Pediatric Cardiology at Siriraj Hospital : Past, Present and Future

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Abstract

The incidence of congenital heart disease (CHD) at Siriraj Hospital for the year 2000 was 4.36 patients per 1,000 livebirths. Types of congenital heart diseases seen by the authors were VSD (18.3%), PDA (16.3%), ASD (16.3%), combined simple left to right shunt lesion (24.7%), tetralogy of Fallot (TF; 6%), D-TGA 2 per cent, other complex congenital heart 8 per cent.

Overall 3 out of 1,000 livebirths will have congenital heart disease that will require immediate intervention including cardiac catheterization and surgical intervention. At the same period of time an average of 750 new cases of children were referred annually for evaluation and treatment of cardiac related problems. Reports of acquired heart disease such as acute rheumatic fever, myocarditis, Kawasaki's disease and arrhythmia problem were summarized here. The Division of Pediatric Cardiology performs both diagnostic and intervention cardiac catheterization in almost 310 children and adults with congenital heart disease yearly. Out of this 35 per cent had interventional procedures including balloon valvuloplasty, balloon angioplasty and stenting, device closure of atrial septal defect and patent ductus arteriosus and radiofrequecy ablation of abnormal conduction pathway. Major progress has been made in pediatric cardiac imaging over the past two decades. At Her Majesty's Cardiac Center, cardiac MRI has been used to evaluate patients with congenital heart disease since June 2000. There were 146 congenital heart disease patients who had cardiac MRI performed for the evaluation of anatomy, function, left to right shunt, and flow regurgitation quantification.

Conclusion : Pediatric Cardiology practice has evolved over the past decade and expanded from clinical practice to therapeutic intervention procedures.

Key word : Pediatric Cardiology, Current Practice, Thailand

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The incidence of congenital heart disease (CHD) varies from 8 to 10 patients per 1,000 livebirths. At the Department of Pediatrics, Faculty of Medicine Siriraj Hospital the authors found that between January 1st 2000 and December 31st 2000, there were 11,245 livebirths, and out of this group of patients, 83 patients presented within 7 days of life with initial abnormal heart sound or other cardiac symptoms such as cyanosis and congestive heart failure[1]. There were 49 patients who were diagnosed to have congenital heart lesion from echocardiographic examination. The incidence of congenital heart disease was 4.36 patients per 1000 livebirths. The ratio of male : female was 0.88 to 1. This incidence appeared to be lower than previous reports [2-5]. The authors did not include patients who might present later than 7 days of life. Types of congenital heart diseases found are shown in Table 1[1].

There were 750 new cardiac patients age 0-13 years, who were diagnosed at Siriraj Hospital between January 1999 and December 1999[6]. Types of congenital heart diseases seen by the authors compared to other studies are shown in Table 2 and 3. In the present study, the five most common types were PDA (25.5%), VSD (24.3%), tetralogy of Fallot (TF; 19.2%), ASD (12.3%) and complex heart disease (8.6%). The five most common forms of CHD reported from the West included ventricular septal defect (VSD; 30.5%), atrial septal defect (ASD; 9.8%), patent ductus arteriosus (PDA; 9.7%), pulmonary stenosis (PS; 6.9%) and coarctation of aorta (CoA; 6.8%)[2-5]. Since Siriraj Hospital is one of the few cardiac centers in Thailand that provides open heart surgery and many patients with complex CHD have been referred. Acquired heart diseases seen by the authors are shown in Table 4. The management of these patients is shown in Table 5 and 6 respectively.

Table 1. Types of congenital heart disease (CHD) diagnosed within the first week of life in our study.

CHD	N	%
VSD	9	18.4
PDA	8	16.3
ASD	8	16.3
PDA/ASD	5	10.3
Common atrium/common ventricle	4	8.3
VSD/PDA/ASD	4	8.3
VSD/PDA	3	6.1
Corrected TGA	1	2.0
DTGA/VSD/ASD/PDA	1	2.0
PA/IVS	1	2.0
PA/VSD/PDA	1	2.0
Single ventricle/subvulvular PS/ PDA	1	2.0
TA/PA/ASD	1	2.0
TF	1	2.0
VSD/Coarctation of aorta /PDA	1	2.0
Total	49	100

PA/IVS = pulmonary atresia with intact ventricular septum;
TF = tetralogy of Fallot; VSD = ventricular septal defect;
ASD = atrial septal defect; PDA, = patent ductus arteriosus;
HLHS = hypoplastic left heart syndrome;
TGA = transposition of great arteries;
Truncus = truncus arteriosus; TA = tricuspid atresia;
PS = pulmonary stenosis; AS = aortic stenosis;
Ebstein = Ebstein anomaly

Interventional cardiac catheterization in congenital heart disease

As previously mentioned the frequency of congenital heart disease has been stable over the past decade. Overall, 3 out of 1,000 livebirths will have congenital heart disease that will require immediate intervention including cardiac catheterization and surgical intervention[7,8]. The first Pediatric cardiac catheterization for intervention was reported as early as 1966 from Rashkind WJ[9] by using balloon atrial septostomy in a newborn with complete transposition of the great arteries to improve mixing.

The major goal of cardiac catheterization in children is diagnostic and intervention. The Division of Pediatric Cardiology has performed both diagnostic and intervention cardiac catheterization for more than two decades. In 2001, almost 310 cardiac catheterizations were performed in both children and adults with congenital heart disease. Of these 110 patients (35.5%) had interventional procedures. The overall cardiac interventional catheterization procedures performed from 1995 to 2002 are shown in Fig. 1. Procedures were divided into four different groups as follows:

1. Balloon valvuloplasty and angioplasty.

Percutaneus pulmonary balloon valvuloplasty (PBPV) was initially performed in 1995[10-12]. Since then over 100 patients have had this procedure done. Their long-term results are excellent. The majority of patients did not require a second procedure for valvuloplasty. Laser-assist perforation of the pulmonary valve in six patients who had pulmonary artresia with intact ventricular septum was also performed[13]. Results of aortic balloon valvulo-

Table 2. Types of congenital heart diseases, ranked in frequency (432 cases).

CHD	N	%
PDA	110	25.5
VSD	105	24.3
Type I	8	7.6
II	71	67.6
III	18	17.1
IV	7	6.7
w/ AR	1	1.0
TF	83	19.2
Pink TF	6	7.2
w/ PA	3	3.6
w/ coronary abn.	4	4.8
ASD	53	12.3
Primum	7	13.2
Secundum	44	83.0
Sinus venosus	1	1.9
Common atrium	1	1.9
Complex*	37	8.6
PS	34	7.9
Valvar	26	76.5
Critical PS	3	8.8
Supra PS	3	8.8
Sub PS	2	5.9
DORV	19	4.4
w/ subAo VSD	7	36.9
w/ subPA VSD	2	10.5
w/ dTGA	3	15.8
w/ dTGA, PS	2	10.5
w/ PS	5	26.3
dTGA	18	4.2
w/ IVS	3	16.7
w/ small VSD	3	16.7
w/ mod, large VSD	4	22.2
w/ VSD, PS	5	27.7
w/ VSD, PA	3	16.7
AVC	16	3.7
Transitional AVC	1	6.3
CAVC	14	87.4
Unbalanced AVC	1	6.3
Dextrocardia**	15	3.5
PA/VSD	15	3.5
Confluent PA's	10	66.7
Non confluent PA's	5	33.3
CoA	9	2.1
w/ hypoplastic Ao arch	2	22.2
Tricuspid atresia	9	2.1
w/ PS	6	66.7
w/ dTGA	2	22.2
w/ LTGA	1	11.1
Branch PS	7	1.6
AS	6	1.4
Bicuspid AV	6	1.4
Coronary AVF	6	1.4
Corrected TGA	5	1.2
DCRV	4	0.9
PAPVC	4	0.9
TAPVC (coronary sinus)	3	0.6
Cortriatriatum	3	0.6
HLHS	2	0.4
IAA (type A)	2	0.4
HRHS	1	0.2
LV-Ao tunnel	1	0.2
Truncus arteriosus	1	0.2
Total	574 lesions	

PDA = patent ductus arteriosus; VSD = ventricular septal defect; w/, with; AR = aortic regurgitation; TF = tetralogy of Fallot; PA = pulmonary atresia; abn = abnormal; ASD = atrial septal defect; complex = *complex congenital heart diseases including common ventricle, heterotaxy; PS = pulmonary stenosis; DORV = double outlet right ventricle; sub Ao = subaortic; sub PA = subpulmonary; dTGA = dextro-transposition of the great arteries; IVS = intact ventricular septum; mod = moderate; AVC = atrioventricular canal; CAVC = complete AVC; **dextrocardia including normal & structural heart defect; PA's = pulmonary arteries; CoA = coarctation of aorta; Ao aorta; LTGA = levo-TGA; AS = aortic stenosis; AV = aortic valve; AVF = arteriovenous fistula; DCRV = double-chambered right ventricle; PAPVC = partial anomalous pulmonary venous connection; TAPVC = total anomalous pulmonary venous connection; HLHS = hypoplastic left heart syndrome; IAA = interrupted aortic arch; HRHS = hypoplastic right heart syndrome; LV-AO = left ventricular-aortic

plasty have been less than optimum[14]. Since a substantial number of patients required a second operation. However, in selected cases the balloon valvuloplasty has become the choice for curative treatment. Other angioplasty procedures were balloon angioplasty and stenting of the pulmonary artery and coarctation of aorta[15,16].

2. Atrial septostomy and atrial septectomy.

One of the earliest procedures performed in congenital heart disease was to create an atrial communication using a balloon catheter which was performed in patients with complete transposition of the great arteries to improve mixing[9]. Over the past few years the authors have developed a technique to use as a palliative treatment for patients who require atrial communication such as patients with Eisenmenger complex. Transatrial septal puncture with balloon dilatation of the atrial septum to create a permanent interatrial communications has also been used.

Table 3. Percentage frequency of congenital heart diseases compared to the other studies[3].

	The present study (n = 432)	ASEAN (n = 10,412)	Taiwan (n = 5,389)	Boston (n = 10,624)	Toronto (n = 6,647)
VSD	24.3	33.5	31.2	19.4	25.0
TF	19.2	21.0	22.0	6.0	10.0
PDA	25.5	15.0	9.9	15.5	12.1
ASD	12.3	11.0	6.3	4.5	4.0
PS	7.9	6.7	8.0	7.5	8.5
TGA	4.2	3.6	5.6	4.7	5.4
AVC	3.7	2.5	-	-	-
PA	3.5	2.4	-	-	-
AS	1.4	1.3	1.1	5.5	5.4
CoA	2.1	1.2	1.5	8.1	5.6

Abbreviations are the same as in Table 2.

Table 4. Types of acquired heart diseases, ranked in frequency (37 cases).

AHD	N	%
Acute rheumatic fever	10	27.0
w/ MR	5	50.0
w/ AR	5	50.0
Pericarditis	9	24.4
Kawasaki disease	7	18.9
Bilateral aneurysm	2	28.6
LCA aneurysm	1	14.3
Ectasia LCA	1	14.3
Rheumatic heart disease	4	10.8
w/ MR	3	75.0
w/ AR	1	25.0
Myocardial disease	4	10.8
Dilated cardiomyopathy	2	50.0
Hypertrophic cardiomyopathy	2	50.0
Infective endocarditis	3	8.1

w/, with; MR = mitral regurgition; AR = aortic regurgitation; LCA = left coronary artery.

3. Vascular coil occlusion.

Coil spring developed as early as the 1970's was used as a occlusion instrument which could be delivered by transcatheter method. The authors have used the coil occlusion in over 90 patients with small PDA (<4 mm). Other indications include occlusion of the collateral artery from descending aorta or coronary arteriovenous fistula[17].

4. Transcatheter device closure of simple congenital heart lesion.

Atrial septal defect (ASD) was found in 10 per cent of congenital heart disease patients. In 1999, the authors reported the first series of patients who had transcatheter closure of ASD using the Amplatzer™ Septal Occluder[18,19]. The authors also compared the result of atrial septal defect closure using Amplatzer™ Septal Occluder with Surgery [20], and performed transcatheter closure of ASD in 80 out of 87 patients compared to 120 patients who had surgical procedure performed. The mean ASD diameter measured was 28.9±9.4 mm in the surgical group compared to 26±5.6 mm in the transcather closure group. ($p = 0.001$). Devices were deployed in 80 patients with sizes from 15 to 38 mm (median = 28 mm). Devices were not successfully deployed in seven patients. One patient had a device embolized into the right ventricle (surgical removal and closure of the ASD). Complications were found in 24 patients in the surgical group and 8 patients in the transcatheter closure group. The benefit for each patient was demonstrated in less morbidity and a shorter time spent in the hospital.

At the same time there were 32 patients who had large PDA closure by using an Amplatzer duct occluder. The PDA diameter ranged from 4 mm to 18 mm. The largest PDA was closed by using the 24 mm ASD device closure.

The future aspect of cardiac catheterization includes a much larger proportion of interventional procedures which will be close to 50 per cent. The ventricular septal defect (VSD) device has been developed over the past few years and is well underway for human trials. Transcatheter implant of the pulmonary valve has also been performed in a few patients. Health care reform over the past few years has yielded an opportunity to concentrate on a more

Table 5. Plan of management of all congenital heart disease 1998-1999.

Plan of management	CHD		AHD	
	N	%	N	%
Medication	184	42.6	25	67.6
Digitalis*	177	96.2	19	76.0
Surgery	180	41.7	3	8.1
Correction	143	79.4	1	33.3
Palliation	37	20.6	2	66.7
Cardiac catheterization	135	31.3	2	5.4
Hemodynamic study	85	63.0	2	100
Intervention	50	37.0	-	
Coil embolization - PDA	19	38.0		
- collaterals	4	8.0		
Pulmonary valvuloplasty	19	38.0		
Balloon atrial septostomy	3	6.0		
Laser pulmonary valvulotomy	2	4.0		
Aortic valvuloplasty	1	2.0		
Pulmonary angioplasty	1	2.0		
Renal angioplasty	1	2.0		
No treatment (follow-up)	99	22.9	8	21.6
Discharge	1	0.2	4	10.8
Total (cases)	432		37	

* Single or combined medication
CHD = congenital heart disease
AHD = acquired heart disease

Table 6. Outcomes of management.

	CHD		AHD	
	n = 432	%	n = 37	%
Improved	63	14.6	8	21.6
Died	20	4.6	1	2.7
No change	11	2.5	5	13.5
Cure	7	1.6	2	5.4
Unknown	331	76.7	21	56.8

effective way to treat congenital heart disease. Transcatheter closure which initially seemed to be more expensive will finally prove to be a complement methods to surgery in the management of congenital heart disease. It is expected that ASD and PDA device closure will replace the surgical procedure in the next decade just like balloon valvuloplasty of pulmonary valve has taken over from open pulmonary valvulotomy.

Acute rheumatic fever

Between 1990 and 2000, there were 88 patients reported of having acute rheumatic fever. It appeared that other than fever, the majority of them presented with carditis (88.6%) and polyarthritis (30.7%). Ninety-six per cent of the patients had elevated erythrocyte sedimentation rate and 74.4 per cent had positive C-reactive protein. However, antistreptolysin O was positive in only 74.4 per cent of patients while 100 per cent positive in antideoxyribonuclease B was found. Chest X-ray was a useful detecting tool for diagnosis of cardiomegaly and pericardial effusion. Prolonged PR interval was found in 40.2 per cent of the patients. Echocardiographic examination revealed pericardial effusion in 57 per cent, mitral regurgitation in 95 per cent and cardiomegaly in 96.3 per cent of the patients. Abnormal myocardial dysfunction was found in one patient.

Absolute bed rest for an appropriate duration was recommended in every patient. Symptomatic treatment of carditis is very important because it is the only cause of death. The anticongestive medications included diuretics, inotropic drugs and vasodilators, such as ACE inhibitor. In the authors' practice, in conjunction with anticongestive medications, antiinflammatory drugs such as corticosteriod were given to every patient with severe carditis (cardiomegaly or congestive heart failure) to improve the well being, however, it is still controversial whether it should be given in these patients.

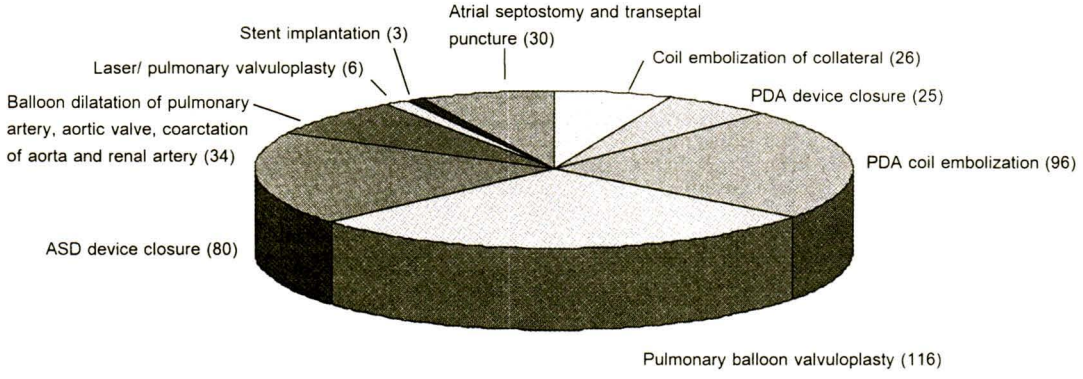

Fig. 1. Details of intervention cardiac catheterization in 413 patients from 1995 to 2002 (Feb).

Kawasaki Disease (KD)

In 1995, the authors reported clinical manifestations in the first 25 patients with KD who were diagnosed between 1981 and 1991[21]. However, intravenous gammaglobulin (IVIG) was not routinely given as a single dose regimen until 1995. If not treated, up to 15 per cent to 25 per cent may develop coronary artery aneurysm (CAA). Treatment of KD using IVIG lowered the frequency of CAA from 20 per cent to between 3 per cent and 5 per cent[22,23]. A subsequent meta-analysis confirmed these findings[24]. However, several cases of KD (up to 23%) [25-27] were unresponsive to the initial treatment with IVIG.

The authors retrospectively reviewed data from KD patients who received single dose IVIG (2 g/kg) treatment between 1995 and 2001. There were 130 patients with KD during the study period. Five patients were excluded; four patients had a late diagnosis with only aspirin treatment and were referred for an echocardiogram, one patient was referred for a dilated cardiomyopathy from a giant coronary aneurysm with a total occlusion of the left anterior descending. There were a total of 125 patients enrolled the present study, comprising 71 boys (56.8%). The patients were divided into group I, comprising 110 patients, receiving a single dose of IVIG and group II, comprising 15 patients (12%), requiring the second dose of IVIG. There were a total of 13 patients (10.4%) who had coronary artery aneurysms. Four patients had giant aneurysms. Six patients underwent cardiac catheterization and two patients underwent coronary bypass graft operation. One patient presented with unstable angina before his bypass operation. One patient had been treated with abciximab during the acute phase to decrease the size of giant aneurysm. The authors found that patients who were anemic (Hb<10 gm/dl), had a high neutrophil counts (>75%) or high band count, and low albumin level were at risk for failure to respond to a single dose of IVIG. As in previous studies[25-32], no association between age, gender or ESR and failure of initial IVIG treatment was found. Earlier, published reports [25,26] made the observation that the likelihood of coronary aneurysm is higher among patients with a greater severity of vasculitis; as reflected by a longer duration of fever and evidence of a more inflammatory response. The Harada score[29] was developed in the early 1990's to select patients who were at higher risk of developing CAA. Their scoring system was weighted towards younger males with anemia (Hct<35%), white blood cell counts >12,000/mm^3, low albumin level (<3.5 gm/dl) and a positive C reactive protein. The authors' current treatment is to give a single dose of IVIG (2 g/kg) to every KD patient in the acute phase. It was found that a scoring system was not particularly helpful in the hospital since the policy is to give IVIG to every KD patient.

Cardiac magnetic resonance imaging in congenital heart disease

Major progress has been made in pediatric cardiac imaging over the past two decades. Echocardiography and cardiac catheterization have assumed a leading role in diagnosing almost all forms of congenital heart disease. However, despite this substan-

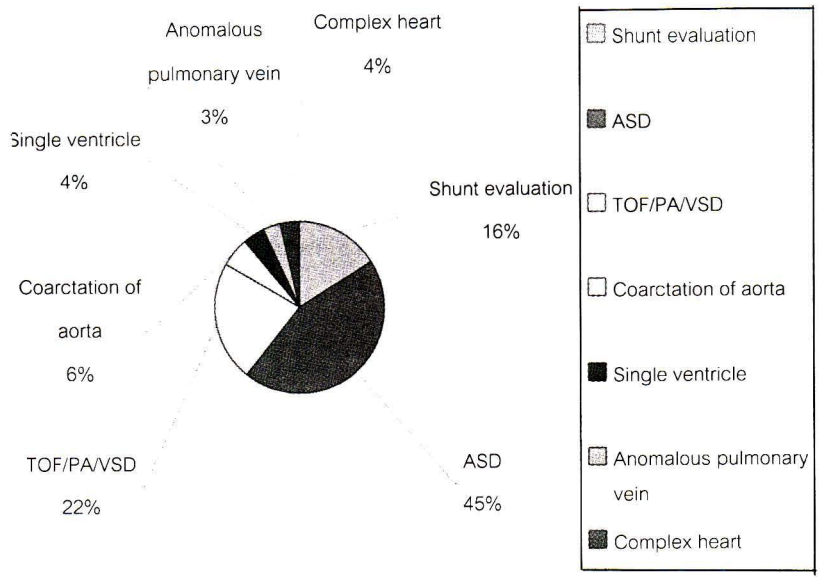

Fig. 2. The types of congenital heart diseases diagnosed by cardiac MRI.

ASD = atrial septal defect; TOF = tetralogy of Fallot; PA/VSD = pulmonary atresia with ventricular septal defect

tial progress, there are some limitations. As patients grow, the acoustic windows become progressively limited. Cardiac catheterization is invasive, expensive and associated with radiation exposure and also carries a risk of complications. There are also some limitations in both techniques to measure blood flow and quantify regional (in some cases global) myocardial function.

Magnetic resonance images (MRI) of the heart were first reported as early as the 1980's. This technique overcomes most of these limitations and bridges a gap between echocardiography and cardiac catheterization. Cardiac MRI has evolved from a technique that produced several static images in an hour-long examination to a modality capable of real-time imaging with 3-dimensional visualization of cardiovascular anatomy. Accurate quantification of blood flow (such as quantification of left-to-right shunt or regurgitation fraction) can be performed within an hour.

At Her Majesty's Cardiac Center, the authors started using cardiac MRI to evaluate patients with congenital heart disease in June 2000. The MRI studies were performed on a Philips 1.5T whole body MR scanner (NT Intera release 7, now upgrade to release 8, with Master gradient). It was performed on 146 congenital heart disease patients, 83 females and 63 males. The patients' mean ages were 25.7± 15.1 years (4-67.7 years) with a mean weight of 45.2±15.3 kg (14.3-99 kg). The diagnoses of the patients who underwent cardiac MRI are shown in Fig. 2.

Pre-operative and post-operative evaluation of congenital heart disease with cardiac MRI

The authors designed a prospective study comparing results obtained from cardiac MRI in patients such as tetralogy of Fallot (TOF), pulmonary atresia with ventricular septal defect (PA/VSD), anomalous pulmonary venous drainage or obstruction and other complex heart anomalies with cardiac catheterization. The usual cardiac MRI study was performed with localizer imaging. These are usually obtained in the axial, coronal, and sagittal planes using fast gradient-echo sequences without ECG gating. The goal is to locate the major structures such as atria, ventricles and the great vessels. Then a series of breathhold multislice, multiple heart phase gradient echo sequence covering the whole interested region was performed in different planes. MR velocity mapping enables measurement of velocities across a stenotic area. Using the modified Bernoulli

equation, the stenotic gradient can be calculated. This technique does not differ from Doppler echocardiography in its practical applications. The main advantage of MR velocity mapping compared to Doppler echocardiography is the unlimited choice of imaging planes with MRI. The authors also tried to demonstrate a close agreement of pulmonary artery measurement and right ventricular outflow tract obstruction in patients with TOF between cardiac MRI and cardiac catheterization. In doing so, it is expected to replace cardiac catheterization in pre-operative evaluation of patients with TOF and PA/VSD. For pre-operative evaluation of great vessel anatomy such as coarctation of the aorta (Fig. 3), vascular rings, and pulmonary artery sling, diagnosis could be made by using both spin echo and gradient echo technique. Contrast-enhanced 3-dimensional angiography in selected cases (Fig. 4) can also be used.

In the follow-up of patients who have undergone repair of TOF, an imaging technique is required to provide adequate information on residual VSD, pulmonary stenosis, the amount of pulmonary regurgitation, and systolic and diastolic biventricular size and function. Because of well-known limitations, transthoracic echocardiography often fails to provide the necessary information, particularly in quantification of pulmonary regurgitation and right ventricular function. The authors are currently using cardiac MRI to evaluate this group of patients, particularly in adult patients who were operated a more than 10 years ago.

Noninvasive quantification of left-to-right shunt using a phase-contrast cine magnetic resonance imaging

Shunt quantification is essential in the management of congenital heart disease. Clinical methods available to yield quantitative flow data have major drawbacks. Doppler ultrasound methods are noninvasive but highly observer-dependent. Radionuclide angiocardiography is restricted to simple left-to-right shunt lesions with normal ventricular function, and patients are exposed to ionizing radiation. Indicator dilution techniques (cardiac catheterization) are invasive, as is oximetry, a method complicated by variations of oxygen consumption and difficulties in estimation of mixed venous oxygen content in atrial-level shunt patients. In contrast, phase-contrast cine MRI (PC-MRI) is noninvasive and can determine aortic and pulmonary flow volumes in patients with congenital heart disease.

MRI imaging techniques consist of a conventional flow-sensitive gradient-echo pulse sequence. Through-plane measurements were performed at the

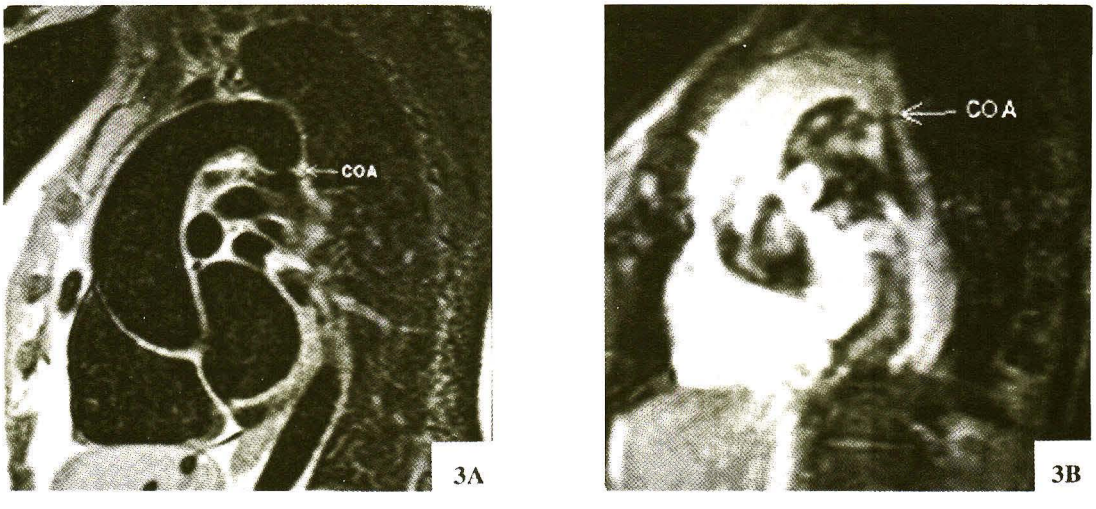

Fig. 3. Cardiac MR1 demonstrated coarctation of aorta in (A) spin echo and (B) Balance FFE technique with flow velocity (black jet) was measured.

CoA = Coarctation of aorta

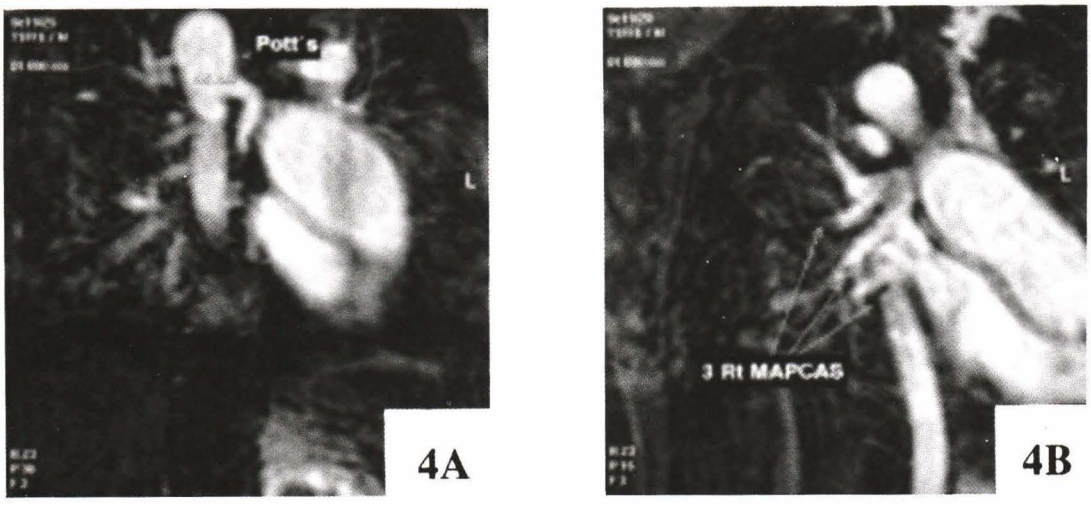

Fig. 4. Cardiac MR1 demonstrated 3D-imaging using of gradient echo small left pulmonary artery from descending aorta to Pott's shunt (A) and three collateral vessels from descending aorta (B).

MAPCA = major aorto pulmonary collateral artery

transaortic and transpulmonary artery (Fig. 5) with velocity-encoded values of 200 to 300 cm/s (aorta), 150 cm/s (pulmonary artery). Data analysis performed by drawing a region of interest (ROI) to all modulus images and subsequently projected to the corresponding phase images. Flow calculation in a single vessel was automatically calculated and completed within 1 to 2 minutes (Fig. 5). From June 2001 to February 2002, the authors prospectively enrolled 26 children with an atrial- or ventricular-level left-to-right shunt. Of these 26 patients, 21 had secundum ASD, two partial anomalous pulmonary venous return, and three patients with ventricular septal defect. Comparison of Qp:Qs measured by both cardiac catheterization and cardiac MRI revealed a fairly good agreement between value measured in both methods. Currently, up to 50 patients are being enrolled to see whether cardiac MRI can be used to replace cardiac catheterization in patients with left-to-right shunt lesion, except in those associated with pulmonary hypertension.

Evaluation of atrial septal defect for transcatheter closure

Atrial septal defect (ASD) accounts for 10 per cent of congenital heart disease at birth and as much as 30-40 per cent of cases seen in adults. It is generally agreed that a large ASD should be closed. Surgical repair of ASD is a safe and widely accepted procedure with negligible mortality. However, it is associated with morbidity, discomfort and a thoracotomy scar. As an alternative to surgery, transcatheter closure of ASD has been developed. The authors initially reported experience of transcatheter closure of atrial septal defect using the Amplatzer™ Septal Occluder with intermediate term follow-up[33-35]. Conventional methods for evaluation of the ASD include transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE). The location of the defect, size of the defect [33], and age of the patient are the major determining factors indicating either surgical correction or transcatheter closure by a device. Three dimensional images[34,35] were constructed to simulate the ASD view as seen by a surgeon, however, it still requires TEE which poses a small but potential life threatening risk with some discomfort. Visualization of ASD by cardiac MRI by spin-echo technique is well established, as is the use of dynamic imaging with gradient-echo technique to visualize ASD shunt flow. The authors have been using cardiac MRI to evaluate patients who are candidates for transcatheter closure of the ASD (Fig. 6). The results have revealed a better agreement between the size of the defect

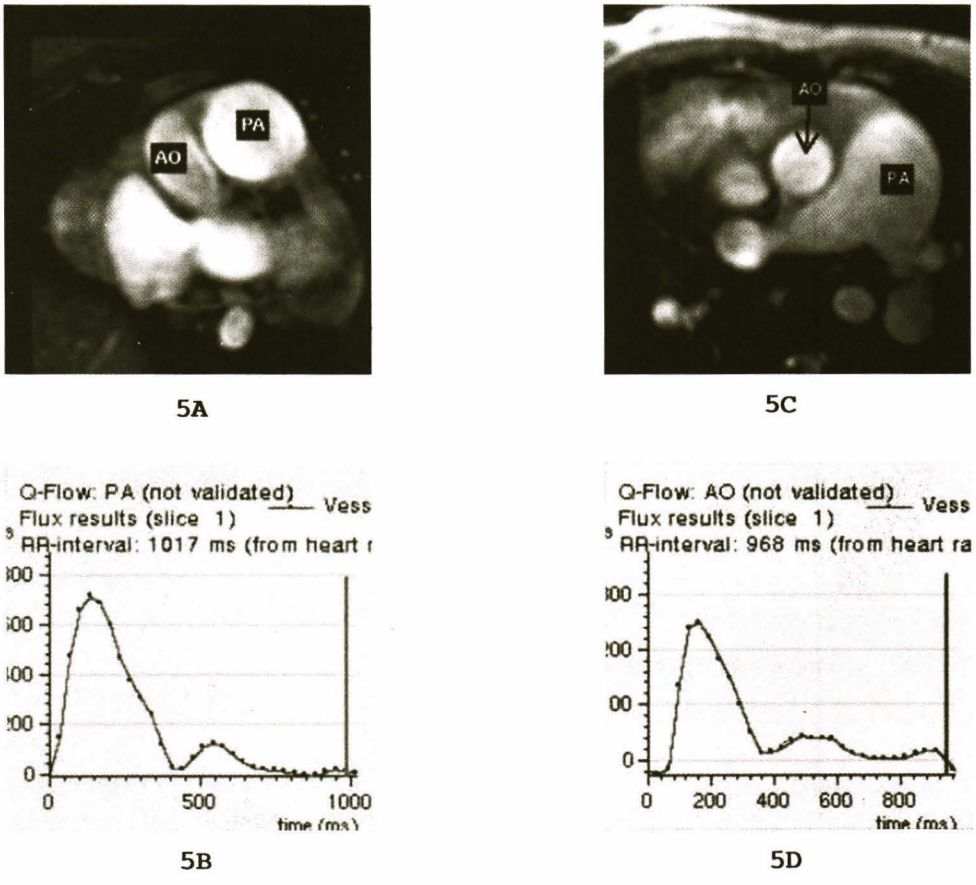

Fig. 5. Left-to-right shunt was calculated by phase contrast MRI measured flow to pulmonary artery A, B compared to aorta C, D. B and C show flow quantification.

PA = Pulmonary artery; Ao = Aorta

measured by cardiac MRI with the size of the device used for ASD closure in each patient when compared with measurement by TEE.

Myocardial diseases in children

Despite the rarity, myocardial diseases are among the important causes of mortality and morbidity in children. Dilated cardiomyopathy (DCM) has been found to be the most common type of cardiomyopathy with the incidence of 8-10 per 100,000 populations in the United States and Europe[36]. Hypertrophic cardiomyopathy (HCM) and restrictive cardiomyopathy (RCM) were reported to account for 20-30 per cent and 5 per cent of cardiomyopathy respectively. It is not that difficult to diagnose these diseases. However, management of myocardial diseases has been a problem for a long time. Without curable means to address the hemodynamic problems, the only option is heart transplantation, which is unavailable in most Thai institutions. Furthermore, post-transplantation care requires close follow-up and endless chemotherapy.

From January 1992 to July 2001, there were 51 cases of primary myocardial diseases diagnosed and treated at the Department of Pediatrics, Siriraj Hospital, Mahidol University, Bangkok, Thailand. This accounted for 0.8 per cent of patients with cardiovascular diseases diagnosed at the same period of time. The distribution of myocardial diseases is demonstrated as a diagram in Fig. 7. Forty seven per cent were male. Median age at the time of diagnosis was 7 months (<1 months-1 year), 4 years (3 months-12 years), 3.75 years (2 months-11 years) and 4 years (4-5 years) for acute myocarditis, DCM,

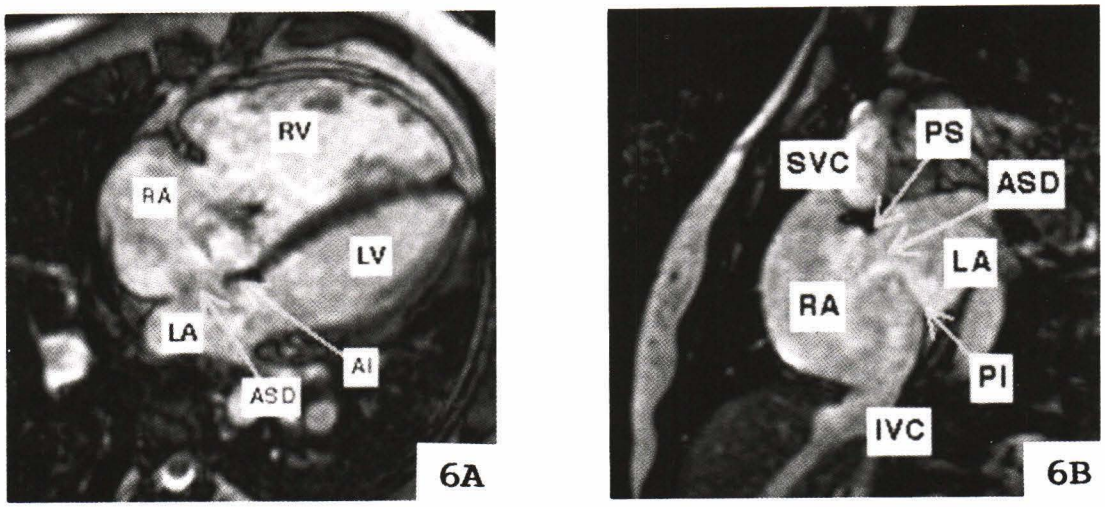

Fig. 6. **Cardiac MR1 demonstrated profile of atrial septal defect (ASD) in four-chamber views (A) and short-axis view (B).**

ASD = Atrial septal defect; RA = Right atrium; LA = Left atrium; LV = Left ventricle; RV = Right ventricle; AI = Anterior inferior rim; SVC = Superior vena cava; PI = Posterior inferior rim; PS = posterior rim; IVC = Inferior vena cava

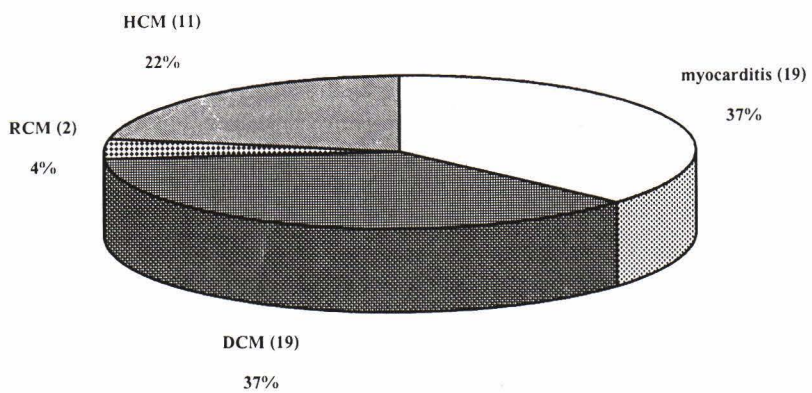

Fig. 7. **Demonstrates the distribution of myocardial diseases.**

DCM = dilated cardiomyopathy;
HCM = hypertrophic cardiomyopathy;
RCM = restrictive cardiomyopathy

HCM and RCM respectively. Almost all patients (>85%) presented with congestive heart failure (CHF) with or without cardiac arrhythmia. Abnormal findings on chest X-ray and electrocardiogram were not specific for myocardial diseases. Echocardiography has been the most reliable, non-invasive diagnostic tool for most myocardial diseases. Ejection fraction in acute myocarditis was signifi-

Table 7. Management in myocardial diseases.

Management	Myocarditis		DCM		RCM		HCM	
	Case	%	Case	%	Case	%	Case	%
Outpatient	0	0	0	0	0	0	9	82
Inpatient	19	100	19	100	2	100	2	18
Admit to ICU	5	26	9	48	2	100	0	0
Medication								
IV inotropic drugs	12	64	17	90	2	100	0	0
Inotropic, vasodilator and diuretics	19	100	9	100	2	100	0	0
IVIG	7	37	0	0	0	0	0	0
CoQ10	0	0	3	16	0	0	0	0
Beta-blockers	0	0	0	0	0	0	10	90
No medication	0	0	0	0	0	0	0	0
Surgical treatment	0	0	0	0	0	0	2	18

ICU = Intensive care unit; IV = intravenous; IVIG = intravenous immunoglobulin; CoQ10 = Co-enzyme Q10; DCM = dilated cardiomyopathy; HCM = hypertrophic cardiomyopathy; RCM = restrictive cardiomyopathy

Table 8. Outcome of treatment after a 3-year follow-up.

Outcome	Myocarditis		DCM		RCM		HCM	
	Case	%	Case	%	Case	%	Case	%
Improve	15	80	15	26	0	0	0	0
Same	2	10	13	63	2	100	10	90
Death	2	10	2	10	0	0	1	10

DCM = dilated cardiomyopathy; HCM = hypertrophic cardiomyopathy; RCM = restrictive cardiomyopathy

cantly higher than in DCM (37% *vs* 20%). In addition, serum cardiac troponin T (cTnT) and creatine kinase MB isoenzyme (CK-MB) levels were significantly higher in acute myocarditis than DCM even with severe CHF[37]. Right ventricular endomyocardial biopsy was performed in 9 cases (acute myocarditis 5 cases, DCM 2 cases, RCM 2 cases) and had 40 per cent sensitivity in acute myocarditis, 100 per cent in DCM and RCM.

Management in each group of patients is demonstrated in Table 7. Outcome of treatment at the follow-up period of about 3 years is shown in Table 8. There was no difference in the mortality rate in acute myocarditis patients with or without intravenous immunoglobulin (IVIG) administration (2 gram/kg).

Up until now, there is no promising medication or means in the treatment of myocardial diseases except heart transplantation which has some limitations e.g. availability of a donor, a good transplantation team and close follow-up with regular invasive check up and high cost etc. Dual chamber cardiac pacing with short atrioventricular interval in the treatment of hypertrophic obstructive cardiomyopathy has been reported not to be beneficial[38]. Recently, carvedilol and pentoxifylline have been reported to improve functional class and ventricular function in DCM patients[39,40].

Cardiac arrhythmia in children

Cardiac arrhythmia is an important cardiovascular problem in children.

Tachyarrhythmia

The majority of cases in children are supraventricular tachycardia (SVT) by re-entry mechanism e.g. Wolff-Parkinson-White syndrome (WPW syndrome), atrioventricular node re-entry tachycardia (AVNRT), concealed accessory pathway, etc. Some types of structural heart diseases are more likely to have SVT e.g. Ebstein anomaly, complex congenital heart diseases and hypertrophic cardiomyopathy etc. Some of the SVT are automaticity mechanism e.g. post-operative junctional ectopic tachycardia (JET) which usually occurs in the acute post-corrective operation period of tetralogy of Fallot (TOF) or

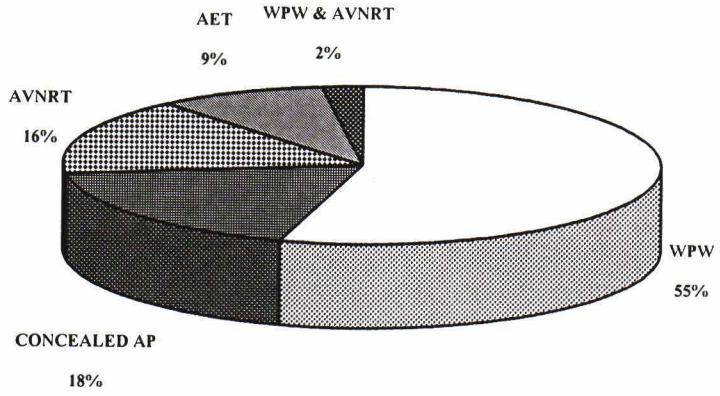

Fig. 8. Diagram shows the diagnoses of patients with tachyarrhythmia who underwent RFCA.

AET = atrial ectopic tachycardia;
AVNRT = atrio-ventricular node re-entry tachycardia;
WPW = Wolff-Parkinson-White syndrome;
CONCEALED AP = concealed accessary pathway

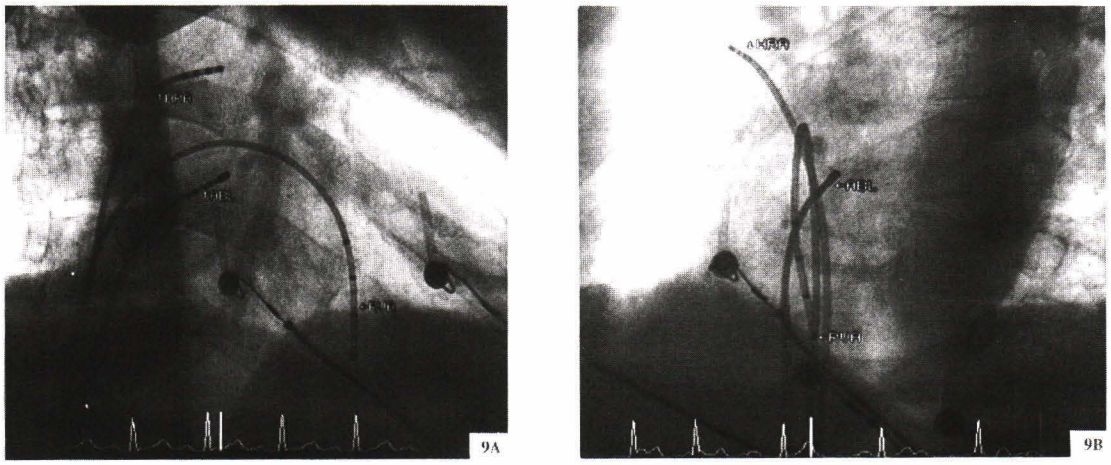

Fig. 9. X-rays of the successful spot in AVNRT (A) Right anterior oblique view (RAO) (B) Left anterior oblique view (LAO).

ventricular septal defect (VSD). Atrial ectopic tachycardia (AET) could occur weeks after open heart surgery or occur in a normal heart. Automaticity mechanism makes these conditions difficult to control and usually ends up with radio-frequency catheter ablation (RFCA) to prevent tachycardia-induced cardiomyopathy. However, this procedure has an increased risk of complications in small children especially those less than 15 kg[41]. Ventricular tachycardia (VT) is quite rare in children. Idiopathic VT e.g. right ventricular outflow tract tachycardia, left ventricular tachycardia from posterior bundle are the

2 most common forms of VT. Long QT syndrome with Torsade de pointe is one of the important causes of death in children. Ventricular tachycardia could also occur in various kinds of myocardial diseases and plays a significant role in sudden death.

Long-term post-operative tachyarrhythmia are atrial flutter in post-operative TOF, as well as in post Mustard and Fontan operation; and VT in post-operative TOF or others with a history of ventriculotomy. The condition affects the quality of life and mortality rate in these groups of patients. Most of them are intractable to potent anti-arrhythmic drugs. Corrective surgery of the impaired cardiac anatomy e.g. repair tricuspid regurgitation and pulmonary insufficiency in post-operative TOF might abolish cardiac arrhythmia.

The mainstay of the management of tachyarrhythmia in children is an antiarrhythmic drug. However, radio-frequency catheter ablation (RFCA) is widely accepted to be the method of choice, which is safe, effective and curable with a high success rate in this era[42,43]. From January 1996 to December 2001, 44 children with SVT underwent RFCA under the conventional mapping method at Siriraj Hospital, Mahidol University, Bangkok, Thailand. All these children met the modified indications of Walsh and Van Hare[43,44]. Median age and weight at the time of the procedure were 11 (1.1-13) years old and 28 (6.8-58) kg respectively. The main presenting symptoms were palpitation (70%). The underlying cardiac arrhythmia is demonstrated in Fig. 8. The median time of the procedure was 100 (60-320) minutes. Acute success rate was 91 per cent. Two patients had a recurrence of tachyarrhythmia. One patient had RFCA redone successfully. The other one with AET is waiting for a repeat RFCA. No major complications were found, except one child with WPW syndrome with antero-septal accessory pathway. This child required a permanent pacemaker implantation after successful RFCA. Example of the catheters' position in conventional mapping is shown in Fig. 9. Long-term post-operative atrial flutter has been treated with RFCA with the initial success rate as high as 75 per cent and recurrent rate of up to 50 per cent[45,46]. This seems to be the future trend owing to the improved survival rate in post-operative complex cardiac surgery.

Bradyarrhythmia

It is uncommon in children. Complete heart block (CHB) without structural heart defects has been reported. One-third of the children with CHB are associated with corrected transposition of great arteries (L-TGA) and complex congenital heart diseases with left isomerism. The rest are secondary from maternal connective tissue diseases[47,48]. Permanent pacemaker (PPM) implantations are required in more than 50 per cent of the later group of patients during the neonatal period due to permanent damage of the conducting system. Fetal echocardiogram is essential for the diagnosis and follow-up of pregnant women with connective tissue diseases. Oral dexamethasone and plasmapheresis whenever second-degree atrioventricular block is detected might improve fetal atrioventricular conduction[47]. The majority of complete heart block in children at our institution occurred after open heart surgery. From July 1998 to December 2001, 18 cases required PPM implantation. Four cases had congenital complete heart block. Of these four, two were diagnosed as neonatal lupus erythematosus as previously reported [49]. There was one child with un-operated L-TGA, one with restrictive cardiomyopathy, and another one in post RFCA antero-septal accessoary pathway. Eleven cases were post-cardiac surgery mainly post-corrective surgery of TOF and VSD. Permanent pacemaker implantation was intervened after 14 days post-operation waiting for resumption of atrioventricular conduction as Class I recommendation of American Heart Association. Dual-chamber PPM should be chosen in patients with residual structural cardiac defects or myocardial dysfunction which requires atrial contraction to optimize the cardiac output. This would result in improved quality of life.

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ความก้าวหน้าในการดูแลรักษาโรคหัวใจในเด็ก : อดีต, ปัจจุบันและอนาคต

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อุบัติการณ์ของโรคหัวใจพิการแต่กำเนิดในเด็กแรกเกิดที่โรงพยาบาลศิริราชคือ 4.36 รายต่อทารกแรกเกิด 1,000 ราย ในผู้ป่วยจำนวนนี้ประมาณ 1 ใน 3 จะต้องการการรักษาเร่งด่วน ในปัจจุบันหน่วยโรคหัวใจเด็กได้ให้บริการผู้ป่วยโรคหัวใจพิการแต่กำเนิดและโรคหัวใจชนิดอื่นในเด็ก ทั้งในด้านการวินิจฉัยโดยใช้เทคโนโลยีใหม่ เช่น Cardiac Magnetic Resonance (MRI) โดยการตรวจสวนหัวใจและให้การรักษาเช่น การขยายลิ้นหัวใจ–เส้นเลือด และปิดรูรั่วของผนังหัวใจ ตลอดจนการรักษาภาวะหัวใจเต้นผิดจังหวะโดยการจี้ทางสายสวนหัวใจด้วยคลื่นวิทยุ จึงอาจกล่าวได้ว่า การดูแลผู้ป่วยโรคหัวใจเด็กได้เริ่มเปลี่ยนจากการวินิจฉัยอย่างเดียวเป็นไปในรูปแบบการรักษาที่จะมีมากขึ้นในอนาคต

คำสำคัญ : โรคหัวใจในเด็ก, ความก้าวหน้าในการดูแลรักษา, ประเทศไทย

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