Introduction

On behalf of the Organising Committee it is a pleasure to welcome you all to the Barbican Centre for the first annual meeting since renaming the association. The original British Paediatric Cardiology Group was formally constituted as the British Paediatric Cardiology Association (BPCA) in 1990 and has become the centre stage for British Paediatric Cardiology and Congenital Heart Disease. At the annual meeting in Dublin last year the association became known as the British Congenital Cardiac Association (BCCA). This year's meeting reflects the seamless approach taken in caring for patients with congenital heart disease from the fetus through to adult life. Of those registering for the meeting 25 have requested to join the association and will be introduced at the member's business meeting.

There has been a last minute change to the lecture venue. It had been planned to use the lecture theatre on the same level as the exhibition hall but a large number of late registrations led to the need for a larger lecture hall and reluctantly we have been forced to use Cinema 1 on level –2. This can be accessed by the stairs or lifts but because of the distance, registrants are requested to move promptly to and from the sessions and meals.

An innovation this year is the holding of a joint session with the Paediatric Cardiac Nursing Association (PCNA) that was suggested by feedback from the meeting in Dublin. In this era of multidisciplinary care, it seems appropriate but so that there is not a complete break from tradition, the PCNA has also organized a separate afternoon session covering Outreach Nursing and Transition to Adult Care. Attendees are requested to evaluate this change in the feedback form to help with planning of future meetings.

It was encouraging to receive 27 abstracts covering a wide range of interests for the young research workers session. The abstract grading committee selected 6 abstracts for oral presentations and 8 abstracts for poster presentations. The grading committee will choose the best oral and poster presentation during the meeting and will announce the results at the annual dinner. Posters will be displayed in the Exhibition Hall on level 3. All selected abstracts will be published in Cardiology in the Young. CPD certificates will be available at the end of the meeting in return for your evaluation form.

The member's business meeting will be followed by the BCCA Annual Dinner at Madame Tussauds. Places for the evening entertainment at Madame Tussauds are limited but there is a waiting list at the reception desk for any returned tickets. For the security of the event, admission will be by production of your ticket. For those who wish to proceed directly from the business meeting to the dinner, there will be coaches but please notify the registration desk by lunchtime if you wish to use the coach.

The success of this meeting is dependent on sponsorship and once again BVM Medical has been our principal sponsor. Together with substantial sponsorship from Actelion and Philips, these 3 companies have covered over half the costs of the meeting. Additional sponsorship has been obtained from Cook, General Electric, Gore, NuMed, St Jude Medical and Baxter. Our sponsors have put on an exhibition that should add to the value of the meeting and you are encouraged to visit the stands.

The Department of Congenital Heart Disease at Guy's Hospital moved to the new Evelina Children's Hospital on the St Thomas' site towards the end of October. While there are areas that have not been fully completed, we are very happy for visitors to see the new hospital and also view the combined X Ray - MRI unit (XMR). During the visit there will be an informal meeting for those interested in MRI of congenital heart disease. Those wishing to be included in this visit should indicate their intent at the registration desk. Visiting will be possible after the meeting ends on Friday afternoon.

Yours sincerely

Eric Rosenthal

Organising Committee
Eric Rosenthal, Reza Razavi, Shakeel Qureshi, Gurleen Sharland, John Simpson, Edward Baker.

Abstract Grading Committee

We would like to thank the following Companies for their Sponsorship of the Meeting

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Thursday 24 November 2005
Cinema 1, Level -2, Barbican Centre

08:30	*Registration & Coffee*	
09:15	Opening remarks	Gurleen Sharland

Session I - Hypoplastic Left Heart Syndrome

Joint BCCA & PCNA

Chairs - Gurleen Sharland & Kerry Cook

09:30	**Anatomy**	Andrew Cook
09:50	**Genetics**	David Wilson
10:10	**Fetal Diagnosis & Counselling**	Sherrida Rollings & John Simpson
10:30	**The parent's view**	Sarah Green
10:50	*Morning Coffee Break & Exhibits*	

Session II - Hypoplastic Left Heart Syndrome

Joint BCCA & PCNA

Chairs - Conal Austin & Debbie Parker

11:30	**The Guy's approach**	David Anderson
11:45	**The Birmingham approach**	Bill Brawn
12:00	**Immediate & long-term results (UKCCAD)**	John Gibbs
12:15	**Discussion**	
12:30	**Debate "TOP is unethical for HLHS"**	*proposer:* Marco Pozzi
		opposer: Lindsey Allan
13:00	*Lunch & Exhibits*	

13:00 - 13.15 *Council Meeting*

Session III Abstracts for Young Investigator Award

Chairs - Owen Miller & John Cheetham

14:30	**Abstracts**	
15:30	**BPCA Survey on Heart Failure**	Rachel Andrews
15:45	*Afternoon Tea Break & Exhibits*	
16:15	**Interventional Cardiology:**	**Chair -** John Gibbs
	What we have learned	Michael Tynan
	What the future holds	Shakeel Qureshi
16:45	**BCCA Business Meeting 16.45 - 1800**	

Evening Entertainment & Dinner

Friday 25 November 2005
Cinema 1, Level -2, Barbican Centre

08:30 *Registration & Coffee*

Session I Interventional Cardiology

Chairs - Thomas Kraseman & Tony Salmon

09:30 **Percutaneous Fontan** — John Cheatham

09:50 **PDA Occlusion in Premature infants** — Neil Wilson

10:10 **PFO closure indications** — Eric Rosenthal

10:30 **Intra-operative intervention** — John Cheatham

11:00 *Morning Coffee Break & Exhibits*

Session II Cyanotic Congenital Heart Disease in the Adult

Chairs - Cathy Head & Gruschen Veldtman

11:30 **Venesection, Anticoagulation, Pregnancy** — Sara Thorne

11:50 **Endothelin antagonists in Cyanotic CHD** — Mike Gatzoulis

12:10 **Heart Lung transplantation** — John O'Sullivan

12:30 **Discussion**

12:45 *Lunch & Exhibits*

Session III MRI

Chairs - Titus Kuhn & Tal Geva

14:00 **How to do cardiac MR in children – practicalities** — Ted Baker

14:15 **Functional assessment of valves** — Albert de Roos

14:30 **Anatomical assessment of intra cardiac structures** — Phillip Beerbaum

14:45 **Replacement of diagnostic cardiac catheterisation** — Tal Geva

15:00 *Afternoon Tea Break & Exhibits*

15:30 **MR guided cardiac catheterisation** — Reza Razavi

15:45 **Debate "Cardiac CT has a major role in children with congenital heart disease?"** — *proposer:* Andrew Taylor; opposer: Tal Geva

16:15 Closing Remarks — Reza Razavi

Optional visit to Evelina Children's Hospital and Interventional MRI theatre

17.00 Reception desk

Thursday 24 November 2005
Nurses Parallel Session
Redgrave Room, Level 4, Barbican Centre

14.30 **Chairs report** - Kerry Cook, Chairman of PCNA

14.50 **Supporting young peoople with congenital heart disease during transition to adult services**

Melinda Edwards & Tracy Sutherland

Consultant Paediatric Psychiatrist, Senior Play Specialist

Both Evelina Children's Hospital Guy's & St Thomas NHS Foundation Trust.

15.20 **Young persons' perspective**

Molly Lloyd - Patient

Tea break

16.05 **Transition to adult services**

Dr Cathy Head

Consultant Cardiologist Guy's & St Thomas NHS Foundation Trust

Young Research Workers Abstracts - Oral

Thursday 24th November 14:30 – 15:30

Cinema 1, Level -2, Barbican Centre

Twenty year trends in recognition of life-threatening neonatal cardiac malformations

Zdenka Reinhardt, Christopher Wren

Department of Paediatric Cardiology, Freeman Hospital, Newcastle upon Tyne.

Background

Babies with cardiovascular malformations are usually asymptomatic at birth. Earlier diagnosis is likely to lead to a better outcome. This study examines trends in the timing of diagnosis of potentially life-threatening cardiovascular malformations.

Methods

Ascertainment of all cardiovascular malformations diagnosed in infancy in the resident population of one English health region in 1985-2004. Babies considered to have a life-threatening cardiovascular malformation included all those with hypoplastic left heart, pulmonary atresia with intact ventricular septum, transposition of the great arteries, or interruption of the aorta; and babies dying or undergoing operation in the first 28 days with coarctation of the aorta, aortic stenosis, pulmonary stenosis, tetralogy of Fallot, pulmonary atresia with ventricular septal defect, or total anomalous pulmonary venous connection.

Results

Of 688167 live born babies in the 20 years of the study, 4444 had cardiovascular malformations diagnosed in infancy (6.5 per 1000). Cardiovascular malformations were potentially life-threatening in 685 (15%). Over the time of the study, 8% were recognised prenatally, 58% postnatally before discharge from hospital, 31% in life after discharge and 3% after death. Malformations most likely to remain undiagnosed at discharge were coarctation of the aorta (63%), interruption of the aortic arch (50%), and total anomalous pulmonary venous connection (47%). Over the 20 years of the study, the proportion of babies diagnosed antenatally increased from around 1% to 20% and no case was first diagnosed after death in the last 10 years. However, the recent proportion going home without a diagnosis remains around 25%.

Conclusions

Overall one third of babies with potentially life-threatening cardiovascular malformations leave hospital undiagnosed. In recent years better antenatal diagnosis has reduced this proportion to one quarter. Better early recognition of such babies is unlikely to be achieved by clinical examination and is more likely to come from further improvements in antenatal diagnosis and more widespread adoption of routine pulse oximetry.

Early and midterm outcomes of Implantable Cardioverter Defibrillator (ICD) implantation in patients with repaired Tetralogy of Fallot and pulmonary stenosis – a retrospective review.

S Viswanathan, K English, MEC Blackburn

Department of Paediatric Department of Paediatric Cardiology, Yorkshire Heart Centre.

Introduction

Repair of Tetralogy of Fallot up until recent decades involved aggressive resection and annular enlargement through a right ventriculotomy. This resulted in ventricular scarring and pulmonary incompetence, with an increased risk of ventricular tachyarrhythmia and sudden death in young adulthood. Following the NICE guidelines, implantation of ICDs as primary prevention in patients with repaired Tetralogy is ever increasing. This study aims to determine the rate of appropriate and inappropriate discharges, the success rate of ICD therapy and the impact of ICD implantation on the use of anti-arrhythmic medication in this population of patients.

Materials and Methods

This is a retrospective review of patients with repaired Tetralogy of Fallot (n=18) and pulmonary stenosis (n=2) with implantable cardioverter defibrillators managed at our tertiary centre. Patients who satisfied the above criteria were identified, their notes and charts were examined and details regarding indication for ICD implantation, device specifications and complications following implantation were collected. Data was also collected on the incidence of appropriate and inappropriate therapies and the success rate of ICD therapy along with the impact of implantation on anti-arrhythmic medication usage in these patients.

Results

Of the 20 patients, 18 had previous repair of Tetralogy of Fallot and 2 had pulmonary valvotomy and infundibular resection for pulmonary stenosis between 1969 and 1989. 70% (n=14) of these patients required reoperation with 10 patients having pulmonary valve replacements (PVR). At the time of consideration for ICD implantation 80% had moderate to severe pumonary incompetence and 60% had more than mild right ventricular dilatation on echocardiography. Indications for ICD implantation were symptomatic ventricular tachycardia requiring cardioversion (n=8), ventricular tachycardia on 24 hr tape/Reveal or electrophysiological study (n=8), ventricular fibrillation (VF)/ pulseless ventricular tachycardia (VT) (n=2), collapse (n=1) and sinus node dysfunction (n=1).

The median age at implantation was 22 years (16.4-43 years). All our patients had dual chamber devices implanted with either dual (n=13) or single coil (n=6) ventricular leads. GEM3 AT (n=5), Marquis DR (n=8) and Maximo DR (n=7) generators (Medtronic Inc.) were implanted in sub pectoral position and both anti-tachycardia pacing and cardioversion modes were programmed as part of individualised VT and VF protocols. Early post procedural complications included atrial lead displacement (n=1) and pneumothorax requiring drainage (n=1).

During a median follow up of 1.6 years (0.03-4.5 years) several episodes of inappropriate sensing were noted in 9 patients (45%) especially early after implantation resulting in inappropriate therapies in 6 patients (30%). This was found to be mainly due to double counting of T waves or inaccurate interpretation of varying PR intervals as AV dyssynchrony which were effectively dealt with by minor changes in device programming. There were 33 episodes of inappropriate anti-tachycardia pacing (ATP) in 4 patients and 19 episodes of inappropriate cardioversion in 5 patients. Appropriate ATP was instituted in 4 patients with successful termination of 19 out of 21 episodes (90% success rate). 2 patients had appropriate cardioversion although in 1 of these

continued overleaf

Early and midterm outcomes of Implantable Cardioverter Defibrillator (ICD) implantation in patients with repaired Tetralogy of Fallot and pulmonary stenosis – a retrospective review.

continued

patients with troublesome ventricular arrhythmia cardioversion was unsuccessful resulting in death at 10 months after AICD implantation having had no detections or therapies during this period.

Prior to ICD implantation 18 patients were on anti-arrhythmic medications with 8 (44%) on amiodarone. At the time of last follow up after AICD implantation all patients were established on anti-arrhythmic agents and of these 6 patients (30%) were still on amiodarone with most of the others being effectively controlled on beta-blockers and/or flecainide.

Late complications of ICD implantation included lead failure in 1 patient requiring replacement 3.3 yrs after implantation and generator replacement in a patient who was pacemaker dependent a year after implantation due to an advisory issued by the manufacturer regarding the risk of sudden battery depletion.

Conclusions

In our study we found a rate of 0.6 appropriate and 1.4 inappropriate therapies per patient-year of follow up which is in keeping with published literature. The mortality in our study group was 5% which is acceptable given the high risk population. Implantation of an ICD allowed switching over from amiodarone to less toxic anti arrhythmic therapy in a proportion of patients. Anti-tachycardia pacing was generally quite successful in terminating tachyarrhythmia with only 2 (10%) out of 21 episodes requiring cardioversion.

Treatment with bosentan in children with an intracardiac communication and severe pulmonary arterial hypertension

P Bonou, AA Hislop, Y Flynn, SG Haworth

Great Ormond Street Hospital and The Institute of Child Health, London UK

Aim

To present the experience of the UK Pulmonary Hypertensive Service for Children at Great Ormond Street Hospital in treating children with "Eisenmenger" type physiology with the dual endothelin receptor antagonist bosentan. Patients and methods: 14 inoperable patients were included in the study. These were selected based on a history of an unrestrictive intracardiac communication and established right to left shunt at rest. There was a female:male ratio 1:0.75 and age range at initiation of treatment of 7-18.6 years (mean 12.5 yrs). 6 patients had Trisomy 21. The cardiac diagnoses were: 6 large VSDs, 6 CAVSD (1 of which was not Trisomy 21) and 2 more complex congenital heart disease cases that underwent palliative surgery (DILV, DOLV, TGA, PA banding and DORV, TGA, VSD, PS, Waterston). 12 patients received antiplatelet/anticoagulation therapy. WHO functional class, haemoglobin (Hb), oxygen saturation, 6 min walk tests, echocardiograms and ECGs were assessed before starting treatment with bosentan and after 6 and 12 months of follow up. 12 patients had a cardiac catheterisation before treatment commenced. The parameters were compared for patients before treatment and at 6 and 12 months post treatment with paired t-test and ANOVA.

Results

All patients were severely symptomatic the mean WHO functional class being 3.2 (range 3-4). The mean Hb was 18.3g/dl (range 13.4-22.9). The mean PVR of all patients was 30.98 units.m2 (7.6-57). The Trisomy 21 patients had a mean PVR of 34.4 units.m2 and non Trisomy 21 patients had a mean of 28.6 units.m2. Mean oxygen saturation at rest was 78.9% (range 67-93), mean 6 min walk test distance was 223.8 m (n=10, range 108-360) and the lowest oxygen saturation during the walk test was 59.7% (42-79) at 6 mins or below. Mean duration of therapy was 15.2 months (range 6-34 months). Bosentan was well tolerated in all patients but discontinued in one due to non compliance with monthly screening blood tests. All patients remained stable and their echocardiogram and ECG findings were unchanged throughout. After 6 months mean WHO functional class had decreased significantly to 2.96 ($p=0.029$), whilst Hb and oxygen saturation at rest had not changed. 10 of the 14 patients were on treatment for more than a year and after 12 months the WHO functional class was maintained at the same improved level. The Hb and oxygen saturation at rest remained unchanged. After 12 months the 6 min walk mean distance had increased from 211 to 253 metres (non significant).

Conclusion

Patients treated with bosentan for up to 1 year showed clinical improvement and remained stable. After 6 months treatment WHO functional class had significantly improved and improvement was sustained for up to 1 year.

Perioperative Red Cell Salvage in Paediatric Cardiac Surgery

S Salam, D Abrams, A Kelleher, J La Rovere

Deptartments of Paediatric Intensive Care & Anaesthesia, Royal Brompton & Harefield NHS Trust, London UK.

Objective

In recent years blood transfusion has become a debated health care issue. To minimise exposure to infectious agents and reduce bank blood transfusion requirements, leucocyte filtration and perioperative red cell salvage (RCS) are increasingly used in paediatric patients. We hypothesised RCS would reduce the need for additional blood products in children following cardiopulmonary bypass (CPB).

Methods

Patients undergoing routine or emergency cardiac surgery requiring CPB over a study period of 3 months were included prospectively in the analysis. Haemoglobin, platelet count, coagulation screen and heparin levels were performed before, immediately after surgery and 24 hours later. RCS was performed in theatre according to surgical and anaesthetic preference. Red cells were salvaged from the surgical site, anticoagulated, washed and following resuspension in saline reinfused into the patient within 4 hours. The incidence of post-operative bleeding (>10ml/kg/hr) was recorded, as was the need for additional red blood cells, platelets and fresh frozen plasma (FFP). The need for blood products was at the discretion of the consultant intensivist. Statistical analysis was performed using student t-test and Chi squared methods. Significance was accepted as $p < 0.05$.

Results

35 consecutive patients (34.54 ± 43.55 months, 13.48 ± 14.39kg) were included in the analysis. A total of 17 infants <12 months were included, 9/24 who received RCS and 8/11 who did not (p 0.052). Cyanotic heart disease was seen in 40%. RCS was performed in 24 of 35 patients, who were significantly older (44.2 ± 44.1 vs. 13.6 ±25.5, p 0.02) and heavier (16.6 ± 16.2 vs. 6.7 ± 4.7,p 0.01). No difference was seen in the prevalence of cyanosis between the two groups. Post-operative bleeding was seen in 21% who underwent RCS and 40% in those who did not (p 0.33). The need for additional red blood cells was significantly reduced in those who received RCS, 37.5% vs. 91%, p 0.003, as was the use of FFP, 8.3% vs. 45.5%, p 0.02. There was no difference in the need for platelet transfusion, p 0.2.

Discussion

In this study RCS was performed on 68.5% of children following CPB. RCS significantly reduced the need for further blood and FFP transfusion, although this was not related to post-operative bleeding. This has important implications for both exposure to infectious agents and health economics. That children who underwent RCS were older and heavier may be related to the complexity of surgery and CPB in younger patients, although infants were represented in both groups. A further analysis of potential health and economic benefits in a homogenous group is needed.

B-Type Natiuretic Peptide (BNP) as a marker of paediatric heart failure.

J. Mangat, C. Carter, Y. Foo, M. Burch

Cardiothoracic Transplant and Biochemistry Deptartment Great Ormond Street Hospital, London, WC1N 3JH

Background

Paediatric heart failure can be difficult to assess. Symptoms vary widely in children who have poor ventricular function on echocardiogram. In addition exercise testing is not possible in young children. BNP has been used in adult heart failure and more recently in paediatrics. We investigated BNP in the setting of a paediatric heart failure and transplantation service to assess its usefulness as a clinical marker of heart failure.

Method

Clinical and echocardiographic data were correlated to 126 BNP samples. Patients were 3 weeks to 16 years of age. Left ventricular end-diastolic dimension (LVEDd) was related to normal values for body surface area (z-score). Clinical status was defined using Ross and NYHA scores. 34 samples were from patients with normal ventricular function. Of the remainder, most were from patients with idiopathic dilated cardiomyopathy (38), anthracycline cardiotoxicity (15), congenital heart disease (25), viral myocarditis (6) and restrictive cardiomyopathy (6). Analysis: BNP was correlated to parametric data (fractional shortening (FS%)) with pearsons correlation coefficient. For non-parametric data (Z-score, NYHA and Ross score), spearmans correlation coefficient was used.

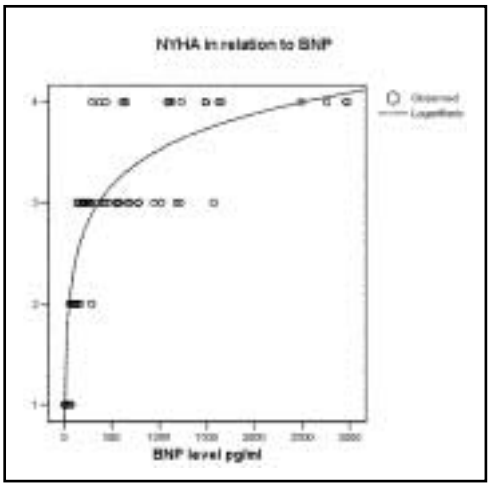
Results

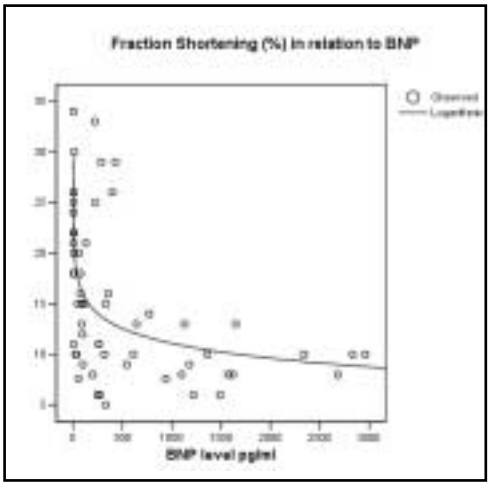
Rising BNP levels correlate to deteriorating clinical status with significance to the 0.01 level (Ross and NYHA), Figure 1. There was also correlation, significant to the 0.01 level to FS%, Figure 2. Increasing BNP levels correlated to the increasing LVEDd z-score significant to the 0.05 level. Mean BNP: in ventricular dysfunction 634pg/ml (S.E.M. =80), in normal function 11.9pg/ml (S.E.M. =1.6).

Conclusions

This is the largest study of BNP in paediatric heart failure. The stronger correlation of BNP to NYHA and ROSS than to LVEDD and FS% suggests a useful role in assessment of children with heart failure. We believe it is useful in the outpatient setting particularly when care is shared with general paediatricians. It also appears to be a useful addition in the assessment of heart failure and perhaps timing of transplantation in a specialist centre.

Figures





Abstract for Young Investigator Award
Omega-3 fatty acids - Pharmacological preconditioning for paediatric cardiac surgical patients

J. McGuinness, J. Byrne, A. Hanly, H. Chen, C. Condron, D. Bouchier-Hayes J.M. Redmond.

Department of Surgery, The Royal College of Surgeons in Ireland.

Background
Inappropriate systemic inflammatory endothelial and leucocyte activation is a major pathogenic component of post-operative low cardiac output syndrome (LCOS) and associated pulmonary oedema, low urine output, and generalised oedema, in paediatric cardiac surgery. We hypothesised that acute pre-treatment with a parenteral nutrition component omega-3 fatty acid infusion in-vitro, would prevent the pattern of leucocyte and endothelial activation seen in paediatric cardiac surgery through induction of preconditioning.

Methods
Endothelial cells were isolated from saphenous vein of 75 adult cardiac surgery patients, grown in monolayers, pre-treated, and then stimulated with either endotoxin (LPS), TNFalpha, or complement C5a (prime mediators of systemic inflammation with paediatric cardiac surgery). Endothelial production of the neutrophil recruiting cytokines IL-6, IL-8, and endothelial expression of the neutrophil adherence receptors E-Selectin, ICAM-1 were assessed. The effects on the inflammatory pro-coagulant response of the endothelium ie. loss of surface thrombomodulin and production of tissue factor were also examined. Similar methods were employed using isolated peripheral blood neutrophils. The effects on endothelial nuclear translocation of the acute inflammatory transcription factor NFkB and the heat shock protein (HSP72) response were investigated for mechanistic insight.

Results
4 hours pre-treatment with the omega-3 infusion prevented endothelial release of neutrophil recruiting cytokines IL-6 and IL-8, also prevented upregulation of endothelial receptors required for neutrophil adherence E-Selectin and ICAM-1, and maintained surface anticoagulant thrombomodulin levels, in response to stimulation with LPS (Table 1) or TNFalpha

	Baseline	LPS Stimulated	Pre-treated LPS Stimulated	P-value
IL-6	50+/-5	2727+/-23	211+/-8	<0.01
IL-8	26+/-16	3678+/-10	2650+/-36	<0.01
E-Selectin	17+/-0.2	109+/-2	13+/-0.4	<0.01
ICAM-1	86+/-12	196+/-1	72+/-0.4	<0.01
Thrombomodulin	73.6+/-0.7	66.1+/-0.3	81.2+/-4.9	<0.01

Units for IL-6, IL-8 are pmol/L
Units for E-Selectin, ICAM-1, and Thrombomodulin are mean channel fluorescence on flow cytometry

The effect of the infusion on adhesion molecule expression was reversible, lasting 18-24 hours. Inflammatory prolongation of neutrophil survival was partially reversed through an increase in apoptotic rate (73%+/-1% of control neutrophils apoptotic at 20hrs vs 77%+/-1% of pre-treated neutrophils, P=0.03). Pre-treatment produced a more than 2-fold increase in endothelial HSP72 levels (P<0.05), and reduced nuclear levels of the p65/p50 subunits of NFkB in response to LPS stimulation (2.1+/-0.2x105 relative luminescence units in controls vs 1.3+/-0.2x105 in pre-treated endothelium, P<0.01).

Conclusions
Acute pre-treatment with an omega-3 fatty acid infusion inhibits multiple components of systemic inflammatory activation seen in cardiac surgery. Induction of HSP72 and subsequent reduction of nuclear levels of NFkB, suggest preconditioning mechanisms of inflammatory transcription factor inhibition. This pharmacological strategy may ameliorate paediatric cardiac surgery associated low cardiac output syndrome and it's sequelae.

Young Research Workers Abstracts – Poster
Thursday 24th and Friday 25th November
Exhibition Hall, Level 3, Barbican Centre

ECMO support for lifethreatening arrhythmia in infancy permits successful radiofrequency treatment

A Rasheed, M D Khan, A K Duke, M Tofeig, A Ng, P Stafford, F A Bu'Lock, R K Firmin, G J Peek

The majority of arrhythmias presenting in infancy cardiovert readily or rapidly respond to conventional medical therapy. A small number prove highly refractory to anti-arrhythmic medications. Myocardial performance may be severely compromised by the combination of fast heart rate and negatively inotropic drugs. Some babies die.

We have recently supported 2 babies with refractory arrhythmias on ECMO, both to pursue drug therapy and eventually, to support the circulation during radiofrequency ablation, with very successful results. The first patient was a 2.5kg neonate presenting with collapse secondary to atrial ectopic tachycardia with a rate of 300 / minute. Myocardial function was severely impaired. The arrhythmia was adenosine resistant and after iv amiodarone loading had no effect, esmolol infusion was started. This produced profound hypotension and the arrhythmia rapidly recommenced after DC cardioversion. In the face of such severe haemodynamic disturbance, VA ECMO was instituted. Further anti-arrhythmics were tried on-circulatory support, but the arrhythmia was incessant despite multiple DC cardioversions. Therefore radiofrequency ablation of the atrial ectopic focus was attempted on ECMO support. This was achieved uneventfully and the myocardial function rapidly improved, with decannulation 24 hours later. Unfortunately the arrhythmia recurred 2 weeks later, but was successfully treated by further ablation without ECMO. The child remains well with normal development on no medication. An 11month old baby presented to the GP with acute onset of lethargy and poor feeding and a heart rate of 350bpm was noted! This was a broad complex tachycardia with independent p wave activity (confirmed with adenosine); ie ventricular tachycardia. Although initially well tolerated, the tachycardia resisted DC 'cardioversion' even up to 60J. Progressive and severe myocardial dysfunction and hypotension ensued, exacerbated by any attempts at drug therapy. The patient was therefore placed on VA ECMO. Some slowing of the ventricular rhythm was achieved with amiodarone and flecainide. The patient was then decannulated but the rapid arrhythmia recurred and ECMO was reinstituted. Electro-physiological mapping was then undertaken on ECMO support. NAVEX mapping identified a right ventricular outflow tract focus. This was resistant to conventional RF energy but was eventually successfully ablated with a 'Cool-tip' catheter. Myocardial function improved rapidly, the patient was decannulated after 48 hours observation and there has been no recurrence of the arrhythmia since discharge.

Although viewed as a very invasive technique, VA ECMO support here has prevented two otherwise unavoidable deaths in babies with conditions readily treated by radio-frequency techniques in older children. Not only did ECMO permit institution of aggressive drug therapy but also safely supported catheter interventions in very small patients. ECMO support should be considered early for small patients with refractory arrhythmias, before irreversible neurological compromise ensues. It could also be used electively to permit radiofrequency ablation in children whose size causes concern for safe catheter manipulation.

Epoprostenol treatment in children with pulmonary arterial hypertension

AE Lammers[1], AA Hislop[2], Y Flynn[1], SG Haworth[1]

Great Ormond Street Hospital for Children[1] and The Institute of Child Health[2], London UK

Background

Continuous intravenous Epoprostenol treatment has been shown to be beneficial in patients with severe pulmonary arterial hypertension (PAH). We report the experience of the UK Pulmonary Hypertension Service for Children treating 39 children with this therapy.

Patients and methods

The patients were aged 4 months to 17 years (median 5.4 years), male: female ratio was 1:1.2. Eleven children were treated with Epoprostenol alone. Twenty-eight were also treated with Sildenafil, Bosentan, Nifedipine or a combination of these. Twenty-five children had idiopathic PAH and 14 had PAH associated with congenital heart disease (9), connective tissue disease (2), chronic lung disease (2) or HIV (1). Cardiac catheter studies made before treatment began showed that in 26 patients the pulmonary artery pressure equalled or exceeded the systemic pressure. Thirty-four patients had a pulmonary vascular resistance (PVR) study with a mean PVR of 23±12 units.m2. After initial evaluation children were monitored by ECG, transthoracic echocardiography, weight, 6-minute-walk test and WHO functional class.

Results

Follow-up was 1-90 months (mean 27). 24 children remain stable and well, 8 have been transplanted and 7 died on treatment.

There was no deterioration in the ECG; the 6-minute walk distance and right ventricular function assessed by echocardiography remained stable. WHO functional class improved from 3.6 to 2.6 during the first year (n=38) ($p<0.001$) and remained stable for up to 3 years (n=17). Weight improved significantly ($p<0.05$). A Kaplan Meier survival curve showed that survival was 94.4% at 1 year, 90.3% at 2 years and 83.9% at 3 years.

Conclusion

Epoprostenol treatment improved survival, clinical status and WHO functional class in children with severe PAH. Survival in the children on intravenous Epoprostenol with and without combination therapy was better than that described in adult studies (McLaughlin VV. et al., Circulation 2002; 106 1477-82).

Heart Transplantation for Failure of the Systemic Right Ventricle Following Atrial Inversion Procedure (Senning/Mustard) for TGA

A.A. Lotto*, M. Chaudari**, J. O'Sullivan**, L. Hamilton*, A. Hasan*, J. Dark***

Regional Cardiothoracic Centre, **Dept. of Paediatric Cardiology, ***Cardiopulmonary Transplant Unit, Freeman Hospital, Newcastle upon Tyne, United Kingdom

Background

Right ventricular (RV) impairment is a well recognized late sequelae of atrial inversion operation. Although conservative interventions as well as anatomic correction have been advocated late after Mustard or Senning procedures, heart transplantation remains the main treatment of patients with end-stage failure of the systemic RV. Aim of this study is to evaluate operative and postoperative results in patients undergoing heart transplantation for end-stage failure of the systemic RV following Mustard/Senning operations.

Methods

Retrospective review of patients referred to the Regional Cardiothoracic Centre for cardiac transplantation was performed.

Results

Since January 1987, 62 out of 567 patients underwent heart transplantation for congenital heart disease. Nine of these had a previous atrial inversion operation performed (6 Mustard, 3 Senning) for TGA. Mean age at time of atrial inversion operation was 16.9±10.1 months. Time interval between atrial inversion and heart transplant was 22.0±11.6 years. Mean age at heart transplant was 23.6±10.8 years. Three patients were on inotropic support before the operation, while one was on ECMO support. Indication for heart transplantation was end-stage RV failure (mean NYHA class 3.6±0.5) with severe impaired function and severe tricuspid valve regurgitation in all patients. Results are shown in Table 1. At a mean follow up time of 21.5±10.09 months, 71.5% of hospital survivors are alive.

Conclusions

Heart transplantation for end-stage RV failure following atrial inversion operation is a high-risk procedure; however it remains the main treatment for this group of patients. The presence of a failing RV and previous surgery increases the operative mortality and exposes patients to late complications.

Table 1

Mean CPB time	260.4±95.3 min
Mean ischaemic time	214.1±25.4 min
Mean ventilation time	79.7±64.7 hrs
Mean ITU stay	6.5±4.8 days
Mean postop. stay	20.5±8.9 days
In-hospital death	2/9 (22.2%)
Late death	2/7 (28.5%)

Assessment of the need for elective balloon atrial septostomy in patients with Tricuspid atresia. A 25 year experience at the Yorkshire Heart Centre

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Introduction

Significant variation exists in the perceived indications for balloon atrial septostomy in children with tricuspid atresia (TA). Many units advocate early, elective balloon atrial septostomy due to potential progressive narrowing of the interatrial opening, especially after the insertion of an arterial shunt. Our practice is to perform an atrial septostomy only in children with clinical and/or echocardiographic features of a restrictive interatrial communication.

The aim of the study was to assess our practice by retrospective evaluation of patients born with tricuspid atresia with particular reference to the clinical fate of the atrial septum. More specifically the frequency of emergency atrial septostomy, the complication rate, the need for delayed septostomy or surgical resection and the longer term outcome were evaluated.

Materials and Methods

The study was a retrospective review (1980- 2005) of the data on 49 live-born patients in our departmental database with a diagnosis of TA. 1 patient was excluded from the study due to referral to another centre and subsequent loss to follow up. By reviewing the medical notes of the remaining 48 patients we collected data on indications for, and timing of balloon atrial septostomy and subsequent complications and long term outcome.

Results

The median time to follow up was 9.5 years (0.7-23.7). The diagnosis group was heterogeneous with 38 (79%) patients diagnosed with TA and normally related great arteries with or without pulmonary stenosis or atresia. The remaining 10 patients had more complex anatomy.

Of the 48 patients, 5 (10%) were deemed to require balloon atrial septostomy for a clinically and/or echocardiographically restrictive atrial septum. 3 procedures were performed within the first 24 hrs of life and 2 within the first month. One of the patients developed NEC following the septostomy and subsequently died of overwhelming sepsis.1 patient required delayed surgical septectomy at 6 years of age, during a bidirectional Glenn operation.

Of the remaining 42 patients there were 7 deaths (16%) but none were attributed to a restrictive atrial septum (1-at birth, severe acidosis and multi organ failure due to interrupted aortic arch and poor response to IV Prostin; 1-5 days with Group B streptococcal sepsis; 1- post op sepsis following insertion of a BT shunt; 1-9 years, acute shunt obstruction; 1-withdrawal of care due to complex anatomy deemed to have a poor prognosis; 2-sudden death at home at 3 _ years and 11 years).

26 patients required palliation with an arterial shunt (19-Modified BT shunt, 6-Waterston Shunt, 1-ductal stent). None of the patients developed a clinically and/or echocardiographically restrictive atrial septum post procedure or during long term follow-up.

Conclusion

We conclude that elective balloon atrial septostomy at presentation may not be necessary in patients born with tricuspid atresia. Accepting the limitations of retrospective data collection we also conclude that progressive narrowing of the interatrial septum at a later date is an unusual occurrence, even after an arterial shunt operation.

We recommend that atrial septostomy should only be performed in patients who show obvious clinical and/ or echocardiographic evidence of restrictive interatrial communication.

Feasibility and Safety of Intravascular Ultrasound for Measurement of Coronary Atheroma Burden in Children after Heart Transplantation.

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Purpose

Although intravascular ultrasound (IVUS) in children has been reported it is not widely used due to concerns over safety. Detailed analyses of measurements outlined by the ACC consensus statement on IVUS have not been reported in children. We report our safety and analysis data to date.

Procedures

IVUS of the left anterior descending was performed with mechanised pullback concurrently with surveillance coronary angiography. Procedure and fluoroscopy screening times were compared with a second group of patients who experienced coronary angiography alone. Analysis was performed to measure vessel and lumen area, vessel and lumen diameters and intimal thickness for serial slices at fixed intervals from a distal identification point. Mean atheroma burden, mean maximum intimal thickness (MIT) and overall maximal intimal thickness were determined for each study.

Results

27 procedures were analysed. Table 1 shows patient demographics and information relating to IVUS analysis. Details of procedure and fluoroscopy time for both IVUS and coronary angiography groups are presented in Table 2. No complications were encountered. Routine coronary angiography was normal in all but one patient.

Conclusions

We have demonstrated the safety of IVUS for coronary surveillance in children. When compared to coronary angiography alone procedure time and screening time are increased but we feel this is outweighed by the increased sensitivity of IVUS. 52% of children have an IMT > 0.5mm despite normal angiography. Such detailed analysis would appear central to future research into paediatric coronary disease.

	Median	**Range**
Age at study (years)	15.0	8.9 – 18.6
Time since transplant (years)	2.3	0.2 – 12.1
Donor age	24.6	6 - 45
	Mean	**SD**
Age at transplant	11.6	4.4
Total length of IVUS pullback (mm)	49	20
Number of slices analysed per study	21.5	6.4
Mean max IMT > 0.5 (patients)	8/27 (30%)	
Max IMT > 0.5 (patients)	14/27 (52%)	
Atheroma burden > 30% (patients)	4/27 (15%)	

	Coronary Angiography (n=31)	IVUS + coronary angiography (n=27)	p value
Procedure time (minutes) (mean ± SD)	31 ± 15	63 ± 32	0.08
Screening time (minutes) (mean ± SD)	7.6 ± 6.0	11.8 ± 6.0	< 0.01

Transcatheter Closure of Ventricle-Pulmonary Artery Communications after Cavopulmonary Shunt or Fontan Procedure

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Background:
Ventricle-Pulmonary artery connections are rare in patients after the Fontan procedure. However, these can cause significant long term problems. Hence the catheter occlusion of the anterograde flow in these patients would be beneficial. Further, in patients with a cavopulmonary shunt anterograde pulmonary blood flow is frequently maintained, but, in some patients, can cause significant volume loading of the heart or can complicate the subsequent Fontan procedure.

Objective:
To evaluate the use of interventional catheter closure of a ventricle-pulmonary artery communication in the setting of a cavopulmonary shunt or after the Fontan procedure.

Patients and Methods:
This was a retrospective study at a tertiary referral centre.
Eight patients (Age: 1.5 -18 years, mean 7.8 years) underwent transcatheter closure of a ventricle-pulmonary artery communication. Indications were cardiac failure or persistant pleural effusions after cavopulmonary shunt (n=2) or after Fontan (n=3) and abolishing the volume load of the single ventricle prior to Fontan completion (n=3).

Results:
Devices used were 17mm Rashkind Umbrella device (n=1), Amplatzer PDA device (n=7) and Amplatzer ASD device (n=1). One patient required two devices. There were no procedural complications.
All 3 patients with prolonged pleural effusions (1 post CP shunt and 2 post Fontan) showed complete resolution of effusions between 4-10 days after catheter closure.Two patients underwent transcatheter occlusion for progressive ventricular dilatation and cardiac failure. The first patient was post Fontan and showed gradual improvement in ventricular function. The second patient after CP shunt died 48 hours post intervention (non procedure related cardiac death). Three patients underwent catheter closure to off-load the systemic ventricle prior to the Fontan procedure. The device had to be removed prior to release in one patient, due to unsatisfactory position. All three patients underwent successful completion of Fontan 6 weeks to 3 months post catheter.

Conclusions:
Transcatheter closure of ventricle-pulmonary artery communication is a safe and effective technique in the treatment of selected patients after CP shunt or Fontan procedure with early or late complications due to inappropriate pulmonary blood flow. This intervention should also be considered prior to the Fontan procedure in selected patients with ventricular overload.

Severe Coronary Ostial Stenosis after Arterial Switch Operation Detected by Transthoracic Doppler Echocardiography

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Background:
Asymtomatic proximal coronary artery stenosis after arterial switch operation (ASO) is rare, but a potentially life-threatening condition, that is reported to appear in up to 7%. Angiography, although considered the state-of-the-art method of diagnosis, is an invasive method, but has limitations for diagnosing ostial stenosis. We report changes in Doppler flow profile and coronary flow reserve (CFR) in two asymptomatic patients (9 and 10 years old) with left main coronary artery (LMCA) ostial stenosis after ASO.

Methods:
Coronary flow was assessed by Transthoracic pulsed and colour-flow Doppler echocardiography (TTDE). CFR was measured in one patient using adenosine infusion (140 mcg/kg/min) over 4 minutes. CFR was calculated as the ratio of reactive hyperaemia to basal average peak velocity (APV). Both children were investigated with coronary angiography. They had myocardial Single-photon Emission Computed Tomography (SPECT), and magnetic resonance imaging (MRI) at rest and after reactive hyperaemia with adenosine infusion. Both patients had balloon dilatation and Cypher select (drug eluted) stent.

Results:
On echocardiogram a flame-like colour-flow diastolic signal was detected at the stenotic coronary ostia. The maximal spectral velocities during baseline conditions over the stenotic ostia were over 1.9 and 2.0 m/sec (normal 30+/-10 cm/s). The post-stenotic CFR was hemodynmically significant with value of 1.3, normal adult range 2.5 - 4. Coronary angiography showed a significant ostial stenosis 90% in both patients. Myocardial SPECT and MRI at rest/adenosine infusion were consistent with severe myocardial ischemia in the territory of the left coronary artery. Normal coronary angiography and coronary flow studies after stenting.

Conclusion:
We suggest that coronary artery flow assessment should be an integral part of the TTDE in the follow up of children with ASO. Serious coronary artery stenosis can be detected with TTDE. Assessment of CFR provides information of the physiological significance of the coronary stenosis.

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8 Years of Fetal Echocardiography in High-risk Mothers: The Birmingham Women's Hospital Experience.

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Introduction

Congenital heart disease (CHD) affects 8 per 1000 live births and it is also responsible for 20% of neonatal deaths. Antenatal diagnosis of major CHD allows appropriate counselling and planning for delivery at a neonatal unit with appropriate intensive care and transport facilities. Birmingham Women's Hospital provides a supra-regional specialist fetal echocardiography in high-risk mothers.

Aim

To evaluate fetal echocardiography findings in high-risk mothers over an 8 year period

Method

We undertook a retrospective review of all pregnant women at high-risk of having a baby with congenital heart disease who underwent fetal echocardiography between 01/01/1997 and 31/12/2004 at Birmingham Women's Hospital.

Results

3,963 mothers were referred for fetal echocardiography and a total of 5,568 fetal echocardiography examinations were carried out during this period. The main reasons for referral were: i) previously affected child – 27% ii) abnormal initial screening scan – 20.7% iii) maternal cardiac condition – 9.5% iv) infant of diabetic mothers – 8% and v) increased fetal nuchal translucency – 3%. 712 (17.9%) echocardiograms were reported as abnormal. The majority of the abnormalities were identified in mothers who had abnormal initial screening scan (62%). In addition, the echocardiogram was also abnormal in 9% of cases with increased fetal nuchal translucency and in 5.7% of infants of diabetic mothers. In those with previously affected child and maternal cardiac condition, the echocardiogram was abnormal in 2.5% and 2.6% respectively.

Conclusion

Abnormal initial screening scans and increased nuchal translucency had the highest yield in identifying CHD in high-risk mothers. Infant of diabetic mothers also have an increased risk warranting fetal cardiac screening for CHD. Normal fetal echocardiogram provides reassurance for the remainder of parents.

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