



DEVELOPMENT OF PAEDIATRIC CARDIOLOGY IN HONG KONG - THE PAST, PRESENT AND THE FUTURE

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Four decades ago, paediatric cardiological service began to develop in Grantham Hospital (GH), which was initially designated as a hospital for treating tuberculosis in 1957. In the late 1960s, the hospital was given a second major task – to develop into a cardiac centre providing cardiological service and cardiac surgery for both adults and children.

The Past

The first closed heart operation was performed in 1967. It was surgical closure of a persistent ductus arteriosus. In 1968, open heart surgery, i.e. operations using cardiopulmonary bypass, was performed in GH. In the first year, a total of 34 cases of cardiac surgery, including congenital heart disease was performed with cardiopulmonary bypass.

In this early period, cardiac operations were concentrated only on older children and adults. After accumulation of experience for several years, attempts to operate on neonates and young infants started from 1971. These operations included atrial septectomy on a ten-day-old neonate with transposition of the great arteries in 1971, repair of truncus arteriosus on a two-month-old infant in 1973 and closure of atrial septal defect in an eight-month-old infant in early 1975. Unfortunately, there were no survivors from these early attempts. It was not until September 1975 that the first infant survived an open heart surgery for closure of ventricular septal defect and atrial septal defect.

In the early years of development of paediatric cardiac service, the majority of cardiac catheterizations in children were performed in Queen Mary Hospital. Surgical cases were then transferred to GH. It was the adult cardiologists who provided care, including post-operative intensive care, to children with heart disease in GH. In 1979, a consultant paediatric cardiologist was appointed to

establish a department of paediatric cardiology in the hospital to take care of children with heart disease. At about the same time, a new cardiac catheterization laboratory was built to improve the diagnostic facilities. Later a few more paediatric cardiologists from the University Paediatric Unit, University of Hong Kong, joined GH. After enhancement of staff, GH gradually developed into a paediatric cardiac centre receiving referrals from the whole of Hong Kong and from Macau as well. Indeed, many children with milder forms of heart disease are treated in all other major hospitals in the territory. Neonates born with more serious heart diseases are often stabilized in individual hospitals before transfer to GH. More and more paediatricians became interested in paediatric cardiology. Thanks to all paediatricians who provided care for our children with heart disease.

The Present

In Hong Kong, we are now able to provide a comprehensive paediatric cardiac service at GH. The service comprises of in-patient, day-patient and outpatient care. The combined adult and paediatric cardiac intensive care unit provides care for post-operative patients. A close link and direct referral system has been established with hospitals and paediatricians in both the public and private sectors.

Clinical Service Re-engineering

In 1994, with the support of the Hospital Authority and community donations, a major re-engineering of clinical service was carried out in the Department of Paediatric Cardiology at GH. The aim was to reduce hospital admission and shorten hospital stay by providing ambulatory service through a day centre. In the day centre, pre-operative and pre-catheterization assessment of patients are then all done on a day basis. An early post-operative discharge policy was adopted and continued post-operative care was provided in the day centre, thus,

shortening children's hospital stay by 5-7 days. At the same time, a high dependency unit was established to relieve the over-crowdedness in the cardiac intensive care unit so that more cardiac operations can be performed.

In 1995, with the addition of one more catheterization laboratory in GH, a catheterization laboratory dedicated for children was materialized. Major advances in diagnosis and treatment of heart disease in children have been achieved since then.

Diagnosis of Congenital Heart Disease

In the early 1980's, the widespread use of the echocardiography has provided a very useful and non-invasive diagnostic tool to investigate heart disease in children. It gradually replaces diagnostic cardiac catheterization as the mainstay of cardiac investigation in children. Nowadays, echocardiography can provide detail anatomical and haemodynamic data necessary for decision making for both the paediatric cardiologist and cardiac surgeon.

Surgical Treatment of Congenital Heart Disease

Surgical treatment has been the conventional treatment of congenital heart disease. In the early years, while simple congenital heart disease could be effectively corrected, the mortality and morbidity of surgery for complicated congenital heart disease remained significant. With improvements in surgical techniques, accumulation of more experience, improved techniques of cardiopulmonary bypass, better myocardial protection and improvements in cardiac intensive care, significant improvement in surgical outcomes even for the more complicated heart disease has been achieved. Today, surgical mortality and morbidity for simple cardiac defects like ventricular septal defects, atrial septal defects and persistent ductus arteriosus are extremely low. Outcomes for cardiac defects of moderate degree of severity like coarctation of aorta, atrioventricular septal defects and tetralogy of Fallot are also extremely good. Even for complex cardiac surgery, like the arterial switch operation for transposition of the great arteries in the neonate, Fontan operations for univentricular heart and repair of complex pulmonary atresias, satisfactory short- and long-term outcomes can also be achieved.

Major advances and new trends in surgical treatment have also occurred. Firstly, a move towards early operation has been advocated. Good results have been reported 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 serious cardiac lesions, like the hypoplastic left heart syndrome that are often not treated previously, have been increasingly performed. Thirdly, use of homografts and various types of valved conduits for connection of ventricles to pulmonary arteries has resulted in better preservation of ventricular function and reduced associated morbidity. Quality of life is excellent for most patients albeit future conduit replacement is usually required. Fourthly, innovative new surgical procedures have been introduced in recent years to improve outcomes. 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 ahead. In Hong Kong, we are keeping up with these trends though in some instances, modifications of strategy are needed to suit the local situation.

Paediatric Interventional Cardiology

Despite surgical success and advancement, its traumatic nature and morbidity resulting from cardiopulmonary bypass and various post-operative 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 the use of less invasive, non-surgical approach to treat congenital heart disease to minimize pain, complications and costs of cardiac surgery. 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 even in young infants. Paediatric interventional or therapeutic catheterizations which are catheter-based techniques have therefore developed rapidly and play an increasing role in the treatment of congenital heart disease. This treatment modality was first introduced to Hong Kong in 1986. The first procedure performed at GH was balloon dilation of a stenotic pulmonary valve. Thereafter, the application has been extended to many other cardiac lesions.

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 replaced surgery and become the standard treatment. They are able to achieve 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 because surgical treatment may be associated with technical difficulties, higher risks or inaccessibility to the site of lesions. 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 catheterization which causes less injury to the immature organs of the newborn. Balloon valvuloplasty for critical pulmonary stenosis and critical aortic stenosis in the neonate is proven to be more effective and safer than surgery. Similarly, in pulmonary atresia with intact ventricular septum, perforation of the atretic pulmonary valve by radiofrequency (an electric 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.

The major advantages of interventional cardiac catheterization over surgery include : being less traumatic and causes less pain and discomfort to children, shorter hospital stay, much faster recovery, cost-effectiveness, and avoidance of a permanent scar on the chest wall. However, these procedures have their own set of complications : vessel rupture, device embolization, 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.

At present, surgical treatment accounts for 60% whereas interventional catheterization accounts for 40% of all intervention in congenital heart disease locally.

Paediatric Electrophysiology and Interventional Procedures to Treat Arrhythmias

Cardiac dysrhythmias 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, electrophysiological studies and interventional

electrophysiology, i.e. catheter-based investigations and treatment, respectively, of cardiac arrhythmias, has developed rapidly and has gained an important role in the management of cardiac arrhythmias.

In Hong Kong, a well-established service of electrophysiological studies and intervention for children was not available until mid 1990s when a paediatric cardiologist returned from overseas training in this specialized field.

Pacemaker implantation for complete heart block and sick sinus syndrome is now usually performed in the cardiac catheterization 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 use of implantable cardioverter-defibrillator, which has now been miniaturized enough to allow transvenous implantation in children at risk of fatal ventricular tachyarrhythmias such as long QT syndrome or hypertrophic cardiomyopathy. In congestive heart failure with ventricular asynchrony, the adult experience of improving cardiac output by synchronization of both ventricles with pacing may be applied to end-stage heart failure in congenital heart disease and cardiomyopathy in children.

Paediatric Cardiac Intensive Care

Advances in cardiac intensive care have significant contributions to improve surgical outcomes and reduction of mortality and morbidity. The use of inhaled nitric oxide since 1999 in GH to treat post-operative pulmonary hypertensive crisis has greatly reduced mortality from this deadly condition, reduced ventilator days, and shortened stay in the intensive care unit.

In recent years, the development of mechanical circulatory support like extracorporeal membrane oxygenation (ECMO) and ventricular assist device (VAD) in GH further saves children's lives from severe post-operative low cardiac output syndrome and intractable heart failure from fulminant myocarditis or cardiomyopathy that are not responsive to conventional inotropic therapy.

Fetal Echocardiography and Prenatal Counseling

With the use of ultrasound in prenatal fetal screening, fetal cardiac abnormalities are increasingly detected by obstetricians. Paediatric cardiologists are frequently asked by obstetricians to help make diagnosis of the more complex cardiac abnormalities and to provide counseling to the parents. In 2004, a fetal echocardiography and counseling clinic has been established at GH and the service is welcomed by obstetricians and parents.

Education and Research

Over the years, more local paediatricians develop interest in paediatric cardiology. Many of them undertake a certain period of postgraduate training in this field and are providing an excellent care to children with heart disease in all hospitals in Hong Kong. In the past two decades, a total of 50 local and overseas paediatricians have been trained in GH. Overseas trainees came from the Philippines, Mainland China and Macau. However, a structured training programme for paediatric cardiologists has not been established in Hong Kong yet.

Since its establishment, high quality research projects with publications in referred journals have been done through the joint effort of the academic and other staff at the GH and other hospitals. Most of these researches are clinical in nature. Research in basic scientific and genetic aspects are still in development.

The Future

The future development of paediatric cardiology in Hong Kong is challenging. We are moving towards using more non-invasive cardiac imaging. This includes use of magnetic resonance (MR) imaging / angiography and high resolution computerized tomography to define not only anatomical details but also certain functional aspects of the heart. They are useful in long-term follow-up of patients. Magnetic resonance-guided cardiac catheterization laboratory may soon be developed. This would help reduce irradiation to both patients and operators but MR-compatible equipments and catheters need to be made available first.

Where treatment of congenital heart disease is concerned, the use of more aggressive and radical surgery performing at a younger age will continue with a view to preserve cardiac function. Tissue engineering may cast hope for producing optimal heart valves and valved conduits for repair of complex heart disease. On the other hand, interventional catheter therapy will continue to be a rapidly advancing field. New catheter procedures will emerge and keep on replacing surgery for treatment of more cardiac defects. Congenital heart surgeons may apparently be operating less on simple lesions or lesions that can be better dealt with by interventional catheter procedures, their

workload, however, has not decreased as they will be operating on the most complex cardiac lesions that would not have been treated previously. Long-term survival in congenital heart disease will also generate a new demand for reoperations.

In recent years, a new strategy called the “hybrid approach” is developed to manage complex congenital heart disease that are difficult to be managed by either surgery or catheter-based techniques alone. In this approach, surgical and catheter-based intervention are combined in the same setting whereby cardiac surgeon may help expose the anatomical structures for the paediatric interventional cardiologist to perform catheter procedures such as closing intra-cardiac defects or occluding unwanted structures in the operating theatre. Alternatively, the paediatric interventional cardiologist may perform some catheter procedures to facilitate difficult cardiac operations. A combined suite where both cardiac surgery and interventional procedures can be performed at the same time is therefore needed in the future to achieve the best therapeutic effect.

As survival of congenital heart disease increase, there will be a growing need to develop an adult (or grown-up) congenital heart program in Hong Kong to provide not only medical care but also counseling service for education, employment, marriage and pregnancy for adults with congenital heart disease. The participation and collaboration of adult cardiologists is extremely important in this regard.

All the aforementioned future development will have significant impact on the future service organization and provision of care in paediatric cardiology. They call for more specialization, a finer division of labour, involvement of multiple disciplines and a centre-based service. A structured training program is necessary for training the future paediatric cardiologists to ensure supply of well-qualified experts, to ensure succession and make the system sustainable in the long term. Similarly, there is a need for more specialized paediatric cardiac nurses to safeguard the quality of care. The existing service has to continue to evolve into a service model in which all parties can participate with partnership and ownership to serve children and adolescents with heart disease in Hong Kong. While we thank all our predecessors for laying down a good foundation that lead to the present achievements in paediatric cardiology, the medical and nursing profession should continue to cooperate to provide a modern and good service to children and adolescents with heart disease.

Further reference

Useful website (community resource from the Children’s Heart Foundation): <http://www.childheart.org.hk>