G81 PARENTAL DECISION-MAKING IN HYPOPLASTIC LEFT HEART SYNDROME: THE ROLE OF A CARING DOCTOR

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[1]S Bertaud, [2]S Suguna Narasimhulu, [2]A Ludormirsky. *[1]Paediatrics, Croydon University Hospital, London, UK; [2]Pediatric Cardiology, NYU Langone Medical Center, New York, USA*

Aims We sought to explore what parents value from their physicians when deciding on treatment for Hypoplastic Left Heart Syndrome (HLHS). We were interested in parental views on the decision-making process, the factors which influence their choices and in the level of physician involvement they desire.

Methods Eight families who had received a diagnosis of HLHS in the last 3 years and had chosen surgical intervention underwent structured interviews lasting between 1–3 hours. Patients were de-identified and results discussed with two Paediatric Cardiologists and a Professor of Bioethics to identify key themes.

Result Parental understanding of HLHS was varied; 6 of the 8 families displayed limited understanding of the long-term complications and quality-of-life issues. All remembered feelings of shock and confusion at the time of diagnosis.

When discussing what motivated their decision to choose surgery the parents all expressed ideas about the ability to give their child hope. When asked how important the following factors were in making their decision, the mean results were: (1: 'not important at all'; 5: 'very important') the doctor's opinion: 3.6; other people's opinion: 2.0; religion: 3.8; financial considerations: 1.9; other commitments: 2.9

Seven families felt the decision was entirely up to them. One mother instead said she, her husband and the doctors were "all together" in making the decision. Seven families said the information was presented to them in a neutral manner by the doctor. Seven families felt their doctors were optimistic about their child's outcome. Three families asked for their doctor's opinion on treatment whilst five did not, stating it was irrelevant to their decision. Seven families said parents should make decisions regarding treatment of HLHS. One father said the doctors should make the decision, whilst his partner felt it should be both parties. All eight families felt content with their decision and none expressed regrets.

Conclusion Our findings offer valuable insight into the parental experience and indicate that most families with a diagnosis of HLHS felt the decision-making was entirely up to them and that information from doctors was neutral but nonetheless optimistic. Whilst parents wish to remain the final arbiters of the decision, some do value the doctor's opinion.

G82 STUDY OF ACUTE LIVER FAILURE IN NEWBORNS AND YOUNG CHILDREN WITH AN UNDERLYING INHERITED METABOLIC DISEASE

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[1]RM Hegarty, [2]A Dhawan, [3]P Gissen. *[1]Neonatal Unit, Guy's and St Thomas' NHS Trust, London, UK; [2]Paediatric Liver Unit, King's College NHS Trust, London, UK; [3]Department of Metabolic Medicine, Great Ormond Street Hospital for Children, London, UK*

Aims To study the demographic, clinical and laboratory findings, diagnoses and outcome of children under 5 years who were admitted with acute liver failure (ALF; INR > 2.0 or INR > 1.5 and encephalopathy) seconday to an underlying inherited metabolic disease (IMD).

Methods A retrospective case note review of children who were admitted between January 2001 to 2012 to a tertiary paediatric liver unit with ALF and a multi-centre review of their long term outcome.

Results A total of 127 children were identified from the database. 36 children (28%; 17 boys; median presenting age 6 weeks, range 1 day-41 months) had an underlying IMD including galactosemia in 17, mitochondrial cytopathy in 7, ornithine transcarbamylase (OTC) deficiency in 4, tyrosinemia type 1 in 4, Niemann-Pick C (NPC) in 3 and congenital disorder of glycosylation type 1 in 1. The remaining aetiologies were: indeterminate in 40 (32%), infectious in 15 (12%), neonatal hemochromatosis in 11 (9%), hemophagocytic syndrome in 8 (6%), drug toxicity in 5 (4%) and other in 10 (8%). Of the 36 children with an IMD consanguinity was present in 16 (44%), developmental delay in 3 (8%), jaundice at presentation in 28 (78%), hepatomegaly in 27 (75%) and encephalopathy in 8 (22%). The median peak (range) INR 4.8 (1.8–15), aspartate transaminase 334umol/L (39–15791) and bilirubin 227umol/L (13–692). Liver biopsy was done in 9 children (25%), neuroimaging in 10 children (28%), bone marrow aspiration in 7 (19%) and muscle biopsy in 5 (14%). 29/36 children with an IMD survived (81%). 4 children with mitochondrial cytopathy (including 1 after transplantation during the postoperative period) and 3 with NPC died. 4 children (1 OTC deficiency; 3 mitochondrial cytopathy) underwent liver transplantation. Follow up data was available for 23 children (mean follow up period, 4 years 3 months) in whom 13 (57%) were identified as having evidence of developmental delay.

Conclusion IMD is a common cause of ALF in children. Indeterminate cases may include undiagnosed metabolic diseases. Survival of children with IMD-related ALF is good, however, long term developmental outcome is less favourable.

G83 NEONATAL ECHOCARDIOGRAPHY BY NEONATOLOGISTS: EXPERIENCE SO FAR

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[1]VM Shetthalli, [1]S Sen, [2]DG Wilson, [1]S Reddy. *[1]David Ferguson Neonatal Unit, Royal Gwent Hospital, Newport, UK; [2]Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, UK*

Background Neonatal Echocardiography is an important investigation for assessment and clinical management of sick preterm neonate and is also essential for diagnosis of congenital heart disease. Not all neonatal intensive care units are equipped with paediatric cardiology services. Many units in UK and internationally have neonatologists doing in house echocardiograms in their units and outpatient murmur clinics. This is one of the largest study which aims to access the validity of echocardiograms done by the neonatologists compared to the cardiologists.

Methods This is a retrospective study over a 7½ year period in a level 3 neonatal intensive care unit. Echocardiograms were performed either in neonatal unit or outpatient murmur clinics. Selected abnormal examinations were referred to regional cardiology services. Database of all the scans were maintained electronically. This neonatal database was compared with regional paediatric cardiology database called Cardiobase® to assess concordance and discordance of the findings in referred patients.

Results A total of 2325 scans were performed on 1639 neonates from 1st September 2003 till 31st December 2011. Cardiobase® data was available for 454 scans. Concordance was found in 91.8% of these scans and partial concordance was found in further 4.4% of scans. 31 infants (1.6% of total infants scanned) had critical structural abnormalities and all of these were identified correctly. 3.7% of the scans had discordant findings and most of these were false positives and they were referred to exclude critical events. Six scans had had false negative findings. Sensitivity and specificity for the scans done by neonatologists was 98%.

Abstract G83 Table 1 Concordance rates

Cardiobase data (Total)	454 scans	100%
Concordance	417 scans	91.8%
Partial Concordance	20 scans	4.4%
Discordance	17 scans	3.7%

Abstract G83 Table 2 Sensitivity and specificitiy rates

Sensitivity	98.4%
Specificity	98.9%
Positive PV	97.2%
Negative PV	99.4%

Conclusion Neonatal echocardiography by neonatologists have high concordance rates and have a high sensitivity and specificity in detecting congenital heart diseases. With appropriate Paediatric Cardiology support Neonatal Echocardiography by neonatologists can be a safe and reliable tool.

G84 A PARENTS VIEW OF CARDIAC SCREENING FOR DOWN SYNDROME

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E Pascall, R Tulloh. *Medical School, University of Bristol, Bristol, UK*

Background Cardiac disorders are common in children with Down syndrome and the Down Syndrome Medical Interest Group (DSMIG) guidelines were updated in 2007. This project aims to review if parents thought these standards were being met.
Methods A survey reflecting the guidelines was posted by the Down Syndrome Heart Group on their webpage and on facebook. Parent responders shared the page in order to more replies. The questionnaire was intended to identify when the diagnosis of Down Syndrome was made, and the time it took for a cardiologist referral and echocardiogram.
Results 98 responses were collected and analysed. 85 responders lived in England (86.7%). 23.65% were diagnosed with Down syndrome prenatally, 70.25% were diagnosed within one week of birth and 6.1% more than one week after birth. 94.45% underwent foetal echocardiography of which 54.1% had the diagnosis confirmed after birth and 94.5% were seen by a paediatric cardiologist within 2 weeks after birth. Of those who did not undergo foetal echocardiography, 71.4% were seen within 6 weeks of birth, in whom 42.9% were found to have congenital heart disease. 73.4% of those diagnosed with Down syndrome within one week of age had an ECG at this time. Only 84% of those with abnormal ECG were referred and seen by a paediatric cardiologist before 2 weeks of age. 14.2% were not seen by a cardiologist or underwent ECG within 6 weeks.
Conclusions The results of this parent led questionnaire show the majority of babies with Down syndrome are diagnosed within one week of birth. Most of the 2007 guidelines set by the DSMIG are being broadly met, however more emphasis should be made on meeting the deadlines for paediatric cardiology review and echocardiogram. This applies to whether diagnosis is made prenatally, within one week of birth or more than one week after birth. In addition only 73% of those that are diagnosed within a week undergo an immediate ECG. More importance needs to be based on adhering to the guidelines and reducing parental uncertainty about congenital heart disease in Down syndrome.

British Paediatric Allergy, Immunology and Infection Group

G85 ADRENALINE USE IN ANAPHYLAXIS IN PAEDIATRIC WARDS IN UK

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[1]M Sundaram, [2]S McGowan-Smyth, [1]B Tharayil, [1, 2]M Samuels. *[1]Academic Department of Paediatrics, University Hospital of North Staffordshire, Stoke on Trent, UK; [2]Keele University School of Medicine, Newcastle under Lyme, UK*

Background Resuscitation Council Guidelines (RC-UK, 2008) for treating anaphylaxis advocate intramuscular adrenaline in doses of 150, 300 or 500 micrograms, according to age bands [1]. However, ALS guidelines recommend weight-based calculations of 10 micrograms/kg IM, leading to administration of a range of volumes [2].
Aims A survey was conducted to evaluate the availability of fixed dose Epipens versus adrenaline vials in paediatric wards and radiology departments in England.
Methods The questionnaire was sent to 105 paediatric pharmacists at the various paediatric units in UK.
Results 60% responding hospitals had adrenaline available, half of them in vials and 85% with prefilled variable-dose syringes. In 53% units, wards stocked adrenaline 1:10,000 and 64% also had 1:1000. 19% hospitals stocked Epipens on crash trolleys and it was available in 48% wards. Adrenaline was given according to a weight-based dose in 57% wards. For contrast studies, adrenaline was available in 68% departments, 9.5% of which stocked Epipen.
Conclusion Most units still use weight-based doses of adrenaline from vials or pre-filled syringes, with Epipens being available in less than half of units. Adrenaline must always be available on wards and in radiology departments, as most arrests from anaphylaxis occur within 10 minutes. To ensure compliance with RC-UK guidelines, either all wards should stock Epipens or the guidelines should reflect practise and recommend weight-based calculated doses.

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G86 ANAPEN, EPIPEN AND JEXT AUTO-INJECTORS; ASSESSMENT OF SUCCESSFUL USE AFTER CURRENT TRAINING PACKAGE

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[1]HM Luckhurst, [2]D Tuthill, [2]J Brown, [2]E Spear, [2]J Pitcher. *[1]School of Medicine, Cardiff University, Cardiff, UK; [2]General Paediatrics, Cardiff and Vale UHB, Cardiff, UK*

Background Anaphylaxis is a severe life threatening allergic reaction. Prompt administration of epinephrine(adrenaline) is the first line treatment. There are currently three epinephrine auto-injector devices available in the UK; original Anapen, new EpiPen and Jext, each of which differ in their advised method of use. International standards recommend training for all patients prescribed epinephrine auto-injectors, we meet these. If families can more successfully use a particular trainer device, this may have important clinical effects.
Aims To assess the effectiveness of the training by evaluating "epinephrine naive" families' ability to successfully use an auto-injector trainer device.
Methods Adults and children over 12, with no experience of auto-injector use were invited to participate in this service evaluation. They were randomly allocated to be trained in the use of one of the available auto-injectors. Their performance was assessed using a ten point marking sheet based on the correct method of administration of epinephrine for the individual device. Six marks were for procedures identical to all three devices (e.g. massage the site of injection) and four were device specific to reflect the differences in administration technique. Success rates were analysed by Chi-square with $p < 0.05$ being deemed significant (http://graphpad.com/quickcalcs/contingency 2).
Results There were 120 participants.