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Pattern of Congenital Heart Disease and Access to Tertiary Cardiac Care in Malaysia

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Summary

This prospective, descriptive study was carried out to determine the pattern and the type of congenital heart disease seen in the Paediatric Departments in 4 government hospitals. The accessibility of surgical or transcatheter interventional treatment was also assessed. Consecutive new patients seen for suspected congenital heart disease from 1/1/95 till 31/12/95 (Group I) were registered. Records of patients seen from 1/1/95 to 31/8/95 (Group Ia) were reviewed 6 months after presentation to determine the nature of treatment received. Group II were cardiac patients who were first seen prior to the year 1995 and had undergone cardiac surgery or transcatheter interventional procedures in 1995.

Of the 375 patients registered in the 4 hospitals, 250 were new patients and 125 were patients first seen prior to 1995 and had surgery or transcatheter interventional procedures in 1995. Of the 250 new patients, 83.2% had acyanotic cardiac lesions and 16.8% had cyanotic lesions. Ventricular septal defect was the commonest lesion, constituting 37.2%, followed by patent arterial duct (18.8%) and atrial septal defects (9.6%). At 6 month review, access to close-heart surgery or transcatheter interventional treatment were readily available. However, for patients with ventricular septal defects, 42.1% who required closure were still waiting for surgery 6 months after presentation. Of the 4 patients with Fallot's Tetralogy who required surgery, 2 had modified Blalock-Taussig shunt and 2 were awaiting surgery. In 1995, a total of 204 cardiac procedures were performed, there were 114 (55.9%) open heart procedures, 50 (24.5%) surgical ligation of the arterial duct, 28 (13.7%) modified Blalock-Taussig shunts, 11 (5.4%) transcatheter duct occlusion and 1 (0.5%) balloon valvuloplasty. The mean age of Fallot's Tetralogy repair was 6.4 years. No corrective surgery was performed for patients with complex cardiac lesions from the 4 hospitals in 1995. In conclusion, the pattern of congenital heart disease was similar to that seen world wide. Except for patent arterial duct, access to surgical treatment was inadequate.

Key Words: Congenital heart disease, Surgical intervention, Transcatheter intervention

Introduction

Congenital heart disease is the most common of the major congenital malformations with an incidence of about 8 per 1000 live births[1,2]. With a total of 486, 871 live births per year in 1987[3], an estimated 38,950 babies would have been born with congenital cardiac malformations. In the early 80's, few diagnostic facilities or cardiac surgery were available for children with congenital heart disease in Malaysia. Over the last decade, there has been rapid development of cardiac services and availability of paediatric cardiologists and paediatric cardiac surgeons. This means that children now have access to definitive treatment for congenital

heart disease. However, a preliminary review of cardiac patients in the Paediatric Cardiology Clinic at Hospital Ipoh in 1993[4] showed that there was a disproportionately high percentage of non-life threatening cardiac lesions seen at Hospital Ipoh compared to the recognized frequency. For example, 55% of the patients with congenital heart disease had ventricular septal defects compared to the recognized frequency of 25-30% world wide[1,2]. The probable reason was that of natural selection where most of the neonates with life-threatening duct-dependent congenital heart lesions were not recognized, or if recognized had no treatment, leading to early death. The other finding was that many of the patients with congenital heart disease were still waiting for surgery beyond the recommended age, for example up to teens for Fallot's Tetralogy[4].

The objectives of the study were to determine the pattern and the type of congenital heart disease seen in the Paediatric Departments in 4 government hospitals; and to determine the access of these patients to surgical or transcatheter interventional treatment.

Materials and Methods

This was a prospective descriptive study. The Paediatric Departments of Hospital Ipoh, Hospital Teluk Intan, Hospital Umum Sarawak and Hospital Queen Elizabeth were involved in the study. All were state General Hospitals with echocardiography services except Hospital Teluk Intan which was a district general hospital with a paediatrician but no echocardiography facilities. The diagnosis of congenital heart disease was made clinically or with echocardiography examination. Each consecutive new patient who was seen at these four paediatric departments from 1st January 1995 till 31st December 1995 for suspected congenital heart disease was registered. Inclusion criteria was all patients with a diagnosis of congenital heart disease and exclusion criteria was cyanosis due to non-cardiac causes.

Two groups of patients were studied. Group I consisted of all new cardiac patients who were seen at the hospitals from 1st January 1995 to 31st December 1995. Group I patients were subdivided into Group Ia and Group Ib. Group Ia included patients seen from 1st January 1995 to 31st August 1995. Their records were reviewed 6 months after their presentation to the hospitals to determine the nature of treatment received. The date of presentation of the patients to the hospitals studied rather than the date the patient was first seen at the tertiary cardiac centre was considered because 2 of the hospitals were in East Malaysia. These 2 hospitals have a fixed number of cardiac surgery slots allocated to them per week. Patients for surgery are prioritized in these hospitals by the paediatricians and referred to the tertiary cardiac centre for assessment and cardiac surgery or transcatheter interventional procedures in a single trip.

Cardiac lesions with the following criteria were considered to be awaiting surgical or percutaneous transcatheter interventional procedures if the patients had not received the appropriate treatment at 6 months follow-up (Table I).

Group Ib were patients seen from 1st September 1995 to 31st December 1995 with no follow-up. To determine the availability and the capacity of tertiary cardiac care, all cardiac patients who were first seen prior to the year 1995 and had undergone cardiac surgery or transcatheter interventional procedures in 1995 were also registered (Group II).

Results

A total of 375 patients were registered in these 4 hospitals. Of these, 250 were new cardiac patients (Group I). Two hundred and forty-six patients (98.4%) had echocardiography examination either by the paediatricians at the general hospitals or at tertiary referral centres to confirm the clinical diagnosis. The diagnosis of 4 patients (1.6%) were made without echocardiography confirmation; they were two ventricular septal defects; one aortic stenosis and one atrial septal defect. Breakdown of the new patients by the individual hospitals were 21.0%, 13.7%, 18.5% and 46.8% from Hospital Ipoh, Hospital Teluk Intan, Hospital Umum Sarawak and Hospital Queen Elizabeth respectively (Table II). There were a total of 125 patients who were first seen prior to the year 1995 and had cardiac surgery or transcatheter interventional procedures in the year 1995 (Group II).

Table I
Criteria considered to be awaiting surgical or transcatheter intervention

Cardiac Lesions	Criteria considered to be awaiting interventional treatment at 6 months follow-up
Ventricular septal defects	•Large ventricular septal defect in intractable congestive cardiac failure and failure to thrive despite antifailure therapy[5,6] or •Evidence of raised pulmonary vascular resistance by echocardiographic examination.
Patent arterial duct	•All patent arterial duct except those who were not in congestive cardiac failure, thriving well and were waiting for umbrella occlusion when a body weight of 10 kilogram was achieved.
Atrial septal defect	•Evidence of raised pulmonary vascular resistance by echocardiography examination.
Fallot's Tetralogy	•Await shunt surgery if one episode of hypercyanotic attack or failure to thrive with SaO_2 of < 60%. •Await total correction if they were more than 24 months of age and had suitable anatomy for one stage corrective surgery by echocardiography examination[6,7].
Simple transposition of the great arteries	•Arterial switch by 6 weeks of age[6] .
Coarctation of the aorta	•Repaired or relieved at 6 months follow-up.
Pulmonary or aortic stenosis	•Pressure gradient was > 60 mm Hg by echocardiography examination and with clinical or ECG evidence of ventricular strain or ischaemia.

Of the 250 new patients, the ages of 246 patients were available. Analysis of the age distribution of patients at presentation showed that 4.9% of patients were diagnosed by the first week of age, 2.8% from the second week to the fourth week of age and 16.7% from 1 month to 3 months of age. The cumulative proportions of infants registered under 1 week, 1 month and 3 months were 4.9%, 7.7%, and 24.4% respectively.

Analysis of the pattern of congenital heart disease showed that of the 250 new patients, 208 patients (83.2%) had acyanotic lesions and 42 patients (16.8%) had cyanotic lesions. Ventricular septal defect was the commonest lesion constituting 37.2% of all congenital heart disease, followed by patent arterial duct 18.8% and atrial septal defects 9.6% (Table III).

A total of 153 patients were seen from 1/1/95 till 31/8/95 (Group Ia). The records of these patients were reviewed 6 months after registration to determine their access to tertiary treatment. After 6 months, 145 patients were reviewed and 8 patients were lost to follow-up. Of these 8 patients, 1 patient with ventricular septal defect died at home; 3 patients with patent arterial duct did not return after referral to tertiary cardiac centre; 4 patients defaulted follow-up. Of these 4 patients, 2 had ventricular septal defects, 1 congenital rubella syndrome with atrial septal defect and 1 Down syndrome with atrioventricular septal defect.

Of the 145 patients reviewed, 66 patients (43.1%) received interventional treatment, 52 patients (34.0%) did not require any intervention, 25 patients (16.3%) were waiting for intervention and 2 (1.3%) were inoperable. Of the 25 patients awaiting intervention; 16 were ventricular septal defects, 3 atrial septal defects and 1 atrioventricular septal defect awaiting surgical correction; 2 patent arterial ducts awaiting occlusion; 2 Fallot's Tetralogy were waiting for surgery and 1 complex cyanotic heart disease was waiting for a systemic-pulmonary shunt. Thirty-eight (60.3%) of the 63 patients with ventricular septal defect required

Table II

Number of patients seen in the 4 hospitals by group

Groups / Hospital	Patients from 1/1/95 to 31/12/95 (Group I)	1/1/95 to 31/8/95) patients with 6 mo follow-up (Group 1a)	Non 1995 patientst treated in 1995 (Group II)
Ipoh	54 (21.0%)	43	23
Teluk Intan	34 (13.7%)	19	14
Umum Sarawak	46 (18.5%)	22	47
Queen Elizabeth	116 (46.8%)	69	41
Total	250 (100.0%)	153	125

surgery; of which 22 patients (57.9%) had their ventricular septal defects repaired and 16 patients (42.1%) were awaiting closure. There were 29 patients with patent arterial duct. One arterial duct closed spontaneously, 26 had surgical ligation/division or transcatheter occlusion and 2 (7.1%) were awaiting occlusion. Of the 8 patients with atrial septal defects; 4 were closed surgically; 3 (42.9%) were waiting for closure and one did not require intervention. Four patients had atrioventricular septal defects. Three of them were associated with Down syndrome and were not considered for surgery; one patient was waiting for surgical repair. Of the obstructive lesions; 4 of the 12 patients with pulmonary valve stenosis had surgical valvotomy, 8 patients did not require intervention; 3 patients had aortic stenosis that did not require intervention. No coarctation of aorta was diagnosed during the first 8 months of the study period. Of the 21 patients with cyanotic lesions, 2 had transposition of the great arteries, 9 Fallot's Tetralogy, 2 double outlet right ventricle, 4 complex cyanotic heart disease and one each of pulmonary atresia with intact ventricular septum, pulmonary atresia with ventricular septal defect, tricuspid atresia and partial anomalous pulmonary venous drainage. Only 2 patients had corrective surgery i.e. one arterial switch repair at 2.5 months old for transposition of the great arteries and one correction of partial anomalous pulmonary venous drainage. The other transposition of the great arteries was associated with VSD and mild pulmonary stenosis. The patient presented at 2.5 years of age, had congestive cardiac failure and pulmonary vascular disease and was inoperable. Eight patients had modified Blalock-Taussig shunts; 2 for Fallot's Tetralogy, 2 for double outlet right ventricle, 2 for complex cyanotic heart disease and 1 each for pulmonary atresia with ventricular septal defect and tricuspid atresia. There were 9 Fallot's Tetralogy, 5 were considered not requiring surgery by our criteria at 6 months follow-up though they would eventually require surgery; of the 4 patients who required surgery, 2 had modified Blalock-Taussig shunt and 2 were awaiting surgery. None of the Fallot's Tetralogy had corrective surgery .

A total of 204 cardiac surgery or transcatheter interventional procedures were performed in the year 1995 for 201 patients from the 4 hospitals (Table IV). One hundred and twenty-six patients (62.69%) were patients first seen prior to the year 1995 (Group II) and 75 (37.31%) were patients seen in the year 1995 (Group I). Of the 204 procedures, there were 114 (55.9%) open heart procedures, 50 (24.5%) surgical ligation of the arterial duct, 28 (13.7%) modified Blalock-Taussig shunts, 11 (5.4%) transcatheter duct occlusion and 1 (0.5%) balloon valvuloplasty (Table IV). There were 3 patent arterial ducts with residual shunts, 2 were closed by occlusion and 1 by surgical ligation. All surgeries were performed at the National Heart Institute, Kuala Lumpur except 6 ventricular septal defects, 3 atrial septal defects and 3 Fallot's Tetralogy were repaired in Singapore. A Blalock-Taussig shunt for a complex cyanotic heart lesion was performed in the United States.

Of the 23 corrective surgery for Fallot's Tetralogy, 20 were performed at the National Heart Institute, Kuala

Table III
Pattern of congenital heart disease in four hospitals in Malaysia in 1995 as compared to the pattern from other parts of the world

ACYANOTIC LESIONS	No. of Patients	Percentage Malaysia	Percentage Liverpool[9]	Percentage United States[1]
Ventricular septal defects	93	37.2	32.5	25-30
Patent arterial duct	47	18.8	11.9	6-8
Atrial septal defects	24	9.6	5.9	6-8
Pulmonary stenosis	21	8.4	7.6	5-7
Atrioventricular septal defects	8	3.2	2.4	-
-Aortic stenosis	4	1.6	5.1	4-7
Mitral valve prolapse	5	2.0	-	-
Tricuspid regurgitation	2	0.8	-	-
Coarctation of aorta	1	0.4	6.3	5-7
Hypertrophied cardiomyopathy	1	0.4	-	-
Anomalous origin left coronary artery	1	0.4	-	-
Corrected transposition of the great arteries	1	0.4	-	-
Subtotal	**208**	**83.2**		
CYANOTIC LESIONS				
Fallot's Tetralogy	22	8.8	5.9	5-7
Transposition of the great arteries	5	2.0	5.0	3-5
Double outlet right ventricle	3	1.2	-	1-2
Pulmonary atresia + VSD	2	0.8	0.8	-
Tricuspid atresia	2	0.8	1.7	1-2
Total anomalous pulmonary venous return	1	0.4	1.4	1-2
Partial anomalous pulmonary venous return	1	0.4	-	-
Univentricular heart	2	0.8	1.7	1-2
Pulmonary atresia intact ventricular septum	1	0.4	-	
Truncus arteriosus	1	0.4	1.1	1-2
Mitral atresia	1	0.4	-	-
Complex cyanotic heart disease	1	0.4	-	-
Subtotal	**42**	**16.8**		
Total	**250**	**100.0**		

Lumpur and 3 in Singapore. The ages at repair of the 20 patients at the National Heart Institute ranged from 1.46 years to 14.72 years, with a mean of 6.4 years. Two repairs (10.0%) were performed when the patients were less than 2 years old, 8 (40.0%) during preschool years from 2-5 years old, 8 (40.0%) at primary school aged from 6-12 years old and 2 patients (10.0%) were more than 12 years old. For the 3 Fallot's Tetralogy repaired in Singapore, their ages ranged from 2.30 - 3.51 years with a mean of 2.71 years of age. No corrective surgery for complex cyanotic heart lesions was performed in the year 1995.

Table IV
Number of cardiac surgeries and transcatheter interventions in the year 1995

Type of cardiac procedure	No. of procedures for 1995 patients	No. of procedures for non -1995 patients	Total number of procedures
VSD closure	23	22	45
ASD closure	6	18	24
PDA ligation	23	26+1*	50
PDA occlusion	5+1*	4+1*	11
PS valvotomy	5	3	8
PS valvuloplasty	0	1	1
AS valvotomy	0	1	1
MS valvotomy	0	1	1
TOF repair	1	22	23
TGA arterial switch	1	0	1
TGA atrial switch	0	4	4
DORV repair	0	2	2
PAVSD repair	0	3	3
T/PAPVR correction	1	1	2
BT shunt	10	18	28
Total	**75+1***	**126+2***	**201+3**

VSD ventricular septal defect; ASD atrial septal defect; PDA patent ductus arteriosus; PS pulmonary stenosis; AS aortic stenosis; MS mitral stenosis; TOF Fallot's Tetralogy; TGA transposition of the great arteries; DORV double outlet right ventricular; PAVSD pulmonary atresia with ventricular septal defect; T/PAPVR total/partial anomalous pulmonary venous return; BT Blalock-Taussig.
**There were 3 patent arterial ducts with residual shunts, 2 were closed by occlusion and 1 by surgical ligation.*

Discussion

The cumulative proportions of infants registered under 1 week, 1 month and 3 months of age were 5%, 8% and 24% respectively. This proportion was significantly lower than the rate in other countries. In the United States of America[1], diagnosis of congenital heart disease is established by 1 week of age in 40-50% of patients and by 1 month of age in 50-60% of patients. In the Baltimore-Washington Infant Study[8], at registration the cumulative proportions of infants registered under 1 week, 1 month, and 3 months were 40%, 58% and 77% respectively. From the Liverpool Registry from 1960-1969, 51.6% of the 884 live born patients with congenital heart disease presented at less than 1 month of age[9].

There were many reasons for the extremely low percentage of diagnosis being established in the neonatal period in this study. Amongst the reasons were inadequate neonatal screening, failure of registration and inclusion into the study, failure of diagnosis or failure of confirmation of clinical suspicion of congenital heart disease before death. The last two reasons could also be partially attributed to the very severe nature of the congenital cardiac lesions presenting in the early neonatal period. These are either arterial duct-dependent pulmonary or systemic blood flow lesions

where the neonates usually present acutely ill, cyanosed or with vascular collapse when the arterial duct closes. These clinical features are non-specific and may be confused with other conditions such as severe bronchopneumonia, sepsis or metabolic disorders. Clinical diagnosis of arterial duct-dependent lesions needs a high index of suspicion and confirmation of diagnosis by an echocardiography examination. During the study period, the 4 hospitals did not have access to diagnostic facilities i.e. colour doppler ECHO machine with paediatric probe and the trained personnel to perform a diagnostic echocardiography examination on the neonatal intensive care ward. The other possible reason was the lack of proper transport facilities. Many of the neonates would require ventilatory and inotropic support in addition to the prostaglandin infusion. After initial stabilization, these neonates would need to be referred to the National Heart Institute or other tertiary cardiac centres for confirmatory investigations and further palliative or definitive treatment. Many of these neonates would deteriorate on the journey and arrive at the tertiary centres very ill. It is believed that the poor general condition of the neonates during transportation and on arrival at the tertiary cardiac centres contributed to the death of these neonates either before definitive diagnosis or before/soon after palliative or definitive treatment. Studies need to be carried out to ascertain this belief. The New England Infant Cardiac Program reported that, excluding premature infants with patent ductus arteriosus, 3 infants of every 1000 live births will need cardiac catheterisation and/or surgery and/or will die with congenital heart disease in early infancy[10].

The pattern of congenital heart disease was comparable to other established data[1,9] (Table III). It is reckoned that the relative frequency of congenital heart defects varies little[11]. Differences in reported frequencies can usually be ascribed to the difference in classification, the inadequacies of enumeration or the means of ascertaining and reporting the nature of defects. Advances in diagnostic methodology would also affect ascertainment or establishment of diagnosis[11]. However, there were relatively higher percentage of acyanotic lesions and non-life threatening cyanotic heart lesions and significantly lower percentage of coarctation of the aorta and complex cyanotic lesions. The high percentage of patent arterial duct could be due to the higher survival rate of premature babies. Ventricular septal defects, including patients with isolated or multiple ventricular septal defects and ventricular defects with an associated atrial defect, patent arterial duct or some valvular abnormalities, remained the most common congenital cardiac defect seen, constituting 37% of all cardiac lesions (Table III). The low percentage of coarctation of the aorta is worrying as patients with coarctation of the aorta are usually asymptomatic and the diagnosis is usually made clinically. This could also be due to an actual low incidence of coarctation of the aorta in this part of the world.

At 6 months follow-up, 26 of the 28 patients (92.9%) with patent arterial duct had surgical ligation or transcatheter occlusion. Two (7.1%) were waiting for occlusion. In 1995, a total of 47 patients had patent arterial duct (Table III) but a total of 61 ligation or occlusion of patent arterial duct were performed in the same year (Table IV). This reflected the greater availability of these procedures in the tertiary cardiac referral centre.

However, 42% of patients with ventricular septal defects who required surgical closure i.e. open heart surgery, were waiting for surgery at 6 month review. Over an 8-month period from January till August 1995 (Group Ia), a total of 38 patients with ventricular septal defects required surgery. Extrapolation would estimate that over a period of 1 year, a total of 57 patients with ventricular septal defects would require surgery. Over the same 12 months period, there were 45 ventricular septal defect repair, 39 of which were performed within the country and 6 overseas. Therefore, at the rate of ventricular septal defect repair for the year 1995, 39/57 (68.4%) patients who required ventricular septal defects repair would have their defects closed within 6 months after presentation and 31.6% of patients would have to wait for more than 6 months for their surgery.

All Fallot's Tetralogy needs surgical correction. In the year 1995, 22 patients presented with Fallot's tetralogy. Over the same year, 23 Fallot's Tetralogy were repaired; 20 of which (90.9%) were performed in the country. Therefore, 9.1% (2/22) of Fallot's Tetralogy would not be repaired at this rate. Of the 23 patients with tetralogy repaired in 1995, only one patient presented in the year 1995 and 22/23 (95.7%) presented prior to the study year. The mean age of corrective surgery was 6.4 years which was far older than the current

recommended approach of one-staged corrective surgery in infancy or before 2 years of age[6]. Therefore, the rate for Fallot's Tetralogy repair was insufficient and there was also significant backlog of patients.

Corrective surgery were performed mainly on simple congenital heart disease. Surgery on complex congenital heart disease is still at its infancy in our country. The National Heart Institute is in its learning stage in the management of complex congenital heart disease[12]. Increased awareness and better recognition of the many complex and duct-dependent lesions would improve the initial survival of these patients and thus increase the demand for surgical or transcatheter interventional procedures further.

Limitations

Neonates suspected to have arterial duct-dependent congenital heart disease and sent to a tertiary cardiac centre might not have been captured in the study contributing to the low number of neonatal patients. At 6 months review, some patients might not return to the referring hospital after being treated but were instead followed-up at the tertiary cardiac centre. For those patients who were first seen prior to the year 1995 and referred to tertiary cardiac centre, further follow-up and treatment might have been arranged directly with the cardiac centre without going through the referring hospital, thus lost in our data capture.

Conclusion

The pattern of congenital heart disease seen in these 4 hospitals was similar to that seen world wide. There were underdiagnosis and underrepresentation of arterial duct-dependent lesions, complex cyanotic heart defects and coarctation of the aorta. This probably reflected the situation in most of the hospitals in our country during the study period. Except for patent arterial duct, access to surgical treatment was inadequate. There was also a significant backlog of patients who required interventions.

Postscript

Much has changed in scope and in the numbers of surgery and transcatheter treatments performed on children with congenital heart disease over the last few years. This study was completed in 1996. Since then, the Ministry of Health has opened 2 additional cardiac surgical centres at Penang and Johor Bahru. At Hospital Sultanah Aminah, Johor Bahru, a total of 80 patients with congenital heart disease received surgical corrections in 1998; there were 6 ventricular septal defects, 43 atrial septal defects and 31 patent arterial duct. Twenty-six patients (32.5%) were aged 10 years or below, 45 (56.3%) between 11-20 years old and 9 (11.3%) were more than 20 years old (personal communication). Meanwhile, percutaneous transcatheter umbrella occlusion of patent arterial duct is now superseded by occlusion with newer devices eg.coils or occluders. At the National Heart Institute, percutaneous closure of patent arterial duct with Amplatzer ductal occluder began in November 1997, and a total of 86 patients benefited by the end of 1998[13]. In addition, percutanteous transcatheter closure of atrial septal defects was also started in 1997[13].

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