

Paediatric Cardiology in Singapore – 1978 to 2008[†]

William CL Yip,^{1,2,3} MD (Singapore), FRCP (Edin), FAMS

This article marks Congenital Heart Defect Awareness Week, 7 to 14 February 2009 (<http://tchin.org/aware/index.htm>). The title, “Paediatric Cardiology in Singapore - 1978 to 2008” reflects the most exciting phase in the development of paediatric cardiology in Singapore. Congenital heart defect (CHD), with an incidence of 1 in 100 live births, is the most important and frequent congenital malformation, causing much morbidity and mortality in infants, children, and even in adults.^{1,2} This article highlights some advances in the science, and the changing patterns in the practice, of paediatric cardiology in Singapore.

Diagnostic Techniques

The clinical presentation of CHD is protean, including cardiac murmurs, abnormal heart sounds, defects in pulses and rhythm, cyanosis, failure to thrive, poor effort tolerance, diaphoresis in infants, chest pain, fainting spells, sudden death during exercise, and the incidental discovery of abnormalities on chest radiography, electrocardiogram (ECG), or fetal sonography.³ In a significant number of patients, the CHD produces no symptoms, and only becomes evident through routine screening.^{4,5}

Although certain acyanotic CHDs may be confidently diagnosed by clinical examination by experienced cardiologists, the exact morphological abnormalities, and functional impairments caused by these lesions, and most cyanotic CHDs, require more sophisticated evaluation.⁶ Before 1976, the only diagnostic modality available, other than the simple ECG and chest X-ray, was cardiac catheterisation, using single plane angiography. M-Mode and B-Mode echocardiography for children was first introduced in Singapore in 1976 at the then University Department of Paediatrics at Mistri Wing, Singapore General Hospital. Cross-sectional (two-dimensional) echocardiography was first available in 1980 from a research grant from the former University of Singapore.^{7,8} Soon, other modalities of echocardiography, namely Doppler interrogation (1984), colour flow mapping (1987),^{9,10} transoesophageal imaging (1988) and three-dimensional real-time echocardiography (2007) were added. Cross-

sectional imaging permits the systematic, sequential and segmental analysis of CHD, using an imaging morphological approach to identify the cardiac segments, ascertain the segmental connections, categorise the defects, and to measure the chamber and vessel sizes. Other echo modalities allow haemodynamic assessment, delineation of abnormal flow pattern, and the quantification of cardiac function. Indeed, echocardiography has replaced cardiac catheterisation as the most important initial diagnostic tool for children with CHD. Singapore was amongst the first countries in the southern hemisphere to send patients for cardiac surgery without cardiac catheterisation in the early 1980s.¹¹

Advances in material science, imaging technology and computer science brought in more sophisticated instrumentation for imaging and non-invasive assessment of CHD from the late 1980s. Bi-plane cine-angiography with movable C- or U-arms allows appropriate projections of the tomographic planes to cater for the special anatomy of the different components of the heart in optimum spatial orientation. More refined instruments, such as smaller introducer sets, special catheters and guide-wires, all facilitate access, navigation within the heart, and safer angiography.

Multi-slice computerised tomography (MSCT) is the later addition of non-invasive imaging that permits the display of exquisite details of cardiac defects, with life-like presentation of reconstructed three-dimensional anatomy. It is especially useful in older patients with complex CHD when echocardiographic imaging may be suboptimal.¹² Magnetic resonance imaging (MRI) provides additional information on cardiac function and haemodynamics. The advantage of MRI over MSCT is that it is radiation-free, although it is more costly and more time consuming, and in young children, general anaesthesia is required to prevent body movement causing artifacts. Radionuclide imaging is also useful in some patients who require assessment of right and left ventricular function and of coronary ischaemia.

Other non invasive tools, including Holter monitoring,^{13,14} high-resolution ECG,¹⁵ and cardiopulmonary exercise stress

¹ Consultant Paediatric Cardiologist, Singapore Baby & Child Clinic, Gleneagles Medical Centre, Singapore

² Adjunct Associate Professor, National University of Singapore, Singapore

³ Visiting Consultant, National University Hospital, Singapore

Address for Correspondence: Dr William Yip Chin Ling, Singapore Baby & Child Clinic, 6 Napier Road, #07-01/03, Gleneagles Medical Centre, Singapore 258499.

[†] Congenital Heart Defect Awareness Week, 7-14 February 2009

testing, are routinely used to provide diagnostic information on rhythm abnormalities and assessment of functional capacity before and after surgery or interventional therapy.

Treatment Modalities

Surgery

Surgery was the only modality of treatment for CHD in Singapore before 1978. In the 1970s, only simple operations, like ligation of patent arterial duct (PAD) and closure of secundum atrial septal defect (ASD), were done. Many babies with more complex lesions were doomed. Operations, more often palliative, were carried out in older children, after diagnostic cardiac catheterisation. The science of surgery progressed, with more effective cardiopulmonary bypass and, in parallel, the local surgeons were better trained in overseas major cardiac centres. Thus, the more complex CHD, like Fallot's tetralogy, transposition of great arteries (TGA) and total anomalous pulmonary venous return, were operated on from the mid-1980s, in younger children, infants, and newborns, often after confirmation by echocardiography alone.

The results of cardiac surgery for CHD continue to improve. The standard is comparable to that of many large centres elsewhere.^{16,17} The only limitation is cardiopulmonary transplantation, which is not available for children in Singapore.

Transcatheter Interventions

Balloon atrial septostomy was the only palliative transcatheter intervention for TGA from 1978 to 1987. This procedure improves the survival of infants with TGA before definitive surgical correction. The first therapeutic transcatheter intervention for pulmonary valvar stenosis (PVS) using balloon valvuloplasty was introduced in a 3-year-old boy in 1987 at the National University Hospital.¹⁸ This opened the era of catheter interventional therapy for patients with CHD in both children and adults. Since then, balloon valvuloplasty has replaced surgery in patients with PVS.¹⁹ The same technique was later employed to treat critical PVS in infants, aortic valve stenosis, and re-coarctation of the aorta.

The next break-through was the introduction of transcatheter occlusion of PAD, using the Rashkind umbrella device (RUD) in 1992 at Gleneagles Hospital.²⁰ Later, after the discontinuation of RUD, Cook's detachable coils for smaller PAD, and the Amplatzer duct occluder for bigger PAD, were used from 1997 and 2000 respectively. After the neonatal period, almost all PAD may be closed by an occlusive device. Singapore participated in the first international trial to close secundum ASD using the Amplatzer septal occluder (ASO) in 1997 at Gleneagles

Hospital.²¹ The result was excellent.²²

Since then, about 80% of secundum ASDs in children and adults can be closed using the ASO. The remaining 20%, with inadequate postero-inferior rim and inadequate septal length, require surgical closure. Other new transcatheter catheter treatments included stenting of peripheral pulmonary artery stenosis, device closure of coronary arteriovenous fistula, device occlusion of aortopulmonary collaterals, stenting of re-coarctation and, in 2004, transcatheter closure of perimembranous ventricular septal defect (VSD) at Gleneagles Hospital when the asymmetrical Amplatzer membranous VSD occluder became commercially available.²³

Patient and Service Load

In the early 1970s, local patients with CHD were mainly referred by general practitioners, paediatricians, polyclinic doctors, school health medical officers and neonatologists. From 1980 onwards, paediatric cardiologists began to see more patients from the surrounding countries. The trend has continued, with Singapore becoming a regional medical hub. Secondly, the establishment from the early 1980s of a fetal echocardiography service, collaborating with obstetricians both in the public institutions and private sector, permitted the early detection of CHD, planned delivery, and in timely intervention.²⁴⁻²⁶

Hence, with improved surgical expertise and a fetal echocardiography service, the demography of local patients changed. The age of detection and operation for patients with CHD has fallen, and more complex lesions are surgically treated. As a result, infants and children who survive the corrective or palliative operations and interventional procedures accumulate, many reaching adulthood. Paediatric cardiologists and adult cardiologists who have special training will have to handle these adult CHD patients, with problems such as postoperative residua, dysrhythmias, cardiac functional impairment, monitoring of coagulation profile in patients with intracardiac prosthesis, and reproductive risk.

In addition, patients with Marfan syndrome have in later years a significant risk of severe mitral regurgitation and aortic rupture, and so require life-long surveillance. Genetic counselling is also important because of different recurrence risks in different patients. Finally, sudden cardiac death during exercise, due to undetected significant cardiac lesions in children and young adults is a tragedy that attracted media attention recently. The issues of pre-participation screening, eg physician expertise, screening methods, and financial resources, are not resolved, because so many individuals regularly take exercise, often in mass events.²⁷

Paediatric cardiology has undergone tremendous changes in the past 3 decades. While there were marked improvements in diagnostic capability and skill, and advances in surgical expertise and in transcatheter interventional therapy, new challenges face paediatric cardiologists as we deal with more complex CHD at a younger age. We strive to better our results, handle problems in the enlarging pool of CHD survivors, improve their quality of life, and prevent sudden death in young individuals who exercise vigorously. We hope that a better understanding of the morphogenesis and development of the electrical system²⁸ of the developing heart can prevent some types of CHD and institute timely treatment.²⁹ Singapore needs more paediatric cardiologists, with adequate support, to achieve these objectives.

REFERENCES

1. Yip WCL, Tay JSH, Tan NC. Congenital heart disease in Singapore present problems and future perspectives. *Singapore Med J* 1982;23:133-9.
2. Wong JCL, Tay JSH, Yip WCL, Wong HB. A 20-year review (1966-1985) of hospital mortality in the University Department of Paediatrics, Singapore. *J Singapore Paediatr Soc* 1987;29(Suppl):73-9.
3. Yip WCL. Congenital heart disease in Southeast Asian infants and children (Part I) clinical diagnostic evaluation. *J Paediatr Obstet Gynaecol* 1987;13:5-12.
4. Chan KY, Yip WCL, Tay JSH, Rajan U, Ng MP. Results of screening for cardiac diseases among school children in Singapore. *J Singapore Paediatr Soc* 1988;30:125-8.
5. Chan KY, Loke KY, Yip WCL, Tay JSH. Cardiac arrhythmias in paediatric practice. *J Singapore Paediatr Soc* 1989;31:32-7.
6. Yip WCL. Congenital heart disease in Southeast Asian infants and children (Part II) diagnostic evaluation by echocardiography and cardiac catheterization. *J Paediatr Obstet Gynaecol* 1987;13:10-7.
7. Tay JSH, Yip WCL, Wong HB. Two dimensional echocardiography. 1. Pre-operative assessment. *J Singapore Paediatr Soc* 1985;27:172-82.
8. Tay JSH, Yip WCL, Wong HB. Two dimensional echocardiography. 2. Post-operative evaluation. *J Singapore Paediatr Soc* 1986;28:79-84.
9. Yip WCL, Ho TF, Tay JSH, Lee CN, Chan KY, Wong ML, Wong JCL. Assessment of ventricular septal defect by colour flow mapping. *Ann Acad Med Singapore* 1991;20:303-7.
10. Yip WCL, Ho TF, Chan KY, Ong KK, Koh BC. Comparison of spontaneous closure rate in children with different types of ventricular septal defect. Ninth Asian Congress of Paediatrics, 1997, Hong Kong. *J Paediatr Child Health* 1997;33(suppl 1):S41.
11. Yip WCL, Tay JSH, Lee CN, Ong KK, Chan KY, Ng CK, et al. Cardiac surgery without invasive investigation in infants and children: a team decision based on clinical and echocardiographic information. Proceedings of the Eighth Asian Congress on Thoracic and Cardiovascular Surgery; Singapore 1987:78.
12. Yip WCL. The role of multislice compute tomography in the diagnosis of congenital heart disease. The First Asia-Pacific Congress of Pediatric Cardiology and Cardiac Surgery; 2006 Nov 1-4; Bangkok, Thailand. Thailand: 2006:74-5.
13. Han L, Ho TF, Yip WCL, Chan KY. Heart rate variability of children with mitral valve prolapse. *J Electrocardiol* 2000;33:219-4.
14. Ho TF, Han L, Yip WCL, Chan KY. Heart rate variability of children after repair of TOF. *European pacing, Arrhythmias and Cardiac Electrophysiology* 2002;3(Suppl):A153.
15. Ho TF, Yip WCL, Chan KY. Age and gender specific criteria for signal averaged electrocardiographic parameters in healthy Chinese children. *Ann Noninvasive Electrocardiol* 1996;1:293-300.
16. Sim EKW, Grignani RT, Wong ML, Quek SC, Wong JCL, Yip WCL, et al. Outcome of surgical closure of doubly committed subarterial ventricular septal defect. *Ann Thoracic Surg* 1999;67:736-8.
17. Sim EKW, Grignani RT, Wong ML, Quek SC, Wong JCL, Yip WCL, et al. The influence of surgery on aortic valve prolapse and aortic regurgitation in doubly committed subarterial ventricular septal defect. *Am J Cardiol* 1999;84:1445-8.
18. Yip WCL, Tay JSH, Chan KY, Lee CN, Wong HB. Percutaneous transluminal balloon valvuloplasty for congenital pulmonary valve stenosis. The first case in Singapore and review of the literature. *J Singapore Paediatr Soc* 1987;29:144-9.
19. Yip WCL, Tay JSH, Chan KY, Wong JCL, Wong HB. Percutaneous transluminal balloon valvuloplasty. The treatment of choice for congenital valvar pulmonary stenosis. *Ann Acad Med Singapore* 1990;19:58-63.
20. Yip WCL. Transluminal devices for treatment of patent ductus arteriosus, atrial and ventricular septal defect. Proceedings of the Sixth Annual Scientific Meeting of the Singapore Cardiac Society; Singapore, 1993:12.
21. Chan KY, Yip WCL, Godman M. Transcatheter occlusion of atrial septal defects: An initial experience with the Amplatzer septal occluder. *J Paediatr Child Health* 1998;34:369-73.
22. Yip WCL, Ang P, Wu WX, Ho TF, Chan KY, Quek SW. Transcatheter closure of secundum atrial septal defect by Amplatzer Septal Occluder: a novel "in-site tulip-bud" method. *Catheter Cardiovasc Interv* 2008;71:S2.
23. Yip WCL, Amin Z, Chan KY, Hijazi ZM. Transcatheter closure of perimembranous ventricular septal defect by Amplatzer membranous VSD occluder in Singapore: Initial experience of the first series in Singapore. Mount Elizabeth Hospital Annual Scientific & Clinical Meeting (incorporating the 18th Annual Seminar & 5th Clinical Meeting); 2004 Nov 6-7; Sheraton Tower. Singapore, 2004:20.
24. Yip WCL. Foetal echocardiography. Proceedings of the Tenth ASEAN Congress of Cardiology; 1994; Bangkok, Thailand. Thailand, 1994:235.
25. Lim AM, Yip VCH, Wu WX, Chan KY, Yip WCL. Foetal echocardiography: indications and outcomes of at risk pregnancies in a tertiary referral centre. Abstracts of the 41st Singapore-Malaysia Congress of Medicine; 2007; Singapore. Singapore: Academy of Medicine Singapore, 2007:108.
26. Yip VCH, Lim AM, Wu WX, Chan KY, Yip WCL. Foetal echocardiography: accuracy and pitfalls in diagnosis. Abstracts of the 41st Singapore-Malaysia Congress of Medicine; 2007; Singapore. Singapore: Academy of Medicine Singapore, 2007:109.
27. Yip WCL. Pre-participation screening for sudden cardiac death in young athletes. "Evidence-based guidelines in clinical practice today". Joint Singapore Paediatric Congress & APAPARI Meeting 2008. Paediatrics, Child & Adolescent Health (Formerly: Singapore Paediatr J) 2008;48: S7-4.
28. Chia EL, Ho TF, Rauff M, Yip WCL. Cardiac time intervals of normal fetuses using noninvasive fetal electrocardiography. *Prenat Diagn* 2005;25:546-52.
29. Chia EL, Ho TF, Wong YC, Yip WCL. Ventricular bigeminy misdiagnosed as fetal bradycardia by cardiotocography – the value of non-invasive fetal electrocardiography. *J Perinat Med* 2004;32:532-4.