Highlights of the Annual Meeting of the Association for European Paediatric and Congenital Cardiology

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Summary of the 47th Annual Meeting of the Association for European Paediatric and Congenital Cardiology
London, UK, 22–25 May 2013

The 47th annual meeting of the Association for European Paediatric and Congenital Cardiology was held on 22–25th May 2013 in London, UK. This is one of the largest scientific meetings in Europe within the field of congenital cardiac disease and was held in association with the Japanese Society of Pediatric Cardiology and Cardiac Surgery, and Asia Pacific Pediatric Cardiology Society. There were 900 submitted abstracts and over 1000 delegates from 57 countries attended. We have summarized some of the highlights of the meeting below.

Preconference training day: update on imaging

This was an excellent day detailing the latest advances in cardiac imaging and how the different modalities can provide guidance for the management and surgical planning of congenital heart defects (CHD).

G Sharland (Evelina Children's Hospital, London, UK) provided a summary and detailed examples of prenatally diagnosed CHDs, persuading the audience that "fetal echocardiography makes everything clear". J Eichhorn (University Clinic, Heidelberg, Germany) demonstrated the increasing importance of MRI and computed tomography in the diagnosis and management of CHDs, as well as some of the current limitations (e.g., spatial resolution in high heart rates). In most cases, MRI and comuputed tomography angiography appear equivalent to conventional catheter angiocardiography and can provide additional information regarding tracheal, bronchial or esophageal abnormalities. Meanwhile, G Butera (Hospital San Donato, Milan, Italy) highlighted that diagnostic cardiac angiography still has a role to play, particularly in specific cases such as major aortopulmonary collateral arteries with dual supply. G Greil (Evelina Children's Hospital) emphasized the fact that hybrid imaging, both MRI and cardiac catheterization in the same session, provides 3D anatomical and physiological information and reduces radiation doses compared with catheterization alone. MRI cardiac catheterization is more accurate in the assessment of pulmonary vascular resistance compared with the assumptions made in the Fick principle and so is rapidly becoming the gold standard [1,2].

Intraoperative imaging

M Vogt (Heart Centre, Munich, Germany) convinced the audience that routine intraoperative imaging was essential after cardiac surgery. The use of intraoperative transesophageal echocardiography (TOE) resulted in a 14% reduction in reoperations in a study from the Mayo clinic. Intraoperative TOE was recommended in every patient undergoing surgery if they are above 3 kg, other than patent ductus arteriosus ligation and repair of aortic coarctation. Although the TOE in pediatric patients has a 1.6–4.9% complication rate (0.2–0.3% in adults), and perforation of the pharynx is the major worry, the complications can be minimized if the TOE probe is introduced during anesthesia induction by the anesthetist. V Tsang (Great Ormond Street Hospital, London, UK) presented data showing that routine intraoperative imaging of stage I Norwood led to a 30% revision rate in the theater and a dramatic reduction in postoperative 'errors'. Therefore, intraoperative imaging is an integral part of the surgical management of CHD and at Great Ormond Street Hospital, approximately 80% of cases have intraoperative imaging.

Future Medicine part of fsg

Cardiovascular imaging beyond routine

J Simpson (Evelina Children's Hospital) detailed advances in 3D echocardiography and how it is a major help in the assessment of the atrioventricular valves, septal defects and complex cardiac anomalies, by permitting views unachievable with 2D echocardiography and understanding structural relationships; for example in patients with complex double-outlet right ventricle [3]. A Taylor (Great Ormond Street Hospital) showed that computational modeling can help to predict which type of operation will provide the optimal result for individual patients, including influence on systemic and cerebral oxygen deliveries, and therefore permit the development of patient-specific therapeutic strategies. TY Hsia (Great Ormond Street Hospital) highlighted how such models can be utilized for preprocedural planning by performing virtual surgery to identify the optimal strategy for each individual prior to the actual operation. T Delhaas (Maastricht University, The Netherlands) introduced the CircAdapt model, which is an educational cardiovascular model simulating beat-to-beat hemodynamics and mechanics. It will be freely available soon for general use on their website [101].

Mannheimer lecture: impact of changing patterns & technologies for CHD long-term outcomes: J Hess (Heart Centre)

The mortality from CHD has decreased (from 30% in the 1960s to <3% today) and the survival into adulthood has increased (20% in the 1940s to >85% today), resulting in a changing profile in the population with CHD. The key question today is not mortality, but morbidity and longer-term quality of life beyond the second decade. As well as the need for more appropriately trained experts in adult congenital heart disease, attention needs to be focused on some of the following issues:

- What is the cause of impaired cardiac output and exercise tolerance in the Fontan circulation? Is it impairment of ventricular function? Is it abnormal preload/afterload or is it reduced E:A ratio with prolonged isovolumic relaxation time, representing reduced ventricular filling during atrial systole as a result of abnormal preload, rather than impaired contractility? Drugs such as enalapril reduce systemic vascular resistance, but do not affect the baseline hemodynamics or improve exercise capacity.
- Coronary flow reserve in patients following the arterial switch operation for transposition of the great arteries is significantly reduced even in asymptomatic patients – what does the longer-term future hold for these patients?
- 50% of patients following repair of aortic coarctation remain hypertensive and this becomes more common as a result of increased aortic stiffness and reduced arterial compliance as patients become older.
- Pulmonary regurgitation following tetralogy of Fallot repair – when should we perform pulmonary valve replacement? Should we use the right ventricular (RV) volume of 150 ml/m^2 or is valve replacement being performed too late, even using this volume criterion? Percutaneous pulmonary valve implantation has been one of the best interventional advances over the past 10 years. Although a technically demanding procedure, it results in significant improvement in hemodynamics and patient status.
- Congenital cardiac MRI is a major advance in providing detailed anatomical and functional information; however, in Hess's view, it should only be performed by specifically trained experts in congenital heart disease.

Hess concluded that all of these issues provide a continuous challenge for practitioners in CHD to which we must adapt.

Plenary session: role of hybrid therapy

G Tulzer (Children's Heart Center, Linz, Austria) gave a lecture on the treatment strategies of borderline left heart. Does fetal aortic valvoplasty prevent hypoplastic left heart syndrome? The anatomical indications for appropriate patients are still not well defined. In his unit in Linz, Austria, 44 fetal aortic interventions were performed with 80% technical success and 10% mortality. The rate of long-term biventricular outcome was 54% at a median age of 3.5 years (range 0–8 years). Various criteria including the Rhodes score (reported in 1991 [4]), Congenital Heart Surgeons Society score (in 2001 [5]), Discriminant Score (reported by Colan *et al.* in 2006 [6]) and Univentricular survival advantage score are all used, but there is still no perfect score to determine biventricular versus univentricular strategy. L Benson (Hospital for Sick Kids, Toronto, Canada) presented the hybrid procedure as an alternative to the Norwood operation, which has the potential advantage of diminishing postoperative

hemodynamic instability and neurological insult [7]. Although data on this are currently still lacking, a major study is underway comparing these strategies, which may clarify the future selection criteria for the hybrid approach. Retrograde aortic arch stenosis may be an important complication of the hybrid strategy, which is managed in Toronto by routinely placing a reversed Blalock-Taussig shunt in patients with aortic atresia [8]. D Anderson (Evelina Children's Hospital) discussed changes in hybrid strategy recently adopted at the Evelina Children's Hospital. The potential problems with the hybrid procedure include retrograde aortic arch obstruction, poor pulmonary artery growth or development of stenosis, obstructed pulmonary venous return leading to increased pulmonary vascular resistance and depressed ventricular function due to increased afterload (poor stent compliance and banded branch pulmonary arteries). Consequently, changes have been made in the approach to using flexible self-expanding stents, such as Sinus Superflex DS (OptiMed, Ettlingen, Germany), similar to the strategy developed in University Clinic, Giessen, Germany [9], and an early stage I Norwood operation at 2–3 months of age, rather than a comprehensive stage II Norwood at an older age. With this strategy, improved results have been observed.

Working group symposium – intervention: coronary arterial fistulas (2013 updates)

Clinical impact, natural history and indication for treatment of coronary fistulas (J Thompson, Leeds General Infirmary, UK). Most (>75%) of coronary artery fistulas are clinically silent and symptoms (such as heart failure) typically occur either in neonates or adults. Complications related to coronary arterial fistulas (CAF) appear more evident in adults; suggesting progressive enlargement of the fistulas over time; however, even large CAFs have been reported to spontaneously close. The major question is when to close a CAF. Should it be closed when another lesion needs treating or when there are symptoms or complications, or when the CAF is medium-to-large, even when patients are asymptomatic in childhood? The long-term complications of CAF closure include coronary thrombosis and myocardial infarction due to thrombus propagation, and so long-term anticoagulation should be considered [10].

Which fistula should be interventionally closed? (S Qureshi, Evelina Children's Hospital). Transcatheter occlusion of CAF is recommended for patients above 10 kg and only considered in patients smaller than this if intractable symptoms of heart failure are present. A variety of devices and coils can be used depending on the morphology of the fistulas. Trial balloon occlusion for 10 min is important to exclude potential myocardial ischemia prior to permanent occlusion with a device. If the coronary artery is dilated all the way back to the aorta, then lifelong anticoagulation is recommended, as these patients are at high risk of coronary thrombosis.

Which fistula should be closed surgically? (G Sarris, Mitera Hospital, Athens, Greece). Transcatheter occlusion is ideal for single, narrow and restrictive fistulas. Fistulas that are inaccessible by catheter or too large for device closure, or have multiple connections, may be surgically closed. Simultaneous bypass grafting can be considered during surgery, if the risk of coronary artery occlusion is high. Long-term complications may also occur with surgical closure, with a recently published study from the Mayo Clinic identifying 24% late mortality after surgical repair of CAF [11].

Working group symposium – imaging: all about right ventricular function in Tetralogy of Fallot

W Helbing-Leiden (Erasmus MC, Rotterdam, Netherlands) described the use of speckle tracking to assess regional RV function after tetralogy of Fallot repair and highlighted the importance of the RV diastolic as well as systolic function. He reviewed a new and very interesting report demonstrating that patients with more functioning *HIF1A* alleles had higher TGF-β1 expression and more fibrosis at initial repair compared with a control population. During follow-up, those patients with more functioning *HIF1A* alleles showed less RV dilation, better preservation of RV function and a greater freedom from RV reinterventions [12].

Pulmonary hypertension (T Delhaas) may be a biventricular disease as the left ventricular (LV) pump function interacts with the RV pump function due to their complex geometry. Early RV free-wall activation via pacing reduces the myocardial fiber strain in a computational model (CircAdapt) with RV free-wall pacing resulting in a 7% decrease in RV end-diastolic pressure and a 6% decrease in RV end-diastolic volume. There may be an improvement in LV filling dynamics mainly due to an increased RV systolic output; that is, it is LV under-filling, not septal bulging, that mainly affects LV function. The septal curvature responds more to

a reduction in pressure overload and so early diastolic stretch of the LV free wall predicts RV systolic dysfunction.

Transposition of the great arteries after atrial correction (C Meierhofer, Heart Centre). Systemic RV dysfunction occurs over time in nearly all of the patients after atrial correction surgery, and severe RV dysfunction may be present in up to 10% of patients. Subnormal RV function develops due to the RV geometry, myocardial fiber arrangement, coronary blood supply, underlying arrhythmia/electrical abnormalities and systemic ventricular work demand. Cardiac magnetic resonance is the gold standard for the assessment of RV function and fibrosis detected on MRI, with late gadolinium enhancement corresponding to poor RV function.

Hypoplastic left heart syndrome after staged Fontan (A Bell, Evelina Children's Hospital). The assessment of RV function is challenging in hypoplastic left heart syndrome due to the variation of morphology, the impact of LV and the impact of different surgical stages. Subjective assessment of RV function on 2D echocardiography correlates with MRI measurements in only approximately 60% of cases, even when performed by the most experienced specialists. Tricuspid annular plane systolic excursion changes with loading conditions. 3D echocardiography is reasonably reproducible, but underestimates the end-diastolic volumes and ejection fraction. Cardiac MRI is the gold standard for the assessment of RV volume and function, but there remains a degree of interobserver error.

Association for European Paediatric & Congenital Cardiology joint session: failing Fontan

Protein losing enteropathy: interventions & prognosis (N Sreeram; University Hospital, Cologne, Germany)

Protein losing enteropathy (PLE) can be precipitated by acute increases in pulmonary vascular resistance (PVR) due to infections. A low-fat diet is part of the treatment and medications used include diuretics, angiotensin-converting enzyme-inhibitors, steroids (usually effective, but transiently with significant side effects), heparin, sildenafil and somatostatin. Sildenafil therapy improves cardiac output and exercise capacity, but there is no evidence for its use in PLE. Catheter intervention should be used to relieve any obstruction in the Fontan pathway. Creation of a fenestration or its enlargement, if already present, can result in an acute improvement, but there are no long-term data available, and a high rate of spontaneous closure of the fenestration has been reported. Pacemaker implantation for sinus node disease and radiofrequency ablation for arrhythmias can be performed, but there are no data available on the effect on PLE. Surgical Fontan revision has generally been disappointing for PLE and most treatments are ineffective or only temporarily beneficial.

Fontan circulation: conversion and transplantation (J Comas, Pediatric Heart Institute, Madrid, Spain). The 20-year survival of total cavopulmonary connection is approximately 80%. Conversion is performed once all other options have failed; however, it is contraindicated in severe ventricular dysfunction. Cardiac transplantation following the failure of the Fontan circuit has higher levels of acute mortality compared with transplantation in other congenital heart lesions and, often, PLE does not improve after transplantation, unless this was secondary to impaired ventricular function preoperatively. It is a big challenge to identify the failing Fontan prior to its true 'failure'.

Working group symposium: aortic coarctation in adulthood

How to test systemic arterial pressure (A Giardini, Great Ormond Street Hospital): Following aortic coarctation repair, 33% of patients may have abnormal 24-h blood pressure (BP) recordings, and 47% of the patients have high BP on stress testing. Isolated BP measurements in the clinic are not sufficient to identify hypertension in many patients. Therefore, 24-h BP monitoring should be performed every 2 years in all patients with repaired aortic coarctation, and exercise testing should be performed every 2–3 years. All hypertensive patients should be investigated for a possible recoarctation, especially if hypertension is resistant to medical therapy. Those patients with hypoplastic aortic arch or prosthetic material are at particularly high risk of hypertension.

Questions:

- Should rest-negative but exercise-positive patients be treated?
- Should modest gradients (<20 mmHg) associated with hypertension (rest or exercise) be eliminated?
- Should the transverse arch be enlarged at the time of primary surgery to reduce risk of hypertension?

Summary

Overall, the 47th annual meeting of the Association for European Paediatric & Congenital Cardiology was a resounding success, not only detailing the latest advances in congenital heart disease, but also focusing on the challenges due to the unsolved queries. The congress fostered a friendly atmosphere throughout the global audience and was of interest to all because of the highly variable scientific program.

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Website

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