

Original Article

Presentation and outcome of patients managed for Rheumatic fever and Rheumatic heart disease at Ahmadu Bello University Teaching Hospital Zaria.

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Abstract

Background: Rheumatic heart disease occurs as a sequel to acute rheumatic fever. Many cases of acute rheumatic fever, however, go undiagnosed as many patients present with otherwise common symptoms and in the absence of overt carditis may go undetected. Most cases present late following the development of valvular damage. A high index of suspicion is key in managing this disease. **Aim:** To determine the Presentation and outcome of patients managed for Rheumatic fever and Rheumatic heart disease at Ahmadu Bello University Teaching Hospital Zaria. **Methodology:** A retrospective study was done using information collected from patient records from the Paediatric cardiology ward and clinic of Ahmadu Bello University Teaching Hospital Zaria over 4 years from March 2015 to March 2019. **Results:** Eight per cent of the total patients seen in the cardiopulmonary clinic had rheumatic heart disease. Fifty-one patients were diagnosed with rheumatic heart disease, forty-five patients presented with established chronic rheumatic valvular involvement and six had acute rheumatic fever. The mean age (\pm SD) at presentation was 10.7 ± 3.2 years. There was a slight female preponderance M: F 1:1.3 and most patients (88 percent) presented late (already having valvular disease). Most patients had mixed mitral valve disease. The clinic dropout rate was 39 percent while the mortality rate was 25 percent. All patients received medical management, the mortality rate was 25 % (13 patients) while only 3.9 per cent received successful surgical management. **Conclusion:** Rheumatic heart disease is the commonest preventable and curable type of acquired heart disease, yet the outcome of care provided in our settings remains poor. There is a need to increase awareness, diagnosis and available treatment of rheumatic heart disease.

Keywords: *Acute rheumatic fever, Outcome, Presentation, Rheumatic heart disease.*

Introduction

Rheumatic fever is a disease caused by group A β -hemolytic streptococcus (GAS) with a sequela of global health significance; rheumatic heart disease (RHD).¹⁻³ The pathophysiology is believed to be explained by molecular mimicry and autoimmunity between the M proteins of the GAS organism and human cardiac myositis leading to carditis. The damage to the heart in RHD may occur following single or multiple infections by GAS on a susceptible host.^{2,3}

Rheumatic heart disease is the most common acquired cardiovascular disease in young people less than 25 years old⁴ and is a disease of poverty that can be prevented.³⁻⁶ Many cases of acute rheumatic fever however, go undiagnosed as many patients present with otherwise common symptoms such as malaise and joint pains and

in the absence of overt carditis may not be detected without high suspicion.^{1,3,6} Most patients present late following the development of valvular damage. The effect of RHD results in poor quality of life, with high morbidity and mortality in low-income countries where the cost of care and availability of needed sophisticated management is scarce or not affordable.

Globally it was estimated that there were 9.6 million to 11.5 million disability-adjusted life-years wasted due to rheumatic heart disease.⁵ The highest morbidities and mortalities were seen in Oceania, Asia and Central sub-Saharan Africa.⁵ West Africa an endemic area for Rheumatic fever had the 7th highest morbidity and mortality recorded.⁵

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Rothenbühler et al⁷ in a meta-analysis of screening studies showed the prevalence of clinically manifest rheumatic heart disease (detected on auscultation) in low-income and middle-income countries of 2.7 cases per 1000 population while clinically silent RH disease (detected by echocardiography) was higher at 21.1 cases per 1000 population. Nigeria is considered an endemic country for rheumatic fever and rheumatic heart disease where an estimated childhood mortality rate of 0.15 million deaths occurs among 100,000 children aged 5-9 years.⁵ This implies that there are far more clinically unmanifested cases in the community. This is buttressed by the study carried out by Ekure et al⁸ in the Niger Delta region of Nigeria who found in a prospective survey a prevalence of RHD of 4.3/1000 following auscultation and 6.5/1000 on echocardiography in children aged 5 – 15 years.⁸ This is very significant in the face of the morbidity and mortality attached to this preventable and curable disease.

This study aims to determine the Presentation and outcome of patients managed for Rheumatic fever and Rheumatic heart disease at Ahmadu Bello University Teaching Hospital Zaria

Material and Methods

A retrospective study was done using information collected from patient records from the Paediatric cardiology ward and clinic of Ahmadu Bello University Teaching Hospital Zaria over 4 years from March 2015 to March 2019. Case notes of children attending the Paediatric cardiology clinic managed for rheumatic fever/rheumatic valvular heart disease were retrieved and reviewed. Information extracted included the biodata, socioeconomic data, clinical presentation and diagnosis as recorded by the attending physician, and laboratory results.

Laboratory investigations included the presence of evidence of streptococcal infection, the erythrocyte sedimentation rate and immunological parameters; ASOT and Rheumatoid factor.

Echocardiographic features included the structural and functional valvular and sub-valvular involvements. Complications of RHD found in the records were included such as the presence of atrial fibrillation, mitral valve prolapse, presence of thrombus and infective endocarditis.

Data was analysed using the software SPSS statistics version 22.0 and presented as means, standard deviations, frequency tables, and percentages.

Results

Demographics

The study showed a mean age of 10.7 ± 3.2 years with an age range of 4 to 18 years. There was a slight female preponderance M: F 1:1.3 which was not statistically significant ($p = 0.7$). About half (51%) of the cases were children aged 10 to < 15 years.

Presentation

Two patients presented with Sydenham's chorea (table 1), one boy and a girl. The latter also had subcutaneous nodules and erythema marginatum (figure 1).

Investigations

Of the fifty-one RHD subjects, only 24 (47.1%) were able to afford an echocardiogram. All subjects had mitral regurgitation (MR). Of the five subjects with tricuspid regurgitation two had moderate TR and three had severe tricuspid regurgitation (TR), one of the subjects with severe TR also had thickening of the tricuspid valve with reduced motion while the other 4 subjects with TR had annular dilatation (Table 2).

Complications

Complications in RHD subjects are shown in Table 3. Two subjects had mild pulmonary hypertension (PHTN), 3 had moderate PHTN and 3 had severe PHTN. Two patients had vegetations and 3 had spontaneous echo contrast in the dilated chambers. The mean age of the subjects with mitral stenosis (MS) was significantly higher than those without MS (13.7 ± 3.8 years; 10.2 ± 3.7 years respectively, $p = 0.0433$).

Two patients were managed for infective endocarditis and 4 had rhythm abnormalities (1 with 1st-degree heart block, 1 with a second-degree heart block and 2 had atrial fibrillation at presentation).

Secondary prophylaxis

All 51 subjects with RHD were placed on penicillin prophylaxis as soon as diagnosis was made. Compliance was, however, noted to be poor as only 22 (43.1%) were regular in receiving their monthly Benzathine penicillin injections. Two of these subjects had to be placed on 3 weekly injections following repeated attacks of rheumatic fever (RF). The use of oral penicillin V in five patients was met with poor compliance due to forgetfulness and irregular availability of the drug and all had to be reverted to intramuscular benzathine penicillin. The remaining 56.9% of the subjects were irregular in taking either enteral or parenteral prophylaxis mainly due to a lack of funds to procure drugs and for transportation to the clinic leading to defaults.

Outcome

Seventeen patients (33.3%) were still on follow-up. At the time of the study, the duration of follow-up varied ranged from 1 day to over 8 years. The clinic drop-out rate

was 39 % (20 patients) while the mortality rate was 25 % (13 patients). One patient (2%) was referred to another facility. All patients (100%) received medical management while only two (3.9%) received surgical management and have been doing well at the time of this study.

Table 1: Clinical presentation of RF/RHD subjects

Presentation	Parameter	Frequency n (%)
Clinical	Carditis	
	Fever	32 (62.7)
	Polyarthritits	12 (23.5)
	Sydenhams Chorea	2 (3.9)
	Subcutaneous Nodules	1 (2)
	Erythema Marginatum	1 (2)
Laboratory	Raised ESR	14 (27.5)
	Raised Aso Titre	10 (19.6)

RHD: Rheumatic heart disease, CCF: congestive cardiac failure, ESR: erythrocyte sedimentation rate, ASO: antistreptolysin O

Table 2: Echocardiographic findings in RHD subjects

Valve	Frequency (n)	Percentage (%)
MR only	16	88.9
MR, MS	8	44.4
AR	3	16.7
TR	5	27.8
PR	1	5.56

MR: mitral regurgitation, MS: mitral stenosis, AR: aortic regurgitation, TR: tricuspid regurgitation, PS: pulmonary regurgitation, RHD: Rheumatic heart disease.

Table 3: Complications in RHD subjects

Complication	Frequency (n)	Percentage (%)
CCF	49	96.1
PHTN	8	15.7
Arrhythmia	4	7.8
Vegetations	2	4
IE	2	4

PHTN: pulmonary hypertension, IE: infective endocarditis



Figure 1: Patient (add the Initials) with erythema marginatum.

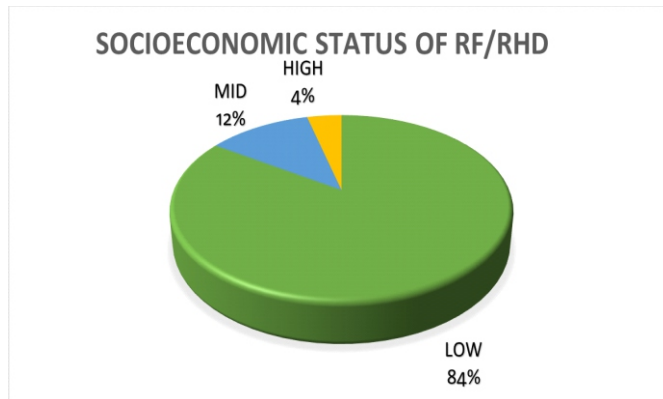


Figure 2: Distribution of RHD subjects by socioeconomic class

Discussion

RHD was found to be the most frequent acquired heart disease in this study similar to other studies done in Nigeria.⁹⁻¹² However, this was not the case in Benin where dilated cardiomyopathy was more prevalent.¹³

While studies done in the Northern parts of Nigeria have reported a persistently high prevalence of RHD, similar studies done in the Southern parts of Nigeria showed a lower prevalence and in fact, a study from Lagos¹⁴ has reported a drop in the prevalence of RHD in adults attributed to the improving health standards and hygiene in the study metropolis.

The subjects with RHD in this study were mostly of low socioeconomic class as reported by Sani et al in Sokoto¹⁵ and other studies from Nigeria and SSA.^{5,16} This is reflected in the inability to carry out important investigations and comply with outlined treatment with the resultant poor outcome recorded. Only 47.1% of the subjects in this study were able to afford an echocardiographic examination and more than a third of the subjects were unable to procure secondary prophylaxis.

Most (96.1%) patients presented with CCF as was reported by other authors^{14,17} RHD is considered to be a major contributor to heart failure in sub-Saharan Africa, especially among the young.¹⁰ Congestive cardiac failure results from chronic valvular injuries most early being mitral valve regurgitation.¹⁴

The commonest affected native valve found in this study was the Mitral valve as reported in a study from this centre and other centres¹⁻⁶ The pattern of valve affectation also was similar to what was documented in previous studies in Nigeria with isolated MR, MR and MS and MR, MS, aortic regurgitation (AR), and MR, MS, AR and TR^{8,9}. This study however did not have any patients with isolated MS. Isolated Mitral stenosis is uncommon in the first decade but the incidence increases with age.¹⁹ More than a third of the patients had mitral

stenosis an indication of increased severity which is attributable to late presentation and the poor adherence to secondary prophylaxis seen in poor socioeconomic settings. Subjects with MS in this study were also found to be older than those with MR. This is consistent with results from Nigeria and other African studies.^{19,20} Primary tricuspid valve disease resulting from RHD was seen in only 1 case and occurred with both mitral and aortic valve lesions as was documented in previous studies. Rheumatic tricuspid valve disease is found alongside mitral and aortic diseases while isolated rheumatic tricuspid valvular disease is a rare occurrence. The finding of 75 % of cases having functional TR is also reported by other authors resulting from pulmonary hypertension.¹⁴ The finding of moderate to severe TR implies a more severe prognosis.^{21,22} Left untreated, severe tricuspid regurgitation is reported to result in RV dilatation, loss of RV reserve despite an apparent preserved RV ejection fraction and complications during tricuspid valve surgery with poor prognosis.¹⁹

It was difficult to determine the exact number of patients presenting with carditis as many patients presented with already established RHD and recurrent congestive cardiac failure from the clinical records. However, all six subjects who were managed for RF had carditis. This is consistent with findings in most low and middle-income countries where more than 50% of cases do not remember the initial occurrence of ARF.

There was a high clinic dropout rate similar to findings from other Nigerian studies.^{15,17} Many patients are unable to afford definitive surgery and ultimately lose hope.

The outcome of patients with RHD remains poor. The mortality rate from RHD in this study was high and only 3.9% of RHD patients were able to access surgical repair. Findings from the same center that involved adults found only 0.7% were able to access surgical care.²¹ This is similar to findings from Sokoto¹⁵ and Kano²⁴ but lower than what was reported from Osun¹⁷ and Lagos.¹⁴ This may be a result of the relatively poor socioeconomic indices, education and health standards in the Northern parts of the country.²³

Conclusion

The outcome of patients managed for rheumatic heart disease remains poor despite the concerted efforts being made by stakeholders. This preventable and curable disease needs increased awareness with appropriate diagnostic and treatment options made readily available.

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