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Diagnosis, Management and Outcome of Children with Heart Disease in a Sudanese Cardiac Centre

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Abstract: To describe the diagnosis, management and outcome of children with heart disease seen at Sudan Heart Centre (SHC). This study reviewed all Children with Heart Diseases (CHD) seen at SHC by one paediatric cardiologist from July 2004 to June 2005. Patients were evaluated clinically and by echocardiography. Cardiac catheterization was done when indicated. Patients were operated on by one paediatric cardiac surgeon from SHC and by a visiting paediatric cardiac surgeon. Five hundred and twenty two patients were evaluated, 435 had abnormal hearts. The median age was 48 months (one day-15 years). Congenital Heart Disease (CHD) constituted 87% and acquired heart disease and rhythm disorders 13%. The frequency of the different CHD was described and in general did not differ from that reported in the literature except for Ebstein anomaly which was noted to be 4 times more frequent than reported. Cardiac catheterization was done for 81 patients, for diagnosis in 61 (75%) and for intervention in 20 (25%) patients. Interventions included balloon dilatation, atrial septostomy septal defect and patent ductus arteriosus closure with the help of a visiting team, the success rate for interventions was 95%. Surgery was done for 125 patients. Seventy three operations were done by the local team and 52 by the visiting team. The 30 day operative mortality was 8.3%. Paediatric cardiac service in Sudan in echocardiography and cardiac catheterization is growing. Cardiac surgical results are comparable to the study.

Key words: Diagnosis, outcome of children, heart disease, cardiac center, SHC, CHD

INTRODUCTION

Sudan Heart Centre (SHC) is a cardiac referral centre established in the year 2000 as one of the first few centres of its kind in Sudan. In the initial period till the year 2001 the service was delivered to children by adult cardiologists. Paediatric cardiology was first started in 2001 by one part-time paediatric cardiologist. A paediatric cardiac surgeon joined the centre in 2002. In July 2004 a full-time paediatric cardiologist joined the centre and established the paediatric catheterization service. The purpose of this study is to review the diagnoses (echocardiographic and cardiac catheterization), management (medical, catheter based and surgical) and its outcome for children seen at the SHC.

MATERIALS AND METHODS

All paediatric patients with heart disease seen at SHC from July 2004 to June 2005 were included in the study. Patients were evaluated clinically and by echocardiography (echo). Cardiac catheterization was done when indicated.

Echocardiography: A complete 2-dimensional/Doppler echo study was done for each patient using MEGAS (Esaote) machine. A standard technique (segmental approach) using the 4 echo views (subcostal, parasternal long and short and suprasternal) was applied. Sedation with chloral hydrate (50 mg/kg/dose) was used in infants who need detailed initial and/or preoperative studies. A written report with printed pictures was initially used for documentation but recently a digital archiving system was established and patients going for surgery had their echo studies recorded digitally.

Cardiac catheterization: Indications for cardiac catheterization were:

- Diagnosis of anatomical lesions not well delineated by echo.
- Diagnosis of hemodynamic abnormalities.
- Treatment of anomalies amenable to interventional catheterization.

Cardiac catheterization was done as day case for diagnostic purposes in older children. Infants and

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children undergoing interventions were admitted overnight for monitoring. General anaesthesia was used for the latter group of patients while for the first group ketamine was used for sedation. Interventional septal defect occlusion was done by a visiting team, while pulmonary valve dilatation, atrial septostomy and patent ductus arteriosus occlusion was done by the local team.

Cardiac surgery: Patients selected for surgery were discussed with the cardiac surgeon in a joint meeting. Our policy is to perform definitive intracardic repair of congenital heart defects(where indicated) in patients weighing more than 6 kg; below 6 kgs our local surgical team performs palliative procedures and definitive closure of patent ductus arteriosus and coarctation of aorta.

Open heart surgery for children weighing below 6 kg and complex repairs were done by a visiting team lead by a paediatric cardiac surgeon.

Statistical methods: Patients' data were entered into an SPSS computer program and frequency tables were generated.

RESULTS

In the study, period 522 patients were evaluated. The age ranged from one day to 18 years with a median of 48 months. Male to female ratio was 1.3:1. Four hundred and thirty five patients (80%) had abnormal cardiac examination and 87 (20%) were normal. CHD constituted 378 (87%) and acquired heart disease and rhythm disorders 57(13%). Rheumatic heart disease was diagnosed in 36 patients (8.2%).

Diagnosis of congenital heart disease: Table 1 shows the frequency of CHD diagnosed on echo and Table 2 the frequency of acquired heart disease and rhythm disorders. The most common acyanotic anomalies were ventricular septal defect (16.1%), atrioventricular septal defect (8.6%), atrial septal defect (6.2%) and patent ductus arteriosus (4.6%). Tetralogy of Fallot was the most common cyanotic anomaly (17.7%) followed by transposition of great arteries (6.6%) and tricuspid atresia (3.2%). Rhythm disorders included 3 patients with congenital atrioventricular block and two patients with supraventricular tachycardia.

Cardiac catheterization: A total of 81 cardiac catheterizations were done in the study period. Table 3 shows the indications for cardiac catheterization and Table 4 the outcome and disposition of patients who underwent catheterization. Common indications for diagnostic catheterization were Tetralogy of Fallot to delineate the pulmonary artery anatomy in 16 patients (23%) and large ventricular septal defect to measure pulmonary vascular resistance in 12 patients (15%). In 2 patients the cardiac catheterization diagnosis was different from echo. In the first patient catheterization

Table 1: Congenital heart disease diagnosed on echo

Lesion	No	(%)
Ventricular septal defect	68	16.1
Small	22	
Large	46	
Atrial septal defect	27	6.2
Secundum	24	
Sinus venosus	2	
Unroofed coronary sinus	1	
Patent ductus arteriosus	20	4.6
Small	2	
Large	18	
Atrioventricular septal defect	37	8.6
Partial	16	
Complete	10	
Unbalanced	9	
With pulmonary valve stenosis	2	
Pulmonary valve stenosis	26	6
Aortic stenosis	6	1.3
Valvular	1	
Subvalvular	4	
Supravalvular	1	
Coarctation of aorta	3	0.7
Anomalous pulmonary venous drainage	4	0.9
Ebstein anomaly	12	2.1
Tetralogy of fallot	77	17.7
Tetralogy of fallot with absent pulmonary valve	2	
Transposition of great arteries	29	6.6
Intact ventricular septum	8	
Ventricular septal defect	10	
Ventricular septal defect/PS	11	
Corrected transposition of great arteries	6	1.4
Tricuspid atresia with	14	3.7
Ventricular septal defect	3	
With ventricular septal defect/pulmonary valve stenosis	10	
With transposition of great arteries	1	
Truncus arteriosus	8	1.8
Double inlet left ventricle	4	0.9
Double outlet right ventricle	12	2.7
Subaortic ventricular septal defect	5	
Subpulmonary ventricular septal defect	4	
Doubly committed ventricular septal defect	2	
Noncommitted ventricular septal defect	1	
Pulmonary atresia with ventricular septal defect	12	2.8
Isomeric heart	7	1.6
Mitral valve anomalies	3	0.7
Total	378	100.0

Table 2: Acquired heart diseases and rhythm disorders

Lesion	No	(%)
Rheumatic heart disease	36	63.15
Mitral regurgitation	17	47.5
Mitral stenosis	4	10.9
Aortic regergitation	4	10.9
Mitral and aortic regurgitation	11	30.7
Cardiomyopathy	7	12.28
Pulmonary hypertension with no CHD	6	10.52
Tumours/miscellaneous	3	5.2
Rhythm disorders	5	8.7
Total	57	100.0

Table 3: Indications for cardiac catheterization

Indication	No	(%)
Preoperative assessment	50	61
Postoperative assessment	8	10
Intervention	20	25
Other	3	4
Total	81	100

Table 4: Outcome of cardiac catheterization

Outcome measure	No	(%)
Suitable for corrective surgery	26	32
Suitable for high risk surgery	10	12
Suitable for palliation	9	11
Not suitable for surgery	6	7
Successful intervention	19	23
Unsuccessful intervention	1	2.5
For re-do surgery	5	6
Conservative treatment	4	5
Insufficient data	1	1.2
Total	81	100

Table 5: Outcome of patients with heart disease

Outcome	No	(%)
Medical treatment	25	6.7
Corrective surgery	96	25.3
Palliative surgery	28	7
Catheter treatment	19	5
Inoperable	39	10.3
Late	21	5.1
Complex	18	6.2
Elective surgery	13	3.5
No treatment needed	29	8
Awaiting surgery	56	15
Expired	12	3.2
Unknown	61	17
Total	378	100

Table 6: Outcome of surgery for congenital heart disease

Surgery	No	No of deaths	% Mortality
Local team			
Open heart	58	5	8.6
Closed heart	14	1	7
Total	72	6	8.3
International team			
Open heart	38	7	18.4
Closed heart	14	2	14
Total	52	9	17.3

Table 7: Operations for congenital heart disease done by the local team

Type of surgery	No.
Tetralogy of fallot repair	20
Ventricular septal defect closure	15
Atrioventricular septal defect repair	6
Patent ductus arteriosus closure	7
Modified blalock-taussig shunt	5
Secundum atrial septal defect closure	3
Secundum atrial septal defect /pulmonary stenosis repair	3
Relief of subaortic aortic stenosis	4
Partial anomalous pulmonary venous drainage (2 with Sinus venosus atrial septal defect	4
Ascending aorta to pulmonary artery shunt	1
Relief of supravalvular aortic stenosis	1
Relief of pulmonary stenosis	1
Pulmonary artery band	1
Permanent pacemaker insertion	1
Total	72

corrected the echo diagnosis of total anomalous pulmonary venous return to the diagnosis of transposition of great arteries with normal pulmonary venous return. The second patient had double outlet right ventricle on echo and catheterization revealed that there was, in addition, severe pulmonary valve stenosis.

Twenty patients underwent interventional catheterization including 11 cases of device occlusion (6 patients with patent ductus arteriosus, 3 with atrial septal defect and 2 with ventricular septal defects). These device occlusions were done in SHC with the help of a visiting team. Other interventions were pulmonary valve dilatation (n = 6) and balloon atrial septostomy (n = 3). One patient had an unsuccessful trial of balloon dilatation for critical pulmonary valve stenosis. This was a syndromic infant with hypoplastic pulmonary arteries where we could not place the exchange wire in the distal pulmonary artery branch. Complications of catheterization included one mortality in a 10 year old boy with pulmonary atresia and ventricular septal defect with major aortopulmonary collaterals who had multiple cerebrovascular accidents. His oxygen saturation before the procedure was 40% and he developed cardiac arrest with induction of anaesthesia. Two patients developed respiratory depression needing intubations and 2 patients had minor bleeding.

Outcome: Table 4 shows the outcome of patients who had cardiac catheterization, Table 5 shows the overall outcome of patients, Table 6 the outcome of patients who had surgery and Table 7 the details of operations done by the local surgical team. Out of 181 patients referred for surgery only 124 were operated on. Seventy two operations were done by the local team and 52 by the visiting team. The local surgeon performed 58 corrective intracardiac procedures and 14 closed procedures of which 6 were palliative. The most common open heart operations were complete repair of Tetralogy of Fallot (n = 20) and ventricular septal defect closure (n = 15); 3 of the ventricular septal defect cases had additional procedures: aortic valve replacement for severe aortic regurgitation (n = 1), mitral valve repair for an isolated anterior mitral valve cleft (n = 1) and relief of pulmonary stenosis in a patient with corrected Transposition of great arteries/pulmonary valve stenosis (n = 1). The median weight was 18.75kgs (range= 3- 45 kg) and the median age was 8 years (range = 0.4- 15 years).The total operative mortality was 8.3% (n = 6). Of these, five cases were corrective heart procedures: Tetralogy of Fallot repair (n = 1), complex ventricular septal defect closures (n = 3) and atrioventricular septal defect /pulmonary stenosis repair (n = 1) while 1 patient had a palliative operation

(Modified Blalock-Taussig Shunt). For the visiting team the median age was 19.8 months (range = 1month-14 years) and the median weight was 7.1 kgs (range = 3-45). The mortality rate was 17.3% (n = 9), 7 were open heart procedures and included transposition of great arteries with abnormal coronary anatomy (n = 1), obstructed total anomalous pulmonary venous drainage (n = 2) one of them was associated with transposition of great arteries, atrioventricular septal defect with Tetralogy of Fallot (n = 1), Tetralogy of Fallot with small pulmonary artery branches (n=1), huge patent ductus arteriosus with pulmonary stenosis in a 3 kg infant (n = 1), conduit change for a child with repaired truncus arteriosus. The closed cases (n = 2) included one modified Blalock-Taussig shunt and one complex Glenn procedure in an infant with isomeric heart.

DISCUSSION

Management of children with heart disease is expensive as it needs specialized personnel and equipment. Working in developing countries where resources are limited and trained personnel scanty is a real challenge.

Few studies were published about the incidence of congenital heart disease detected by echocardiography in Sudan. El Hag (1994) looked at 179 patients from 1991-1993 and found that CHD constituted 56% and rheumatic heart disease 39%. In Sudan, until the year 2001 echocardiography for children used to be done by adult cardiologists which lead to many limitations as the standard echo methodology (segmental approach) and nomenclature for congenital heart disease were usually not applied. During our study period we reviewed 40 echocardiograms for CHD done by adult colleagues and the diagnosis was incorrect in 45% and incomplete in 30% of patients. Only in 25% the diagnosis was accurate and in all of these patients the diagnosis was either a ventricular or atrial septal defect (unpublished data), findings that are consistent with that of Stanger *et al.* (1999) and Ward and Purdie (2001)

In this study, we described 19 echo diagnoses with their subdivisions and their frequencies, many of these lesions had not to our knowledge been investigated in Sudan. The frequency of most CHD did not differ from that published in the literature However, for Ebstein anomaly we found a frequency of 2.1%, more than 4 times the reported frequency of 0.5% in the western literature (Sulafa and Nuha, 2006).

Rheumatic heart disease continued to be the leading cause of acquired heart disease in our region, a prevalence of 3/1000 was reported in Sudan in 1992 (Ibrahim *et al.*, 1992). Compared to El Hag (1994) who reported a frequency of rheumatic heart disease of 39%, our frequency (8.2%) is much less which might reflect some improvement in socioeconomic standards. Of the 7 patients with cardiomyopathy 2 had noncompaction of the ventricular myocardium; a cardiomyopathy that we recently reported in a large series in Saudi Arabia and we think is largely under-diagnosed (Sulafa and Godman, 2004)

Indications for cardiac catheterization did not differ from literature. Catheterization corrected echo diagnosis only in 2 patients indicating the reliability of echo even for complex anomalies. Interventional catheterization was first started by the adult cardiologist then taken over by the paediatric cardiologist. Visiting teams then helped starting a program for device closure of atrial and ventricular septal defects and ductus arteriosus. Patent ductus arteriosus and atrial septal defect closure are being done but ventricular septal defect closure is a technically demanding procedure and is not planned at this time by the local team. Although the cost of these devices is high (2-3000 US Dollars) it is still comparable to the cost of surgery and non-surgical closure is now requested by many families.

Only 69% of patients requiring surgery were operated on indicating a long waiting period which is directly proportional to the socioeconomic status of the family. The total cost of surgery (4000 US Dollars) is too high for many families especially those with rheumatic heart disease. Funds had been developed that can mostly pay 50% of the cost. On the other hand 5% of our patients presented when they were already inoperable. This is not unexpected in a country where paediatric cardiac services just started but should alert the general paediatricians about the importance of early diagnosis.

The institutional 30 day operative mortality of 8.3% for congenital heart operations is consistent with figures reported by Jenkins and Gauvreau of 2.5-11.4% (median, 5.6%) for unadjusted mortality in 22 large institutions in the United States. The patients who died were all in risk category 2; the risk-adjusted mortality for the latter category is 0-8.8 % (median, 2.5%) as reported by Jenkens and Gauvreau (2002) thus yielding an acceptable mortality for our patients.

The visiting team helped to treat many children with small weight and do complex repairs like Rastelli repair, Ross-Kono procedure, arterial and atrial switch operations. In addition there was significant consolidation of the experience of our intensive care staff with management of this group of children. The relatively high mortality of the international team is directly related to the complexity of lesions of the patients who died. These operations were all done during three 10- day visits with the rate of 2-3 operations per day which added to the postoperative management difficulties.

CONCLUSION

We managed to consolidate the service of paediatric cardiology at the SHC both in echocardiography and cardiac catheterization. We can confidently perform definitive intracardiac repair of the most common congenital cardiac defects and palliate the more complex congenital cardiac defects in the well selected patients.

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