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EDITORIAL

Guest editor, Sara Thorne
Consultant cardiologist
Cardiology - University Hospital Birmingham NHS Foundation Trust
Queen Elizabeth Hospital, Queen Elizabeth Medical Centre, Birmingham

Congenital heart disease is growing up

Congenital heart disease is growing up: adult survivors, reaping the benefits of early surgical and percutaneous intervention, outnumber children in many developed countries. Many have little in the way of residual defects and can look forward to a long and healthy life. However, many others reach adulthood with increasingly complex surgically-modified disease – their truly long term outlook is unknown, but patients with complex anatomy and physiology will require careful and creative management, as described in the articles in this edition.

What principles should be followed to allow adults with congenital heart disease to live a long and healthy life? First of all they must not be lost to follow up at their transition from paediatric to adult services. All patients should be seen at least once by a specialist in adult congenital heart disease and appropriate long term care planned. With this in place, the overriding aim of follow up must be to preserve ventricular function.

The cardiologist must consider the different challenges faced by the ventricle. It may be the volume loaded right ventricle in the patient with repaired tetralogy of Fallot and severe pulmonary regurgitation, or a large unrepaired atrial septal defect. It may be the pressure loaded right ventricle in the adult with high pulmonary vascular resistance due to abnormal pulmonary vasculature or pulmonary arterial hypertension, or it may be the pressure loaded systemic right ventricle in the post Mustard/Senning or congenitally corrected transposition patient. The at risk left ventricle may have been chronically volume loaded: such as in an adult whose left to right shunt was not repaired until late in childhood, or who had longstanding left atrioventicular valve regurgitation. Or it may have faced a long term pressure load, such as in the patient with the difficult to repair small left ventricular outflow tract. The single ventricle is at high risk and appears to have a limited capacity to maintain normal function in the long term. The cyanotic "unrepaired" single ventricle eventually fails under the strain of chronic volume loading, whereas the single ventricle in a Fontan circulation may fail under the burden of a chronically high afterload. Whether it will be possible to help the majority of adults with a single functional ventricle maintain a good quality of life beyond their forties and fifties is in question.

These complex patients need monitoring to determine if and when to re-intervene in order to maintain the best ventricular function for the longest time. High quality non invasive imaging – in particular echocardiography and magnetic resonance imaging (MRI) – is crucial to decision making. New advances in percutaneous intervention will reduce the number of open operations patients need in their lifetime, limiting the repeated insults of cardiopulmonary bypass on the ventricles and preserving function.

Guest editor,
Sara Thorne

Along with ventricular dysfunction, arrhythmia is responsible for significant morbidity and mortality. Ventricular tachycardia is a well recognised cause of sudden death in those with abnormal right ventricles. However, atrial arrhythmia, in particular atrial flutter probably causes more morbidity and more deaths. Those at greatest risk of sudden death are the post Mustard/Senning repair of transposition: atrial flutter is common and frequently conducts 1:1, causing cardiovascular collapse. Urgent electrical cardioversion is required, and ablation has a high success rate, despite the distorted atrial anatomy. The patient with a Fontan circulation tolerates atrial flutter (often atypical) similarly badly, but ablation can be more challenging – or impossible, particularly in those who have undergone extracardiac total cavopulmonary connection.

Despite residual defects, most adults have a good quality of life and are able to work, have relationships and families. Pregnancy can be a great – but welcome - challenge for the patient and her cardiologist. It is a marker of the success of early management, and for many, proof that they can truly lead a normal life. The article by Roos-Heeslink in this edition of The Journal, ably describes the management issues that surround pregnancy.

Many of the techniques described in this edition are very expensive: magnetic resonance imaging, 3 dimensional echo, advanced electrophysiological mapping systems and percutaneous valves. Unchecked, any health service can become a financial bottomless pit. After the 2007 banking crisis, the UK economy has struggled to stay out of recession and its National Health Service (NHS) is increasingly cash-strapped. Service developments and new treatments vying for funding need to demonstrate their cost-effectiveness threshold and the cost per year of life gained in an attempt to ensure that restricted resources are used to the greatest benefit.[1]

The affordability of health care in a developing country is another question altogether. The demands on a limited health budget mean that the provision of good basic health care to the whole population will always take precedence over the needs of relatively small numbers of adults who have survived with complex and uncommon congenital heart conditions.

To improve cardiovascular health in South Africa, the emphasis is on investment in "Best Buy" measures that will improve the health of large sections of the population: the prevention of rheumatic heart disease, the development of multidrug regimes like combined antihypertensives, legislation to reduce salt and fat in food and antismoking measures. Measures to strengthen health service structures are also needed, developing district based services and regional centres of excellence with good communication between them, as well as on developing surveillance systems for acquired heart disease, stroke and diabetes.[2]

So, can a low-tech, low cost approach to care for the adult with congenital heart disease work? The answer is yes, with careful thought and some compromise.

The burden of congenital heart disease in Africa is significant, with up to 13% of new cardiac diagnoses in children and adults being congenital.[3] These patients are natural survivors with diagnoses like ventricular septal defect and tetralogy of Fallot; many of those with critical obstructive lesions will have died undiagnosed. Late presentation, with heart failure, pulmonary hypertension and the complications of cyanosis, makes effective treatment more difficult. So, the first task is to make the diagnosis of a congenital heart condition. Efforts to detect congenital heart disease need to be integrated into other programmes, such as the ones designed to pick up subclinical rheumatic heart disease, or to do oximetry on all newborns.

Working with non governmental organisations has helped to set up and maintain some cardiac surgical centres; however a lack of trained medical staff can present a problem. Where surgical access is limited

a percutaneous approach to balloon valvotomy and atrial septal defect and patent arterial duct closure might be realistic, using a simple C-arm for imaging.

Where funding and the ability to provide good ongoing follow up are limited, surgical tactics should aim to be definitive rather than palliative, an approach that works well for relatively simple conditions like ventricular septal defect and tetralogy of Fallot. Likewise the arterial switch procedure for transposition of the great arteries leaves the majority free of significant residual lesions, although they still require follow up to watch for the development of aortic root dilation and aortic regurgitation. Similarly it is preferable to develop surgical techniques for valve repair, rather than replacement, thus avoiding the need for repeat valve replacement or warfarin anticoagulation. Difficult decisions have to be faced: careful consideration is needed before embarking on a surgical programme for patients with a single ventricle, including hypoplastic left heart syndrome. Healthcare professionals must clearly communicate to the families of babies with a single ventricle that a series of operations will be required in the 1st years of life, needing prolonged inpatient stays and frequent outpatient visits. The Fontan patient is likely to need long term medicines, and after a period of relatively infrequent medical input in young adulthood, will need more intensive follow up and medication and will likely face a decline in function by their 30s.

Is access to a scarce resource like MRI essential to assess right ventricular size and function? The answer is no, although it does help to provide objective and reproducible measurements. Serial assessments by a skilled clinician and echocardiographer will allow sensible planning of the timing of further interventions. Can the adult with congenital heart disease get to see their specialist adult congenital cardiologist? In a small heavily populated country like the UK, the cardiologist may have to travel 50 miles to run an effective outreach clinic, but in South Africa the distances may be prohibitive and patients easily lost to follow up. The "Best Buy" approach should permit improvements in national congenital heart disease management. Adult congenital heart disease patients should be included in cardiac surveillance systems, and links between a designated cardiologist or other health care professional in a district based hospital and the specialist adult congenital cardiologist in the specialist centre should allow a high standard of care without the need for doctor or patient to travel long, expensive distances. This hub and spoke model of care was recommended for UK patients with congenital heart disease in 2006,[4] and the ongoing English National Review of Congenital Cardiac Services[5] continues to develop it.

Despite the best care, many patients with congenital heart disease face premature deterioration and death. The need to develop palliative and end of life services – that can run in parallel with exploration of interventions like heart transplantation – is explored in Bowater's article in this edition.

Whichever health economy we work in, learning more about the natural and unnatural history and life expectancy of our patients with complex congenital heart disease will enable us to plan their future needs. We should develop low cost structures that allow effective surveillance and planning, from the timing of interventions to preserve ventricular function, to symptom control and quality of life.

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