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ORAL PRESENTATION

IRISH NEONATAL MORTALITY STATISTICS FOR 2006

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Aims: To determine the exact cause of neonatal deaths in Ireland for 2006.

Methods: All Paediatric and maternity Units in Ireland were contacted by post and phone, through the Faculty of Paediatrics in the College of Physicians, Ireland. Paediatricians were requested to fill a questionnaire which dealt with data pertaining to all live-born infants who died within the first 28 days of life. Details requested included gestation, birth weight, sex, age at death, mode of death and principle cause of death. Data was entered into an Access Database. Each hospital and infant was given a unique ID number prior to computer entry. Deaths were categorized into as per Wigglesworth classification into Lethal Malformation, Prematurity, Asphyxia and “Specific”.

Results: The response rate was 98%. The number of births in 2006 in Southern Ireland was 64,237. The overall number of neonatal deaths for 2006 was 128. 21% had post mortem performed. The principle causes of death were Congenital Malformations 34% ($n = 43$), Prematurity 49% ($n = 63$) and Asphyxia 10.9% ($n = 14$). This compares to figures of 39, 35 and 8% for 1987 and 39, 30 and 12% for 2001. The overall Neonatal Mortality Rate for 2006 was 2.0/1000 compared to 3.1/1000 for 2005. The Corrected NMR was 1.3 compared to 1.8/1000 for 2005.

Conclusions: There is a slight reduction in the overall Neonatal Mortality and the corrected NMR rates in Ireland in the past 12 months compared to previous years. This may be due to an increase in the total number of births or due to increased reporting, which is more likely with good outcome. The number of deaths attributable to asphyxia remains 10%.

AUDIT OF TWIN PREGNANCIES PRESENTING AT THE COOMBE WOMEN'S HOSPITAL

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Introduction: Since 1970, the prevalence of multiple births has been increasing because of more widespread use of assisted reproductive technologies to treat infertility. The overall prevalence of twins in Ireland is approximately 12 per 1000 births. Multiple pregnancies are high-risk pregnancies with numerous associated fetal and neonatal complications, and are known to provide additional management challenges in the perinatal period. Infants from multifoetal pregnancies are more likely to be born prematurely and are more likely to

require neonatal intensive care. The fetal mortality rate for twins is 4 times the fetal mortality rate for single births. The neonatal mortality rate for twins is more than 5 times greater than the neonatal mortality rate for single births. Approximately 50% of twin deliveries occur before 37 weeks' gestation.

Aim: The aim of this study was to audit the twin pregnancies presenting for admission to NICU/SCBU in the Coombe during July, August and September 2007.

Method: An audit was undertaken of all twin deliveries in the Coombe requiring admission to the NICU and SCBU from July to September 2007 using the Euroking K2 clinical information computer system. The parameters assessed were gestation, BW, IVF pregnancies, duration of stay and respiratory supports required. Charts of the twin babies delivered in peripheral units and postnatally referred up to our unit for tertiary care were also analysed and the above parameters assessed.

Results: There was a total of 68 twins deliveries between July and September 2007 in the Coombe Hospital, of which 40 babies (58.8%) were admitted to the Unit. An additional 2 babies were referred from peripheral units for specialist care. The mean duration of stay was 16 days with 5 babies still inpatients in the Coombe requiring ongoing care. Three were transferred to Crumlin hospital for further specialist treatments and 2 babies were transferred back to peripheral units for stepdown care.

Gestation ranged from 29+3/40 to 40+1/40. Mean gestation was 34 weeks. Mean birth weight was 1.99 kg. Low birth weight (<2.5 kg) was the main reason for admission. 13 of the 40 babies (32%) delivered had assisted fertility treatments; clomiphene and/or IVF. 6 of the babies (8%) were documented as MCDA twins. 23 babies did not need any additional ventilatory supports and further ventilatory supports varied significantly according to gestation and other associated complications.

Conclusion: Therefore given the degree of prematurity, reduced birthweight and higher incidence of complications in twin gestations (in particular respiratory distress) they result in significantly increased resource use in NICU.

NEUROMETABOLIC DISORDERS IN THE 'NEW' IRISH POPULATION

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Background and aims: The 2006 Government of Ireland census reported a 10% increase in our population since 2002, the majority of these being of non-Irish ancestry. Our aim was to survey this 'new' Irish population as it is represented in our service at the NCIMDs in order to identify trans national minority groups and associated

inherited metabolic disorders (IMDs). Chelating this epidemiological data is important for two reasons:

1. It has implications for service planning and provision of care.
2. Racial-cultural issues are potential obstacles for those seeking treatment for genetic disease.

Methods: Case notes of patients seen at the NCIMDs between January 1st and December 2007 were reviewed and those of non-celtic ancestry identified. Cultural background and details such as diagnosis, genotype, phenotype, history of consanguinity and need for an interpreter service were recorded. Patients of mixed race were excluded. A literature search was then conducted to determine common and unique recessive disorders harboured by the most prevalent minority groups.

Results: We found 51 patients, representing 3.7% of our patient population. The most prevalent groups were Nigerian (13 patients or 1% of total patient population), Roma gypsy (5), Lithuanian (3) and Polish (4). There were three sibship pairs and 10% of patients were products of a consanguineous union. No novel IMDs were identified. However genotypes and phenotypes of common and familiar IMDs differ from those of the 'traditional' Irish population. The presence of a translator was required in 20% of cases. Our literature search revealed several publications on genetic disorders in the Roma but little information on recessive metabolic disease in other nationalities.

Conclusion: Our study strengthens current awareness of the wide range of cultural customs, social attitudes, health and religious beliefs that exist in a society undergoing rapid change and is of direct relevance to health care.

THE NEED FOR EARLY DISCHARGE GUIDELINES

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Background: There has been a large increase in the childbearing age population in North Dublin, e.g. the population has increased by a quarter in Dublin 15. In order to tackle the problems of bed and personnel shortages there has been a growing trend towards early hospital discharge. It is now becoming common for mother and baby to be discharged 24 h or less postnatally. There is however concern about this and strategies must be in place to ensure the safety of early discharge.

Aims: To review the literature regarding early discharge planning, with particular reference to hospital readmission rates, outpatient referral and presentations to A&E. To develop guidelines for early discharge of newborn infants for the Rotunda hospital based on the American Academy of Paediatrics Guidelines.

Methods: A literature review was performed. Data was collected on the number of births and outpatient appointments in the Rotunda hospital during 2006 and 2007. The services available for early discharges in the Rotunda Hospital were reviewed and an early discharge policy has been developed based on evidence based guidelines.

Results: In recent years numerous studies have been published to ascertain the safety of early discharge policies. These studies looked mainly at hospital readmission rates to determine if early discharges are safe and cost effective. However even with careful screening and early follow up, several studies have observed potentially clinically significant increases in adverse outcome.

Within the Rotunda hospital, a midwifery led early discharge service, called DOMINO is already established, consisting of 8 midwives who, in combination with the local GP manage the care of low risk pregnancies both antenatally and postnatally. This ensures community follow up of infants discharged home at 6–12 h of age, and daily review. On average 10–14 babies are discharged weekly under the care of the DOMINO scheme. A similar service the Early Transfer Home (ETH) service is currently being established.

Last year in Ireland, there were 64,237 births. Over the last 10 years the number of births has increased by 27%. Already this year, up to end of September, there has been 6089 deliveries in the Rotunda, which is an increase of 15% compared to the same period last year. With only 72 postnatal beds available, bed shortages and nursing manpower shortages have put increased pressure on paediatric NCHDs and consultants to discharge infants at barely 24 h of age, outside a DOMINO or ETH arrangement and without any formal follow up support services in place. With limited community services available, especially at the weekend, ensuring that these infants are appropriately followed up is putting additional stresses on an already strained paediatric service. This is particularly evident in paediatric outpatients department where during the period June to July 2007 the average monthly attendance was 1008, 16% of which were reviewed at the weekend, when services are additionally limited.

Conclusion: Early discharge policies without appropriate support and services are potentially dangerous. Based on the AAP guidelines, we propose the following recommendations.

- Mothers and newborns considered for early discharge must be either under the care of the DOMINO or ETH service or the infant must be 24 h of age who fulfills all the early discharge check list criteria, including term birth, normal vital signs, and passed urine.
- Appropriate early follow up must be specifically arranged for all discharged infants.
- Services must be put in place to ensure that there is adequate numbers of trained staff available to manage the care of these infants in both the emergency and paediatric OPD.
- More community services must be established.
- Hospital admission policies must readily facilitate the readmission of babies to hospital for at least the first 14 days of life.

ANALYSIS OF INPATIENT DERMATOLOGY REFERRALS: TO OPTIMISE THE EDUCATIONAL NEEDS OF PAEDIATRIC TRAINEE DOCTORS

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Introduction: Inpatient consultations form a significant component of the dermatologists' workload requiring adequate time for the initial assessment and subsequent review of patients. We present our data of inpatient children seen on consultation by a Dermatologist.

Aim: To identify the educational needs of junior / trainee doctors by analysis of paediatrics inpatient consultation referrals to the Dermatology Department.

Methods: We reviewed the consultation data of inpatient children referred to the Dermatology Department between August 2001 and August 2007 that had been stored on a Filemaker Pro database. These were a randomised subset and represented approximately 50% of all inpatient consultations to the Dermatology Department in that time period.

Results: 220 consultation records were identified. 95% were for patients at Limerick Regional Hospital and 5% from the Maternity Hospital. The age ranged from a 33-week premature baby with skin necrosis from candidal septicaemia to a 16-years old female with atopic dermatitis.

152 (69%) children were admitted primarily because of their acute skin condition and this was the primary reason for their admission. In 78 (35%) cases, the Consultant Dermatologist's diagnosis was different to the inpatient referral diagnosis on the consultation referral form. The seven commonest dermatoses diagnosed were atopic eczema (33%), urticaria (10%), impetigo (5%), staphylococcal scalded skin syndrome (5%), seborrhoeic dermatitis (4%), psoriasis (4%) and tinea infection (3%). Even among one diagnostic group (tinea infections), there are interesting trends emerging with indigenous children developing scalp ringworm infection in immigrant population with unusual trichophyton like trichophyton tonsurans infections. 75 patients (34%) required one consultation visit but 145 (66%) required 2 or more review visits from the Dermatology Team during that admission. 93 patients (42%) required follow up in Dermatology Outpatients after their admission.

Discussion: The Dermatological conditions seen ranged from minor to life threatening dermatoses and acute skin failure. Over half the referrals were admitted via the Accident and Emergency department primarily because of their acute skin failure indicating that a significant proportion of these dermatology patients were seriously ill. That 33% of referrals were for children with the atopic dermatitis reflects poorly on facilities available for their care in the community and hospital outpatient setting. In more than one third of the referrals the Dermatologist's diagnosis was different to the admitting team. This has important implications on clinical outcome as appropriate treatment and short length of stay would be dependent on accurate and timely diagnosis. The complex nature of some of their dermatoses is reflected in 66% requiring more than one consultation during the admission and 42% needing follow up in Dermatology Outpatients.

Conclusions: This study emphasises the need for both junior Dermatology and paediatric trainees to undertake extra training in the Dermatologic conditions of paediatric patients as they form a significant component of the inpatient referrals to Dermatology (over quarter of all referrals). Further teaching is required of dermatoses affecting the dark skin. It is also valuable for the patients with acute skin failure to have access to inpatient beds. Our data supports the need for expansion of service provision of Dermatology in the Region to help contribute timely diagnostic and therapeutic advice.

WHAT ARE THE DUTIES OF NEONATAL SENIOR HOUSE OFFICERS: COULD INTERNS UNDERTAKE THE ROLE?

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Objectives: This project was undertaken to document the duties of the Neonatal Senior House Officer (SHO) and to determine whether an Intern could fulfill the role.

Population: Neonatal SHOs working in the Department of Neonatology, National Maternity Hospital, Holles Street, Dublin, Ireland.

Methods: The daily duties and activities of the Neonatal SHOs were documented and timed over a 5-week period. Activities were grouped into three categories: clinical, procedures, clerical/communication tasks. All tasks were subsequently rated on a four point scale of

difficulty: Easy, Standard, Difficult, Very Difficult. It was then determined whether an Intern could effectively perform the task.

Results: A total of 2,088 min of SHO work was studied. Of this 740(35.4%) minutes was spent in clinical tasks, 695(33.3%) minutes undertaking procedures and 652 (31.2%) in carrying out clerical/communication work.

Of the 2,088 min studied, 1,729(82.8%) were spent undertaking tasks deemed to be Easy or Standard. The other 359(17.1%) minutes relate to tasks evaluated as being Difficult or Very Difficult.

The tasks reported as Difficult were clinical examination in the A&E, intravenous drip insertion, doing an ECG. The task placed in the Very Difficult category was performing a lumbar puncture.

Conclusion: This study provides a comprehensive picture of the duties undertaken by the Neonatal SHO. Over 80% of the tasks are considered to be easy or standard. The small number of tasks that are considered difficult can be readily addressed. The matter of clinical evaluation in the A&E requires a more supervision, while further training is needed for some of the procedures.

The findings in this study indicate that Interns could undertake the role of a Neonatal SHO. We suggest that rotations of 3–4 months duration in Neonatology should be introduced into a number of Intern posts.

PROVIDING AN INTRODUCTION TO CARBOHYDRATE ESTIMATION EDUCATION IN A GROUP SESSION

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The introduction of multiple daily injection (MDI) regimens and insulin pump technology has seen a revival in the interest in using varying forms of carbohydrate estimation in the dietary management of T1DM.

The Nutrition Subcommittee of the Diabetes Care Advisory Committee of Diabetes UK, the ADA and the WHO, recommend adjustment of pre-meal insulin doses for the carbohydrate content of the meal, BDA 2003.

Children and young people using multiple daily injection regimens should be offered education about insulin and dietary management as part of their diabetes care package, to enable them to adjust their insulin dose to reflect their carbohydrate intake (Nice guideline 2004).

Carbohydrate counting can be effectively taught in small groups using real life examples and practical interactive activities (FACTS 2005).

Aims:

- To design and pilot group education sessions on carbohydrate counting to all children and their families, who attended the diabetes with Type 1 Diabetes clinics in Cavan/Monaghan.
- The educational content aimed to provide a revision of the basics of glycemic control and diet, outline the benefit of increasing flexibility in relation to both eating habits and insulin injections and teach the skill of carbohydrate counting using practical examples.
- To pilot new ways of delivering information with the aim of improving the education process and maximizing the level of engagement both parents and children in the education process.
- To promote an emphasis on teamwork and improved communication among children and families attending.
- To encourage supported problem solving and sharing of the complicated tasks involved in a child's daily diabetes

management routine with the aim of fostering less dependence on health professionals.

- To facilitate peer support and improved relationships among the group.

Methods: There were a total of 13 afternoon and evening sessions, two per month from November 2006 to July 2007. Sessions were allocated by grouping of ages 1–12 and 13–18 years and lasted on average 2 h 15 min. The sessions were facilitated by the paediatric dietitian.

Session format:

1. Informal introductions/icebreaker exercise.
2. Brief review of basic dietary knowledge using group brainstorming.
3. Brief overview of the role of diet and insulin using story telling/question format with toys/props.
4. Outline of carbohydrate estimation and its use with mdi therapy.
5. Family task: carbohydrate estimation exercise using props and carbohydrate tables with group discussion.
6. Family task: food labels reading exercise with explanation and support from facilitator.
7. Summary and question and answer session.
8. Written evaluation.

Results: Group attendance averaged approximately 7 people per session. In total 73 people partook-representing 34 children/adolescents with T1DM and one/two parents/carers and siblings. Seven children were educated individually in the clinic setting. Evaluation showed an improvement in confidence ratings regarding the management of blood sugar using diet and insulin pre/post session. Qualitative feedback and suggestions for future activities were recorded.

Discussion: The group sessions and evaluation have generated practical learning points for future planning regarding: organization, content and format. Ideas to enhance and support these sessions include the need for prompt review, increased phone support and incorporation of the sessions into the clinic format to increase participation.

Suggestions for development of multidisciplinary group sessions regarding carbohydrate counting and other topics were noted.

REACH OUT AND READ

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Background: Early childhood language development requires exposure to spoken language. Children exposed to broader vocabularies and frequent reading aloud tend to have better language development and an easier transition to school. Conversely poor exposure is associated with delays in language acquisition, future difficulty with reading and a negative impact on academic success.

In 1989 paediatricians in the U.S. implemented a programme to promote early childhood literacy known as “Reach out and Read” (ROR). This programme involves giving parents developmentally appropriate books and counselling at various developmental checks ranging from 6 months to 5 years of age. Parents are encouraged to incorporate reading aloud into their daily routines.

In autumn of this year the Rotunda Hospital implemented the first Reach out and Read programme in Ireland.

Aims: To evaluate the effectiveness of the programme, prior to its initiation we surveyed the baseline reading and literacy activities of

families being followed in our POPD. Follow up surveys will be performed at regular intervals to assess the impact of the programme.

Methods: During July and August 2007, patients’ families attending POPD consultant clinics were surveyed. The survey assessed activities of daily living focussing on those associated with language development and sleep routines.

The outcomes to be evaluated included: frequency of reading aloud, number of books in the home, inclusion of reading as a favourite shared activity, sleep/bedtime routines and any previous encounters with a healthcare professional about language development education.

Results: 47 surveys were fully completed. The average age was 9.0 months (corrected gestation). Ages ranged from 2 to 36 months.

24 (51%) out of 47 parents read to their children. The average age of commencement was 5.9 months. 18 (38%) parents read to their children daily with a mean daily reading time of 24 min. 5 included it as part of their bedtime routine. 29 (62%) parents had books for their children at home with a median number of 6 books. 5 (11%) parents listed reading as one of their child’s top 3 favourite activities or top 3 joint parental activities.

4 (9%) of 47 families had received advice from a doctor or nurse with regard to their child’s language development and 38 (81%) of the 47 were keen to receive more information.

Conclusion: Our results suggest that a significant number of children attending our POPD are not being exposed to reading aloud at an early age. This may impact on future academic success. There is also a clear need for healthcare professionals to provide more information at routine visits about language development.

The ROR programme has commenced at the Rotunda Hospital. We anticipate that the programme will increase the frequency and enjoyment of reading aloud, decrease the age when parents start reading to their children and improve parents’ satisfaction with the healthcare visit. Furthermore the programme should raise awareness among healthcare professionals of the importance of language development in this age group.

Reference: www.reachoutandread.org.

AN AUDIT ON PAEDIATRIC ANALGESIA IN AN IRISH EMERGENCY DEPARTMENT

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Introduction: Inefficient management of pain in the Emergency Department (ED) is a well-documented problem, particularly in children. While this has been shown in other countries, little is known of the success of Irish EDs in providing prompt analgesia for their paediatric patients.

Aims: To describe, and improve, the provision of analgesia for children in an Irish ED through the use of a nurse-initiated analgesia protocol.

Methods: 95 children aged 1–16 presenting to the ED over 7 days were categorised into those with major fractures, and those with other diagnoses. Time from triage to analgesia in both groups and the rate of analgesia provision in the major fracture group were recorded. The results were circulated amongst staff and a nurse initiated analgesia protocol was introduced. A second audit including 145 patients over 10 days assessed the effect of the interventions.

Results: 55.6% of patients with major fractures received analgesia after a mean time of 54 min, which insignificantly improved to 61.1% ($p = 0.735$) after 7 min ($p = 0.004$). There was no improvement in time to analgesia for children without a major fracture. No child had a

documented pain score, which slightly improved to 19.3% following interventions.

Conclusions: The provision of analgesia for children in this Irish ED was inadequate and poor compliance with the protocol resulted in an unsatisfactory improvement following interventions. All Irish EDs should audit their provision of analgesia for children, and work to ensure compliance with any interventions made.

ARE WE ADEQUATELY PROTECTING CHILDREN FROM SOLAR ULTRAVIOLET RADIATION?

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UCC

Introduction: Skin cancer is the most common cancer type in Ireland. Sun exposure is the most important risk factor. 50–80% of lifetime sun exposure occurs in childhood.

Objective: The primary aims of this study were: to estimate sun exposure in hours of children in Cork during the summer months; to examine sun protection measures used by children and their parents and to explore parental knowledge of sun exposure and protection.

Design: Cross-sectional study, using a semi-structured questionnaire, conducted in June 2006 in primary schools, pre-schools and crèches in Cork City and County.

Results: Parents of 250 children aged less than 12 years were sampled. Response rate was 180 (72%). Thirteen children with eczema were excluded, leaving a study group of 167 children. 99 (59.3%) were of skin type 1 or 2. Mean sun exposure of Cork children is 40.9 h per week in the summer months, with 77 (46.1%) children developing sunburn. 95 (57%) children on weekdays and 137 (82%) children on weekends are outdoors between 11 am and 3 pm. Sunscreen and hats/caps are the most common protection measure used. A minority used protective clothing, sunglasses or sought shade. Only 31 (30.5%) children have sunscreen re-applied every 2 h. 109 (65.3%) parents are confused by sun protection factors. One hundred and thirteen (67.7%) parents think sunscreen protects, so their children can stay outdoors for longer.

Conclusion: Irish children spend substantial periods exposed to strong sunlight without adequate protection. Parents need better information about what actual protection sunscreens offer their children.

PAEDIATRIC MULTI-DRUG RESISTANT TUBERCULOSIS IN IRELAND—THE FIRST OF MANY?

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We present the nine child contacts identified on contact tracing of a highly infectious HIV positive adult female with multi-drug resistant tuberculosis (MDR-TB) including pulmonary cavitary disease and laryngeal disease. These children were also a contact of another adult with fully sensitive TB. This group includes the first case of MDR-TB in a child in Ireland.

Child 1, aged four years and child of MDR-TB index case was asymptomatic. Examination was normal. She had not received BCG and was HIV negative. Chest radiograph revealed mild hilar enlargement. Mantoux test (2 TU) was 20 mm. Gastric washings revealed *Mycobacterium tuberculosis* resistant to isoniazid, rifampicin, streptomycin, and ethambutol. The organism's sensitivities included amikacin, capriomycin, ciprofloxacin, clarithromycin, cycloserine, ethionamide and linezolid. She was commenced on directly observed treatment (DOT) with five agents including intravenous amikacin for six months necessitating portacath insertion and treatment was planned for 1 year with oral ofloxacin, ethionamide, cycloserine and high dose isoniazid.

Child 2 aged 18 months and child of index case. She was asymptomatic and examined normally. She was HIV negative and had received BCG. Chest radiograph was normal and Mantoux was 17 mm. She had negative gastric washings and a positive quantiferon test. She received ethionamide, ofloxacin, and high dose isoniazid for latent MDR-TB for six months.

Child 3 was a domestic contact with normal chest radiograph and negative mantoux. He received similar treatment to case 2 for latent TB. Child 4 was a domestic contact who had a negative mantoux and normal chest radiograph and she received no treatment.

These children had no adverse effects to date. Hematology and biochemistry remained normal. Cases 1 and 2 developed mild elevation in TSH but with normal T₄. Audiology remained normal.

Five further children had limited contact (2–3 h) with index case with MDR-TB but were a domestic contact of an individual with pan-sensitive TB. Three had positive mantoux, negative gastric washings normal chest radiographs and received INH prophylaxis. One had negative mantoux and normal CXR but was put on INH in view of her age. One had an abnormal chest radiograph with lobar disease and positive mantoux but negative washings and was started on usual treatment for TB as clinicians were not aware of MDR-TB contact at that point. This child responded to treatment for pan-sensitive TB.

Management of MDR TB and MDR-TB exposure in children presents a number of challenges including choice and duration of treatment and risk of side effects. If a lone parent is the index cases ongoing risk of exposure due to poor compliance with DOT may mean children require foster care. All of the children in this group remain currently well but will require long term follow up.

TB OUTBREAK IN CORK CRECHE: WHAT WAS THE SPECTRUM OF TUBERCULOSIS DISEASE MANIFESTATIONS IN CHILDREN WITH ACTIVE DISEASE?

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Background: In March 2007, an infectious case of sputum smear positive pulmonary tuberculosis (TB) was notified in a Cork crèche worker. In immune-competent children, age is the most important variable that determines risk of developing disease following *M. tuberculosis* infection; young children (<3 years of age) are at highest risk. Of the 268 children exposed, 18 cases of active TB were diagnosed. We describe the spectrum of TB disease manifestations at presentation in a cohort of young children.

Methods: The children diagnosed with active TB disease (diagnostic criteria: contact with infectious case, positive mantoux (2TU)

≥5 mm, abnormal chest radiograph and positive microbiology) were identified. The medical records of these cases were reviewed.

Results: There were 18 cases of active TB (results follow for 17); aged 2.25–4.5 years. None had received BCG. 16/17 were symptomatic. 12 had cough, all non-productive. 6 had fever, 5 had night sweats, 4 were coryzal, 2 had weight loss, 2 had decreased appetite, and 1 child had lethargy. 9/17 children had ≥2 symptoms. On examination 4 had cervical lymphadenopathy, 1 had crackles, 1 had erythema nodosum. Ten children had positive mantoux ≥5 mm (13–20 mm), 1 had a negative mantoux (0 mm) 6 weeks after exposure but a positive mantoux (18 mm) 4 weeks later, and 6 had negative mantoux (0 mm). Regarding radiographic features: 5 had hilar adenopathy, 5 had lobar consolidation and 7 had hilar adenopathy and consolidation (1 had pleural effusion). 11/17 had gastric washings; 5/11 were culture positive (45%).

Conclusions: There were no systemic manifestations of TB. Initial false negative rate for mantoux test was high (41%). Protective effect of BCG against lung disease may be greater in young children as no child with BCG developed active disease.

COMPLICATED PNEUMONIA—IS THERE A RISE IN INCIDENCE IN CORK?

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Background: An increase in the incidence of complicated pneumonia over the last two decades has been documented internationally. The main cause of complicated pneumonia is *Streptococcus Pneumoniae* although other organisms such as *MRSA* have been implicated. There has been a rise in complicated pneumonia in some areas since the introduction of pneumococcal vaccination, possibly related to sero-type shift.

Aims:

- (1) To document the incidence of complicated pneumonia in Cork University Hospital over the last decade prior to introduction of pneumococcal vaccination.
- (2) To examine whether there has been a rise in incidence.

Methods: Complicated pneumonia was defined as pneumonia (clinical illness, cough, fever, lethargy, dyspnoea with radiological changes on chest radiograph) associated with any of the following: (i) Empyema or pleural effusion (ii) Lung abscess (iii) Necrotising Pneumonia (iv) Requiring chest tube and/or thoracotomy. Hospital records were examined for time period 1996–2006. Search for 1996–2000 still in progress. Population included children aged 4 weeks to 16 years and 364 days.

Results: Search revealed 18 cases of which seven met eligibility criteria. Two cases presented in 2006, four cases in 2005 and one case in 2001. All presented in spring or winter. Three had antibiotics from GP prior to admission. Mean age 10 years was with five females and two males. All seven patients had chest drains inserted, three required decortication surgery by thoracotomy, one required decortication via thoracoscopy and mini thoracotomy. Mean length of stay (mean 17 days) with one patient requiring re-admission and one patient transferred to tertiary referral centre. Different organisms were

isolated in three separate cases—*Streptococcus pyogenes* *Streptococcus Pneumoniae* and *Staphylococcus aureus*.

Conclusion: The incidence of complicated pneumonia is estimated to be low compared to international figures. Based on our limited data there appears to be a recent rise in incidence in Cork. However we cannot exclude a case ascertainment bias which could impact on result. We feel that the incidence of complicated pneumonia should be prospectively monitored on a national basis following introduction of pneumococcal vaccination in January 2008.

PARENT WELL-BEING AND FAMILY SUPPORT ARE ASSOCIATED WITH BETTER METABOLIC CONTROL, ADOLESCENT QUALITY OF LIFE AND LOWER FAMILY BURDEN IN 2,062 ADOLESCENTS WITH TYPE 1 DIABETES

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Objectives: To assess and determine the roles of parent well-being and family support in glycemic control and quality of life of adolescents (QOL) with Type 1 Diabetes.

Methods: Clinical data and centrally analysed HbA1c were collected on 2,062 adolescents, aged 11–18 years and from 1994 parents from 21 centers in 19 countries in Europe, Japan, North America and Australia. Adolescents completed QOL, well-being and Life Ladder questionnaires. Parents completed WHO5 well-being and Family Burden questionnaires.

Results: Mean HbA1c was 8.2 % (±1.4%), higher in girls 8.3 (±1.5%) than boys 8.1(±1.3%, $p < 0.0001$). Good parent self rated wellbeing scores were associated with lower HbA1c ($p < 0.005$), greater adolescent well being ($p = 0.001$), less impact of diabetes ($p < 0.001$), less worries ($p < 0.001$), less adolescent perception of parent over involvement ($p < 0.01$), greater health perception ($p < 0.001$) and life ladder ($p < 0.001$), less physical and psychological symptoms ($p < 0.001$). Greater family burden was associated with lower parent well-being, higher HbA1c values ($p = 0.000$), poorer adolescent well being ($p = 0.000$), greater impact of diabetes ($p = 0.000$), more worries ($p = 0.000$), more parent over involvement ($p = 0.000$), poorer health perception ($p = 0.000$) and life ladder ($p = 0.000$), more physical and psychological symptoms ($p = 0.000$). Adolescents accompanied to clinic by parent had lower HbA1c 8.1 vs. 8.6% ($p < 0.007$), greater well being ($p = 0.000$), less worries ($p = 0.000$), greater health perception ($p < 0.003$) and life ladder ($p = 0.000$).

Conclusion: Good parent well-being is associated with lower family burden relating to diabetes, better metabolic control and adolescent QOL. Parent attending diabetes clinic is associated with better glycemic control and QOL in the adolescent. Parent QOL and family burden assessment form an important part of diabetes care.

INHERITED METABOLIC DISORDERS MIMICKING NON-ACCIDENTAL INJURY

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Background: Several inborn errors of metabolism have been reported to present as non-accidental trauma (NAT). Parents were erroneously accused of harming children subsequently found to have subdural hematomas secondary to Glutaric aciduria type 1, 1-hydroxyglutaric aciduria and Mucopolysaccharidosis type 111 (San Filippo syndrome). Other presentations were osteoporotic femoral fractures due to protein deficiency in propionic aciduria and bruising secondary to vasculopathy in Menkes disease. In addition presentations mimicking non-accidental injury have been reported with Gaucher's disease (bruising and epistaxis), congenital disorders of glycosylation (intracranial hemorrhage), homocystinuria and methylmalonic aciduria (retinal hemorrhages)

However, children with neurometabolic disorders are known to be at increased risk of NAT and therefore concerns regarding a child's safety and protection should not conclude when the diagnosis is made.

Aims: We aimed to identify patients with metabolic disorders presenting with apparent non-accidental injury in our cohort, to estimate the prevalence of child abuse in patients with metabolic disease in Ireland and to formalize our approach to the "metabolic screen" for non accidental injury.

Methods: We reviewed the files on all patients <16 years seen between 1 July 2005 and 1 July 2007 to document cases in which the constellation of symptoms and signs evoked consideration of NAT.

Results: 11 cases were identified 6 of whom were proven to have been genuinely injured by a carer and were not found to have any evidence of metabolic disease on screening. 5 patients presented with clinical signs and symptoms suggestive of non-accidental injury which were explained by complications of an underlying metabolic disorder.

No case of physical child abuse was found in any patient with established metabolic disease during the study period.

Conclusion: Inherited metabolic disorders should be considered in the differential diagnosis of non-accidental injury and appropriate screening/evaluation/investigation performed. As a group they present a difficult diagnostic challenge and there are huge social and legal implications to an erroneous diagnosis.

THE ECONOMIC IMPLICATIONS OF RSV BRONCHIOLITIS ON THE MID-WEST

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Background: Prophylaxis for RSV bronchiolitis is an important but costly means of reducing admissions to paediatric wards. The product literature, based on clinical trials, recommends that children born at less than 35 weeks gestation who are <6 months at the start of the RSV season should be given a course Palivizumab. The high cost of this monoclonal antibody has huge implications on local financial allocations. The data for its use however largely originates in the

United Kingdom and United States of America and our literature search did not reveal studies performed on its cost-effectiveness in the Irish paediatric population.

Aims: The aims of the study were to determine;

1. the economic impact of RSV bronchiolitis in a 7 year cohort, and
2. whether broadening the inclusion criteria for prophylaxis would be cost-effective.

Method: A retrospective HIPE-data analysis provided information on in-patient cases of diagnosed bronchiolitis over a seven year period from October 1999 to January 2007. Only patients who had been delivered in the Mid-Western Regional Maternity unit were included, and gestational age reached was retrieved from labour ward records and added to the data set. Each patient was defined by variables including gestational age at delivery, age at diagnosis of bronchiolitis, and length of stay during RSV-admission. Those with negative RSV swabs or who were aged over one year at the beginning of vaccination season were excluded from the cohort, as were those with incomplete data. Information was compiled in a data file, cleaned, coded and analysed using SPSS version 14.0. Cost was calculated at per-bed-day rates on the premise that the babies admitted were subject to negligible cost of intervention.

Results: The final number in the cohort was 553. The total cost of RSV admissions during the period of the study was 2,469,901.44€, and these patients occupied 2489.82 bed-days over the 7 years and 4 month period. The cost of vaccinating under the current guidelines was found to be greater than the cost of their in patient stay. The cost of extending the guidelines to (a) all babies born at <35 weeks gestation and <1 year of age at the start of the RSV season or (b) all babies born at <37 weeks gestation did not prove to be cost-effective.

Conclusion: The economic impact of RSV prophylaxis has not, to date, been a deciding factor in the recommendations of its use. However, if economic 'net-loss' is taken as the baseline zero, a reasonable case could be made for widening the inclusion criteria.

THE CHANGING FACE OF PAEDIATRIC TRACHEOSTOMIES

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Aims: Tracheostomy placement remains a major issue. In 1972 only 30% of children receiving tracheostomy were under 1 year and the most common indication for tracheostomy in children was acute inflammatory airways obstruction (acute epiglottitis, croup and diphtheria). Current indications more commonly seen include ventilator dependence associated with congenital malformations affecting the lungs or airways and premature infants with chronic lung disease or tracheal stenosis due to prolonged ventilation. We reviewed the current trends in a tertiary paediatric hospital in Dublin.

Methods: A retrospective analysis was performed of all children who underwent a tracheostomy in a tertiary paediatric hospital in the study period January 2003–September 2007. Patients were identified using HIPE data; this was cross-referenced with airways specialist database and theatre records. We performed a retrospective review of clinical records, operative notes and laboratory data. Data collected included:

- i. Demographics iv. Elective or emergency procedure
- ii. Indications for tracheostomy placement v. Date of decannulation
- iii. Associated syndromes vi. Mortality

Results: During the study period (57 months) 26 children had a tracheostomy inserted. 24 cases were elective and 2 were emergency procedures.

The mean age of the study population on insertion of the tracheostomy was 13.5 months (0–169 months). 19/27 cases (73%) were <1 year old. The mean age of insertion for the <1 year group was 3.2 months old.

The indications for tracheostomy were divided in two groups:

- (i) Upper Airways Obstruction (UAO)
- (ii) Ventilator Dependence/Neurological

In 14/26 cases (53.8 %) the indication was UAO. In the <1 year infants, 11/19 cases (57.9%) had UAO as the indication for tracheostomy. In the over 1 year infants ventilator dependence was main indicator (4/7 cases 57.1%). Pierre Robin sequence was related to UAO indicated tracheostomy in 7/26 cases (26.9%). 5/7 cases (71.4%) of Pierre Robin sequence related cases were <1 year.

In 12/26 cases (46.2%) the indication was ventilator dependence/neurological. Exomphalus major was seen in 5/26 cases (19.2%) all were <1 year old on tracheostomy insertion with ventilator dependence being the indication for tracheostomy. A syndromic association was seen in 8/26 cases (30.7%). Central/Neuromuscular disorder was associated in 6/26 cases (23%). Decannulation was accomplished in 10/22 cases (45.4 %) in the study period.

4/26 patients died in the study period. 2/19 (10.5%) mortality rate <1 year old infants. 2/7 (28.5%) mortality rate over 1 year old group. 2 cases were central/ neuromuscular conditions, 1 aspiration and 1 irreversible pulmonary hypertension. No mortality was directed related to the tracheostomy complications.

Hospitalisation was often prolonged because of difficulties in the placement of patients back to local district hospitals or provision of local services. 19/26 patients had a local hospitals providing care. 6/13 patients living outside the Dublin area had step down hospital care.

Conclusions: An evolving and increasing indication for tracheostomy has been ventilator dependence. Changes in the epidemiology of infectious diseases and the capabilities of medical technology (vaccines, intensive care etc) have altered the indications for, and implications of, tracheostomy in children. A national plan for provision of further education and supports for parents and for local hospitals would aid in management of these patients.

A TWO YEAR RETROSPECTIVE SURVEY OF INTER-HOSPITAL TRANSPORT OF CRITICALLY ILL CHILDREN

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UCC

Background: Most paediatric tertiary care services are centralised in Ireland in accordance with universally accepted best practice. Many children each year are transferred to Dublin for critical and tertiary care from paediatric units outside Dublin.

Aims: To quantify and describe the cases sent from Cork University Hospital (CUH), identifying areas of potential improvement in services provided locally and centrally.

Methods: HIPE data gathered at CUH identified all paediatric admissions transferred to a tertiary care centre in 2005 and 2006. Chart review was undertaken using a piloted questionnaire. Data were analysed using SPSS.

Results: 70 out of 75 identified cases met the inclusion criteria. These were roughly equal in sex and had a mean age of 4.34 years. 67/70 (96%) went to OLHSC, Crumlin. Of these >50% required haematology/oncology, renal or gastroenterology services. 15/70 (21%) required intensive care services for transfer. 14 babies were eligible for transfer by the National Neonatal Transfer Service but only 3 were transferred by that service. One child died soon after arrival in Dublin and 1 child died much later of his index illness. Significant events during transfer were only documented in 3 cases. Most transfers, for which data were recorded, left after 15:00, meaning that they arrived in Dublin outside normal working hours. The time from decision to transfer to departure varied greatly due to many factors. Standards of documentation were very inconsistent.

Conclusions: Transport services for critically ill children are only provided ad hoc, consuming human and transport resources that are not geared to their immediate needs. National neonatal transfer services only assisted with <25% of suitable cases. A national transport service for all critically ill children is urgently needed in Ireland.

THE USE OF ETANERCEPT IN THE TREATMENT OF JUVENILE IDIOPATHIC ARTHRITIS (JIA): A PRELIMINARY REPORT OF THE IRISH EXPERIENCE TO DATE

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Introduction: Etanercept is a recombinant human soluble tumour necrosis factor alpha (TNF α) receptor fusion protein. It binds TNF and thereby blocks its interaction with the cell surface TNF receptors rendering TNF biologically inactive. JIA is an umbrella term for heterogeneous group of childhood inflammatory joint disorders that affects 1 in a 1,000 children <16 years. Etanercept has been licensed for the treatment of JIA since 2002.

Aim: To assess the use and tolerability of etanercept in children with juvenile arthritis attending our centre.

Method: This study was a retrospective chart review of all children treated with etanercept at our centre to date. Disease subtype (ILAR Classification, 2001), indication for usage, age of diagnosis, frequency of dosage and duration of usage were recorded. Concomitant therapy with methotrexate and any adverse reactions were noted.

Results: A total of 22 patients (16 female, 6 male) have been treated with etanercept since 2004, of whom 18 commenced therapy in the last 12 months. The median age at diagnosis and commencement of therapy was 5.5 years (range 0.25–13.10) and 10.25 years (range 3.77–18.33) respectively. Of the 22 patients, 14 were noted to have polyarticular JIA, 2 had systemic onset JIA, 3 had enthesitis related arthritis (ERA), 2 had extended oligoarticular JIA, and 1 had psoriatic JIA. Indications for the use of etanercept was recorded as lack of response to methotrexate in 21 patients and intolerance of methotrexate in 1 patient only. All patients received the twice weekly subcutaneous dosage of 0.4 mg/kg. Combination therapy with methotrexate was administered in 20 of the 22 patients. Etanercept was discontinued permanently in 6 patients. In 4 patients this was as a result of severe adverse reactions such as new onset uveitis (2), neutropenia (1) and low mood (1). Lack or inappropriate response

was documented in 2 children. Milder side effects (4) were also noted such as nausea and transient injection site reactions.

Conclusion: The use of biologic therapies in JIA is increasing dramatically, and this is reflected in this small study of our use of etanercept at our centre. This study is the first report to our knowledge of the use of etanercept in the Irish paediatric population. Adverse reactions were higher than expected with serious adverse reactions noted in 18% of the children.

'RECENT ADVANCES IN THE GENETICS OF CHILDHOOD ONSET EPILEPSY. OUR RESULTS OF SCREENING SO FAR'

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Background: Intractable early onset epilepsy is a devastating neurological condition with major effects on patients and their families. Recently mutations of the neuronal voltage-gated sodium channel alpha subunit type 1 gene (SCN1A) have been identified as a cause for severe myoclonic epilepsy of infancy (SMEI), generalized epilepsy with febrile seizures plus (GEFS+) and intractable childhood epilepsy with generalized seizures (ICEGTC). Following this discovery we commenced screening selected patients looking for SCN1A gene mutations.

Aims: To review results of genetic analysis for SCN1A mutations on samples sent since the commencement of screening on patients with a phenotype suggestive of SMEI/GEFS+/ICEGTC.

Methods: Patients fulfilling the clinical and EEG criteria for SMEI/GEFS+/ICEGTC were tested for SCN1A mutation. Case notes review looking at specific areas of clinical history, was carried out for all patients positive for gene mutation.

Results:

- Twenty samples were sent over an eight month period.
- Results on twelve samples have returned. Further eight results currently pending.
- Six results are positive for SCN1A gene mutation (50%).
- Of the six positive patients average age of first seizure was eight months (range 5–15 months).
- Four patients have a clinical diagnosis of SMEI and two GEFS+ with characteristic seizure type's e.g. febrile convulsion, hemiconic and polymorphic seizures.
- Five of the patients have poor seizure control despite trial of many anti-epileptic drugs (AED) (range 4–10 medications).
- Four patients have significant developmental delay and behavioural issues.
- All six have had abnormal EEG recordings characteristic with syndrome diagnosis.
- No patient has a history of abnormal MRI brain scans or any underlying structural brain abnormality that could contribute to their seizures.
- All patients have normal chromosomes.
- Both patients with a diagnosis of GEFS+ have a parent with a history of epilepsy since childhood and one affected father has been confirmed as positive for SCN1A mutation.

Conclusions:

Detection of SCN1A mutations is a significant advance in the genetics of epilepsy.

- For families it may offer a clearer cut diagnosis and also have genetic implications in the familial forms of epilepsy.
- Mutation identification can help Taylor AED choice and treatment as Lamotrigine may be contraindicated while Stiripentol and vagal nerve stimulation effective.
- It may be implicated in cases of seizure disorders previously attributed to vaccine damage.

THE IMPACT OF THE ORGAN RETENTION CONTROVERSY ON POST MORTEM RATES IN THE CHILDRENS UNIVERSITY HOSPITAL: 1997–2004

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Background: There has been a decline in the rate of post-mortem (PM) examinations worldwide. Although consent for post-mortem is more likely to be given for fetal and paediatric deaths, there has been a steady decline in paediatric post-mortems in Ireland, which accelerated even further in the wake of the 'Organ Retention controversy'. The Dunne Post Mortem Inquiry was established in March 2001 and was superseded by the Madden Report in 2005. Their purpose was to review the post mortem examination policy, practice and procedures with particular reference to the issues of organ removal, retention, storage and disposal in the State since 1970. It was hoped an inquiry would help restore public confidence in hospitals, emphasise the necessity of post-mortems, provide recommendations on best autopsy practise in Ireland and reassure the public that procedures surrounding autopsy practise would be open and transparent.

Aim: To determine whether the Organ Retention controversy impacted upon post mortem rates in CUH.

Method: CUH policy on post mortem consents was reviewed. Data was collected on all deaths in CUH during the years 1997–2004. Information was gathered on whether PMs were coroner or non-coroner cases and whether consent for a limited or a full PM was obtained in hospital cases.

Results: Over the eight year period from 1997 to 2004 there were a total of 345 deaths in CUH.

On average there were 43 deaths per year. The highest number of deaths occurred in 1999 and 2001 (51 cases), with the lowest number of deaths in 2003 (30 cases). Each year the percentage of coroner's cases averaged 52%. The highest rate was in 1997 (63.6%), the lowest in 2003 (40%). Prior to the outbreak of the 'Organ retention controversy' in 1999, the hospital post-mortem rate for non-coroner cases in 1997 and 1998 was 37.5 and 42.85% respectively. In 1999 this figure dropped to 27%. As the crisis escalated to new height in 2000 the in-hospital (house) post-mortem rate dramatically plummeted to 4.5%. Since 2000 this figure has slowly risen, averaging 14.86%, during the years 2001–2004. The highest rate was in 2002 when the figure rose to 18.18%. This is in stark contrast to the pre-controversy era. The rate in 2004 is still only a third of the figure for 1998. In parallel with the drop in full house post-mortem figures, there has been a rise in the rate of limited post-mortems being performed. Between the years 1997–2000, only one limited PM was performed in 1998. The ratio of full house PM to limited PM was 6:1. In 2000, this ratio was reversed with 4 limited PMs performed and only 1 full PM. This trend continued in 2001. In 2004 the limited PM rates was still higher than the

'pre controversy era' but the ratio of full house PM to limited PM is now 3:2.

Conclusion: The Organ Retention controversy had a negative impact on PM rates in CUH. There are an increasing number of limited PMs being performed, as many parents will not consent to full postmortem examinations. The declining rate in postmortem rates is a cause for concern. Undoubtedly they are of proven benefit to the families and to medical education, research and audit. Faith in the medical profession has been severely damaged and rebuilding that trust is crucial in order to advance medical knowledge on why children are dying.

CIRCUMSTANCES SURROUNDING WITHDRAWAL AND LIMITATION OF LIFE SUPPORT IN A PAEDIATRIC INTENSIVE CARE UNIT

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Background: Withdrawal and limitation of life support in a Paediatric Intensive Care Unit (PICU) is a fundamental aspect of clinical practice and yet remains an emotive and highly challenging issue. During the past decade 40–60% of deaths in PICUs follow withdrawal and limitation of life support¹. Few studies report how these decisions were reached.

Aim: To assess the frequency of end-of-life decisions preceding child death and the characteristics of the decision making process in an Irish PICU.

Methods: Retrospective analysis of all children (<16 years) dying in CUH PICU from September 2006 to September 2007. Diagnosis and end-of-life data were determined from the medical and nursing records. Patients who were transferred and died outside of the unit were excluded.

Results: There were 391 admissions and 24 deaths during the study period; overall mortality rate was 6%. Of the 24 deaths, 14 (58%) were preceded by an end-of-life decision. Of these; 9 deaths followed active withdrawal of life support (cessation of mechanical ventilation and removal of endotracheal tube), 1 followed limitation of life sustaining treatment (LST), 1 was following a "do not resuscitate" (DNR) order, 3 were brain dead. End-of-life decisions were made when it was a "no chance situation" in 7/14 deaths, a "no purpose situation" in 4/14 deaths and 3/14 were clinically brain dead. Admission diagnoses were classified as acute illness unrelated to any chronic disease; 7/14 cases, chronic illness including known complications of; 6/14, uncertain diagnosis (at the time of death); 1 case. Consultant Paediatrician and/or Intensivist initiated discussions about forgoing LST in 13 cases; in 1 case the parents requested palliative extubation. Consensus between families and staff about forgoing LST as the best approach was reached after the first meeting with 7/14 of families and 7/14 required ≥ 2 meetings. In the 1 DNR death, the time to death after consensus was 1 day and for withdrawal/limitation of LST was a mean of 11 min (2–1,170 min) however was not documented in 2 cases. The family were present in 10/14 cases when LST was forgone, not present in 1 case and not documented in 3 others. The dying patient was held by the family in 9 of these occasions.

Conclusion: In the CUH PICU, end-of-life decisions preceded child death in approximately 60% of cases. In every case, the decision making process involved the multidisciplinary caregivers including the Consultant Paediatrician and Intensivist and the parents (natural and foster in 1 case). Consensus between families and staff about forgoing LST as the best approach was reached in all cases. The majority of children died soon after the end of life decision was made.

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BIOCHEMICAL MARKERS MAY IDENTIFY PRETERM INFANTS WITH A PDA AT HIGH RISK OF DEATH OR SEVERE IVH

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Background: The presence of a patent ductus arteriosus (PDA) in preterm infants is associated with increased risk of intraventricular haemorrhage (IVH), and death. It is difficult to identify PDAs that may lead to potential complications using echocardiography alone. Elevated levels of cardiac troponin T (cTnT) and N-terminal-pro-B type Natriuretic Peptide (NTpBNP) predict outcome in adults with cardiac disease and stroke.

Aims: To evaluate the association between NTpBNP and cTnT levels and death or severe IVH in preterm infants.

Methods: Infants <1,500 g underwent simultaneous echocardiographic and cTnT/NTpBNP assessments at 12 and 48 h of life. They were divided into 3 groups, those with a closed PDA at 48 h, infants with a PDA and IVH III/IV (severe IVH) and/or death, and infants with a PDA and no severe IVH or death. cTnT and NTpBNP levels at 48 h were compared using ANOVA test.

Results: A total of 80 infants with a median gestation of 28 weeks [IQR 26.1–29.5] and median birth weight 1.06 kg [IQR 0.87–1.21] were included. At 48 h, infants with a PDA and poor outcome had significantly higher median NTpBNP and cTnT levels compared to infants with a PDA and good outcome and those with spontaneous PDA closure: (NTpBNP: 9282, 5121, and 740 pmol/L respectively, $p = 0.008$, and cTnT: 0.66, 0.25, and 0.13 $\mu\text{g/L}$ respectively, $p = 0.027$). Infants with a PDA had significantly higher left ventricular contractility (mVcfs: 1.21 vs. 1.05 circ/s, $p = 0.04$), left ventricular output (240 vs. 172 ml/kg/min, $p < 0.001$) and significantly lower celiac artery blood flow (23 vs. 31 ml/kg/min) There were no significant differences in echocardiographic parameters of PDA size, systemic blood flow or myocardial contractility between the good and poor outcome PDA groups.

Conclusions: cTnT and NTpBNP may be superior to echocardiography in predicting the occurrence of severe IVH and death in preterm infants. The use of biochemical markers such as NTpBNP and cTnT in conjunction with echocardiography may provide a basis for targeted treatment in infants with a PDA.

IDENTIFYING AND TREATING MUDA (ACTIVITIES WITHOUT VALUE) IN NEONATAL INTENSIVE CARE UNIT, CORK UNIVERSITY MATERNITY HOSPITAL

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Introduction: One way of increasing work efficiency is by eliminating activities that are not productive or do not add value. MUDA is the Japanese term used to describe “activities without value”. Opening a new maternity hospital with new workers in a completely new environment is a major management project with potential for many MUDAs even in processing simple and uncomplicated processes.

Objective: To identify MUDAs throughout the new Hospital, with a special focus on NICU, that might lead to Health Care personnel working more efficient and with greater satisfaction.

Method: A cross sectional, randomly chosen sample of CUMH staff, were approached. The term MUDA was explained to them. They were asked to identify three MUDAs along with possible practical solutions. The data were analysed qualitatively using thematic content approach.

Results: 87 staff agreed to participate, resulting in a total of 261 MUDA. Double work MUDA (e.g. rewriting patient information in more than one place) along with waiting MUDAs (e.g. waiting for ward round to take place) were common among nurses. While doctors and allied medical staff were more likely to complain of MUDAs involving unnecessary steps in a process (e.g. searching for tools to complete a certain task).

	Double work (%)	Unnecessary steps (%)	Waiting (%)	Others (%)
Doctors ($n = 17$)	20	67	11	1
Nurses ($n = 61$)	39	30	26	5
Allied staff ($n = 9$)	13	78	1	5

Conclusion: Simple processes can become prolonged and inefficient in a larger, complex, and completely new environment. Despite the recognition of MUDAs in their work practice, few practical solutions were offered by the staff. Recognition of MUDAs in the work environment is the first step toward increase productivity and satisfaction.

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COOL IT!!!! WHO IS EXCLUDED FROM HYPOTHERMIA TRIALS FOR NEONATAL ENCEPHALOPATHY?

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Introduction: Pooled analysis of 8 RCTs ($n = 638$) shows that hypothermia for hypoxic ischaemic encephalopathy reduces the combined outcome of death or major disability and the individual outcomes of mortality and neurodevelopmental disability in survivors (1). Hypothermia is now proposed as standard management of neonatal encephalopathy with future infants included on prospective clinical registers.

Aims: To evaluate the number of infants with neonatal encephalopathy admitted to a large Irish perinatal centre who would have met the inclusion criteria for large RCTs of cooling and define the excluded population.

Methods: Retrospective chart review using the Holles St neonatal encephalopathy database from Jan 2005 to Sept 2007. Each case was assessed using the individual trial criteria (1) and reasons for inclusion and/or exclusion recorded.

Results: 71 term infants had moderate or severe neonatal encephalopathy excluding major congenital abnormalities during the study period. Only 15 were eligible for individual hypothermia trials and 4 were excluded for the following reasons: 2 outborn and admitted >6 h of age, 1 predicted poor outcome, 1 chorioamnionitis. Fifty-six were ineligible due to delayed onset of abnormal neurological signs, transfer from peripheral hospital >6 h or insufficient clinical data. Therefore 11 (15.4%) infants would have been randomised pending parental consent. If hypothermia becomes standard management for neonatal encephalopathy and the ICE inclusion and exclusion criteria alone were used, 10 infants were eligible and 3 would have been excluded. Therefore only 7 (10%) infants from the NMH cohort would be included in a cooling registry. Our findings contrast with an increased proportion of screened infants meeting inclusion criteria in the large RCTs. Shankaran et al. screened 798 infants and although 239 were eligible only 208 were enrolled (26%). Gluckman et al. reviewed 541 infants and 234 were enrolled (43%).

Conclusion: Hypothermia appears to be beneficial for the management of neonatal encephalopathy. However significant numbers of our infants did not meet the inclusion criteria of the RCTs thus reducing the applicability of the trials and meta-analysis.

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USE OF EXPRESSED BREAST MILK AND FORMULA IN VERY PRETERM AND VERY LOW BIRTH WEIGHT INFANTS

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Background: Very preterm (<32 weeks' gestation) and very low birth weight (VLBW, <1,500 g) infants are at increased risk of

necrotizing enterocolitis (NEC), an uncommon condition which is potentially fatal and associated with significant long-term morbidity. Though many strategies have been advocated to avoid NEC, the only intervention consistently associated with reduction in the risk of developing NEC is feeding with expressed breast milk (EBM). We wished to determine how many infants <32 weeks or <1,500 g born at our hospital received EBM and what proportion of their feeds it constituted.

Methods: We conducted a prospective observational cohort study of all infants <32 weeks or <1,500 g admitted to the neonatal unit at the National Maternity Hospital between 1 January 2007 and 31 June 2007. We determined from the bedside nursing observation charts whether infants were fed and what type of feed they received on each day of their nursery stay.

Results: We identified 55 very preterm or VLBW infants admitted during the study period. One infant died before feeding was commenced; another died having received <2 mL of formula. Twelve (22%) infants received EBM exclusively during their nursery stay; 18 (33%) infants were exclusively formula fed and 24 (44%) were fed a combination of EBM and formula. EBM constituted a mean of 59% of the total feeds received by infants who were fed a combination of EBM and formula.

	Feed on first day of feeding	Feed on discharge from nursery
EBM	27 (49%)	20 (36%)
Formula	23 (42%)	29 (53%)
Both	4 (7%)	4 (7%)

Ten of the 27 infants who were commenced on EBM changed to formula during their admission and were subsequently discharged on formula. The mean day of life these infants switched from EBM to formula was day 24. Three of the 23 infants who were commenced on formula were discharged home on EBM. The mean day of life on which infants changed from formula to EBM was day 4. Eight of the 24 infants who received a combination of EBM and formula in the nursery were discharged home receiving EBM exclusively.

Conclusion: Many infants at increased risk of NEC do not start on EBM in our unit and fewer than half are still receiving breast milk at discharge.

THE CONTINUING PROBLEM OF NEONATAL ABSTINENCE SYNDROME

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Background: Neonatal abstinence syndrome (NAS) is the term used to describe the symptoms and signs of drug withdrawal experienced by infants of mothers with drug dependencies. Infants often require admission to the neonatal unit for the management of their symptoms, especially with the increasing maternal doses of methadone being used antenatally. Duration of treatment and admission to the unit can be extensive.

Aim: The aim of this study was to review infants with NAS who were admitted to our neonatal unit during a one year period, in particular to look at the treatment required, duration of treatment and length of stay.

Method: A search of the hospital computer database was performed to obtain a list of infants with a diagnosis of NAS. Those admitted to the neonatal unit between the 30 June 2006 and 30 June 2007 were included. The medical chart of each infant was reviewed and appropriate data collected.

Results: 29 infants with NAS were admitted to the neonatal unit. The average maternal age was 28.2 years (range 21–42). 17 mothers (60.7%) were Hepatitis C positive. All mothers were cigarette smokers. One infant was excluded from the study (chart unavailable). Of the 28 infants included, 4 infants (14.3%) were born preterm and 19 (67.9%) had a birth weight on or below the 10th percentile. Two infants did not require any pharmacological treatment. Of the remaining 26 infants, 14 (53.8%) required phenobarbitone alone, 10 (38.5%) oromorph and phenobarbitone and 2 infants (7.7%) phenobarbitone together with oromorph and diazepam. The average duration of treatment was 43.5 days of phenobarbitone, 21 days of oromorph and 25 days of diazepam. The average duration of stay on the unit was 21.9 days. Over half (53.6%) did not attend for their four month developmental review and of the 13 who did attend, 6 (46.2%) had abnormal development.

Conclusion: Neonatal Abstinence Syndrome remains a significant cause of neonatal morbidity. It is associated with prolonged hospitalisation and the need for poly drug use to control symptoms. Further work is needed to encourage attendance at follow up clinics.

ASSESSING THE NEED FOR POST-DISCHARGE HAEMOGLOBIN MONITORING IN PRETERM INFANTS

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Aim: Premature babies are more susceptible to anaemia. It has been the practice in NMH to routinely perform a haemoglobin 2 weeks post-discharge on all infants ≤ 32 weeks' gestation. The aim of this study is to audit the usefulness of this practice and, by doing so, to attempt to define a sub-group of infants who may benefit most from this surveillance.

Methods: All infants ≤ 32 weeks gestation admitted to the NICU between January 2006 and June 2007 who were subsequently discharged home with a plan to follow up in NMH were identified. Computerised clinic records were then examined to determine the number of infants who were booked to have a routine haemoglobin performed. The number of tests done, in addition to the results and the subsequent need for transfusion was recorded. Factors which could potentially increase the likelihood of transfusion, such as gestational age and pre-discharge haemoglobin, were also noted.

Results: A total of 55 babies of ≤ 32 weeks gestation were discharged home with planned follow up in NMH during the time period specified. Of these, 30 (55%) were booked for routine haemoglobins. 5 (36%) of the 14 infants <28 weeks gestation were tested, 11 (68%) of the 16 infants of 28–29 weeks gestation were tested and 14 (58%) of the 24 infants of 30–32 weeks gestation were tested. Most infants underwent serial testing and a total of 80 haemoglobins were performed on these 30 patients, an average of 2.7 blood tests per patient. Only 2 patients were re-admitted to the NICU for a blood transfusion as both infants had a haemoglobin of <8 g/dL. No infant with a pre-discharge haemoglobin >9 g/dL required readmission for a blood transfusion irrespective of gestational age or day of discharge. In addition, no infant with a pre-discharge haemoglobin of <9 g/dL but with a reticulocyte count of >5% required readmission for a blood transfusion.

Conclusions: Our study suggested that routine haemoglobin testing in preterm infants ≤ 32 weeks gestation is not indicated. The only infants that may benefit from a follow up haemoglobin post-discharge are those in whom the predischARGE haemoglobin is < 9 g/dL and the reticulocyte count is $< 5\%$.

CEREBELLAR HAEMORRHAGE IN EXTREMELY PRETERM INFANTS IS AN OMINOUS FINDING

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Abstract: The importance of cerebellar haemorrhage as an under-diagnosed and potentially life-threatening complication of preterm birth has been emphasized in current literature. Mata M et al. first described the improved technique for sonographic imaging of the cerebellum through the mastoid fontanelle in 1996. However this practice is not carried out universally. The high morbidity associated with CBH has been highlighted in a recent article by Limperopoulos et al in *Pediatrics*. In particular they conclude that cerebellar injury may also contribute to long term non-motor sequelae such as cognitive, learning and behavioral disability in survivors. We have reviewed a recent series of ELBW infants with CBH.

Methods: CBH were identified by viewing the posterior fossa through the mastoid fontanelle in ELBW infants undergoing routine cranial ultrasound scanning. The presence and the extent of CBH were documented.

Results. In the past 12 months there were six documented cases of CBH. The clinical details of these cases are described in Table 1 below.

Table 1. Clinical and ultrasound findings in ELBW infants with CBH

Cases	Gestation outcome/ age (weeks)	BW (g)	Sex	Extent CBH (h)	Timing	Seizures IVH	Associated	Hydrocephalus
1	23 + 6 Death 54	610	M	Extensive	72	Yes	Grade 3	No
2	24 + 0 Death 3	520	M	Extensive	48	Yes	Grade 4	Yes
3	24 + 0 Death 3	800	M	Extensive	48	Yes	Grade 4	Yes
4	25 + 0 Death 36	860	M	Extensive	72	Yes	Grade 3	No
5	24 + 2 Death 12	680	M	Extensive	192	Yes	Grade 4	Yes
6	25 + 4 Death 28	650	M	Extensive	72	Yes	Grade 3	Yes

The mean gestational age was 24 + 3 weeks (23 + 6 to 25 + 4). The mean birth weight was 687 g (520–860 g).

All of the cases were male and all had extensive CBH on US imaging. The majority (5/6) cases of CBH occurred early in the first 72 h of life and all had associated severe IVH. Seizures uniformly occurred and hydrocephalus developed in $\frac{3}{4}$ cases. All infants died.

Discussion: CBH is clearly a major complication of CBH for preterm ELBW infants. When it occurs it is extensive in nature. It develops

early usually in the first 72 h of life and appears to be invariably associated with IVH. Clinical manifestations, in particular seizures are common. It appears to be a condition confined to extreme prematurity and ELBW and there seems to be a male predominance. The outcome is devastating as all of the infants in our series died. There is no clear explanation as to the pathogenesis of these injuries. The cerebellum's blood supply is provided by branches from the vertebral basilar system. Perhaps CBH results from reperfusion injuries as is proposed in IVH as the timing of these insults is similar; however this is unproven.

Conclusion: CBH in extremely preterm infants is a worrying sign and is likely to be a marker for very poor prognosis. Its presence is a further useful finding when counseling parents in the NICU. Improved visualization of CBH in the posterior fossa has only become feasible in recent years due to better US resolution and higher frequency probes. We recommend mastoid views as part of the routine cranial US in all preterm infants < 32 weeks gestation to be carried out during the first 72 h of life.

AUDIT OF THE POST-DISCHARGE MANAGEMENT OF DCT POSITIVE INFANTS

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Aim: Infants who are DCT positive are frequently subjected to weekly haemoglobin testing post-discharge from hospital. The aim of this study was to audit our practice in NMH with a view to making a recommendation regarding the most appropriate outpatient management strategy.

Methods: All infants ≥ 35 weeks gestational age born in the National Maternity Hospital between January 2005 and June 2007 who were DCT positive were identified. The total number of blood tests performed post-discharge in addition to the number of transfusions administered post-discharge was recorded. Factors predicting those infants most likely to need transfusion after discharge were also determined.

Results: 125 babies ≥ 35 weeks who were DCT positive were identified. A total of 515 haemoglobins post-discharge was performed, an average of 4.5 blood tests per baby. Of these, only 4 resulted in a blood transfusion.

The DCT was positive due to Rhesus antibodies in 83 cases (66%), ABO incompatibility in 38 cases (30%), and other antibodies associated with haemolytic disease of the newborn in 5 cases (4%).

3 of the 4 infants who needed a post-discharge transfusion were affected by Rhesus disease. All of these infants had received either an in utero transfusion or a simple transfusion prior to discharge. Infants were transfused if the haemoglobin level fell below 8 g/dL. The other baby to receive a post-discharge transfusion was a non-identical twin of 37 weeks GA with ABO incompatibility who only required 3 days of phototherapy (maximum SBR of 220 $\mu\text{mol/L}$) and was transfused at 6 weeks of age for a haemoglobin of < 8 g/dL. The impact of gestational age, level of antibody titre detected, timing of sensitization and predischARGE haemoglobin on the need for post-discharge monitoring is currently being assessed.

Conclusions: These results clearly highlight the fact that far too many blood tests are routinely performed on DCT positive infants which has implications both in terms of time, the appropriate use of resources and impact on infants and families. Babies with Rhesus disease who have had in utero or pre-discharge blood transfusion clearly benefit from surveillance. Equally, it may be appropriate to screen babies of 37 weeks gestational age or under. For all other

infants, routine testing does not appear to be indicated and clinical assessment should suffice.

ANTIFUNGAL PROPHYLAXIS FOR THE PREVENTION OF NEONATAL CANDIDIASIS

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Background: Randomised controlled trials suggest prophylactic administration of antifungal agents reduce the rate of colonisation and invasive *Candida* infection in a subgroup of high-risk very low birth weight neonates. The extent of antifungal prophylaxis use in the United Kingdom and Ireland is unknown.

Methods: A postal questionnaire was administered in February 2007 to neonatologists practicing in UK and Ireland caring for very low birth weight infants. Information was requested on the prophylactic agents used, dosing schedules and duration of therapy. The rationale for reported practices was also ascertained.

Results: The response rate was 55% (125/228). Antifungal prophylaxis use was reported by 66 (53%) respondents. First line agents utilised included oral nystatin (53%) and intravenous fluconazole (41%). Marked variations in dosing schedules were reported. The most frequent indications for antifungal prophylaxis included antibiotic administration 45 (68%), decreased birth weight 33 (50%) and central vascular access 28 (42%). The majority of respondents who did not use antifungal prophylaxis felt that the rate of invasive fungal disease within their unit was not high enough to justify its use (Median Likert score 4 vs. 5 $p = 0.001$). Concerns regarding the need for further efficacy studies and potential development of antifungal resistance were also prevalent.

Conclusions: Many clinicians caring for very low birth weight neonates do not routinely use prophylaxis. This reflects the wide variation in the incidence of invasive disease, lack of conclusive evidence supporting a role for antifungal prophylaxis and concerns related to emergence of resistant strains.

A COMPARISON OF INVASIVE AND NON-INVASIVE ARTERIAL BLOOD PRESSURE MEASUREMENTS IN THE NEONATE

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Background: Blood pressure monitoring is an essential part of neonatal intensive care. The two different methods used are (a) invasive through an indwelling arterial catheter and (b) the non-invasive oscillometric method using a blood pressure cuff.

Aims: To compare the results of invasive (Marquette and Dash monitors) and non-invasive oscillometric (Procare, Marquette and Dash) blood pressure recordings in sick newborn infants.

Methods: Neonates who had indwelling arterial catheters, admitted to NICU between August and December 2006 were enrolled in the

study. All participants had an arterial cannula in either their umbilical, radial or posterior tibial arteries. The child's blood pressure was recorded at admission and subsequently at six hourly intervals for the first 72 h of life or until the indwelling catheter was no longer required. Blood pressure was recorded simultaneously using an invasive (Marquette or Dash) and two different non invasive (Marquette and Procare, or Dash and Procare) monitors. The two non-invasive readings were taken simultaneously, one from the upper limb and the second from the lower limb. The two different monitors were then alternated for the following reading and this alternating fashion continued for subsequent recordings. All readings were taken while the child was settled and quiet.

Results were analysed using excel statistics package to construct a Bland Altman plot.

The study was approved by the local ethics committee. Informed consent was obtained from parents prior to enrolment.

Results: 25 babies were enrolled in the study. Birth weight and gestation ranged from 560 to 4,500 g and 24 + 1 to 40 + 5 respectively. 166 matched invasive and non-invasive recording were obtained.

All three non-invasive monitors tended to over read blood pressure. The average differences in mean BP being—Dash 2.4 mm Hg, Procare 4.5 mm Hg and Marquette 8.4 mm Hg. In all three non invasive monitors the higher the mean blood pressure the larger the difference between the invasive and non invasive recordings. There was no significant difference between the non invasive recordings taken from the upper or lower limbs.

Conclusion: In sick infants who are at risk of hypotension, invasive monitoring remains the gold standard for measuring blood pressure.

QUANTIFYING PRETERM BRAIN MATURATION USING THE AMPLITUDE INTEGRATED EEG

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Background and aim: The preterm infants brain undergoes rapid development in the early weeks after birth. The nature and the rate of this maturational process has been difficult to quantify. In this study we attempted to document the rate of neurological change using electroencephalographic measurements in a cohort of very preterm infants.

Methods: 30 infants ranging from 24 to 32 weeks gestation were examined in this study. Each of the infants had an amplitude integrated EEG carried out in the first week of life. The tool employed was the Olympic 6000 CFM, software 2.0. This is a one channel aEEG monitor. Numerical data was exported from the CFM in the form of csv files and entered into Labview, chart analyser software. This is a one channel aEEG monitor. Infants included had no intracranial pathology seen on ultrasound. At the time of aEEG recording none of the patients received sedation and all were clinically stable.

Results: Table 1: Infants were subdivided into 24–27 weeks and 28–31 weeks gestational age. Labview was used to analyse the parameters that appeared to be most associated with gestational age. These were, median amplitude and %activity >20 μ V. In Table 2 the group were further subdivided into 24–26, 27–29 and 30–32 weeks.

Discussion: The aim of this study was to document maturational processes in a cohort of preterm infants in their first week of life using

Table 1 aEEG parameters (median amplitude and % activity >20 μ V) recorded for a group of infants at 24–27 and 28–31 weeks gestation

Parameters	Gestational age	
	24–27 weeks (<i>n</i> = 14)	28–31 weeks (<i>n</i> = 16)
Amplitude (median) μ V		
Mean	9.7	12.8
S.D.	2.1	1.7
%Activity >20 μ V		
Mean	18.2	25.5
S.D.	6.5	7.7

Table 2 aEEG parameters (median amplitude, % activity >10 and >20 μ V) recorded in infants at 24–26, 27–29 and 28–32 weeks gestation

Parameters	Gestational age		
	24–26 weeks (<i>n</i> = 10)	27–29 weeks (<i>n</i> = 10)	30–32 weeks (<i>n</i> = 10)
Amplitude (median) μ V			
Mean	9.6	12.2	12.4
S.D.	2.1	2.7	1.7
%Activity >10 μ V			
Mean	45.8	58.5	61.8
S.D.	11.2	12.1	7.7
%Activity >20 μ V			
Mean	18.3	24.9	23.5
S.D.	8.5	9.8	6.9

aEEG. Our findings show clear electroencephalographic changes between 24 and 32 weeks gestation both for median amplitude and %activity >20 μ V. Both parameters increased with gestational age. This indicates rapid maturation occurring in the early weeks after birth. When the data is subanalysed the maturational changes are more evident between 24–26 weeks and 27–29 than they are between 27–29 and 30–32 weeks. Although this is preliminary data, it does suggest that maturation may be stepwise rather than linear. Findings in this study provide further insight into the maturation process in the brains of the preterm infant. In addition it examines a quantitative method of analysis of the preterm aEEG. By setting down normal reference values for aEEGs in preterm infant this may allow easier interpretation for the neonatologist. In addition it may identify infants that deviate from normal patterns as those at most neurological risk

THE ROLE OF THE ADVANCED NEONATAL NURSE PRACTITIONER (ANNP) IN NEONATOLOGY

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Aim: Advanced Nurse/Midwives Practitioners roles were first introduced in Ireland in the early 1990s to provide holistic,

autonomous and timely care for patients (NCNM 2005). The goal of advanced practice within neonatal nursing is to develop a comprehensive, seamless structure of providing care, which can meet the requirements of the neonatal population. This can be achieved through innovation and the provision of a patient-centered high quality service. The main aim of the ANNP role is provide expert care to the high risk neonate and improve continuity of care. The first ANNPs were accredited in 2005. However, while the role of the ANNP is well established internationally, the aim of this study was to explore the ANNPs' experiences of their role within the context of Irish healthcare.

Methods: A qualitative design was used for this study as the numbers were small and little published literature exists in this area. In keeping with the tenets of the methodology purposive sampling was used and the authors conducted four in-depth interviews with experienced ANNPs employed in three maternity teaching hospitals in Ireland. Data collection involved tape-recorded semi-structured individual interviews utilising open-ended questions. Thematic analysis was the chosen method for analysing data generated from the study.

Results: Three main themes emerged from the data. Firstly, essence of the role conceptualised the idea of holism and the consistency of care which is achieved from a total quality approach to neonatal care. Incorporated into this was issues regarding autonomy, expert practice, job satisfaction, accountability, advanced decision making and diversity. Secondly, being a professional and clinical leader focused on leadership, teamwork, and support networks, being visionary, developing standards and bridging the gap between medicine & nursing. Thirdly, challenges of their role identified included balancing care with other demands that the role entailed, time management, organisational change, increasing ANNP workforce and future planning. ANNPs regarded the holistic nature of their role as being pivotal to their existence. The high level of job satisfaction expressed by the participants is related to direct comprehensive patient care which they provide within a holistic family-centred care framework. The findings showed that the essence of their role was in the consistency of their care that they were able to provide, being in a position to provide professional and clinical leadership to colleagues and other disciplines. Time constraints impede greatly on ANNPs fulfilling all the requirements of their job description. The participants described that they have experienced acceptance of their role. However, there are some areas where this remains problematic which can impede their ability to provide comprehensive care, such as radiography and prescriptive authority.

Discussion: The findings of this study highlight the commitment and expertise provided by ANNPs and provides new insights into the experiences of the ANNP within this role. Themes which emerged from this study were the uniqueness and inherent strength of the ANNP role which lies in their ability to command many skills incorporating a vast knowledge base. Considering their diversity of their experiences, the study concludes that the ANNP experience of their role is rewarding and all participants were positive and optimistic about the development of the role and its future. It is anticipated that the role will grow in numbers and address complex and dynamic healthcare system needs that reflects the demands of our current society today. This can be achieved by promoting collaborative practice and teambuilding, developing individualized, holistic, culturally sensitive management strategies which are cost effective and meet the needs of the neonate and the family. If the development of this role continues, promotion of collaborative practice, improved teambuilding, development of individualized, holistic, cost-effective family-centred care will prevail. Furthermore, initiating, leading, implementation and evaluation of potentially better practices, increased research utilization and increased access to education for staff and parents will also be enhanced.

FEEDING VERY PRETERM AND VERY LOW BIRTH WEIGHT INFANTS

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Background “The management of acute neonatal illnesses and gradual advancement of feeding to minimize the risk of feeding-related complications, such as necrotizing enterocolitis, may conflict with the nutritional goal of obtaining rapid growth in preterm infants” (AAP 1998).

Aim: To describe the current feeding practice amongst very preterm (<32 weeks) and very low birth weight (VLBW, <1,500 g) infants in our unit. Specifically we wished to determine:

1. When enteral feeds were commenced
2. The time taken to reach feeds of 150mls/kg/day; and
3. Why feeds are held during their time in the nursery.

Methods Prospective observational study of all infants < 32 weeks or < 1500 g admitted to the intensive or special care nurseries of the National Maternity Hospital (NMH) between 1 January and 31 July 2007. For each day of the nursery stay, we determined from the bedside nursing records:

- Whether infants received enteral feeds
- If not, the reasons why feeds were withheld
- The volume of feeds given in millilitres per kilogram bodyweight (mL/kg)

Data collection continued until the infant was discharged from the nursery or full breast feeding was established.

Results Fifty-five neonates were enrolled. Enteral feeding was started at a mean (SD, range) of 3.6 days (2.6, 0–13). Feed volume of

Table 1 Reasons for feeds not being commenced in premature neonates

Number of infants (%)	Mean number of days	Reason identified
20 (36)	1.6	Prematurity
12 (22)	1.6	Respiratory distress
10 (18)	1.0	No EBM available
10 (18)	2.4	None identified
8 (15)	3.0	Suspected Sepsis
9 (17)	1.9	Antenatal factors
7 (12)	2.6	Medications
4 (7)	2.5	Aspirates/Suspected NEC/NEC

Table 2 Reasons for feeds subsequently held following initial feeding of neonate

Number of episodes (%)	Reason identified
4 (30.7)	Suspected sepsis
3 (23.1)	Unidentified
2 (15.4)	Abdominal distension
1 (7.7)	Radiological NEC
1 (7.7)	No EBM available
1 (7.7)	Inotropes
1 (7.7)	Nitric oxide

150 mls/kg was reached at 13.8 (8.2, 5–44) days. The reasons for not starting feeds are shown in Table 1. Twelve (22%) infants subsequently had feeds held for a mean (range) of 3.7 (1–28) days. The reasons for feeds subsequently held are shown in Table 2.

Conclusion In preterm infants admitted to our unit, enteral feeding is frequently delayed; once started, it is frequently stopped; and it takes about 2 weeks for infants to receive a reasonable volume of enteral feeds.

PATENT DUCTUS ARTERIOSUS (PDA) LIGATION IN VERY LOW BIRTH WEIGHT INFANTS IN IRELAND

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Introduction: Patent Ductus Arteriosus (PDA) is a significant cause of morbidity and mortality in premature neonates, resulting in congestive heart failure, prolonged ventilator dependency, and an increased incidence of chronic lung disease (CLD). PDA ligation in premature infants has been shown to have low surgical morbidity. However there is substantial late mortality and a high incidence of morbidity in the survivors, especially if ligation performed >3 weeks of age. Recently, PDA ligation has been associated with poorer neurosensory outcomes compared with medical treatment alone. In addition the systemic inflammatory response associated with surgery has been postulated to contribute to poor neurodevelopmental outcomes.

Methods: All infants who had a PDA ligation in Ireland were identified using HIPE data excluding infants with other congenital abnormalities.

Results: 93 infants <1,500 g had a PDA ligation in Ireland in a 4-year period. The median birth weight was 847 [interquartile range (IQR): 720–1,050]g and gestation 26.6[25–28] weeks. The day of diagnosis of PDA was 3[2–6] days and 16 were treated with ibuprofen and 44 with indomethacin. 11 hospitals referred infants for ligation during this period and there was no difference in time to ligation depending on inside versus outside pale. There was no operative mortality or morbidity and infants were discharged postoperatively on day 1 [1–2]. Age at PDA ligation was 22[15–29] days and 51 infants were >3 weeks old. Platelets, urea, creatinine and albumin were significantly altered postoperatively but urine output, FBC and other electrolytes were unchanged.

Conclusion: Access to PDA ligation in Ireland has no geographic variation. Prospective data collection is vital to assess longterm outcomes in preterm infants associated with PDA ligation.

THERAPEUTIC POSITIONING (TP) CONVENTIONAL POSITIONING (CP) TO IMPROVE FLEXOR TONE IN PRETERM INFANTS: A CONTROLLED STUDY

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Background: The term baby gets abundant and varied experience in the womb in preparation to meet the world. The preterm baby however starts out with a decided disadvantage in that is not physically

ready to adapt to the world outside the uterus. If full gestation is interrupted before 36 weeks the infant will not experience natural flexion. This hypotonic baby will suffer the full effect of gravity resulting in excessive extensor tone. This is more likely to occur if the baby is positioned supine as in conventional positioning (CP).

The aim of this study was to position the baby to encourage flexion and engagement of the hands in midline and to reduce adverse effects of gravity on the immature musculoskeletal system. The objective was to compare CP to therapeutic positioning (TP) by means of a customized positioner developed by the researcher (KN) using insulated wire, bandage, cotton and polythene cover.

Methods: Preterm infants who were below 37 weeks of gestational age, Apgar > 6 at 5 minutes and birth weight > 1,500 grams were randomized to CP or TP. Minimum of 12 hours positioning was administered per day for a period of 4 weeks in supine position. Heel to ear, popliteal angle, ankle angle scarf sign were compared, weekly by an Amiel-Tison neurological assessment by the researcher (KN).

Angles	Conventional positioning		Therapeutic positioning		<i>p</i> value
	Mean	SD	Mean	SD	
Adductor	87	13.80	77	12.07	0.019
Heel to ear	101.5	13.08	89.25	9.21	0.001
Popliteal	103.5	12.68	92	12.07	0.005
Ankle	10.5	6.048	6.15	3.99	0.010

Results: The TP group had significant lower adductor, popliteal, ankle, heel to ear angles compared to the CP group, indicating increased flexor tone in TP group.

Conclusion: TP, by simple customized devices, was associated with significantly better tone compared to conventional positioning. Further studies are needed to determine if TP is associated with improved, long-term neuro-developmental outcome in premature infants.

NORMAL TROPONIN IS ASSOCIATED WITH SEVERE ENCEPHALOPATHY AND POOR NEURODEVELOPMENTAL OUTCOME IN INFANTS FOLLOWING PERINATAL ASPHYXIA

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Introduction: Troponin is a sensitive marker of asphyxia in term infants mirroring the myocardial injury sustained in global hypoxia-ischemia. However these studies compare infants with asphyxia to normal neonates. Troponin has also been suggested to be a sensitive marker of stroke in adults.

Aims: To examine the relationship between troponin in infants with asphyxia and neurological outcome.

Methods: Infants with evidence of neonatal encephalopathy admitted to neonatal intensive care unit of a tertiary referral centre were evaluated and serum troponin was sent. Infants were retrospectively divided into those with normal and abnormal troponin results and birth history, clinical parameters, brain imaging were evaluated.

Neurodevelopmental outcome was assessed by a consultant paediatrician.

Results: 27 term infants with asphyxia had serum troponin measured of which 7 were excluded due to hemolysed troponin sample (5) and congenital abnormalities (2). 10 infants with normal troponin (mean $\pm$ SD: 0.07 ± 0.4 ng/mL) all had seizures and 7 had abnormal MRIs and neurodevelopmental delay. In contrast none of the 10 infants with elevated troponin results (0.6 ± 1.0 ng/mL) had seizures despite multiorgan involvement and all were developmentally normal. Gestation, Birthweight and Apgars were similar in both groups.

Conclusions: Troponin is a sensitive marker of severity of asphyxial injury in neonates. However infants with asphyxia and normal troponin all had seizures and increased neurological complications. Further exploration of the significance of troponin in this group is indicated.

Troponin	Normal (<i>n</i> = 10)	Abnormal (<i>n</i> = 10)
Troponin (ng/mL)	0.07 ± 0.04	0.6 ± 1.0
CK	$1,391 \pm 1,095$	$3,766 \pm 5,065$
LDH	$1,102 \pm 551$	$1,708 \pm 968$
Cord pH	7.0 ± 0.2	7.1 ± 0.2
CrUSS abnormal	6	4
MRI abnormal	7	0
Seizures	7	0
Neurodev abnormal	7	0

IS THERE EVIDENCE OF LUNG DAMAGE ON HRCT IN WELL CHILDREN WITH NORMAL LUNG FUNCTION WITH CYSTIC FIBROSIS, CHRONICALLY COLONIZED WITH PSEUDOMONAS?

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Background: In cystic fibrosis (CF), current practise involves eradication and maintenance antibiotic treatment of patients chronically colonized with *Pseudomonas aeruginosa*. Some asymptomatic patients are reluctant to start additional therapies. This study investigates whether there is lung damage on High Resolution Computed Tomography (HRCT) in patients with normal lung function, chronically colonized with pseudomonas.

Methods: Patients with CF who are asymptomatic [normal lung function (FEV₁ $\geq$ 80% predicted), no chronic cough, no exacerbations requiring intravenous (i.v) antibiotics], chronically colonized with pseudomonas (>3 positive cultures/year), who haven't been treated with nebulised/ i.v anti-pseudomonal antibiotics (last 2 years), were included. An ultra low dose HRCT thorax (the dose of radiation is equivalent to 2 chest radiographs) was performed and assessed by two experienced radiologists.

Results: Conclusion: In this small cohort of CF patients with chronic pseudomonas colonisation and normal lung function, 2/4 had bronchiectasis. While FEV₁ underestimated the presence of lung disease; HRCT was a more sensitive tool to assess lung damage. Information about lung structure may allow patients reluctant to start additional therapies, to make informed decisions. Further investigation of the

Patient characteristics	Patient 1	Patient 2	Patient 3	Patient 4
Age	16.2 years	16 years	15.2 years	9.9 years
Sex	M	F	M	F
Genotype	$\Delta F508/\Delta F508$	$\Delta F508/\Delta F508$	$\Delta F508/\Delta F508$	$\Delta F508/\Delta F508$
Age colonised pseudomonas	11.1 years	0.5 years	7.2 years	5.8 years
Duration colonised	5.1 years	15.5 years	8 years	4.1 years
Mucoid	No	No	Yes	No
Sensitivity	Resistant x1 antibiotic	Resistant x1 antibiotic	Resistant x1 antibiotic	Resistant x2 antibiotics
FEV1 % predicted	81%	92%	95%	82%
Pancreatic sufficient	Yes	No	Yes	No
HRCT report	Mild bronchiectasis in all 5 lobes.	Bronchial wall thickening	Bronchiectasis in all 5 lobes	Bronchial wall thickening

factors which results in slower progression of disease despite pseudomonas colonisation would be useful.

IS VARICELLA VACCINATION REQUIRED FOR CHILDREN WITH INHERITED METABOLIC DISORDERS?

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Background: Childhood varicella infection ('chickenpox') is a highly contagious self limiting disease generally considered to have low rates of secondary complications. Children with inherited defects of intermediary metabolism may potentially have more complications due to concomitant secondary metabolic decompensation with the infectious catabolic process and immunodeficiency associated with a number of these disorders. Immunization may decrease morbidity in this patient group. Presently children with Inborn Errors of Metabolism in Ireland follow the current national vaccination schedule in Ireland with precautionary guidelines.

Aims: (1) We sought to determine the frequency of varicella infection and associated complications in the cohort of patients with inherited disorders of intermediary metabolism or increased susceptibility to infection actively followed at our centre, (2) to identify the proportion of susceptible patients in this group who are varicella naïve and (3) to administer varicella vaccine to the latter group and monitor for any side effects.

Methods: Patients aged between 1 and 16 years on 1 July 2007 fulfilling the above criteria were identified from the NCIMD national database. A questionnaire designed to ascertain whether the child had a prior history of varicella infection, complications or vaccination was mailed to parents. Those who did not reply in writing were contacted by telephone.

Results: 126 subjects were identified and received the questionnaire. Responses were received from 122 subjects, (97% response rate). There was a history of previous varicella infection in 64 cases (53%) and of varicella vaccination in 5(4%). Fifty-three (43%) patients were

found to be varicella naïve. Of the 64 children who had a history of varicella infection, 5 patients required hospitalization for complications including life-threatening lactic acidosis in one patient with mitochondrial disease, and metabolic decompensation in the other 4 patients. 4 patients with mitochondrial disease developed secondary bacterial infection. In total, 9/ 64 (14%) patients had a complicated course including hospitalization. Prophylactic 'unwell dietary regimes' were administered to 59 children.

Conclusion: The complication rate observed in this study (14%) is much higher when compared with 0.01% in comparable age matched apparently healthy children. We thus propose that varicella vaccination be routinely administered to this cohort of children.

IMPROVING MANAGEMENT OF CHILDREN WITH ACUTE GASTROENTERITIS IN A REGIONAL PAEDIATRIC DEPARTMENT—COMPLETION OF AUDIT LOOP

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Aim: Acute gastroenteritis is one of the commonest presentations to emergency and paediatric departments. Careful clinical assessment of hydration status as well as evaluation of the risks of dehydration is crucial. It was anticipated that the closure of a dedicated Children's Gastroenteritis Unit in St Finbarr's Hospital, Cork in March 2007 would lead to increased admissions of children with acute gastroenteritis at Cork University Hospital. We recognised that the management of gastroenteritis needed to be reviewed and updated to optimise medical care.

Methods: A retrospective audit was conducted between March and June 2006, examining the clinical documentation, assessment of dehydration and management of 50 children admitted with acute gastroenteritis to Cork University Hospital. A prospective re-audit was performed using between March and May 2007 following the implementation of a new guideline, assessment form and parental information leaflet. The standards in audit were derived from a published evidence based guideline for management of gastroenteritis.

Results The results in 2006 showed that there were poor documentation and assessment of risk factors, signs and degrees of dehydration, inappropriate investigations especially blood urea and serum electrolytes, as well as inappropriate use of IV fluids and its unnecessarily prolonged duration.

Following implementation of the new strategy there was a remarkable overall improvement in clinical documentation, management and duration of admission. Recording of risk factors for dehydration and clinical signs associated with dehydration improved dramatically. The percentage of children who had blood urea and electrolytes sent during admission had reduced from 66 to 42%. The number of patients who were treated with intravenous fluids has reduced significantly from 50 to 30%. The duration of intravenous fluids has also reduced dramatically with the 80% of the patients received intravenous fluids for less than or equal to 12 h and none of the patients received intravenous fluids for more than 24 h, compared to previously where 64 and 92% of patients receiving intravenous fluids for less than or equal to 24 and 48 h respectively. There has been a significant reduction in the duration of admission with 98% of the patients admitted less than or equal to 48 hours, compared to 68% previously. There were no readmissions.

Conclusion: The adoption of a clinical guideline and assessment form which are agreed between senior clinicians leads to better assessment and management by trainees. Apart from desirable improvement in overall clinical practice, the reduction in the duration of intravenous fluids and admission is essential as part of saving of hospital costs.

SEEN BUT NOT HEARD: CONGENITAL CYTOMEGALOVIRUS IN AN IRISH COHORT

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Introduction: Congenital cytomegalovirus (cCMV) has been identified as the most prevalent congenital infection worldwide. It is a leading cause of neurological and sensory impairment, particularly sensorineural deafness, in children. Only 5–10% children with cCMV will be symptomatic at birth. However, most of these children suffer long-term sequelae. A further 10–15% of asymptomatic children with cCMV, will also develop long-term sequelae.

Aim: Our aim was to assess the incidence and outcomes of symptomatic cCMV in babies born between December 2004 and September 2007 in the National Maternity Hospital, Holles Street.

Method: The National Viral Reference Laboratory database was used to identify babies who tested positive for CMV. A retrospective chart review was performed to compile clinical details of these patients. Congenital infection was defined as a positive CMV culture, DEAFF test or PCR from urine or the presence of CMV IgM in blood within 3 weeks of birth.

Results: 19,521 babies were delivered during the study period. 11 infants were identified with CMV infection; 8 of these were classified as congenital infection and records were unavailable for one infant. Infants were screened for the following indications: 5 for intra-uterine growth retardation (IUGR), 1 for jaundice, hepatosplenomegaly and petechiae, and 1 as part of screen as infant of a HIV positive mother. Mean age at time of diagnosis was 5.5 days. Two babies were thrombocytopenic and 1 had abnormal liver function tests. 6 had cranial ultrasound changes suggestive of congenital infection, 2 of

these infants also were microcephalic. Two babies were treated with ganciclovir and 3 babies died of CMV-associated complications. At follow-up, one infant had sensorineural hearing loss identified by audiometry at age 3 months, 2 had early developmental delay and 2 infants were lost to follow-up.

Conclusion: Congenital CMV infection represents a significant cause of morbidity worldwide. Early diagnosis of cCMV in infancy may help us to identify neurological and sensory sequelae at a young age, permitting appropriate intervention in early childhood. The incidence of this disease in an Irish population is unknown. A collaborative study is taking place between the National Viral Reference Laboratory and the National Maternity Hospital.

RADIOFREQUENCY ABLATION FOR CARDIAC ARRHYTHMIAS: EFFICACY AND SAFETY IN THE IRISH PAEDIATRIC POPULATION

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Introduction: Radiofrequency ablation (RFA) of the accessory pathways causing cardiac arrhythmias has been conducted in children with success in many countries since its first use in North America. RFA has been conducted for this indication in the Irish paediatric population since 2001, but to date no data as to efficacy and complications has been collated. The study was carried out in Our Lady's Hospital for Sick Children, Crumlin which is at present the only centre in Ireland to conduct this procedure. The study therefore comprises all paediatric RFA data available in Ireland.

Aims: To identify the demographic and pathologic characteristics of the target population receiving this procedure. To assess short- and long-term success of RFA, and also complications which arose. To determine whether any subgroup of the target population was more predisposed to recurrence of arrhythmia or to complications of RFA.

Methods: A retrospective audit was conducted of all radiofrequency ablation procedures done in OLHSC from the first procedure in 2001 to date. Demographic data was gathered along with identification of diagnosis on ECG and electrophysiological studies. Data was collected from patient chart review and from catheter laboratory records. Data was analysed using Enstat III software.

Results: 40 patients met the inclusion criteria, of which data was available on 35. 30 had SVT (85%), and 3 (8%) had VT. 70% of those with SVTs were on Flecainide prior to RFA. Mean age at time of procedure was 9.83 years (SD 3.181, range 0.83–15.8). Mean age at onset of symptoms was 8.22 years (SD 3.32). Of the VTs ablated, 100% failed at first ablation but all were successfully ablated on repeat. 66% of SVTs were successfully ablated on the first attempt. 96% were ablated including repeat procedures (only 1 SVT refractory). Complications included equipment failure, atrial fibrillation post-procedure and myocardial perforation (1 case each), and heart block (3 cases).

Conclusion: Radiofrequency ablation in our population had an overall success rate of 96% and obviated need for further medical therapy in 88%. Repeat ablation was required in 34%. Complications included heart block, Afib and perforation. There was no statistically significant difference in success rates between males/females or age groups. Collection of further data prospectively might yield more information as to which subgroups respond best to RFA treatment.

PULMONARY FUNCTION IN PAEDIATRIC PATIENTS WITH TYPE I DIABETES MELLITUS

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Introduction: Several studies have been published to date on the topic of pulmonary function in patients with Type I Diabetes Mellitus with varying results and conclusions, many of which have been contradictory^{1,2}. However, some good evidence does exist to suggest poor performance in PFT's in these patients, which warrants further investigation^{1,3,4}.

Objective: To assess pulmonary function in children with Type I Diabetes Mellitus and to compare their performance to the internationally accepted values of their unaffected peers.

Methods: We performed a prospective, observational study of our affected cohort using standard spirometry equipment and techniques (PFTs). A standardised proforma was collated for each patient detailing their medical history and physical examination findings, including Tanner Staging. Blood samples were taken for full blood count, renal and thyroid function tests and glycosylated haemoglobin. Urinalysis was performed to detect microalbuminuria or glycosuria. Written consent/assent was obtained from parents/patients in advance of participation. Ethical approval was granted by the appropriate regional authority prior to initiation of the study.

Results: Fifty-one patients had PFTs performed. The male:female ratio was 1:1.13. The average age was 11.8 years, range 6–17 and the mean time since diagnosis was 4.5 years, range 0–13. Thirty-six (70.6%) of these children were in a state of diabetic ketoacidosis at presentation and the average number of hospital admissions was 3.54 per patient, range 1–25. Only 1 patient had a documented known complication of DM to date (early retinopathy), however, almost 20% reported minor complications with their insulin injections sites. The mean dose of insulin required was 0.88 mg/kg/day, range 0.44–1.63. Glycaemic control was satisfactory in our cohort with an average HbA1c of 8.4%, range 7.2–10.8.

Four patients in the study group had been previously diagnosed with asthma by their primary care physicians; 3 of these were receiving inhaled medication on an as required basis. However, we detected an incidence of restrictive lung disease consistent in 14 of 51 cases (27.5%). The mean FEV1:FVC ratio pre- and post-inhaled salbutamol were 91.0 and 92.5, respectively. Adjusted DLCO ranged from 15.9 to 37.1, with an average value of 25.5.

Discussion: Recent articles in the *American Journal of Medicine*⁵ and the *American Journal of Respiratory and Critical Care Medicine*⁶ cited the importance of recognizing respiratory involvement in patients with diabetes mellitus. Our study has confirmed that children with Type I DM have reduced pulmonary function when compared to their peers, this may have relevance with the planned introduction of inhaled insulin.

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HYPONATRAEMIA IN INFANTS WITH CONGENITAL HEART DISEASE

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Introduction: In recent years hyponatraemia has become an increasing problem in acute paediatric medicine and can be life threatening.^{1,2} As a consequence multiple guidelines have been developed to minimise the risk of hyponatraemia. However, there is a paucity of reports regarding hyponatraemia in the non-acute inpatient setting.

We observed an increase in mild hyponatraemia (130–133 mmol/L) in infants with congenital heart disease admitted to the RBHSC. The aims of this audit were to determine if there was an actual increase in incidence of hyponatraemia between the study periods, frequency of venesection and sodium supplementation; and to identify possible contributory factors.

Methods: This was a retrospective chart and biochemical database audit. We included infants (<12 months old) admitted to the paediatric cardiology department RBHSC in two 12 month time periods (2000 and 2005).

Results: In 2000 and 2005 respectively 62 and 66 infants were admitted. 40/62 and 50/66 charts were available. Age, weight and sex were similar in both study periods. There was a significant decrease in mean serum sodium from 140.3 to 136.6 mmol/L and an increase in the number of patients with hyponatraemia 24% (15/62) and 64% (42/66) (*p* values <0.05).

In 2005 86% (212/247) of hyponatraemic samples were between 133 and 130 mmol/L. Mean number of venesections per patient significantly increased from 9.9 to 16.4 (*p* value <0.05). The proportion of patients receiving sodium supplementation increased from 5% (2/40) to 38% (19/50) (*p* value <0.05). Diuretic and fluid therapies were similar in both eras. No seizures in either time period.

Conclusions: Mild hyponatraemia has increased in infants with CHD in our unit. In our patient group the only adverse outcomes have been iatrogenic in terms of venesection and sodium supplementation. We feel that a fresh appraisal of hyponatraemia in the non-acute setting is required to optimize patient management.

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EVALUATION OF THYROID SCREENING SERVICE IN DOWN'S SYNDROME POPULATION IN DONEGAL

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Introduction: Thyroid disorder occurs more frequently in people with Down's syndrome than in the general population.¹ About 10% of children of school going age have hypothyroidism.² Diagnosis on clinical grounds is unreliable therefore biochemical screening is essential. All babies born in the UK and Ireland have neonatal screening for hypothyroidism. Guidelines for surveillance for thyroid disorder in children with Down's syndrome were introduced in 2001.³ Biochemical testing, including estimation of T4, TSH, and thyroid antibodies should be carried out at least once every 2 years from age 1 year and throughout life. Mildly raised TSH (5–10 mU/l) or the presence of antibodies with normal T4 and no clinical evidence of hypothyroidism may not warrant treatment.

Objectives: There are 77 children up to age 18 years with Down's syndrome attending a dedicated clinic at Letterkenny General Hospital. The aim of the study was to evaluate thyroid screening service in this population in Donegal before and after the guidelines.

Study design: Retrospective study

Subjects and methods: All Children with Down syndrome born between 1 January 1994 and 31 December 2005 at Letterkenny General Hospital were identified using the HIPE system. A chart review was conducted in conjunction with electronic laboratory results. A simplified proforma was created to record all the following clinical detail.... The surveillance for thyroid disease for those children born before the introduction of the guideline (Group 1) was compared with those children born after 2001 (Group 2). The data was analyzed using SPSS 14.

Results: Forty children with Down syndrome were identified. Nineteen were born before the guidelines (Group 1) and 21 after the guidelines (Group 2). Testing for thyroid disease occurred every 2 years in 42% in Group 1 and in 86% in Group 2. (*p* value = 0.017). One child in Group 2 and none of the children in Group 1 had hypothyroidism detected on neonatal screening. None of the children in either group had symptoms attributable to hypothyroidism. Two children in both Group 1 and Group 2 were hypothyroid and on treatment. There were 8 children (42%) with transiently high TSH in Group 1 and 10 children (48%) in Group 2.

Conclusions: We observed an increase in thyroid function screening with the introduction of the surveillance guidelines in 2001. This improvement may be in part due to a dedicated Down syndrome clinic based in Letterkenny General Hospital. Despite better surveillance, there was no increase in detection of hypothyroidism. The prevalence of transiently high TSH was high in both groups. Further longitudinal studies on thyroid disease in Down's syndrome are required to evaluate whether testing every 2 years throughout life as suggested by the guidelines is justified.

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POSTER BOARD PRESENTATION

SPONTANEOUS SPLENIC RUPTURE IN THE NEWBORN

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Introduction: Splenic rupture in the newborn is a very rare entity. In the absence of predisposing factors, a high index of suspicion is required to diagnose and manage this potentially life-threatening condition.

We report 2 cases of Nigerian male infants who presented to our Neonatal Intensive Care Unit within a 6-month period, with acute splenic rupture. Both were full term and neither had any risk factors for splenic rupture.

The first baby was born by an emergency caesarean section and presented at 2 h of age with pallor, abdominal distention, a generalized petechial rash, hypotension and severe metabolic acidosis. His Haemoglobin was 10.5. He was ventilated, given multiple blood products and inotropic support. However the baby died at 27 h of age. Post-mortem revealed a large splenic laceration with massive intra-peritoneal haemorrhage. No underlying primary coagulopathy was identified.

The second baby was born by spontaneous delivery and presented at 33 h of age with pallor, poor feeding, abdominal distension, tachypnoea, hypothermia and severe metabolic acidosis. His haemoglobin was 6 g/dl. He responded to ventilation, inotropic agents and multiple blood products. Following transfer to Crumlin Hospital, he had an emergency splenorrhaphy at day 5, followed by a splenectomy at the age of 10. He made a full recovery.

Discussion: Splenic rupture is rare in neonates with <50 reported cases worldwide. The pathogenesis remains poorly understood. Predisposing factors include traumatic delivery, macrosomia, splenic parenchymal pathology and coagulopathies. Only 10 cases of neonatal splenic rupture in an otherwise healthy baby have been reported and none have occurred in infants of Nigerian parents. Despite its rarity, these 2 cases reinforce the need to consider splenic rupture as a possible diagnosis in unexplained circulatory collapse in a neonate.

MENKE'S DISEASE: A CHILLY DIAGNOSIS AND A CLINICAL CHAMELEON

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Introduction: Kinky Hair Disease is a fatal X-linked recessive neurodegenerative disorder of copper transport, first described in 1962 by John Menkes There is an estimated incidence of 1 in 250,000 live

births. It is caused by a mutation in the Menkes gene (ATP7A), a 23 exon gene, encoding Copper-transporting ATPase. The resultant faulty protein (MNK) is 1,500 amino acids transmembrane copper transporting P-type ATPase and is located in trans-Golgi network.

There are numerous mutations discovered so far unique to any given family. There is a massive clinical variability on presentation. We describe two cases of Menke's disease in males.

First case report: An 8-day-old term male presented with a 2 day history of poor feeding, pallor, lethargy and cold extremities. Birth history is uneventful. Family history is not significant. His parents were nonconsanguineous.

On examination, the patient had extreme hypothermia (25.2°), bradycardia (60 B.P.M) and acidosis (Ph 7.16) with decreased peripheral perfusion. Sepsis and endocrine abnormalities were excluded.

Investigations revealed caeruloplasmin 34 mg/L (200–400), copper 2.8 µmol/L (13.1–19.7). Ultrasound revealed bilateral uncomplicated duplex kidneys. MRI revealed a large right subependymal heterotopia. Later MRA revealed tortuous elongated vessels, both intra and extra-cranially with a double loop configuration of middle cerebral arteries bilaterally. Electron microscopy of hair shaft revealed pili torti and trichorrhexis nodosa. Mutation analysis revealed a deletion on exon 6–9 of ATP7A gene on the X chromosome.

Second case report: A 5-month-old term male presented with a history of afebrile seizures (staring unresponsive episodes lasting 30–40 s), hypotonia, delayed milestones and recurrent infections. Birth history is uneventful and family history is not significant. His parents are nonconsanguineous.

On examination, the patient had hypotonia, developmental delay and hypopigmented brittle hair.

Investigations revealed plasma copper 1.6 µmol/l (13.1–19.7) and caeruloplasmin 75 mg/L (200–400). MRI brain had cerebral atrophy and MRA revealed elongated tortuous cerebral arteries. Electron microscopy of hair revealed pili torti and trichorrhexis nodosa. Mutation analysis of ATP7A gene showed mutation c.1784_1785insG in exon 7.

Summary: Menke's Disease has a constellation of presentations. The disease may present in the neonatal period with hypothermia, hypoglycaemia, poor feeding, fatal haemorrhagic diathesis, multiple congenital fractures, jaundice, hypotonia, cephalohaematoma and hair is often normal. Later it may present with seizures, delayed development or regression, failure to thrive, hypotonia, poor head control, face is cherubic appearance, depressed nasal bridge, ptosis, reduced facial movement, hair is colourless and hypopigmented.

Our first case reveals the extremely early age of onset of symptoms with our second revealing a more classical presentation of Menkes Disease.

IDIOPATHIC PULMONARY HAEMOSIDEROSIS IN CHILDHOOD. A CASE SERIES

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Background: Idiopathic pulmonary haemosiderosis (IPH) is rare. Reported incidence is 0.24–1.0 per million children/year. 5-year survival varies from 50 to 86%. Treatment is controversial, but response to steroids, azathioprine and hydroxychloroquine has been described.

We discuss three cases of IPH diagnosed in OLCHC.

Case 1; referred aged 4 following investigations for recurrent anaemia and recurrent bilateral pneumonia. Bronchoalveolar lavage (BAL) and biopsy confirmed haemosiderosis. She responded to pulsed methylprednisolone but had recurrent haemorrhage. She is currently stable on hydroxychloroquine and azathioprine.

Case 2; a 3 years old became acutely unwell following general anaesthesia with bilateral infiltrates on CXR and fall in haemoglobin. BAL revealed haemosiderin laden macrophages. She responded to IV methylprednisolone and azathioprine but recurrent haemorrhages occurred on weaning of prednisolone.

Case 3; 4 years old with trisomy 21 and congenital heart disease presented with recurrent anaemia, chest symptoms and CXR infiltrates. Lung biopsy confirmed pulmonary haemosiderosis. He responded to azathioprine and steroid therapy but further haemorrhage occurred on weaning. He is currently on hydroxychloroquine. All children presented following referral from other specialty services with severe anaemia and responded to steroid treatment. Two required subsequent addition of hydroxychloroquine. Only one child had chest signs in association with haemorrhage. IPH remains a challenging condition to diagnose and treat.

A CASE REPORT AND REVIEW OF CUTIS APLASIA SECONDARY TO ANTENATAL EXPOSURE TO CARBIMAZOLE

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Aim: To report a case of cutis aplasia secondary to antenatal exposure to carbimazole and highlight the side effects associated with its use in pregnancy.

Case Summary: A 19-year old primagravida lady gave birth to a male infant at term by spontaneous vaginal delivery with three small well defined scalp lesions. The pregnancy was unplanned and at the time of conception Mum had been on carbimazole 5 mg BD for Grave's Disease which was diagnosed in 2002. She remained on a reduced dose of 5 mg OD until delivery. Baby H had no other associated anomalies and had normal thyroid function. The lesions healed well without any treatment and at four months of age he was doing very well with three very small areas of scarring and alopecia.

Discussion: Congenital defects of the scalp are uncommon; occurring in 1 in 2000 deliveries and despite several case reports in the literature, their association with in utero exposure to antithyroid drugs still remains controversial. Although our baby had isolated cutis aplasia there are case reports in the literature of carbimazole embryopathy, which encompasses choanal atresia, gastrointestinal anomalies, athelia or hypothelia, developmental delay, hearing loss, aplasia cutis and dysmorphic facial features. Poor control of thyroid disorders during the antenatal period is associated with adverse perinatal outcomes and thionamides appear to be the mainstay of drug therapy for Grave's Disease during pregnancy. Transplacental passage of propylthiouracil is less than carbimazole and it appears to be a safer drug to use in pregnancy.

Conclusion: This case report again highlights the likelihood of a direct association between in utero exposure to carbimazole and cutis aplasia. Propylthiouracil is preferred over methimazole or carbimazole during pregnancy.

MISSED VACCINES, MISSED OPPORTUNITY

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Background: Paediatric inpatients have an increased risk of incomplete vaccination compared to the general population. Opportunistic

vaccination of children in hospital is recommended in the UK and Australia but is not common practise in Ireland. Barriers to opportunistic vaccinations are lack of knowledge of the current immunisation schedule, lack of specific personnel with responsibility for coordinating vaccination of inpatients and also differing attitudes among medical staff as to whether opportunistic inpatient vaccination is appropriate.

Objective: To review the incidence of incomplete vaccination among paediatric inpatients and to assess practice among medical and nursing staff in identifying patients who were not fully immunised.

Methods: A prospective chart review and individual parent interview of 115 patients on a paediatric ward was performed detailing the patient's immunisation status.

Results: All patients had vaccination status documented by nursing staff on the ward and at triage, when admitted via the emergency department. Children were incompletely immunised in 26% of cases. The six month 5 in 1/Men C vaccine was the most commonly missed vaccine. A vaccination history was taken by on-call medical staff in only 77% of patients. Specific documentation of vaccination status was limited to "up to date" in 52% of case notes reviewed, but this was inaccurate in 10% when verified by individual parent interview. Only one child with incomplete immunisations was vaccinated during their inpatient stay.

Conclusion: Incomplete vaccination status is commonly found but very little is done to facilitate completion of the vaccination schedule in hospital. Accurate and detailed history taking is crucial in order to facilitate opportunistic vaccination. With the proposed addition of the Hep B and pneumococcal vaccines to the schedule in 2008, it is increasingly important that medical staff are educated about current childhood vaccinations and the valid contraindications to immunisation.

DISORDERS OF CHOLESTEROL BIOSYNTHESIS IN IRELAND

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Seven disorders of post-squalene cholesterol biosynthesis are recognized of which Smith Smith-Lemli-Opitz syndrome (SLOS) is the most common. These disorders are associated with major developmental malformations that are unusual for metabolic diseases and represent the first true merging of dysmorphology and biochemical genetics. Studies have reported SLOS, the prototypical example of this group, to be the third most common genetic disorder in American Caucasian populations after phenylketonuria and cystic fibrosis.

We reviewed the databases at the National centre for Inherited Metabolic Disorders (NCIMD) and the National Centre for Medical Genetics (NCMG) in order to identify all patients with inborn errors of cholesterol synthesis diagnosed in the 10-year period between 1 June 1997 and 1 June 2007. Clinical features (congenital malformations, behavioral phenotype, growth and developmental profile), biochemical features (plasma sterol profile at diagnosis), ethnic background and treatment were noted.

7 patients (age range 3–45 years) attend the NCIMDs making it the 13th most common condition treated here. An additional 6 deceased patients were identified from the database at the NCMGs.

All patients had SLOS. None of the other disorders of post-squalene cholesterol biosynthesis have been found in Ireland to date.

Cholesterol biosynthetic disorders are rare disorders in the Irish population but important to recognize as they are partially treatable.

VORICONAZOLE IN THE TREATMENT OF ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS IN CYSTIC FIBROSIS

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The incidence of allergic bronchopulmonary aspergillosis (ABPA) in children with cystic fibrosis (CF) aged over 6 years is 10%. This is associated with a deterioration in lung function and clinical status. The mainstay of treatment is oral corticosteroids, however antifungals may reduce the fungal load and have a "steroid-sparing" effect. Itraconazole has been superseded by voriconazole because of improved absorption and bioavailability.

Aim: To review the efficacy and safety of voriconazole in the treatment of ABPA in a cohort of children with CF.

Method: A retrospective chart review of all cystic fibrosis patients attending our hospital who have received voriconazole.

Results: 16 patients received 17 courses of voriconazole, with a mean age of 14.4 years. The mean increase in FEV1 post treatment was 8.8% of baseline. There was a mean drop in Total IgE of 37.6% from baseline and a mean decrease in prednisolone of 35%. Two patients reported side-effects—one had photosensitivity and visual disturbance, the other developed dry, itchy skin.

Conclusion: Voriconazole is a relatively safe and efficacious treatment for ABPA in our population. However a RCT of voriconazole in the treatment of ABPA in CF is needed.

TRIAL OF NEURALLY ADJUSTED VENTILATORY ASSIST (NAVA) IN THE PAEDIATRIC INTENSIVE CARE UNIT

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Introduction: Neurally Adjusted Ventilatory Assist (NAVA) is the first form of assisted ventilation to enable the patient's respiratory centre to assume full control of mechanical support regardless of changes in respiratory drive, mechanics and muscle function.¹ The electrical impulse which elicits diaphragm contraction (Edi) is sensed by electrodes positioned on a nasogastric tube. Computer analysis of this impulse then adjusts frequency, size and form of assisted breaths.

Sample case: We present the case of a 2-year old male who suffered smoke inhalation injury and subsequently developed ARDS which required high frequency oscillation for 10 days. By day 16 of admission he was eligible for a trial of NAVA.

Results: During the 3 h trial of NAVA there was a reduction in peak pressures and tidal volumes compared to those recorded on pressure support CPAP and normocapnia was maintained (Table 1). Pressures

delivered with each NAVA breath varied in proportion to the Edi impulse detected in contrast to the fixed pressure supplied by conventional ventilation.

Table 1

	Peak insp. pressure (cmH ₂ O)	Respiratory rate (b/min)	EtCO ₂ (%)	Minute vol. (L/min)
PS CPAP (PS 12/PEEP5)	18	38–48	5.2–5.7	3.7–4.6
NAVA (1.3 cmH ₂ O/ μ V)	10–18	30–39	4.7–5.2	2.7–3.8

Conclusion: Current ventilation strategies provide consistent, preset assistance per breath in the face of a variable respiratory drive. This may result in iatrogenic hyperinflation, respiratory alkalosis, and haemodynamic impairment.

NAVA is a more physiological method of ventilation that improves synchrony by providing mechanical support that varies in accordance with changes in the patient's respiratory drive.² By allowing the patient's diaphragmatic potential to trigger the ventilator, tidal volumes and airway pressures are consistently reduced, despite maintaining similar minute ventilations between the two strategies.

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ACCURACY OF REPORTING OF EXTREMELY PRETERM AND EXTREMELY LOW-BIRTH WEIGHT BIRTHS IN IRELAND

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Background: Extremely preterm (<28 weeks' gestation) and extremely low birth weight (<1,000 g) infants constitute a small proportion of births annually in Ireland. However, they are an important group as they have increased mortality, increased morbidity amongst survivors and consume a disproportionately large amount of health care resources. All births in Ireland are reported; this data is collected by the Register of Births, Marriages and Deaths of the Central Statistics Office (CSO) and the National Perinatal Reporting Service (NPRS) of the Economic and Social Research Institute (ESRI). Detailed information on all infants <32 weeks or <1,500 g admitted to nurseries in 9 hospitals in Ireland is voluntarily submitted to the Vermont-Oxford Network (VON). Extremely preterm infants born in hospitals without neonatal intensive care facilities have increased mortality and morbidity.

Aims: To compare the numbers of extremely preterm and ELBW infants identified by the CSO, NPRS and VON for the years 2000–2004 inclusive.

Methods: We contacted the CSO directly to request the number of infants born <28 weeks or <1,000 g. From VON we determined the

number of infants <28 weeks and <1,000 g. We compared the annual reports from NPRS.

Results: There were discrepancies in the numbers of infants identified by each method (see Tables 1, 2).

Table 1 No. of infants born <28 weeks' gestation

Year	CSO	NPRS	VON
2003	192	242	196
2004	210	n/a	201
2005	172	n/a	189

Table 2 No. of babies born <1,000 g

Year	CSO	NPRS	VON
2002	186	261	n/a
2003	187	267	177
2004	196	n/a	180

Conclusions: There appears to be significant and consistent variation in the number of extremely preterm and ELBW infants born and treated annually in Ireland. These factors have major implications for the planning and provision of tertiary neonatal care in Ireland.

BRONCHIAL CARCINOID TUMOURS—A RARE PAEDIATRIC ENTITY

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Bronchial carcinoid tumours, albeit rare, still constitute the most frequent primary pulmonary neoplasm of childhood and have been documented in those as young as three. The varying ways in which they can present, coupled with the fact that they are infrequently seen, means that initial clinical suspicion can be low and hence diagnosis and definitive management delayed.

Case report—a boy, aged 11, presented with left sided chest pain and mild cough. Initial X-rays revealed lingular opacification and a subsequent CT Thorax demonstrated a persistent lobulated soft tissue mass involving the region of the oblique fissure. This was felt to be secondary to previous infection in an underlying congenital lesion and he was initially treated with antibiotics and physiotherapy.

He re-presented acutely, aged fourteen, with cough, chest pain and haemoptysis. Repeat CT confirmed opacification in the left upper lobe and lingula with an area of calcification and raised the possibility of bronchial obstruction in left upper lobe bronchus.

Bronchoscopy showed an intraluminal well encapsulated lesion which was suggested to be a carcinoid tumour on endoscopic biopsy. A left upper sleeve lobectomy took place some three years later after initial presentation with histology confirming the diagnosis of a typical carcinoid tumour. Subsequent follow-up has shown no recurrence of tumour. This case will be detailed and the paediatric literature on pulmonary carcinoid tumours will be reviewed.

ATTITUDES OF THE HEALTH CARE PROVIDERS TOWARDS THE EXTREME PRETERM NEWBORN

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Background: Decision making at the limits of viability remains a challenging process. Outcome may be perceived differently and this may influence the decision making process.

Aims: To explore the opinions of healthcare providers on the use of interventions and outcome in the <28 week newborn.

Methods: An anonymous postal questionnaire was sent to various health care providers (obstetricians, neonatologists, paediatricians, SpRs in Paediatrics and Obstetrics and gynaecology, midwives and neonatal nurses) working in maternity units in the Republic of Ireland. Questions related to antenatal, intra partum and neonatal management of expectant women from 22 to 28 weeks gestation.

Results: The response rate was 48% to date (20% SpR in Paediatrics, 70% amongst neonatologists). 20% of the respondents would advocate administering antenatal corticosteroids at 23 weeks, 85% at 24 weeks. The majority (80%) felt that a neonatologist should counsel an expectant mother at 24 weeks. The majority provide verbal information only on outcome at less than 28 weeks. Neonatologists were more likely to advocate cesarean section at 25 weeks for both fetal distress and breech presentation. Wide variation between different healthcare provider groups and within groups was identified at gestations <25 weeks. Obstetricians were more pessimistic about survival and longterm outcome compared to neonatologists.

Conclusion: Different professional groups have different views on outcome and management of extremely premature infants. Greatest variation exists in the grey zone (<25 weeks). Establishment of national outcome data may help the decision making at the limits of viability.

AN UNUSUAL CASE OF RENAL STONES, FAILURE TO THRIVE AND HYPOTONIA IN A MALE INFANT

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Case report: A full term baby boy (birthweight 2.9 kg), second child to a non-consanguineous Irish couple, was delivered normally after a significant lack of growth from 29 weeks gestation. Family history was unremarkable.

At 3 months of age, he was admitted for failure to thrive with vomiting and poor feeding. On examination, he had plagiocephaly, a small triangular face, blue sclera, atopic eczema and an unusual scalp hair distribution. He had mild head lag and central hypotonia. He was noted to have transient renal tubular acidosis which resolved after a week course of sodium bicarbonate. Renal ultrasound showed presence of bilateral renal stones. Gastrostomy insertion and Nissen fundoplication were performed at 6 months of age for severe gastro-

esophageal reflux. His investigations did not reveal any underlying metabolic condition.

However, cytogenetic analysis was performed on this patient and revealed an abnormal male karyotype 46 XY, del(3)(q27.3q28).ish del(3)(q27.3q28)3q29(D3S4560+) including loss of material from the long arm of chromosome 3 with an interstitial deletion of the bands q27.3q28. In situ hybridization using Vysis probe D3S4560 mapping to the subtelomeric region of long arm of chromosome 3 (3q29) showed no evidence of deletion in this region, suggesting the deletion is interstitial.

Discussion: We report our case with clinical photographs and a review of the literature. The phenotype includes mild to moderate mental retardation, slightly dysmorphic features namely long and narrow face, short philtrum and high nasal bridge. Neurodevelopmental features that may be manifested are autism, gait ataxia and microcephaly. Other clinical manifestations include chest wall deformity, long and tapering fingers, cleft lip and palate, ligamentous laxity, recurrent middle ear infections, horseshoe kidney and hypospadias and abnormal pigmentation¹. Microdeletion of chromosome 3q has been associated with decelerations in head growth (which may be a useful diagnostic clue) nasal voice, six lumbar vertebrae, lower limb contractures and cerebral sigmoid venous thrombosis². An association with renal stones has not been reported.

Conclusion: There are <10 cases of distal interstitial deletion of chromosome 3q with variable clinical abnormalities reported. They may present as common neonatal or paediatric problems. Identification of a distinct 3q phenotype remains elusive.

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OVERDOSE OF INTRAVENOUS INTRALIPID IN A PREMATURE NEONATE

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Introduction: We would like to report a premature infant who inadvertently received a large infusion of intralipid (approximately 30 times the normal dose) in a 12-h period. This case illustrates the short-term physiologic effects and the long-term outcome of a significant intralipid overdose in an infant.

Case report: A boy was born by emergency Caesarean section at 30 weeks gestation (birthweight 1.1 kg) because of intrauterine growth restriction associated with absent placental end diastolic flow and a non-reassuring cardiotocograph. On Day 3, administration of parenteral nutrition (PN) was started. Intralipid 20% was introduced on Day 4, beginning at 1 g/kg/day, increasing to 2 g/kg the following day. Gastrointestinal priming with expressed breast milk was also started on Day 4. Full gastric feeds were established by Day 19. The following day, feeds were withheld because of abdominal distension. Parenteral nutrition including intralipid 20% (1 g/kg/day) was ordered to be infused at 0.2 ml/h. At 4:30 A.M., the night-shift nurse noted that the intralipid and PN infusions were reversed, with the intralipid infusion running at 6.7 ml/h and the PN at 0.2 ml/h. Following a chart review, it was concluded that the error occurred at 5:30 P.M. the

previous day (Day 20), when fresh PN and intralipid infusions were sited.

Both the point-of-care blood and central laboratory gas analyzers were unable to process the blood samples. Similarly, an urgent triglyceride level was unprocessable, while a repeat level 27 h later was normal (1.9: normal 0.3–1.7 mmol/L). Minimal elevation of liver enzymes with insignificant variation of total and direct bilirubin occurred, but the infant had persisting hyponatremia for five days following the incident, with serum sodium values ranging from 126 to 130 mmol/L. At 9 months old, the infant was noted to be appropriate for his developmental milestones. He had three admissions after discharge from the neonatal unit for investigation of failure to thrive and a suspected attachment disorder.

Discussion: This case highlights the potential for human error when the infusion rate is checked but the type of infusion is not, despite the difference in colour of the infusion (intralipid: white; PN solution: clear). Drug changeover should not be done during evenings or late in the day when human fatigue is prone to set in or during staff shift changes. Line crossings could also be the potential serious problem. Verification with a second person should precede medication administration and help ensure the seven rights of medication—the right patient, the right drug, the right dose, the right time, the right route, the right documentation, and the right programming of medication administration¹. Another cause of human error could be what has been described as a new socio-psychologic phenomenon called involuntary automaticity (IA), a cognitive mechanism that causes people to miss cues that are right before their eyes despite double-checking. In IA, the main problem is not making errors but missing them. One method to prevent IA is the verbal response double-checking protocols system².

Conclusions: This report highlights once more the potential for serious error with intralipid infusions. Although the intralipid overdose was not lethal, there were immediate systemic effects.

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IDIOPATHIC THROMBOEMBOLISM LEADING TO AMPUTATION OF THE LEFT UPPER LIMB IN A TERM NEWBORN

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Background: Arterial thromboembolism (TE) in the absence of current or recent history of a central catheter is a rare condition in the newborn period. We report a case of severe upper extremity arterial TE of unclear aetiology in a full-term neonate.

Case: Baby L was a male infant born at 36+4 weeks gestation following elective section for IUGR. Antenatal scan had shown oligohydramnios and a multicystic dysplastic kidney. Mum was 21 years old, Para 0; her serology was negative for CMV HIV Hepatitis and syphilis. She was rubella and varicella immune. HSV IgG was positive. At birth Baby L's left forearm and hand was noted to be black in colour below the antecubital fossa, with peeling of the skin of the left hand. There was a clear line of demarcation on the forearm. The brachial pulse was palpable but the radial pulse was not. Apgars were 9¹ and 10⁵. Birth weight 2.06 kg. The rest of the systemic exam was unremarkable. No amniotic bands were noted at delivery and the umbilical cord did not encircle the arm. Baby L was transferred to the NICU and subsequently to OLHSC for extensive investigations.

Haematology: prolonged PT, normal APTT, FBC and CRP Thrombophilia screen was normal. TORCH screen and blood cultures were negative. Herpes and varicella IgG were positive. Imaging: MRI of neck, c-spine, left upper lobe of lung and myelogram showed no mass lesions or signal abnormalities. MR angiogram showed the origin of the left subclavian vessel, however it was not visible beyond this and was not visible in the axilla. There was delayed contrast enhancement of the brachial and forearm arteries. Echo revealed a small PDA but no intracardiac thrombus. Cranial ultrasound was normal. Kidney ultrasound confirmed a duplex left kidney with a dysplastic multicystic lower pole. The right kidney was normal.

The placenta had been sent for histological analysis. No amniotic bands were seen. There was an active villitis of moderate to marked severity. It contained intranuclear cowdry inclusions that were suggestive of herpes infection. There was widespread secondary villous sclerosis due to vascular occlusion. Active and organising thrombi were present in the mainstream villi and chorionic vessels. Focal chorionic villous haemorrhage was also noted a sign of impaired fetal circulation. The thrombi in the vessels and or a vasculitis could account for the embolus in the infant.

Baby L was deemed not suitable for thrombolytics. He subsequently had a through elbow amputation at 1 month old with delayed closure of the skin. He was discharged two weeks later and has been attending for regular dressings since.

Discussion: This case represents a rare entity Cases reported to date have been associated with maternal diabetes mellitus¹, in utero twin-to-twin transfusion syndrome^{2,3} or amniotic bands⁴ and iatrogenic causes e.g. UAC⁵. In this case there was no maternal diabetes, the pregnancy was singleton, no amniotic bands were present and there was no invasive procedures performed. The only positive finding was IgG for herpes in infant and mother and histological findings in the placenta consistent with same, a previously unreported association. The degree of infarction and poor perfusion meant limb salvage was not an option.

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CASE SERIES OF HYPERPROLINEMIA TYPE II IN IRELAND

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Introduction: There are two inherited disorders affecting proline catabolism. Proline Oxidase deficiency results in moderately elevated plasma proline levels range 600–2,000 $\mu\text{mol/L}$ (normal $<350 \mu\text{mol/L}$). This is known as Hyperprolinemia Type I and is asymptomatic. However in Pyrroline-5-Carboxylate (P5C) Dehydrogenase Deficiency, known as Hyperprolinemia Type II, proline levels exceed 2,000 $\mu\text{mol/L}$. This condition is associated with seizures and mental retardation.

Biochemical analysis: Ion exchange amino acid chromatography shows markedly elevated proline levels in plasma and iminoglycinuria (elevated proline, hydroxyproline and glycine in urine). Urinary organic acid analysis detects the glycine conjugate of P5C (Pyrroline-5-Carboxylglycine).

Patients: There have been 12 cases of Hyperprolinemia Type II diagnosed to date in Ireland. Of these only 4 are attending the clinic at the National Centre for Inherited Metabolic Disorders. The ages range from 3 to 28 years. There is a wide spectrum of clinical manifestation ranging from the profoundly handicapped to the apparently asymptomatic.

Therapy: Previous literature suggested that either Hyperprolinemia Type II was (1) a benign condition or (2) that there was no effective treatment. We have attempted a treatment regime of reduced dietary protein intake. Plasma proline levels were monitored monthly.

Results: In two of the four patients the attempt to introduce protein restriction failed. The remaining two patients (siblings) reduced their natural protein intake by approx 33%. Three months into treatment their plasma proline levels have decreased significantly. The younger sibling is reported by parents and teachers to show improved behaviour and reduction of tremor. The older sibling is currently asymptomatic but it is felt that reduction of proline may have future preventative benefits.

Conclusion: Initial trials of dietary protein restriction in Hyperprolinemia Type II show promising reduction in plasma proline levels resulting in improved clinical outcome for affected patients.

STUDY OF THE KNOWLEDGE OF INHERITED METABOLIC DISORDERS AMONG PATIENTS AND THEIR FAMILIES IN THE IRISH POPULATION

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Galactosemia and Maple Syrup Urine Disease (MSUD) are among the common recessively inherited diseases in Ireland. Diagnoses are established on newborn screening. Patients are managed at the National Centre for Inherited metabolic disorders. Our aim is to assess the genetic knowledge among affected families to see if a referral to a genetic counsellor and further information is warranted.

Adult patients and parents of affected children were interviewed in person using a questionnaire including 4 demographic, 8 knowledge,

2 information and 5 impact questions. To date, 26 interviews have been conducted -8 adults patients (7 galactosemia, 1 MSUD) and 18 parents (12 galactosemia and 6 MSUD).

All parents of children with MSUD correctly answered questions on MSUD and recurrence, but did not know the risk or implications of carrier status. 9 of 12 parents of children with galactosemia scored 6/8 or better on knowledge, while all adults with galactosemia correctly answered 3–5 of the 8 knowledge questions. 16/26 study participants requested more information about their condition and transmission information. Affected adults identified a need to meet others with the same condition.

Our study indicated that parents of children with these genetic disorders are well informed, but adult patients would benefit from further genetic counselling. This may reflect a reluctance to transmit genetic information within families.

A CASE OF PERSISTENT PULMONARY HYPERTENSION IN A NEWBORN WITH COSTELLO SYNDROME

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A baby boy, the third child of a non-consanguineous Congolese couple was born at 37 weeks weighing 3.6 kg. Pregnancy was complicated by polyhydramnios necessitating induction of labour. He was in good condition at birth (Apgars 8₁, 9₅), however at 15 min of age he rapidly developed severe respiratory distress necessitating intubation and ventilation. Persistent pulmonary hypertension of the newborn was diagnosed based on echocardiographic findings plus the absence of other cardiopulmonary pathology. The baby was noted to have coarse features and lax skin. Ventilatory requirements weaned over the first few days and extubation was trialled on day 3 but failed because of upper airway collapse. He was however successfully extubated day 7. The infant made slow progress but was discharged aged one month still requiring tube feeds.

At four months he presented with rapid onset respiratory failure secondary to a severe left sided collapse and consolidation and again required intubation and ventilation. He was also noted to have severe failure to thrive and typical features of Costello syndrome including cutis laxa and deep palmar and plantar creases.

Ventilatory requirements weaned reasonably quickly but he failed extubation on two occasions apparently because of upper airway collapse. Microlaryngoscopy was performed and revealed a laryngeal inflammatory polyp which was removed and tracheomalacia which was not felt to be severe. Due to repeated failed extubations a tracheostomy was sited with good result.

Mutation analysis of the HRAS gene identified the common pathogenic mutation in exon 2; c.34G>A(p.Gly12Ser) confirming the clinical diagnosis of Costello syndrome.

At 8 months of age, the baby remains tracheostomy dependant. Efforts to decanulate have not been successful.

Persistent pulmonary hypertension of the newborn has not previously been described in Costello syndrome. Respiratory problems have been occasionally reported including pulmonary fibrosis, bronchomalacia, laryngeal collapse and subglottic stenosis.

This report adds to the range of respiratory problems which feature in Costello syndrome and suggests that Costello syndrome be considered in a dysmorphic baby with persistent pulmonary hypertension of the newborn.

DIAGNOSTIC OUTPUT OF SKIN BIOPSIES IN INHERITED METABOLIC DISORDERS

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Background and aims: The performance of a skin biopsy with culture of skin fibroblasts and enzymology studies is an important step in the diagnosis of many inherited metabolic disorders. In this retrospective analysis, the skin biopsies done over a 24 month period at the NCIMDs were evaluated to determine the diagnostic output from and main indications for this invasive procedure.

Methods: All patients who had skin biopsies performed between January 1 2005 and December 31 2006 at the request of a Metabolic Paediatrician were identified from the database at the Department of Pathology. Case records were reviewed. Age at time of investigation, clinical features, adverse incidents (infection, bleeding, specimen loss, cell failure) and biopsy results were recorded.

Results: 130 patients (83 male, 48 female) underwent skin biopsies during the study period. Age ranged from 2 weeks to 56 years. There were no adverse incidents. Clinical indications in order of frequency were, developmental delay 41 (31.5%), hypoglycaemia 20 (15.4%), positive family history of an inherited metabolic disorder 11 (8.5%), seizures 8 (6.2%), cardiomyopathy 7 (5.4%), ALTE 6 (4.6%), hypotonia 5 (2.3%), liver disease 3 (2.3%), Autism 2 (1.5%) and others 27 (20.8%). A formal report was available on all patients. In 29 of the 130 biopsies the results were diagnostically relevant, 11 patients were assigned a definite diagnosis (6 fatty acid oxidation defects and 5 respiratory chain defects), 7 a probable/possible diagnosis and 11 patient's enzymology studies out ruled a particular familial diagnosis, 3 of which were glutaric aciduria type 1. In 101 cases the diagnosis was established by other means or remained unclear.

Conclusion: The diagnostic output was 22.3%. The main factor for a conclusive result is the indication for the biopsy. This investigation remains a cornerstone in the pathway to a diagnosis of an inherited metabolic disorder.

SEASONAL VARIATION OF VITAMIN D AND ITS RELATIONSHIP WITH VITAMIN D SUPPLEMENTATION IN CHILDREN WITH CYSTIC FIBROSIS (CF)

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Aim: To assess vitamin D levels among children with CF, to determine the seasonal variation of Vitamin D and ascertain if any relationship exists between vitamin D level and vitamin D supplementation (IU), Pancreatic Enzyme Replacement therapy (PERT), (IU/kg), Age and FEV 1%.

Methods: A retrospective review of Vitamin D levels from the years 2003–2007 was obtained from medical records ($n = 204$ vitamin D levels, taken from 86 CF patients). Serum 25 (OH)D (vitamin D) had been measured using a radioimmunoassay (DiaSorin Stillwater, Minnesota). Further information (Vitamin D supplementation, PERT, age and FEV1 %) was obtained from the 86 patients and compared with their most recent vitamin D level, which had been measured within the previous year (2006–2007). Only patients on Cholecalciferol (vitamin D3) were included and were on current dosage for at least 1 year. Minitab Statistical Package was used to analyse the data. Regression analysis was carried out and box plots of seasonal variation in Vitamin D were drawn.

Results: Twenty-one percent and 58% of Vitamin D levels were >75 and 50 nmol/L respectively. Vitamin D levels varied each Month with the highest levels seen in August, September and October with median results of 69, 71 and 64 nmol/L respectively. The lowest levels were seen in December, January and February with medians of 40, 38 and 40 nmol/L. No correlation between Vitamin D levels and vitamin D supplementation, PERT or FEV1% was found with r of -0.18 ($p = 0.119$), 0.12 ($p = 0.285$) and 0.2 ($p = 0.072$) respectively. There was a significant negative association between Vitamin D level and age ($r = -0.37$, $p = 0.000$).

Conclusion: These results show that low vitamin D levels are prevalent in children with CF as only 21 % of levels were above 75 nmol/L as recommended by the CF Foundation. It also highlights the seasonal variation of Vitamin D and the possible latent effect of sun exposure as Ireland's sunniest Months are May and June (Met Eireann) yet the highest Vitamin D levels were observed in Autumn. The lack of positive correlation between Vitamin D Supplementation and Vitamin D levels show that current levels of Vitamin D supplementation (up to 2,000 IU/ day) may be inadequate to correct low levels in all Children with CF and thus an alternative source of vitamin D needs to be established.

MATURITY ONSET OF DIABETES IN THE YOUNG IN AN IRISH PEDIGREE

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Background: Heterozygous mutations of hepatocyte nuclear factor-1 alpha (HNF1A) genes are the most common cause maturity onset diabetes of the young (MODY). HNF1A mutations result in severe impairment of insulin secretion and an early onset of diabetes. Young patients with MODY are often mistakenly diagnosed as having type 1 diabetes mellitus (T1DM).

Case report: We describe a family with diabetes in three generations. The eldest sibling presented at the age of eleven when urinalysis (performed because of abdominal pain) revealed glycosuria. She had no osmotic symptoms. Her HbA1c was 8.9%. She required only a very small dose (.06U/kg) of NPH insulin subcutaneously (sc) daily. Her mother and grandmother were diagnosed as having T1DM as young adults but always had low insulin requirements. Her sister presented at the age of 12. She had had several skin and dental infection over a 6-month period and random blood sugar levels taken at home were high. Her HbA1c at presentation was 7.1%. Her diabetes was well controlled with small doses of subcutaneous NPH insulin. Their brother presented at the age of 13 years, one month later when he complained of polydipsia. In light of the family history further investigations were carried out and he was also commenced on low dose sc insulin.

The Oral Glucose Tolerance Tests for the siblings showed the following:

Time	Case 1	Case 2	Case 3
0	21.9	10.1	7.6
30	16.3	13.9	12.8
60	21.9	18.8	14.8
90	21.2	18.5	14.6
120	19.8	18.2	10.1

Anti-insulin, pancreatic islet cell and GAD antibodies were negative in all three children. Thyroid function and lipid profiles were normal. Genetic analysis found a hotspot mutation on exon 4: gly292fsX25. Their mother died of breast cancer prior to the genetic testing. The children's father and only other sibling tested negative for this mutation.

Conclusion: Rare forms of diabetes constitute probably at least a few per cent of all patients seen in diabetic clinics and should be considered where there is a strong family history, low insulin requirement and negative antibodies. The identification of the molecular cause helps to optimize treatment in specific clinical situations. The correct diagnosis also helps to predict the progress of diabetes in affected individuals and defines the prognosis in the family.

COLONISATION OF PREVIOUSLY "PREPARED" ENDOTRACHEAL TUBES IN THE DELIVERY ROOM

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Background: Endotracheal intubation is frequently performed during delivery room (DR) resuscitation. Endotracheal tubes (ETTs) are packaged as sterile, single-use items. Many practitioners prefer to use an introducer (or stylet) placed in the lumen of the ETT when attempting intubation in newborns. ETTs are frequently prepared (ie. introducer placed in the lumen, with or without prior shortening of the ETT) in the DR when the need for resuscitation is anticipated. Frequently, these ETTs are not immediately used. We noticed that many of these prepared ETTs remained on resuscitation trolleys in our institution and were thus available for later use.

Aim: To determine whether ETTs which had been previously been prepared were colonised with bacteria. To compare these to unopened ETTs and introducers found on DR resuscitation trolley.

Methods: On two days we collected all prepared ETTs from the resuscitation trolleys in the delivery suites and operating theatres of the National Maternity Hospital. We selected a total of five unopened ETTs and five unopened introducers from resuscitation trolleys in the DRs to act as controls. All ETTs were plated in the laboratory. Sections of the ETT tubing and connectors were incubated separately in Cooked Meat medium at 35°C overnight. The broths were sub-cultured onto Chocolate, Blood and Neomycin Agar. The plates were incubated overnight in CO₂, aerobic and anaerobic conditions respectively. All organisms isolated were identified.

Results: We collected 15 prepared ETTs from the delivery rooms, 13 of which had introducers. Thirteen (87%) ETTs were colonised with bacteria; the most frequently identified organisms were Coagulase Negative Staphylococci and Bacillus species. No organisms were cultured from the unopened ETTs and introducers.

Conclusion: ETTs which are found open on resuscitation trolleys are frequently colonised with bacteria which are potentially pathogenic. Such ETTs should be discarded and a new ETT opened if intubation is to be attempted.

AUTOANTIBODY POSITIVITY AND TYPE 1 DIABETES: THE TEMPLE STREET EXPERIENCE

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Background: Serological markers of an autoimmune pathologic process including islet cell (ICA), glutamic acid decarboxylase (GAD) and insulin autoantibodies (IAA) are present in 85–90% of individuals with Type 1 diabetes mellitus (T1DM) when hyperglycaemia is first detected¹. The prevalence of autoantibody positivity in our tertiary population of children with T1DM was reviewed to compare to international data.

Method: Any child who presented with new onset T1DM between January 2004 and June 2007 had blood taken for ICA, GAD and IAA at first presentation as per protocol. Annual assessments included immune function testing for hypothyroidism and coeliac disease. A retrospective study reviewed the findings for this cohort of children.

Results: Eighty seven children (male 53, female 34) were diagnosed in this time period. Median age at diagnosis was 8.4 years (range 1.3–13.7 years). A full antibody profile was available in 50/87. Twenty six out of 50 (52%), 6/50 (12%) and 3/50 (6%) children were positive for one, two and three antibodies respectively. Fifteen of the 50 (30%) were negative for all three antibodies. The most common autoantibody identified in new onset diabetes was ICA which was positive in 26/50 (52%). All the children who tested positive for more than 1 antibody were female. Children <5 years old had a lower incidence of antibody positivity. Of the eight children <5 years for whom a full profile was available, five (62%) were negative for all three antibodies.

Only two of the eight were in DKA at presentation, one of whom was antibody negative. All eight had symptoms typical of T1DM at the time of diagnosis and none were <6 months of age.

No child had a co-existing autoimmune disease at the time of diagnosis but four subsequently tested positive for hypothyroidism and one for coeliac disease.

Conclusion: Autoantibody presence in the diagnosis of Type 1 diabetes in an Irish population does not reflect international data, but screening for autoantibodies and comorbid autoimmune diseases remains an important part of diabetes management.

(1) Craig ME, Hattersley A, Donaghue K. ISPAD Clinical Practice Consensus Guidelines 2006–2007 Definition, epidemiology and classification. Pediatric Diabetes 2006;7:343–51.

GLUCOSE MONITORING IN A TERTIARY NICU

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Background: Studies on critically ill adults demonstrate the benefits of tight glycaemic control on short and long term outcomes. There is a lack of data in this regard in the paediatric or neonatal setting. Indeed,

there is not yet a consensus on the normal glucose value in either preterm or term infants. Review of the literature confirms that while bedside measurement of glucose with devices such as the Haemacue machine can be useful in the NICU, they should be used as a screening tool only. Confirmation with a laboratory measurement, particularly at the extremes of what is considered to be normal, should precede intervention.

Aim: Our initial aim was to assess the effect of glycaemic control on developmental outcomes in the preterm population. The first stage of this project was to assess frequency and accuracy of glucose monitoring in our unit, to establish whether or we comply with best practice.

Method: The laboratory data on all infants admitted to Unit 8 during 2006 was examined, with particular emphasis on glucose measurements.

Results: A total of 7473 laboratory specimens were sent during 2006. Of these, a glucose measurement was requested on only 168(2.25%) samples. 80(47.6%) of these were requested out of hours.

Discussion and conclusion: The amount of laboratory measurements of glucose during this period is unacceptably low. We conclude that practice in our unit relies too heavily on glucose measurement from our bedside testing, in this case the Haemacue machine, without confirmation of values in the lab setting. The Haemacue values should be used as a screening tool only for abnormalities in glucose control. In June 2007, a blood gas analyser with the capacity to measure whole blood became available in our NICU. This machine has been shown to correlate with laboratory measurement of glucose in a preliminary study.

We plan to commence a study formally comparing the glucose readings of the Haemacue meter, blood gas analyser and laboratory values, to include both term and preterm infants. From this, we hope to establish guidelines for best practice.

AEROBIC EXERCISE CAPACITY IN PRE-TEENAGE CYSTIC FIBROSIS CHILDREN

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Children with cystic fibrosis (CF) are observed to have diminished lung function, nutritional status and aerobic exercise capacity. All three parameters have been linked to overall survival. There is little information on the relationship between these parameters in pre-teenage patients.

This was a prospective case control study. Our study group consisted of patients from our CF clinic aged from 10 to 12 years. A control group of normal children were matched for age, height and gender. Pulmonary function tests were measured in all subjects. Aerobic capacity (VO_2 max) was measured using a cycle ergometer.

Twelve CF patients and an equal number of controls were recruited. The two groups, study versus control, were closely matched for age (10.5 years vs. 10.4), height (142.8 vs. 143 cm), weight (34.6 vs. 37.5 kg) and gender ratio. VO_2 max did not differ significantly between the study vs. control group (46.4 vs. 47.25 ml/kg, $p = 0.84$). FEV_1 was significantly better in the control group versus the study group (93.7 vs. 71%, $P < 0.001$).

Children with CF aged between 10 and 12 have comparable aerobic exercise capacity when compared to their peers despite significant deterioration in their lung function.

NON-CF BRONCHIECTASIS IN IRISH CHILDREN

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Non-CF bronchiectasis (NCFB) has been reported to be rare in developed countries

High resolution CT scanning has greatly facilitated the earlier recognition of bronchiectasis.

The epidemiology and aetiology of NCFB has not been documented in an Irish paediatric population.

Aim: To determine the aetiology, co-morbidity, severity and the lobar distribution of NCFB.

Methods: Retrospective review of clinical, radiological, immunological and microbiological data from 92 non CF patients with HRCT diagnosis of bronchiectasis.

Results: Of 92 patients (50 female), the mean age at diagnosis was 6.4 years.

The aetiology of bronchiectasis was as follows; Idiopathic 29 (32%), Chronic aspiration 18 (20%), Post-pneumonia 16 (17%), Immune deficiency 15 (16%), Chronic aspiration with immune suppression 5 (5%), Primary ciliary dyskinesia 8 (9%) Obliterative bronchiolitis 1 (1%).

Bronchial asthma and Gastroesophageal reflux disease (GORD) were concurrently present in 18 (20%) and 10 (11%) respectively.

Left lower lobe was commonly involved followed next by the middle lobe.

The common sputum isolates were *Haemophilus influenza* (50), *Streptococcus pneumoniae* (34) and *Staphylococcus aureus* (14), *Moraxella catarrhalis* (9) and *Pseudomonas aeruginosa* (8).

Surgical interventions were performed in 23 (25%) of patients, lobectomy 11(12%), Pneumectomy 2 (2%), laryngeal cleft repair 4(5%).

Conclusions: NCFB is under-recognised in Irish children and diagnosis is often delayed. Bronchial Asthma and GORD are common co morbidity.

A high index of suspicion and early HRCT can expedite the diagnosis.