Mcrae A[2], Steinbeck K[2,3]

[1]The Department of General Medicine, The Children's Hospital at Westmead, Sydney, Australia
[2]The Academic Department of Adolescent Medicine, The Children's Hospital at Westmead, Sydney, Australia
[3]Discipline of Paediatrics and Child Health, Sydney Medical School, University of Sydney, Australia

Introduction: Requests for assistance in menstrual management and menstrual suppression are a common, emotive and sometimes controversial aspect of disability care. Little has been published since the introduction of the levonorgestrel releasing intrauterine system (LG-IUS) has become available to the therapeutic armamentarium.

Methods: A retrospective review and detailed data collection from the medical records of all girls with physical and intellectual disability referred for menstrual management to the Paediatric and Adolescent Gynaecology clinic for the three year period between January 1, 2010 and January 1, 2013.

Results: Eighty girls were referred. A third (28) of girls were premenarchal at first review with caregivers seeking anticipatory advice. Of the postmenarchal girls the median age of menarche was 12 (range 10–15). Caregiver concerns of both groups were explored in detail, as were the menstrual characteristics of the post menarchal girls. We investigated the first line and second line interventions trialled and reasons for change.

Our population differ from similar previously published groups in the marked absence of the use of depot medroxyprogesterone acetate or the subdermal etonorgestrel-releasing device. The combined oral contraceptive pill (OCP) was the most often used therapy (67%) and 18 girls in total had a levonorgestrel releasing intrauterine system inserted (30%).

Conclusion: Premenarchal concerns are important to address as a paediatrician, it can be an opportunity to allay caregiver concerns and provide them with appropriate advice. Our study supports the use of the OCP as good first line management in achieving menstrual suppression. The LG-IUS appeared to be a satisfactory second line option. Further investigation into longer term outcomes and complications of the device insertion should be performed to determine its viability for the future.

P&CHD – NEONATES

HIGH-FLOW NASAL CANNULAE FOR THE TREATMENT OF PRETERM INFANTS: WHAT IS THE EVIDENCE?

Brett Manley[1,2]

[1]Neonatal Services and Newborn Research Centre, The Royal Women's Hospital, Melbourne
[2]Department of Obstetrics and Gynaecology, The University of Melbourne

Heated and humidified high-flow nasal cannulae (HFNC) are being used to treat preterm infants in the majority of neonatal intensive care units (NICUs) in the USA, United Kingdom, and Australasia. Until recently, this practice was not supported by evidence from clinical trials, as shown by the 2011 Cochrane Review on the topic. In recent times, non-tertiary centres have also begun to treat newborn infants with HFNC.

This talk will cover the following:

- The background to HFNC use in preterm infants and the proposed mechanisms of action of this therapy.
- Clinical scenarios in which HFNC are being used to treat preterm infants, including:
  - post-extubation support
  - primary respiratory support after birth
  - 'weaning' infants from nasal CPAP
- Results of clinical trials of HFNC, with particular emphasis on the three recently published randomised trials of HFNC vs. nasal CPAP in preterm infants:
  - Collins *et al* 2013
  - Yoder *et al* 2013
  - Manley *et al* 2013

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

- Recommendations for HFNC use in NICUs, including:
  - important safety principles
  - safe and effective gas flows
  - evidence-based clinical indications
- The use of HFNC in non-tertiary centres, including important factors to be considered when using non-invasive support in these centres
- Trials of HFNC which are proposed or underway, including randomised trials of HFNC as primary support for preterm infants with early respiratory distress in NICUs and non-tertiary special care nurseries.

TBC

Dargaville, Peter – 935

ABSTRACT NOT AVAILABLE AT THIS TIME

TBC

Malcolm Battin

ABSTRACT NOT AVAILABLE AT THIS TIME

P&CHD – RESPIRATORY

TBC

Vyas, Julian – 936

ABSTRACT NOT AVAILABLE AT THIS TIME

TBC

Dalziel, Stuart – 937

ABSTRACT NOT AVAILABLE AT THIS TIME

HIGH FLOW NASAL CANNULA, PHYSIOLOGICAL PRINCIPLES AND CLINICAL EVIDENCE

Schibler, Andreas – 938

ABSTRACT NOT AVAILABLE AT THIS TIME

P&CHD – NEUROLOGY

NEW GENES IN FOCAL EPILEPSY

Lynette Sadleir

3 ••

The focal epilepsies which are the most common epilepsies have not traditionally been considered genetic in origin. Recent discoveries show that genes have a role to play in both familial and sporadic cases of focal epilepsy and are revealing opportunities for better diagnosis, prognostication, genetic counselling and potential therapies.

EARLY DETECTION OF CEREBRAL PALSY

Alicia Spittle[1,2,3]

[1]Murdoch Childrens Research Institute, Parkville, Victoria, Australia
[2]The University of Melbourne, Parkville, Victoria, Australia
[3]The Royal Women's Hospital, Parkville, Victoria, Australia

Cerebral Palsy (CP) is an umbrella term which "describes a group of disorders of the development of movement and posture, causing activity limitations, which are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain."[1] Despite CP being the most common physical disability in childhood, the majority of children with CP are not diagnosed early in infancy, with the average age of diagnosis in Australia recently reported from CP registers as 19 months of age. Early detection of CP is important not only for counselling families but also to ensure that intervention is commenced in a timely manner. Research has shown that delays in diagnosis of CP are associated with dissatisfaction and grief for families. Further, emerging evidence from the neuroplasticity literature suggest that intensive, repetitive, task-specific intervention for CP ought to commence very early while the brain is most plastic

Identifying children at risk of CP early in development has improved over the past decade with greater understanding of the predictive value of early motor assessments and brain imaging. General Movements Assessments, which involve observation of an infant's spontaneous movement patterns, have been shown consistently in systematic reviews of early detection of CP to be the most predictive.[2] This presentation will provide an overview of the evidence for early detection of CP, with an emphasis on General Movements Assessments

1. Bax M, Goldstein M, Rosenbaum P, et al. Proposed definition and classification of cerebral palsy, April 2005. *Developmental medicine and child neurology*. Aug 2005;**47**(8):571–576.
2. Spittle AJ, Doyle LW, Boyd RN. A systematic review of the clinimetric properties of neuromotor assessments for preterm infants during the first year of life. *Developmental medicine and child neurology*. Apr 2008;**50**(4): 254–266.

P&CHD/AMD – INFECTIOUS DISEASES

TUBERCULOSIS. DIAGNOSIS AND CHALLENGES

Lesley Voss

Starship Children's Hospital, Auckland, New Zealand

Control of tuberculosis remains one of the millennium development goals with Stop TB Paternship and World Health Organisation establishing a target to reduce annual incidence to less than one case per million population by 2050. In Australia and New Zealand rates of TB have been stable or declining, both for adults and children, for many years with the large burden of disease in both these countries now from the foreign born population. Both countries have close links with a number of the WHO recognized high burden countries. To achieve the target of TB eradication a variety of new strategies including new diagnostics drugs and vaccines will be required.

An important component of TB management is diagnosis of the patient in a quick and efficient manner. This includes recognition of symptomatology by both the patient, primary and secondary medical care. Then rapid and effective diagnostic tools need to be available to confirm the diagnosis and provide susceptibility results in a timely manner.

This talk will go through these issues and discuss some of the new diagnostic techniques, including Xpert MTB/RIF test, that have become available in recent times and describe their use in both adult and paediatric population.

THE QUICK AND THE DEAD: CLINICAL IMPACT OF RAPID MICROBIOLOGICAL DIAGNOSTICS

Stephen McBride

•• 1

The management of infection is a critical part of medical practice, and in critically unwell patients delay in antimicrobial therapy active against the infecting organism is associated with poor patient outcome. Recent years have seen massive advancements in rapid microbiological diagnostics and their proliferation from the research laboratory to the clinical laboratory and hospital practice.

This case-based presentation will explore how real-world application of rapid microbiological tests can impact on patient management and outcomes, and how these technologies can be applied in different hospital settings.

© 2014 The Authors

MENINGOCOCCAL DISEASE: AN UPDATE

Mark Thomas

••

Disease due to *Neisseria meningitidis* is among the most terrifying of all infectious diseases. The presentation may be dominated by the non-specific features arising from diffuse intravascular infection or by the typical features of acute meningeal infection. Often features of septicaemia and of meningitis are present.

The outcome is worst in those with uncontrolled intravascular infection – fulminant meningococcaemia, and is strongly correlated with the concentration of bacteria in the blood, which may be as high as 10E11/L.

Measurement of serum pro-calcitonin has been recommended as a diagnostic aid, but is not widely used. The diagnosis is confirmed by visualizing Gram negative cocci in CSF or blood, by culturing the organism from blood or CSF, or by detecting meningococcal DNA in blood, CSF or tissue.

Treatment is with benzyl penicillin 12 MU / day for 3 days. Corticosteroids may offer some benefit.

Immunisation provides a high level of protection against disease due to group A and C strains, but provides only modest protection, for a short duration, against disease due to group B strains.

TBD: OBESITY AND EPIGENETICS, DIABETES AND ENDOCRINE AMD/PCHD

THE PRACTICAL MANAGEMENT OF CHILDHOOD OBESITY

Matthew Sabin

Approximately 1 in 4 children and adolescents are either overweight or obese. With this comes a heavy workload for paediatricians – not only in terms of screening for obesity and its related complications, but also instituting appropriate and effective management approaches. This talk will outline the main causes and consequences of childhood obesity, with practical advice and tips for the busy general paediatrician.

Allan Sheppard

<Text TBC>

AN UPDATE ON BARIATRIC SURGERY FOR PHYSICIANS

Grant Beban

Bariatric Surgery is increasingly used for the treatment of severe obesity and its associated illnesses.

Although studies show good long term results in most series with acceptable complication rates, there can be uncertainty around when to consider bariatric surgery and which operation may be best.

Who may (or may not be) appropriate for surgery, usual outcomes of surgery regarding weight and other conditions, and the complications physicians need to be aware of will also be presented.

The pros and cons of the main procedures that are currently available will be discussed, along with how these may be taken into account when considering surgery for a particular patient. I will also touch on some of the evolving indications for bariatric surgery, and the interaction with usual models of care.

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

Physicians as Medical Experts – P&CHD Poster Abstract

TRANS CRANIAL DOPPLER (TCD) SCANNING IN CHILDREN FOR STROKE PREVENTION

Abdulla C[1], Telfer P[2]

[1]BHR University Hospitals, London, United Kingdom
[2]Barts & London Hospitals NHS Trust London, United Kingdom

Introduction: Stroke is among the top ten causes of death in childhood. the incidence of stroke is around 2 per100,000-population (approximately 30–40 cases per year in RCH Melbourne)1 majority ends up in long term neuro-disabilities.

Stroke Prevention Studies on Sickle Cell Disease (SCD) children, has helped in primary prevention of stroke in children by early identification of cerebral arterial blood flow changes using TCD Scanning. TCD scanning with treatment is cost effective 2

In Australia, SCD, although uncommon is now increasing because of the migration of genetically susceptible population. TCD scanning is used in Australian Intensive Care Units. This presentation is to share our experience to explore the use of TCD scan to prevent stroke in high risk Australian children.

Methods: All SCD children who had TCD scan during the period 2004–2009 in Royal London Hospital were analysed retrospectively.

Results: A total of 360 children between 2 and 16 yrs. old had TCD scanning; 6% had high velocities (high risk) of which 3% showed silent/+ clinical signs of infarction. Blood flow velocity of children on treatment returned to normal standard.

Conclusion: TCD is a useful investigation tool in early recognition and prevention of stroke in high risk SCD children. Use of TCD scanning can be explored in all high risk children for stroke in Australia.

References:

1. Stroke Foundation Fact Sheet – Dr.Mark Mackay, Paediatric Neurologist, Director Children's Stroke Program, Children's Neuroscience Centre, Royal Children's Hospital Melbourne.
2. The clinical effectiveness and cost-effectiveness of primary stroke prevention in children with sickle cell disease: a systematic review and economic evaluation. MG Cherry, J Greenhalgh etc.

ANALYSIS OF LOW ACTIVE VITAMIN B12 LEVELS IN A PAEDIATRIC POPULATION IN A TERTIARY CENTRE

Alhucema P[1], Greenway A[1], Campbell J[1]

[1]The Royal Children's Hospital (RCH), Melbourne, Victoria, Australia

Background: Vitamin B12 (vit-B12) or cobalamin is essential for normal blood and neurological development. Early recognition and treatment is important and therefore a sensitive marker to assess vit-B12 status is essential. A new method to detect levels involves active vitamin B12 (Holotranscobalamin) instead of total serum vit-B12. This is suggested to be a more sensitive marker and may provide an earlier marker of deficiency (1) (2)

Objective: To review the clinical presentation and risk factors for a population of children with low active vitamin B12 (active-B12). Are we detecting patients with subclinical or early deficiency by utilizing a more sensitive assay?

Methods: This is a descriptive retrospective analysis of children with measured low active-B12 over a 3 year period at RCH, Melbourne.

Results: The total low active-B12 results were 109. Children at higher risk for developing vit-B12 deficiency were: ≤ 1 year of old and patients with malabsorption >1 year old. Our study suggests active-B12 is a more sensitive assay, with 49.5% of patients being asymptomatic; 23.3% with macrocystosis on film and only a quarter with evidence of tissue deficiency, however this cannot be accurately concluded until we have established a paediatric reference range for serum homocysteine and all children with low active-B12 are investigated for signs of tissue deficiency with total homocysteine and methylmalonic acid.

References:

1. Nexo E, Hoffmann-Lucke E. Holotranscobalamin, a marker of vitamin B-12 status: analytical aspects and clinical utility. *The American journal of clinical nutrition*. 2011;**94**(1):359S–65S.
2. Herrmann W, Obeid R, Schorr H, Geisel J. Functional vitamin B12 deficiency and determination of holotranscobalamin in populations at risk. *Clinical chemistry and laboratory medicine: CCLM / FESCC*. 2003;**41**(11): 1478–88.

DO TEXT MESSAGE REMINDERS IMPROVE ATTENDANCE AT A PAEDIATRIC CLINIC?

Vicki Burneikis

Central Coast Local Health District, New South Wales, Australia

Aim: To ascertain whether sending text messages to parents' mobile phones the week before routine paediatric outpatient appointments can reduce rates of non-attendance.

Method: For 23 weeks, from the start of school term 3 to the end of term 4 of 2013, text message reminders were sent to parents or carers whose children had appointments in 12 randomly selected weeks, but not the remaining 11 weeks. Those not sent text messages received no reminders, but all appointments were written on cards at the time of booking (typically 3–6 months prior). The time taken to send the messages was recorded.

Results: 93 parents received text messages, and of these 84 children (90%) attended appointments. 82 parents received no confirmation, and of these 58 (71%) attended appointments. Text messages took an average of 45 seconds each to send.

Analysis and Conclusion: Text messages improved attendance from 71 to 90% (chi squared = 10.9, $p < 0.01$). The Number Needed to Text to get one extra attendance (NNT) was 5.1, meaning just under 4 minutes of work led to one more attendance. Average revenue generated per patient was estimated from the previous years' income as $110, making text messages an extremely cost effective intervention.

THE IMPACT OF SOCIAL DISADVANTAGE AMONG CAREGIVERS ON THE QUALITY OF LIFE OF CHILDREN WITH CHRONIC DISEASES – A SYSTEMATIC REVIEW

Didsbury M[1], Kim S[1,2], Medway M[1], Tong A[1,2], Craig J[1,2], Wong G[1,2]

[1]Sydney School of Public Health, The University of Sydney, Sydney, New South Wales, Australia
[2]Centre for Kidney Research, The Children's Hospital at Westmead, Westmead, New South Wales, Australia

Introduction: Quality of life (QoL) is a critical outcome in the management of chronic diseases in children. The aims of this study were to summarise the association between social disadvantage of the caregivers and the quality of life of children with chronic disease; and to determine the specific social and economic factors that impact on the QoL of these children.

Methods: We systematically reviewed all studies that had measured and reported on the association between at least one measure of social disadvantage in caregivers and at least one QoL measure in children with asthma, chronic kidney disease (CKD), type 1 diabetes and epilepsy. MEDLINE, Embase and PsycINFO were searched to January 2014. These conditions were included as they represent the four most prevalent and debilitating non-communicable childhood diseases.

Results: From 7748 records, 24 eligible cross-sectional studies were identified and included (n = 5,969; 5 asthma [n = 462], 3 CKD [n = 593], 10 Epilepsy [n = 1450], 6 T1D [n = 3464]).

Overall, 19 (79%) studies reported a significant association between at least a single SES determinant and QoL across all disease groups. Significant factors included the level of parents' education, family income, parents' occupation, level of health insurance and the parents' marital status. Children with type 1 DM whose mother had completed higher school certificate experienced better overall QoL compared to those who completed primary education (standard mean difference (SMD) 0.52, 95% CI: 0.01 to 1.03, $I^2 = 85\%$, $P = 0.05$). Household income did not have a significant impact on QoL (SMD: 0.50, 95% CI: –0.37 to 1.37, $I^2 = 85\%$, $P = 0.26$).

Conclusion: Among children with chronic diseases, those who are from economically disadvantaged backgrounds reported poorer quality of life than

© 2014 The Authors

Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)
Journal of Paediatrics and Child Health **50** Suppl. 2 (2014) 9–15

their wealthier counterparts. Strategies to improve the socioeconomic gradient of the caregivers may prevent or reverse the unfavourable health outcomes in children with chronic illness.

HYDROCEPHALUS DUE TO BLAKE POUCH CYST, AND ECTOPIC ANUS

Whitehall JS[1,2], Karunaratne N[1], Ambler R[3], Chaseling R[4], Edwards MJ[1,2]

[1]Department of Paediatrics, School of Medicine, University of Western Sydney
[2]Department of Paediatrics, Campbelltown Hospital, NSW Australia
[3]Mount Druitt Hospital, Sydney, NSW, Australia
[4]Neurosurgery, Children's Hospital, Westmead NSW Australia

Introduction: Atresia of the foramen of Magendie causes Blake pouch cyst (BPC), a posterior dilatation of the fourth ventricle, sometimes leading to non-communicating hydrocephalus involving all ventricles. A 5-year-old girl presented for genetic assessment of hydrocephalus with BPC, diagnosed by MRI for ataxia, speech problems and macrocephaly, and treated by ventriculoperitoneal shunt. The pregnancy was conceived by donor-egg IVF, previous embryos having died early in culture. A child of the egg donor had an unspecified bowel problem. Vaginal bleeding occurred in the first trimester, weight gain was 7 kg, fetal ultrasound at 26 weeks showed renal pelvis dilatation but was normal at 36 weeks. Birth weight at 36/40 was 3.41 kg (90th percentile), an ectopic anus was repaired at 4 months. The patient had recurrent febrile and possible afebrile seizures and urinary tract infections but postnatal renal ultrasound was normal. Spinal MRI of the spine was normal.

Methods and Results: Weight and height were above the 99th percentile, head circumference 55.2 cm was further above the 99th percentile. There were no features of VACTERL syndrome apart from the ectopic anus. CGH microarray and chromosome breakage studies for Fanconi syndrome were normal

Conclusion: The differential diagnosis included VACTERL with hydrocephalus, Townes-Brock, Currarino, Pallister-Hall syndromes and Fanconi anaemia. This patient has no known syndrome. BPC has been reported with biliary atresia in one boy, but not with ectopic anus. There has been little mention of BPC in the genetic literature. It is thought to be a sporadic abnormality with no significant recurrence risk for siblings.

RUSSELL-SILVER SYNDROME AND HYPERCALCAEMIA WITH MATERNAL UNIPARENTAL DISOMY OF CHROMOSOME 7

Whitehall JS[1,2], Vosu J[1], Allgood C[1,2], Mendoza-Cruz C[2], Dunstan R[1,2], Burgess T[3], Edwards MJ[1,2]

[1]Department of Paediatrics, School of Medicine, University of Western Sydney
[2]Department of Paediatrics, Campbelltown Hospital, NSW Australia
[3]Molecular Cytogenetics, Victorian Clinical Genetics Services, Murdoch Children's Research Institute

Introduction: A baby girl born weighing 2155 gm at 37 weeks gestation was referred for genetic assessment because an elevated risk for trisomy 18 had been found on first trimester screening, she had poor postnatal growth and had hypertension and hypercalcaemia treated with a low calcium formula. There were no other hazards during the pregnancy and no relevant family history. At age 3 months the weight 3.48 kg was far below the 3rd percentile, length 54.2 cm was between the 1st and 3rd percentile and head circumference 38.5 cm was between the 3rd and 25th. One ear was outstanding from the scalp, and there was bilateral 5th finger clinodactyly otherwise there were no morphological features of a syndrome. A search of the literature identified a child with uniparental disomy of chromosome 7, Russell-Silver syndrome and hypercalcaemia[5].

Methods and Results: CGH microarray (Agilent) was normal. Parathormone levels were normal or elevated at the time of hypercalcaemia. Illumina HumanCoreExome-12 v1.1 single nucleotide polymorphism microarray was performed on DNA from the patient, showing she had a long continuous stretch of homozygosity of chromosome 7q21.11q36.1 (nucleotides 80,474,046–151,846,195). SNPTrio analysis of the patient and her parents showed that both copies of chromosome 7 were maternally inherited. The patient's mother had normal calcium levels on several occasions during pregnancy.

Conclusion: This is the second child with Russell-Silver syndrome and hypercalcaemia reported to be associated with uniparental disomy of chromosome 7, although the previous patient had suppressed PTH levels, in contrast to the parathormone levels in our patient.

[5]**Reference:** Stark Z et al (2010). Atypical Silver–Russell Phenotype Resulting From Maternal Uniparental Disomy of Chromosome 7. *Am J Med Genet* **152A**:2342–2345.

WHY ABORIGINAL HEALTH NEEDS A NEW DIRECTION

Bret Hart

Unity of First People of Australia, Western Australia, Australia

A recent return to the Kimberley after an absence of 3 decades gave the impression that Aboriginal health has deteriorated despite more resources being allocated over the same period.

This prompted a review and confirmed that there is no shortage of evidence and reports regarding the poor state of ATSI people compared to other Australians over many years. This is realised in ATSI communities with one describing a "Dark Cloud" overshadowing future prospects for their health and the health of their children. This concept is consistent with Jilek's syndrome of 'anomic depression' induced by the loss of a community's socio-cultural identity. This was before the concept of 'biological embedding' was conceived and predated an appreciation of the epigenetic influence of past trauma. "The stolen generation" exacerbated their adverse past history. Also the social determinants of health are compromised in many ATSI people. All these factors help explain why improving ATSI health has been a challenge and will be in future due to the foetal and childhood origins of adult disease.

With most of the multiple determinants of ATSI health lying outside the health care system, any improvement will require a new coordinated multisectoral approach. This whole of government response is the basis of 'Health in All Policies (HiAP)' – a growing movement in Europe and the USA. As the Prime Minister has taken responsibility for Indigenous Affairs, he is uniquely placed to instruct his Ministers and their departments to ensure their policies and programs are ATSI health enhancing.

By selecting the early years as a priority and adopting an ATSI HiAP approach, there is the potential to prevent further transgenerational transmission of trauma and disadvantage and to finally achieve the elusive goal of improving Aboriginal health.

CLINICAL DIAGNOSIS OF CONGENITAL ADRENAL HYPERPLASIA IS UNRELIABLE IN BOTH GENDERS – NEW ZEALAND NEWBORN SCREENING DATA FROM 1994–2012

Heather N[1], Seneviratne S[2], Webster D[4], Jiang Y[3], Derraik J[2], Carll J[4], Jefferies C[1,2], Cutfield W[1,2],Hofman P[1,2,]

[1]Starship Children's Hospital, Auckland, New Zealand
[2]Liggins Institute, University of Auckland, New Zealand
[3]Department of Statistics, University of Auckland, New Zealand
[4]Newborn Metabolic Screening Programme, LabPlus, Auckland City Hospital, Auckland, New Zealand

Background: New Zealand was one of the first countries to include congenital adrenal hyperplasia (CAH) in its newborn screening programme. Severe CAH is a rapidly evolving, life-threatening disorder that becomes apparent within the first month of life.

Aims: To evaluate the efficacy of the New Zealand newborn screening programme for CAH.

Methods: Infants diagnosed with CAH in the newborn period from 1994–2012 were identified from National Testing Centre records. Case characteristics were reviewed and compared for those diagnosed clinically or by screening.

Findings: Over this period, 41 neonates (25 female, 16 male) were diagnosed with CAH, giving an incidence of 1: 26,316. Almost 50% (n = 20) of newborns with CAH were identified only by newborn screening, including 20% of females. All clinically detected cases (n = 21) had abnormal virilisation and 25% had an affected sibling.

Overall, whole blood 17-hydroxyprogesterone sampling occurred at a median of 2 (range 0–8) days, screen result notification at 7 (0–19) days, and treatment commenced at 6 (1–30) days.

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

Compared to those diagnosed by screening alone, those diagnosed clinically were predominantly female (95% vs 25%; $p < 0.0001$), were diagnosed earlier (5.2 vs 8.8 days; $p < 0.001$), had higher serum Na (136 vs 131 mmol/L; $p < 0.001$) and lower serum K concentrations (5.4 vs 6.0 mmol/L; $p < 0.05$).

Vomiting and failure to thrive were present in 15% of screening detected cases at diagnosis, but none had hypotension or collapse. Serum electrolyte concentrations showed a linear correlation with age at diagnosis: Na: $r^2 = -0.64$, ($p < 0.0001$) and K: $r^2 = 0.38$ ($p = 0.0055$).

Conclusions: Newborn screening alone accounted for nearly 50% of CAH detected in the newborn period, including a fifth of all girls, indicating that clinical diagnosis is unreliable in both genders. No babies presented with adrenal crisis. This study adds further argument for CAH screening internationally.

LIFE THREATENING LARYNGOSPASM AS A PRESENTATION OF CHILDHOOD EPILEPSY

Humblet M[1,2], Nolan M[3]

[1]Univeristy Hospitals Leuven, Belgium
[2]Paediatric Intensive Care Unit, Starship Hospital, Auckland, New Zealand
[3]Paediatric Neurology Department, Starship Hospital, Auckland, New Zealand

Background and Aims: We present a case of life threatening recurrent laryngospasm, as an isolated manifestation of epilepsy.

A 22 month old girl presented with rapidly progressive choking episodes, initially only nocturnal but progressing to frequent unprovoked episodes day and night. They were associated with severe oxygen desaturation and dystonia whist being fully aware, requiring paediatric intensive care. Two thirds of the episodes occurred from sleep. Investigations (MRI brain and neck, CSF and EEG x 2, which captured events) could initially not confirm a diagnosis of epilepsy. However, anti-epileptic treatment was continued, whilst further non-neurologic investigations were ongoing, including laryngoscopy and bronchoscopy. An extensive repeat video EEG demonstrated the episodes to be cortical seizures, focality over the right posterior temporal region. To achieve seizure control, high doses of Carbamazepine (50 mg/kg/day) and levetiracetam (90 mg/kg/day) were necessary.

Methods and Results: A literature search was performed and this degree of epileptic laryngospasm in children has not previously been described. Review of choking episodes during sleep debate the diagnostic challenge that stems from the frequent absence of recorded epileptiform activity, often causing a delay in adequate seizure treatment. Two recent reports on severe neonatal episodic laryngospasm describe a phenotype of seizure disorder related to sodium channelopathy. In these patients, high dosages Carbamazepine were necessary to reach seizure control.

Conclusions: Despite being a rare phenomenon, severe laryngospasm episodes in children should be suspicious as a presentation of epilepsy, even without correlation with EEG. Treatment should not be postponed while excluding other aetiology. Carbamazepine may be effective in monotherapy or combined with levetiracetam and high doses may be necessary to achieve seizure control.

BRIEF AUDITS WHILE PRESCRIBING "BICILLIN" FOR ARF SECONDARY PREVENTION; A PRACTICAL QUALITY IMPROVEMENT TOOL FOR ARF/ RHD AREAS WITHOUT REGISTERS

Malcolm John

Bay of Plenty District Health Board, Whakatane, New Zealand

Background: Short audits are quick and yield useful insights to inform practice. Paediatricians are presented with 15–30 three monthly benzathine penicillin IM prescriptions in District Nursing Service (DNS) folders to sign at the end of a working day. Prescribing becomes an audit window giving insight into primary, secondary and tertiary prevention of Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD).

Objectives: To audit the ARF cohort from three rural towns, for improvable elements, either causal pathway, co-morbidities, confounding factors for patient diagnosis. To audit analgesia, and factors in "Bicillin" adherence for secondary prevention, and clinical follow-up as contributors to timely management, surgery when indicated, moderating morbidity and mortality.

Methods: Benchmarks are Cardiac Society ANZ endorsed National Heart Foundation (NHF), ARF guidelines/ algorithms on treatment of Group A Streptococcus pharyngitis, diagnosis/management ARF, clinical follow-up intervals by cardiac severity. Follow-up appointments offered, and attended, were compared to standard and clinician recommendation. Reviewing clinician contacted patient's whanau (extended family) clerical staff, medical and nursing clinicians, as indicated.

Conclusions: Short audits with ARF RHD focus conducted over 5 years confirmed ARF distribution by age, ethnicity, geography, and deprivation decile. ARF was associated at times with non presentation for sore throats, short treatment prescribed and adherence issues. Some RHD patients presented in cardiac failure with preceding sub-clinical ARF. At times diagnosis was delayed by non-admission, confusing co-morbidity, non-steroidal use, undue orthopaedic focus, and limited echocardiograph access. Clinician turnover, unfamiliarity, non-adherence to NHF guidelines impacted on diagnosis and management. Clinical Follow-up duration was limited, sometimes with long intervals, (some not on IM Penicillin). Complex concurrent family challenges were ameliorated by whanau family support. Rapid audits while prescribing "Bicillin", address aspects of ARF/ RHD quality improvement.

ACQUISITION OF KNOWLEDGE AND SKILLS AFTER "ANAPHYLAXIS EDUCATION" IN A REGIONAL CENTRE TO IMPROVE PATIENT CARE

Kaur N[1], McCrossin T[2]

Introduction: Allergies are the fastest growing chronic diseases in Australia[1]. **Food-induced anaphylaxis** has doubled in the last 10 years and **10% of Australian infants have proven food allergy**[2]. Deaths due to anaphylaxis, especially food related, occurring in community highlight the issue of increased awareness and anaphylaxis education of patients by health professionals.

Aim: We hypothesized that Anaphylaxis education of staff using Adrenaline auto-injectors would improve patient care and surveys were used to evaluate knowledge retention.

Methods: Quality improvement methodology was used. Multi-voting by a team of medical and nursing staff from paediatrics and emergency identified the categories for inadequate education which were: lack of confidence and knowledge of access to resources, Lack of education and inadequate equipment. Education sessions for staff **highlighting the different techniques to use Adrenaline auto-injectors** and raising awareness of resources such as ASCIA website were undertaken over a period of 8 weeks. Anaphylaxis education Kits including Adrenaline auto-injector trainers were developed. The effectiveness of sessions was measured by a pre and post education survey.

Results: A total of 55 staff participated in the study, which included (medical-38%,nursing-42% , medical and nursing students -18%).The confidence in writing an anaphylaxis action plan increased from pre-education 34% to post education-89%. The confidence in demonstration of adrenaline auto-injector use increased from 51% to100%. The post education experience showed improved patient education and demonstration of auto-injector use.

Conclusion: Anaphylaxis education led to significantly improved knowledge and skills of staff which improved patient care. Follow up at 12 months will study knowledge retention and use of anaphylaxis education kits.

References:

Australasian Society of Clinical Immunology and Allergy[1] (ASCIA), Allergy and Immune Diseases in Australia report 2013[2]

GENERAL SYSTEMS THEORY (GST). A GENERAL BIOLOGICAL THEORY BASED ON TWO ASSUMPTIONS OF BIOCHEMICAL RESPONSE PROVIDES A BIOCHEMICAL MODEL OF UNDERSTANDING EPI-GENETICS, DISEASE, BEHAVIOUR, AGING, DEATH AND EVOLUTION

John B Myers

Wellspring's Universal Environment P/L, Balaclava, Victoria, Australia

Aim: To promulgate a GST that provides a unifying theory of understanding of biological response including behaviour, disease, aging, epi-genetics and evolution.

Introduction: Claude Bernard is considered the father of physiology. His theory/concept of the constancy of the milieu interieur helps to explain the

© 2014 The Authors

migration of sea animals to land, as fluid bathing cells within the organism is composed of water and sodium primarily as found in sea.

Methods and Results: Experimental findings in humans showed that plasma sodium concentration remains constant to within or <1% on a two week diet of added sodium, whereas cell sodium increased by 30%.

Discussion: On the basis of these results Myers (Medical Hypotheses.1982;9:241–57) proposed a general biological theory (General Systems Theory (GST)) termed "the constancy of the milieu exterieur", which is the same as Claude Bernard's except viewed from the cellular perspective rather than the multi-cellular organism's, based on two assumptions of biological response. A. Innermost membranes maintain the fluids external to that membrane within an optimal range. This occurs at all outer levels of biological organisation. B. A persistent disturbance of the external fluid at any biological level of organisation will result in change in the inner fluids. According to Hochachka and Somero, physiological response may be rapid (escape) or occurs by adaptation. Thus adaptation may be adequate and appropriate leading to evolution if breeding and survival is enhanced, and epi-genetics in utero; to disease if adaptation is either inadequate or inappropriate; to aging if the change in environment is not sensed i.e. not responded to; and to death if the environmental change is sudden or overwhelming. Likewise, behavioural response is an attempt to control the external environment. Ethical response reflects appropriate and/or adequate inner change and adequate or appropriate response to environmental changes/challenges.

Conclusion: GST provides a general understanding of biological responses.

THE PSYCHIATRIC COMORBIDITIES OF AUSTRALIAN CHILDREN WITH MOVEMENT DISORDERS

Michelle Nicholson[1], Isobel Heyman[2], Benjamin Baig[2], David Dossetor[1], Clara Marecos[3], Manju Kurian[3], Mary-Clare Waugh[1], Ruth Evans[1], Joshua Burns[1], Russell Dale[1]

[1]The Children's Hospital at Westmead, New South Wales, Australia
[2]South London and Maudsley NHS Foundation Trust, UK
[3]Great Ormond Street Hospital for Children NHS Foundation Trust, London UK

Background: Although it has been well described that children Tourette Syndrome have higher rates of psychiatric comorbidities than community controls, there is little known about the mental health problems that affect children with non-Tourette movement disorders.

Objective: The aim of this study is to describe the prevalence of psychiatric co-morbidities in children (5–16 years) with movement disorders from an Australian clinic population and to compare this rate with normative data using the same psychiatric assessment tool.

Methods: Children were recruited through secondary and tertiary clinical settings when they attended for review with a Paediatric Neurologist or Rehabilitation Specialist.

Participation in the study required completing a detailed psychiatric assessment questionnaire called the DAWBA (Development and Wellbeing Assessment) tool.(1)

The DAWBA is a screening questionnaire that collates information relating to emotional and behavioural symptoms and their impact and derives DSM-IV psychiatric diagnoses.

Approximately half (51%) of children in the study had Tourette disorder. The remaining 49% had other movement disorders including dystonia (n = 38), chorea (n = 13), tremor (n = 3), stereotypy (n = 3), myoclonus (n = 2), mirror movements (n = 1), and psychogenic (n = 1).

Results: The rate of DSM-IV psychiatric disorders in the Tourette group was 26%. In the non-Tourette group the rate was a similar 29%. These figures represent an almost three fold increased prevalence compared to population rates of 9.5% found in a UK community cohort of 7912 children.(1).

Conclusion: This study suggests that children with movement disorders other than Tourette Syndrome are almost three times more likely to meet threshold for a psychiatric diagnosis than children in the general population. These preliminary findings contribute to an important aspect of movement disorder phenomenology.

Reference:

1. Goodman A, Heiervang, E., Collishaw, S., Goodman, R., The 'DAWBA bands' as an ordered-categorical measure of child mental health: description and validation in British and Norwegian samples. *Soc Psychiatry Psychiatr Epidemiol.* 2011;**46**(6):521–32.

ABORIGINAL CHILD HEALTH INDICATORS IN A COHORT OF URBAN ABORIGINAL INFANTS RECEIVING A SUSTAINED NURSE HOME VISITING INTERVENTION IN LIVERPOOL, BANKSTOWN, FAIRFIELD LGAS AND THE INNER WEST

Noble J[1], Yoong T[1], Pollack J[1], Eastwood J[1]

[1]Department of Community Paediatrics, Sydney & South Western Sydney Local Health Districts (LHD), NSW

Introduction: Our aim is to describe health and developmental outcomes in the first year of life in urban Aboriginal infants in a sustained nurse home visiting (SNHV) program in disadvantaged local government areas (LGAs) in Sydney.

Methods: The *New Directions* program is a model of care consisting of health promotion, early access to antenatal care, SNHV and well-child care coordination, aimed at better outcomes for Aboriginal children. It is based in Central and South Western LHDs of Sydney. All children receive health and developmental assessments at 12 months of age. Data collected was used in this analysis study.

Results: Maternal smoking (51%) was comparable to the prevalence in Indigenous females and pregnant Indigenous mothers, but was higher than the general female population ($p < 0.001$). There was significantly lower prevalence of breastfeeding at discharge (59.1%) and at 6–8 week (35.3%) compared to Indigenous mother ($p < 0.001$) and the general female populations ($p < 0.001$). Immunisations were up-to-date in only 81% at 12 months; lower compared to the general population ($p < 0.05$).

Our infants were lighter and had smaller head circumferences at birth, and gradually became heavier and had larger head circumference than the Center for Disease Control and Prevention (CDC) comparison by 12 months old.

Our most significant findings were our infants' development at 12 months of age, which was mostly comparable to all Griffiths Mental Development Scales (GMDS) subquotient means, and was higher than infants in the Gudaga study in communication ($p < 0.001$), fine motor ($p < 0.05$) and overall quotient ($p < 0.05$)[1].

Conclusion: This was a pilot evaluation which raised many issues. Further research is required to establish if our positive findings were associated with the SNHV intervention. It also highlights the areas where further improvements could be made to optimise child health indicators to close the gap for the Aboriginal children in our areas of service.

Reference:

1. McDonald J, Comino E, Knight J et al. Developmental progress in urban Aboriginal infants: A cohort study. *J. Paediatr. Child Health.* 2012; **48**: 114–21.

INVESTIGATING FRAGILE X SYNDROME IN PAEDIATRICS – CAN WE RATIONALISE OUR CURRENT PRACTICE?

Noovao RJ, Graham D

Waikato Hospital, Hamilton, New Zealand

Background/Aims: Fragile X Syndrome (FXS) is the most common monogenetic cause of Intellectual Disability and Autism Spectrum Disorder currently known. Genetic testing is however expensive and produces a low yield of positive results. This study sought to identify potential methods that could reduce testing costs without jeopardising prompt and accurate diagnosis.

Methods: A three-tiered approach involved a local 200-child clinical audit, an Australasia-wide questionnaire responded to by 224 paediatricians and a systematic review of the literature to identify a FXS screening checklist to assist in reducing the number of unnecessary FXS tests requested.

Results: The clinical audit showed that the majority (86%) of Waikato paediatricians used appropriate indications when requesting FXS tests. However, there was potential to reduce testing costs by 24% by using 'best practice' guidelines, a stepwise approach to investigations or a second review before requesting investigations in cases of diagnostic doubt.

The clinician questionnaire revealed over-investigation of children with Attention Deficit Hyperactivity Disorder or Specific Learning Disorders alone by some clinicians, while others under-investigate girls and those with mild Intellectual Disability. The questionnaire also demonstrated that 60% of clinicians would consider using a FXS screening checklist, provided certain conditions were met.

Lastly, a systematic review of the literature suggested the possibility of reducing FXS testing costs by up to 60% in boys. However, due to borderline

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

validity and generalisability, such a checklist would need a rigorous local trial to ensure that diagnostic accuracy is upheld before its use could be recommended. No valid checklists can currently be recommended for girls.

Conclusion: Adherance of all practitioners to 'best practice' guidelines, a stepwise approach to investigations, re-review of patients in whom there is diagnostic doubt and a locally validated FXS screening checklist in boys all have potential to improve diagnostic efficiency and cost utility in the investigation of FXS.

A PUZZLE. WAS RENAL VEIN THROMBOSIS AND HEART FAILURE ASSOCIATED WITH INTRAHEPATIC CHOLESTASIS OF PREGNANCY (ICP)?

Trainee name: Julia Parmeter

Supervisor names: Dr Catherine Allgood, Dr Raymond Chin, Professor John Whitehall

Hospital: Campbelltown Hospital, NSW.

Description of Case

We present the case of a neonate born with normal Apgars to a mother with ICP who developed

biventricular failure, acute renal vein thrombosis and hyperbilirubinaemia requiring exchange transfusion within the first few days of life.

The neonate was delivered by Caesarian section at 36 weeks gestation from a mother ICP because of decreased foetal movements and non-reassuring cardiotocography. The mother had pruritis from

25 weeks gestation and was treated with ursodeoxycholic acid from 30 weeks gestation. Maximal bile acids were 40 μmol/L at 32 weeks gestation and were associated with mildly elevated liver enzymes. The birth weight was 3401 g and Apgars were 9 and 9 at 1 and 5 minutes respectively. Respiratory distress developed at two hours of age with haemoglobin 198 g/L, white cell count 20.5 x10∧9/L, platelets 94 x10∧9/L and metabolic acidosis (pH 7.27 pCO2 42, HCO3 18.9, BE -7.7). Broad- spectrum antibiotics were commenced.

At 25 hours of age the baby developed haematuria and hepatomegaly was noted, but there were no cardiac murmurs and peripheral pulses were normal with a blood pressure 80/67 mmHg (75% centile). The chest xray was unremarkable. Ultrasonography revealed left renal vein thrombosis (RVT). At 32 hours of age echocardiography revealed biventricular failure with an ejection fraction of

15%. He was intubated, and transferred for tertiary care, including clexane and inotropy. By Day 5, cardiac function was normal, but hyperbilirubinaemia developed (peak bilirubin 461 μmol/L) and exchange transfusion was required. Subsequent progress was uneventful. Thrombophilia screen done subsequently was normal.

Issues of Interest in the Case

1. What caused the renal vein thrombosis? None of the classical causes are present including perinatal asphyxia and the thrombophilia screening is normal. Is renal vein thrombosis an unreported complication of ICP, or did an undetected cardiomyopathy cause renal vein thrombosis?
2. What caused respiratory distress? Did excess bile acids, as has been reported in the literature?
3. What caused the cardiomyopathy? ICP is associated with sudden foetal death, arrhythmias and mitochondrial suppression but why was presentation delayed? Was there failure from in- utero bile acids despite normal Apgars and cardiac signs? Did renal vein thrombosis lead to cardiomyopathy?
4. Did hepatocyte damage impairing conjugation from the raised bile acids along with absorption of thrombus cause hyperbilirubinaemia?
5. We propose ICP caused cardiomyopathy with secondary respiratory distress, acidosis and RVT and advise early echocardiography.

MUTATION RESPONSIBLE FOR PRIMARY CILIARY DYSKINESIA (PCD) CAUSING IDIOPATHIC TACHYPNOEA IN NEWBORN IDENTIFIED BY SINGLE NUCLEOTIDE POLYMORPHISM (SNP) ARRAY AND SEQUENCING OF LRRC6 GENE

Whitehall JS[1,2], Allgood C[1,2], Legendre M[3], Burgess T[4], Edwards MJ[1,2], Chin R[1,2], Dunstan R[1,2], Freelander M[1,2], Amselem S[3], Morgan L[5], Pervez T[1]

[1]Department of Paediatrics, School of Medicine, University of Western Sydney
[2]Department of Paediatrics, Campbelltown Hospital
[3]U.F. de Génétique, moléculaire, Hôpital Armand Trousseau, Paris, France
[4]Molecular Cytogenetics, Victorian Clinical Genetics Services, Murdoch Children's Research, Institute
[5]Department of Thoracic Medicine, Concord Hospital

Background: PCD is an autosomal recessive disease with disrupted ciliary structure and/or function in several organs, including chronic sinopulmonary disease. 50% of patients have situs inversus/dextrocardia because of random determination of situs. A full term boy weighing 3725 gm after an uncomplicated pregnancy had Apgar scores of 9/1 and 9/5. His parents were first cousins of Lebanese ancestry. His P6G9 mother had a cousin with dextrocardia. He had mucopurulent rhinorrhoea and tachypnoea treated with oxygen from age 31 hours for 7 days. Chest X ray showed dextrocardia and situs inversus.

Methods: Nasal cilia were obtained using a cytology brush. Ciliary motility was assessed via light microscopy and photometry; ciliary ultrastructure was assessed using electron microscopy. Illumina HumanCoreExome – 12 v1.1 was used for the SNP microarray. Sanger sequencing of *LRRC6* coding exons and flanking intronic sequences.

Results: All cilia were completely immotile with a deficiency of inner and outer dynein arms. A younger brother with chronic lung disease of prematurity had normal ciliary structure and function: SNP microarray identified long contiguous stretches of homozygosity (LCSH) in him, a 12-year-old unaffected brother, and the proband with PCD. Autozygosity mapping identified unique regions of homozygosity in the proband. One of these included the LRRC6 gene (chromosome 8q23.3-q24.3), whose mutations were found in 2012 to cause ciliary dyskinesia[1]. Sequencing identified a homozygous or hemizygous c.230dupA (p.Asn77Lysfs*3) mutation in exon 3. Both parents were heterozygous for the mutation.

Conclusion: SNP array is a very effective method for identifying areas of homozygosity that simplify the search for mutations in consanguineous families with autosomal recessive diseases.

Reference:

1. Kott E, Duquesnoy P et al. (2012). Loss-of-function mutations in LRRC6, a gene essential for proper axonemal assembly of inner and outer dynein arms, cause primary ciliary dyskinesia. *Am J Hum Genet.* **91**(5):958–64

A WARNING ABOUT TB

Sgarlata J[1], Edwards M[1,2], Allgood C[1], Bent G[1], Whitehall J[1,2]

[1]Campbelltown Hospital, New South Wales, Australia
[2]University of Western Sydney, New South Wales, Australia

Introduction: An alert, well grown 5 month old infant presented to Campbelltown Hospital, Sydney SW, with minimal cough for two weeks. Although not 'sick', she had severe tuberculosis, providing warnings: it can occur with minimal contact, in an ethnic community lesser known for tuberculosis (TB), and, without any major clinical signs, be widespread to include meningitis and be complicated by sudden demise.

History: She was born in Campbelltown to Tongan parents but a maternal cousin from Victoria visited for three individual weeks, as recently as 2 months ago. The cousin had sputum positive TB.

On admission, she was afebrile, RR 66, HR 140, saturations 96% in air with mild recession. She had decreased air entry on the right, and hepatosplenomegaly. CXR revealed right-sided consolidation, bilateral nodules and a widened pericardium. Hb was 108 g/L, WCC 15.8x 10^9/L, CRP 36 mg/L. Community acquired pneumonia was diagnosed and treated with penicillin and subsequently cefotaxime and lincomycin, without success. Fever rose to 39.3°C and work of breathing increased.

On day 2, ultrasound confirmed hepatomegaly with multiple hypo-echoic foci (to 1 cm) and uncomplicated splenomegaly.

© 2014 The Authors

Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)
Journal of Paediatrics and Child Health **50** Suppl. 2 (2014) 9–15

Suspecting TB or malignancy, she was transferred to a tertiary centre where gastric aspirate AFB and positive PCR confirmed TB and four drug therapy was commenced. QuantiFERON-TB Gold In-Tube was later positive.

LP revealed one polymorph, three monocytes and four indeterminate cells. Glucose was 3.1 and protein was 0.5. No AFB. PCR negative. Two days later she collapsed with reduced consciousness. CT showed multiple tuberculoma with mild hydrocephalus and meningeal enhancement. She improved with steroids and was discharged in 2 weeks without discernible neurological signs.

Discussion: Paucity of clinical signs was indirectly proportional to gravity. The case demonstrates the higher rate of TB in children of migrant parents, though Tonga is not highly burdened.

MOVING ON FROM 'NOT ENOUGH TIME': FACTORS INFLUENCING RESEARCH PARTICIPATION IN CLINICIANS WORKING AT A TERTIARY CHILDREN'S HOSPITAL

Paget SP[1,2], Caldwell PHY[2,3], Murphy J[4], Lilischkis KJ[5], Morrow AM[1,2]

[1]Kids Rehab, The Children's Hospital at Westmead, New South Wales, Australia
[2]Sydney Medical School, University of Sydney, New South Wales, Australia
[3]Centre for Kidney Research, The Children's Hospital at Westmead, New South Wales, Australia
[4]Service Improvement Unit, The Children's Hospital at Westmead, New South Wales, Australia
[5]Kids Research Institute, The Children's Hospital at Westmead, New South Wales, Australia

Background: Whilst clinicians are often cited as being in the best position to identify research priorities, their research participation is poor, particularly in paediatrics. An online questionnaire was sent to clinicians working at our hospital to explore attitudes to research participation[1]. The low response rate (17%) prompted us to conduct focus groups to better understand the factors involved.

Aims/Objectives: To improve understanding of the factors influencing and inform strategies to improve research participation in clinicians at a tertiary children's teaching hospital.

Methods: Six focus groups (two each for doctors, nurses and allied health professionals (AHPs)) were conducted between December 2010 and April 2011. Audio recordings of the focus groups were transcribed and the data analysed using Excel. Thematic analysis was conducted through an inductive process based in grounded theory, and passages of text were coded according to theme using the constant comparative method. Additional analysis examined differences in responses between professional groups.

Findings: Forty clinicians participated in the focus groups (17 doctors, 12 nurses and 11 AHPs). Research participation was identified as important but a core part of workload. Factors influencing research participation were grouped into three major themes (Personal, Cultural and Logistic). Professional groups differed in the content of subthemes. Solutions suggested for increasing research participation were grouped into seven subthemes (culture change, relevance, protected time, communication and collaboration, accessible funding, information and education, and research mentors / champions).

Conclusions: The results of this study provide a useful insight into the factors influencing clinicians' research participation. Differences between professional groups suggest that a one-size-fits-all approach will not be sufficient to improve research participation across all groups.

Reference:

1. Paget SP, Lilischkis KJ, Morrow AM, Caldwell PHY. Embedding research in clinical practice: differences in attitudes to research participation among clinicians in a tertiary teaching hospital. *Intern Med J.* 2014; **44**:86–89.

THE FAMILY HOME VISITING PROGRAM IN SOUTH AUSTRALIA: EXPERIENCES OF ABORIGINAL FAMILIES

Spurrier NJ[1,2], Sawyer A[2], Chong A[2], Axleby-Blake T[3], Ah Kit J[3], King R[4], Sawyer MG[2]

[1]Department for Health and Ageing, South Australia, Australia
[2]University of Adelaide, South Australia, Australia
[3]Child and Family Health Service, South Australia, Australia
[4]The Aboriginal Health Council of SA, South Australia, Australia

Introduction: The Child and Family Health Service offers a two-year, nurse-delivered family home visiting (SA-FHV) program to new mothers in South Australia. Having an Aboriginal infant is one of four key population criteria considered for entry to the program. To assess whether the program has been equally successful for Aboriginal families, we compared outcomes for Aboriginal and non-Aboriginal families.

Methods: The full evaluation of the SA-FHV program[1] included 77 Aboriginal and 393 non-Aboriginal infants. At 24 month follow-up, data from 40 Aboriginal infants and 209 non-Aboriginal infants was available. Linear regression was used to compare outcomes between Aboriginal and non-Aboriginal families enrolled in the SA-HV program.

Results: At baseline, mothers of Aboriginal infants had scores indicating better functioning on the Parenting Stress Index Competence and Attachment scale compared to mothers of non-Aboriginal infants. The association remained for the Competence scale after adjusting for confounders. Mothers of Aboriginal infants continued to score better on the Competence and Attachment scales at 24 months. Mothers of Aboriginal infants also reported fewer concerns about child feeding at 9 months. Aboriginal infants had higher rates of accidents/injuries at baseline and at 24 months. There were no significant differences on other outcomes assessed; maternal mental health, child development and health service use (preventive or emergency).

Conclusion: The SA-FHV program appears to achieve comparable outcomes for mothers of Aboriginal and non-Aboriginal infant. The program has developed a high level of cultural competency including nurse visits being supported by Aboriginal cultural consultants. Because one of the entry criteria is Aboriginality, it may be that some Aboriginal families who take up the offer of FHV are already functioning well in terms of parenting and extended family support.

Reference:

1. Sawyer et al. Effectiveness of nurse home-visiting for disadvantaged families: results of a natural experiment. *BMJ Open* 2013

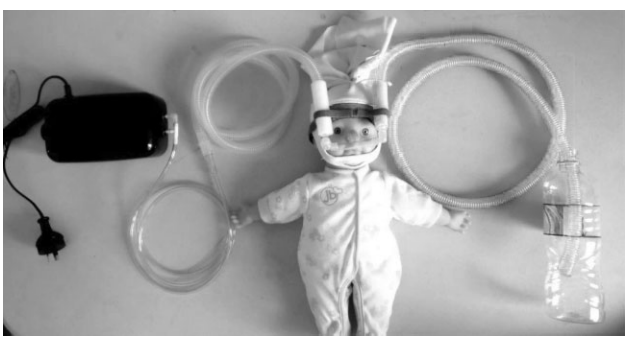
CHEAP CPAP FOR THE DEVELOPING WORLD

Ho D[1], Whitehall J[1], Maday J[2], Edwards M[1]

[1]University of Western Sydney, New South Wales, Australia
[2]Campbelltown Hospital, New South Wales, Australia

Acute respiratory distress is a major cause of infant mortality. In developed countries, bubble CPAP is employed with Fisher&Paykel (F&P) devices, costing up to AUD$8000 for installation and AUD$200 for consumables. Cheaper CPAP is needed for developing countries We compared CPAP from F&P with an over-the-counter fish tank pump (FTP).

Methods: Pressures generated by FTP and F&P at various flow rates were measured. The FTP system includes: fish-tank-pump (Sunsun Air Pump HP-1116), 11 mm diameter tubing, Teleflex Hudson RCI nasal cannula and a water bottle. F&P readings were obtained from a hospital set-up which correlated with data from their product brochure[1]. The water level gauge was set at 5 cm and 10 cm to create corresponding system pressures. The pressures were measured at the CPAP nasal cannula using a Fluke 922 Airflow Meter.



© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

Results:
Table 1: CPAP generated by F&P and FTP at various flow rates and gauge depths.

Water Gauge set	5 cm		10 cm	
Flow Rate [L/min]	F&P[1]	FTP	F&P[1]	FTP
5	5.3	5.4	10.1	10.2
7	5.6	5.9	10.4	10.7
11	6.5	6.4	10.9	11.3
12	6.8	6.8	11.1	11.6
14	7.3	7.0	11.4	11.8

Discussion/Conclusion: FTP generated consistent and comparable pressures with F&P (Pearson correlation coefficient 0.997) at a unit cost of approximately AUD$50, which is also less than a recent 'cheap' African system (USD$350/unit)[2]. FTP does not require hospital gas. It draws room air with attendant warmth and humidity, and worked efficiently for over 14 days. Oxygen can be added using a T-piece connected to the input tubing.

References

1. Bubble CPAP system set-up guide. Fisher & Paykel Healthcare; 2011. Available from: http://www.fphcare.com.au/products/bubble-cpap-system/
2. Brown et al. A high-value, low cost bubble (CPAP) for low resource settings. *PLOS One*. 2013. **8** (1). E53622

THE HIGH IMPACT OF RARE CHILDHOOD DISEASES: FREQUENT USE OF MULTIPLE HEALTH PROFESSIONALS AND SERVICES

Zurynski Y[1,2], Deverell M[1,2], Christodoulou C[2,3,4], Leonard H[5,6], Phu A[1,2], Dalkeith T[2,3], Johnson SLJ[2], Elliott E[1,2,4]

[1]*Australian Paediatric Surveillance Unit*
[2]*Sydney Medical School, The University of Sydney*
[3]*Western Sydney Genetics Program*
[4]*Sydney Children's Hospitals Network*
[5]*University of Western Australia*
[6]*Telethon Institute for Child Health Research*

Background: There are over 8000 different rare diseases. Most have onset in childhood, are chronic, complex, and require frequent specialist care. Few studies have analysed the impact of rare childhood diseases on health services.

Aims/Objectives: We aim to describe health service use in a large group of Australian children living with rare diseases.

Methods: A survey was sent to families who have children aged <18 yrs with a rare disease. The survey asked about health services use over the last 12 months. Families were recruited from the Steve Waugh Foundation, The SMILE Foundation, the Association of Genetic Supports of Australasia and the Genetic Metabolic Disorders Service, at the Children's Hospital at Westmead. Surveys were completed by a parent or carer.

Findings: 443 surveys have been completed. The 443 children represented ~300 different rare diseases; 24 had no final diagnosis. The children were evenly distributed across age groups (median 9 yrs) and 95% were Australian born. In the last 12 months 367 patients visited a general practitioner and accounted for a total of 3011 visits; 260 patients visited a paediatrician- total 1057 visits. Other specialists were also frequently accessed: geneticists (151 patients; 363 visits), surgeons (131 patients; 382 visits), and neurologists (127 patients; 343 visits). There was a substantial number of visits to allied health professionals: occupational therapists (183 patients; 1737 visits); physiotherapists (178 patients, 2566 visits) and speech pathologists (154 patients, 2060 visits). 242 patients had at least one hospital admission.

Conclusions: The use of primary care, specialist and allied health services is very high and the associated costs are likely to be significant. We are currently analysing the cost impacts of rare childhood diseases on in-patient services. Families called for integrated multidisciplinary clinics to avoid fragmentation of care, to save costs to the family and to improve their experiences.

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)

AUTHOR QUERY FORM

Dear Author,
During the preparation of your manuscript for publication, the questions listed below have arisen. Please attend to these matters and return this form with your proof.
Many thanks for your assistance.

Query References	Query	Remarks
1	AUTHOR: Please provide an affiliation link for author Stephen McBride.	
2	AUTHOR: Please provide an affiliation link for author Mark Thomas.	
3	AUTHOR: Please provide an affiliation link for author Lynette Sadleir.	

Author Index

© 2014 The Authors
Journal of Paediatrics and Child Health © 2014 Paediatrics and Child Health Division (Royal Australasian College of Physicians)