

*British Paediatric Cardiac Association: Abstracts*

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## Abstracts for British Paediatric Cardiac Association Annual Meeting: St John's College, Cambridge, 15–16 December 2003

### Results in the mid-term of an axillary artery approach for balloon valvoplasty of severe aortic valvar stenosis in early infancy

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Severe aortic valvar stenosis in early infancy has generally been treated by surgical valvotomy or balloon valvoplasty. Due to previous femoral arterial complications, we changed our practice in infants <5 kg by using an axillary arterial cut down. Balloon valvoplasty using this approach was performed in 22 consecutive infants aged from 1 to 77 days, with a mean age of 17 days, all having severe aortic valvar stenosis. Their weight was between 2 and 4.46 kg, with a mean of 3.28 kg. Left-ventricular function was normal in 13, mildly reduced in 3, and moderately reduced in 6 infants, with 3 requiring inotropic support. Aortic valvar diameter, as measured echocardiographically, varied from 5.2 to 8.5 mm with a mean of 6.43 mm, and the mean velocity across the aortic valve was 4.10 m/s, with a range from 1.6 to 5.5 m/s. Balloon valvoplasty was performed via a right axillary artery cut down using a 4 French sheath, though occasionally a bare Tyshak balloon catheter was inserted directly. The diameter of the balloon was equal to or within 1 mm of the aortic valvar diameter. The arteriotomy was repaired with interrupted 6/0 Prolene sutures. Minimal or no angiography was undertaken, with a mean screening time of 9.8 min. *Results:* Balloon valvoplasty was achieved in all the infants. The gradient prior to intervention ranged from 50 to 125 mmHg, with a mean of 77 mmHg and was reduced to between 10 and 60 mmHg, with a mean of 30 mmHg, following balloon valvoplasty. Echocardiography after the procedure showed 9 patients with no aortic regurgitation, 2 with trivial, 10 with mild, and 1 with moderate to severe aortic regurgitation. The volume of the pulse in the right arm was normal in the 13 of the 22 infants immediately following intervention. Only 2 of the remaining 9 infants required an intravenous infusion of Heparin to maintain patency of the axillary artery. One patient experienced axillary arterial transection requiring microsurgical repair. There was 1 death. This infant had significant left-ventricular hypoplasia and, in retrospect, may have been better managed by a Norwood operation. All patients have been followed up for a mean of 37.3 months, with a range from 7 months to 9 years. Repeat valvoplasty was necessary in 3 patients. Subsequent surgical intervention was needed in 3, consisting of 2 surgical valvotomies, one combined with mitral valvar replacement, and 1 Ross procedure at the age of 3 months. The brachial pulse in the right arm was decreased in 2 infants, and absent in 2. *Conclusion:* Aortic valvoplasty via an axillary arterial cut down is a safe and effective method, comparing favourably to access via the femoral artery. This approach aids rapid crossing of the aortic valve in these haemodynamically unstable patients. Axillary arterial compromise may occur, but appears to be less important than femoral arterial compromise, and needs further evaluation.

### Is screening for congenital cardiac disease useful?

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Over the last 20 years, there have been major advances in the detection of congenital cardiac disease, with antenatal and postnatal echocardiography augmenting the postnatal examination. Congenital cardiac disease can now be diagnosed during foetal life or prior to discharge from hospital. Earlier diagnosis may have a benefit on

a measurable outcome. The aim of this study was to ascertain, first, whether congenital cardiac disease is being diagnosed earlier, second, the current rate of detection for congenital cardiac disease and, third the impact on clinical presentation. *Methods:* Patients were identified from the database of a specialist cardiac clinic in a single centre where their mothers had received obstetric care and been delivered. A retrospective case-note review was performed. Diagnosis of congenital cardiac disease was made either antenatally, prior to, or after discharge from hospital. The mode of clinical presentation was recorded. Patients were grouped according to year of birth, which corresponded to the level of screening available at that time. From 1985 to 1989, this was by clinical postnatal examination. From 1990 to 1994, a foetal 4-chamber view was obtained at the anomaly scan and combined with clinical postnatal examination. Then, from 1995 until 2002, a 5-chamber view was obtained during foetal scanning, along with an echocardiographic study prior to discharge if indicated clinically from the postnatal examination. *Results:* 238 children fitted the criteria for the study. With the introduction of each new technique, a greater proportion of children were diagnosed earlier, particularly in those with severe disease in whom intervention was required in the first year of life.

Time period	Antenatal (%)	Predischarge (%)	Postdischarge (%)
1985–1989	N/A	50	50
1990–1994	10	55	35
1995–2002	56	26	18

Comparing the latest period with those before it, significantly fewer children were diagnosed after initial discharge from hospital ( $p = 0.001$ ), significantly few children presented symptomatically ( $p = 0.02$ ), with  $p < 0.001$  in the severe subgroup, and fewer required preoperative inotropic support. *Conclusions:* New screening techniques have resulted in earlier diagnosis of congenital cardiac disease. Over the period 2000 through 2002, 87% of children with severe congenital cardiac disease were diagnosed either antenatally or prior to discharge. When mode of presentation is used as a measure of outcome, screening for congenital cardiac disease is useful.

### Cardiopulmonary exercise testing in children referred for cardiac transplantation

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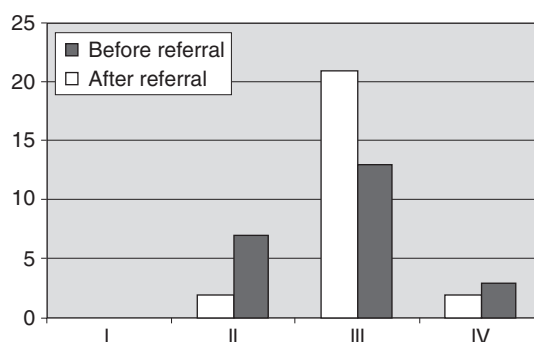
*Introduction:* Measurement of peak consumption of oxygen is widely used to determine need for cardiac transplantation in adults with severe heart failure, with a level below 14.0 ml/kg/min indicating that a patient should be listed. No data exists, however, on its use in children. *Hypothesis:* We sought to determine whether measurement of peak consumption of oxygen is reproducible, and useful for rationalising decisions over the listing for transplantation of children with end-stage cardiac failure. *Methods:* We attempted to measure peak consumption of oxygen in all children referred for assessment for transplantation at Great Ormond Street Hospital between July, 2002 and October, 2003. Peak consumption of oxygen was derived from respiratory gas analysis with a medical mass spectrometer. Minimal height for successful use of the equipment was 120 cm. We performed 24 tests in 19 children at a mean age of 12.5, with a standard error of 0.6 years, weight of 40.4 kg, with standard error of 3.3, and height of 147.4 cm, with standard error of 3.0. Of the children, 10 had dilated cardiomyopathy, 1 had restrictive cardiomyopathy, 3 had anthracycline cardiotoxicity, 4 had undergone palliative surgery for complex congenital cardiac disease, and 1 had arrhythmogenic right-ventricular dysplasia. *Results:* Tests were successful in 16 children, of whom 3 were tested twice, and one was tested three times. Testing was unsuccessful in 3 children, 2 due to lack of co-operation, and 1 due to equipment failure. Of the 21 successful tests, mean peak consumption of oxygen was 21.0 ml/kg/min, with standard error of 1.5. Results correlated well with class in the grading system of the New York Heart Association, with a correlation coefficient of 0.86, though not with echocardiographic assessment of left-ventricular function by fractional shortening measurement. Peak consumption of oxygen was  $<14.0$  ml/kg/min in 4 children, and all have been successfully transplanted. One child with a peak consumption of 22.4 ml/kg/min deteriorated acutely 2 months after testing, and was then too unwell to be retested before a heart became available. The other children, with peak consumption of oxygen  $>14.0$  ml/kg/min, were listed for a medical follow-up, and will be retested as indicated by their clinical condition. All 19 have survived to date. *Conclusion:* Peak consumption of oxygen can be measured in the majority of children with end-stage heart failure above a height of 120 cm and, as in adults, can be used to provide objective evidence to help in decisions over listing for transplantation.

## Experience in children with severe pulmonary arterial hypertension using Bosentan (Tracleer)

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Pulmonary arterial hypertension is a rare but debilitating disease, resulting in progressive deterioration that is characterised by an increase in pulmonary vascular resistance. The most effective therapy to date has been continuous intravenous infusion of epoprostenol, a relatively complicated therapy with risk of sepsis. Other options are limited in small children with severe disease. Bosentan, or Tracleer, an endothelial receptor antagonist, has shown promising results in adults, but little is known about its efficacy in children. *Aims:* To examine the safety and efficacy of Bosentan in children with pulmonary arterial hypertension. *Characteristics of patients treated:* We treated 25 children aged between 10 months and 16 years with Bosentan for periods from 2 to 16 months, with a mean of 7 months. In 13 patients, the pulmonary hypertension was primary and secondary in the other 12. Mean weight at initiation of treatment was 21.8 kg, and the patients were treated with a dose of 15–125 mg, with a mean of 45 mg. All patients had evidence of right atrial and right-ventricular hypertrophy, with three-fifths having evidence of strain. Echocardiograms showed tricuspid regurgitation and right-ventricular hypertrophy in all, and impaired right-ventricular function in 44%. Pulmonary arterial pressure was equal to or greater than systemic in 76%. Pulmonary vascular resistance was elevated in all patients, with measurements of more than 11 Wood units in 84%. Pulmonary vascular resistance did not fall in response to vasodilators, including nitric oxide in 92%. 5 patients were already on Epoprostenol at an average dose of 45 ng/kg/min. *Results:* Most children showed clinical improvement as indicated by improved exercise tolerance, and a beneficial shift in the classification of the New York Heart Association (see Figure below). Rescue Epoprostenol therapy was needed in 3 children after 8 months of treatment with Bosentan. No side effects have been reported so far. *Conclusion:* Chronic administration of an oral rather than an intravenous drug is an attractive option, particularly in children. Although our limited experience with Bosentan in treating severe primary and secondary pulmonary hypertension in young children is encouraging, one needs to monitor these patients carefully. Our current indication for Bosentan is, first, for patients in the second or third classes of the New York Heart Association at presentation, second, high pulmonary vascular resistance which is not reactive to acute vasodilators, and third, patients in whom treatment with Epoprostenol is not an option.



## Clinical role and technical aspects of cardiac magnetic resonance imaging in infancy

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*Background:* Magnetic resonance imaging has been demonstrated to be an important non-invasive diagnostic modality in patients with congenital cardiac disease. Few studies, however, have focused on young children. This study examined the clinical utility and technical challenges of cardiovascular magnetic resonance imaging in infants. *Methods:* All patients aged under 1 year undergoing cardiac magnetic resonance imaging at Boston Children's Hospital, between January 1999 and July 2002, were identified, and their magnetic resonance imaging records and clinical data were retrospectively reviewed. *Results:* Cardiac magnetic resonance imaging examinations, 99 in all, were performed in 91 patients with a median weight of 4.9 kg, and a range from 1.2 to 16.3 kg,

at a median age of 102 days, with a range from 1 to 358 days. All studies were performed under general anesthesia without any complications, using a head coil in 57, a cardiac scanner in 19, a multi-purpose array in 19, or a surface radio frequency receiver coil of 5 in. in 4. The infants, seen as outpatients, were discharged the same day. The primary questions for referral were delineation of the thoracic vasculature in 54, assessment of possible compression of the airways in 25, evaluation of cardiac tumour in 6, and other reasons in 14. For those referred for assessment of thoracic vasculature, findings at surgery in 37, and catheterisation in 16, were all concordant with the mode for resonance imaging diagnoses. In the children with suspected compression of the airways, findings at surgery in 16, and catheterisation in 2 were also in agreement with findings at resonance imaging. The typing of tumour by resonance imaging in the 3 patients with preoperative studies were all confirmed by pathologic results. Systematic review demonstrated that the anatomy of the airways was best shown with fast spin echo sequences, and the thoracic vasculature by Gadolinium-enhanced 3-dimensional magnetic resonance angiography. **Conclusion:** Cardiac magnetic resonance imaging plays a limited but important diagnostic role in infants with congenital cardiac disease. It is a useful and safe imaging modality in delineating the thoracic vasculature, evaluating possible compression of the airways, and characterising cardiac tumours. In selected cases, cardiac resonance imaging may obviate the need for cardiac catheterisation or bronchoscopy.

### Disruption of the myocardial force–frequency relationship following cardiopulmonary bypass: first human data and implications for postoperative care

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**Background:** The force–frequency relationship is an innate property of the myocardium to alter contractile force due to the changes in calcium cycling related to heart rate. The well-described changes in calcium cycling following cardiopulmonary bypass may, therefore, have important implications for postoperative ventricular performance. **Methods:** We studied neonates undergoing the arterial switch procedure. Baseline force–frequency relationship was studied using a new tissue Doppler index of contractility, isovolumic acceleration, during transoesophageal atrial pacing prior to sternotomy. Tissue Doppler imaging was performed using a System V ultrasound scanner (GE Vingmed). The left-ventricular free wall was interrogated in an apical 4-chamber view. Postoperative assessments were made during atrial pacing using epicardial pacing wires. **Results:** We studied 11 neonates undergoing the arterial switch procedure for transposition. Preoperatively maximal isovolumic acceleration was  $3.6 \text{ m/s}^2$ , and this occurred at an optimal heart rate of 190 beats/min. Postoperative force–frequency relationships showed a much flatter response with significantly depressed values of isovolumic acceleration ( $p < 0.001$  ANOVA). There was a slight improvement in contractile force at 24 h as compared with 6 h postoperatively, though this was not significantly different. **Conclusion:** For the first time, these data show non-invasively the disruptive effects of cardiopulmonary bypass on the force–frequency relationship. There is depression of both the maximal contractile force generated and also the slope of the force–frequency relationship. Heart rates greater than the optimal heart rate would be clearly detrimental to ventricular performance. These novel data may allow postoperative manipulation of contractile performance by modulation of heart rate.

