

ECHOCARDIOGRAPHIC PROFILE OF RHEUMATIC HEART DISEASE AT A TERTIARY CARDIAC CENTRE

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Background: Rheumatic Heart Disease (RHD) continues to be a major public health problem in developing countries like Pakistan. Objective of this cross sectional-analytical study was to analyze the severity of valvular lesions on echocardiography in patients pre-diagnosed with RHD. **Methods:** The transthoracic echocardiographic records of RHD patients from 2004 to 2008 were retrospectively reviewed for type and degree of valvular involvement according to AHA/ACC guidelines. **Results:** A total of 13,414 patients [7,219 Males (53.8%), 6,195 Females (46.2%)] ranging from 11 to 90 years with a mean age of 42.33 ± 18.976 were studied. On echocardiography, 7,500 (56%) had mitral regurgitation (8.8% severe MR), 6,449 (48.2%) had tricuspid regurgitation (7.1% severe TR) and 5,550 (41.4%) had aortic regurgitation (4.8% severe AR). MS was detected in 2,729 (20.3%) patients (15.3% severe MS), AS in 102 (0.8%) and TS in 31 (0.2%) patients. Mixed mitral valve disease was seen in 3,185 (23.7%), mixed aortic valve disease in 222 (1.7%) and mixed tricuspid valve disease in 47 (0.4%) patients. All three valves were involved in 2,826 (21.06%) patients, combination of mitral and aortic valves in 3,103 (23.13%), mitral and tricuspid in 3,784 (28.2%), and mitral only in 3,701 (27.59%) patients. There was some mitral valve abnormality in all patients. **Conclusion:** Mitral valve was most commonly affected, while regurgitant lesions were more common than stenotic lesions, and most severe in younger patients. All valvular lesions had almost an equal distribution among the sexes, except aortic regurgitation, which was more common in females. Therefore, echocardiography should be done routinely for patients with RHD, focusing on younger population, to facilitate diagnosis and definitive treatment before complications set in.

Keywords: Rheumatic heart disease, Echocardiography, Valvular heart disease

INTRODUCTION

Rheumatic heart disease (RHD), an uncommon cause of valvular heart disease in the modern world, is still a leading cause of valvular heart disease in third world countries like Pakistan,¹⁻⁴ where RHD remains endemic.⁵ The prevalence of RHD in Pakistan was found to be 22/1000 in inner Lahore and 5.7/1000 in rural Pakistan in recent studies.^{6,7} This is in concordance with the previously available data putting Pakistan among the high risk countries for RHD.⁸⁻¹⁰ The prevalence of RHD in neighbouring India according to some studies varies from 1.0 to 5.4/1000¹¹ while it is 1.2 per 1000 in Bangladesh.¹² In the developed countries the scenario is totally different, with RHD no longer being a reportable disease, having a prevalence of less than or equal to 0.7%.¹³⁻¹⁴

Although history constitutes the basis of diagnosis of any disease, an accurate history of rheumatic heart disease is difficult to obtain and cannot be solely relied upon for diagnosis.¹⁵ Two-dimensional colour doppler echocardiography is the current gold standard for accurately identifying and quantifying the type and severity of valvular involvement in rheumatic heart disease.¹⁸⁻²² As most valvular lesions with timely intervention hold a good prognosis for the patient, the knowledge of the severity of valvular lesions in RHD patients is very

important.¹⁵ We conducted this study to describe the severity of valvular involvement in rheumatic heart disease patients at a tertiary cardiac centre.

MATERIALS AND METHODS:

Transthoracic 2D echocardiographic records of rheumatic heart disease patients presenting in the cardiology and cardiac surgery departments at the Punjab institute of cardiology (PIC) were analysed retrospectively over a five year period (2004–2008). The types and degrees of valvular involvement were defined according to AHA/ACC guidelines (Table-1).

For this study purpose we classified a valvular lesion with combination of any degree of stenosis and regurgitation as a mixed lesion. We examined the reports of 40,000 patients and only those with echocardiographic features peculiar to rheumatic involvement were included. Congenital heart disease patients, post operative cases, and also collagen vascular disease, myxomatous and old age degenerative disease patients were excluded from the study. A total of 13,414 patients qualified under the inclusion criteria.

The data were entered in SPSS version 13.0, which was also later on employed to derive the descriptive frequencies regarding the distribution of valvular lesions among the patient population in terms of age and sex.

Table-1: Classification of the Severity of Valve Disease in Adults according to AHA/ACC 2006 guidelines for the management of patients with Valvular Heart Disease

| A. Left-sided valve disease | | | |
|---|--|---|--|
| Aortic Stenosis | | | |
| Indicator | Mild | Moderate | Severe |
| Jet velocity (m/s) | Less than 3.0 | 3.0–4.0 | Greater than 4.0 |
| Mean gradient (mm Hg)* | Less than 25 | 25–40 | Greater than 40 |
| Valve area (cm ²) | Greater than 1.5 | 1.0–1.5 | Less than 1.0 |
| Valve area index (cm ² /m ²) | | | Less than 0.6 |
| Mitral Stenosis | | | |
| | Mild | Moderate | Severe |
| Mean gradient (mm Hg)* | Less than 5 | 5–10 | Greater than 10 |
| Pulmonary artery systolic pressure (mm Hg) | Less than 30 | 30–50 | Greater than 50 |
| Valve area (cm ²) | Greater than 1.5 | 1.0–1.5 | Less than 1.0 |
| Aortic Regurgitation | | | |
| | Mild | Moderate | Severe |
| Qualitative | | | |
| Color Doppler jet | Central jet, width less than 25% of LVOT | Greater than mild but no signs of severe AR | Central jet, width greater than 65% LVOT |
| Doppler vena contracta width (cm) | Less than 0.3 | 0.3–0.6 | Greater than 0.6 |
| Quantitative (cath or echo) | | | |
| Regurgitant volume (ml/beat) | Less than 30 | 30–59 | Greater than or equal to 60 |
| Regurgitant fraction (%) | Less than 30 | 30–49 | Greater than or equal to 50 |
| Regurgitant orifice area (cm ²) | Less than 0.10 | 0.10–0.29 | Greater than or equal to 0.30 |
| Mitral Regurgitation | | | |
| Qualitative | Mild | Moderate | Severe |
| Colour Doppler jet width | Small, central jet (less than 4 cm ² or less than 20% LA area) | Signs of MR greater than mild present but no criteria for severe MR | Vena contracta width greater than 0.7 cm with large central MR jet (area greater than 40% of LA area) or with a wall-impinging jet of any size, swirling in LA |
| Doppler vena contracta width (cm) | Less than 0.3 | 0.3–0.69 | Greater than or equal to 0.70 |
| Quantitative (cath or echo) | | | |
| Regurgitant volume (ml/beat) | Less than 30 | 30–59 | Greater than or equal to 60 |
| Regurgitant fraction (%) | Less than 30 | 30–49 | Greater than or equal to 50 |
| Regurgitant orifice area (cm ²) | Less than 0.20 | 0.20–0.39 | Greater than or equal to 0.40 |
| B. Right-Sided Valve Disease | | | |
| Characteristics | | | |
| Severe tricuspid stenosis: | Valve area less than 1.0 cm ² | | |
| Severe tricuspid regurgitation: | Vena contracta width greater than 0.7 cm and systolic flow reversal in hepatic veins | | |

RESULTS

A total of 13,414 patients 7219 (53.8%) males, 6,195 (46.2%) females with rheumatic heart disease were analysed in this retrospective study. Their age ranged from 11 to 90 years with a mean age of 42.33±18.976 for the study population; 44.3±19.58 for males and 39.99±17.96) for females. All three valves were involved in 2,826 (21.06%) of the patients, combination of mitral and aortic valves in 3,103 (23.13%) of the patients, mitral and tricuspid in 3,784 (28.2 %), and mitral only in 3,701 (27.59%) of the patients. In this study population 7,540 (56.2%) had normal aortic valves, 6,887 (51.3%) had normal tricuspid valves and none had normal mitral valve.

Majority of the patients had MR 7,500 (56%) patients, followed by TR 6,449 (48.2%) and AR in 5,550 (41.4%) patients. MS was found in 2,729 (20.3%), AS in 102 (0.8%) and TS in 31 (0.2%) of the

study population. Overall, valvular regurgitations remained commoner than valvular stenoses. Mixed valvular disease was most prevalent in the mitral valve 3,185 (23.7%) patients, followed by aortic 222 (1.7%) and tricuspid valves 47 (0.4%) patients (Table-2).

The data was analysed with respect to age and sex, as distribution of various valvular lesions has been seen to differ accordingly. Gender based differences in the frequencies of various valvular lesions are shown in Table-3. Mitral stenosis was found in 1,484 (20.5%) of males (15.5 % severe MS), mitral regurgitation in 4,080 (56.6%) with 8.2 % having severe mitral regurgitation. Mixed mitral valve disease was seen in 1,655 (22.9%) of the patients. In females, mitral stenosis was found in 1,245 (20.1%) of the patients (15.1% severe MS) and mitral regurgitation in 3,420 (55.1%) with 9.5% having severe mitral regurgitation. Mixed mitral valve disease was seen in 130 (24.7%) of the patients.

In the male population, 2,262 (31.3%) had aortic valve involvement. Some degree of aortic stenosis was seen in only 42 (0.6%) patients. Aortic regurgitation was detected in 2,116 (29.3%), and only 3.6% had severe aortic regurgitation. Mixed aortic valve disease was seen in 104 (1.4%) males. Amongst females, 3,612 (58.3%) had aortic valve involvement; aortic stenosis in only 60 patients (0.9%), and aortic regurgitation in 3,434 (55.5%) with 6.2% having severe AR. Mixed aortic valve disease was seen in 118 (1.9%) of the female population.

In the age group analysis it was seen that left sided valvular lesions were fairly common from 11–70 years of age (Figures-1, 2). Majority of the male patients with moderate and severe MS were seen in the age group of 51–60 years (Table-4). The highest number of male patients with severe MR and mixed mitral valve disease were in the age groups 11–20 years and 51–60 years respectively. Majority of the female patients with moderate and severe MS were seen in the age groups of (21–30 years), severe MR (21–30 years) and mixed mitral valve disease amongst 11 to 30 years. There was only a small number of aortic stenosis patients in the whole study population and severe AS was mostly seen in the older age group of 51–70 years. The highest number of patients with severe AR were seen in the age groups of 21–30 years for males and 11–20 years for females. Whereas, the largest number of patients with mixed aortic valve disease were seen in the age groups of 11–20 years and 21–30 years for males and females, respectively.

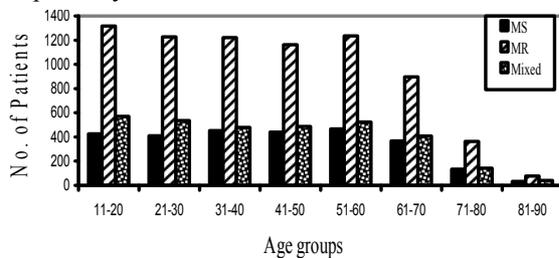


Figure-1: Mitral Valve Involvement

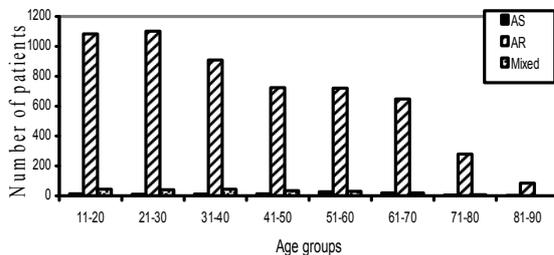


Figure-2: Aortic Valve Involvement

Figure-3 shows the pattern of involvement of tricuspid valve in the total study population with respect to age. In this study population, 6,527 (48.8%) of the patients had tricuspid valve involvement with tricuspid stenosis seen in only 31 patients. Tricuspid regurgitation

was the most predominant lesion, detected in 6,449 (48.2%) of the patients, with severe TR in 6.3% and 7.6% of males and females respectively. Mixed tricuspid valve disease was seen in 47 patients (0.4%) of the study population. Majority of the patients with moderate and severe tricuspid regurgitation were seen in the age group of (11–20 years).

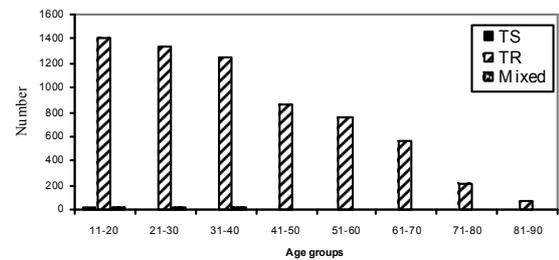


Figure-3: Tricuspid valve involvement in study population

Juvenile Rheumatic heart disease (patients less than 20 years of age), a severe form of RHD, constituted a large proportion 2,314 (17.3%); 1,209 males and 1,105 females. We found that severe MR, severe AR and severe TR were commonly found in the age group of 11–20 years along with mixed mitral and aortic valve disease. The distribution of significant valvular lesions with respect to age and gender is shown in (Table-4).

Table-2: Distribution of valvular lesions in total study population

| Valve Condition | Mitral Valve | Aortic Valve | Tricuspid valve |
|-----------------|---------------------|--------------|-----------------|
| Normal valves | 0 (0.0%) | 7540 (56.2%) | 6887 (51.3%) |
| Stenosis | 2729 (20.3%) | 102 (0.8%) | 31 (0.2%) |
| Regurgitation | 7500 (55.9%) | 5550 (41.4%) | 6449 (48.2%) |
| Mixed Disease | 3185 (23.7%) | 222 (1.7%) | 47 (0.4%) |
| Total | 13414 (100%) | | |

Table-3: Frequency distribution of valvular lesions among males and females

| Valvular lesions | Males | Females |
|------------------|-------|---------|
| Mild MS | 4.9% | 4.9% |
| Mod MS | 0.1% | 0.1% |
| Severe MS | 15.5% | 15.1% |
| Mild MR | 30.7% | 26.5% |
| Mod MR | 17.7% | 19.1% |
| Severe MR | 8.2% | 9.5% |
| Mild AS | 0.5% | 0.9% |
| Mod AS | 0.1% | 3% |
| Severe AS | 1 | 3 |
| Mild AR | 6.8% | 15.0% |
| Mod AR | 18.9% | 34.3% |
| Severe AR | 3.6% | 6.2% |
| Mild TR | 11.2% | 7.6% |
| Mod TR | 31.6% | 31.3% |
| Severe TR | 6.3% | 7.9% |
| Mixed MV disease | 22.9% | 24.7% |
| Mixed AV disease | 1.4% | 1.9% |
| Mixed TV disease | 0.3% | 0.5% |

Table-4: Age group (years) distribution of significant valvular lesions

| Valvular lesions | Males | females |
|------------------|----------------------|-----------|
| Severe MS | 51-60 yrs | 21-30 yrs |
| Severe MR | 11-20 yrs | 21-30 yrs |
| Severe AS | 61-70 yrs | 51-60 yrs |
| Severe AR | 21-30 yrs | 11-20 yrs |
| Severe TR | 11-20 yrs, 31-40 yrs | 21-30 yrs |
| Mixed MV disease | 51-60 yrs | 11-30 yrs |
| Mixed AV disease | 11-20 yrs | 21-30 yrs |
| Mixed TV disease | 31-40 yrs | 21-30 yrs |

DISCUSSION

Rheumatic heart disease continues to be a major health problem in the developing countries. It accounts for a large percentage of cardiovascular disease related admissions and is an important indication for cardiac surgery in third world countries like Pakistan. Recent studies conducted in this country continue to show the high prevalence of the disease, which is in contrast to the virtual extinction of the disease in the developed world.^{7,14,15}

Our study revealed that the most common lesions seen in patients with rheumatic heart disease were of regurgitant type with mitral valve leading, followed by tricuspid and aortic valves (MR>TR>AR). This is consistent with previously reported data from different countries.^{13,15,24} Whereas, mixed mitral, mixed aortic and mixed tricuspid valvular lesions were commoner than their respective stenotic valve lesions.^{13,25}

The study found TR to be commoner than AR.^{15,24} However, it was predominantly functional TR secondary to pulmonary hypertension. Tricuspid regurgitation because of rheumatic involvement is uncommon, but is always associated with mitral valve disease.^{26,27}

We also found that MR, TR and all stenotic lesions had almost an equal distribution among males and females, while aortic regurgitation was more common in females. Recent studies show inconsistency; with some showing typical picture of equal prevalence of RHD amongst males and females, while others depicting females as the dominant patient population.^{24,28-32} This may be because in the rural areas of a country like Pakistan where health care facilities are scarce, women are at a higher risk due to social and cultural constraints and thus, delay in management of a relatively benign lesion can lead to aggravation of the disease causing significant morbidity and mortality. We believe that this problem can be tackled by educating our masses via regular visits of health care professionals, newspapers, radio and television, stressing on the importance of better outcomes and quality of life with timely diagnosis and intervention.⁷

Our study also showed that Juvenile rheumatic heart disease is very common in South Asia as reported in earlier studies.^{7,27,33} Patients under 20 years constituted a major portion (17.3%) of our study population. We found that severe MR, severe AR and severe TR were most commonly found in the age group of 11-20 years.

The high prevalence and severity of rheumatic heart disease in our part of the world is a great cause of concern and calls for immediate and effective prophylactic measures to ensure a symptom free life span for the patient. The data suggests that recurrences of RF are higher in patients with RHD and each recurring disease episode further damages the heart.³⁴ In order to counter these recurrent attacks effective secondary prevention is required, which relies on accurate case detection for the appropriate use of prophylactic antibiotics and regular medical surveillance. Here, echocardiography plays an important role firstly, in early detection of RHD. Secondly, monitoring of medical therapy because RHD at this stage would warrant lifelong prophylaxis.¹⁴ Therefore, echocardiography should be recommended as a routine screening tool for investigation of RHD, and also for follow up to guide in the timely intervention for severe valvular lesions.

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