Cardiology

**CONGENITAL HEART DISEASE IN PREMATURE INFANTS**

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Aims: To assess the prevalence at live birth and spectrum of congenital heart disease in premature infants.

Methods: Ascertainment of all congenital heart disease (CHD) diagnosed in infancy in the resident population of one health region in 1987-2000. All babies with PDA or ASD were excluded to avoid selection bias. All babies with a gestation at birth of <37 weeks were identified. Data from our region and from the literature predict that 5.6% of all babies will be live born at 32-36 weeks, 0.85% at 28-31 weeks, and 0.35% at <28 weeks. Thus 6.8% of live born babies will be pre-term.

Results: Of 492,559 live births in the 14 years of the study, there were 2795 with CHD (excluding PDA and ASD), a prevalence at live birth of 5.7/1000. CHD was present in 5.1/1000 of term babies and 13.4/1000 of pre-term babies. The odds ratio for CHD in prematurity birth of 5.7/1000. CHD was present in 5.1/1000 of term babies and be pre-term.

Conclusions: There is a greater than two-fold excess of congenital heart disease in pre-term compared with term babies. 16% of all babies with CHD are pre-term. This excess is more marked for specific malformations, particularly pulmonary atresia with VSD and complete AV septal defect.

**THE RELATIONSHIP BETWEEN THE QT AND QTc INTERVAL AND TEMPERATURE DURING WHOLE BODY MILD HYPOTHERMIA IN NEONATES**

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Background: Prolongation of the QT interval during accidental hypothermia is well described and is considered an ominous sign because of its association with ventricular tachycardias exhibiting the specific configuration known as torsade de pointes. Few human studies have reported the effect of mild hypothermia on the QT interval. The aim of our study was to explore the changes in the QT and QTc interval during prolonged whole body mild hypothermia in neonates receiving extracorporeal membrane oxygenation (ECMO).

Methods: Twenty seven neonates (median gestation 40 weeks; range 33-41 weeks) referred for ECMO at the Glenfield Hospital, Leicester were enrolled in the study. Five groups (N=5 per group) were each studied for the first five days of ECMO. The first group was maintained at 37°C throughout the study period. Subsequent groups were cooled to 36°C, 35°C and finally 34°C respectively for the first 24 hours and the final group for the first 48 hours of ECMO before being rewarmed to 37°C. Using a 24 hour digital monitor, the QT and QTc values were recorded continuously during the cooling and rewarming period. A standard 12 lead ECG was recorded daily during the same time period and the QT and QTc interval measured.

Results: Regression analysis of the digitally measured QTc interval for groups 2-4 during the first 48 hours of ECMO showed that there was a small effect of temperature. However, the intragroup variance was large compared with the intergroup variance. The coefficient of determination was -3.1x10^-2°C with an intercept of 564x10^-2 seconds. From the digital recordings during the first 24 hours of cooling, the median (range) QT interval measurements for groups 2-4 were 326无缘了seconds (263-395), 335ms (260-425), 328ms (208-444) and 349ms (273-408) respectively.

Conclusions: The QTc interval changed with temperature. However, the large intragroup variance suggests that there are factors other than temperature which influence the QT interval.

**PREVALENCE OF COMPONENTS OF THE INSULIN RESISTANCE SYNDROME (IRS) IN A MULTI-RACIAL SAMPLE OF OBSESE CHILDREN AND ADOLESCENTS**

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Aims: The insulin resistance syndrome (IRS) is strongly predictive of increased risk of diabetes and cardiovascular disease in adults. Components of the IRS include abnormal glucose homeostasis (AGH) (hyperinsulinism or impaired glucose tolerance), hypertension (HT) and dyslipidaemia (DL). Little is known regarding the prevalence of components of the IRS in obese children and adolescents in the UK, particularly in ethnicities at high risk of type 2 diabetes.

Methods: Assessments of glucose homeostasis, sitting blood pressure, and fasting blood lipids were performed on 95 children and adolescents aged 3-18 years referred for assessment of obesity. All were obese (BMI >95%) and none were known to have diabetes. Ethnicity: White (56%), Asian (23%), Black (17%), Mixed (5%). Mean age 11.4yr; 65 females. 77 had a standard oral glucose tolerance test (and 18 had fasting insulin and glucose samples. The prevalence of AGH (fasting insulin ≥20μU/L, fasting glucose ≥6.1mmol or glucose >7.8mmol/L at 120min), HT (systolic BP >95%) and DL (Total cholesterol >95% or HDL cholesterol <0.9mmol/L) were assessed. Logistic regression was performed based on risk of ≥2 IRS components.

Results: 36% had AGH; 37% had HT; 25% had DL. 36% had 1 IRS component, 21% had two and 4% had all 3 components. Higher risk of ≥2 IRS components was associated with older age (OR=1.3, p=0.003) and insulin insensitivity (calculated using the homeostatic model (HOMA) (OR=1.2, p=0.015). Ethnicity, family history of IRS, birth-weight and sex were not associated with IRS risk.

Conclusions: Nearly one-quarter of obese children and adolescents of all ethnicities have ≥ 2 components of the IRS. The longitudinal stability and progression of the IRS is unknown. Greater clinical focus on cardiovascular risk is needed in obese children & adolescents.

**PREVALENCE AND SPECTRUM OF CARDIAC ABNORMALITIES IN PAEDIATRIC PATIENTS WITH INTESTINAL MALROTATION**

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Paediatric surgical problems are often encountered in patients with congenital heart defects like tetralogy of Fallot. Likewise isomerism of atrial appendages is well known to be associated with malrotation of gut with its attendant potential complication for intestinal volvulus. We retrospectively analysed the case notes of 37 consecutive patients with an age range 1 day to 9.5 years (mean 1.44 years, median 0.08 years) who had undergone ladd’s procedure for intestinal malrotation in our institution during the 4 years period from march 1996 to may 2000. Twenty (54.15%) were operated upon in neonatal period. In ten (27%) patients the initial presentation was with volvulus involving the small intestine. Twenty three (62.2%) had the primary diagnosis of malrotation of the gut. Other surgical defects were – meckle’s diverticulum(3), right descending colon(1), pyloric stenosis(1) and dysplastic kidney(1). Only ten (27%) of the 37 patients had a paediatric cardiology review by the time of surgery. In only one (2.7%) of these patients was the review initiated by the paediatric surgeons. The primary cardiac diagnoses amongst the ten patients reviewed cardiologically were right atrial isomerism(3), left atrial isomerism(3), supracardiac total anomalous pulmonary venous connection(1), scimitar syndrome(1) and hypertrophic obstructive cardiomyopathy(1). This study underscores the importance of cardiac assessment of any paediatric patients presenting with intestinal malrotation. There should also be a low threshold for entertaining the possibility of malrotation of gut in congenital heart disease patients with gastrointestinal symptoms, particularly those with suspected or confirmed atrial isomerism.

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VALUES PLACED ON THE QUALITY OF LIFE OF CHILDREN WITH CONGENITAL HEART DISEASE: COMPARING PARENTS WITH HEALTH PROFESSIONALS

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Aim: The childhood outcomes of congenital heart disease (CHD) vary widely and include spontaneous resolution, surgical correction and a spectrum of cardiac and neurological disability. It is important for clinical decision-making, to understand the perspectives of both parents and health professionals (HP) towards the quality of life of children with CHD. Neonatologists, parents and adolescents value health state descriptions of low birth weight survivors differently, but this has not been explored for CHD.

Method: We developed 8 health state descriptions for cardiac and neurological disability resulting from CHD and presented these with a self-administered anonymous questionnaire. The questionnaire asked respondents to 1) rank the health state descriptions from best to worst, 2) score each health state using a visual analogue scale (VAS) and 3) mark death on the scale.

Results: 109 HPs (72% female, median age 38 years) and 106 parents (82% female, median age 37 years) completed the questionnaire. HPs and parents agreed in the order of ranking health states from best to worst (Spearman rank correlation r=1). The lowest VAS scores were assigned to health state descriptions with severe neurological disability. Scores did not differ significantly by age, sex or whether respondent was in the HP or parent group. 59.6% thought that health states worse than death were possible. VAS scores assigned to health states worse than death were significantly lower than death.

Conclusion: Parents of children with CHD and health professionals who care for them place similar values on the quality of life outcomes of children with CHD. Both groups are more averse to neurological than cardiac disability. Further exploration of the values assigned by children and adults with CHD, for clinical decision-making, and assigned by the general public, for resource use allocation, is required.


IS FOUR LIMB BLOOD PRESSURE COMPARISON CLINICALLY USEFUL IN EXCLUDING COARCTATION OF THE AORTA?


Aims: To assess whether comparing arm and leg blood pressures in neonates can be used to confirm normality of the aortic arch.

Methods: Infants were recruited from the post-natal wards. Each infant underwent echocardiography and later had blood pressures measured once in each limb using a Dinamap Compact T 482210. This methodology was guided by a telephone survey of 40 UK neonatal units with 4 or more intensive care cots.

Results: Thirty eight of 40 units used an aciometric method when measuring blood pressure in all limbs and 33 of these took one measurement in each limb. Thirty nine neonates were examined. One had a small ventricular septal defect. The remainder had structurally normal hearts and aortic arches. Three had a blood pressure in the arms that was 20 mmHg higher than the legs, suggestive of coarctation of the aorta. This gives a specificity of comparison of the arm and leg blood pressures of 36/39 (92.3% (95% CI 79.98%)) or a false positive rate of 3/39 (7.7% (1.6-20.9)). The standard deviation in measurements between arms was 15.7 mmHg, 14.5 mmHg between legs and 11 mmHg when comparing the nearest arm and leg values.

Conclusions: Prevalence at live birth of CoA is 1 per 4000. With a false positive rate of 7.7% for upper and lower limb blood pressure comparison only one baby in 300 with a positive result would have a CoA (assuming no false negatives). This is not sufficiently specific for the test to be clinically useful. If coarctation of the aorta is suspected it can only be excluded by skilled echocardiography.


STUDY TO REVIEW THE CHANGING PATTERN IN THE DIAGNOSIS AND PRESENTATION OF STRUCTURAL HEART DISEASE

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Introduction: Antenatal diagnosis of structural heart disease has been possible for over a decade. Nationally there is great variation in the rate of antenatal diagnosis. Echocardiography is increasingly available outside specialist cardiac centres. The aim of this study was to review the impact of these changes on the timing of diagnosis and the mode of clinical presentation of infants with cardiac anomalies.

Methods: Patients were identified from the specialist cardiac clinic database and a retrospective case-note review performed. The timing of diagnosis was either Antenatal(AN), Postnatal pre-hospital discharge(pre) or Postnatal post-hospital discharge(post). The mode of presentation was recorded. Patients were separated into groups according to date of birth: Group A 1985-1989; Group B 1990-1994; Group C 1995-1999 and Group D 2000-2002.

Results: 164 patient records have been reviewed. Over the study period there has been a steady rise in the number of cases diagnosed antenatally with a steady fall in cases diagnosed after discharge form hospital (see table). In the 90 infants with severe heart disease (intervention needed in first year of life) there has been a steady fall in the proportion presenting with either collapse, cyanosis or heart failure, group A 75%, B 62%, C 34% and D 16%.

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<th>Group</th>
<th>AN</th>
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<tr>
<td>A n=25</td>
<td>0</td>
<td>10 (40%)</td>
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<td>B n=44</td>
<td>1 (2%)</td>
<td>16 (36%)</td>
<td>27 (62%)</td>
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<td>C n=57</td>
<td>14 (24%)</td>
<td>21 (37%)</td>
<td>22 (39%)</td>
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<tr>
<td>D n=38</td>
<td>17 (45%)</td>
<td>11 (26%)</td>
<td>10 (29%)</td>
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Conclusions: Fetal and early postnatal echocardiography has resulted in the earlier diagnosis of structural heart problems. Fewer babies are presenting with symptomatic heart disease.