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Where There is No Echocardiography: an Overview of the Diagnosis of Paediatric Heart Diseases in a New Paediatric Cardiology Unit at the University of Port Harcourt Teaching Hospital

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Summary

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Background: The Paediatric Cardiology unit of the University of Port Harcourt Teaching Hospital (UPTH) was set up in March 2005 and a consultant paediatrician interested in Cardiology was mandated by the Paediatric Department to attend to the increasing number of patients with cardiac diseases.

Objective: To report our experience in the clinical diagnosis of heart diseases without modern diagnostic tools, in the first two years of a new Paediatric Cardiology unit.

Methods: This was a prospective study carried out between March 2005 and February 2007 using detailed history, physical examination and a Flow Table.

Results: Forty-nine children (29 males, 20 females), M: F= 1.5:1, with age range of eight days to 13 (mean 3.50±4.52) years, were seen. Ventricular septal defect was the commonest cardiac defect in 36.7 percent followed by Tetralogy of Fallot in 20.4 percent. The diagnosis was 68 percent accurate when compared with echocardiography (ECHO) diagnosis in 19 patients. Six patients had surgery, 12 were lost to follow-up, and seven (14 percent) have died. The remaining 24 are either receiving medications and/or are awaiting surgery.

Conclusion: History and clinical examination remain the mainstay of cardiac diagnosis in the absence of specialized tools such as an echocardiogram. Training of Fellows in Paediatric Cardiology and equipping of Federal and State government hospitals are of utmost necessity.

Key words: New cardiology unit, paediatric patients, echocardiography.

Introduction

THE gold standard for the diagnosis of a structural cardiac disease is an echocardiographic evaluation.[1] However, history taking and examination remain at the core of evaluation of any patient with suspected cardiac disease.[2] This is especially important in resource-poor and low technology settings of developing countries. The information obtained from these allows an immediate decision to be taken on the need for and urgency of further investigations such as chest radiography, electrocardiography (ECG) and echocardiography (ECHO) when available. Although ECHO has revolutionized the practice of Paediatric Cardiology, it is still not readily available in resource-poor countries because of its exorbitant cost. Additionally, there are very few training opportunities with consequent lack of training facilities and trained personnel for its use when available. Therefore, history taking, clinical examination and chest radiology remain the mainstay of diagnosis in the absence of an ECHO.

The Paediatric Cardiology unit of the University of Port Harcourt Teaching Hospital (UPTH) was created in 2005 with a new consultant paediatrician interested in Cardiology but who had no previous tutelage and no formal training in the subspecialty. Other members of the unit consisted of a senior

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registrar, a registrar and two house officers who rotated among other units, every 2-3 months. Prior to this time, children with heart diseases were managed under the various admitting units of the Department of Paediatrics. During the period of this report, provisional diagnoses of cardiac diseases were made from the history, physical examination and chest radiography of all patients, as there were no facilities for paediatric electrocardiography or echocardiography in the hospital. Patients whose parents or caregivers could afford the cost were sent to Ibadan, western Nigeria, for an ECHO and possible surgery in India or Israel.

Patients and Methods

This was a prospective study of all patients presenting to the Paediatric Cardiology unit having been referred to the clinic, or were seen on the wards between March 2005 and February 2007. Once suspected to have a cardiac disease, a proforma tagged 'Cardiac Recruitment Protocol' including biodata of the patient, was filled and updated continually.

The proforma contained records of presenting symptoms and timing of presentation, maternal medical history, pregnancy and birth history, gestational age, birth weight, neonatal history, and relevant family history. In addition, the presence or absence of dysmorphism, normal growth or failure to thrive, digital clubbing, oedema, presence or absence of cyanosis, character of all pulses, respiratory pattern, and normal or abnormal blood pressure values were noted on examination. Other information recorded were the presence and position of cardiac impulses, evidence of cardiomegaly, type of apical impulse (right or left ventricular), thrills, heart sounds, timing, pitch, location and radiation of murmurs when present. The murmurs were graded into innocent or pathological, using Nadas' criteria.[4] Results of investigations were also entered in the proforma. On chest radiography, the pulmonary vasculature was assessed to determine the degree of pulmonary blood flow - whether normal, decreased or increased. To arrive at a provisional diagnosis, a 'Flow Table' (Table 1) that was modified from a Flow Diagram[3] was used. The diagnosis of acquired heart disease was based on known criteria.[5] The aim was to diagnose common congenital and acquired heart diseases and commence treatment as appropriate.

The Flow Table differentiated acyanotic and cyanotic defects based on history of cyanosis, chest radiological finding of normal, increased or decreased blood flow, and the radiological and/or physical examination finding of right, left or biventricular hypertrophy. The severity of each defect was based on the history of failure to thrive, occurrence of recurrent chest infections, congestive cardiac failure (CCF), frequency of admissions, and for cyanotic congenital heart disease (CHD), history of a poor suck in infancy, worsening cyanosis with crying or defecation, exertional dyspnoea, squatting in an older child and hypoxic spells. Rheumatic heart disease was diagnosed in the presence of clinical evidence of mitral and aortic valvular disease, following present or past evidence of acute rheumatic fever using the Jones criteria.[5] The diagnosis of arrhythmias and syncope was based on a history of palpitation and or giddiness and examination finding of abnormal pulse rate and rhythm in the presence or absence of a cardiac disease. Patients who could afford to travel to Ibadan, which is about ten hours journey by road from Port Harcourt, were referred for further assessment including ECG and ECHO, and for possible recruitment for surgery in India or Israel. Others continued to receive medical treatment on inpatient and outpatient basis. Patients had medical management appropriate for their diagnoses with anti-failure regimen (frusemide, *Aldactone*, digoxin and occasionally, captoril), steroids, anti-inflammatory drugs, iron supplements and secondary prophylaxis for infective endocarditis as necessary.

Statistical analysis

Data were computed and analysed using SPSS version 11.0. The following age groups were considered: 1 to 30 days, 31 days to 2 years, 2 to 6 years, 6 to 12 years and > 12 years.[6]

Results

Forty-nine children were seen in the Unit during the period of study. Of these, 29 were males and 20 females, a ratio of 1.5:1. Their ages ranged from eight days to 13 (mean 3.50±4.52) years with a median of 10 months. Thirty (61.2 percent) of the children were aged two years or less. Seven (14.2 percent) had dysmorphic features with a diagnosis of Down syndrome,[7] two (four percent) had congenital rubella syndrome and one was diagnosed with foetal alcohol syndrome. The commonest presenting symptom was difficulty in breathing, encountered in thirty (61.2 percent) of the patients (Table II). Chest pain and palpitations were the least common symptoms noted. At least one variety of murmur was heard in 94 percent of the patients (Table III).

Of the eighteen patients diagnosed with ventricular septal defect (VSD), 55.6 percent were males. Conversely, there was a preponderance (58.3 percent) of rheumatic heart disease in females. Only two patients each were diagnosed as cases of patent ductus arteriosus (PDA) and atrial septal defect (ASD)

Table I

Flow Table for Congenital Heart Defects

Cardiac defect	*Cyanosis*	*↑BPF*	*↓PBF*	*Normal PBF*	*LVH*	*RVH*	*BVH*
Ventricular septal defect	-	+	-	-	+	-	+
Patent ductus arteriosus	-	+	-	-	+	-	+
Endocardial cushion defect	-	±	-	-	+	-	±
Atrial septal defect	-	+	+	-	-	+	-
Partial anomalous pulmonary venous return	-	+	-	-	-	+	-
Coarctation of the aorta (COA)	-	-	-	+	+	-	-
Aortic stenosis	-	-	-	+	+	-	-
Endomyocardial fibroelastosis	-	-	-	+	+	-	-
Mitral regurgitation	-	-	-	+	±	-	-
Pulmonary stenosis (PS)	-	-	-	+	-	+	-
COA (infants)	-	-	-	+	-	+	-
Mitral stenosis	-	-	-	+	-	+	=
Truncus arteriosus	+	+	-	-	+	-	+
Single ventricle (SV)	+	+	-	-	+	-	+
TGA + VSD	+	+	-	-	+	-	+
TGA +PS	+	-	+	-	+	+	+
TA + hypoplastic pulmonary artery	+	-	+	-	+	+	+
SV+PS	+	-	+	-	+	+	+
Tricuspid atresia	+	-	+	-	+	-	-
PA + hypoplastic right ventricle	+	-	+	-	+	-	-
Tetralogy of Fallot	+	-	+	-	-	+	-
Pulm vascular obstructive disease	+	-	+	-	-	+	-
Ebstein's anomaly	-	-	+	-	-	+	-

Modified from Flow Chart [3]

respectively, and both were males (Table IV). There was no case of coarctation of the aorta.

Table V shows that 57.9 percent of the 19 patients who could afford echo evaluation in Ibadan were males. Isolated VSD and tetralogy of Fallot accounted for 21 percent each of the echo diagnosis. One patient each had arrhythmias, cardiomyopathy, double outlet right ventricle (DORV) and pulmonary stenosis (PS) with patent foramen ovale (PFO). When the echo diagnoses in the 19 patients were compared with the provisional diagnoses made by clinical methods only, it was found that all cases of VSD, Tetralogy of

Table II

Presenting Symptoms of Cardiac Disease in 49 Children

Symptom	*Number of Patients*	*% of Total*
Difficulty in breathing	30	61.2
Fatigue	22	44.9
Fast breathing	20	40.8
Dark lips	15	30.6
Poor feeding	11	22.4
Excessive sweating	9	18.4
Palpitations	3	6.1
Chest pain	3	6.1

Table III

Presenting Signs of Cardiac Disease in 49 Patients

Signs	*Number of Patients*	*% of Total*
Cyanosis	5	10.2
Finger clubbing	5	10.2
Chest deformity	5	10.2
Polycythemia	5	10.2
Murmurs	46	93.9

Table 1V

Types of Cardiac Disease by Clinical Diagnosis

Type	*Male*	*Female*	*Total*	*Percentage*
VSD	10	8	18	36.7
RHD	5	7	12	24.5
Tetralogy of Fallot	8	2	10	20.4
Syncope/Arrhythmias	2	3	5	10.2
PDA	2	0	2	4.1
ASD	2	0	2	4.1
Total	29	20	49	100

RHD = Rheumatic heart disease

Table V

Cardiac Diseases Diagnosed by ECHO in 19 Patients

Type	*Male*	*Female*	*Total*	*Percentage*
Isolated VSD	2	2	4	21.1
Tetralogy of Fallot	3	1	4	21.1
PDA	1	0	1	5.26
ASD +VSD	0	1	1	5.26
VSD +PDA	1	1	2	10.5
RHD	1	2	3	15.8
Arrhythmias	1	0	1	5.26
Cardiomyopathy	1	0	1	5.26
PS + PFO	1	0	1	5.26
DORV	0	1	1	5.26
Total	11	8	19	100

DORV = Double outlet right ventricle

Fallot, PDA, rheumatic heart disease (RHD) and arrhythmias were correctly diagnosed. However, DORV was wrongly diagnosed as a large VSD with pulmonary obstructive vascular disease (POVD), PS + PFO as a VSD and the child with cardiomyopathy as having RHD. The two patients who had combined lesions of VSD +PDA and VSD + ASD were each diagnosed as a case of a large VSD. In all, there was 68.4 percent accuracy (13/19) between the clinical and echo diagnoses.

Six (31.6 percent) of the patients who had echo confirmed diagnosis have had surgery. The patients with DORV+PS, VSD+ASD, PS+PFO and Tetralogy of Fallot respectively, had surgery in India, One patient with VSD+PDA had surgery in Israel while one with Down's syndrome and ASD+VSD had surgery in America. Seven (14.3 percent) of the patients have died while 12 (24.5 percent) were lost to follow-up. All the patients who had surgery except two, are seen regularly in the clinic. One died one month after intra-cardiac repair (ICR) for Tetralogy of Fallot while riding on his bike while the second had successful surgery but had not returned to the country. The remaining 24 (59 percent) patients awaiting financial assistance for surgery are being stabilised on appropriate medical management meanwhile.

Discussion

Heart diseases constitute a major cause of childhood morbidity and mortality in Nigeria[8] and is among the top 10 non-communicable diseases according to recent studies in the country.[9,10] The incidence is largely unknown because of absence of large multi-centre studies. However, it is estimated to be high because of the high incidence of rheumatic fever in many developing countries.[11] The male preponderance in this series is similar to those reported from Nigeria[12] and other countries.[13,14] The finding that VSD was the commonest lesion seen, accounting for 36.5 percent of the cardiac disease, is similar to those reported in two other echocardiological studies reported from Nigeria in which VSD accounted for 37.5 percent[15] and 30.8 percent[12] of cases respectively, and was thus the commonest lesion encountered. The natural history and clinical course of VSD were explained to parents depending on the timing and severity of presentation since effective size of the VSD was not determined. Patients presenting in heart failure in early infancy were unlikely to have spontaneous closure. The four cases of isolated VSD who benefited from echo diagnosis in Ibadan had been correctly diagnosed without echo, while two others who were thought to have isolated VSD were

subsequently found by echo to have associated ASD and PDA.

Tetralogy of Fallot (TOF) accounted for 20.4 percent of cases diagnosed, and was the only cyanotic CHD detected in this study. The incidence was higher than the 17.1 percent[12] and 15 percent[15] recorded in northern Nigeria, probably because other causes of cyanosis such as hypoplastic left heart syndrome and TGA might have been missed and diagnosed as TOF. The importance of echocardiography in differentiating TOF from other cyanotic CHD cannot be overemphasised.[15] It is gratifying to note that the four patients who were diagnosed at Ibadan had earlier been correctly diagnosed without echo in our centre. Patients with rheumatic heart disease accounted for 24 percent of all cases. This was less than 29.5 percent[12] and 39.5 percent[15] reported earlier, probably because other reports were based on echo findings with better view of valvular involvement. However, rheumatic heart disease is still a major public health problem in Nigeria due to poor living conditions, and inadequate health care, among others. It is known to be a sequel of acute rheumatic fever which is controllable by improved living conditions and prompt and adequate treatment of sore throat.[12] It is noteworthy that 60 percent of the 19 patients who could afford echo evaluation in Ibadan were males. This is expected in this country where male preference is still high.[16]

The 68 percent accuracy of our clinical diagnosis in this study is noteworthy considering the absence of formal training in Paediatric Cardiology. This report highlights the fact that, in the absence of special tools in evaluating cardiac patients, the age long practice of history taking, physical examination and chest radiography still play a major role in effective diagnosis. Many centres in Nigeria do not and may not have such relatively sophisticated equipment for the foreseeable future because of the exorbitant cost of the machine. Even if the equipment is available, personnel will still need to be adequately trained in its use. The onus then is on the paediatrician to make the best use of the readily available methods. Referral to other centres with trained cardiologists and modern equipment is encouraged for patients who can afford it.

Conclusion

History and clinical examination remain the mainstay in the diagnosis of cardiac disease in the absence of specialised tools such as an echocardiogram. These are essential pre-requisites before a meaningful echo can be done. Training of Fellows in paediatric cardiology and adequate equipping of Federal and State government hospitals is an urgent necessity. The Federal Government should complement the efforts of non-governmental organisations like the Save a Child's Heart Nigeria while plans should be expedited to set up at least, two cardiac centres in the country.

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