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Editorial

A New Era in the Treatment of Congenital Heart Disease

The spectrum of congenital heart disease ranges from simple lesions like a persistent arterial duct to lethal conditions like the hypoplastic left heart syndrome. Conventional treatment in the past five decades has been surgery.

New Trends in Paediatric Cardiac Surgery

Today, surgical mortality and morbidity for simple lesions are extremely low. Complex cardiac surgery, like the arterial switch operation in the neonate, Fontan operations and repair of complex pulmonary atresias, when performed in well-equipped and experienced centres, can also achieve excellent short- and long-term outcomes.

The surgical success are attributed to improvement in surgical techniques and cardiopulmonary bypass, better myocardial protection and advances in post-operative intensive care, such as the use of inhaled nitric oxide in post-operative pulmonary hypertensive crisis and mechanical circulatory support like extracorporeal membrane oxygenation and ventricular assist device to treat post-operative low cardiac output syndrome.

The new trend in surgical treatment is five-fold. Firstly, a move towards early operation for some cardiac lesions has been advocated. Good results have been achieved in neonatal repair of Tetralogy of Fallot and cavopulmonary connections in univentricular repair of complex cardiac lesions in infancy. The rationale for early operation is to preserve ventricular function and promote growth of young children. Secondly, complicated cardiac operations on severe cardiac lesions, like the hypoplastic left heart syndrome, that are often not treated previously, have been increasingly performed. Thirdly, increasing use of homografts and various types of valved conduits for connection of ventricle to pulmonary arteries has resulted in better preservation of ventricular function and reduced associated morbidity.[1] Quality of life was excellent for most patients but future conduit replacement is usually required. The quest continues for an optimal conduit. Tissue engineering may cast hope for producing better valved conduit with a longer life span for human use in the future. Fourthly, many innovative new surgical procedures have been introduced in recent years. In hypoplastic left heart syndrome, Sano et al[2] modified the Norwood stage I operation by using a right ventricular to pulmonary artery shunt instead of the standard aortopulmonary shunt to achieve a more favourable post-operative haemodynamics and improve operative survival (92% versus 70%). Many centres have switched to this

modification for first stage palliation of this complex condition. Similarly, a radical new approach for an early one-stage repair instead of the conventional staged approach for the complex pulmonary atresia with ventricle septal defect and multiple aortopulmonary collaterals has achieved initial success.[3] Fifthly, with increasing survival of patients with congenital heart disease, a growing population of patients who require reoperation has emerged. Pulmonary regurgitation in post-operative tetralogy of Fallot causing right ventricular dysfunction, failure and arrhythmia is now a major concern and this large group of patients will need pulmonary valve replacement. This situation will certainly pose new challenges to surgeons and may lead to an increase in demand for heart transplantation in the years to come.

Paediatric Interventional Cardiology

Despite the surgical success and advancements, its traumatic nature and morbidity associated with cardiopulmonary bypass and various postoperative complications have been well documented. Furthermore, adverse neuro-developmental sequelae in children undergoing cardiopulmonary bypass at an early age is also a major concern. It is with this background that a new trend develops in the past two decades toward use of less invasive, non-surgical approach to the treatment of congenital heart disease to minimise pain, complications and costs of cardiac surgery. Paediatric interventional or therapeutic catheterisations which are catheter-based techniques have developed rapidly and play an increasing role in the treatment of congenital heart disease.

Advances in medical technology led to the production of various balloon catheters, devices and equipments, making a lot of transcatheter interventional procedures of congenital heart disease possible. To date, interventional procedures, including balloon dilation of valvar pulmonary and aortic stenosis, native coarctation of aorta, post-operative re-coarctation, and branch pulmonary artery stenoses; device occlusion of persistent arterial duct and secundum atrial septal defect, have all become the standard treatment and achieved comparable, if not better, results compared with surgery.

Other procedures like balloon dilation and stent implantation in various vascular stenoses or device occlusion of residual cardiac defects and unwanted vessels like shunts and collateral vessels have also become the preferred method of treatment. Currently, interventional procedures have been widely incorporated into the management strategy of complex congenital heart diseases.

It is worth mentioning that while some neonatal congenital heart diseases definitely require surgical repair, many others can be treated more effectively and safely by interventional cardiac catheterisation which causes less injury to the immature organs of the newborn. This is made possible by miniaturisation of catheters and devices. Balloon valvuloplasty for critical pulmonary stenosis and critical aortic stenosis in the neonate is proven to be more effective and safe than surgery. Similarly, in pulmonary atresia with intact ventricular septum, perforation of the atretic pulmonary valve by radiofrequency or laser energy followed by balloon dilation to establish continuity between the right ventricle and the pulmonary arteries has become the preferred initial treatment in the management

of this complex neonatal cardiac lesion.

In the recent few years, paediatric interventional cardiology continues to expand and new procedures are introduced at a rapid pace. Percutaneous transcatheter closure of perimembranous ventricular septal defect with a specially designed occluding device[4] has been successful. This procedure has the potential to benefit a large number of children. Percutaneous pulmonary valve replacement using a percutaneous stent-based expandable pulmonary valve is now possible.[5] Likewise, percutaneous transcatheter aortic valve replacement, is also under clinical trials. A new method of using transcatheter techniques instead of surgery to complete the second stage of the Fontan circuit in univentricular repair for complex congenital heart disease has also been introduced.[6] It is expected that many more innovative procedures will be introduced in the future, making treatment of congenital heart disease less invasive.

Interventional Electrophysiology

Cardiac dysrrhythmias may be associated with congenital heart disease and occur in its postoperative period or in a structurally normal heart as a primary disease. Traditional treatment with anti-arrhythmic drugs alone is often limited by the inability to achieve a full control, the side-effects of drugs and patients' compliance. In the past two decades, interventional electrophysiology, i.e. catheter-based treatment of cardiac arrhythmias, has developed rapidly and has gained an important role in the management of cardiac arrhythmias. Pacemaker implantation for complete heart block and sick sinus syndrome is now usually performed in the cardiac catheterisation laboratory using the transvenous route instead of surgical epicardial placement.

The greatest advance in the past decade is the introduction of radiofrequency catheter ablation to cure tachycardia that are related to accessory pathways like the Wolff-Parkinson-White syndrome or atrioventricular nodal re-entrant tachycardia. More recently, the use of cryoablation instead of radiofrequency ablation allows the technique to be performed in a more controlled and reversible manner, thus reducing the risk of procedure-induced heart block.

Another important development is the implantable cardioverter-defibrillator, which has now been miniaturised enough to allow transvenous implantation in children at risk of fatal ventricular tachyarrhythmias, as in long QT syndrome or hypertrophic cardiomyopathy. In congestive heart failure with ventricular asynchrony, the adult experience of improving cardiac output by synchronisation of both ventricles with pacing may be applied to end-stage heart failure in congenital heart disease and cardiomyopathy in children.

Advantages and Limitations of Interventional Cardiology

The major advantages of interventional cardiac catheterisation over surgery include : it is less traumatic and causes less pain and discomfort to patients, shorter hospital stay, much faster recovery, cost-effective, and avoidance of a permanent scar on the chest wall. However, these procedures have their own set of complications: vessel rupture, device embolisation, arterial or venous thrombosis and radiation exposure. These complications are usually uncommon and the overall risks have been proven to be lower than that of cardiac surgery. They can be reduced by careful patient and device selection, meticulous techniques, low dose pulse fluoroscopy and most importantly, operator experience.

The Hybrid Approach

Management strategies for complex congenital cardiac lesions are complicated and often challenging. Both cardiac surgeons and paediatric interventional cardiologists, standing alone, may encounter problems while providing treatment. There is a growing need of combining both surgical and catheter-based intervention to achieve the best treatment outcome. The "hybrid" approach involves a close collaboration between paediatric cardiac surgeon and interventional cardiologist to perform cardiac procedures together. Examples are intraoperative stenting, perventricular device closure of ventricular septal defect without cardiopulmonary bypass, balloon dilation or occlusion of vascular structures during surgical repair and the percutaneous approach to Fontan palliations. With the increasing complexity of treating congenital heart disease, the hybrid approach will be playing a more important role.

The Future

The treatment of congenital heart disease has changed from being primarily surgical to a combination of

interventional cardiology and surgery. Paediatric interventional cardiology is a rapidly advancing field. It has not only replaced surgery for treatment of a large number of defects but also provided an additional and complimentary treatment to surgery for complex cardiac lesions and complicated post-operative residual lesions. Congenital heart surgeons may apparently be operating less on simple lesions or conditions that are better dealt with by interventional procedures, their workload, however, has never decreased. They are increasingly operating on more complex cardiac conditions, which previously would not have been treated. With the increase in survival of congenital heart disease, cardiac surgeons are also facing an increasing demand for reoperations on survivors. Both cardiac surgery and interventional cardiac catheterisation have advantages, disadvantages and inherent limitations but we are expecting important progress in both fields in the years ahead. Paediatric interventional cardiologist and cardiac surgeons will have more opportunities to cooperate and work together either in the operating theatre or the cardiac catheterisation laboratory in the hybrid approach to treat congenital heart disease. We are entering a new era of management of congenital heart disease in which judicious application of different treatment modalities is important to improve outcome and quality of life.

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