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Contaminated breast milk leading to neonatal sepsis and death

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Introduction

The advantages of human breast milk over formula in preventing neonatal sepsis and necrotising enterocolitis has been well established in medical literature, hence the widespread use of breast milk to feed preterm infants. However, breast milk can occasionally transmit serious viral and bacterial diseases to the neonate. We report three cases of late onset neonatal septicaemia (including one death) in our unit that were likely caused by contaminated expressed breast milk.

Case studies

Twins were born at 32 weeks gestation via emergency caesarean section to a 34-year-old primigravida lady who had prolonged rupture of membranes for 16 days and received a course of antenatal steroids 2 weeks prior to delivery.

Twin 2, a female, required nasal CPAP and IV antibiotics for the first 6 days. She was fully established on expressed breast milk by day 9 of life. On day 14 she became suddenly unwell with clinical evidence of septicaemia. Despite immediate treatment with IV antibiotics, ventilation and cardiovascular support, she continued to deteriorate and died 2 days later. **Twin 1**, a male, had a similar initial clinical course and was established on expressed milk in the first week. On day 15, within 24 h of his twin becoming ill, he became vaguely unwell with a low-grade temperature. He was commenced promptly on IV antibiotics, remained stable and made a full recovery.

Twin 2 died from overwhelming *E. coli* septicaemia with evidence of meningitis. Twin 1 also grew *E. coli* from blood culture. A sample of mother's breast milk was sent for culture and grew *E. coli*. Serotyping revealed an identical strain of *E. coli* in the blood cultures and the mother's ebm.

In a third case, a singleton female was born via emergency caesarean section at 25 weeks gestation. Her birth weight was 790 g. She was intubated and given IV antibiotics. On day 19 she was well and was commenced on expressed maternal breast milk. On day 21 she developed signs and symptoms of sepsis and disseminated intravascular coagulopathy. She was treated with IV antibiotics. Blood cultures subsequently grew klebsiella pnuemoniae. Despite appropriate management, a repeat culture on day 27 continued to grow klebsiella pnuem. Mother's ebm was sent for culture and also grew klebsiella pnuem. Although serotyping was not done, this seemed the most likely source of the infant's septicaemia. This infant required significant intervention but eventually made a full recovery.

Discussion

We believe that the cause of all three episodes of late onset sepsis was contaminated maternal breast milk. There have been several reports of contaminated breast milk causing disease in the neonate. Despite this, there is little consensus on what constitutes significant contamination and how the issue of bacteriological screening of unbanked breast milk should be addressed. In the absence of evidence-based guidelines and in light of the

increasing number of such reports, we would urge the screening of breast milk prior to administration to preterm infants.

Obesity in childhood cystic fibrosis: a new phenomenon?

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Obesity has become one of the most important public health problems in child and adulthood. As the prevalence of obesity increases so does the prevalence of the comorbidities associated with obesity. The term ''obesity'' refers to children with BMI >98th percentile for age and sex and ''markedly overweight'' refers to children with BMI between the 91th and 98th percentile for age and sex. Heretofore the main concern in paediatric cystic fibrosis was nutrition and maintaining adequate weight gain. Now we are seeing a new trend of overweight children attending the department. Our aim is to comment on the prevalence of obesity in the Limerick CF group.

Methods

We performed a review of growth charts of the 68 children (0–16) attending the Midwestern hospital Paediatric cystic fibrosis unit. BMI was calculated for all patients and plotted over time. Characterisics of the patients were identified, including Δf508 homozygosity, age at presentation, BMI at presentation, average birth weight. Evidence of adverse effects of obesity were examined, HbA1c, lipid profiles, liver function tests and albumin coagulation studies and liver ultrasound appearances were recorded.

Results

Of the 68 children reviewed 4 (5.9%) were obese—between 98 and 99% percentile on the BMI chart.

Five (7.4%) children fell into the 91–98th percentile—markedly overweight. Of note also the patients who veer into obesity and overweight categories plateau later into more acceptable BMI categories.

The four obese patients in more detail:

Steatohepatitis	1/4
Abnormal liver function	1/4
HbA1c	All normal
Genotype	3/4 Δf508
Premorbid overweight at diagnosis	2/4
Fat soluble vitamins	All normal

Conclusion

Advances in nutrition combined with genetic factors and lifestyle influences, mean we are seeing more overweight children in the cystic fibrosis population. A previous study showed obesity exerts a positive effect on FEV1. Potential long term adverse effects on blood pressure, type 2 diabetes and steatohepatitis merit further consideration. As good nutrition has positive prognostic effects, is a period of obesity in childhood an acceptable phenomenon in cystic fibrosis?

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Quality of life and glycaemic control in a population of children with insulin dependent diabetes mellitus

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Introduction and aim

Diabetes mellitus is a chronic condition with an increasing prevalence worldwide. By its nature, insulin dependent diabetes mellitus (IDDM) has a significant impact on an individual's quality of life (QOL) and sense of well being.

An individual's self perceived QOL will influence their emotional attitude to their diabetes and will thereby influence their compliance, motivation and hence glycaemic control. The reverse is also likely to be true, i.e., poor glycaemic control contributes to a reduced perceived QOL and good glycaemic control contributes to an improved perceived QOL.

There have been a number of studies published looking at QOL and depression score in patients with diabetes but the majority of these have focused on the adult population. The Hvidore study looked at children 10–18 years of age but not under 10 years of age.

A better understanding of the relationship between perceived QOL and glycaemic control in children will contribute to our therapeutic skills in managing these patients.

Method

Quality of life questionnaires were conducted on 31 out of 40 children with IDDM, whose parents agreed to take part in the study. Three different QOL measurement tools

were administered to children and families during the study. These were the Behavioural and Emotional Rating Scale (BERS-2), the Diabetes Quality of Life Instrument for Youth and the Locus of Control rating scale. All assessments were conducted by the same clinical psychologist. All HbA1c measurements on each patient over previous 2 years were collected and documented.

Statistical analysis was done to explore the relationship between quality of life scores and standards of glycaemic control (HbA1C).

Results

HbA1c results for all 40 patients for the previous 2 years were reviewed. Average HbA1c for the group as a whole was 8.2%. Average HbA1c for children < 10 years was 7.6%; from 10 to 15 years was 8.3% and > 15 years was 8.9%.

The BERS-2 scale proved the most useful tool in assessing QOL. The mean scores for 'Affective Strength' subscales were above 12 indicating a 'very low' probability of having an emotional or behavioural disorder and there was a significant correlation between this subscale score and HbA1c levels ($p < 0.008$).

The Diabetes Quality of Life for Youth scale is a self-reported QOL scale and in our group of patients, 84.6% of those with a HbA1c of 8% or less reported 'excellent or good' quality of life.

Diet in children with cystic fibrosis related diabetes

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Introduction

There is little data available on dietary management of Cystic Fibrosis related diabetes (CFRD). Standard CF dietary intake is diabetogenic with recommendations of high simple carbohydrate 50% and fat 40% of total energy intake.

Aims

To assess dietary intake of children with CF and determine differences in dietary intake between normal glucose tolerance CF (NGT), IGT (pre-diabetes) and CFRD, 3 day food diaries were assessed. Anthropometric data was collected on all children. Data was analysed with WISP and SPSS.

Methods

About 100 consecutive attendees with CF; aged 9.5–19 years participated, (53 male and 47 females). Children with CF were categorized by Oral Glucose Tolerance Testing as: normogylcaemic (NGT) 65; impaired (IGT) 15 and diabetic (CFRD) 20.

Results

Boys was significantly lighter; when compared to the general population aged 11–18 years, $p < 0.05$. There was no difference noted for females with CF females. Mean % FEV1 was significantly lower for CFRD (51%) versus other mean values (71%) for glucose tolerance groups, $p < 0.05$. A significantly lower difference in % FEV1 and FVC was noted for ($n = 53$) males versus ($n = 47$) females, $p < 0.006$. All patients irrespective of glucose tolerance were below the recommended CF Estimated Average Requirements (EAR < 150%), $p < 0.001$. There was no difference for males and females. There was no difference in either: total energy intakes or carbohydrate intakes across the three groups. Total percentage carbohydrate as energy for NGT, IGT and CFRD was 54.5, 55.5 and 53.9%, respectively. We report a high simple sugar diet in all 3 groups, but no difference in quantity of sugars in diet for NGT, IGT and CFRD was 30, 26.6 and 24.2% respectively. About 31% were receiving nutritional supplements, with 15% on PEG feeding. A higher percentage of children with IGT (27%) and CFRD (25%) required PEG feeding versus NGT patients (9%). There was no difference in the percentage carbohydrate in the PEG in the three groups. Oral nutritional supplements were higher in the IGT (27%) group.

Conclusions

There were no statistically significant differences in anthropometry between the three glucose tolerance groups. There was a statistically significant difference for CFRD and %FEV1, $p < 0.05$. The current CF diet is no different in NCF, IGT nor CFRD. Further research is required on specific dietary guidelines for children with CFRD and non-diabetes.

Genetic aspects of cystic fibrosis related diabetes

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Cystic fibrosis related diabetes (CFRD) is increasing. Class III alleles of the variable number tandem repeats in the insulin gene (INS-VNTR) are associated with reduced insulin production. We tested whether INS-VNTR class III alleles are associated with CFRD, and whether the CFTR genotype influenced the development of CFRD.

DNA of 105 children with CF (64 f and 51 m, 9.5–19 years), and of 300 non-CF individuals was analysed. We used a restriction enzyme polymorphism (-23 Hph1) in the insulin gene as a surrogate marker for class I and class III alleles of the insulin VNTR. The -23 Hph1 polymorphism was analysed by PCR, enzymatic digestion and agarose gel electrophoresis. CF children were categorised based on OGTT results as: 72 (69%) normal glucose tolerant (NGT) and 33 (31%) CFRD.

INS-VNTR genotype

The control population INS-VNTR allele frequency was almost identical to other European allele frequencies. There was no significant difference between overall CF patients INS-VNTR allele frequencies and controls. There was a higher frequency of class I/III heterozygotes in CFRD compared to CF NGT, which did not reach statistical significance.

	Class I alleles (%)	Class I/III heterozygotes (%)	Class III alleles (%)
Control: 300	53	41	6
CF patients: 105	49	48	3
CFRD: 33	36.4	60.6	3
CF NGT: 72	54.9	42.3	2.8

CFTR genotype

The commonest CFTR genotype was homozygous delF508 (58%); 35% delF508 heterozygotes, 7% others R117H and G551D and 0.7% rarer CF genotypes. There was a statistically significant association between the delF508 homozygotes 23 (70%) and CFRD, (Fischer exact test, $p < 0.011$). The INS-VNTR genotype CF did not affect CFRD in delF508 homozygotes,

We report a non-significant trend towards a higher frequency of Class III INS-VNTR alleles in CFRD, suggesting that the insulin gene may influence CFRD. In addition, homozygosity for delF508 in the CFTR gene was associated with the development of CFRD. Larger population based studies are warranted to confirm these results.

Continuous glucose monitoring enhances the detection of cystic fibrosis related diabetes in children with cystic fibrosis

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Background

Cystic Fibrosis Related Diabetes (CFRD) and impaired glucose intolerance (IGT) are now frequent complications with improved survival in children with cystic fibrosis (CF). The increased morbidity and mortality associated with CFRD emphasizes the need for accurate screening. Many of the screening tools for the diagnosis of CFRD rely on standarised thresholds in the oral glucose tolerance test (OGTT) derived from epidemiological studies based on non-CF populations for the diagnosis of type 1 and 2 diabetes. Current screening tools are inadequate for the diagnosis of CFRD in children.

Aims

to evaluate continuous glucose monitoring (CGMS) in CF children; compare results with OGTT and HbA1c and determine if CGMS enhances the detection of CFRD.

Materials and methods

About 101 CF children had paired screening with OGTT and CGMS: patients commenced CGMS after 1 h calibration, underwent OGTT and continued CGMS for 48 h. Assessment was repeated after 6 months. **CGMS analysis:** All cases were individually analysed by experienced physicians in the management of diabetes in children. All CGMS glucose values were carefully analysed for mean glucose, variability

(MAGE, MODD and CONGA), duration and degree of hyperglycaemia. Clinical assessment was considered essential and individualised in every case.

Results

Children were categorized as normoglycaemic (NGT), IGT and CFRD. About 101 children with CF were screened at baseline and 100(99%) underwent repeat assessment at 6 months. Paired screening identified 12 new cases of CFRD. Insulin therapy was commenced in 12 cases following assessment of OGTT, CGMS and clinical status of each child.

Glucose tolerance category	Baseline GTT	Baseline CGMS	Baseline paired diagnosis	6 Months followup OGTT	6 Months followup CGMS	6 Months followup paired diagnosis
Screening children with CF with OGTT and CGMS at tme 0 and 6 months						
NGT	71	60	65	70	65	61
IGT	14	12	11	15	14	11
CFRD	16	29	24†	15	21	28†
Totals	101	101	101	100	100	100

Correlation coefficient $r = 0.88$, $p < 0.001^{\dagger}$)

Over a total 12 months time period 8% converted from NGT to IGT; 88% were unchanged and 5% progressed from IGT-CFRD. CGMS correlates well with OGTT at all five time points ($r = 0.75$–0.88). CGMS correlated less well with HbA1c [($r = 0.27$, $p > 0.05$) mean 5.5% range 3.8–9.0%]. Twenty percent of cases had abnormal HbA1c results; 50% of these were defined as NGT on OGTT and CGMS screening.

Conclusion

HbA1c and OGTT alone are not sensitive screening tools in CF. CGMS enhances early detection of abnormalities of glucose tolerance by revealing important trends that facilitate diagnosis. This report highlights the need to redefine the diagnostic criteria for CFRD, as HbA1c and OGTT are missing up to 75 and 27% of glucose intolerance in CF children, respectively. We propose annual paired screening OGTT and CGMS in all children with CF over 10 years if there is any clinical suspicion of diabetes development.

Retrospective review of the perioperative management of children with diabetes mellitus in Cork University Hospital

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Introduction

There is no consensus on the optimal metabolic management of children with diabetes mellitus (DM) undergoing surgery. The recommendations of International Bodies [1–3] are based on guidelines which recommend the use of intravenous fluids (IVF) and insulin to provide the most stable glycaemic control, and hourly blood glucose (BG) measurement (half hourly during surgery) to maintain BG in the ''optimal range'', 5–12 mmol/l. Improved peri-operative BG control using IV insulin compared to subcutaneous (SC) intermediate acting insulin has been demonstrated [4]. However long acting insulin has not yet been compared to IV for surgery.

Aims

This review aims to evaluate different treatment protocols used in the peri-operative management of children with DM from January 2004 to December 2006 and specifically to consider pre-operative glycaemic control, peri-operative insulin and fluid regimes, monitoring and BG levels and any post-operative complications or increased hospital stay.

Methods

All children with DM aged less than 18 years who had a surgical procedure requiring anaesthetic and fasting were eligible. The patients were identified using HIPE discharge summaries and a review of surgical ward admission books. The charts were subsequently analysed using a proforma to evaluate the aspects of patient management.

Results

Six children (age range 5–15 years) with DM had surgery. Pre-operative HbA1C ranged from 6.5 to 10.8% (an elective case) with mean 8.4%, indicating suboptimal control. The admitting surgical doctor did not review the diabetic control in any of the cases. The diabetic team were not involved until post-operatively in one emergency case (acute appendicitis), but were involved in all other cases. Urine was not analysed in four cases.

Case 1 and 2 (day cases on basal bolus regimes) had their usual SC long acting insulin and required no additional insulin. The mean BG level was 8.5 (range 8.2–8.8) and 11.5 mmol/l (7.9–16.9).

Case 3 (day case on two injection regime) received an insulin infusion (sliding scale) and IVF from admission. Mean BG level was 11.8 mmol/l (4.7–18.2).

Case 4 (acute appendicitis) was fasting for 30 h before theatre. She was successfully maintained on her usual dose of SC long acting insulin (0.48 U/kg/day) and IVF. Mean BG level was 6.7 mmol/l (4.1–10.1) with 23% of values outside the range. During and after surgery she was given IV insulin as per sliding scale. Post-operative mean BG was 4.4 mmol/l (2.9–11), with 81% values less than 5 mmol/l.

Case 5 (Alveolar bone graft) was also successfully maintained on her usual SC long acting insulin (0.1 U/kg/day). After surgery she was fasting for 48 h and receiving maintenance IVFs. Mean BG level was 10.2 mmol/l (5.2–13.8), with 30% of values outside the range.

Case 6 (acute appendicitis) was commenced on an insulin infusion (sliding scale) and IVF on admission. His mean BG was 9.2 mmol/l (6.1–13.4) with 15% of values outside the range.

The children had hourly BG monitoring pre and post-operatively; however three did not have BG levels documented during surgery (times ranging 75–120 min). Those who did have levels checked during surgery also had long intervals (75–110 min) with no BG level. It was difficult to assess the fluids and insulin given in theatre. There were no hypoglycaemic episodes and no post-operative complications or prolonged hospital stay.

Conclusions

This review provides evidence to suggest that children with type 1 DM may be treated with SC long acting insulin during fasting and surgery rather than conventional insulin infusions or sliding scale. It also demonstrates the need for better communication between the paediatric diabetic, surgical and anaesthetic teams and improved intra-operative monitoring and recording.

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Basiliximab induced noncardiogenic pulmonary oedema in two paediatric transplant recipients

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Objectives

To report early noncardiogenic pulmonary oedema related to Basiliximab induction therapy in young paediatric transplant patients

Methods

Retrospective review of all paediatric transplant cases performed in the National Paediatric Transplant Centre, Temple Street, Dublin

Results

Twenty-eight transplants were performed in 28 children between 2003 and 2006. Five were live related (LRD) and the remaining cadaveric (CAD). Six were pre-emptive (PET). All received Basiliximab induction 2 h prior to surgery. In addition, induction immunosuppression consisted of Tacrolimus and Methylprednisolone. In all but two patients, Basiliximab was re-administered at day 4.

Two patients, aged 11 and 6 years (one CAD and other LRD, respectively) developed acute noncardiogenic pulmonary oedema 6–48 h after transplantation. Both children had renal dysplasia as primary cause of renal failure. Both required delayed ventilation and were ventilated for 4–6 days, respectively. Both grafts had primary function, but the CAD transplant subsequently developed acute tubular necrosis, and was eventually lost within 3 weeks due to thrombotic microangiopathy and severe acute antibody mediated rejection despite immunosuppression with sirolimus, mycophenylate, steroids and plasma exchange therapy.

Conclusion

We report a rare but serious side effect of Basiliximab. To our knowledge, this is the first report of Basiliximab induced noncardiogenic oedema so early post transplantation and in such young children. Early recognition and aggressive appropriate supportive therapy is vital for patient and where possible, graft survival.

Food challenges are easy, safe and important procedures

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Aim

Food challenges are the gold standard in diagnosis of paediatric food allergy. In the absence of definitive diagnosis, exclusion diets may remain in place for many years after they should have been reassessed. Food challenges are easy to perform but require intensive supervision. We report our early experience with food challenges in Ireland's only Allergy Clinic in Cork.

Method

Prospective collection of data regarding each food challenge performed from September 2006 to date.

Results

Thirty open food challenges were performed in 29 children. Nine subjects were < 5 years, 15 were 5–10 years and 5 were 11–14 years. Eighteen challenges were negative and 11 were positive. One child was uncooperative. One wheat challenge, five of seven peanut challenges and 5 of 12 egg challenges were positive. All four milk challenges were negative. No child required adrenaline and every child was discharged the same day.

Conclusion

Open food challenge is easy and safe to perform. These first Irish data are consistent with international data regarding resolution of milk allergy and relative persistence of egg and peanut allergy. Many children tested appear to have been on unneccesary elimination diets for many years because the persistence or resolution of allergy had not been objectively demonstrated.

A survey of imported childhood malaria in Dublin: 1999–2006

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Aim

To describe the epidemiology and survey the inpatient management of imported childhood malaria in Dublin between 1999 and 2006 and compare it to current best practice guidelines.

Methods

Cases were identified through laboratory archives from the three Paediatric Hospitals in Dublin. Demographic and laboratory data and details of clinical outcome and management were extracted retrospectively from the clinical case notes.

Results

Sixty-seven cases were identified (37 male, 30 female). The average age at presentation was 78 months (range 7–185 months). Twenty-nine cases were among new immigrants from malaria endemic areas, 38 among travellers from Ireland to endemic regions. The commonest reason given for travelling to a malaria endemic region was to visit relatives and friends (23). The country most commonly visited was Nigeria.

P. falciparum alone was responsible for 58 cases (88%) identified, *P. vivax* for three cases. Six children had mixed infection. Among those who travelled from Ireland to malaria endemic regions, only 10 (26%) had taken regular malarial prophylaxis.

Fifty-eight children were admitted, giving a total of 280 inpatient days (mean in-patient stay of 4.8 days). Fourteen children were admitted to intensive care unit with a mean stay of 2.5 days. A total of 13 different anti-malarial regimens were used in this cohort. Almost 80% of patients received combination anti-malarial therapy. The average duration of anti-malarial therapy was 9.3 days.

Only one child had a documented relapse. Follow-up blood smears were performed after discharge in 27 cases (40%). No child in this cohort died from malaria or from its complications.

Conclusions

Imported childhood malaria remains relatively uncommon, but is an important differential in children recently arrived from or returning from endemic areas. The results show good outcome from imported childhood malaria in Dublin since 1999. The poor uptake of anti-malarial prophylaxis, however, indicates that we urgently need to improve preventive measures among travellers to high-risk regions. The wide variety of regimens employed, and the disparity in practice between the three different hospitals also suggests that an agreed management protocol would be beneficial, and might lead to shorter in-patient stays. Post discharge follow-up also needs to be improved.

Ursodeoxycholic acid use in cystic fibrosis

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Background

Cystic fibrosis-related liver disease peaks in adolescence and up to 20% of adults develop chronic liver disease. A Cochrane review in 2000 suggested insufficient evidence existed to justify the routine use of ursodeoxycholic acid in cystic fibrosis. In recent years evidence has emerged to support the increasing use of UDCA in cholestatic liver diseases Routine ultrasonography has been shown to be an early marker of liver disease.

Aim

To examine ursodeoxycholic acid (UDCA) use in the paediatric population in the Cystic Fibrosis Unit in the Midwestern Regional Hospital and also to describe the characteristics of those on UDCA. We review the results from our clinical evaluation for detecting and monitoring the progression of liver disease, including biochemical tests of liver function and abdominal ultrasonography, in those on UCDA and those not on UCDA. We also remark on the incidence of overt liver disease in this population.

Methods

A retrospective chart review was undertaken, including all patients between 0 and 16 years.

Results

Data on 68 children was collected. All patients were included. The average age at diagnosis of cystic fibrosis was 10 months, 43/68 cases were ΔF508 homozygous and 44/68 (65%) of our patients are on ursodeoxycholic acid. The average age of commencement was 5.8 years ranging from 15 months to 13 years. In the group on UDCA the main indication for commencement was abnormal ultrasound findings 39/44 (88%), 10/44 (23%) has abnormal biochemistry and 3/44 (7%) were commenced prophylactically.

All patients on UDCA had an annual assessment consisting of ultrasound and biochemistry. The follow up ultrasound was abnormal in 39/44 cases. Follow up liver function tests were normal in 82% and abnormal in 11% of those on UDCA. Further examination of the follow up ultrasound scans showed a stable appearance in 28/42 (66%), disimprovement in 8/42 (19%) and improvement in 5/42 (12%).

About 3/68 (4.4%) developed overt liver disease (abnormal LFTs, splenomegaly, cirrhotic nodular changes on ultrasound). None of these was on UDCA. One required liver transplant and 2 have compensated liver disease.

Conclusions

Interestingly on UDCA, the majority of abnormal ultrasound appearances remained stable or in some cases reverted to normal.

Although the evidence that UDCA protects against CF related liver disease lacks rigorous multi centre double blind controlled trials, our experience in a single centre suggests that early use of UDCA prevents the development of CF related liver disease.

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A 14-year study (1993–2006) of invasive neonatal sepsis and meningitis at the Rotunda Hospital, Dublin

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Aim

An audit of invasive sepsis in infants admitted to a neonatal intensive care unit during a 14 year period from 1993- 2006.

Method

A retrospective audit was performed of infants with positive cultures from sterile sites. Because of the difficulty of interpreting the significance of isolates that are normal skin flora, coagulase negative staphylococci and diphtheroid species were excluded from the analysis. Data were collected on organism type, antibiotic susceptibility pattern, site of infection, age at onset of infection, gestation at birth, birth weight, sex, multiple birth and outcome. Early-onset sepsis (EOS) was defined as onset of invasive infection in a neonate < 7 days old and late-onset sepsis (LOS) onset at ≥7 days old.

Results

There were 88,491 livebirths (LB) over the study period. In all there were 97 episodes of EOS (incidence, 1.09/1,000 LB). Bacteraemia alone was the manifestation in 92.8% of cases and 7.2% had meningitis ± bacteraemia. Group B Streptococcus (GBS) was the most common cause of EOS (0.43/1,000 LB). Infants born preterm accounted for 48.4% of EOS cases. There were 11 deaths (11.3%) from EOS, 74 infants survived and the outcome is unknown in 1 infant. There were 133 episodes of LOS (incidence 1.5/1,000 LB), of which 112 occurred in hospitalised infants and 21 in infants admitted from the community. Bacteraemia alone was the manifestation in 88.7% of cases, 10.3% had meningitis ± bacteraemia and $< 1\%$ had septic arthritis. The commonest causative organisms were *E. coli* (21%) and *S. aureus* (19.7%). Other important organisms associated mainly with LOS in hospitalised infants were *Klebsiella* species ($n = 11$), and *Candida* species ($n = 8$). *Listeria monocytogenes* was isolated from only 1 infant (meningitis, LOS). 75% of LOS occurred in pre-

term infants and 85.2% in infants of < 3 kg birthweight. There were 5 deaths (3.75%) directly attributable to LOS. Results from susceptibility testing showed that all GBS isolates remained sensitive to penicillin, and all S. *aureus* isolates were sensitive to methicillin/flucloxacillin, only 36% of *E. coli* were sensitive to ampicillin and >95% of *E. coli* were sensitive to gentamicin and cefotaxime.

Conclusions

Although GBS is the most common cause of EOS, the incidence (0.43/1,000) remains below that in many other countries. Similar to the experience in other countries, most LOS in hospitalised infants occurred in preterm LBW infants. Antibiotic resistance is uncommon in our NICU population; factors contributing to this include a strict antibiotic policy and an active ongoing screening programme for colonisation with antimicrobial resistant organisms in infants in the NICU.

Death and decision making in neonatal intensive care

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Introduction

Technological and pharmacological advances have greatly expanded treatment options for critical care of newborn infants. However, with this increased ability comes an increased responsibility as to whether these interventions are appropriate in all circumstances.

Aims

To assess end of life decision-making in a tertiary neonatal intensive care unit.

Methods

This study is a retrospective chart review of all neonatal deaths in a tertiary neonatal intensive care unit over a 2-year period. There are approximately 8,000 deliveries each year and approximately 1,000 admissions to NICU each year. Maternal, antenatal, perinatal and neonatal factors were reviewed for each case. We sought to answer specific questions, namely the cause of death, whether active intervention was withheld or withdrawn, the existence of formal do-not-resuscitate (DNR) orders documented in patient charts, parental involvement in the decision making process and use of analgesia. Deaths were categorised according to Wigglesworth classification.

Results

All charts of neonates who died between January 2005 and December 2006 (inclusive) were reviewed. A total of 80 patients were identified, 41 deaths occurring in 2005 and 39 in 2006, mortality rates of 5.12/1,000 and 4.87/1,000, respectively. The corrected neonatal mortality rate was 2.825/1,000 over the 2-year period. The median day of death was 2 (1–24 days). Extremely low birth weight infants accounted for 46% of all deaths. The treatment decisions were made either antenatally (10%), at the time of delivery (22.5%) or following the initial evaluation in the intensive care (67.5%). The treatment decisions were clearly documented in 62.5% of cases prior to enactment of decision. The presence of a formal DNR order was documented clearly in 35% of cases prior to death. Withholding life-sustaining treatment occurred in 37.5% of cases, due to extreme prematurity or a known lethal congenital malformation. Comfort care was provided in all cases. Care was actively withdrawn in 42.5% of cases following discussion with parents. The main reason documented was poor prognosis and adverse longterm outcome. Severe intraventricular haemorrhage, confirmed lethal malformation and severe hypoxic ischaemic injury accounted for the majority of cases where care was withdrawn. In 11.2% of cases death occurred despite maximal intensive care. Analgesia was administered to 26% of patients where care was being withdrawn in the intensive care setting and morphine was the chosen agent in all cases. The post-mortem rate was 16%.

Discussion

Current practice reflects a shared decision making process and highlights the importance of effective communication between healthcare providers and parents both antenatally and postnatally. Quality of life issues play an important role in this decision.

Asphyxia in full term infants: is it preventable?

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Intra-partum asphyxia (I.A) in mature infants causes 10–15% of cases of cerebral palsy. Over the last 30 years there has been a massively increased intervention rate in pregnancy and labour, with widespread ultrasound fetal assessment, almost universal adoption of continuous fetal heart rate monitoring and a more than quadrupling of the caesarean section rates from 5 to 25%.

The most reliable indicator that I.A. has occurred in a term infant is the presence of an encephalopathy in the 48 h following delivery, plus evidence of other organ damage (kidney, heart, liver, muscle). Seizure rates in term infants are reported on an annual basis by all three Dublin maternity hospitals. These figures show that the rate of seizures in the early 1980s was 0.87/1,000 live births, at a time when the caesarean section rates were 5–7%. The seizure rate has worsened to $>1/1{,}000$ over the intervening years despite current caesarean section rates of 18–28%.

Aims

To determine attributable risk individual factors associated with the development of neonatal encephalopathy in a case/control study. Secondary aims were to assess the predictive value of laboratory and neuroimaging investigations in particular MRI scanning for neurological outcome.

Methods

All infants admitted to the Rotunda NICU between Jan 2001 to present with neonatal encephalopathy within the first 24 h of life were included. A grade of mild, moderate or severe was assigned. (Sarnat and Sarnat). Two controls (the infant born before and after each case) were chosen for each case. Exclusion criteria were gestation <36 weeks or any congenital anomaly. Detailed maternal antenatal history, resuscitation history, neonatal clinical course laboratory and neuroradiology imaging findings were studied and correlated to outcome.

Results

To date 150 cases (95 babies with grade 1 encephalopathy; 55 babies with grade 2 or 3 encephalopathy) and 296 controls have been analysed. Primagravity, nationality, APH, oligohydramnios, grade 2 or 3 meconium, maternal pyrexia in labour, mode of delivery and venous cord blood gas were significant risk factors for I.A. On logistic regression (controlling for parity, IOL & length of stage one of labour) odd ratios for assisted delivery was 3.2, emergency c-section was 8 and assisted & emergency c-section was 13.4. Of laboratory markers raised LDH levels were the most sensitive. A normal MRI was more sensitive for normal outcome than normal cranial ultrasound. When abnormalities on imaging are present a combination of MRI and cranial ultrasound are the most useful.

Conclusion

Most assisted deliveries\emergency sections are preformed in an effort to prevent birth asphyxia. It is clear that by current means we are identifying babies at increased risk. However we have failed to improve outcome despite our intervention. Earlier, increased, targeted risk factor reduction may lead to improved identification of those at risk of I.A.

Efficacy of intravenous immunoglobulin in the management of haemolytic disease of the newborn

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Introduction

Intravenous immunoglobulin (IVIG) is indicated for use in Rhesus and ABO hemolytic disease of the newborn (HDN) to reduce the need for exchange transfusion, to decrease hospital stay and the duration of phototherapy.

Aim

To evaluate the change in bilirubin levels following intravenous immunoglobulin in neonates with haemolytic disease of the newborn.

Methods

Cohort study of infants who received intravenous immunoglobulin for the management of severe hyperbilirubinaemia from 1st July 2005 to 31st December 2006. Clinical data were extracted from patient records. The rate of rise of Total serum bilirubin (TSB) was calculated over approximately 6 h immediately prior to IVIG administration, and compared with the rate of rise in the 6 h immediately after IVIG administration. IVIG was administered (1 g/kg) when despite intensive phototherapy, TSB level rose to within 34–51 μmol/l of the exchange level and if required a repeat dose was given.

Results

11 infants received IVIG and the effect of IVIG on the total serum bilirubin (TSB) level, and its effect on the rate of rise of TSB was quantified. There was a statistically significant decrease in bilirubin levels before and after treatment with IVIG from 234 to 219 mmol/l ($p = 0.001$). In addition, the rate of change in bilirubin level significantly altered from an upward to a downward trend ($p = 0.001$). The Number Needed to treat (NNT) to prevent an exchange transfusion was 2.75—comparable with the recent systematic review of IVIG with a NNT of 2.7.

Conclusions

The ''number needed to treat'' in order to prevent one exchange transfusion is very low and the financial saving from the shortened inpatient stay and duration of phototherapy offsets the cost of IVIG. We would advocate the use of IVIG in hyperbilirubinaemia secondary to haemolytic disease in accordance with the recent AAP guidelines to avoid exchange transfusion.

Vasopressor inotropes lead to impaired myocardial function and systemic blood flow in preterm infants

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Introduction

The widespread use of vasopressor inotropes in preterm neonates in response to hypotension stems from the assumption that hypotension reflects low systemic blood flow. These agents subject the preterm myocardium to an increasing afterload and may impair contractility, leading myocardial ischemia and worsening systemic blood flow. We hypothesised that dopamine may worsen myocardial performance in preterm infants.

Methods

Echocardiographic measurements of myocardial function, cardiac output, and systemic blood flow were performed during dopamine administration and compared to controls on days 1 and 3 in infants >1,500 g. Troponin T and NT-pro B type natriuretic peptide (NTpBNP) levels were measure at the same time as echo.

Results

Thirty-three infants were included. Five infants received a dopamine infusion on days 1 and 3 compared to 28 controls of similar gestation, birth weight and antenatal background. Table 1 illustrates the effect of dopamine on different echocardiographic and biochemical parameters of myocardial function on day 1. We will also demonstrate the effect of dopamine on myocardial function using cine loop echocardiography.

Table 1 Dopamine infusion is associated with lower shortening fraction, LV and RV outputs, and lower celiac artery blood flow

	No Dopamine (n = 28)	Dopamine (n = 5)
Mean BP (mmHg)	34 [31–39]	31 [28–33]
Shortening fraction (%)	33 [24–38]	20 [7–26]
RV stroke volume	1.13 [0.71–1.72]	0.77 [0.42–1.14]
RV output	169 [114–208]	96 [80–200]
LV stroke volume	1.08 [0.74–1.46]	0.86 [0.38–0.92]
LV output	155 [120–194]	108 [80–172]
Celiac artery blood flow	25 [16–39]	19 [12–30]
Troponin (pg/ml)	0.21 [0.11–0.37]	0.69 [0.16 – 1.22]
NTpBNP (pmol/l)	1,271 [745–3,486]	1,267 [660–2,163]

Troponin levels were also higher in the dopamine group. Stroke volume was expressed in ml, Output and flow in ml/kg/min

LV left ventricle; *RV* right ventricle

Conclusion

We have demonstrated the potential detrimental effect of dopamine on the preterm myocardium and circulation. More research is needed examining this effect and long term outcome associated with the use of inotropes.

Management of patent ductus arteriosus in the very low birth weight infant

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Background

Controversy surrounds the management of patent ductus arteriosus (PDA) in the very low birthweight infant (no treatment, prophylactic, early targeted or late treatment). Late diagnosis of PDA may be associated with a reduced rate of successful medical closure, necessitating surgical ligation. Recently we have adopted an earlier targeted treatment approach.

Aims

To audit the management of PDA within our unit.

To compare early versus late treatment of PDA.

Methods

A retrospective chart review of all infants less than 1,500 g diagnosed with PDA in the Coombe Women's Hospital between 1/1/04 and 31/10/06 was performed. Patients were excluded if they had a structurally abnormal heart or lethal congenital abnormality. Primary outcome was the need for surgical ligation. Secondary outcome measures included mortality, incidence of intraventricular haemorrhage, chronic lung disease, necrotising enterocolitis, retinopathy of prematurity and pulmonary haemorrhage. Information compiled included: weight and gestations of the babies, day of life of PDA diagnosis, treatment option used, timing of treatment, rate of successful closure, rate of surgical ligation and rates of morbidities of prematurity including death, IVH, CLD, NEC, ROP and pulmonary haemorrhage. Institutional Review Board approval was obtained.

Results

The overall incidence of PDA was 20.8% (69 cases) in < 1,500 g. The rate of surgical ligation was 3.6%. Nineteen were treated conservatively and 44 were medically treated with ibuprofen. Of the 44 babies treated with ibuprofen, 15 were treated by day 3 (early group) and 29 were treated later (late group). The

remainder (6) were either treated with indomethacin or ligated early. The median (range) weight and gestation of the conservative group was 1,120 g (650–1,570 g) and 28 + 3/40 (25 + 1 to 31 + 6); the early group was 800 g (540–1,335 g) and 27 + 6/40 (25 + 0 to 30 + 0); the late group was 920 g (490–1,500 g) and 27 + 2/40 (24 + 0 to 31 + 2).

Rate of successful closure in the conservative group was 87, 80% in the early group and 79% in the late group. The incidence of grade 3–4 intraventricular haemorrhage was 13% in the early group versus 31% in the late. There was no significant difference in mortality, the rate of chronic lung disease, necrotising enterocolitis, retinopathy of prematurity and pulmonary haemorrhage.

Conclusions

An early-targeted approach to the management of patent ductus arteriosus did not improve the rate of medical closure or reduce need for ligation, but did demonstrate a lower incidence of severe IVH. The significance of this warrants further investigation.

Kell isoimmunisation: a condition of increasing importance

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Introduction

The relative importance of Kell antibody has increased since the reduction in Rhesus isoimmunisation. Although uncommon it is potentially a serious problem for the affected fetus because in addition to haemolysis it also causes bone marrow suppression. Women may become sensitised following a blood transfusion.

Aims

There is very little Irish data on Kell isoimmunisation. In particular we do not have information on how frequently the fetus is affected. We carried out this study in order to establish the frequency of the condition and the spectrum of severity in those affected.

Methods

The records of the Blood transfusion department NMH were examined for the period 1984–2004 and those with anti-Kell antibodies identified. The case notes of any affected infants were subsequently examined

Results

During the study period there were 160,000 births. Of these there 97 women who were anti-Kell antibody positive. Among the 97 anti-Kell antibody positive pregnancies there were 9 affected fetuses. Of these nine affected fetuses the outcome was as follows: one intra-uterine death, two fetuses required intra-uterine transfusions, one required a top-up transfusion after birth, one infant had prolonged hyperbilirubinaemia requiring phototherapy, one infant had long-term neurological sequelae.

Discussion

This study gives an insight into the uncommon but important problem of Kell isoimmunisation. It is encountered in 1 in every 1,600 pregnancies. The data indicates that 10% of women with anti-Kell antibodies will have an affected fetus. In the 20 year epoch, nine affected fetuses were encountered. If this were extrapolated to the whole country it would suggest that we can expect three affected fetuses annually.

The recent introduction of Kell negative blood for girls and women in the reproductive age group will help to reduce this problem in the future.

Conclusion

This study highlights the problem of Kell isoimmunisation. As its relative importance has increased, those involved in perinatal medicine, both obstetrics and neonatology, need to have an understanding of its clinical significance and likely outcome.

Presentation of congenital heart disease in the neonatal period at the national maternity hospital

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Aim

The NMH provides routine antenatal ultrasound screening in addition to fetal ECHO in selected cases. Since March 2005, it also performs routine predischarge lower limb pulse oximetry on all infants. The purpose of this study was to determine when cases of congenital heart disease (CHD) are first suspected and what factors are helpful in supporting or confirming the diagnosis.

Methods

All cases of CHD diagnosed in inborn babies within the first 28 days of life from January 2003 toMarch 2007 were identified through the clinical information system. Infants with recognisable chromosomal and/or genetic syndromes were excluded from the analysis.

Background

Sixty-three cases of CHD were diagnosed, over a 4 year period of which four cases were excluded—Trisomy 18 [1], Trisomy 21 [1], Outborn infants [2]. Of the 59 cases, we have analysed 39 cases to date. 14 cases (36%) were identified antenatally. The diagnoses included

Transposition of Great Vessels [2], Hypoplastic Left Heart [4], Coarctation of Aorta [2], Complete AVSD [2], Ebstein Anomaly [1], Tetralogy of Fallot [1], VSD/ASD/PDA [2]. Of these infants 11 (79%) were transferred to OLHSC for further evaluation, 1 died prior to transfer (decision not to treat), and two were discharged home with cardiology OPD follow up, both of which were in the VSD/ASD/PDA group. Of the infants transferred, all were transferred by DOL 2. Thirteen (93%) of the 14 cases diagnosed antenatally were confirmed on postnatal ECHO. One case suspected antenatally to be a Hypoplastic Left Heart was subsequently diagnosed with a VSD/PDA/PFO. Eight (73%) infants were transferred on prostaglandin infusion and three were intubated on transfer. Nine (82%) had oxygen saturations >85% at the time of transfer, and the remaining two had oxygen saturations of 82% (HLHS) and 64% (complete AVSD). The latter infant died soon after transfer. Postnatally, a further 25 cases were diagnosed. The diagnoses here included Transposition of Great Arteries [4], Coarctation of Aorta [5], TAPVD [2], Critical Pulmonary Stenosis [2], Hypoplastic Left Heart [1], Dextrocardia [3], Ebstein's Anomaly [1], Aortic Stenosis [1], Tetralogy of Fallot [1] VSD/ASD/PDA [4], Complex Cardiac Disease [1]. Of these cases 19 (76%) were suspected prior to discharge. Twelve cases (48%) were suspected due to cyanosis. Six cases (24%) were detected due to respiratory distress. One case was suspected on the basis of mottling and poor perfusion. Three cases (12%) were detected on routine day 2 physical and one case was diagnosed on routine predischarge oxygen saturation monitoring. Of the six cases (24%) which presented post discharge, three had Coarctation, one had TGA, one had Aortic Stenosis and one had TAPVD. All of these cases presented in the era prior to routine predischarge pulse oximetry. Twenty-one (84%) of the postnatal cases had abnormal chest X-rays. Upper and lower limb BP was measured in all but one case but in no case did it suggest or alter the diagnosis. ECGs were performed in 21 (82%) of cases, 11 (52%) of which were reported as normal. The ECG was not found to be helpful in diagnosis. A hyperoxic test was performed in only one case.

Conclusion

36% of cases of CHD are now diagnosed antenatally and in all, but one case, the diagnosis was confirmed postnatally. A further 49% are diagnosed predischarge. The use of routine pulse oximetry helped to detect one infant with CHD that would not have been suspected clinically prior to discharge. 15% of cases were only identified after discharge but, interestingly, no late diagnosis has occurred since the introduction of routine predischarge pulse oximetry. CXR is invariably helpful in diagnosis but ECG or four Limb BP did not contribute much. Continuing audit of this area is important in an effort to minimise delayed diagnosis of significant heart disease.

Extubation failure and reintubation criteria in very low birth weight preterm infants in a tertiary neonatal intesive care unit: a retrospective study

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Introduction

Up to 30% of very low birth weight (VLBW) infants require intubation and mechanical ventilation. Although ventilator technology and monitoring of premature infants have improved over time, selecting the optimal time for extubation is challenging. Studies quote extubation failure rates of up to 30% in VLBW with RDS, rising to 40% in those weighing < 1,000 g. There are no established criteria for successful extubation, nor have the reasons for failure of attempted extubation been described in recent literature, despite the potential for adverse effects on the VLBW infant during such episodes.

Aim

To quantify the extubation failure rate among VLBW infants in our institution. Secondary aims were to describe the clinical indications for reintubation and to the role of interventions such as CPAP in preventing early reventilation.

Methods

A retrospective chart review was performed looking at eligible infants born between December 2005 and December 2006 at the National Maternity Hospital. Infants were eligilbe if they had a birth weight of ≤1,500 g, a gestation of ≤32 weeks and required mechanical ventilation over the first 48 h of life. Exclusion criteria included any severe congenital pulmonary or cardiac abnormality or withdrawal of care on ventilation. Background data on birth weight, gestational age, use of antenatal steroids, postnatal surfactant therapy and duration of ventilatory support were recorded. A diagnosis of extubation failure was made if infants required reintubation and ventilation within 48 h of the original trial of extubation. The subsequent level of respiratory support (e.g., CPAP

settings, use of a backup rate, etc.) and any co-morbidities were documented, as well as the clinical indication for reintubation where it occurred.

Results

Sixty eligible infants were identified. 26% ($n = 16$) required reintubation within the first 48 h. Of these, 43% ($n = 7$) were reintubated on multiple occasions. A further 16% of all babies ($n = 10$) were intubated at more than 48 h, most of whom deteriorated because of sepsis. 97% of included infants had been treated with caffeine, 95% were started on CPAP and of those 66%($n = 40$) had a backup non-invasive positive pressure rate set.

Of the 16 infants with early intubation failure, 11 had documented severe apnoea and 8 were hypercarbic (range of PCO_2 from 8 to 12 kPa, their average pH was 7.16 (range 7.0–7.3). Four had a clinical diagnosis of increased work of breathing and three were reintubated for lack of respiratory effort. Five infants had a combination of the factors listed above.

The average duration of mechanical ventilation before attempted extubation was 188 h in infants who subsequently failed and 127 h in those who were succesfully extubated. The average time to reintubation was 4 h in those who failed. Measures instituted in an attempt to avoid reintubation included increasing the PEEP, FiO_2 and back up rate on CPAP.

Conclusion

This study found a 26% reintubation rate among extubated VLBW infants. Severe apnoea, hypercarbia (with acidosis), poor respiratory effort and increased work of breathing were the clinical indications for reintubation. Severe apnoea was the commonest reason for reintubation. Most infants who failed were reintubated within the first few hours following extubation. This study also demonstrated that those who failed extubation were on mechanical ventilation for a longer period of time. Predicting the optimal time to extubate premature VLBW infants remains a challenge in the neonatal intensive care unit.

Patient misidentification in the neonatal intensive care unit: quantification of risk. a comparison between data from Boston (USA) and Cork Unified Maternity Services (Cork, Ireland)

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Objective

To look at the potential risk of misidentification among patients in Cork Unified Maternity Services NICU resulting from similarities in patient name or hospital record numbers (MRNs) and compare results with a NICU in Boston.

Methods

A list of all patients who received care in the NICU in Cork during 1 calendar year (2005) was obtained from the NICU Badger system.

A patient day was considered at risk from misidentification errors when the index patient either shared a surname, had a similar sounding surname or similar MRN with another patient also in the NICU on that day.

Results

During the 1 year study period, 6,282 days of patient care were provided to 613 patients in the 26 bedded NICU. Only 7 calendar days were free of risk from patient misidentification errors in 2005 once similarities in patient identifiers due to multiple births were excluded and only 2 calendar days were free of risk before these multiple births were excluded.

The most common cause for potential misidentification were similar appearing MRNs, of which there were 269 patients at risk due to this in 1 year, once multiple births were included this figure dropped to 207. One hundred and twenty-nine patients were at risk of misidentification due to having the same surname as another patient on the NICU, once multiple births were excluded this figure dropped to 22. Thirteen patients were at risk due to similar sounding surnames. Multiple births contributed 17.46% of patients and 27.1% of days of patient care in the NICU. Also data was collected to compare the number of NICU admissions with both native Irish and non-native Irish names. There were 98 patients with non-native Irish names out of 613 patients, this means that 15.99% of NICU admissions in 2005 had non-native Irish names.

Conclusion

Patients cared for in the NICU in Cork are at a high risk of patient misidentification which could lead to dangerous errors occurring. Even after multiple births have been excluded from this data the risk is still significantly high. As similar MRNs seems to produce the most substantial risk to patients maybe a different method of assigning MRNs to patients should be considered to remove this risk.

Paediatric tuberculosis in the midwestern region from 1997 to 2006

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Aims

To present the findings of a 10 year review of paediatric tuberculosis (TB) in the Mid-Western region.

Methods

A retrospective case note analysis of patients aged 0–16 years diagnosed with active or latent TB in the Mid-Western region between 1997 and 2006. There is an estimated population of 100,000 children in the Mid West area. Patients were identified by the HIPE discharge system, the department of public health database and the microbiology laboratory database for positive culture results.

Latent TB was defined as a positive mantoux test and no clinical features or chest radiograph changes.

Active TB was defined as positive mantoux test with clinical features and/or chest radiograph changes and/or microbiological results suggestive of TB.

Positive mantoux test was defined as per the guidelines from American Academy of Paediatrics.

Results

A total of 24 patients were identified by the above methods. Twelve children were diagnosed with active TB in the past 10 years. There were only two children diagnosed with latent TB within the hospital system. It was not possible to ascertain the incidence of latent TB in the community as these children are often treated by individual medical officers and not identified on a database.

Of the patients with active TB there was a 2:1 male to female ratio. The age at diagnosis ranged from 2 years 4 months to 13 years with a median age of 6.5 years. 75% of these children received the BCG vaccine. 66% (8) of the subjects had contact with a TB infected family member. Symptoms of pulmonary TB were the most prevalent (50%) but lymphadenitis, erythema induratum and systemic symptoms such as fever, night sweats and irritability were also present. There were no cases of military TB or meningitis. Two patients were asymptomatic but had abnormal chest radiographs and strongly positive mantoux tests. The mantoux test was positive in all subjects and greater than 15 mm in all with symptoms. No early morning gastric aspirate or sputum cultures were positive for acid fast bacilli and the only positive laboratory investigation was a lymph node aspirate. The recently developed whole blood interferon-gamma release assay test was not used in any patients. Chest X ray was reported as abnormal in 50% of cases.

Conclusion

Despite the limitations of this retrospective review, a number of conclusions can be drawn: (1) Paediatric TB is uncommon within Mid-Western Region and in the majority of cases there is a history of contact with an infected family member. (2) Most cases of active TB had been immunised with the BCG vaccine at birth. (3) Diagnostic confirmation of TB in children is challenging but the whole blood interferon-gamma assay may be helpful with the diagnosis of latent TB. (4) A national database of all patients with TB is essential for accurate surveillance and ongoing management of these children.

Can neonatal interhospital transport be minimised?

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Introduction

Many tertiary and quaternary neonatal units in Europe and USA are equipped with all necessary imaging modalities and medical subspecialty teams to manage neonatal surgical and medical complications. Neonatal transfers are associated with increased neonatal morbidity and require the involvement of experienced neonatal transport teams.

Aims

We aimed to study the number and nature of neonatal transfers from a tertiary neonatal unit over a year.

Methods

All neonatal transfers from National Maternity Hospital between January and December 2006 were included. Both nursing records of neonatal transports and our central computer system for data on neonatal transports were used. Patient records were reviewed for patient demographics, hospital to which infant was transferred and reason for transfer either from the nursing notes or review of patients charts.

Results

One hundred and ninety-one infants were transferred to other units during this time period (15% of total admissions to Neonatal Unit during this time period). One hundred and six (55%) infants were transferred to a tertiary paediatric hospital. Thirty-three infants required transfer for surgical conditions while 19 were transferred for Cardiology review/investigations. Seventeen transfers were made for MRI while eight babies needed barium studies. Other less common reasons for transport being ENT 5, Ophthalmology 3 and 4 infants were transferred to adult hospital for CT Purposes. Nine infants were trans-

ferred on ventilator support and 10 infants transferred were less than 1 kg birth weight. 67 infants were transferred back to peripheral neonatal units once stabilized and two infants were transferred to Sweden on ECMO.

Conclusion

Significant numbers of neonates are transferred to other units for cardiology or surgical review, imaging and other investigations. Interventions to reduce neonatal transport by providing more essential investigations and treatment modalities to neonates in the tertiary or quaternary neonatal unit need to be explored.

Barriers to discharge from transitional care in a tertiary paediatric hospital

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Introduction

Advances in neonatal and paediatric intensive care have improved mortality and increased the number of children requiring longterm ventilation. However the intensive care environment is unsuitable for the long-term developmental needs of children and resulting in the creation of transitional care units (TCU) to cater for the needs of children and their families.

Methods

Retrospective observational cohort study of children in the TCU of a tertiary care paediatric hospital.

Results

Children (864) were admitted to the Intensive Care Unit in 2006 and 12 were ultimately transferred to TCU. We excluded three patients from the study who were admitted because of critical ICU bed shortages. The age at transfer to TCU ranged from 3 months to 18 years. Of the nine eligible patients the diagnoses were as follows: Quadriplegia post Hib Meningitis, congenital Hypoventilation Syndrome, Ex 27/40 with severe BPD, Relapsed Medulloblastoma and GB type Polyneuropathy, Diabetic Embryopathy, Congenital tracheal stenosis, Marden Walker Syndrome, two children have no specific diagnosis to date despite extensive investigation. All patients were managed by a multidisciplinary team including an average of eight specialist consultant teams and at least four therapists' teams as well as nurses and health care assistants. Average length of hospital stay was 15.5 months. Four patients have been discharged from TCU, two of these following 1 year of discharge planning and two were readmitted for RTI. Two of the patients discharged are cared for at home, one is in a Rehab unit and the other in another district general hospital. Children spent up to 10 months in hospital after funding was approved at a significant cost. Major barriers to discharge were delays in funding and necessary staffing for home care. In addition the lack of resources and staffing for ventilated children delayed transfer to local hospitals.

Conclusion

The major barriers to discharge are related to delays in funding and lack of community support services. Streamlining funding and improving community care structures are vital to enhance the discharge process.

First febrile convulsions: unnecessary hospital admissions?

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Background

In Northern Ireland all children with first febrile convulsion are admitted to hospital. Reasons for admission vary from historical, to parental anxiety and concern regarding underling clinical diagnosis. This practice varies from current practice throughout the UK and Ireland. This audit looks at admissions over a 1-year period with a view to changing practice.

Aims

To look at all hospital admissions with febrile convulsion over a 1 year period in order to review current practice, study patient outcomes and to make decisions regarding future management protocols.

Methods

Retrospective case note analysis of all children coded as presenting with febrile convulsion to the emergency department, RBHSC in the time period 1 September 2003 to 31 August 2004 were obtained. Clinical information was gathered from current care pathway.

Results

One hundred and fifty-five children were admitted with first febrile convulsion. There was a slight male predominance with the median age of admission 20 months. There was a positive family history in 30%. The median temperature on arrival was 38.7°C. Blood sugar was recorded on arrival in 92% of cases with median BM 6.6. Median seizure duration was 5 min with 72% described as tonic clonic in nature.

57% of children had bloods taken during the admission with MSSU being performed on 86%. Anticonvulsants were required in 12% of cases prior to or on arrival to A&E. 98% of children received antipyretics,

41% were discharged home on antibiotics. Median admission duration was 1 day with URTI being the commonest diagnosis (65%)There were no cases of meningitis or septicaemia. There were no PICU admissions and no deaths.

Conclusions

Admission policies will be reviewed and an alternative care pathway derived following this study. Results indicate that it is not necessary to admit all children presenting with first febrile convulsion. Children who have a confirmed clinical diagnosis and in whom there is no parental anxiety and adequate education could be discharged home after a period of observation.

There were no cases of meningitis or septicaemia presenting as first febrile convulsion with the most common underlying diagnosis being URTI.

References

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Postural orthostatic tachycardia syndrome (pots) in children: a regional review of confirmed cases

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Aims

1. To describe the incidence, physiological characteristics and haemodynamic profiles of all children aged 16 years and under, diagnosed with Postural Orthostatic Tachycardia Syndrome using tilt-table testing in the Mid-Western region of Ireland.
2. To highlight POTS as a differential diagnosis in the evaluation of fainting (syncope), palpitation, pre-syncope and related non-convulsive paroxysmal events in children.

Method

A retrospective review of all patients aged 16 years and under attending the syncope laboratory for prolonged tilt-table testing over a 6 year period. Data was compiled from a customised database (Filemaker Pro). Diagnostic criteria required the presence of a persistent tachycardia (either an increase from baseline of up to 30 beats per minute (bpm) or an increase in heart rate of up to or greater than 120 bpm within the first 10 min of head up tilt, associated with symptoms of orthostatic intolerance in the absence of significant hypotension, as defined by Grubb et al. (2006). The unit policy requires all patients to have structural cardiac abnormalities ruled out prior to undergoing prolonged tilting.

Results

Over a 6 year period 56 children were referred for prolonged tilting (M = 45%, F = 55%). A significant proportion of these children had extensive neurological and cardiac investigations prior to referral for tilt test. In all children referred, syncope was cited as a presenting symptom. 45% were diagnosed with POTS after tilt-table testing. A female preponderance was identified (male:female ratio = 41:59%). The average age of diagnosis for females was 15.2 years and for males was 15.3 years, (mean age 15.3 years). 37% patients diagnosed with POTS had concomitant Neurocardiogenic Syncope (NSC) diagnosed of tilting. 21% of all patients had normal tilt-tests. The average height and weight for POTS patients was 170.5 cm (M = 179 cm, F = 164 cm) and 61 kg (M = 70 kg, F = 55), respectively compared to 165 cm and 57 kg, respectively in the non-POTS group. The average heart rate was 110 bpm after 30 min of active tilting was compared to 95 bpm in those not diagnosed with POTS and 87 bpm after prolonged tilting compared to 85 bpm in the non-POTS patients.

Conclusion

Even though well described in adults, the epidemiology and clinical characteristics of POTS is perhaps under-reported among children. The presenting symptoms can often be misinterpreted as symptoms of other cardiovascular and neurological conditions leading to misdiagnosis and unnecessary medication, a situation which we identified within our study group. The childhood population of the Midwestern region being 100,000, our study proposes the first reported Irish prevalence of confirmed POTS in children. We hope that this study will raise awareness of the diagnosis and thus prevent affected children undergoing unnecessary investigation and inappropriate treatment.

Investigation of global developmental delay

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Guidelines were produced in 2006 to assist in the investigation of global developmental delay. These guidelines were compiled from the best available evidence—mainly grade III and IV studies

Aims

- To audit the investigations undertaken in patients presenting to the Paediatric Developmental Outpatient Service with Global Developmental Delay

- To compare these results to the best available Evidence based guidelines formed on this topic. Arch Dis Child 91:701–705 (2006)

Methods

A retrospective chart review was performed on all patients diagnosed with global developmental delay who had their first presentation to developmental paediatric out patient services during the time period 1st August 2005–30th September 2006. Clinical information compiled included family history, dysmorphic features and neurological signs. All investigations performed and their results were recorded.

Results

Thirty-five patients with global developmental delay presented during the study period. Two of these had a known cause for developmental delay—prematurity with interventricular haemorrhage and congenital CMV. Of the remaining 33 patients three were diagnosed with a likely cause for the developmental delay. In all three chromosomal analyses were abnormal revealing one 10 q deletion and two unbalanced translocations. These three children had dysmorphic features. A further child was diagnosed incidentally with cystinuria. A total of 189 investigations were performed. First line investigations performed included karyotype, fragile X, metabolic screen and organic acids, FBC, urea and electrolytes, calcium, liver and thyroid function tests. Further investigation including FISH for 22Q11 and William's syndrome, CK, lactate, carnitine, biotinidase, transferrins, radiology including MRI brain and EEG were performed on some children

Discussion

First line investigations as recommended are very similar to what was performed but would also recommend routine screening for lead levels, CK, ferritin and Biotinidase. The usefulness for routine screening of lead levels in Ireland is questionable. The guidelines only recommend metabolic screening if there is family history, consanguinity, regression, organomegaly or coarse features. In the Irish population the incidence of metabolic conditions is higher therefore it is reasonable to include the metabolic screen as part of the first line.

Despite relevant investigations 85% of the patients have no identifiable cause for the global developmental delay.

Conclusions

The available UK guidelines are useful as a guide to the investigation of global developmental delay. Further study in this area is necessary to develop guidelines relevant to the Irish population.

Child obesity and child protection

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Introduction

There is increasing recognition that there is an "obesity epidemic" both in Ireland and worldwide, with an increase in associated comorbidities such as type 2 diabetes mellitus, hypertension, cardiovascular disease, arthritis, dyslipidaemia and sleep apnoea. Treatment with a multicomponent regime involving diet, exercise and behavioural change agreed between medical staff and family is prescribed. Does the family's inability or unwillingness to implement the essential changes in diet and lifestyle constitute a serious child protection concern and demand the appropriate response? The literature is unhelpful in this regard. We present three cases to demonstrate child protection concerns and interventions in grossly overweight children.

Case reports

Case 1: A 14 year old boy admitted in June 2005 as the community services were concerned about his morbid obesity and risk of sudden death. He is the eldest of three brothers, living with parents (also obese), attends a special needs school and functions in the low range of intellectual ability with little speech, communication or social skills. Community services had for years been unable to effect change in his diet and lifestyle which was marked by "junk" food, "addiction" to TV, cartoons and video, and absence of exercise.

Admission weight was 131.6 kg, BMI 51.4, BMI SDS +4.14. On supervised diet and exercise regime in hospital he lost 11 kg in 11 weeks. Parents continued to demonstrate no insight as to the cause of his problems. Following extensive discussions with the multidisciplinary team and community services he was discharged to foster care where he continues to show dramatic weight loss and major improvements in behaviour, social and emotional development. He remains in foster care, with home visits and appears happy.

Case 2: Brother of case one, 13 years, also obese with more severe learning, behaviour, and communication problems, he has no speech and negligible social skills. He was admitted because of refusal to weight bear and was found to have compression fractures of dorsal spine and osteoporosis/osteomalacia. Biochemical profile was normal but vitamin D levels were low—27 nmol/l (range 53–150). Further inquiry revealed that his

diet was largely bread, butter and crisps and similar to his brother there was no exercise and constant exposure to TV, cartoons and video. Admission weight was 70 kg, BMI 35.2, BMI SDS +3.63. On supervised diet, vitamin D and calcium treatment, and increasing exercise he regained mobility and lost 4 kg in 8 weeks. Behaviour and eating habits improved, ''screen addiction'' lessened, though to date there is no improvement in communication ability. Discharge is planned with a Court supervision order.

Case 3: 7-year old girl, from the travelling community, now settled. She was admitted to hospital as an emergency because of morbid obesity, daytime somnolence and sleep apnoea. Admission weight was 50.6 kg, BMI 32.4, BMI SDS +4.51. Emergency adenotonsillectomy was required and a period of night time BIPAP. Parents are overweight, father morbidly so and it is likely food ''rewards'' were used at home to mange tempers and behaviour.

Current management involves the hospital and community multidisciplinary teams, with regular meetings involving the family. Improvements have been noted—dietary change, more exercise, absence of further weight gain and family insight. Continuing close supervision, including social work is planned.

Conclusion

The three cases presented may be ''extreme'' but demonstrate that obesity is now a major malnutrition in Ireland and that child protection measures may be needed to insure the child's immediate and longer term survival and well being.

IV Rehydration in the paediatric emergency department

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Abstract

Aims

To benchmark our current practise for preadmission IV rehydration in the paediatric emergency department and to determine subsequent rates of reattendence.

Methods

This restrospective study incorporated all children under the age of 14 years commenced on IV rehydration, attending an inner city paediatric emergency department in a Tertiary Referral centre between 1/11/2006 and 30/11/06. The decision to commence IV rehydration was at the discretion of the treating physician. Prehydration weight, vital signs, baseline renal function and blood glucose were recorded. As per hospital protocol, 12 h maintenance fluid requirements were calculated according to the patient's weight. This total was then infused as a solution of 0.45% Saline and 2.5% Dextrose over 3 h.

Results

Of the 4,109 attenders to the Emergency Department between 01/11/06 and 30/11/06 inclusive, 62 patients (33 male: 29 female) were admitted for IV rehydration. Of those admitted 37 (60%) were less than 3 years old. The most common reasons for attendance were: Persistent Vomiting 39 (63%), and Diarrhoea 19 (30%), and a short history of refusing oral intake 4 (6%).53 patients had a renal profile performed prehydration, and 17 (32%) had a urea above the normal reference range. Fifty-five patients (88%) were discharged home directly from the Emergency Department. Of these, nine (14%) subsequently reattended with a similar complaint, and one of whom was subsequently admitted. There were no adverse events documented.

Conclusions

Preadmission IV rehydration in the paediatric Emergency Department is a safe, acceptable and effective alternative to in-patient admission in patients presenting with mild to moderate dehydration.

Audit of the emergency management of status epilepticus (SE)

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Objectives

1. Characterise the clinical features of patients presenting with SE to OLH.
2. Outline the pre-hospital management of seizures.
3. Audit the emergency management of Status Epilepticus (SE) in hospital.
4. Compare the results of those needing ICU care versus ward care.
5. Provide a framework for the development of strategies that will decrease the requirement for intensive care.

Design

Retrospective chart review.

Methods

Ten admissions to ICU, over a 12 month period, with SE were identified from an ICU database and 10 other admissions needing ward care were identified from a neurology database. There were 7 patients with 10 admissions to ICU and 9 patients with 10 admissions to the wards.

The clinical and demographic data were obtained from the charts. Treatment of SE was compared with the hospital SE protocol.

Results

	ICU admissions	Ward admissions
Previous diagnosis of epilepsy	1/7	2/9
Previous history of febrile seizures	2/7	3/9
Pre-hospital duration of seizures: mean	32 min (5–70)	37 min (5–120)
Pre-hospital seizure management	1/7	3/9
Ambulance as mode of transport	7/10	6/10
Treatment as per SE protocol	2/10	6/10
>2 doses of benzodiazepines given	8/10	5/10
Phenytoin given in first 30 min	3/10	3/7

Summary

Only 20% of patients had treatment as per the protocol in the ICU group versus 60% in ward group. More than 2 doses of benzodiazepines were administered in 80% of patients in ICU group versus 50% in the ward group. Pre-hospital management was undertaken in 14% of patients in ICU group versus 33% in the ward group.

Conclusions

1. Ambulance crews managing acute seizures in children should be permitted to administer rectal or buccal benzodiazepines.
2. There is a need to educate all doctors treating SE to adhere to SE management protocol.
3. The administration of intravenous phenytoin should be mandatory once 2 doses of benzodiazepines have been given.

Redefining markers of a haemodynamically significant patent ductus arteriosus in preterm infants. A prospective observational study

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Background

Determining the significance of a patent ductus arteriosus (PDA) remains difficult in preterm neonates and current echocardiographic markers of ductal significance give little information about ductal steal. We hypothesized that the relationship between celiac artery flow (CAF) and left ventricular output (LVO) in the presence of a PDA may determine ductal significance.

Objectives

To examine CAF to LVO ratio (CAF:LVO) in the presence and the absence of a PDA in preterm neonates.

Methods

This was a prospective observational study of neonates < 1,500 g. Echocardiography was performed at 12 h and day 3. PDA was identified and LVO and CAF were measured by echocardiography. The infants were divided in to those who developed a significant PDA and controls. Following successful PDA treatment, a further assessment was carried out in the two groups.

Results

A total of 33 infants were enrolled. Nineteen infants had a PDA (Median gestation 27 weeks, birth weight 915 g), and 14 controls (gestation 28.7 weeks, birth weight 1,110 g). At 12 h, there was no difference in CAF, LVO or CAF:LVO. On day 3, there was a statistically significant difference in CAF:LVO between PDA versus control groups (Fig. 1). CAF:LVO significantly correlated with conventional markers of ductal significance. Following successful PDA treatment CAF:LVO returned to levels similar to controls.

Conclusion

CAF:LVO may be a useful marker of ductal significance. It may be used to determine which PDA warrants treatment and serve as a marker of treatment success. It may provide an accurate quantitative assessment of ductal steal in the presence of a PDA, and improve understanding of a physiologically significant PDA.

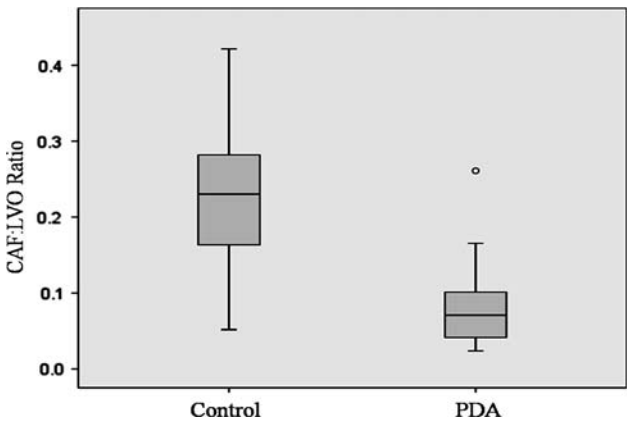

Fig. 1 CAF:LVO ratio is significantly lower in the PDA group compared to controls on day 3 (0.07 vs. 0.21, $p < 0.0001$); CAF:LVO: celiac artery flow to left ventricular output ratio

Predictive value of mutation analysis for glutaric aciduria type 1. Helping to prevent dyskinetic cerebral palsy

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Background

Glutaric aciduria type 1 (GA1) is a rare, lifelong, life threatening, recessively inherited disorder of amino acid metabolism. This affects breakdown of the amino acids lysine and tryptophan which are impaired by deficiency of the enzyme glutaryl CoA dehydrogenase. Diagnosis is suggested by finding elevated levels of the metabolites glutaric acid, glutaconic acid and 3 hydroxyglutaric acid in urinary organic acid analysis and raised glutaryl carnitine on acylcarnitine profiles. The diagnosis is confirmed by detecting low glutaryl CoA dehydrogenase activity in skin fibroblasts. Treatment consists of excluding lysine from the diet and consuming low levels of tryptophan and carnitine supplementation.

Presentation is usually from 6 months to 2 years. Dyskinetic cerebral palsy is often the presenting feature and usually occurs following a viral infection which has put the child in a catabolic state. Other features include encephalopathy, aphasia, macrocephaly and subdural effusions

Aim

GA1 is known to occur in the Irish Traveller community. We aimed to look at the genotype and phenotype correlations that occur in the Irish Traveller community and to assess complications.

Methods

All Irish patients with a diagnosis of GA1 were identified from the NCIMD database. Case notes and family pedigrees were reviewed and the mutation, if known, recorded.

Results

There are currently 13 living cases of GA1. Six of these 13 patients are from the Irish Traveller community. These six patients are from five families. All have severe forms of GA1, ie high excretion of metabolites at the time of diagnosis. Three of these families have had mutation analysis testing. All are homozygous for the E365K mutation. Although different surnames, all six patients are part of the same extended family.

Complications encountered in these patients include axial hypotonia in one patient and mild developmental delay in two patients.

Conclusion

Testing for GA1 currently occurs on clinical suspicion after the child has presented with one of the devastating complications of the condition, in particular dyskinetic cerebral palsy. Once damage has occurred, it is irreversible. Prevention offers a favourable outcome. Knowing the mutation of GA1 in Traveller families allows for high risk screening, targeted screening and early intervention.

An audit of referrals to a genetics clinic for the investigation of developmental delay

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Aim

To ascertain the number of individuals, both children and adults, referred to a consultant clinical geneticist for the investigation of developmental delay in which a diagnosis was made, and to describe the diagnoses.

Methods

All consecutive case notes from patients referred to the genetics clinic for the investigation of developmental delay over a 2 year period (2004–2005 inclusive) were reviewed. These cases were all seen by one Consultant Clinical Geneticist in multiple centres (Crumlin, Temple Street and Cork). Exclusion criteria included individuals with deafness, isolated cleft palate and skeletal dysplasias; neonatal referrals were also excluded.

Results

A total of 119 individuals ($n = 119$) were referred and evaluated, of which 51/119 were male (42.86%) and 68/119 were female (57.14%). The age range at the time of the first review at the genetics clinic was from 0.17 to 37.25 years. A diagnosis was reached in 35/119 (29.41%) of the cases referred. Of the 35 diagnosed cases, 14/35 (40%) were male and 21 (60%) were female. 22/35 (62.86%) were dysmorphic and 13/35 (27.14%) were non-dysmorphic. A new diagnosis was reached in 20/35 (57.14%) cases and confirmation of a diagnosis made in 15/35 (42.86%) cases. In 5/119 (4.20%) cases, we removed a previous diagnostic label. No diagnosis was made in 84/119 (70.59%) cases. The diagnosis made most commonly was a chromosomal disorder (4 cases) and Noonan syndrome (4 cases), followed by teratogenic syndromes (total of 3 cases) such as fetal alcohol syndrome (2 cases) and fetal valproate syndrome (1 case). Rare diagnoses included Cohen syndrome in a

child from the Irish Traveller population which led subsequently to 4 other children being diagnosed with this same condition. Of the total study cohort (n = 119), 63/119 (52.94%) were dysmorphic and 56/119 (47.06%) were non-dysmorphic. We were more likely to make a diagnosis in individuals with dysmorphism (22/63 = 34.92%) compared to those without dysmorphism (13/56 = 23.21%).

Conclusion

Our diagnostic pick-up rate for individuals referred to the genetics clinic for the investigation of developmental delay is similar to other studies from international genetics centres. Referral to the genetics clinic for the investigation of developmental delay can be helpful, particularly in those children and individuals with dysmorphism.

Is one A&E department adequate to meet to meet the needs of the greater dublin area?

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Background

The HSE is planning the development of a single tertiary Paediatric Hospital in Ireland. The tertiary centre would provide care for all the secondary needs of Greater Dublin. The exact plans of the HSE regarding A&E services and urgent care centres has not yet been clarified, but it is unlikely that all centres would be staffed on a 24 h basis as out of hours staffing has major financial implications. It is possible that the A&E department in the new National Children's Hospital would be the only centre operating from midnight to 8 a.m.

Aim

Our aim was to assess the patient profile, investigation/intervention rates and admission rates in those attending the A&E Department in Temple Street between midnight and 8 a.m in order to assess the likely demands on a single A&E centre in terms of staffing and other resources. We were also interested in the impact the recently developed after-hours GP service might have on Paediatric A&E attendances in our inner-city centre and whether or not the caseload could be managed in an 'out of hospital' setting.

Methods

Two 1 week periods were assessed (Nov 13–19, Dec 11–17). Week 2 coincided with the commencement of the DubDoc GP out of hours service.

Symphony, the A&E computer programme and database was used to identify patients attending the department between the hours of midnight and 8 a.m. during these weeks. A&E doctors' notes, patient medication records and patient demographics were examined for each patient. We excluded those attending for prearranged clinics in the Department.

Results

During the 2-week period, the total number of attendances from midnight to 8 a.m was 171(male = 112, female = 59). (In week 1, this represented 8.8% of the total daily attendance rate (79/895), and in week 2 it represented 10.5% (92/875). 91.8% of all attendances were self-referrals. GP referral rates were 5% (4/79) and 2.1% (2/92), respectively. The total number of children who presented via the ambulance services during the 2 week period was 33 (19.29% of the total). We found that 56% (96/171) of patients attending the department required only physical examination with no other intervention. Interventions included X-rays (9.94%, 17/171), IV fluids (4.67%, 8/171), Nebulised medications (16.3%, 28/171) and minor procedures egglue (0.05%)/ neuro-observations (1.1%). One child presented in cardiopulmonary arrest during the 2 week observation period.10.5% of total attendances were admitted (18/171) in the 2 week period. The majority were discharged with no further follow-up (77.8%, 133/171).

Conclusion

Based on this 2 week study, it can be seen that the number of children attending A&E overnight is variable, from an average of 10 children per night to 11.5 per night. The majority of overnight attenders were discharged with no medical intervention, and a significant number required care which could be delivered in a non-tertiary setting, e.g., nebulised medications. The DubDoc initiative did not appear to affect either GP referral rates or attendance numbers—however this must be interpreted in the context of lack of public awareness of the service in its early stages. The majority of children were brought to A&E by private means, which would imply that the location of overnight services may not be relevant. To fully assess whether a single A&E service is feasible in Dublin city, attendance and intervention rates from all three Paediatric centres must be examined.

A high diagnostic yield in children attending a metabolic centre with developmental delay

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Aims

Inborn errors of metabolism (IEMs) are reported to be a rare cause (1–5%) of developmental delay, (Shevell et al. 2003). The aims of this study were to determine the etiological yield of metabolic investigation in children referred to a tertiary metabolic service when developmental delay was a significant feature in the presentation, and to determine the variables that make achieving a diagnosis more likely.

Methods

Cases were identified retrospectively from the database at the National Centre for Inherited Metabolic Disorders over a period of 3 years (Jan 2004–Dec 2006). All children 0–16 years of age referred with developmental delay or when developmental delay was a significant feature at referral were included in the study. Details of patients were obtained from case notes. Referral information, history, examination findings, investigations and diagnoses were studied.

Results

One hundred and six cases (64 male, 42 female) with an age range between 0 and 176 months were identified during the 3 year study period. The source of referrals was from General Paediatricians (55.7%), Paediatric Neurologists (34%), General Practitioners (2.8%) and others (7.5%). Of the 106 cases, 40 (37.7%) had a proven definitive metabolic diagnosis, including 8 (7.6%) that had a diagnosis at referral. A metabolic diagnosis of mitochondrial respiratory chain defect was probable in a further 19 patients. The definitive diagnoses included: Mitochondrial respiratory chain disorder (20), SCAD (1), SCHAD (1), Organic acidurias (3), Aminoacidopathies (4), Lysosomal disorders (2), PDH deficiency (2), UMP deficiency (1), Menkes disease (1), late diagnosed Galactosaemia (1) and other genetic syndromes (4).

Variables associated with a higher diagnostic yield included referral from a Pediatric Neurologist (47% yield of investigation versus 29% in General Paediatrician group), microcephaly ($p < 0.05$), abnormal eye examination ($p < 0.05$), and biochemical markers including an elevated serum lactate ($p < 0.05$), abnormal plasma amino acids ($p < 0.0005$) and abnormal urine organic acid analysis ($p < 0.05$). We noted a higher diagnostic yield when autism was noted as a feature than comparable studies.

Conclusion

We conclude that the diagnostic yield of metabolic investigation of patients referred with developmental delay in our study group (at least 37.7%) is significantly higher than observed in other studies, probably reflecting our documented very high prevalence of IEMs in Ireland (birth prevalence of at least 1:500 births for 2006) and high referral rate. Achieving a metabolic genetic diagnosis has important implications for the appropriate management, counselling and treatment of treatable metabolic genetic disorders in children presenting with developmental delay. This study also highlights the need for appropriate testing and for guidelines for investigation that reflects our specific population.

Eosinophilic esophagitis from a limerick perspective

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Aims

1. To present the incidence and clinical features of eosinophilic eosophagitis diagnosed at Limerick from 2004 to 2007
2. To discuss the clinical characteristics and endoscopic features of this recently recognised condition.

Background

Eosinophilic esophagitis (EE) is an emerging diagnosis in both paediatric and adult medicine. It has been described as ''asthma of the gullet'' and the histological hallmark is accumulation of eosinophils within the oesophagus. Symptoms include abdominal pain, vomiting and dysphagia and it typically presents in boys with a history of food allergies and asthma. Diagnosis is by typical endoscopic appearance (Figs. 1—3) and biopsy of the oesophagus with >20 eosinophils per HFL being diagnostic (Figs. 4, 5). The mainstay of management is swallowed corticosteroids and there is some evidence that elimination diets are beneficial. It is important to diagnose and treat this condition as progression to oesophageal stricture is a recognised complication.

Methods

A retrospective case review of confirmed cases of EE between November 2004 and March 2007.

Results

Seven children were diagnosed with EE. The majority (6) were male and all were aged between 9 and 14 years at the time of diagnosis. The presenting symptoms were abdominal pain, indigestion, vomiting

and dysphagia. In all cases except one there was a history of atopy, in particular asthma. The findings at endoscopy were all typical of EE with longitudinal fissures or circular rings and the diagnosis was confirmed by biopsy in all but one case. There was associated *H. pylori* gastritis in two cases and a non-specific gastritis in one patient. Four patients were treated with swallowed corticosteroids. Five patients are asymptomatic on no medication and two patients have ongoing symptoms.

Conclusion

EE is an increasingly recognised paediatric disorder and should be suspected in children with unexplained abdominal pain, dysphagia or vomiting and a past history of allergy.

Management of acute asthma in children: should ireland be in the space age?

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Aims

To determine the effect of implementing an evidence-based clinical practice guideline, for the inpatient management of children with acute exacerbation of asthma needing hospitalisation in a regional paediatric department.

Methods

A retrospective analysis including patients aged 2–16 years referred to the Paediatric Department, CUH with an acute asthma exacerbation. Data were collected for identical 2-month seasonal periods before (2005) and after (2006) implementation of departmental asthma education, admission proforma, clinical practice guideline with emphasis on use of spacer delivered bronchodilator therapy and specific discharge criteria. Cases were identified via the Hospital In-Patient Enquiry (HIPE) system and supplemented by review of ward admission data. Two researchers extracted the relevant data and reviewed patient charts. Institutional approval was obtained prior to commencement.

Results

The number of referrals with acute exacerbation of asthma needing hospitalisation had increased in 2006 (52) compared to 2005 (35). Most patients were referred by GP (31/35 in 2005, 39/52 in 2006) between 5 p.m. and 8 a.m. or on weekends (25/35 in 2005, 37/52 in 2006). The majority of patients had known asthma and were on inhaled corticosteroids (18/35; 2005, 39/52; 2006). There was a raised threshold for admission after (2006) implementation with 11/52 patients having mild exacerbations, 33/52 moderate and 8/52 severe compared to 2005 when 21/36 admissions were mild. Accordingly, a greater proportion of admissions in 2006 (47/52) met international admission criteria than in 2005 (17/35). Assessment of asthma control was complete in a greater proportion of cases using admission proforma: (40/52) in 2006, (5/35) in 2005. Duration of admission was significantly less in the post-implementation group for equivalent exacerbation severity, e.g., for mild (30 h; 2005, 22 h; 2006), moderate (28 h; 2005, 23 h; 2006), severe (50 h; 2005, 46 h; 2006). Average time between being fit for discharge and actual discharge was longer pre-implementation of specific discharge criteria; 20 h in 2005 compared to 5 h in 2006. Despite shorter length of stay in 2006, there were no re-admissions within 14 days. Duration of bronchodilator therapy was shorter in 2006 and more likely to be given by spacer device earlier for equivalent levels of severity, e.g., for moderate exacerbations, in 2006: average length of $\beta 2$ agonist therapy: 18 h with 12 h by spacer device, in 2005: average length of therapy: 25 h with 3 h by spacer. While the rate of administration of oral steroids was similar pre (85%) and post (89%) guideline implementation, the average time to administration was significantly faster in 2006 (56 min) than in 2005 (227 min). There was an improved documentation of asthma education in 2006 (e.g., inhaler technique reviewed in 37/52 in 2006, 21/35 in 2005 and action plan given in 23/52 in 2006, 7/35 in 2005) but this needs further improvement.

Conclusion

With appropriate education, an evidence-based clinical practice guideline with emphasis on use of spacer delivered bronchodilator therapy for children with exacerbations of acute asthma decreases their duration of hospitalisation without increased re-admissions. Reduction in time to oral steroid delivery and specific discharge criteria that reduce inter-physician practice variations are key factors.

Improving acute asthma assessment utilising an evidence based template

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Background

The recognition and treatment of acute asthma attacks is an essential requirement for paediatric senior house officers (S.H.O.)

Aim

To evaluate the impact of an evidence based structured template on the assessment of acute asthma attacks in the emergency department by paediatric SHOs.

Methods

A Medline search was used to identify published papers which assessed paediatric asthma severity. From these papers a template was constructed which consisted of asthma symptoms (5), triggers (3), medication usage, risk factors for severe attack (7), physical exam (11), ancillary assessment (2).

The quality of acute asthma assessment was evaluated initially by a retrospective analysis of all paediatric asthma admissions for 2003. A prospective analysis was then undertaken for patients admitted in 2005–2006. Children greater than 2 years were eligible if they presented to the emergency department with symptoms of acute asthma.

Results

Retrospective analysis of 153 charts (65 < 5 years, 88 > 5 years) indicated adequate history 33%, adequate medication usage 45% and adequate physical examination 37%.

In the prospective analysis, documentation was excellent (>90%) as regards symptoms, risk factors for acute attacks and physical assessment. Medication use in the 24 h prior to Emergency Department attendance was documented in 75% and rescue plans were outlined in 33%.

Conclusions

This template can be used to facilitate an improved acute asthma assessment. It can highlight parents' beliefs or behaviours where specific interventions are required. Paediatric SHOs need to have a clear understanding of asthma rescue plans to counteract the deficiencies detected.

Bed burden of bronchiolitis: regional review and rational options

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Aims

(1) To analyse the epidemiology and in-patient bed utilization by bronchiolitis over a 7 year period in a regional centre. (2) To try to define 'season of RSV bronchiolitis' with particular reference to RSV prophylaxis timing and duration.

Methods

A retrospective HIPE data analysis of confirmed inpatient cases of bronchiolitis during a 7 year period from October 1999 to January 2007. Detailed length of stay (LOS) analysis based on gestation and birth weight of the defined birth cohort of the region was compiled based on Regional Maternity labour ward records. All nasopharyngeal aspirates (NPA) tested for RSV during the study period was analysed with special emphasis on seasonal trends. LOS was calculated to minutes and separately analysed for RSV positive and negative patients. Only infants born in the regional maternity were included in the final analysis. Analysis of illness severity/intensive care requirements/therapeutic intervention were not part of the remit of our review.

Results

A total of 1,850 children were admitted in the 7 year period and the clinicians requested NPA for RSV in 940 (50.81%). Out of the 940 swabs 287 (30.53%) were reported as RSV positive 653 were negative for the RSV virus. Male to female ratio was 1.3:1 (males 1,051 and females 800). RSV +ve (287) babies stayed 1,226 days in total in hospital with the mean of 4.495 days and the duration of stay in RSV negatives show the total of 2,670 days with mean of 4.5 days. Our proportional birth weight and gestation based analysis did not reveal significantly increased bed utilization by LBW, VLBW or ELBW infants among a defined birth cohort. Interestingly all our < 25 weeks gestation bronchiolitis were RSV negative, perhaps due to the RSV prophylaxis. We specifically identified wide spectrum of case spread during winter and specific rational measures to be implemented to reduce length of stay and optimise RSV prophylaxis timing.

Discussion

Our series of bronchiolitis is one of the largest reported in Ireland within a specified birth cohort. This initial work is part of an ongoing case based economic impact analysis of bronchiolitis in collaboration with University of Limerick.

The CAT that prevents asthma? The relationship between acute asthma and air pollution

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Background

The prevalence of asthma and admission to hospital with acute asthma in Childhood increased steadily in the latter part of the twentieth century. In Europe and North America admissions peaked in 1995 and thereafter have fallen remarkably. The reason for the rise and subsequent fall are unclear. Possibly explanations include improved managements in the community, changing threshold for admission to hospital, or an environmental factor. Based on observations of patterns of air pollution at Mace Head over 20 years, we hypothesized that this reduction could be the result of improved air quality consequent on the introduction of tighter vehicle emission controls, the advent of the catalytic converter, and latterly the ban on bituminous domestic fuels.

Methods

Admissions of children aged 1–14 with acute asthma have been measured at the Paediatric Department, UCHG since 1985. This is the sole paediatric in-patient unit serving a well defined population in mixed urban/rural settings. The birth rate has been stable with little net migration until recently. There is no heavy industry in Galway, and the prevailing wind is from the Atlantic.

Black smoke levels were measured using standard methodology from the mid eighties to date. This measurement reflects small particle (< 5 μm) air pollution. Particles of this size disperse widely and remain suspended in the air for several days. These were measured at three different sampling stations all within a one mile radius in central Galway. Black smoke levels measured in μg per m^3 were measured on a daily basis. These measurements were averaged over 1 year time frames and compared with the numbers of asthma admissions.

Results

Black smoke levels rose steadily from 1984 through to 1995. Thereafter levels declined remarkably. Asthma admissions followed a near identical pattern. The number and rate of childhood asthma admissions was closely correlated with average smoke levels in the same year.

Discussion

These data document the remarkable improvement in air quality from the mid 1990s. Though the number of vehicles on the roads has increased steadily, black smoke levels have declined remarkably. Legislation required that catalytic converters were fitted to all new cars from 1993. Since that time the number of vehicles without catalytic converters has declined steadily and there are now few if any petrol engine cars on the road without these devices. Bituminous coal was banned in the Dublin area in 1990 and this ban was extended to the Galway area in the late 1990s. This may have had a further effect in reducing black smoke levels. These data suggest that the decline in acute asthma in children is related to improved air quality.

Potentially better practices in reduction of bronchopulmonary dysplasia

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Aim

To compare management practices in prevention and management of bronchopulmonary dysplasia (BPD) in the three regional maternity hospitals in Dublin. To identify the practices the hospitals are currently considering implementing from the Vermont-Oxford Network (VON) recommendations.

Methods

1. Questionnaire. The questionnaire was based on the Potentially Better Practices to reduce chronic lung disease, as developed and updated by VON in three quality improvement collaboratives.
2. Benchmarking and Site Visits. Questionnaire was administered to neonatology consultants, registrars, and senior neonatal nurses. The 2005 annual clinical report was used for statistics for each hospital.
3. Responses. Responses from each hospital were unified into a single response, which was verified by the hospital.

Results

There were 17 questionnaires distributed and all 17 were returned. Comparisons of selected variables (9 of 30) are shown in Table 1. Procedures the 3 hospitals are planning to implement are listed in Table 2.

Conclusion

This study reported many similarities and some differences in prevention and management of BPD in participating hospitals. Vermont Oxford Network incidence of BPD was 36% in this population for 2005. Practices are being implemented to try and reduce BPD by the individual hospitals. Increasing awareness of potentially better practices in reducing BPD, with cooperative reviews of each other's practices within neonatal units in Ireland, should help to reduce chronic

Table 1 Comparisons of selected variables among the three participating hospitals

Hospital	Hospital A	Hospital B	Hospital C
Incidence of BPD (same diagnostic criteria)	12%	18%	25%
Routine intubation in delivery room			
Yes/No	No	Yes	Yes
Ventilation in D.R.	Self-inflating bag	Self-inflating and NeoPuff	Anaesthetic bag and manometer, or NeoPuff
Inspiration pressure monitoring in D.R.	No	Yes	Yes
Oxygen saturation monitoring in D.R.	No	Yes	Yes
Use of oxygen blender in D.R.	No	Yes	Yes
Antenatal steroids (same indication criteria)	81%	72%	74%
Systemic postnatal steroids for BPD	5%	4%	4%
Postnatal steroids indication criteria			
Age	3–4 weeks	>6 weeks	>6 weeks
FiO2	>0.5	>0.3	0.8–1.0
PIP	23–28 cm H2O	22–25 cm H2O	22–25 cm H2O
Inhaled postnatal steroids	No	No	Yes

Table 2 Procedures hospitals are planning or considering implementing

Hospital	Hospital A	Hospital B	Hospital C
Procedure	Oxygen blender and NeoPuff at delivery	Vitamin A supplementation	Oxygen blender, weaning strategy and volume guaranteed ventilation

lung disease, as has occurred successfully with other quality improvement projects of the Vermont Oxford Neonatal Network.

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Harvesting neonatal heart valve homograft. Can we do better?

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Background

In adults, heart valves are retrieved from multi-organ system failure cadavers and explants. Cadaveric heart valves are used in the reconstruction of congenital heart diseased valves in infants and children. They need to be retrieved from neonatal/paediatric cadaveric donors. The object of this study was to compare the number of potential cadaver donors that could be retrieved from the Irish Neonatal Mortality database to the number of neonatal donors retrieved by the National Transplant Team (Mater hospital).

Method

Eligible babies for heart valve retrieval include >36 weeks, negative serology for maternal/fetal infection and retrievable harvesting within 6–8 h of death. Infants with congenital heart disease were excluded. Donors arise from infants born with anencephaly or asphyxia. Those infants were retrieved from the Irish Neonatal Mortality Database from 2000 to 2004. The National Transplant Team (Mater Hospital) provided data on the use of infant homografts from 2000 to 2004.

Results

Forty-one babies with term asphyxia and 12 babies with anencephaly were documented in the Irish Neonatal Mortality Database giving a total of 53 potential donors. During the same time period, only five heart valves were harvested by the National Transplant Team.

Conclusion

Less than 10% of potential neonatal cadaveric donors were harvested over a 5 year period. This, according to the National Tissue Bank meets the demand for homografts for the present. Nevertheless, surplus valves may be needed to match increasednational demands envisaged in the nearest future or alternatively exported to the countries where demand exceeds supply.

An audit of successful extubation of VLBW infants

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Aim

To document extubation success rates of VLBW infants in a level III NICU.

Methods

Retrospective chart review of all neonates born between Jan 2005 and Dec 2006 with a birth weight ≤1,500 g and gestational age ≤32/40 who were ventilated for greater than 12 h. Successful extubation was defined as remaining extubated for greater than 72 h. Each infant's demographics were documented including gestation and birth weight. Ventilation settings, patients' vitals and last blood gas prior to extubation and ventilatory support given post extubation were recorded. Use of caffeine pre extubation was also recorded.

Results

A total of 39 infants were studied of which 31 were successfully extubated giving a successful extubation rate of 80%. Reasons for reintubation were bradycardia and desaturations in two infants, respiratory distress in four infants including one infant with stridor and suspected sepsis in two infants. Mean gestational age of babies studied was 27.7 weeks with a mean birth weight of 1,150 g. Babies who failed extubation were of similar mean gestation 28.8 and birth weight at 1,130 g. The average length of intubation was 1150 h with standard deviation of 39.6. The ranges of peak inspiratory pressures at extubation were 12 to 20 with an average of 14. Ventilator rate being provided just prior to extubation ranged from 10 to 30 with a mean of 15. With regard to vital statistics all infants had a respiratory rate above that provided by the ventilator. All infants were commenced on NCPAP at extubation. In total 2 of the 45 infants did not receive caffeine prior to extubation neither of whom failed extubation.

Conclusion

Our extubation success rate is satisfactory. However there was no apparent difference in ventilator settings, vital signs and blood gases just prior to extubation between the successful and unsuccessful groups making it difficult to predict accurately which babies will extubate successfully. At present there is significant amount of research directed at developing tests for predicting successful extubation. One example is the spontaneous breathing test. Infants once deemed to be ready for extubation are placed on ET CPAP at the same PEEP setting for 3 min. A failed SBT is recorded if the infant either had a bradycardia for more than 15 s and/or a fall in SpO2 below 85% despite a 15% increase in FiO2. In order to close the loop we hope to reaudit our rates of successful extubation with this additional test.

Audit on multicultural diversity in paediatric attendances in the emergency department, children's university hospital, temple street

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Aim

To audit the population of paediatric attendances to the Emergency Department to ensure appropriate planning of services and provision of care.

To analyse the pattern of attendances of different age groups and type of presentation in the non-native and Irish paediatric population.

Background

The changing population in Ireland and the influx of migrants accounts for around 10% of the Irish population according to the latest National Census.

Conditions like sickle cell, malaria, thalasaemia and other diseases have become more prevalent and paediatricians need to be aware of it when treating patients.

Different cultural backgrounds also influence parents' view of certain illnesses or injuries that may require immediate medical attention.

Methods

By using the Emergency Department computerised database, SYMPHONY all the attendances to the Emergency Department (ED) over a 3 month period from 1st October– 31st December 06 were analysed.

Hospital policy does not allow patients to be identified according to their ethnicity, therefore documentation of non-native patients were done by looking at their names and surnames and cross-checking with ED records.

Patients were also divided according to their age group (< 1, 1–5, 5–10, > 10 years) and presenting complaint (illness/injury).

Data was collected using excel spreadsheet and entered into a pivot table.

Results

There were a total of 12,220 attendances in the 3 month period. Almost a quarter, 2,777/12,220 (23%) were of non-native names. 2,019/2,777 (73%) of non-native attendances were under 5 years of age, compared to 5,539/9,443 (59%) of the Irish attendances. Children

under 5 years of age comprise 6,212/8,790 (71%) of all the attendances in the 'illness' category, which is 6,212/12,220 (51%) of the overall workload. The overall total attendances under 5 years of age are 7,558/12,220 (62%). 2,202/2,777 (80%) of the non-native population attended with an 'illness' while 575/2,777 (20%) attended with an injury, compared to 6,588/9,443 (70%) of Irish attendances were in the 'illness' category and 2,855/9,443 (30%) attended with an injury.

Conclusion

There is a high usage of the Emergency Department among the non-native paediatric population. It is highest in the under 5 years of age category, in keeping with the recent influx of immigrant population over the last few years. We have to ensure ready access to interpreter services and that NCHD staff are trained to deal with families and diseases from different ethnic and cultural backgrounds.

Most non-native patients present with an 'illness' rather than an 'injury'. The patterns of illnesses are similar to the general paediatric population and only a small number present with ethnic-specific diseases.

The majority of our workload in the ED is with children under 5 years of age. Close liason with Public Health Nurses are crucial as they are our main community link with children of this age group.

Ongoing audit needs to be done to ascertain the workload and resources required to facilitate provision of healthcare and support to non-native children in our catchment area.

A proposal is being made to collect data on ethnicity, using the same categories as the National Census at the ED reception to improve the accuracy of the data collected and to make provision for better services in the future.

The pied piper of hamelin: an unusual case of leptospirosis

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Introduction

Haemorrhage in Leptospirosis is well described in the literature, however disseminated intravascular coagulopathy (DIC) is not. We present a case where Leptospirosis was associated with DIC and massive pulmonary haemorrhage.

Case details

A 14.5 year old boy was GP referred for a 1 day history of crippling bilateral calf pain. This symptom was preceded by a prodrome of mild gastroenteritis and pyrexia. Upper limbs ecchymoses had been noted, but attributed to sport injury. He had had no recent foreign travel and no ill contact. On initial examination, he was afebrile, mildly tachypneic, tachycardiac with a blood pressure of 100/64. His calves were tender and warm. Residual systemic examination was normal. The initial impression was viral myalgia/ rhabdomyolysis and to outrule out deep vein thrombosis. Initial platelet count was 37, Prothrombin time = 16.7 and INR = 1.6.

Within a period of a few hours, he became hypotensive, mottled and icteric. Further history illicited that he had gone swimming 2 weeks previously in a local river. The impression was of an infectious hemolytic process. Titres were sent for hepatitis, EBV, Leptospirosis and Lyme disease. He was then commenced on IV Penicillin and Ceftriaxone.

When 100 ml of blood exuded from the oral cavity, this was initially thought to be hemetemesis. He developed decompensated hypotension and acute respiratory distress. On intubation, approximately 1 litre of bright fresh blood was suctioned through the endotracheal tube. His chest radiograph confirmed the presence of massive bilateral pulmonary haemorrhage. Ventilation was problematic and his oxygen index was high, extracorporeal membrane oxygenation (ECMO) criteria were fulfilled. With his coagulopathy corrected he was transferred to Great Ormond Street Hospital.

His respiratory distress responded to high frequency ventilation. Multi-organ involvement included myocarditis, pancreatitis, uveitis and hepatitis were treated symptomatically. He made a full recovery. Leptospirosis was subsequently confirmed.

Discussion

Adolf Weil was a German Professor of Medicine (1886) who described this disease 30 years before Inada & colleagues identified the causal organism ie a Gram negative spirochete [1]. (Greek: *Lepto*: slender. *Spira*: spiral [2]). Rats are notoriously known to be the vector to animals and humans, who may be directly or indirectly in contact with the urine of an infected animal.

Leptospirosis presents in 90% cases as an acute febrile illness with an excellent prognosis. However in 10% of cases, the more severe form is known as Weil disease that classically includes fever, jaundice, renal failure and hemorrhage. The pulmonary, cardiac and the central nervous systems are also frequently involved.

Bilateral exquisite calf pain is one of the pathognomonic features for leptospirosis [3]. It is due to haemorrhage into the muscle bed secondary to myositis that often begins during the first week of symptoms and may last into the third and fourth week.

This case highlights the dangers of recreational swimming in freshwater. Leptospirosis is not a disease confined to sewage workers and veterinarians. It is a statutory notifiable disease in Ireland. There were 14 confirmed cases in 2005, 10 cases in 2004, 9 cases in 2003, 8 cases in 2002, 9 cases in 2001 and 7 cases in 2000 [4].

Conclusion

Although Leptospirosis may often mimic viral symptoms, the more severe form proves difficult to manage and can be deleterious even when appropriate actions are taken. A possible key to correct diagnosis is a thorough history, focusing on the patient's recreational activities and animal exposure.

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A case of spontaneous linear fracture of the skull in a neonate

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Background

Linear skull fractures, in the absence of instrumental deliveries are extremely rare. We present such a case, which was complicated by idiopathic hypercalcaemia and congenital heart disease.

Case history

The female infant was born at 40 weeks and 3 days gestation by emergency lower-segment caesarean section for failure to progress in labour. It was a non-instrumental delivery and no trauma was documented at the time. She did not require any resuscitation and apgar scores were 9 at 1 min and 9 at 5 min. The pregnancy had been uneventful and there was no significant maternal history.

On day 2 of life she was admitted to NICU due to tachypnoea and difficulty with feeding. A murmur was detected on examination and an echocardiogram demonstrated multiple small ventricular septal defects. She was commenced on diuretics. A boggy swelling over her left parietal bone, which was thought to be a cephalhaematoma, was also noted. On day 9 her feeding deteriorated and a cranial ultrasound scan was performed, which elicited a fracture of the parietal bone. An MRI scan confirmed "a *non- depressed, linear fracture*" with a 6 mm overlying, cephalhaematoma. Of note, there was no evidence of underlying brain parenchymal injury, therefore, conservative management was instituted. Other investigations revealed the presence of hypercalcaemia with normal parathyroid and vitamin D levels. At 3 months the infant is neurologically normal.

Discussion

A large, multi-centre study in the New England Journal of Medicine in 1999, estimated the incidence of intracranial damage to be 1 in 664 and 1 in 860 for forceps-assisted and vacuum-extraction deliveries; respectively. However, the incidence was as low as 1 in 1900 for non-instrumental, vaginal births. An extensive search of all current medical databases revealed only 2 previous reports of spontaneous *linear* fractures similar to that which we have described here.

Conclusion

Intrapartum, spontaneous, linear skull fractures are extremely rare. In our case there does not appear to be any neurological deficits to date. Although rare it is important to remember that these injuries can occur spontaneously and this should be taken into account when considering forensic investigations in neonates with skull fractures.

Significant RSV infection in neonatal ICU graduates 30–34 weeks gestation

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Background

Studies have suggested that preterm infants born at up to 35 weeks gestation who require hospitalisation for RSV lower respiratory tract infection have morbidity comparable with that of less mature preterm infants.

Palivizumab is a costly intervention, which has not been shown to reduce mortality in these infants. Current Rotunda palivizumab guidelines limit prophylaxis to infants less than 1 kg birth weight, or < 30 weeks gestation and < 6 month old at the start of the RSV season. Extension of the programme to infants up to 34 completed weeks would mean immunizing approximately 70 additional infants per year at a cost of up to 6,700 Euro per infant.

We wished to examine the rate of significant RSV infection in our infants 30–34 weeks' gestation and 1–2 kg weight at birth to determine whether palivizumab prophylaxis would be justified in this population.

Study design

All living infants 30–34 weeks gestation, born in 2004–2005 was identified. A letter was sent to parents of these infants notifying them of the study and alerting them to a phone call interview. Those not wishing to participate could opt-out via phone/letter at this point. Phone interview then occurred to determine whether the infant had a likely episode of RSV bronchiolitis requiring admission to hospital. If so, a detailed information sheet was sent along with a consent form for access to the infant's chart in the relevant hospital. This procedure was approved by the ethics committees of the Rotunda Hospital, the Children's University Hospital Temple St. and Our Lady's Hospital, Crumlin. Data collection and storage methods were approved by the ethics committees.

Methods

153 infants were born in the Rotunda in 2004–2005 between 30 and 34 weeks gestation and 1–2 kg birth weight. Five deceased infants were excluded as were seven others for various reasons. A further 25 had to be excluded as no phone number was available. This left 116 infants eligible in total after exclusions. Parents were then contacted as per the study protocol.

Results

Of 116 infants, 47 could not be contacted (due to incorrect/disconnected number, or no reply after at least 5 attempts) leaving 69 contacted (59% of eligible infants). 6/69 had admission to hospital with suspected bronchiolitis (by parental report). On chart review four had nasopharyngeal aspirate (NPA) for RSV which was negative, the remaining two did not have clinically suspected RSV infection. None had proven RSV bronchiolitis. Seven infants were given synagis in other units post discharge, although they did not meet internationally accepted guidelines for administration of palivizumab.

Discussion

This small study demonstrates that RSV infection was not a significant cause of hospitalisation in our population studied. This contrasts with published research which has shown hospitalisation rates of 1.8–9.8% for RSV bronchiolitis in infants born less than 35 weeks gestation.

We acknowledge bias may have occurred as a number of parents could not be contacted. It is possible they may represent a different population subgroup at higher risk of RSV infection.

We feel further larger studies are needed to determine the incidence of significant RSV infection in infants 30–34 weeks gestation before routine palivizumab prophylaxis can be justified.

Emergency department use by paediatric patients in a district general hospital

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Background

Cavan General Hospital serves a population of approximately 110,000 residents of counties Cavan and Monaghan and parts of counties Meath, Leitrim and Longford.

The paediatric service in Cavan general hospital cares for children from birth to their 15th birthday. The hospital recently appointed the first paediatric accident and emergency trained clinical nurse specialist to the Emergency Department (ED) in order to improve clinical expertise and facilitate training of ED staff. The ED is the single route of entry for all paediatric admissions. We sought to prospectively evaluate all children attending the ED service from 0 to 14 years over a 5-month period. This was the first attempt to evaluate patients with both medical and non-medical problems attending the service. We sought to compare our results with that of a previous audit of medical patients, in light of an increasing local population and plans to amalgamate the two current acute paediatric units into a new North East regional hospital site. We also identified a number of other areas of interest to be examined.

Aims

To examine demographics of patients presenting to the ED. To examine waiting times in comparison to accepted best practice. To look at pre-ED advice seeking behaviour and referral sources. To compare the above with results of previous ED audit in 2002. We also wished to look at any language difficulties encountered in light of an expanding immigrant population. We wished to evaluate current analgesia/antipyretic administration by parents, GPs and staff with a view to a new ED analgesia protocol.

Methods

Sequential capture of all children, 0–14 years attending the ED of Cavan General Hospital from August 1 to December 31, 2006. A one page questionnaire was completed by the doctor or nurse attending to patient. Completed questionnaires were collected and analysed.

Results

Eight hundred and twenty-six completed questionnaires were received. This represented 40% of the total paediatric attendances for this period. The information

received is a valid indicator of aspects of ED activity. The busiest weekday was Monday. The busiest time of day 0800–1700; 60% of attendances. Seventy percent of attendees resided in Co. Cavan but 39% travelled 11–20 miles to ED. Forty-six percent were GP referrals, only 3% of others sought advice before presenting to ED. There was poor awareness of need for antipyretics and analgesia amongst parents. There was a low rate of analgesia/antipyretic administration by GPs. The average waiting time for children to be seen in the ED was 33 min while 6.6% of patients waited longer than 1 h and these were non-medical patients in the majority. This was supported by the previous audit which found longer waiting times associated with patients referred for surgical review.

Conclusions

This study although limited by difficulties with staff compliance in completing questionnaires reflects paediatric activity levels in the ED. It demonstrates the need for a paediatric trained nurse staffing of the ED at all times. We plan to introduce a protocol for initiation of analgesia/antipyretics at triage. There is a clear need to consider initiation of ''fast track'' system for minor injuries to reduce waiting times in this group.

The genetic basis of autosomal recessive osteogenesis imperfecta in the irish traveller population

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Osteogenesis Imperfecta (OI) is usually an autosomal dominant disorder, and is clinically classified according to the Sillence classification of 1-IV. However, the Irish Traveller population has an autosomal recessive form of severe OI, which fits with type II/III in the Sillence classification. We have identified 16 patients in 5 extended Traveller families, where almost all the affected children are born with severe limb and thoracic deformities due to multiple fractures, including in utero fractures. Most have died within 6 months, of respiratory compromise. However, there are two surviving affected children at ages 5 years.

No type I collagen abnormality has been described in the Irish Traveller OI (Pope et al. 1989). Recently, the genetic basis of one form of autosomal recessive OI has been found, due to homozygous mutations in a gene CRTAP or cartilage associated protein (Morello et al. 2006). CRTAP is homologous to a family of prolyl 3-hydroxylases which modify collagen, and mutations affected the modification of collagen fibrils. A partner protein for CTRAP, LEPRE1 or prolyl 3-hydroxylase 1 (P3H1) has also found to be the basis of another autosomal recessive form of OI in people of African origin (Cabral et. al. 2007).

Samples from three affected Irish Traveller children were analysed for mutations in CRTAP and P3H1. No mutations were found in CTRAP, but all three were homozygous for a frameshift mutation c.232delC in exon 1 of the P3H1 gene. Cultured fibroblasts from one affected case were analysed by mass spectroscopy for prolyl 3-hydroxylation of type I collagen. The level of hydroxylation was markedly reduced, at a level of 15%, compared to 95–98% seen in normal controls.

These findings have now identified the genetic basis for autosomal recessive OI in the Irish travellers. This will now lead to improved OI diagnosis and genetic counselling for Travellers who have a family history of severe or lethal OI. In addition, these findings give further insights into the biology of bone collagen.

Trampolining as an adjunct to regular physiotherapy in children with cystic fibrosis

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The benefit of exercise in patients with cystic fibrosis (CF) has been recognized for as long as CF has been identified as a clinical syndrome [1]. Physical activity can have important, and even life-saving effects as secondary prevention of disease [2].

Trampolining has become increasingly popular both in the CF and general population. In light of recent publications in both medical journals [3] and national press [4] highlighting dangers associated with trampolining we carried out a questionnaire on a sample of our outpatient CF population.

We surveyed 34 patients seventeen male and seventeen female between the ages of 4 and 16 years (average 8.7 years). Twenty-nine patients (85%) have a trampoline, nineteen of which (56%) use the trampoline every day. Seventeen of the twenty-nine children were supervised by an adult when trampolining. The number of

children on the trampoline at one time ranged between 1 and 4 (average 2.6). There were no reports of any injuries that required medical attention.

Thirty children in the group perform regular formal physiotherapy averaging 21 min/day. Two children used trampolining as a sole alternative to physiotherapy. Over half of our group (55%) thought that exercise is a good replacement for physiotherapy, with 38% sometimes using trampolining instead of physiotherapy. When asked which they preferred, 55% said physiotherapy, 35% preferred trampolining, and 10% had no preference.

Trampoline use has not been proven to benefit patients with CF and considering the risk of injury, it has been suggested that trampolining should not be recommended [5]. However, trampolining is very popular among children with CF. Some parents report that trampoline is the only alternative when compliance with physiotherapy is a problem. The results show a tendency among both children and parents to use exercise or trampoline as an alternative to physiotherapy. Fortunately no children surveyed have been injured during trampoline use, which may be due to high levels of supervision and smaller numbers using the trampolines at one time.

Trampolining is very common in our CF population. We plan to encourage continued use of trampolines with the introduction of clear safety guidelines, reinforcing that trampolining is an adjunct to and not a replacement for regular formal physiotherapy.

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Effectiveness of palivizumab prophylaxis less than 32 weeks gestation

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Background

Letterkenny General Hospital has a birth rate of 1,700–1,800 per year with an average of 15–18 pre term babies (< 36/40).

Pre term babies less than 32 weeks gestation and less than 6 months of age at the onset of RSV season and up to 2 years of age with BPD receive Palivizumab. Also babies with gestation between 32 and 35 weeks with immunodeficiency or cyanotic congenital heart disease receive Palivizumab during the RSV season.

Aim

To determine the effectiveness of Palivizumab (synagis) prophylaxis in preventing admissions with RSV positive bronchiolitis.

Methods

Retrospective cohort study from 2002 to 2004. Data was gathered on all children who received Palivizumab prophylaxis during this period.

Result

19 babies less than 32 weeks gestation (range 28–32 weeks) received Palivizumab and none of these babies were admitted with RSV positive bronchiolitis.

Conclusion

International studies would suggest that expected admission rates for RSV positive bronchiolitis in infants delivered between 28 and 32 weeks would be up to 20%.

Our study has found an admission rate of 0% for babies born between 28 and 32 weeks gestation who were given RSV prophylaxis.

This study was limited due to small population size. Debate continues regarding most appropriate gestational age at which to give Palivizumab prophylaxis.

Our results would suggest significant benefit in giving RSV prophylaxis less than 32 weeks.

Congenital cutaneous mastocytosis presenting as neonatal rash

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Aims

1. To raise awareness of congenital cutaneous mastocytosis (CCM) as a differential diagnosis of neonatal rash.
2. To highlight the risk of anaphylaxis in CCM and the importance of allergen/medication avoidance which could trigger histamine release.
3. To suggest the need for long term follow up.

Background

Mastocytosis is a condition characterised by the accumulation and degranulation of mast cells within the skin. It is a spectrum ranging from a solitary mass (mastocytoma) to diffuse rash with systemic involvement and manifestations of histamine release. Congen-

ital cutaneous mastocytosis occurs in neonates and infants and may be mistaken for other neonatal conditions such as congenital leukaemia or congenital CMV infection (Table 1). It is important to avoid certain medications in these patients as there is a risk of overwhelming histamine release and anaphylaxis. Patient education about trigger avoidance (Table 2), histamine blockers, availability of pre-filled adrenaline autoinjector and long term follow up is the mainstay of management. The prognosis is good with the lesions resolving in 50% of patients by adolescence.

Methods

Prospective patient selection and analysis in a defined birth cohort over 5 years.

Results

Six patients were identified during the neonatal period with a hyper-pigmented rash characteristic of CCM (Figs. 1–3). The diagnosis was confirmed by skin biopsy (Figs. 4–5). This incidence of 1 per 3,530 live births is higher than previously reported. In all patients the rash has persisted (Fig. 6) and none developed evidence of systemic mastocytosis during follow up.

Conclusion

CCM is a rare paediatric dermatological condition which can present in the neonatal period. It is important to suspect and confirm the diagnosis early and organise appropriate follow up.

Gender differences in severe intraventricular haemorrhage in preterm infants

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Background

Male infants have a survival disadvantage and increased risk of adverse neurodevelopmental outcome compared with females [1]. High-grade intraventricular haemorrhage (IVH) is an important cause of severe cognitive and motor neurologic impairment in very low birth weight infants and is associated with a high mortality rate. Recently homozygosity for an estrogen receptor-alpha (ER-alpha) gene Pvull pP polymorphism has been associated with increased IVH in premature male infants [2]. In addition, prenatal steroids have been shown to be less effective in male compared with female infants in decreasing RDS [3].

Aim

We aimed to identify gender differences in risk factors and outcomes of preterm infants with Grade3–4 IVH.

Methods

We performed a retrospective study from January 2005 to December 2005 to identify all neonates < 1,500 g with Grade 3–4 IVH. We evaluated the antenatal management, specifically the use of antenatal steroids and the outcome of these patients.

Results

One hundred and thirty-five infants < 1,500 g were admitted to the NICU during the study period. Twenty-one infants had Grade 3-4 IVH of which 11 were female and 10 were male. All the male infants had bilateral IVH despite antenatal steroids. In contrast only one girl out of five who received antenatal steroids developed a Grade 4 haemorrhage and the rest had a lesser grade. There were no significant differences between gestation, birthweight and Apgars between male and female infants. The mean gestation was 27 + 5 weeks with a mean birthweight of 1,324 g.

Conclusion

Prenatal corticosteroid therapy may be less effective in male infants than in female infants in reducing grade 3–4 IVH. Ensuring adequate antenatal steroid treatment is vital to ameliorate severe neonatal morbidity in preterm infants. Further study of the mechanisms mediating gender differences in neonatal outcome is warranted.

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An initial report of a novel widespread cutaneous adverse drug reaction to cefaclor occurring in children

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Aims

(1) We identified and analysed the 'haemorrhagic' or 'bruise-like' skin lesions secondary to cefaclor, a cephalosporin antibiotic, for a 5 year period after observing the index case in 2001. (2) To highlight the importance

of drug reaction as a differential diagnosis while evaluating more important conditions such as non-accidental injury or meningococcal disease.

Methods

Four children (mean age of 5 years, range 2–7) who received cefaclor from general practitioner for upper respiratory infection were admitted with a bruise like rash on the arms, axillae, thighs, groin and buttocks during the study period. Early lesions were deeply haemorrhagic looking and the rash was mildly itchy. Neither palpable purpura nor associated arthritis/arthralgia or tonsillitis was noted. None of the patients were thought to be at risk of non-accidental injury. One child was febrile at admission and two of them received intravenous antibiotics after septic work-up. Three children had negative viral and mycoplasma titres. One of them received prednisolone and chlorpheniramine maleate.

Discussion

Antibiotics are the most frequently prescribed paediatric drugs and also the cause of most drug-induced adverse reactions in children [1]. Cefaclor is an orally absorbed broad spectrum cephalosporin and is effective in the treatment of upper, lower respiratory and urinary tract infections. Commonly described rashes are urticaria, maculopapular eruption, serum sickness and erythema multiforme, while serum sickness, Stevens Johnson syndrome and toxic epidermal necrolysis are rare. The frequency of rashes documented for cefaclor had been shown to be higher than penicillin, sulfonamides, and other cephalosporins [2].

We describe a distinctive, unusual ecchymotic like rash which could be easily mistaken for bruising induced by non-accidental injury or meningococcal septicaemia. The features of the rash were large bruise like patches of non-blanching erythema. The sites affected included thighs, axillae and abdomen. Unusual features in one patient were lesions on the back of ears and periumbilical region. These lesions were fixed unlike urticaria. Two patients had urticaria adjacent to the bruise like rash. Children were not in pain and if itch occurred it was minimal, again in contrast to urticaria. The rash occurred after few days of initiation of cefaclor. Once involution of the rash started to occur, the lesions subsided within 2–3 days.

Histology was not performed on these patients, chiefly because they were not toxic. We are considering skin biopsy in future patients to try to characterise the pathology of these lesions. Intradermal and oral rechallenge has not been performed. This adverse reaction has not been previously reported by the manufacturing company and our observation is reported to Irish Medicines Board.

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Paediatric scalp skin conditions: some diagnostic and therapeutic tips

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Abstract

Scalp conditions in children are sometimes difficult to diagnose and treat. Partly this may be because they are not always what they seem on first assessment. Often the initial appearance suggests a form of infection—however the true or actual underlying cause of infection may not be apparent. We present three patients who illustrate these diagnostic and treatment dilemmas.

The first patient was a 10 year old boy from County Limerick who was admitted with a 1 week history of a tender mass in the parietal scalp. This prurulent, boggy mass with associated hair loss did not respond to broad spectrum intravenous antibiotics. Surgical colleagues were contemplating drainage procedure under general anaesthetic and possible grafting. Dermatologic review elicited that the boy lived adjacent to a cattle farm and the diagnosis of kerion due to cattle ringworm was confirmed by mycology testing using novel retrieval of affected skin and hair utilising a sterile domestic toothbrush. He responded well to systemic oral griseofulvin and steroids (used to decrease scarring alopecia caused by the scalp abcesses).

Accurate diagnosis of fungal disease is important for two reasons. Firstly, we are aware of children in other health units who have had inappropriate and unnecessary extensive surgery, including plastic surgery and grafting, for scalp ringworm treatable by appropriate oral antifungal medicine. Secondly, with increased immigration and racial diversity the spectrum of cause of scalp fungal disease is changing in Ireland. This has therapeutic implications as some fungi spread more rapidly from

person to person and with less effective treatment response so knowing the cause is important particularly in the community setting.

The second patient was an 5 month old infant recently adopted from Vietnam and admitted with an acute scalp cellulitis. Her occipital scalp cellulitis was responding poorly to intravenous antibiotics. On further skin examinatiion, it emerged that she had scabies. Although scabies does not normally occur on the scalp it can do so in infants under 12 months and in crusted scabies. The scabies infection had both caused celulitis from scratching and had maintained the cellulitis. This infant had the classic sign of burrows manifesting as pustules on the soles of the feet. She had previously been ''treated'' with Derbac M® (a malathion preparation). Malathion is a scabistatic rather than scabicidal preparation which led to therapeutic failure and admission with cellulitis.

The third patient is a young 7 year old girl presenting with a therapeutic scalp problem. She had severe scalp lice infection with secondary pyogenic infection and poor response to topical anti lice measures. No viable lice were found after 3 days treatment using mayonnaise held in place on the scalp by cling film sheet and tubifast. This proved curative as the mayonnaise occlusion of the scalp asphyxiated the lice. Vaseline can be used in a similar manner.

In summary:

- In children presenting with signs of scalp ''infection''—pustular discharge, alopecia, cellulitis—we advocate looking for an underlying fungal, mite or louse infection.
- Toothbrush sampling is proving to be a good method for detecting the causes of fungal infection and is viewed as a non-threatening sampling tool by the children.
- Scabies and lice can present with pyogenic infections of the scalp.
- Severe head lice can be treated with non-toxic occlusive therapy.

Novel gene mutation in neonatal hyperekplexia with mild clinical phenotype

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Hypertonia in a neonate may be a sign of neurological dysfunction, for example secondary to hypoxic-ischaemic insult. Rarely it can be due to hyperekplexia (startle disease), a familial neurological disorder characterized by abnormally enhanced startle response, followed in most cases by momentary generalized muscular stiffness. The gene implicated in this condition is GLRA1 (human glycine receptor 1 subunit gene). This codes for an inhibitory glycine receptor clustered on the postsynaptic membrane of inhibitory glycinergic neurons. Mutation leads to neuronal hyperexcitability by impairing glycinergic inhibition. A number of mutations in the alpha-1 subunit have been described in familial and sporadic cases of hyperekplexia.

We report a neonate who presented on day 1 of life with hypertonic flexed posture and exaggerated startle reflex. Head lag was reduced. Neurological examination was otherwise normal. There were no antenatal risk factors and delivery had been uncomplicated. Family history was significant; the baby's father had presented with similar signs in the newborn period and had subsequent normal developmental milestones.

Brain imaging was normal. EEG was also normal. CSF showed raised amino acids and normal free gamma-aminobutyric acid (GABA), with a decrease in GABA in a second sample 1 month later. Genetic analysis revealed heterozygosity for the novel allele GLRA1(G160R) present in the affected infant and her father.

While the long-term prognosis of hyperekplexia is relatively benign, sudden death due to severe spasms have been reported. This new mutation, with associated mild clinical phenotype, will be studied further in the context of effect on receptor function and may provide additional prognostic information.

A rare cause of bronchiectasis in children

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Introduction

Primary Ciliary Dyskinesia (PCD) is a disorder characterised by impaired ciliary function, leading to an array of clinical manifestations such as chronic bronchiectasis, chronic sinusitis and infertility. A 13 year old female presented to her GP with recurrent ''soft and clicky cough''. She was born at full term, but subsequently developed respiratory distress on her second day of life, which warranted admission to the NICU. There, she required IV antibiotics for 3 weeks. Further investigations, including a sweat test were negative. There had been no major respiratory problems until 13 years later when she grew *Pseudomonas* sp. in her sputum. A CT Thorax was done, which showed bronchiectatic changes. A full investigation work-up for ''non-Cystic Fibrosis (CF) bronchiectasis'' was carried out, including Nasal

Brushings and Exhaled Nitric Oxide (ENO). Electron microscopy of nasal brushings revealed marked cilial aplasia, abnormal/ misalignment of neighbouring cilia and absence of cilial dynein arms which is consistent with Primary (PCD). The fractionated ENO was decreased; two parts per billion (ppb); normal range (5–25) ppb.

Summary

PCD is a rare autosomal disorder with an incidence of 1/15,000 – 1/ 30,000 across all ethnic borders. This case is to highlight PCD as a differential diagnosis in non-CF bronchiectasis and its respiratory manifestation in the early hours of life. In addition, the poster would also emphasise on the use of ENO as an important diagnostic screening tool.

Group b streptococcus prevention: an audit

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Introduction

Group B Streptococcus (GBS) is recognised as the most frequent cause of severe early onset sepsis in newborn infants. The US Centres for Disease Control and Prevention (CDC) have published extensive guidelines on the prevention of GBS disease via bacteriological screening of pregnant women between 35 and 37 weeks gestation. In Ireland there is little collected data on which to base a consensus guideline. In our neonatal unit we use the risk factor approach provided in previous CDC guidelines to decide on intrapartum prophylactic antibiotics and subsequent management of the new born. In our hospital, women who have one or more clinical risk factors for GBS disease or have GBS bacteriuria in pregnancy are given antibiotics in labour. If they do not receive adequate treatment (i.e., antibiotics >4 h during labour) their babies have blood cultures taken and are treated with IV antibiotics for 48 h pending the results of the cultures.

Aims

(1) To quantify the work load generated for the neonatal unit by treating babies under this protocol. (2) To identify possible areas of improvement in our practice to help minimize this work load.

Methods

We undertook a prospective audit of all term babies treated with antibiotics for the prevention of GBS sepsis. This included all babies born to mothers with one or more risk factors for GBS disease who were treated inadequately in labour. It also included cases where the mother was adequately treated but the baby was symptomatic. The data was collected every day over 2 consecutive calendar months. The baby's details, maternal risk factors, length of stay in the neonatal unit, clinical course and test results were recorded on a proforma by the admitting doctor.

Results

During the period of the audit, 45 babies were admitted to the neonatal unit for treatment under the GBS protocol. This represented 27.1% of all babies admitted to the neonatal unit over the same period. 24 babies, 53% of the cohort, were admitted overnight. The length of admission ranged from 1 to 72 h with an average of 14.9 h. 39 babies (86%) had antibiotics for just 48 h with the maximum length of treatment 5 days. In 27 (60%) of cases there was maternal GBS bacteriuria. A further 10 had prolonged rupture of membranes >18 h and 11 women had pyrexia >38 degrees in labour. 12 babies developed symptoms. There were no positive blood cultures and none of the babies were significantly unwell. In some women there was an unexplained delay in the administration of intrapartum prophylactic antibiotics.

Conclusions

Babies admitted to our neonatal unit under the GBS protocol represent a significant burden. In addition, it causes separation anxiety as babies are removed from their mothers for significant amounts of time. Strategies to reduce the number of infants requiring treatment are desirable. Identification of women with GBS bacteriuria through a chart labelling system may enable earlier administration of prophylactic antibiotics in labour. A rapid antigen screening test performed at the onset of labour might decrease further the number of babies requiring treatment.

Botulinum toxin injections under local anaesthesia and sedation in children with cerebral palsy

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Introduction

Botulinum toxin is a potent neurotoxin that causes a temporary chemical denervation of spastic muscles in children with CP. It is predominantly used to improve gait, prevent contractures, ease nursing care and delay orthopaedic surgery. In children with severe spasticity it can also be used for painful disabling spasms and cosmesis. Treatment with botulinum toxin must be combined with a planned targeted programme of

physiotherapy. It has been shown that injections can be safely given without general anaesthesia (GA), which is the practice in most European centres. Topical anaesthesia, midazolam sedation and analgesia can be safely used in combination. Accuracy of injections is enhanced with use of EMG or ultrasound guidance. In Ireland injections to date have mostly been given by orthopaedic surgeons or paediatric neurologists.

Aim

To review safety and practice of a newly established, hospital based, Community Paediatric Service.

Method

An audit of the first year of service from November 2005 to October 2006, Community Paediatrics, National Children's Hospital, Tallaght. Medical records of children who had received botulinum toxin injections as a day case were reviewed.

Results

Sixty children had botulinum toxin injections during the first year. Five children had two sets of injections within the year. The total number of children injected was 55. Dysport® was used in all children and all received doses within the recommended range of < 30 U/kg. All children had been jointly assessed by the paediatrician and physiotherapist prior to injections. Most children were ambulant and injections were given to improve gait. The age range of children injected was 2.2–15.7 years. Of the 12 (21%) children receiving their first injection, 5 were under 3 years. Eleven children had multi-level injections; the remainder single-level. EMG was used for injection of the tibialis posterior muscle. Ultrasound was not available. Fifty two children (98%) had local anaesthesia and sedation. Parents stayed with the child during the procedure and hospital stay did not exceed 4 h. There were no serious adverse events. The physiotherapists in the referring centres received a copy of the treatment sheet and commenced therapy within 2 weeks of injections.

Conclusions

Single- and multi-level botulinum toxin can be safely and efficiently administered by experienced community paediatricians using local anaesthesia and sedation. Joint assessments with physiotherapists and the treating physician enhance the delivery of a coordinated treatment programme.

Reduced bone mineral density: a follow up study

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Introduction

Reduced bone mineral density (BMD) is a known complication of Cystic Fibrosis (CF). Its pathogenesis is poorly understood and the most effective treatment for prevention and treatment of reduced bone mineral density are unclear.

Aim

The aim of this study was to compare BMD in patients with CF after therapeutic intervention and to review variables that could be associated with a change in BMD.

Methods

Follow up data for Dual–Energy X-ray Absorptiometry (DEXA) scans on Lunar DPX IQ, Lunar Prodigy and Lunar Prodigy Advance bone densitometers was available on 13 patients over a 7 year period from 1/1/00 to 31/3/07. Change in BMD was evaluated by comparing z scores for L2-L4. Clinical data for each patient was reviewed for variables including change in Body Mass Index (BMI), FEV1 (% predicted), serum vitamin D3 and treatment over the time period between the 1st and 2nd DEXA scan (average 2 years). Of the 13 patients reviewed all were pancreatic insufficient and had been treated with oral vitamin D3.

Results

Five patients showed no improvement in their Z score. One of the five patients showed a deterioration over 2 years from initial $z = -1.7$ at 13.8 years to $z = -4.1$ with a decrease in BMI from 18.2 to 15.6 with unchanged predicted FEV1. Three patients with a slightly increased or unchanged BMI showed mean decrease 5.6% (range 4–7%) predicted FEV1. The fifth patient showed z score deterioration from 0.9 to 1.9 with increase in FEV1 and static BMI. All 5 patients showed an average decrease in serum vitamin D3 of 22.5 (range 4–32) nmol/l despite being on high vitamin D3 supplementation (mean 0.64; range 0.31–1.11 μg/kg). Four of the five patients all with z scores between –3 and –4.1 have subsequently been treated with oral alendronate.

Of the seven patients four showed an average increase of 10% predicted FEV1, two remained unchanged and the seventh showed deterioration in predicted FEV1 of 11% and subsequently died. Five of the seven patients showed mean increase in serum vitamin D3 levels of 12 nmol/l (range 5–30 nmol/l) on a mean vitamin D3 supplementation of 0.32 (range 0.17–0.625) μg/kg. Two patients were treated with oral bisphosphonates. (One required intravenous palmidronate because of poor response to oral alendronate.)

Conclusion

Patients with CF and pancreatic insufficiency are at risk of malabsorbtion of fat soluble vitamins, and those with low vitamin D3 levels have a higher risk of low bone

mineral density and long term skeletal complications. The optimum dose of vitamin D has not yet been established but in our cohort those with no improvement in BMD were treated with twice the amount of oral vitamin D3 but showed no corresponding increase in serum levels. Our data suggests there was poor correlation between oral vitamin D3 supplementation and improved BMD. Further studies are needed.

$n = 75$	Normal, $n = 39$ (52%)	Osteopenia, $n = 26$ (34.7%)	Osteoporosis, $n = 10$ (13.3%)
Mean age (years)	13.4	14.3	15
Range	(10–16.8)	(10–19.2)	(13–16.9)
Male (M)	15 (38.5%)	15 (58%)	6 (60%)
Female (F)	24 (61.5%)	11 (42%)	4 (40%)
Homozygous ΔF508	19 (48%)	13 (50%)	8 (80%)
Steroids	4 (10.2%)	8 (30.8%)	5 (50%)
Pancreatic Insufficiency	33 (84.6%)	23 (88.5%)	10 (100%)
Liver Disease	8 (20%)	3 (11%)	3 (30%)
Diabetes Mellitus	6 (15%)	6 (23%)	3 (30%)
Chronic Pseudomonas aerginosa	14 (35.9%)	12 (46%)	8 (80%)
Mean FEV 1 (% predicted)	79.1	72.8	52.3
Standard Deviation	±20.9	±21.9	±22.3
Mean BMI	19.8	18.3	17.6
Standard Deviation	±3.1	±3	±2.9

Prevalence and risk factors for bone demineralization in cystic fibrosis

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Introduction

Increased survival in cystic fibrosis patients has led to recognition of its skeletal complications. Decreased Bone Mineral Density (BMD), i.e., osteopenia and osteoporosis are an emerging problem for clinicians who care for children with Cystic Fibrosis (CF).

Aims

The aim of this study was to determine the prevalence and to assess risk factors for reduced BMD in a child population with CF.

Methods

A retrospective analysis was performed on all Dual–Energy X-ray Absorptiometry (DEXA) scan performed on children with cystic fibrosis with mean age 13.9 (10–19.2) years. DEXA scans over 7 years between 1/1/2000 and 31/3/2007 ($n = 78$) on Lunar DPX IQ, Lunar Prodigy and Lunar Prodigy Advance bone densitometers were examined. 3 patients were excluded because of insufficient data. Patient records were reviewed for clinical data. See Table.

Results

Bone Mineral Density (expressed as a z score) of L2-L4 spine and was reduced in a total of 48% patients. Osteopenia was defined as a z score ≥ -1 and osteoporosis as a z score of ≥ -2.5.

Conclusion

Our data suggest that reduced BMD is common (48%) and it is likely to start in early childhood. Reduced bone density in our sample was strongly associated with diminished lung function, chronic colonization with Pseudomonas aerginosa, homozygous ΔF508 mutation, pancreatic insufficiency and treatment with oral steroids. Of the patients with osteoporosis ($n = 10$) there was significantly poorer outcome. One patient died and one underwent successful double lung transplantation for end stage lung disease.

Acute motor axonal neuropathy with severe monomelic residual weakness: an unusual childhood manifestation of guillain barre syndrome

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The authors describe two cases of Acute Asymmetrical Lower Limb weakness as a result of Guillain Barre syndrome, a manifestion that has rarely been reported in Western Countries. Both cases had strikingly focal

unilateral limb involvement with EMG readings suggestive of an acute motor axonal or monomelic neuropathy. Both cases occured in the North West within a 9 month time period.

Case 1: A 16 month old boy was referred from a peripheral hospital with a 1 week history of lower limb weakness. His initial complaint was that of inability to weight bear his left leg and fever. He quickly developed progressive generalised weakness affecting his left leg more than right. There was a history of a recent Lower Respiratory tract infection (LRTI) but he was in no respiratory distress. His vaccinations were up to date and he had not received any recent vaccinations. Clinical examination was noted for a flaccid weakness of his left leg some weakness of the right leg. He had an absent knee and ankle jerk on the left with a tight achilles tendon. Sensation appeared to be intact. MRI of pelvis and spine was unremarkable. Lumbar puncture had a normal cell count but a raised CSF protein at 830 mg/dl. Initial EMG was suggestive of an Acute Axonal Guillain Barre. Stool for Campylobacter jejuni was negative and no anti-Glycolipid antibodies were detected. Metabolic screen was unrevealing and viral and bacteriological serology including polio, lyme and mycoplasma were negative. Blood and urine heavy metals was also negative. There was no clinical improvement despite iv Gammaglobulin and later Plasmapheresis. A repeat EMG showed progressive evidence of axonal degeneration of the left lower limb with multiple fibrilliations. Follow up shows persistent flaccid left lower limb weakness.

Case 2: A 2.5 year old boy was referred from a peripheral hospital for investigations of acute motor regression 3 months earlier, rash and two tonic clonic seizures. He had a history of a LRTI 3 months earlier after which he went off his feet. He had not been walking since then. Examination showed a flaccid weakness of both lower limbs, right more than left, with a foot drop on the left. Reflexes were absent. Sensation was intact. EMG showed an active axonal neuropathy of left lower limb. MRI brain and spine, CSF, metabolic work up including heavy metals were all normal. Follow up shows persistant flaccid left lower limb weakness.

Discussion

Guillain Barre (GB) is the commonest cause of acute motor paralysis in childhood. Two disease patterns are known to exist: Demyelinating and Axonal forms. Acute Inflammatory Demyelinating Polyneuropathy or (AIDP) is characterised by segmental demyelination of peripheral nerves in association with infiltration of inflammatory cells. Acute Motor Axonal Neuropathy or (AMAN) occurs without demyelination or inflammation. The primary axonal form was first described by Feasby et al. (1986). AMAN is rarly reported in western countries with studies showing an incidence of 3–7% of GB. This type is more common in China. The term AMAN was coined in early 1990s by McKhann et al. He noted epidemics during summer months, an association with *C jejuni* infection and many having antiglycolipid antibodies [1]. Two pathophysiological disease mechanisms are postulated: (i) Degeneration restricted to distal motor nerve terminal, immune mediated conduction block at the axonal membrane and complement mediated inflammation characterised by a rapid recovery. (ii) Wallerian-like degeneration of motor fibres on necropsy, myelin disruption at nodes of Ranvier, macrophage extrusion into periaxonal space at the paranode, separation of the axon from adjacent Schwann cell membrane and subsequent degeneration of Schwann cell cytoplasm and axon tending to cause a slower recovery. Our cases fit better with the latter group.

These two patients have very significant persistent unilateral lower limb weakness which will likely result in lifelong disability. Hiraga et al. showed that most of severely disabled AMAN patients who were unable to walk at 6 months may still show improvement over a period of years [2]. However, little is known about the long term prognosis of AMAN in children.

The focal nature of presentation in our cases is unusual and the temporal and geographical clustering raises the question of other unidentified pathogenic factors.

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Children with convulsive status epilepticus (CSE) presenting to Cork University Hospital: an audit of documentation and management

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Aim

To identify the characteristics of children presenting to Cork University Hospital with convulsive status epilepticus (CSE) and to identify the timing, management,

and effectiveness of each of the therapeutic interventions and finally to audit the documentation of paediatric CSE at our institution.

Methods

A retrospective audit of all children presenting in CSE over the 1.5 year period between July 1 2004 and December 31 2005. Cases were identified through the discharge database and discharge summaries.

Results

19 cases of CSE were identified. Male to female ratio was equal. No cases were under 6 weeks of age, 1 case was between 6 weeks to 1 year and 18 cases greater than 1 year. 73.6% had a known seizure history. 57.8% had a first episode of CSE.

Documentation of out of hospital seizure duration and total seizure duration was poor (50% of each). The average documented seizure duration pre hospital was long at 47 min, range 0–70 min. Only 38% receiving pre hospital treatment. Treatment on arrival tended to overuse various benzodiazepines and there was poor compliance with protocol.

Cessation of CSE was achieved in 15 cases (83.3%, 15/18) with a combination of first and second line treatment. Eleven responded to first line treatment with benzodiazepines. Four responded to second line treatment with phenobarbitone or phenytoin. Six underwent rapid sequence induction. Three cases were intubated for respiratory complications. Two cases were induced for seizure control with thiopentone. One case is unclear from documentation reason for intubation (transfer). 44.4% (8/18) were admitted to ICU. There was one death from prolonged CSE.

Conclusion

Protocolised management of prolonged seizures is essential in order to prevent the morbidity and mortality associated with CSE. Our results show a lack of seizure management in the community and poor protocol compliance in the hospital setting. Given the long journey time to hospital and prolonged seizure duration pre hospital early adequate seizure management within the community is imperative. Following the implementation of a major education drive both in the community and hospital by the paediatric neurology team and the widespread use of buccal midazolam we are confident that a re audit will show improved results.

The culpit in post vaccine related seizures; SCN1A mutations

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The gene for the voltage-gated sodium channel alpha-subunit type 1 (SCN1A) has been implicated in a rapidly expanding variety of epilepsy syndromes, including generalised epilepsy with febrile seizures plus (GEFS +), severe myoclonic epilepsy of infancy (SMEI) and idiopathic childhood epilepsy with generalised tonic-clonic seizures (ICEGTC). These clinically related but diverse conditions have been associated with de novo mutations in the SCN1A gene. Post vaccine related seizures – previously attributed to pertussis vaccine, are now known to be associated with SCN1A gene mutations.

We report the case of a 6 month old infant who presented with a prolonged (> 1 h) focal seizure, not associated with pyrexia, occurring less than 24 h after the second dose of the ''5 in 1'' vaccine.

The infant had been developmentally normal prior to admission. There was no family history of epilepsy or febrile convulsions.

On initial presentation at 6 months three focal seizure episodes on alternating sides, with no recorded fever, were reported.

Over the next 2 months, our patient presented on three more occasions with alternating left or right sided focal seizures. On her second admission, her first febrile seizure, with rhythmic jerking of the right side and left facial twitching, was noted. She subsequently was admitted to ICU after a 30 min episode of right arm jerking associated with hypoxia.

She presented to A/E 10 days later with a history of a 4 min tonic clonic seizure and 9 days later returned with a prolonged 55 min febrile seizure, which commensed with right hand fisting and proceeded to generalized tonic-clonic activity.

Her interictal neurological exam has remained within normal limits, apart from the finding of mild pigmentary retinopathy. A Todd's palsy post-ictally was noted on her ICU admission.

Her investigations to date include normal brain MRI. EEG showed focal slowing in the right hemisphere. Extensive metabolic investigations are normal to date, awaiting skin and muscle biopsy. SCN1A gene analysis revealed a positive splice site mutation associated with the clinical phenotype of SMEI.

Severe seizures/encephalopathy occurring post vaccination was previously attributed to pertussis vaccine. This is the first reported Irish case of such seizures, now known to be due to SCN1A gene mutations.

The values of the ultrasound scan as a diagnostic tool duringthe pre-and postnatal period in a patient with osteogenesis imperfecta

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Aim

To explore the value and limitations of the ultrasound scan (USS) both ante- and postnatally in a severe form of Osteogenesis Imperfecta (OI).

Method

Based on an index case—Review of antenatal USS in a severe case of OI, and the prospective evaluation of USS as a diagnostic tool in detecting long bone fractures in the postnatal period.

Results

1. Antenatal: USS in the antenatal period initially only detected bilateral rhizomelia and asymmetrical femoral shortening. However, on postnatal review of the scans, long bone fractures could be demonstrated on certain views.
2. Postnatal: Following the lead of clinical suspicion, three long bone fractures were identified postnatally using USS. X-ray all later confirmed these.

Discussion

Where a significant right-to-left discrepancy exists between long bone measurements on antenatal USS, and where additional features such as antenatal fractures are present, the alarm towards possible OI should be raised. Unfortunately, as demonstrated in this case, there are shortcomings in the use of the antenatal USS regarding the detection of fractures, seeing the view at a given time is dimension limited and not all views of the same long bone will demonstrate an existing fracture. In this particular case the femur appeared to be hypo plastic due to overlapping of the long bone segments, and because of the sonographic window, appearance of the long bone was as if continuous. Suspecting OI *antenatally* by recognising features as proposed above is of great value in improving the outcome of the infant.

USS used during the postnatal period to confirm suspected fractures in OI can potentially be of great value in dramatically reducing the use of X-rays and thus the associated risks of repeated exposures of infants to radiation. Even though postnatal US is not the gold standard for fracture confirmation, in an infant with current and future potential x-ray exposure, perhaps USS could play a key role in detecting major fractures especially when a particular long bone is clinically in question.

Low superior vena cava flow (SVC) states in very low birth weight infants (VLBW), flow measurements in anterior cerebral artery (ACA) and adverse outcome

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Aims

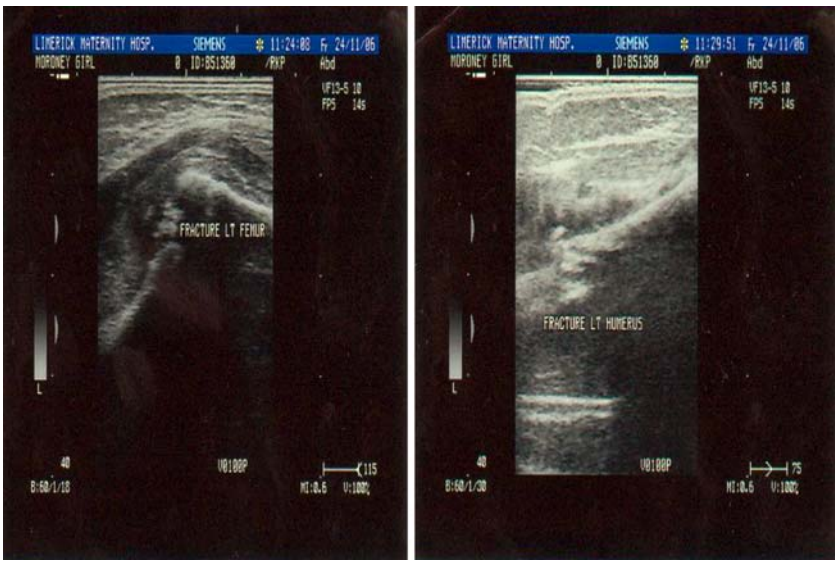
(i) To document relation between low SVC flow states and intracranial haemorrhage and/or early neonatal mortality (ii) To determine the relationship between superior vena cava (SVC) flow and blood flow velocity patterns in anterior cerebral artery (ACA).

Methods

A prospective observational cohort study. Neonates with birth weight less than 1,500 g were eligible for enrollment. Newborns with congenital heart disease (excluding patent ductus arteriosus) or major congenital malformations were excluded. Echocardiographic evaluation of flow (SVC, left and right ventricular output, patent ductus arteriosus) was performed in the first 24 h of life. Cranial ultrasound examinations including blood flow velocity measurements at ACA (including systolic, diastolic and mean velocities, pulsatility index/PI/and resistance index/RI/) were performed simultaneously. The primary endpoint was the relationship between low SVC flow states (less than 40 ml/kg/min) and adverse outcome defined as intraventricular haemorrhage grade ≥ II and/or early neonatal death. The secondary goal was to assess ACA blood flow velocity values as a predictor of SVC flow.

Results

Between November 2006 and April 2007 27 VLBW neonates were enrolled following parental consent. Three patients were excluded after enrollment (perimembranous VSD, muscular VSD, trisomy 21 with muscular VSD). Five babies had low SVC flow states (less than 40 ml/kg/min). There were no differences between the baseline characteristics of both groups (low SVC



versus normal SVC) –mean birth weight 1.12 versus 1.02 kg ($p = 0.5$), gestational age was 26.8 versus 27.7 weeks ($p = 0.43$) and hours of life at exam were 18.2 versus 19.8 ($p = 0.49$). The incidence of IVH ≥ grade II and/or early neonatal death was 16.7%. The combined outcome of IVH ≥ grade II and/or early neonatal death was 40% low SVC state versus 10.5% normal flow ($p = 0.18$, OR 5.67, 95% CI 0.28 to 99.66). There was a poor correlation between SVC flow and ACA PI. Mean ACA RI was 0.764 versus 0.728, respectively ($p = 0.45$, 95% CI –0.06 to 0.13).

Conclusion

There was a tendency towards a higher incidence of early neonatal death and/or intraventricular haemorrhage with low SVC states in first 24 h. These measurements may aid in determining which patients may benefit from cardiovascular support. ACA blood velocity measurement seems to be of no benefit to predict low SVC flow states.

Procedure related scalded skin in neonates: primum non-nocere

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Introduction

Literature suggests that Staphylococcal Scalded Skin Syndrome (SSSS) can manifest following inoculation of an area of broken skin such as umbilical cord stump or surgical scar. The incidence is reported as between 0.09 and 0.13 cases per million in the general population, occurring in the majority of cases between the ages of 2 and 5 years. It is recognized to occur in clusters in the intensive care, however, reports of SSSS in the neonatal setting among full-term healthy babies is scanty. When the infection has an identifiable site of original inoculation, there is an opportunity to isolate the common source.

Method

We report a cluster of cases of SSSS in a 1 month period. A case was defined as an infant with blistering or peeling skin, and with exfoliative toxin A *Staphylococcus aureus* positive cultures. Four babies with no common relationship, who were previously discharged home well from the post-natal wards, presented to the Accident and Emergency Department within a short period of time with a blistering soft tissue infection. Staphylococcal scalded skin infection was immediately suspected in each case. All cases developed into a widespread infection. Upon investigation of each case by Paediatric and Infection Control Team, the site of inoculation was clearly identified as skin interruption in aid of national health protection schemes in cases 1, 2 and 3. These occurred following the site of a heal prick done as part of the Guthrie card national screening process in cases 1 and 2; and in the case 3 following administration of vitamin K intramuscular injection. Both of these procedures are performed as part of National Health Protection Schemes. In case 4 the blistering began on chin and perineum.

Swabs were sent for microbiological analysis and an epidemiological investigation was commenced following the first three cases. This included date of

birth, type of delivery, ward and room occupied during inpatient stay in the Maternity Hospital, Obstetric and Paediatric teams involved in patient delivery and discharge checks, and persons involved in National Health Protection Scheme procedures.

Results

Preliminary investigation by the infection control team identified that none of these babies were admitted to the Neonatal Unit. Two babies shared the same date of birth, two were delivered by caesarean section (1 emergency). None of the babies shared the same consultant, however one member of the team was linked with two of the cases, and a common ward was linked to two of the cases.

Analysis of the initial preliminary testing cultured *Staphylococcus aureus* from all four patients. In patients 1, 2 and 3 toxin Type A Group II phages were deciphered using molecular epidemiological typing. Pulse Field Gel Electrophoresis (PFGE) demonstrated indistinguishable patterns compelling the suggestion of a common point source of infection. Cultures from Case 4 were found to be a different strain.

Discussion

Side effects of skin interruption in aid of health protection are well documented. It is scarce, however, for such a sinister infection to result. Our literature search did not reveal previous reports of staphylococcal scalded skin in full-term otherwise healthy babies evolving as a complication of public health measures. Although the exact source of infection has not yet been determined for each case, this cluster of staphylococcal infection re-emphasizes the importance in practicing universal precautions and clean technique in all procedures including standard measures to which all babies are subject.

Unusual cause of developmental delay and failure to thrive

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A 16 month old Irish-born child of Nigerian origin was referred for evaluation of developmental delay, and failure to thrive. He was born at full term, birth weight was 2.8 kg. He was breastfed and had weaned to solids. Development was within normal limits until 7 months. Thereafter developmental milestones were delayed.

Initial assessment showed weight was on the 0.4th centile, height on the 9th centile and head circumference was on the 0.4th centile. Development was at a 9 month level, there was firm hepatomegaly of 5 cm below the costal margin and bilateral cataracts.

A complete workup for developmental delay, failure to thrive, cataracts and hepatomegaly was done. Investigations revealed iron deficiency anaemia with neutropaenia, elevated liver function (AST- 56 U/l, ALT 34 U/l), urine was negative for reducing substances. Newborn screening was reviewed and repeated and both were negative for galactosaemia. Beutler test, however, showed no galactose-1-phosphate uridyl transferase (GALT) activity and Galactose-1-Phosphate was high (1.88 umol/gmHb), consistent with untreated galactosaemia. Enzyme analysis revealed GALT activity to be < 0.5 umolsub/H/gmHb confirming classical galactosaemia. Glucose-6-dehydrogenase (G-6-PD) assay showed deficient G6PD. Magnetic resonant imaging (MRI) brain showed delayed myelination.

Molecular sequencing for the GALT gene showed homozygosity for the GALT mutation p.Ser135Leu which is found almost entirely in individuals of African origin and accounts for approximately 48% of African –American and 91% of African Negroid GALT alleles, respectively. This mutation is associated with 10% residual activity in some tissues such as the liver and leucocytes thus explaining the late presentation and the negative newborn screening.

G6PD deficiency is a cause of false positive Beutler as G6PD is required for the Gal-1-PUT enzyme activity in this assay.

Diagnoses of G6PD and classical galactosaemia were made and galactose free diet was commenced and advice given on what drugs and food to avoid to prevent haemolysis.

On follow up he is now thriving however little developmental progress has been made.

In summary this 16 month old Irish-born Nigerian who presented with failure to thrive, gross developmental delay, cataracts and hepatomegaly was found to have abnormal liver function, G6PD deficiency and a late diagnosis of classical galactoaemia.

This presentation would be very unusual in the indigent Irish population where the common GALT mutation is Q188R. It is important to note that immigrant children may have unusual presentations of classical metabolic disorders due to different genetic mutations.

An audit of nitric oxide therapy provision during neonatal transports

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National Neonatal Transport Programme (NNTP)

Introduction

The use of inhaled Nitric Oxide (iNO) therapy in neonatal centres in Ireland necessitates the continuation of this therapy when infants require transfer. The provision of iNO in the transport environment involves additional logistical and safety concerns which the NNTP has addressed and since January 2006 the availability of iNO therapy on NNTP transports has been routine. The necessary equipment for iNO therapy is kept in the NNTP ambulance and a bioengineer travels with the team where possible. Staff training and criteria for the use of iNO therapy in transport are in place. iNO is initiated in consultation with the referring and receiving hospital consultants based on infant diagnosis, oxygenation index and ventilatory requirements. One commenced, iNO therapy is continued for the duration of the transport.

Objective

To review the use of iNo therapy by the NNTP since its routine availability.

Methods

The audit takes the form of a retrospective study of all NNTP transports that have involved the use of iNO, dating from January 2005 to March 2007. Details recorded on the NNTP's database and individual transport case notes were reviewed. Parameters assessed were: mobilisation times, diagnosis on transport, mode of transport and demographics. The clinical status of the infants involved were also reviewed by noting: the gestational age, age on transport, iNO therapy initiation times, pre and post iNO ventilation and oxygen requirements and stabilisation times. Any other associated problems were also identified.

Results

A total of 21 transport during the study period involved the use of inhaled Nitric Oxide therapy (iNO). 16(76%) referrals for iNO were from regional neonatal centres and the remaining 5 (34%) were within Dublin. One of these transports was by air and the remaining 20 by road ambulance Although the iNO system had been set up in anticipation of being used, in 5 (23%) of cases iNO was not required. Since the NNTP has been equipped to provide iNO routinely, the mobilisation times have improved from a mean of 81 min in 2005 to a mean 43 min in 2006. Fourteen (66%) infants were a gestational age of >37 weeks when transferred and 14(66%) were in the first 48 h of life. The five infants who did not require iNo were diagnosed on transport with sepsis in 3 cases and Congenital Heart Disease (CHD) in 2. The diagnosis of those who travelled on iNo therapy were: Persistent Pulmonary Hypertension of the Newborn, 6 (37.5%), Meconium Aspiration Syndrome, 4 (25%), Congenital Diaphragmatic Hernia, 3 (19%), Patent Ductus Arteriosis, 2 (12.5%) and Respiratory Distress Syndrome, 1(6%0. Of the 16 infants who were transported on iNO, 6 (37.5%) had commenced iNO therapy prior to NNTP team's arrival and in 10 (62.5%) of cases the NNTP initiated the treatment. The ventiallatory requirements decreased in 6 (60%) infants following the initiation of iNo by the NNTP team. Oxygenation also improved in 6 (60%) of these cases. There was no change in the respiratory status of 3 (30%). Of those infants who were already receiving ino on arrival of the NNTP team, their status remained unchanged for the duration of the transport.. Three (14%) infants were deemed too unstable to proceed with transportation. The team remained at the referring hospital overnight on two of these occasions and were able to proceed with the transport the following day.

Conclusions

In line with the easier access to iNO on NNTP transports, the utilisation of this therapy has increased. The NNTP mobilisation times when using nitric oxide are half of those recorded in 2005. There have been no major incidences associated with the use of iNO during NNTP transports to date. The availability of iNo during NNTP transports has enabled the timely transfer of infants who require specialist care.

Metronome use may optimize ventilation with the neopuff resuscitator device

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Introduction

A self-inflating(Laerdal) bag, a flow-inflating(anaesthetic) bag or a T-piece device(Neopuff) are all acceptable devices which can be used to ventilate newborn infants either via a face mask or endotracheal tube [1]. The Neopuff maintains functional residual capacity (FRC) by providing a consistent positive end expiratory pressure (PEEP) throughout the resuscitation process. Previously we have demonstrated a major increase in inspiratory time (IT) is associated with operator inexperience using the Neopuff device. In addition a long IT

is associated with a significant increase in the rates of all air leak (pneumothorax, pulmonary interstitial emphysema, pneumomediastinum, pneumoperitoneum) [2].

Aims

To compare bag and mask to Neopuff ventilation by different operators on inspiratory time, respiratory rate (breaths per minute), tidal volume and minute volume (expiratory tidal volume times the respiratory rate in breaths per minute). To examine the impact of metronome use on these indices using both devices.

	Self-inflating Bag		Neopuff		*p* value		
	Control	Metronome	Control	Metronome			
n	230	230	230	230	a	b	c
IT (s)	0.5 ± 0.9	0.3 ± 0.1	0.7 ± 0.8	0.5 ± 0.4	< 0.0001	< 0.0001	< 0.0001
RR (bpm)	40.0 ± 21.0	43.2 ± 30.5	42.6 ± 26.8	39.9 ± 15.7	0.016	0.6	0.21
TV (ml)	15.5 ± 17.9	16.7 ± 7.9	11.9 ± 2.1	12.1 ± 1.8	0.003	< 0.0001	< 0.0001

IT inspiratory time; *RR* respiratory rate; *TV* tidal volume

a Bag con vs. met; b Neo con vs. metro; c Con bag vs. neo

Design

Twenty-three staff members of a tertiary neonatal unit (4 Consultant Neonatologists, 2 Neonate Research Fellows, 4 Registrars, 4 Senior House Officers, 8 Neonatal staff nurses including 3 sisters, and 1 midwife) participated in the study. Each participant was asked to setup the Neopuff device at a target PIP of 20 cmH_2O and PEEP of 5 cmH_2O. They were then directed to perform ventilation at a rate of 40 breaths per minute. Once the subject was satisfied that they were ventilating at approximately a rate of 40 bpm for 1 min the metronome was commenced and the effect observed. This was repeated using bag and mask ventilation.

Results

Data analysis was performed on segments lasting 10 breaths allowing examination of 230 datapoints in each category: Self-inflating bag compared with Neopuff either alone or with metronome. Inspiratory time was significantly increased using Neopuff compared with the Self-inflating bag but normalised using the metronome. Similar results were found for tidal volume but respiratory rate was comparable in all groups.

Conclusions

Metronome use improves inspiratory times especially when using the Neopuff device. Routine use a metronome in the delivery room could decrease the risk of airleak related to increased inspiratory times.

References

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Fetal lung volume by three-dimensional ultrasound and prediction of neonatal respiratory outcome

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Background

Abnormal antenatal lung growth can occur as the result of a number of conditions, but has been difficult to diagnose. Three dimensional ultrasonography has made it possible to measure fetal lung volumes and it has been established that lung volumes increase exponentially with gestation. It is important to determine whether fetal lung volumes in conditions associated with abnormal lung growth are predictive of outcome.

Aim

To examine the relationship between fetal lung volumes (FLV) measured by three dimensional ultrasound and postnatal measures of respiratory outcome (lung volume and duration of oxygen dependency) in patients with congenital diaphragmatic hernia (CDH) or anterior abdominal wall defects (AWD).

Methods

Fetal lung volumes were measured in nine fetuses (four with left sided CDH [three liver-up, one liver-down], four with gastroschisis and one with exomphalos) at a median of 31 (range 24–35) weeks of gestation. Fetal lung volumes were expressed as a ratio of the observed to the expected for gestational age. Lung volume was assessed by measurement of functional residual capacity (FRC) using a helium gas dilution technique at a median of 8 (range 3–32) days after birth. FRC was related to

bodyweight at the time of measurement. The durations of oxygen dependency were obtained by review of the patients' notes.

Results

The ratio of observed to expected fetal lung volumes correlated significantly with both FRC ($r = 0.667$, $p = 0.05$) and the duration of oxygen dependency ($r = –0.8$, $p = 0.01$).

Conclusion

These results suggest assessment of fetal lung volume in cases suspected of abnormal lung growth is predictive of neonatal respiratory outcome and therefore may be useful regarding counselling parents and determining whether therapeutic interventions might be helpful.