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ORIGINAL ARTICLE

Pediatric Echocardiography in University of Nigeria Teaching Hospital, Ituku-Ozalla: "Real-World Cardiology – Experience from a Center in Nigeria"

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Objectives: The main objective is to showcase the overall pattern and distribution of cardiac diseases diagnosed by echocardiography and to highlight the difficulties encountered in delivering a Pediatric Cardiology/Cardiac Service at University of Nigeria Teaching Hospital. **Materials and Methods:** There were 308 children seen from July 2011 to June 2016. Their echo reports and case notes were retrieved and analyzed using SPSS version 20 (Chicago). **Results:** Their age range was from 3 days to 18 years mean (6.5 ± 4.2 years). One hundred and seventy-five (57.0%) were male and 133 (43.0%) were female with male: female ratio of 1.3:1. Of these, 211 had structural heart disease with 154 being congenital heart disease (CHD) and 57 were acquired heart disease, giving the prevalence of 0.44% and 0.16%, respectively. Isolated ventricular septal defect was highest acyanotic CHD with 44.1%, while tetralogy of Fallot was highest with 14.3% in cyanotic group. Some rare anomalies were also seen; one case of left ventricular noncompaction syndrome, Eisenmenger's syndrome, and Ebstein's anomaly representing 0.6%, respectively. The outcome was satisfactory with 35.5% receiving surgery with 7.7% mortality. The cost of procedures and dearth of interventional equipment were some of the challenges encountered. **Conclusion:** Although there exists low prevalence of echo diagnosed cardiac disease, a good number of them could not get surgical intervention due to cost.

Key words: Echocardiography, children, heart lesions, challenges

INTRODUCTION

Echocardiography has shown consistently to have a high sensitivity and specificity in diagnosing most cardiac defects.[1] Pediatric echocardiographic services became widely available in Nigeria in the early 2000's and since then most reports on the spectrum of pediatric heart disease had been either on congenital or acquired entity.[2-5]

The prevalence of congenital heart diseases (CHDs) in children is 4–10/000 live births and contributes to cardiac cause of childhood morbidity and mortality worldwide.[6-8] In Nigeria and Sub-Sahara Africa, this is confounded by the double burden of disease: noncommunicable and communicable diseases attributable to the high incidence of acquired rheumatic heart disease (RHD).

Although the overall incidence is largely unknown in much of the developing worlds, it may be twice that of developed countries.[6-8]

Pediatric echocardiography has been available consistently for over 6 years at the University of Nigeria Teaching Hospital Enugu (UNTH). Most of the equipment used was either acquired by the hospital or donated to the hospital by our overseas partners and friends and serve both adults and pediatric patients. However, difficulties ranging from patients' relation not been able to afford the fees for echo even when the fees are very meager, poor transportation facilities since

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majority of this patient come from villages where poor road network abound, lack of political will, etc. Furthermore, other challenges may even arise from the machine itself, for instance, we had the challenge of inappropriately sized probes for neonates. However, to the best of the authors' knowledge, no report of the pattern of echocardiographic findings among pediatric patients in our hospital has been published since this takeoff, 5 years ago from July 2011 to June 2016.

During this time, two standard immobile ultrasound imaging systems and a mobile system were in use. Standard two-dimensional (2D), M-mode and color flow imaging capabilities, pulsed-wave (PW) and continuous wave (CW) Doppler, and contrast echo techniques were all employed fully and provided more specific cardiac diagnoses than had hitherto been available to us.

Many other patients in Lagos, and in Nigeria, remained with their morbid conditions and had to make do with mere medical treatment of the various complications associated with their cardiac ailment including recurrent chest infections, heart failure, and endocarditis.[9,10] Earlier successful efforts at open heart surgery across the West African subregion were mainly at the UNTH, Enugu; [10] Korle Bu Teaching hospital in Accra, Ghana;[9] as well as cases done in Abidjan, Ivory Coast. Cases of congenital heart disease have been reported in our center.[11] However, much has not been done on echo diagnosis and overall pattern of heart disease in children from this perspective. This therefore necessitates the study.

MATERIALS AND METHODS

Three hundred and eight children with clinical indications for echocardiography, and whose caregivers were able to pay (N8, 500:00 [~ $24 USD] for inpatient and N10,000.00 [~ $28 USD] for outpatients) for the procedure or had an insurance coverage underwent transthoracic echocardiographic examinations.

The examinations were performed using the Hewlett-Packard model SONO 2000 Ultrasound Imaging System. The machine has a transducer with multifrequency in the range 5.5–12MHz for children, and this was used for the study. For each examination, the child laid supine or on the left lateral decubitus position. The younger patient who was not cooperative in the presence of their caregiver was pacified with toys or sedated with a mild short-acting sedative as appropriate. This was given half an hour before the procedure. Aqua sonic gel was used to ensure effective contact of the transducer with the chest wall. For each patient, intracardiac anatomy was studied using the standard 2D echocardiographic views, intracardiac dimensions, and valve motion was studied using standard M-mode technique, while valvular incompetence and cardiac shunts were qualitatively and quantitatively assessed using color flow and PW and CW Doppler techniques.[12] Some real-timed images were also taken.

In this study, lesions were classified as congenital when structural defect of the heart or great vessels are present at birth or detected later in life; isolated abnormalities when it does not coexist with any other cardiac anomaly; complex lesion when it occurs as sets of associated malformations; and acquired lesions stemming from insults on the heart occurring after birth.

The prevalence rate of diseases seen at UNTH, Enugu was estimated as the arithmetic percentage of the total number of children attending outpatient and inpatient care during the study period.

Data were analyzed using IBM SPSS version 20 (Chicago, IL, USA).

RESULTS

A total of 35, 234 children were seen at the general pediatrics clinics and wards during the study period, of which 308 children had echocardiography. Of these, 211 had structural heart disease (SHD) with 154 and 57 of them having CHD and acquired heart disease (AHD). This gave a prevalence of 0.44% and 0.16%, respectively. Their age range was from 3 days to 18 years; mean (6.5 ± 4.2 years). Of the 211 children with SHD, 120 (57.0%) were male and 91 (43.0%) were female with male: female ratio of 1.3:1.

Children with trisomy 21 were examined and 10 were found to have endocardial cushion defect and 11 had ventricular septal defect (VSD). Marfan syndrome (MS) was present in one child who had aortic root dilatation with or without aortic insufficiency as well as mitral valve prolapse with or without regurgitation.

Diagnosis by echocardiography in these 308 children is shown in Table 1.

Echocardiographic findings

Major abnormal echocardiographic findings were CHD in 154 (50.0%), with their mean age of 4.9 ± 3.0 years; AHD in 57 (18.6%) with the mean age of 7.1 ± 4.2 years.

The children with CHD were presenting later after 1 year 9 month of symptoms. There was no diagnosis of CHD *in utero*, although 95% of the mothers had ultrasound scans in pregnancy. This was done for gestational age and fetal sex assessment. A normal echo scan was seen in 70 (22.8%) with the mean age of 4.2 ± 6.3, while trivial abnormalities; minimal pericardial effusions and minimal tricuspid and pulmonary valvular regurgitation were seen in 27 (8.7%) of them. This is shown in Table 2.

Among the 154 with CHD, acyanotic CHD was seen in 111 (72.0%) and cyanotic heart disease in 43 (27.9%). Isolated

Table 1: Diagnosis by echocardiography in 308 children

Clinical indication	Male, *n* (%)	Female, *n* (%)	Total, *n* (%)
CHD (acyanotic)	74 (24.0)	52 (16.9)	126 (40.9)
CHD (cyanotic)	44 (14.2)	23 (7.5)	67 (21.8)
Adenoidal disease	41 (13.3)	14 (4.5)	55 (17.8)
Cardiac murmur	5 (1.6)	3 (0.9)	8 (2.6)
Systemic hypertension	2 (0.6)	1 (0.3)	3 (0.9)
Arrhythmia	2 (0.6)	2 (0.6)	4 (1.3)
Pericardial effusion	3 (0.9)	0 (0.0)	3 (0.9)
RHD	15 (4.8)	10 (3.2)	25 (8.2)
Preoperative - cardiac function evaluation	7 (2.8)	5 (1.6)	12 (4.4)
Others	3 (0.9)	2 (0.7)	5 (1.5)
Total	196 (63.5)	112 (36.5)	308 (100.0)

Others=DCM/endomyocardial fibrosis, 4 cases for cardiac evaluation in preopthalmic surgery, 8 cases of multiple congenital abnormalities for pediatric cardiac evaluation. CHD=Congenital heart diseases; RHD=Rheumatic heart disease; DCM=Dilated cardiomyopathy

Table 2: Summary of echocardiographic findings in 308 children and mean ages

Major findings/number of cases (%)	Male/female ratio	Mean age±SD
Normal echocardiogram/70 (22.8)	2.6:1	4.2±6.3
Trivial abnormalities/27 (8.7)	1.7:1	3.2±4.3
CHD/154 (50.0)	1.1:1	4.9±5.0
AHD/57 (18.6)	1:1	5.1±10.2
Total/308 (100.0)	1.8:1	6.5±7.2

SD=Standard deviation; CHD=Congenital heart disease; AHD=Acquired heart disease

VSD was highest in the acyanotic group in 68 (44.1%) and tetralogy of Fallot (TOF) was highest in 22 (14.3%) in cyanotic group. Some rare anomalies were seen in both categories; a case of left ventricular (LV) noncompaction in 1 (0.6), Eisenmenger's syndrome 1 (0.6%), Ebstein's anomaly in 1 (0.6%), and total anomalous pulmonary venous connection (TAPVC) in 1 (0.6%). There was no documentation of cardiac tumors in our study. This is shown in Table 3.

The outcome for these children was very good as shown in Table 4.

Rheumatic heart disease

Specific valvular and other disease in 57 children with other SHD includes RHDs was seen in 46 children (80.7%) of 57 children with AHD, with isolated magnetic resonance (MR) seen in 18 children (31.5%), the remaining 28 children had a combination of valvular lesions, mainly regurgitation, which included MR in most of the cases. Isolated tricuspid regurgitation was seen in six children (10.5%) and all have severe pulmonary hypertension, with features such as flattened interventricular septum and paradoxical septal motion.

Mitral and aortic regurgitation occurred in 10 children (17.5%), while six children (10.5%) had all the four valves affected in a regurgitant lesion. We noted 2 children (3.5%) with MS in combination with other valvular acquired lesions.

Other acquired heart diseases

Eleven other children apart from the 46 children with RHD had other structural lesions. These were dilated cardiomyopathy (DCM) in 4 children (7.1%) had marked dilated chambers with poor contractility and markedly reduced ejection fraction and fractional shortening of the LV, suggesting DCM, pericardial effusion in 5 children (8.7%); out of these, 3 of them has features suggestive of trisomy 21 and 2 others has nephrotic syndrome. It was mild-to-moderate effusion as it was mostly posterior located, with no swinging cardiac motion and no late diastolic collapse of the right atrial free wall. Hypertrophic cardiomyopathy mostly of the concentric type was seen in 2 children (3.5%). One of them had obstructive features caused by systolic anterior motion of the mitral valve against the hypertrophied septum.

Intervention/outcome

Seventy-five children of the 211 children with SHD (35.5%) benefited from open and close heart definitive surgery. Of the 75 children, most had acyanotic heart diseases seen in 46 (61.3%), these had the lowest mean age 2.1 ± 1.1 years, 20 children were from the cyanotic group (26.6%), while the remaining 9 (12.0%) children the mean age 10.2 ± 2.5 years, had various valvular repair for RHD, except for 2 of them who had valvular replacement; a 15 and 17-year-old males that had MS in combination with other lesions. Of the 46 from the acyanotic group, 25 children had VSD closure, 7 children had atrial septal defect closure, 8 had patent ductus arteriosus (PDA) ligation, while 6 had repaired for Endocardial Cushion Defect. Total correction for TOF was done in 10 children and 3 children had arterial switch operation for dextrotransposition of the great arteries (D-TGA). The highest mortality outcome was 7.7%, recorded in the acyanotic group. Almost all, 70 (93.3%) of the operation were done in the study center [Table 5].

The remaining majority of the children (64.5%) requiring surgery and cannot afford it are on follow-up and being managed conservatively. We recorded 15% default on follow-up and 35% mortality in this vulnerable group of children. The time

Table 3: Specific lesions in 154 children with congenital heart disease

Lesions	Number of patients	
	Percentage of CHD (*n*=154)	Percentage of SHD (*n*=211)
Acyanotic CHD	111 (72.0)	52.6
VSD (isolated)	68 (44.1)	32.2
ASD	6 (3.8)	2.8
PDA	16 (10.5)	7.5
ECD	10 (6.4)	4.7
LV noncompaction	1 (0.6)	0.4
Combinations		
ASD + PDA	3 (1.9)	1.4
VSD + PDA	7 (4.5)	3.3
Cyanotic CHD	43 (27.9)	20.3
TOF	22 (14.2)	10.4
DORV	9 (5.8)	4.2
D-TGA	6 (3.8)	2.8
Ebstein's anomaly	1 (0.6)	0.4
TA	3 (1.9)	1.4
TAPVC	1 (0.6)	0.4
Eisenmerger's syndrome	1 (0.6)	0.4
Total	154 (100.0)	72.3

TOF was the most common cyanotic lesion. It represented 51.1% of the cyanotic CHD and 14.2% of all the CHDs. Some rare abnormality were seem; a case of LV noncompaction in 1 (0.6); Eisenmenger's syndrome 1 (0.6%); Ebstein's anomaly in 1 (0.6%); and TAPVC in 1 (0.6%). CHD=Congenital heart disease; SHD=Structural heart disease; VSD=Ventricular septal defect; ECD=Endocardial cushion defect; OS ASD=Ostium secundum atrial septal defect; TOF=Tetralogy of Fallot; D-TGA=Dextrotransposition of great arteries; DORV=Double-outlet right ventricle; TA=Truncus arteriosus; TAPVC=Total anomalous pulmonary venous channels

from diagnosis until death would be revealing and contribute to natural history data.

DISCUSSION

Echocardiography has become the most commonly used diagnostic tool in CHD.[13] It is the investigation of choice when compared to chest radiograph and cardiac catheterization as it is noninvasive and radiation-free. It provides comprehensive information about cardiac structure/function and quantifies the severity of involvement with 95%–97% sensitivity even in asymptomatic patients. It also gives real-time results and interpretations with chances for reviews and updating.[12]

Table 4: Details of specific valvular and other disease in 57 children with other structural heart disease

Lesion(s)	Number of patients	Percentage of total
RHDs	46	80.7
MR only	18	31.5
Mitral and aortic regurgitation	10	17.5
Tricuspid regurgitation	6	10.5
Mitral, aortic, tricuspid, and pulmonary regurgitation	6	10.5
MR, MS, and aortic regurgitation	2	3.5
MR with tricuspid and pulmonary regurgitation	4	7.1
Other structural lesions		
DCM	4	7.1
HOOC	2	3.5
Pericardial effusion	5	8.7
Total	57	99.9

DCM=Dilated cardiomyopathy; MR=Mitral regurgitation; MS=Mitral stenosis; RHD=Rheumatic heart disease; HOOC=Hypertrophic obstructive cardiomyopathy

Pediatrics echocardiology in our center has recently been established and is gaining proficiency. Previous data on pediatrics heart diseases were from adult cardiology, with some competency in pediatrics echocardiography. Hence, this gives an opportunity to reassess the pattern of SHDs in our center and to compare it with local and global patterns. Male preponderance was found in our study, and this is similar to some local reports;[2-5,11,14] however, some international studies showed almost equal gender distribution.[6-9] This may be due to some genetics interplay or bias of male preference in health-seeking behavior in some African regions.

SHD which includes CHD and AHD represents one of the major causes of cardiovascular disease morbidity and mortality among African children. Our study found a prevalence of 0.44% and 0.16% for CHD and AHD, respectively. The higher prevalence of CHD and low prevalence of AHD in our study is in contrast with some local studies and that of other countries in the region[2,8,15,16] and compares to a few local studies and that of Western countries.[6,10,14]

The reduction in the prevalence of AHD is interesting as the age of study population and epidemiological risk factors, such as poor socioeconomic factor, which favors the spread of Group A β-hemolytic streptococcal throat infection and rheumatic fever (RF) still exist.[2,14] However, there seems to be changing trend as some recent hospital-based studies found a high prevalence of RF in the Northern parts of Nigeria but declining prevalence in the South.[5,10,17] This may be attributed to improvement of health services in the South.

Table 5: Profile and outcome of 65 children who had surgery

Type of lesion/Number cases (mortality)	Male/female ratio	Mean age±SD	Outcome mortality (%)
Acyanotic CHD/39 (3)	1.7:1	2.1±1.1	7.7
Cyanotic CHD/17 (1)	1.5:1	5.6±2.0	5.8
Valvular (RHD)/9 (0)	2.0:1	10.2±2.5	0.0
Total/65 (4)	1.7:1	6 0.8±3.2	6.1

SD=Standard deviation; CHD=Congenital heart disease; RHD=Rheumatic heart disease

Of the children with CHD, the spectrum of conditions in our study is similar to that from other parts of Nigeria,[2-4,10,14] Africa, and the world,[6,18,19] with VSD and TOF being the most common acyanotic and cyanotic heart lesions. These conditions represent those that allow survival beyond infancy. Surprisingly, a few with high mortality such as d-TGA were also observed. They do not usually survive and many of these early infant deaths are erroneously diagnosed as pneumonia or "sepsis;" hence, underestimating the prevalence of cardiac disease in Nigerian Children.

The six cases of d-TGA were all above 7 months and 2 of them were 5 and 9 years, respectively. Surprisingly, these two older children with d-TGA had small ventricular shunt, which has been known to prolong survival though. Before this modern era of corrective surgery, mortality was > 90% in the 1st year.[20] The place of 2D echocardiography cannot be overemphasized in differentiating the condition from double-outlet right ventricle (DORV), with the demonstration of transposed ventricular-arterial connections in d-TGA and aortomitral discontinuity in DORV. There was no documentation of cardiac tumors in our study as supported by other local and regional studies.[2-6,10,11]

The late presentation found in our study was also noted by some authors.[11,14] This may be attributed to poor health-seeking behavior and financial constraints, coupled with delayed diagnosis at the secondary center, reflecting the paucity of cardiac services at secondary level in Nigeria. Some of our children had been misdiagnosed and treated for recurrent or chronic respiratory tract infections rather than cardiac failure. Delayed presentation may also be due to dearth of personnel with competencies in prenatal diagnosis of CHD, as 95% of the mothers had ultrasound scans in pregnancy, but none was diagnosed prenatally. It was done for gestational age and fetal sex assessment. Late presentation has implication for prognosis, as complication of the disease may have occurred. For example, in this study, we unfortunately documented a case of Eisenmerger's syndrome in an 11-year-old female with large 7-mm PDA shunting right to left with cyanosis. She had been managed for recurrent chest infection at a secondary health facility before her presentation. She is currently on conservative treatment as her case is inoperable due to suprasystemic pulmonary pressures, when she could have benefitted from off-pump PDA ligation.

Preventive strategies based on increasing health-seeking behavior by creating community awareness, financial empowerments, and improving diagnostic yield at the secondary center through their adoption by tertiary centers, for training and retraining will help to reduce late presentation. Furthermore, a multidisciplinary approach, with involvement of pediatric cardiologist for routine fetal cardiac screening in the perinatal period will aid early detection and intervention.

The finding of adenoidal disease as an indication for 2D Echo is of interest and had not been noted by earlier studies.[2,10,11] This may be a new trend by the ear–nose–throat surgeons for preoperative assessment and should be encourage. It aids early detection of cardiac involvement in obstructive sleep apnea and for follow-up if there was an evidence of pulmonary hypertension preoperatively. The finding of dysmorphic features associated with CHD in our study had been observed by some authors.[2,4,10,14] This may be due to genetic influences and can be a pointer for routine cardiac screening in these populations. We documented some rare cardiac anomalies, for example, include a case of LV noncompaction, Eisenmenger's syndrome, Epstein's anomaly, and TAPVC, which were rarely reported in our environment.[2,4,10,14]

The number of definitive surgeries and 30-day mortality rate outcome was 35.5% and 7.7%, respectively, showing an improving trend when compared to previous reports locally and in Sub-Saharan Africa.[15,20] This improving trend is evidenced by the previous work done by Bode-Thomas *et al*. 14 years ago, which noted only minimal off-pump definitive surgeries were done as at that time and most heart surgeries where done outside Nigeria.[2] Again a more recent study by Chinawa *et al*. in 2013 noted that only 6.2% of children had some form of surgery, however, mortality was not documented.[12] Sani *et al*. in Sokoto in 2015 noted an increase to 9.8% of cardiac surgeries, with 16.7% mortality, [4] while Falase *et al*. in noted a 17.6% mortality rate.[21] Kennedy and Miller in 2013 from Malawi noted minimal cardiothoracic surgical service in Malawi, which performs only off-pump PDA ligation at an earlier age.[8] Our improving trend may be attributed to increasing competencies in cardiothoracic services, enhanced cardiac facilities, and collaborative efforts of our local team with foreign partners on a medical mission basis. We are expectant that in the nearest future, this transfer of skills will reach the full solution stage, where tertiary cardiac services will be provided mainly by our team.

The major management challenges are inadequate financing, continuing human resources development, and dearth of diagnostic and interventional equipment. Inadequate

financing severely limited the number of operations that can be done. This is a call to our health policy-makers to scale-up the coverage of National Health Insurance Scheme to informal sector and to include such special cases as treatment of congenital abnormalities/organ transplant, terminal illnesses, e.g., cancer and chronic renal failure, as these had been on total exclusion list.[22-24] This will help prevent the scourge of our double burden of disease; prevalent communicable disease and increasing prevalent of noncommunicable diseases such as CHD in our health system.

Another challenging issue was establishing standard International Normalized Ratio (INR) for anticoagulation. Three out of 9 of the children who had valve replacement for RHD had this issue. Their INR varies from laboratory to laboratory on the same blood sample, hindering optimal dosing adjustment for warfarin. Our hematologists are leaving no stone unturned in this regards, and it calls for regulation and setting of a standard operational procedure for the laboratories, with a reference laboratory.

Limitation

This work would have been more elaborate and ideal if it involves many other cardiac centers.

CONCLUSION

Although there exists low prevalence of echo-diagnosed cardiac disease, a good number of them could not get surgical intervention due to cost.

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Conflicts of interest

There are no conflicts of interest.

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