

SPECTRUM OF RHEUMATIC HEART DISEASE IN ZARIA, NORTHERN NIGERIA

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Abstract

Background: Rheumatic heart disease (RHD) is a disease that is still prevalent in the developing countries and still poses a major medical and cardio-thoracic surgery challenge in Nigeria.

Method: A study of data from echocardiography laboratory from October 1999 – September 2002.

Results: Forty-seven (47) out of 600 echocardiography scans had a diagnosis of rheumatic heart disease from October 1999 to September 2002. They were made up of 32 females and 16 males (female/male ratio of 2:1). The ages of the patients ranged from 5 to 52 years with a mean of 19.51 +/- 1.4. There was no statistically significant difference in the mean age of female and male patients. Nearly 90 % of the referring Physician made the correct diagnosis of rheumatic heart disease. Forty eight percent of the patients had depressed left ventricular function (EF < 50%). The mitral followed by the aortic valves were the most commonly affected. Sixteen percent of the patients had severe mitral stenosis. Mitral valve disease whether pure regurgitation, stenosis or combined disease was more common in patients below the age of 25 years. The mean Wilkins score of patients with severe mitral stenosis was 6.8 +/- 1.2. Two patients had aortic valve disease and were above the age of 25 years. The complications included mitral valve prolapse in 19.6%, atrial fibrillation, endocarditis, pericardial effusion and rupture of chordae tendinae. Only 0.7% had surgical intervention but the rest were managed medically. Percutaneous transluminal balloon mitral commissurotomy is not part of management armamentarium.

Conclusion: The mitral and the aortic valves are most commonly affected valves. Patients with critical mitral stenosis have a favourable anatomy for PTMC but it is not available, and a negligible number have the opportunity to have surgical intervention. The need to develop some cardio-thoracic centres as matter of urgency is recommended so as to alleviate the patients' suffering.

Key words: Rheumatic, heart, disease, spectrum

Introduction

Rheumatic heart disease (RHD) is a disease that is still prevalent in the developing countries and its prevalence is put at 0.3-18.6%.¹ In the developed countries the scenario is a different one, with prevalence of less or equal to 0.7%.²⁻⁵ The description of rheumatic heart disease in Nigeria and the northern parts of the country in particular was based on clinical, radiological findings and M-mode echocardiography. Rheumatic heart disease still constitutes a major medical and cardio-thoracic surgery challenge in Nigeria. Real time imaging has given more insight into the nature of the disease and which valves are affected. We describe the spectrum of rheumatic heart disease that is seen in Ahmadu Bello University Teaching Hospital (ABUTH) echocardiography laboratory.

Materials and Methods

The data in the echocardiography laboratory was studied. The patients that had the diagnosis as rheumatic heart disease in their echocardiographic finding from October 1999 to September 2002 included in the study. The parameters recorded include personal data, symptoms of presentation, requesting Physician's diagnosis and echocardiographic findings.

The echocardiographic parameters analysed included, left atrial and aortic root diameter (along the long parasagittal view). The left ventricular dimensions in diastole and systole were also recorded and analysed which included IVSd and IVSs (septum thickness in diastole and systole), LVIDd and LVIDS (left ventricular internal diameter in diastole and systole), LVPWd and LVPWs (left posterior wall thickness in diastole and systole), ejection fraction (EF) and (FS) fractional shortening.

The valves and subvalvar apparatus (mitral) that were involved structurally and functionally were analysed. Complications including atrial fibrillation, chordae tendinae rupture, thrombosis, mitral valve prolapse and endocarditis were studied. The outcome of management of some of the patients was also studied.

The data was reported as mean plus standard error and, percentages in non-quantitative parameters. Student t test and chi-square were used to compare some subgroup (male and females). The other statistical instruments used included correlation coefficient and multiple regressions.

Results

Forty-seven (47) out of 600 echocardiography scans had a finding of rheumatic heart disease. They were made up of 31 female and 16 males with a female/male ratio of 2:1. The age of the patients ranged from 5 to 52, with a mean of 19.51 ± 1.4 . The age of the female patients ranged from 5 to 35 with a mean of 19.1 ± 8.2 . The age of the male patients ranged from 6 to 52 with mean of 20.4 ± 11.8 . There was no statistically significant difference in mean ages between the male and female patients ($t = 0.44$, $F = 0.20$, $p = 0.7$).

The Wilcoxon two- sample test (Kruskal – Wallis test for two groups) an equivalent of chi – square was $X^2 = 0.002$ and $P = 0.9$ did not show statistically not significant difference.

Table 1 shows the distribution of physician's request diagnosis with, 89% making the right diagnosis. Table 2 shows the echocardiographic finding of the subjects studied. Twenty percents of patients had evidence of LVH using the septal thickness of ≥ 1.2 cm. Forty-eight percent of the patients had ejection fraction (EF) below 50% and 12% below 30% (advance cardiac failure), which depicted a restrictive pattern on Doppler tracing. Twenty seven percent had fractional shorting (FS) below 18% (accepted normal ≥ 18.0). Table 3 shows valves involved in regurgitation, with 18% of the patients having combined mitral, aortic, tricuspid and pulmonary regurgitation. The combination of mitral, aortic and pulmonary regurgitation was the least (1%). Mitral valve disease was made of pure regurgitation or stenosis, or combination of both regurgitation and stenosis (97%). Mitral valve disease is seen through out all ages. There were more males than females with mitral regurgitation below the age of 25 years (24 out of 40). Pure mitral valve regurgitation was seen in 15.7%. The combination of mitral and aortic valve was seen more in patients above the age of 25 years (12 out of 21). Four patients had critical mitral stenosis below the age of 25 years against 3 above the age of 25 years (Figure 1). There were similar

findings in mild to moderate mitral stenosis. Figure 2 shows an apical four chamber view with thickened anterior mitral valve leaflet and mosaic jet into the left atrium- MR (mitral regurgitation). The mitral valve area in patients with stenosis ranged from 0.9 to 3.6 cm^2 with a mean of $1.9 \pm 0.16 \text{ cm}^2$. Twenty two percent (7) had critical mitral stenosis ($\text{MVA} \leq 1.0 \text{ cm}^2$). Aortic valve disease was seen in 20 (< 40%). Aortic valve alone was seen in only two patients with combined stenosis and regurgitation.

The echocardiographic complication noted included 9 patients with mitral valve prolapse (19.6%); endocarditis and pericardial effusion had two each (8.7%) and one with rupture of the chordae tendinae. Atrial fibrillation was seen in six out of 47 (12.9%) patients who had larger left atrial diameter. There was no statistical significant difference between male and female patients in terms of left atrial diameter, complications and mitral valve area. There was a statistically significant difference between male and female as regards ejection fraction. ($t = 2.005$, $F = 4.02$, $p < 0.05$), but fractional did not show similar differences ($t = 1.3$, $F = 1.7$, $p < 0.2$). Age seemed to have affected mitral valve area significantly ($F = 4.2$, $p < 0.04$) in patients with mitral stenosis, the older the patient the more the stenosis. There was no statistically significant relationship between mitral valve area and aortic root diameter ($F = 1.1$, $P < 0.5$). The assessment of mitral valve and its apparatus in patients with stenosis using Wilkins score⁶ showed a range of 4 to 9 with a mean of 6.8 ± 1.2 . Thirty-two patients out of the forty-seven (68%) were on prophylactic penicillin (benzethine penicillin) monthly injection for prevention of rheumatic fever

Four patients (0.9%) had surgical intervention in terms of MVR (mitral valve replacement) out of which one patient had combine mitral and aortic valve replacement. Two of the patients that had MVR presented with syncopal and seizure attacks before surgery and had to be placed on anti-convulsants and anticoagulants.

Table 1: Pre-echocardiographic scanning diagnosis

Diagnosis on request for echo scan	No.	%
1. Rheumatic heart disease	36	76.6
2. Rheumatic heart disease plus CCF	6	12.8
3. Ventricular septal defect	2	4.3
4. Congenital heart disease	4	2.1
5. Rheumatic heart with endocarditis	2	4.3
Total	47	100

CCF = Congestive cardiac failure

Table 2: Echocardiographic features of the patients

Features	Range	Mean \pm SE		t-test	P value
		M	F		
Left atrial diameter	2.3 – 8.1	4.7 \pm 1.2	4.6 \pm 1.2	0.3	0.7
Aortic root diameter	1.2 – 3.2	2.2 \pm 0.5	2.1 \pm 0.5	0.3	0.7
IVS	0.5 – 1.3	1.05 \pm 0.2	0.97 \pm 0.3	0.9	0.3
IVSs	0.4 – 1.2	1.03 \pm 0.2	1.02 \pm 0.3	0.17	0.8
LVIDd	3.0 – 7.3	5.2 \pm 0.8	5.4 \pm 0.7	0.6	0.5
LVIDs	2.8 – 5.1	3.7 \pm 0.5	4.1 \pm 0.5	1.8	0.08
LVPWd	0.4 – 1.2	0.9 \pm 0.2	20.82 \pm 0.2	1.1	0.7
LVPWs	0.4 – 1.4	1.02 \pm 0.2	1.09 \pm 0.2	0.9	0.3
Ejection fraction	21.9 – 78.7	47.1 \pm 15.7	58.6 \pm 15.7	2.0	0.05*
Fractional shorting	7.5 – 44.3	23.6 \pm 9.6	28.3 \pm 10.6	1.3	0.10
Mitral valve area (stenosis)	0.9 – 3.6	1.9 \pm 0.7	1.8 \pm 0.81	.04	0.7

* P value statistically significant

Table 3: Echocardiographic regurgitation (incompetents) in all the patients

Regurgitation	No.	%
Mitral / aortic/ tricuspid/ pulmonary	8	18.2
Mitral/ tricuspid	7	15.9
Mitral alone	7	15.9
Mitral/ aortic	6	13.9
Mitral/ aortic/ tricuspid	6	13.6
Mitral/tricuspid / pulmonary	9	20.5
Mitral/aortic/ Pulmonary	1	2.3
Total	44	100

more females than males as reported from South Africa ¹⁰.

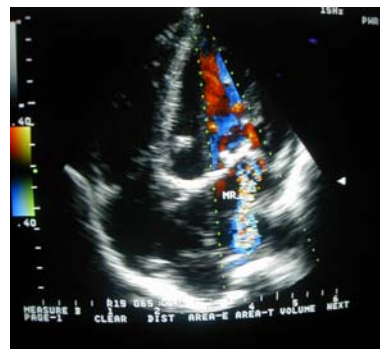
Figure 1: Parasternal short axis view, with a section through mitral valve. It shows severe mitral stenosis with thickened valve leaflets and fused commissures (mitral valve area 0.9 cm²). MV = mitral valve



Discussion

The developing countries are still having fulminate course of rheumatic heart disease because the predisposing factors to rheumatic fever and inadequate prophylactic penicillin therapy. ^{1, 7-9} This is the reverse in the Western countries especially the United state of America where the disease is almost extinct. ¹ The situation in some countries (developing) is also compounded by lack of well-equipped medical set up to manage such condition. It is of recent that real time imaging to study the anatomy of the valves involved and haemodynamic effect of the disease process is being acquired in Nigeria. Comprehensive autopsy studies to match the clinical finding/ imaging features and necropsy reports are not readily available in Nigeria as reported from South Africa. ¹⁰ In Nigeria most descriptions of rheumatic heart disease are from the southern part of the country ⁷ and a lot of the literature showed clinical and radiological diagnosis. The mean age of our patients were similar to that of other developing countries ^{10,11} but lower than the series reported from South Africa. ¹⁰ There were no differences between the mean age male and female patients in our series. As reported by other authors the disease process seemed to present at early age and has a rapid progress. ^{1, 10,11} There were

Figure 2: Apical four-chamber view, showing thickened anterior mitral valve leaflet and a mosaic colour envelope into left atrium MR (mitral regurgitation) in a patient with mixed mitral valve disease (stenosis and regurgitation)



A community survey might bring out the true sex distribution because of non-proper reflection of community prevalence by hospital data. A school children survey done in Egypt, showed more prevalence of rheumatic heart disease in females (4.2/1000) than males (2.2/1000)¹² and in the report of 714 cases from South Africa, females were twice the population of males.¹⁰ A similar report from one of the WHO study centres (Zambia) for the prevention of rheumatic fever, showed more females than males.¹³ Nearly 70% of our subjects were on prophylaxis for recurrent rheumatic fever, the short fall was because some of the patients were referred from neighbouring hospital that have no trained physicians to manage the condition appropriately. Most patients presented with cardiac decompensation, but two patients with severe mitral stenosis in addition had syncope and seizure disorder. Because of the prevalence of the disease process in our clinical setting nearly 90% of the physicians made the right diagnosis before echocardiography scan. In a few number of patients congenital heart disease was thought to be the problem because of the age of the patients involved. The echocardiography scan helped in the assessment of the structure of the affected valves and haemodynamic status of the heart (left ventricular function) and complications. The absence of transoesophageal echocardiography (TEE) would have made for absence of thrombus in the left atrial appendage, which cannot be assessed well with trans-thoracic scan (TTE).

The valves organically involved were the mitral and aortic, with the mitral valve being the most commonly affected in more than 95% of the cases. This finding is similar to other findings elsewhere.^{9,10,14,15} Combined mitral stenosis/regurgitation was seen more than pure mitral stenosis or regurgitation. The finding is similar to imaging and surgical findings from South Africa.¹⁰ A combined mitral and aortic valves disease seemed to have severe hemodynamic compromise. Tricuspid and pulmonary valves were involved functionally but organic involvement was not proven. Pure mitral stenosis is seen more in female (female/ male ratio of 2:1). Similarly a combine mitral valve disease is more common in female, the predomination of female in number could explain this findings. These findings were similar to findings from South Africa and Egypt.^{10,12} The two patients that had aortic valve disease were both males and they had combined stenosis and regurgitation lesions. Haemodynamically 48% of the patients had depressed cardiac function measured by ejection fraction and fractional shortening. Twelve percents had severe cardiac decompensation, which showed Doppler tracing studies of restrictive pattern. The report from South Africa showed that the patients were in New York heart Association (NYHA) class 3 and 4,¹⁰ whereas our patients ranged from NYHA class 1 to 4, this difference might have been due to special group nature of South African series.

Twenty two percent of the patients with mitral stenosis had critical lesion that would require surgical

intervention immediately or Intervention Cardiology i.e. open digital valvotomy or percutaneous transluminal mitral commissurotomy.^{6,16,19,20} The left atrial diameter was similar in both sexes but got larger with age, which shows the worsening disease and haemodynamic changes with time. The mitral valve area in both male and female patients was not different. The males seemed to have better ejection fraction, which might be explain by their lower number. There was no relationship between aortic root diameter and mitral valve area.

Nearly 100% of patients were managed medically on drugs, even those that had critical mitral stenosis could not have surgical intervention except 0.9%. It has been reported that there is less than one cardiology department per 1 million population in nearly all developing countries²¹ and the situation of cardiac surgery is even worse. Asia and sub-Saharan countries have less than 0.2 per I million populations, and some Africa countries have none at all.²¹ The mean Wilkins score in patients with mitral stenosis made them suitable candidates for percutaneous transluminal commissurotomy (PTCM).^{6,16,19,20,22} Percutaneous transluminal mitral balloon valvuloplasty a minimally invasive intervention by cardiologist is not part of our management armamentarium. In the early 70^s and 80^s digital open valvotomy was done in many centres in Nigeria, which seemed to have disappeared with the economic down turn. In addition the only cardio-thoracic centre has problems taking off in properly. Rheumatic fever prevention is yet to be integrated into the country's primary health care as done in other developing countries that noted a drop in the prevalence of the disease.²³ Surgery is basically beyond the reach of the common man or woman who are mostly afflicted with the disease. We might have improved in making the diagnosis of the disease process but intervention has not marched the former.

Rheumatic heart disease is still prevalent in developing countries and affects mainly the younger age group. The mitral and the aortic valves are most commonly affected valves. Patients with critical mitral stenosis have a favourable anatomy for PTMC but it is not available, and a negligible number have the opportunity to have surgical intervention. The need to develop some cardio-thoracic centres as matter of urgency is recommended so as to alleviate the patients' suffering.

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