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ORIGINAL RESEARCH

An echocardiographic study of Rheumatic Heart Disease amongst children in a Tertiary Centre

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Abstract

Background: Rheumatic Heart Disease (RHD) is a major form of acquired heart disease amongst children in developing countries, where it continues to be a cause of childhood morbidity and mortality.
Objective: To describe the prevalence and spectrum of valvular affectation in children with RHD from the echocardiography laboratory.
Methods: The records of echocardiographic scans conducted over a 10-year period in a tertiary health facility were reviewed. Children with RHD, diagnosed using standard criteria were identified. The pattern and severity of valvular involvement and other associated cardiac abnormalities were recorded.
Results: Forty-one of the 2742 (1.5%) children who had echocardiography had RHD. Their mean age was 10.9±3.1 years; 28 (68.3%) were aged ≥10 years while 21 (51.2%) were males. Mitral valve was the predominantly affected valve in 40 (97.6%) children and mitral regurgitation was the commonest valvular abnormality in 19 (46.3%) cases. Nineteen (46.3%) had severe valvular damage, 5 (12.2%) had pericardial effusion while 7 (17.1%) died. Only 2 (4.9%) children had valvular repair surgery.
Conclusion: Although the prevalence of RHD in this study is low, it is noteworthy that RHD still affects Nigerian children. The poor access to surgery emphasizes the need to strengthen both primary and secondary prevention of RHD while enhancing facilities for surgical intervention.

Keywords: Acquired Heart Diseases, Children, Echocardiography, Rheumatic heart disease, Valvular disease.

Introduction

Rheumatic Heart Disease (RHD) is a cardiac valvular complication of poorly managed rheumatic fever (RF) which is a post-infectious outcome of pharyngitis caused by Group A β-Haemolytic Streptococcus in genetically susceptible individuals. [1] The valvular damage usually occurs from repeated RF but may occur after a single episode. The damage affects mostly the mitral and aortic valves leading to complications such as heart failure and thromboembolic phenomena. [2]

In developing countries, RHD is a common cause of acquired heart disease, especially in sub-Saharan Africa. [2,3,4] In 2015, it was estimated that RHD affected about 33 million

people worldwide and was responsible for 319,000 deaths that year. [5] If untreated, RHD is a chronic debilitating condition with progressive valvular damage and eventual death. With mild disease, penicillin prophylaxis prevents further damage. However, severely damaged valves require either surgical repair or valvular replacement, often at huge costs. [5]

RHD is prevalent among the poor, and where overcrowding with poor personal and environmental hygiene occur. [6] With the improvement in living conditions and environmental sanitation in the developing world, coupled with the improved access to healthcare and implementation of control programmes, the prevalence of RHD will continue to decline in affected communities. [7]

In Nigeria, there have been various reports on the prevalence of RHD among children in the echocardiography laboratories. [4, 8] In the study reported by Bode-Thomas and co-workers [8] in children with RHD in Jos, Nigeria, RHD was the commonest acquired heart disease, accounting for 57.7% of acquired heart diseases. In a multi-centre study involving Benin City, Abuja, and Lagos, RHD was the third commonest acquired heart disease after myocarditis/dilated cardiomyopathy and pericardial disease. [4] These findings may suggest that the burden of RHD varies in different localities.

In this retrospective study, the prevalence of RHD among children presenting for echocardiography in a tertiary health facility serving Benin City, Edo State, and environs, was determined. The study also described the pattern of valvular involvement and other echocardiographic findings in children with RHD.

Methods

This study was a retrospective review of the records of paediatric echocardiographic scans performed between January 2008 and December 2017; a ten-year period. Every child who had an echocardiographic scan had their biodata and other socio-demographic information in the scan report. These information were also documented in a database (Microsoft® Excel spreadsheet) in a computer meant for that purpose.

The echocardiography was done with two machines: Aloka ProSound SSD-4000 SV® (Aloka, Meerbush, Germany) was used between 2008 and 2011, while Phillips HD7 XE® (Phillips, Bothel WA, USA) was used between 2012 and 2017. The machines were equipped with probes with a frequency range between 3.5 and 8 MHz. Each subject had echocardiographic interrogation using the two-dimensional (2D) studies according to the recommendations of the American Society of Echocardiography (ASE). [9] M-mode and Doppler studies were also done according to the recommendations of ASE. [10,11]

Images were acquired from the apical, subcostal and the parasternal windows to study the heart valves, notably the left-sided heart valves. Regurgitations and stenosis of the mitral and aortic valves were recorded. The following morphologic abnormalities of the valves were noted: restricted mobility of the leaflets, focal or generalised valvular thickening, coaptation defect prolapse and thickening of the subvalvular apparatus. The diagnosis of RHD into the definite or borderline disease was based on the recommendations of the World Heart Federation. [12] The diagnosis of definite RHD was made when there was pathologic mitral regurgitation with at least two morphologic features of RHD of the mitral valve, mitral stenosis mean gradient ≥ 4mmHg, pathologic aortic regurgitation with at least two morphologic features of RHD and borderline disease of aortic and mitral valves. Pathologic mitral and aortic regurgitations were defined

by regurgitant jets that are pan-systolic, visible from two different views, with a velocity of ≥3m/s and jet lengths of ≥2cm and ≥1cm for mitral and aortic regurgitations respectively. Borderline RHD was diagnosed in the presence of at least two morphologic features of RHD of mitral valve without pathological mitral regurgitation or mitral stenosis when there was pathological mitral or aortic regurgitation.

The quantification of the valvular disease into mild, moderate and severe was based on the recommendations of the American College of Cardiology/American Heart Association's quantification of valve diseases. [13] Based on the colour Doppler jet area, Doppler vena contractile width, regurgitant orifice area, left atrial and ventricular enlargement for severe disease and the mean gradient across the mitral valve for mitral stenosis. The predominantly affected valve was either mitral or aortic. In situations where both valves were involved, the valve with the more severe damage was the predominantly affected valve. The diagnosis of pulmonary hypertension was made when the pulmonary systolic pressure was >35mmHg, [14] while pericardial effusion was diagnosed in the presence of echo-free space between the pericardium and epicardium of the heart. Heart failure was diagnosed based on the fulfilment of the diagnostic criteria described by Omokhodion and co-workers. [15] Arrhythmia was detected using electrocardiographic tracings. A socio-economic class (SEC) was assigned to each subject using the methods described by Olusanya *et al.* [16]

Statistical analysis

The echocardiographic scan data entered into the Microsoft® Excel spreadsheet was transported to IBM-SPSS 20.0 (Chicago IL) spreadsheet with which the statistical analysis was done. The results were presented as frequencies and percentages. Continuous variables were presented as means and standard deviations and the differences in means were tested with Student's t-test while differences in proportions were compared using the Chi-Square test. *P* values <0.05 were considered statistically significant.

Results

Over the study period, 2742 children had echocardiography, 41 had definite RHD, giving a prevalence of 1.5% or 15/1000 children. These 41 children comprised 20 (48.8%) males and 21 (51.2%) females. The mean age of the children was 10.9±3.1 years with a range of 5-17 years. Thirteen (31.7%) of the children were aged <10 years while 28 (68.3%) were aged ≥10years. The mean age of the males, 9.3±2.6 (range: 5 – 15) years was lower than that of the females, 12.5±2.7 (range: 6 – 17) years; p <0.0001. Twenty-seven (65.9%) children belonged to the low socioeconomic (SEC) class. Table I shows the SEC distribution and other socio-demographic characteristics of the children.

Pattern of valvular involvement

The predominantly affected valves were the mitral valves in 40 (97.6%) and aortic valves in 1 (2.4%). Mitral regurgitation was the commonest valvular disorder in 19 (46.3%) children, followed by mitral stenosis in 9 (22.0%). The distribution of the other valvular abnormalities is shown in Table II. Eleven (26.6%) children had mitral and aortic valve involvement while there was no abnormality of the tricuspid and pulmonary valves.

Severity of the valvular damage

Of the 41 children, 19 (46.3%) had severe valvular damage, 17 (41.5%) had moderate damage while 5 (12.2%) had a mild variety. The distribution of the severity of valvular damage according to the age category is shown in Table III. The older children had worse valvular damage compared to the younger children (p = 0.002).

Table I: The age group and socioeconomic distribution of the study population

Characteristics	Male	Female	Total	*P* values
	n (%)	n (%)	n (%)	
Socioeconomic Classes				
High	4 (20.0)	1 (4.8)	5 (12.2)	
Middle	6 (30.0)	3 (14.3)	9 (22.0)	0.10
Low	10 (20.0)	17 (80.9)	27 (65.8)	
Age				
< 10 years	11 (55.0)	2 (9.5)	13 (31.7)	
≥ 10 years	9 (45.0)	19 (90.5)	28 (68.3)	0.002

Table II: Spectrum of valvular abnormalities in the children studied

Valvular abnormality	Frequency	Percentage
Isolated MR	19	46.3
MR + MS	9	22.0
MR + AR	7	17.1
MR + MS + AR	2	4.9
Isolated MS	1	2.4
Isolated AR	1	2.4
MS + AR	1	2.4
MS + AS	1	2.4
Total	41	100.0

MR - Mitral regurgitation, MS - Mitral stenosis, AR - Aortic regurgitation, AS - Aortic Stenosis

Other findings on echocardiogram

Five (12.2%) children had pericardial effusion; 2 (4.9%) of them requiring drainage while 3 (7.3%) with severe mitral valve disease had pulmonary hypertension. The mean fractional shortening (FS) and ejection fraction (EF) were available for 31 (75.6%) children. The mean FS was 30.5±9.2% (range: 11.9 – 45.3%) while the mean EF was 60.4±11.7% (range: 36.6 – 83.0%) Six (19.3%) of the 31 children had low FS and EF values.

Co-morbidities

Twenty (48.81%) children had congestive heart failure during the echocardiographic scan, while one (2.4%) each had Patent Ductus Arteriosus (PDA) and Atrial Fibrillation. The subject with atrial fibrillation was a 13- year old girl, whose arrhythmia responded to digoxin and was also placed on low dose aspirin. The subject with PDA was a 6-year old girl, who had thickened anterior mitral valve and mitral regurgitation.

Table III: Relationship between severity of valve damage and age category

Age Group (years)	Severity of valvular damage			*P* values	
	Mild	Moderate	Severe	Total	
	n (%)	n (%)	n (%)	n (%)	
< 1 0	5 (100.0)	4 (23.5)	4 (21.1)	13 (31.7)	0.002
> 10	0 (0.0)	13 (76.5)	15 (78.9)	28 (68.3)	
Total	5 (100.0)	17 (100.0)	19 (100.0)	41 (100.0)	

Outcome

Only 2 (4.9%) of the 41 children had surgery. Both had severe combined mitral and aortic valve diseases and had mitral and aortic valve replacements. Seven (17.1%) mortalities were recorded while 18 (43.9%) were lost to follow-up. Of the 18 that were lost to follow up, 8 (44.4%) had severe disease. Sixteen (39.0%) of the subjects are still being followed up in the clinic.

Of the seven that were known to have died, 2 (28.6%) were males and 5 (71.4%) were females. The mean age was 12.9±2.1 years. This age was not statistically significantly different from the age of those that lived 10.5±2.8 years, p = 0.067. The differences in the mean fractional shortening (FS) and ejection fraction (EF) between those who died and those that survived were significant, p = 0.048 and 0.017 respectively. Six (85.7%) of those that died, had severe valvular disease requiring surgery.

Table IV. Comparison of the mean ages, fractional shortening and ejection fraction between children who died and those that survived

Parameters	Died	Survived	*P* values
Mean Age (years)	12.9±2.1	10.5±2.8	0.067
Mean FS (%)	24.4±7.5	33.5±7.6	0.048
Mean EF (%)	51.4±13.0	63.9±10.1	0.017

FS = Fractional shortening, EF = Ejection Fraction

Discussion

The prevalence of RHD of 1.5% amongst children presenting for echocardiography in this study is lower than other similar hospital-based echocardiographic studies conducted amongst Nigerian children. In the study by Sani *et al,* [17] in Kano City, 9.8% of children presenting for echocardiographic scan had RHD. This value is larger than what was obtained in the present study. Similarly, a higher value of 17.4% was reported in another hospital-based echocardiographic study conducted in Jos, Nigeria by Bode-Thomas and co-workers [8] in children with RHD. The difference in prevalence rates of RHD between the Kano and Jos studies in comparison with the current study suggests a low RHD burden amongst children in Benin-City metropolis.

Previous studies conducted among children in Benin-City metropolis suggested a low level of Group A β Haemolytic Streptococcal activities; the organism responsible for the causation of rheumatic fever and subsequently, rheumatic heart disease. Two earlier studies that evaluated Group A β Haemolytic Streptococcal throat carriage amongst school children in Egor Local Government Area (LGA), part of the Benin metropolis, did not identify a single case of Group A Streptococcal throat carriage. [18, 19] This was at variance with the finding of Group A Streptococcus among children with a sore throat in Abeokuta, Nigeria. [20] Furthermore, in a community-based study that screened primary school children within the Benin-City metropolis, using the auscultatory method with echocardiographic confirmation of suspected cases for RHD, a low prevalence of 5.7/100,000 was reported. [21] The latter further buttressed the low Group A Streptococcal activity and therefore, the low burden of RHD in the area.

The low prevalence of RHD in this study compared to the rates reported from other centres may be due to the improvements in the living conditions in Benin-City, with less crowded houses and better personal and environmental hygiene. Another plausible

reason for the low prevalence of RHD in the present study could be the ready access to antibiotic treatment of sore throat in children in the study locality. This practice has been demonstrated in the study locality in an earlier study, [22] in which health workers were reported to prescribe various antibiotics to children with a sore throat. [22] Another study [23] demonstrated parental pressure for an antibiotic prescription for their children with a sore throat. It may be argued that the difference in prevalence between this study and those from Jos and Kano cannot be wholly accounted for by the improvement in living conditions, as the improvement cuts across the nation. It is also possible that other yet to be identified characteristics in the children and the environment could be at play. It is possible that genetic factors may have contributed to the observed differences as there are reports of genetic influences on the inheritance of RHD and the association of certain Human Leukocyte Antigens with the risk of developing rheumatic fever and RHD. [24]

There was no gender difference among the subjects in this study; this is at variance with other studies that have shown female preponderance. [2,17] The females in the present study were significantly older than the males. The reason for this finding is not obvious. It could be due to delay in the presentation of the female children to health facilities as male children are often preferred in our culture, prompting early presentation when they are ill. There were significantly more subjects aged ten years and older compared to those aged less than 10 years. This finding is likely due to the fact that RHD is a chronic disease, whose course worsens with time. The damage to the heart valves tends to occur from repeated rheumatic fever episodes, thus the disease becomes more manifest with age.

Mitral valve disease was the commonest valvular lesion in this study and it is consistent with the findings in other studies. [2,4,8,17,21] There was no child with pulmonary valvular involvement in the present study. The absence of pulmonary valvular abnormality is similar to observations made in previous studies of children with RHD. [8] However, functional tricuspid valvular regurgitation and damage to the pulmonary valves have been reported in studies involving adults. [17] Since RHD is a chronic disease, the condition is prone to complications as the disease progresses. The commonest complication seen in the present study was heart failure. Heart failure is prevalent in other series, especially in studies involving adults. [2,8,17] Other complications observed in the present study included pericardial effusion and pulmonary hypertension which tend to occur as the disease progresses. The prevalence of atrial fibrillation in the present study is low compared to other studies conducted among children. It would appear that the complications tend to set in as the disease advances. The low prevalence of atrial fibrillation may explain the paucity of embolic phenomena in this study since atrial fibrillation predisposes to embolism.

The low proportion of children who had surgical intervention speaks to the fact that it is a disease of the poor, as shown in the high proportion of patients belonging to the low SEC. These are families who cannot afford the high cost of valvular repair or replacement. This observation buttresses the need for governmental and non-governmental assistance for surgical interventions in children with surgically-amenable heart diseases. The mortality of 17.0% recorded in this study may be attributed to the late presentation of children with the disease, a common practice in our environment and the unaffordability of surgical treatment by the sufferers as depicted by the low number of subjects who had surgery. Late presentation leads to more damage and advancement in the severity of the disease which is associated with poor cardiac functions as exemplified by significant lower EF and FS in the children that died compared to those who survived.

Therefore, this study may have demonstrated that low EF and FS are associated with mortality in children with RHD.

The proportion of children lost to follow-up in this study is high. This may have been due to a number of reasons: some patients residing outside the state of study (Edo State) might have sought care in other facilities close to them and inability to afford the cost of transportation and care as 65.8% of patients in this study belonged to low SEC. Some children with severe valvular disease may have died outside the study centre since 44.4% of them had severe disease.

Conclusion

The prevalence of rheumatic heart disease among children presenting for echocardiography in this tertiary centre is low compared to reports from similar studies done in other parts of the country. Mitral valve disease is the commonest disorder while most of the children were older than 10 years of age. Low EF and FS are associated with mortality in children, who mostly have poor access to intervention. This buttresses the need for improved governmental and non-governmental efforts to increase access to surgical interventions in children with RHD.

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