When God does not want me to die

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In August 2010, an internationally renowned plastic surgeon aged 47 collapsed in his operating theatre and was asystolic and effectively dead. The circumstances over the ensuing 2 hours were remarkable and involved 2 cardiologists who were able to try to resuscitate him, emergency femoro-femoral access to an ECMO machine being immediately available and immediate coronary stenting of a totally occluded left main, all performed within a short period.

It left him in dire cardiac straits on an ECMO machine, respiratory failure on a ventilator, renal failure on dialysis and a subsequent stroke causing neurological disorder. Despite all these setbacks and intermittent infections, he survived and 6 weeks later, it was decided that he would never survive without an orthotopic heart transplant. He was discussed on 5 October prior to his transplant by the multidisciplinary team who felt that he was felt that he was a candidate for transplantation.

Three nights later, an ideal donor became available. Should the donor go to a 47-year-old colleague on life support or should it go to a possibly more worthy recipient who had been on the waiting list for a long time? This is the ethical dilemma.

It was decided by the team to proceed with his transplant, which has been remarkably successful. Apart from requiring dialysis for another 2 months, numerous infective episodes, which originated prior to the transplant and 1 episode of mild rejection, his recovery has been truly remarkable.

The series of circumstances from the day of his collapse to the present time would certainly suggest that “it was not his time to die” and the multidisciplinary team made the correct assessment to offer him the donor heart on 9 October 2010.

The patient has made a remarkable recovery from his initial illness, is compliant with all medication, but has made a decision at this time not to work as a plastic surgeon. It is my privilege to present his story in the Ethics Section of the SA Heart Congress. This is truly a unique story worthy of telling.

Rheumatic heart disease in pregnancy

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Rheumatic heart disease, though virtually eradicated in the first world, remains a common debilitating disorder in developing countries and in our setting is the commonest cause of cardiac disease in pregnant women.

Pregnancy is associated with significant haemodynamic changes that occur as early as the first trimester, peaks in the second and plateaus until delivery. Underlying valvular heart disease is often unmasked in the second trimester but may go unnoticed due to the overlap of normal physiological pregnancy-related physical signs and that of heart failure.

The risk to mother and foetus depends on the degree of symptoms and functional class of the patient, the type and severity of the valve, left systolic function and pulmonary pressures.

Regurgitant murmurs are low risk and usually well tolerated. The problem arises with stenotic lesions though. Of these, mitral stenosis is the commonest. Mild to moderate mitral stenosis is tolerated well but when the valve area is <1.5cm², decompensation usually occurs even if the patient was previously asymptomatic. Aortic stenosis is usually well tolerated if mild with a normal ejection fraction.
Management of these patients involves a multi-disciplinary approach including a cardiologist, obstetrician and anaesthetist. Ideally these patients should be assessed prior to pregnancy to see whether or not the pregnancy would be safe and to plan antenatal and delivery management. Regular follow-up by both obstetricians and cardiologists during the pregnancy is required in high risk patients.

The recognition and surgical management of chronic pulmonary thromboembolic disease

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Chronic pulmonary thromboembolism resulting in pulmonary hypertension (CTEPH) is truly the “aortic stenosis” of the pulmonary circulation. The development of pulmonary hypertension and subsequent right heart failure is a progressively debilitating process which is often misdiagnosed or diagnosed late. The unresolved thromboembolic material undergoes chronic fibrotic changes causing an obstructive lesion that is unresponsive to medical therapy.

The initial deep vein thrombosis or pulmonary embolus is often silent and there is a subsequent “honeymoon period” before symptoms and signs of pulmonary hypertension develop. Exertional dyspnoea should alert the diagnosis and patients with pulmonary hypertension should have the diagnosis of CPTEH excluded.

A VP scan will suggest the diagnosis, but not the extent of disease. Echocardiography, CT scan and Pulmonary angiography confirm the diagnosis and the anatomic extent and sites of obstruction. Jamieson, Thistlethwaite and colleagues have advocated an intraoperative classification of CTEPH into 4 groups according to location of thrombus and vessel wall pathology. This classification correlates well with postoperative outcome. Possible angiographic predictors of haemodynamic improvement after surgery have been suggested by Takashi, Funihari and colleagues.

Treatment is surgical via a median sternotomy and requires bilateral pulmonary thromboendarterectomy under profound hypothermia and circulatory arrest. Possible surgical candidates depend on:

- Anatomic variables (extent and proximal location of the pulmonary vascular obstruction).
- Haemodynamic variables (severity of pulmonary hypertension, PVR and the function of the right ventricle).
- Patient variables (comorbid conditions – severe parenchymal lung disease with obstructive or restrictive dysfunction).

Best outcomes found where there is a match between PVR and degree of angiographic occlusion on CT and angiography. Postoperatively there is an improvement in haemodynamics, which causes reverse RV remodelling with reduction in tricuspid regurgitation and return of RV systolic and diastolic function towards normal levels. Functional status improves to Class I or II in 90% of patients, mean reduction in PVR may be 50 - 65%.

Life-long anticoagulation is necessary and investigations and surgery should probably be referred to specialist centres for treatment.
An update of pharmacogenetics

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Pharmacogenetics is the search for heritable genetic polymorphisms that influence responses to drug therapy. The most important application of pharmacogenetics is to guide choosing agents with the greatest potential of efficacy and smallest risk of adverse drug reactions. Many studies focussing on drug-gene interactions have been published in recent years, some of which led to adaptation of FDA recommendations, indicating that we are on the verge of the clinical application of genetic information in drug therapy.

Besides the genetic variation depicted in the table below, the evidence for most other pharmacogenetic associations is not strong enough for clinical application. Nevertheless, currently several guidelines on drug prescription already contain pharmacogenetic information. Very recently, an update was published of the current Dutch guidelines implementing pharmacogenetics in several therapeutic recommendations. In addition, over 60 FDA-approved drug labels contain pharmacogenetic information. For instance, in 2007, the FDA recommended initiation warfarin doses for carriers of certain CYP2C9 and VKORC1 polymorphisms.

Moreover, in 2009, the FDA added a warning to the clopidogrel prescribing information to highlight the impact of the poor metaboliser genotypes of CYP2C19. Finally, the pharmacogenetic score developed in the PERGENE trial seems to be able to identify patients that will not benefit from ACE inhibitor treatment and the clinical consequences of this knowledge, whether indeed patients with this specific risk profile should not receive ACE inhibitor treatment, should be prospectively tested. Ongoing large clinical trials, primarily designed as pharmacogenetic studies, and GWAS with large study populations promise to provide evidence that is more convincing.

Overall, there are enough reasons to remain optimistic that, even though we are taking small steps at a time, we are heading to an era where we can finally use pharmacogenetics in clinical practice to optimise treatment for the individual patient.


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<tr>
<th>Drug class</th>
<th>Problem</th>
<th>Evidence</th>
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<tr>
<td>Statins</td>
<td>Variable lipid lowering</td>
<td>Mediocre</td>
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<td></td>
<td>Risk of myopathy</td>
<td>Strong</td>
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<td>Antiplatelet drugs</td>
<td>Resistance</td>
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<td>Anticoagulants</td>
<td>Dosing problems</td>
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<td>Beta blockers</td>
<td>Variable response</td>
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<td>ACE inhibitors</td>
<td>Variable blood pressure reduction</td>
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<td></td>
<td>Variable benefit to decrease risk of events</td>
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Update on women and cardiovascular disease

**Naomi Rapeport**
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Heart disease is the leading cause of death in women. Mortality rates in women over the age of 60 years are more than double in low- and middle-income countries than in high-income countries. Exposure to risk factors starts earlier in life, and therefore preventive interventions need to target younger women. The first women-specific clinical recommendations for the prevention of CVD were published in 1999, even though there was little gender-specific research data. Since the late 1990s, increasing numbers of women have participated in CVD studies resulting in gender-specific analyses. Major randomised controlled clinical trials in women have changed the practice of CVD prevention.

“Evidence-based guidelines for cardiovascular disease prevention in women” have been published and updated over the past 7 years. Emphasis has been placed on preclinical detection of disease to identify asymptomatic individuals at high risk, who could benefit from early intervention. Despite these guidelines, there are problems with risk stratification of women, with a general under estimation of CHD risk, which has focused on short-term (10-year) risk and on MI and CHD death. The 2011 guidelines have addressed these anomalies. The focus is now on long-term risk for CVD rather than solely on 10-year risk for CHD. The new cut point for defining “high risk” is a risk of 10% or more of death from any cardiovascular event in the next 10 years.

Other modifications include the use of new risk stratification scores (the updated Framingham CVD risk profiles and the Reynolds risk score for women). New major risk categories include patients with systemic autoimmune collagen-vascular disease and those with a history of preeclampsia, gestational DM or pregnancy-induced hypertension.

A new concept of “ideal cardiovascular health” has been proposed. This is the absence of clinical CVD and the presence of ideal levels of cholesterol, blood pressure, blood glucose, body mass index, and a lifestyle that includes smoking abstinence and physical activity. The major hurdle is to implement these guidelines early. This is particularly so in low- and middle-income countries, where there is a looming pandemic of CVD.

Anti-coagulation in pregnancy

**Vera Regitz-Zagrosek**
Institute of Gender Medicine, Centre for Cardiovascular Research, Charité – Universitätsmedizin, Berlin

Anti-coagulation in pregnancy is a major, partially unsolved problem. It is of particular clinical relevance in patients with mechanical valve prostheses, with severely depressed ejection fraction, with arrhythmia and at risk for thrombembolism. Vitamin K antagonists are safe for the mother, but may cause embryopathy in the first trimester. Heparin is safe for the baby but less efficient to prevent maternal thrombembo-lism. Adequate prospective studies on both regimes are lacking.

The ESC Task Force on the management of cardiovascular disease during pregnancy gave the following recommendations in September 2011:

In patients with mechanical valve prostheses, anti-coagulation with vitamin K antagonists is the safest therapy to prevent valve thrombosis and the choice of therapy during the second and third trimester. During the first trimester, if anti-coagulation is needed, oral anti-coagulants (OAC) should be considered when the required daily dose is low (equivalent to warfarin <5mg). If higher doses are required, unfractionated heparin (UFH) or LMWH with strict dose adjustment according to APTT or anti-factor Xa levels should be considered. At the 36th week of gestation OAC should be discontinued and replaced by dose-adjusted UFH or LMWH. In patients with severely impaired left-ventricular ejection fraction or atrial fibrillation, LMWH or OAC should be used according to stage of pregnancy. LMWH is the drug of choice for prophylaxis of VTE in pregnancy. In suspected pulmonary embolism, treatment with LMWH should be given until the diagnosis is excluded by objective testing. If the diagnosis is confirmed intravenous administration of UFH is recommended. For postpartum management, if no signif-icant bleeding has occurred, prepartum heparin should be restarted with subsequent overlap with OAC.
**Gender in Pharmacotherapy**

**Vera Regitz-Zagrosek**  
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Gender aspects in pharmacological therapy are of major relevance since they affect a large number of patients with all kinds of diseases and contribute to a significant number to emergency hospital admissions. More than half of emergency departments' visits by elderly patients in internal medicine are due to adverse effects of pharmacotherapy. These are much more frequent in women than in men and are frequently due to overdosing or not considering interaction between sexual hormones and a given drug. Pharmacodynamic and pharmacokinetic effects contribute to gender specific adverse effects. In the field of cardiovascular medicine, relative overdosing of Digitalis caused increased mortality in women in the Dig-study. Relative overdosing of anti-coagulants or thrombolytics leads to more adverse effects and bleeding in women than in men; anti-arrhythmic drugs with QT-prolongations have more pro-arrhythmic effects in women. A gender specific spectrum of adverse effect has also been reported for beta-blockers, ACE-inhibitors, and glitazones. The aldosterone antagonists eplerenone appeared to be more efficient in women than in men in 1 clinical study and animal experiments.

Efficiency for non-cardiovascular drugs also varies according to sex and gender: opioid analgesics, neuro-muscular blocking agents, anti-depressants and a number of anti-cancer drugs also exhibit differences in efficiency profile and adverse effects in women and men. Thus, adequate investigation of all drugs in male and female animals and in early phase studies before marketing is mandatory.

**Female sex and oestrogen receptor-beta attenuate cardiac remodelling and apoptosis in pressure overload**

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Sex differences in the manifestation of myocardial hypertrophy (MH) in patients with aortic stenosis can be reproduced in an animal model of pressure overload (PO). We investigated the influence of ERβ on the pathophysiological and molecular mechanisms leading to sex-differences in PO induced cardiac hypertrophy and heart failure.

We performed transverse aortic constriction (TAC) or sham surgery in male and female C57Bi/6 (WT) mice and with genomic deletion of ERβ (ERβ−/−). The animals were echocardiographically and haemodynamically characterised and sacrificed 2 or 9 weeks after surgery. Left ventricles (LV) were analysed by microarray, histology, RT-PCR and immunoblot. Nine weeks after TAC, MH was present and was significantly more pronounced in WT males than in females. In the ERβ−/− animals males developed a more eccentric form of MH than females after TAC. It was accompanied by the impairment of systolic and diastolic function and the beginning of left ventricular dilation. Male WT TAC animals showed a strong increase in cardiac fibrosis whereas female WT hearts showed no changes of collagen content after TAC. In contrast, ERβ−/− female animals developed more severe fibrosis after TAC. Only ERβ−/− male TAC animals went into the apoptotic gene programme. Microarray analyses were in agreement with functional changes.

In summary, WT animals, the hypertrophic response in pressure overload induced MH is characterised by severe cardiac fibrosis in males, while mitochondrial activity is better maintained in females. Female ERβ−/− animals develop a more severe cardiac phenotype similar to WT males 9 weeks after TAC. ERβ contribute to the maintenance of energy homeostasis in female mice and limits the development of eccentric cardiac hypertrophy, fibrosis and apoptosis in female and male mice.
Update on lipid management in South Africa

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Hypercholesterolemia is recognised as a major risk factor for the development of coronary artery disease. The degree to which patients in South Africa are being treated to accepted international LDL-cholesterol (LDL-C) targets has been previously studied, but factors responsible for the undertreatment of hyperlipidaemia are largely unknown. The CEntralised Pan South African study on tHe Undertreatment of hypercholeSterolemia (CEPHEUS) looked at 3 000 subjects across the ethnic spectrum of South Africa, who were on lipid-lowering therapy, to determine the number of subjects achieving LDL-C goals and to search for reasons for failure to achieve goal.

Whilst patients of African ancestry were just as likely to be undertreated as their Caucasian counterparts (58% at goals vs. 54.5%), they were more likely to be obese (BMI 32.6 vs. 29.8), hypertensive (87.5% vs. 64.7%), diabetic (73.5% vs. 31.3%) and have features of the metabolic syndrome (82.9% vs. 59.9%). Data from the 1 424 women in the study showed similar results between the ethnic groups, but overall women were less likely to reach the goal than men (OR 0.81). Responses to patient questionnaires reveal a reluctance to change medication type and/or dose, with 64% of subjects on the same drug and same dose as the initiation drug and dose after a mean of 5.8 years of therapy. 87.2% of participating practitioners felt constrained by funder reimbursement formularies in their choice of treatment.

These results suggest the need for greater education amongst patients and practitioners about the importance of LDL-C goals, and for a greater input and effort from medical aid funders/administrators to facilitate the attainment of these goals.

Genetic testing for sudden death: Who, Why, and When?

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Many arrhythmogenic diseases associated with increased risk for sudden cardiac death have a genetic origin and, thereby, it is possible to identify the individuals (or some of them) who carry a disease-causing mutation by genetic screening. This is particularly true for the group of diseases usually referred to as channelopathies and cardiomyopathies. Among them, without any doubt the 1 for which more progress in understanding the complex relationship between genotype and phenotype, is the congenital long QT syndrome (LQTS). Accordingly, this presentation will focus on the many issues surrounding genetic testing for LQTS.

This year 2 consensus statements dealing specifically with genetic testing for diseases associated with sudden cardiac death were published and are likely to have a significant impact. I will discuss the main aspects related to genetic testing for LQTS independently of what those documents have stated, which is largely correct. Instead, based on 40 years of personal experience with this intriguing and often lethal disorder, I will focus on what is most relevant to the practising cardiologist. My main objective will be to provide clinicians who may encounter these patients rather seldom with simple tools to correctly decide when it is appropriate to recommend genetic testing and when not to do so would represent an inexcusable error.
Aortic valve and the advances in the role of imaging (ECHO & CMR)

Joseph Selvanayagam
Department of Cardiology, Flinders Medical Centre, Flinders University of South Australia, Adelaide, Australia

This talk will focus on the role of echocardiography and cardiovascular magnetic resonance in the assessment of the aortic valve. Particular emphasis will be made on its role in the pre-procedural evaluation of aortic stenosis. Comment will be made on the recent advances in the use of 3D echocardiography and cardiovascular magnetic resonance on assessment of valvular function structure and aortic dimensions. The complimentary roles of the various imaging techniques will be discussed.

Cardiac function – how does CMR measure up to ECHO?

Joseph Selvanayagam
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This talk will focus on the technique of Cardiovascular Magnetic Resonance (CMR), and how it assesses both systolic and diastolic left ventricular function. Comment will also be made about CMR assessment of right ventricular function.

CMR has rapidly become the imaging method of choice and the goal standard in the assessment of cardiac function of both normal and abnormal ventricles. With regard to the measurement of global left ventricular systolic function, given its 3D nature and order of magnitude greater signal to noise ratio, CMR is highly superior to 2D echocardiography. This imaging is typically performed in, although not limited, to the conventional serial short axis views and the 3 cardinal long axis views. The ability of CMR to imaging any plane without the need for optimal imaging windows allows for unprecedented flexibility for the interrogation of abnormal heart structures. CMR is also considered to be the most accurate imaging method for the evaluation of right ventricular (RV) volumes. CMR measurement of RV volumes has been validated with course correlation between RV and LV stroke volumes and between RV stroke volumes and tricuspid flow measurements. The inherent 3D nature of CMR makes it particularly well suited to study the RV given its complex and variable (even in normal volunteers) morphology. Cine images are obtained using steady state free precession (SSFP) sequence, which provide images of higher signal to noise (than older sequences such as gradient echo), and hence exceptional delineation of blood to myocardium interface. This allows for accurate and reproducible quantitative assessment of chamber dimensions and systolic function using manual or semi-automated preliminary techniques. These will be discussed during the talk briefly.

CMR has traditionally been considered inferior to echocardiography for the assessment of left ventricular diastolic function. However, with improvements in both hardware and software, and in particular rapid imaging and hence improved temporal resolution, CMR now does offer several approaches to the measurement of diastolic function.

CMR assessment of myocardial viability – the new kid on the block

Joseph Selvanayagam
Department of Cardiology, Flinders Medical Centre, Flinders University of South Australia, Adelaide, Australia

Viability assessment can be defined practically as detecting myocardium that shows severe dysfunction at rest, but which will improve function, either spontaneously with time (stunned) or following revascularisation (hibernating). The identification of residual myocardial viability is critical to the management of patients with ischaemic heart disease. Delayed enhancement CMR with gadolinium chelates was first described as far back as 1984, in a canine model of acute myocardial infarction. However, initial studies were hampered by insufficient image contrast between normal and injured myocardium due to technical and sequence limitations.
This talk will describe the practical aspects of delayed enhancement imaging, its validation in large animal models, the large array of clinical studies that have occurred both in acute and chronic CAD settings, as well as more recent modifications of the protocol including the addition of T2 weighted imaging for the detection of myocardial oedema (and hence myocardial salvage in the acute setting). Comparison will be made with already existing modalities, in particular nuclear techniques and stress echocardiography. Advantages and disadvantages of the methods will be discussed. Ancillary information provided by the CMR technique would also be discussed during the talk.

**CMR assessment of unusual cardiomyopathy**

**Joseph Selvanayagam**

Department of Cardiology, Flinders Medical Centre, Flinders University of South Australia, Adelaide, Australia

This talk will concentrate on the CMR assessment of non-compaction cardiomyopathy (LVNC). LVNC, although rare, is being increasingly recognised, particularly with the use of cardiovascular magnetic resonance and its associated high spatial resolution. Comment will be made during the talk about the CMR characteristics of this rare cardiomyopathy, its distinguishing features with respect to other cardiomyopathies (such as hypertrophic and dilated cardiomyopathy), as well as physiological causes of hypertrophy such as athletes heart. A seminal study in this area would be discussed. Finally, its remaining challenges in its diagnosis will also be outlined.

**Cardiomyopathy in pregnancy**

**Karen Sliwa**

Hatter Institute for Cardiovascular Research, Department of Medicine, Groote Schuur Hospital and the University of Cape Town, South Africa

Women can have a pre-existing cardiomyopathy or develop cardiomyopathy pre- or postpartum. Women with pre-existing cardiomyopathies have a limited cardiovascular reserve. The haemodynamic challenges of pregnancy, labour and giving birth pose unique risks to these patients, which can lead to decompensation presenting as heart failure, arrhythmias and maternal death. The clinical course and management of pregnant women with hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, left ventricular non-compaction cardiomyopathy, as well as peripartum cardiomyopathy will be presented.

Asymptomatic patients with inherited cardiomyopathies generally tolerate pregnancy well, but worsening of the clinical condition could occur during pregnancy even if intensive medical treatment is provided. Managing heart failure in pregnancy requires special considerations. Angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor II blockers are contraindicated on pregnancy because of teratogenicity, neonatal anuric renal failure, and neonatal death. Use of spironolactone is not recommended. Atenolol is associated with a higher incidence of foetal growth restriction compared to other beta-blockers and its used is not advised.

Anaesthetic considerations in pregnant women with cardiomyopathy require a specialised approach and, when possible, should be planned in the antepartum period. The risk of maternal cardiac complications during pregnancy is significantly increased if prior cardiac events, poor functional class (NYHA Class III or IV) or advanced left ventricular systolic dysfunction are present. Although no long-term follow up studies have been reported, in general the postpartum condition is no worse than the antepartum condition. Counselling and preconceptual evaluation are important factors in the management of women with inherited cardiomyopathies.
Assessment of pulmonary hypertension in the catheter laboratory

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Department of Cardiology, Royal Children’s Hospital, Melbourne, Australia

Introduction: Pulmonary hypertension may be the consequence of congenital or acquired heart disease or of primary pathology affecting lung vasculature. When produced by congenital heart disease the usual cause is a septal defect either at ventricular or arterial level, which exposes the pulmonary circulation to systemic arterial pressure. In the presence of acquired heart problems such as mitral stenosis, the problem results from elevation of pulmonary venous pressure leading to raised pulmonary artery pressure. Primary disease of pulmonary arteries/arterioles or pulmonary veins may also lead to pulmonary hypertension.

In all these situations, the pulmonary arterioles respond with a sequence of pathological changes which result in gradual increase in vascular resistance and reduction in the extent to which recovery can occur. Reversibility depends on the severity and the duration of pulmonary hypertension and also on its cause.

Decisions about treatment, including surgery, require careful assessment with measurement of pressure, flow and resistance in the pulmonary circulation.

Methods: Catheter laboratory equipment will document pressures and provides a range of calculations, which will use oxygen saturation data, coupled with haemoglobin measurement and oxygen consumption to measure flow and resistance. It is important however that the cardiologist and technicians who use such equipment have a clear understanding of the basis for these calculations and look at the data critically rather than merely accepting the numbers that the equipment generates.

There are a number of assumptions implicit in the calculations and several of these have potential for serious errors. Awareness of these is vital if the measurements are to be useful. An obvious example is the likelihood of significant amounts of dissolved oxygen in pulmonary venous blood when the patient is breathing gas containing high oxygen concentrations (as employed to assess lability of pulmonary hypertension). Unless this can be measured and factored into the calculations the flow measurements and resistance calculations will be incorrect.

Another source of error may be the use of assumed, rather than measured, oxygen consumption.

Conclusion: Measurement of pulmonary blood flow and resistance is subject to a range of technical difficulties and potential errors. Awareness of these is important.

Cardiac function long-term after the Fontan operation

James L. Wilkinson
Department of Cardiology, Royal Children’s Hospital, Melbourne, Australia

Introduction: More than 40 years have passed since the first Fontan operation was performed. During that time the procedures has been modified many times and the indications have been clarified and extended. A wide range of patients with “functionally univentricular hearts” are now managed with the procedure and results have improved steadily. Early survival is now excellent in many centres but late morbidity and mortality have been recognised as an inevitable threat for this cohort of patients. Twenty years ago, in an analysis by Fontan and colleagues, there was an expectation that survivors after a “perfect Fontan” would experience significant late mortality with anticipated survival at 15 years of 73%.

Methods: A cohort of more than 300 patients who had Fontan procedures between 1980 and 2000 were reviewed to assess survival and complications with different modifications of the operation.

A group of long term survivors (17 ± 4 years post Fontan) were studied to assess cardiac function and quality of life using several methods which included cardio-pulmonary exercise testing, echocardiography, QoL questionnaires and measurement of neuro-humeral factors.

Results: Early mortality dropped from 6.3% in the first decade of the study period to 0% in the second. Survival at 15 years was 81% for the atrio-pulmonary Fontan patients and 94% for lateral tunnel. Extracardiac Fontan patients appear to be doing better than the lateral tunnel group with 100% survival at 9 years.
Freedom from late complications including SVT and heart failure showed a similar trend with extracardiac Fontans being superior to lateral tunnel patients, who fare better than atrio-pulmonary Fontans.

Exercise testing demonstrates that Fontan patients have a reduced exercise capacity, with associated reduction of VO2 Max, which was less marked in the Lateral Tunnel group than in the Atrio-pulmonary Fontan patients. Anaerobic threshold was also reduced, but to a lesser degree, and was again better in the lateral tunnel group.

Quality of life was generally perceived as being “normal” in long-term survivors without arrhythmias.

**Conclusion:** With newer variations of Fontan, long-term survival and quality of life have improved substantially over the last 3 decades.

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**The pursuit of excellence - Setting the bar high**

*James L. Wilkinson*

Department of Cardiology, Royal Children’s Hospital, Melbourne, Australia

**Introduction:** Excellence may be defined as “The quality of being exceptionally good. A quality that confers superiority” (dictionary definition). In medicine, it may be applied to such areas as clinical skills, technical ability, research, teaching or outcomes.

Most doctors would probably aspire to achieving high levels of skill, potentially excellence, in at least 1 of these areas, though few will achieve it in all. From our patients’ perspective, we are judged mainly on “Outcomes”. This particularly involves minimising the risks and maximising the benefit of our treatments. We must work to improve safety and we must be constantly aware that patients and their families quite rightly fear, knowing from media publicity the high frequency of errors!

The achievement of excellence depends on our ability to minimise errors and to seek always to employ treatment strategies that are based on the best available evidence. At medical school and throughout our training as paediatric cardiologists we listen to our teachers as they imbue us with dogma and doctrine, some of which is soundly based but much of which is more eminence based than evidence based? Many of our teachers seem blissfully unaware that much of what they teach is likely to be proved incorrect. A quote from 1 distinguished teacher, with more insight than most, runs “Remember that half of what I tell you is wrong – the problem is that I don’t know which half?”

Excellence is not dependent simply on resource availability but on careful and critical assessment of risk and benefit of the treatments that we adopt for our patients. This basic philosophy is applicable in every medical environment whether in highly developed “Western” medicine or in the developing world.

**Conclusion:** In the pursuit of excellence, we need to:

- Ensure optimal training and continuing education (CME).
- Encourage critical review and discussion of literature (evidence).
- Audit outcomes on an ongoing basis and compare with results from elsewhere (audit).
- Make treatment decisions after appropriate inter-disciplinary discussion (teamwork).
- Work together – cardiologists and surgeons are complimentary not competitors (collegiality).