Cardiology

G121 THE PROVISION OF CARDIOLOGY SERVICES IN A NON-CARDIAC PAEDIATRIC INTENSIVE CARE UNIT SETTING

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Background: Approximately 1% of patients with cardiac problems are admitted to non-cardiac paediatric intensive care units (PICU) in the UK every year.[1] The Paediatric Intensive Care Society standards document specifies that PICU should have access to specialty services such as paediatric cardiology within 2 h and have a 24-h on-site cover.[2]
Aims: To review local workload and conduct a survey of other similar, non-cardiac units across the UK to assess compliance with recommended standards.
Methods: We retrospectively reviewed the indications and outcomes of children referred for cardiology opinions over a 4-year period in our non-cardiac PICU. Patients were identified from the unit database. We then conducted an e-mail survey of the lead consultants in other UK non-cardiac PICU to determine the provision of cardiology services to critically ill children.
Results: Over the study period there were 1835 admissions to our PICU, 62 (3.4%) were referred to the single paediatrician with expertise in cardiology who provides cardiology services locally. Of these referrals, 26 (42%) patients had abnormal echo findings. 13 (21%) were transferred to a cardiac unit and underwent surgery. All nine (15%) cases with arrythmias were managed locally. 15 primarily non-cardiac PICU were identified from the PICAnet database. All 15 units had either a paediatrician with cardiology interest, consultant intensivist, echo technician or radiologist available to perform echocardiography. Their immediate availability was dependent on being on call. Only one unit had round-the-clock cover. A further seven (47%) units could manage paediatric cardiology input within 24 h. Telemedicine links were only used in one unit.
Conclusions: The local admission rate is in line with national figures. Very few PICU meet the recommended standards and services are dependent on chance. Whereas telemedicine has been utilised in other areas, it is poorly utilised in PICU.

1. **Paediatric Intensive Care Network**. *National report*. January 2005–December 2007.
2. **Paediatric Intensive Care Society**. *Standards document*. 2001.

G122 COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT, SEX AND SURVIVAL

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Introduction: Complete atrioventricular septal defect (CAVSD) is one of the most common serious congenital heart defects (CHD), affecting approximately 35 children per 100 000 live births. In approximately 50% of cases, it is associated with Down syndrome. Although at birth girls are more likely to be affected than boys (in contrast to most CHD types in which boys predominate), it is unclear whether there are sex differences in longer-term childhood survival.
Aim: To investigate, for children with CAVSD, sex differences in survival to 15 years as well as early life factors predicting survival.
Methods: Data were obtained from the hospital records of 447 children (208 boys (47%)) with CAVSD within the UK Collaborative Study of Congenital Heart Defects, a UK-wide prospective cohort of 3903 children (2148 boys (57%)) with serious CHD born between 1992 and 1995. Of those with CAVSD, 39 were born preterm (<37 weeks' gestation), 17 died before cardiac surgery, 220 had non-cardiac congenital anomalies (of whom 197 had Down syndrome) and 79 had additional intracardiac structural defects. We estimated sex-specific survival up to 15 years of age and investigated factors that might predict survival using life tables, Kaplan–Meier survival curves and Cox proportional hazards models (Stata SE version 10).
Results: At birth, 47% (95% CI 42% to 51%) of those with CAVSD were boys. Overall, 65% (290/447; 95% CI 60% to 69%) of children with CAVSD survived to 15 years of age. Girls were more likely than boys to survive to 1 year (girls 77% (95% CI 71% to 82%); boys 63% (95% CI 56% to 70%)) and to 15 years of age (girls 67% (95% CI 62% to 75%); boys 49% (95% CI 41% to 56%)). In a Cox proportional hazards model, male sex was associated with a higher risk of death (hazard ratio 1.51, 95% CI 1.06 to 2.14) after adjustment for preterm birth, associated intracardiac or non-cardiac anomalies and age at first intervention.
Conclusion: Sex is an independent predictor of survival from CAVSD, with boys faring worse than girls. This finding is not explained by preterm birth, associated anomalies or timing of cardiac intervention. The female survival advantage was evident from birth, not confined to the postoperative period and did not attenuate during puberty, implicating early life factors.
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G123 ACCURACY OF ECHOCARDIOGRAPHY DIAGNOSIS BY PAEDIATRICIANS WITH EXPERTISE IN CARDIOLOGY

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Background: Children with suspected cardiac problems are often seen by paediatricians with expertise in cardiology in local hospitals. The most complex of these patients may then be referred to a joint clinic with a visiting paediatric cardiologist. Practice varies widely across the UK.
Aim: To determine the accuracy of the paediatrician with expertise in cardiology diagnosis in referrals to the joint clinic.
Methods: A retrospective case note review was conducted for all children attending the joint clinic for the first time between March 2004 and September 2008. Children with antenatally diagnosed cardiac problems were excluded. The echocardiography diagnoses of the paediatrician with expertise in cardiology and the visiting paediatric cardiologist were compared.
Results: Over the 55-month study period, 1545 patients were seen in paediatrician with expertise in cardiology clinics. 161 (10%) were referred to the joint clinic. The median age of the patient at first appointment was 1.5 years (range 2 weeks to 16.75 years). The sex distribution was 93 (58%) female and 68 (42%) male. In 136 (84%) cases, there was complete diagnostic agreement between the paediatrician with expertise in cardiology and the visiting paediatric cardiologist. In 17 (11%) patients the main diagnosis was unchanged but the visiting paediatric cardiologist noted an additional finding. In four (2%) patients a completely different echocardiography diagnosis was made by the visiting paediatric cardiologist. The abnormality had resolved in four (2%) patients at clinic attendance and these patients were immediately discharged. 66 (41%) of the patients referred to the specialist clinic have required either surgical or catheter intervention. Of the patients, 106 (66%) are currently being followed in the clinic, 34 (21%) have been discharged, seven (4%) have been referred to adult services and 11 (7%) have moved away. Three (2%) children have died.
Conclusion: In this institution, paediatricians with expertise in cardiology make accurate diagnoses of abnormality when referring patients to a specialist clinic. High quality local services allow the

vast majority of patients with normal scans to be discharged and most patients with minor abnormalities to be managed in the local paediatrician with expertise in cardiology-run clinics. Appropriate referral of children with complex cardiac abnormalities allows the best possible use of specialist paediatric cardiology expertise.

G124 ANTENATAL MANAGEMENT AND OUTCOME OF FETAL TACHYARRHYTHMIA: A 10-YEAR EXPERIENCE IN A TERTIARY FETAL MEDICINE UNIT

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Aims: Our aim was to review the antenatal management and outcomes of fetal tachyarrhythmias.
Methods: A retrospective review of the management and outcomes of all fetal tachyarrhythmias at a tertiary fetal medicine centre between 1 January 1997 and 31 December 2006.
Results: During this period, 382 mothers were referred for fetal echocardiography for fetal dysrhythmia. Median gestation for referral was 30 weeks (range 19–41). The rhythm disturbance in the majority was ectopic beats that required no treatement. 29 fetal tachyarrhythmias were identified. These were not associated with structural heart disease. Of the tachyarrhythmias, there were 24 cases of supraventricular tachycardia (SVT), three cases of atrial flutter and two sinus tachycardia. In the SVT group, five fetuses were hydropic. Four of the hydropic babies were resistant to medical transplacental therapy and needed direct fetal therapy with amiodarone or adenosine. One died in the neonatal period as a result of prematurity and hydrops. 21 cases of SVT were successfully managed antenatally with medical transplacental therapy. From 2001 onwards, maternal flecanide therapy was rapidly introduced when there was poor response to digoxin (within 24 h). All three cases of atrial flutter required DC cardioversion postnatally. The two cases of sinus tachycardia were managed conservatively and were normal on postnatal examination.
Conclusion: In our experience, the overall prognosis for antenatally treated SVT was good. Medical transplacental therapies with digoxin or flecanide were safe and successful in reverting the SVT to sinus rhythm. Hydropic babies were likely to require direct fetal therapy.

G125 MANAGEMENT OF PULMONARY HYPERTENSION IN DOWN'S SYNDROME

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Background: Children with Down's syndrome (DS) are known to be more at risk of pulmonary arterial hypertension (PAH) than the general population. This is multifactorial but is partly due to upper airway obstruction and to congenital heart disease (CHD).
Aims: To report our experience of children with DS and PAH and to document the cardiorespiratory management.
Design: Retrospective review in a tertiary paediatric cardiac centre.
Patients: Included were those with DS and PAH (tricuspid regurgitation (TR) jet velocity >2.7 m/s) who were referred for assessment from March 2005 to October 2008. Those with a large left to right shunt were excluded as were those with established Eisenmenger syndrome.
Results: 17 children were included in the study (nine boys). The median age at assessment was 567 days (range 106–5734) and median TR jet was 3.4 m/s (2.7–5.2). At cardiac catheterisation under baseline conditions, median systolic pulmonary artery (PA) pressure was 39 mm Hg (20–95) and median pulmonary vascular resistance (PVR) was 4.12 U.m^2 (1.9–13.35) at baseline. All children received inhaled nitric oxide (10 and 20 parts per million) and 100% oxygen to determine pulmonary vascular response. All children had respiratory assessment; six required bronchoscopy, which showed bronchomalacia in three; one was followed for tracheo-oesophageal fistula and two were normal. Eight underwent sleep studies, and as a result three have had adenotonsillectomy performed. Two children underwent lung biopsy showing plexigenic arteriopathy. Those who had low PVR and CHD underwent corrective surgery. If there was a low PA pressure under anaesthesia, then upper airway obstruction was diagnosed. If there was elevated PVR, oxygen therapy was prescribed along with sildenafil, bosentan or both. In all, seven children received sildenafil therapy before successful cardiac surgery. Repeat cardiac catheterisation was undertaken in three, showing improvement of the PVR in all.
Conclusions: In order to manage this complex group of patients successfully, a combined cardiological, respiratory and surgical approach is required.

G126 REFERRAL PATTERNS AND FETAL ECHOCARDIOGRAPHY FINDINGS IN HIGH-RISK MOTHERS: A 10-YEAR EXPERIENCE

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Aims: To evaluate the referral patterns and fetal echocardiography findings in high-risk mothers over a 10-year period in a tertiary fetal medicine centre.
Methods: A retrospective review of all pregnant women who underwent fetal echocardiography between 1 January 1997 and 31 December 2006.
Results: 5182 mothers were referred for fetal echocardiography and a total of 7252 fetal echocardiography examinations was carried out during this period. The main reasons for referral were: previously affected child, 26%; abnormal initial screening scan, 22%; maternal cardiac condition, 10%; infant of diabetic mother, 8% and increased fetal nuchal translucency, 3%. 985 (19%) echocardiograms were reported as abnormal. The majority of the abnormalities were identified in mothers who had an abnormal initial screening scan (62%). In addition, the echocardiogram was also abnormal in 9.5% of cases, with increased fetal nuchal translucency and in 4.9% of infants of diabetic mothers. In those with a previously affected child and maternal cardiac condition, the echocardiogram was abnormal in 2.5% and 1.8%, respectively.
Conclusion: Abnormal initial screening scans and increased nuchal translucency had the highest yield in identifying congenital heart disease (CHD) in high-risk mothers. Infants of diabetic mothers also have an increased risk warranting fetal cardiac screening for CHD. Normal fetal echocardiogram provides reassurance for the remainder of parents, especially those with a previous child with CHD or in those mothers with a cardiac condition.

G127 DIAGNOSIS MANAGEMENT AND OUTCOME OF ADOLESCENT VASOVAGAL SYNCOPE

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Introduction: Spontaneous syncope occurs in approximately half of individuals during their lives. However, vasovagal syncope (VVS) has a different spectrum in childhood. We aimed to analyse the number of patients diagnosed with VVS in our department in the past 10 years with respect to its management and outcome.
Methods: This is a 10-year retrospective study and the data were obtained from the cardiac database between 1997 and 2007. Any

individual presenting with syncope without a structural heart abnormality was included in the study. Patients with a structural heart defect were excluded from analysis. The analysis included patient demographics, the mode of presentation, type of investigations, medications, duration of follow-up, status on discharge with regard to morbidity and mortality.
Results: 105 patients were referred with adolescent VVS in the past 10 years. All of them had echocardiogram and a 12-lead ECG as the baseline investigations, but 43% of patients had no other investigations. Commonly used other investigations were the head-up table tilt test, 24-h Holter and treadmill exercise test. Of those who had the tilt test (19), 58% were positive but only 8% of the study group required drug treatment and two with asystole required permanent pacing. All patients were given advice on salt and fluid optimisation and the avoidance of triggers; 74% of patients were discharged at the time of study with this advice alone and the mean discharge period was 9 months. No patient has died as a result of syncope.
Conclusion: The commonest cause of adolescent syncope is vasovagal in origin. The diagnosis can almost always be established clinically with a thorough history and further investigations are only required in atypical cases or if there is an abnormality on the 12-lead ECG. All of the patients would require reassurance, positive feedback and education to avoid triggers. Medications are rarely needed and most patients would benefit from positive feedback, simple salt and fluid optimisation and the avoidance of triggers. Morbidity is negligible and no mortality directly related to syncope was encountered.

G128 DRUG PROVOCATION TESTS IN UNMASKING INHERITED CARDIAC CHANNELOPATHIES IN CHILDHOOD

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Introduction: Drug provocation tests have recently been introduced to unmask suspected Brugada and long QT syndrome. The commonly used tests are the ajmaline-flecainide challenge test and the epinephrine challenge test. We analysed the number of drug challenges carried out in our department in the past 6 years and reviewed their indications, outcome and complications.
Methods: Data were collected retrospectively from the period 2002–7. Information was obtained from the cardiac database and/or ward admission book and from individual patient's notes. Indications for test, resting ECG, test results and complications were analysed.
Results: 26 patients underwent drug challenge tests in the past 6 years, of whom 14 had epinephrine challenge, seven were given ajmaline challenge and five were studied with flecainide challenge. Three exhibited a positive response to epinephrine and two had a positive response to ajmaline. Indications for testing were a family history of sudden death/cardiac arrest or a family history of Brugada syndrome or long QTS and an unexplained history of dizziness/collapse with suspected abnormal ECG. 15 of the 26 patients had a suspected abnormal resting ECG. All five patients who had a positive test had a suspected abnormal ECG. There were no complications nor was there any provocation of arrhythmia during the testing.
Conclusion: Drug challenge tests are safe and good tools in investigating patients with suspected Brugada or long QTS. In borderline cases diagnosis can be facilitated. Even if we did not encounter any complications in the 26 patients tested in the past 6 years, these tests should be performed under continuous medical surveillance with advanced paediatric life support facilities due to the potential induction of ventricular tachycardia.

G129 AN AUDIT OF THE MANAGEMENT OF HEART MURMURS ON POSTNATAL WARDS

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Background: Heart murmurs detected incidentally during newborn examination are often innocent but may signify an underlying cardiac malformation. Investigation and management of these murmurs varies widely and is often dependent on local resources. In order to standardise the management of heart murmurs in our hospital a guideline (based on clinical examination with selective cardiology review) was introduced. This aimed to ensure safe management while not overwhelming the cardiologists with unnecessary referrals.
Aims: To establish adherence to and safety of the guideline. To quantify workload implications. To establish the causes of murmurs in our population.
Methods: Patients were prospectively identified over a 2-year period (August 2006 to July 2008). Case notes were reviewed and examination findings, investigations, follow-up and diagnosis recorded.
Results: 91 babies were identified. The guideline was generally well adhered to. In total, 50 babies (55%) were referred for cardiology assessment: 31 before discharge from hospital and 19 following review in the neonatal outpatient clinic. In 43 babies this assessment included an echocardiogram. 31 babies (34%) had an underlying cardiac malformation, of whom 26 were identified before discharge home. No baby discharged from follow-up without cardiology review subsequently presented with a cardiac problem.
Conclusion: A significant minority of babies with a heart murmur have an underlying cardiac malformation. Our guideline appears to ensure the timely identification of these babies and rationalises our use of specialist services.

G130 EFFECT OF EXERCISE TEST ON RATE-CORRECTED QT INTERVAL IN CHILDREN INVESTIGATED FOR SUSPECTED LONG QT SYNDROME

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Introduction: The exercise test is known to prolong the rate-corrected QT interval (QTc) in children with long QT syndrome (LQTS), compared with their baseline ECG and compared with controls. Some recent studies suggest that the QT interval does shorten with exercise in LQTS, but remains shortened in recovery thus forming a QT hysteresis loop.
Aim: To study the effect of peak exercise and early recovery on QTc of children investigated for suspected LQTS and to compare this with the effect of the hyperventilation test and the 1993 Schwartz diagnostic score.
Methods: Twenty-five children (10 males and 15 females, aged 8.3–17.6 years) investigated with the exercise test on modified Bruce protocol for suspected LQTS were included. Twelve-lead ECG was recorded at rest, after one minute of hyperventilation, at peak exercise and into one minute of recovery, and the QTc calculated using Bazett's formula was compared against each other.
Results: The mean QTc at rest was 425 ms (SD 31). Compared with baseline QTc, both hyperventilation and peak exercise caused a significant increment—mean (95% CI) increment: 35 ms (19 to 51) and 21 ms (4 to 37), respectively. There was no significant change from baseline at recovery. The mean increment in QTc at peak exercise was 14, 26 and 45 ms, respectively, in low (n = 14), intermediate (n = 9) and high (n = 2) probability groups as per Schwartz diagnostic criteria, whereas the early recovery produced a mean change of −1, −14 and 10 ms,

respectively. There was no statistical difference between the low and intermediate probability groups. There was considerable overlap between the low probability group and the intermediate/high probability group in terms of their peak exercise QTc and the clinical outcomes were also variable.

Conclusion: A change in QTc at peak exercise or early recovery did not particularly help in confirming or rejecting the diagnosis in our cohort of children investigated for suspected LQTS.

G131 CONGENITAL HEART DEFECTS ASSOCIATED WITH TURNER SYNDROME: A POPULATION-BASED 23-YEAR STUDY

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Background: Cardiovascular malformations are known to occur in live-born Turner syndrome, typically coarctation of the aorta in infants and children and aortic valve problems in adults.

Aim: To examine the total prevalence and spectrum of cardiovascular malformations in Turner syndrome in an English health region during a 23-year period and to evaluate the influence of antenatal diagnosis.

Methods: A retrospective analysis of data maintained in the regional congenital anomalies registry from 1985 to 2007, which included all cases diagnosed antenatally and postnatally. Early fetal losses were excluded.

Results: Among 790 000 registered births we identified 197 with Turner syndrome (25 per 10^5), leading to 109 terminations, eight stillbirths and 80 live births. The karyotype was 45X in 128 (65%), 45X/46XX in 57 (29%), 45X/47XXX in five (3%) and unknown in eight (4%). Cardiovascular malformations were present in 70/197 (36%) overall and in 53/117 (45%) of those not live born and 17/80 (21%) live births (see table). Of 123 cases of Turner syndrome diagnosed antenatally, only 13 were born alive. Antenatal diagnosis increased progressively during the study period, from 13/31 (42%) in the first 5 years of the study to 30/39 (77%) in the last 5-year period, with a corresponding increase in the proportion terminated electively. 74 cases were diagnosed after birth, and included 67 live births, four stillbirths and two terminations.

Abs G131 Table Distribution of congenital heart defects in Turner syndrome

Cardiac diagnoses	ToP and SB	Live births	Total
Coarctation of aorta	35 (66%)	7 (41%)	42 (60%)
Hypoplastic left heart	12 (23%)	1 (6%)	13 (19%)
Interrupted aortic arch	2 (4%)	2 (12%)	4 (6%)
Other left-sided lesions	0	5 (30%)	5 (7%)
Others	4 (8%)	2 (12%)	6 (9%)
Total	53	17	70

SB, stillbirth; ToP, termination of pregnancy.

Conclusion: This retrospective analysis of all cases of Turner syndrome in a circumscribed English population has confirmed a high prevalence of cardiovascular malformations. In addition to aortic coarctation and bicuspid aortic valve, we identified a significant proportion with hypoplastic left heart syndrome and interrupted aortic arch. Antenatal diagnosis of Turner syndrome increased during the course of the study period, with a corresponding increase in elective terminations and a decrease in more serious cardiovascular malformations among those born alive.