



**Research Article**

**A STUDY OF SPECTRUM OF RHEUMATIC HEART DISEASE IN SAWAI MAN SINGH MEDICAL COLLEGE, JAIPUR**

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**ABSTRACT**

**Background:** Rheumatic heart disease remains a major public health problem in developing countries like India affecting the young population imparting huge familial, social and manpower burden. While acute rheumatic fever is on the decline even in the developing world, there are still a large number of chronic rheumatic heart disease cases, often complicated by chronic congestive heart failure and recurrent thrombo-embolic phenomena, both posing greater challenges for management. We report on the prevalence and pattern of valve involvement in RHD from our centre.

**Materials and methods:** This is a hospital based descriptive cross-sectional study during December 2013 to January 2016 over a period of 25 months at Sawai Man Singh Medical College, Jaipur including 940 patients with clinical and/ or echocardiographic evidence of definite rheumatic heart disease.

**Results:** The age of the patients ranged from 8 to 74 years with mean age  $39.82 \pm 4.2$  years with female preponderance (F:M = 2.1:1) ( $p < 0.01$ ). Majority of the rheumatic heart disease patients belonged to 30–44 years (28.78%) followed by 15–29 years (25.75%) and 45–59 years (25.00%). Majority belonged to the low socioeconomic status (60.60%) ( $p < 0.05$ ). The predominantly involved isolated valve was mitral in 440 patients (46.80%) followed by isolated aortic valve in 88 patients (9.36%) and 316 (33.62%) had dual valvular involvement. The common rheumatic valvular lesions were pure mitral stenosis in 128 (13.61%), isolated mitral regurgitation in 232 (24.68%), combined mitral stenosis/regurgitation in 144 (15.32%), combined mitral/aortic regurgitation in 92 (9.78%) and combined aortic stenosis/regurgitation in 72 (7.66%) patients with few overlappings.

The common complications encountered were heart failure in 360 (38.30%) and arrhythmias in 496 (51.00%) patients. 520 patients (55.32%) received injectable benzathine penicillin whereas 180 patients (19.15%) preferred oral penicillin V. Surgical intervention was done in 216 (22.97%) patients. 48 (5.10%) expired in the CCU during the course of treatment.

**Conclusion:** RHD is a leading cause of heart failure among young populations with requirement of prolonged duration of medical treatment and many of them requiring surgery.

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**INTRODUCTION**

**Background**

Rheumatic heart disease (RHD) is one of the most common cardiac diseases in developing countries like India. According to the WHO, at least 15.6 million people worldwide have RHD. Of the 500,000 individuals who acquire acute rheumatic fever (ARF) every year, 300,000 go on to develop RHD and 233,000 deaths annually are attributable to ARF or RHD [1, 2].

RHD leads to early onset of heart failure in the young population along with multiple complications with lots of mortalities and morbidities each year. It also imparts huge economic and social burden in Rajasthan.

We do not have adequate studies on the spectrum and related complications of RHD in Rajasthan.

This belt is located in the western part of the country covering many patients from nearby zones like Agra (Uttar Pradesh), Sheopur (Madhya Pradesh), Hissar (Haryana). This study is carried out in our tertiary level hospital in Jaipur to study the magnitude of RHD in the overall cardiac outpatients and inpatients and the spectrum of cardiac involvement along with complications.

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**Aims and objectives**

This study is designed to study the overall burden of RHD in the cardiac department out and inpatients along with spectrum of cardiac involvement with special focus on pattern of valvular involvement. This study will give a real glimpse of burden of RHD in Rajasthan.

**MATERIALS AND METHODS**

This is a hospital based descriptive cross-sectional study during December 2013 to January 2016 over a period of 25 months at Sawai Man Singh Medical College, Jaipur including 940 patients with clinical and/ or echocardiographic evidence of definite rheumatic heart disease.

**Inclusion criteria**

Consecutive 940 patients with echocardiographic evidence of definite rheumatic heart disease were included in the present study. 2012 World Heart Federation criteria for the echocardiographic diagnosis of rheumatic heart disease was applied to reach the diagnosis [3].

**Definite RHD (A, B, C, or D)**

- A. Pathological MR and at least 2 morphological features of RHD of the MV
- B. MS mean gradient  $\geq 4$  mmHg
- C. Pathological AR and at least 2 morphological features of RHD of the AV
- D. Borderline disease of both the AV and the MV

**Borderline RHD (A, B, or C)**

- A. At least 2 morphological features of RHD of the MV without pathological MR or MS
- B. Pathological MR
- C. Pathological AR

**Criteria for pathological mitral regurgitation**

(All 4 Doppler echocardiographic criteria must be met).  
 Seen in 2 views: In at least 1 view, jet length  $\geq 2$  cm, velocity  $\geq 3$ m/s for 1 complete envelope, pan-systolic jet in at least 1 envelope.  
 Morphological Features of RHD.  
 Features in Mitral Valve (MV): MV leaflet thickening  $\geq 3$  mm (age specific), chordal thickening, restricted leaflet motion, excessive leaflet tip motion during systole.

**Pathological aortic regurgitation**

Seen in 2 views: In at least 1 view, jet length  $\geq 1$  cm, velocity  $\geq 3$ m/s in early diastole, pan-diastolic jet in at least 1 envelope.  
 Features in aortic valve (AV): Irregular or focal thickening, coaptation defect, restricted leaflet motion, prolapse.

**Exclusion criteria**

Those not fitting into the echocardiographic criteria for diagnosis of definite RHD were excluded from the study. Verbal consent was taken from each patient during the study period. Data was regularly entered into the SPSS-20 software.

**Statistical analysis**

The datas were analysed by the statistician. Number and sex distribution were expressed in mean and standard deviation. Crosstab analysis was done wherever required. P value of  $<0.05$  was considered significant.

Data was collected from both cardiology outpatients and inpatients (admitted in cardioward/Cardiac surgery unit). Relevant data and information were entered into the pre-structured proforma and then analysed by SPSS-20 software.

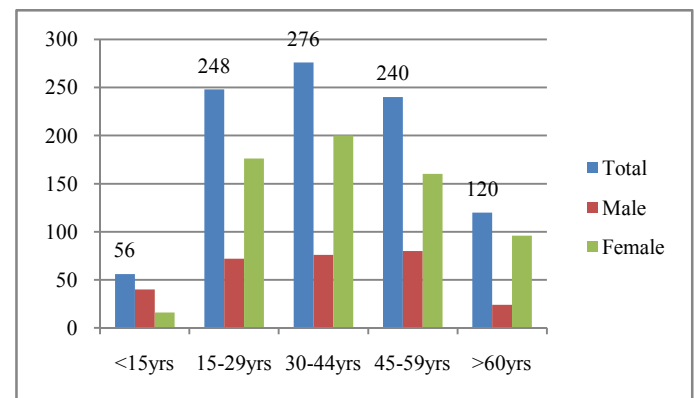
Ethical clearance was taken from the Institutional review Board Committee of Sawai Man Singh Medical College, Jaipur

**OBSERVATIONS AND RESULTS**

Out of total 1106 patients who were evaluated with 2-dimensional transthoracic echocardiography, 940 patients were categorized to be having definite RHD and 166 patients had possible or borderline RHD.

Those with definite RHD were enrolled in our study for further statistical analysis.

The age of the patients ranged from 7 to 76 years with mean age  $39.82 \pm 4.2$  years with female preponderance (F:M=2.1:1) ( $p < 0.01$ ). The majority of the RHD patients belonged to 30–44 years (29.36%) followed by 15–29 years (26.38%) and 45–59 years (25.53%) as shown below in the Fig. 1 highlighting the involvement of productive age group.



**Fig 1** RHD distribution with respect to age and sex.

Geographical distribution: The majority of the patients in this study were from Shekhawati area (344–36.59%) followed by Hadoti area (224– 23.83%), Bharatpur belt (104–11.06%), Hanumangarh area (56–5.96%), Dausa belt (40– 4.25%), West Rajasthan (32–3.40%) and few from outside Rajasthan (40– 4.25%). Majority belonged to low socioeconomic status (63.83%) ( $p < 0.05$ ). 324 (34.47%) had middle SES and only 32 (1.70%) had good SES. History of acute rheumatic fever was elicited only in 64 patients (6.80%). Overcrowding (family members  $N_6$ ) was noticed in 560 patients (59.57%) ( $P < 0.01$ ). 88 patients (9.36%) were pregnant all of whom had uneventful antenatal and postnatal history.

Isolated mitral valve was the most commonly affected valve in our study (46.80%) followed by isolated aortic valve. 33.62% had dual involvement of mitral and aortic valve. Only 7.66% had involvement of tricuspid valve and 2.55% had pulmonary valve involvement along with mitral and aortic involvement as shown in Table 1.

**Table 1** Patterns of valvular involvement of RHD (n=940)

Valve involved	Percentage	Number of patients
Isolated mitral valve	46.80%	440
Isolated aortic valve	9.36%	88
Mitral & aortic	33.62%	316
Mitral, aortic and tricuspid	7.66%	72
Mitral, aortic and pulmonary	2.55%	24

Our patients in this study had both isolated and combined valvular lesions with majority being regurgitant lesions (MR in 692 patients-73.61%). There were many overlappings of the stenotic and regurgitant lesions in a single valve or combined lesions. These are illustrated above in Fig. 2. While assessing the severity of individual valvular lesions, 104 patients (11.06%) had severe MS, 244 patients (25.96%) had severe MR, 36 patients (3.83%) had severe AS and 104 (11.06%) had severe AR.

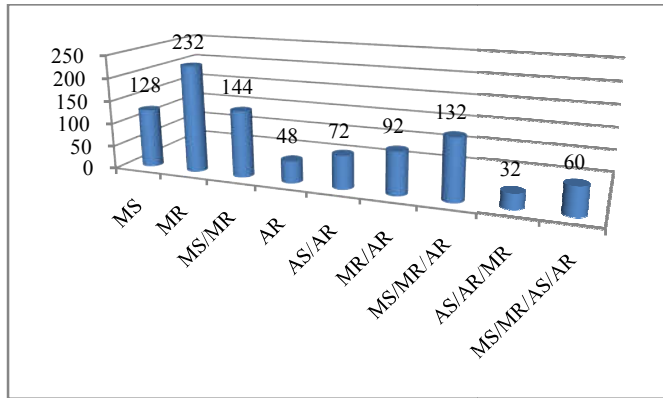


Fig 2 Distribution of valvular lesions

The Table 2 illustrates the severity of valvular lesions in our study. Out of total 940 patients enrolled in this study, secondary pulmonary hypertension (PASP +30 mm Hg measured by TR jet velocity) was detected in 480 patients (51.06%). Giant atrium (atrial dimension >7.5 cm) was noted in 144 patients (15.32%) out of whom 8 had upper GI bleeding due to chronic pressure and erosion of esophagus. 32 patients (3.40%) had left atrial appendage clot predisposing to risk of thromboembolism like embolic stroke and peripheral lower limbs arterial ischemia.

Table 2 Distribution of severity of valvular lesion (n=940)

Valve Lesion	Percentage	Number of patients
Mitral Stenosis	49.36%	464
Mild	15.31%	144
Moderate	22.97%	216
Severe	11.06%	104
Mitral regurgitation	73.62%	692
Mild	28.93%	272
Moderate	18.72%	176
Severe	25.96%	244
Aortic Stenosis	17.44%	164
Mild	11.91%	112
Moderate	5.96%	56
Severe	3.83%	36
Aortic regurgitation	46.38%	436
Mild	14.89%	140
Moderate	20.42%	192
Severe	11.06%	104

38.30% of the patients had heart failure and 51.00% had cardiac arrhythmias which is illustrated in the Figure 3. Atrial fibrillation was the most common arrhythmia in our study (280/940–29.78%) as shown in Fig. 3 and was more common with valvular MS (60.24%) (P<0.05) followed by MR (26.34%). Majority of the patients were managed medically. 216 patients (22.97%) had undergone surgical intervention. Out of 940 patients, 48 (5.10%) expired in hospital during the study period and the majority of the patients are on regular follow-up. The causes of death were: refractory heart failure in 28 patients (2.97%), acute pulmonary edema in 16 patients (1.70%) and malignant ventricular arrhythmias in 4 patients (1.27%).

Table 3 Complications (n=940)

Complications	Number of patients	Percentage
Secondary PAH	480	51%
Giant atrium	144	15.3%
LA clot	32	3.4%
IE/Vegetation	28	2.9%
Valvular cardiomyopathy	48	5.1%
Arrhythmia	360	38.29%
Prosthetic valve thrombosis	12	3%
Cardioembolic stroke	56	5.9%
Acute limb ischemia	20	2.1%
Heart failure	360	38.3%
Acute rheumatic fever	20	2.1%
Carditis	96	10.21%

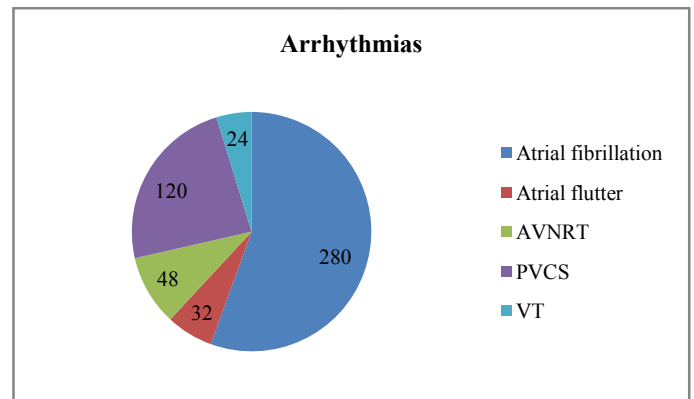


Fig 3 Distribution of different arrhythmias (n=940)

## DISCUSSION

Rheumatic heart disease (RHD) is a common cardiac disease in children and young adults in developing countries especially in the rural areas. RHD is caused by an abnormal autoimmune response to a Group A streptococcal infection in a susceptible host.

There is marked global variation in acute rheumatic fever incidence and RHD prevalence and the sub-Saharan African and Asian-Pacific regions are identified as high disease burden areas [4,5].

Globally there are around 34 million patients suffering from RHD and majority resides in Asian countries where burden is estimated to be 10.8–15.9 million patients [6]. These estimates indicate that the global burden is significantly higher than the earlier estimates for children as well as all age groups.

According to the Global Burden of Disease 2010 study, 10 million disability-adjusted life years (DALYs) are lost per year, almost all in low and middle income countries [7]. Lozano *et al.* [8] has reported 345,100 deaths due to RHD in 2010, which represents a quarter reductions from 1990, with ages-standardized death rate of 5.2 per 100,000, which was a 53.10% reduction from 1990.

Globally, approximately 25%–50% of newly diagnosed cases, deaths, hospitalizations and burden of RHD are from India [9]. The prevalence rates have been found to be 1.3-4.5/1000 population among children in age group 5-15 years [10]. In Studies done by Indian Council of Medical Research in school children in late 1980s, RHD prevalence of 1.0/1000 in Delhi, 2.9/1000 in Vellore and 5.4/1000 in Varanasi has been reported [11]. Parimal *et al* found the prevalence of RHD clinically as 16.7/1000 in school children, however echo prevalence was quite low (0.67/1000) [12].

The females were predominantly affected by RHD than males probably because of medical negligence during childhood and lack of availability of health care. The social stigmata and disparity among the genders still prevails in the rural community.

In our study, mitral valve (54.55%) is most commonly involved in RHD followed by aortic valve and combined valvular lesions Isolated Mitral regurgitation along with combination (73.62%) was the most common RHD in our study followed by mitral stenosis, aortic regurgitation and aortic stenosis.

Sanyal S, Berry A, Duggal S, *et al.* [14] in a prospective study from North India described the chronic MR to be more prevalent of RHD in children and young adults whereas MS is increasingly common in patients in the fourth to sixth decades of life.

Severe chronic MR eventually leads to ventricular dysfunction causing reduced ejection fraction, elevated end-systolic volume and elevated left heart filling pressures and finally congestive heart failure.

Rheumatic fever resulting in chronic RHD is the most common cause of mitral stenosis. Symptomatic rheumatic MS may occur as early as second decade of life in children from developing countries of the world. Isolated pure MS was diagnosed in 128 (13.61%) patients but the combined MS lesion with MR contributed 49.36% in our study.

Pure MS occurs in about 25.00% of adults and another 45.00% have combined MS and MR. Women are more likely than men to develop rheumatic mitral stenosis [15].

In mitral stenosis, progressive obstruction to LV inflow develops due to fibrosis and partial fusion of the mitral valve leaflets. Approximately 30.00% of outside Rajasthan RHD patients aged 10–19 years have mitral stenosis, and the mean age of those with mitral stenosis is 33 years.

Chronic rheumatic AR occurs because of leaflet thickening, fibrosis and leaflet contracture resulting in abnormal leaflet coaptation and a regurgitant orifice. During compensatory phase, ventricular dilation occurs to maintain forward stroke volume and cardiac output and ejection fraction remains normal. Similar to chronic MR patients, those with chronic severe AR may remain asymptomatic for years. Over time, decompensation may occur resulting in decreased LV function and symptoms.

Aortic stenosis occurs 30–40 years after the acute illness as calcification, leaflet thickening, fibrosis, commissural fusion and adhesions develop over time. Rheumatic AS and AR often occur concurrently usually along with rheumatic mitral valve disease. So, mitral valve should be carefully evaluated in all patients with chronic rheumatic aortic valve disease since coexistent mitral valve involvement is common.

Asymptomatic patients with valvular heart usually remain stable for years. Medical management includes serial evaluation to detect interval change and the onset of symptoms, prevent complications and to detect change in valvular function, chamber size and ventricular function.

The presence of significant symptoms is a logical and accepted indication but the optimal timing of intervention for asymptomatic children is unclear. Percutaneous balloon

valvotomy is effective in many patients with rheumatic mitral stenosis. Mitral valve and aortic valve repair/replacement are performed for adults with severe MR/AR with symptoms and/or left ventricular dysfunction and marked LV enlargement.

The surgical approach to rheumatic tricuspid valve disease is based on the underlying abnormality. A tricuspid ring annuloplasty may be performed for tricuspid regurgitation owing to annular dilation. Tricuspid commissurotomy is reserved for rheumatic tricuspid Stenosis [17].

Many patients in our study had multivalvular involvement. Similar finding is reported from other states of India. Both the mitral and aortic valves may be involved (e.g. aortic regurgitation and mitral stenosis or aortic and mitral regurgitation). Multivalve involvement is seen in 38.00% of patients with MS, with the aortic valve affected in approximately 35.00% and the tricuspid valve in approximately 6.00%. The pulmonic valve is rarely affected. Two thirds of all patients with rheumatic MS are female. The interval between the initial episode of rheumatic fever and clinical evidence of mitral valve obstruction is variable, ranging from a few years to >20 years [16,18].

The management is usually that of the dominant lesion. However, the proximal valve lesion may modify the effects of the distal lesion e.g. severe mitral stenosis may prevent the development of significant LV dilatation secondary to aortic regurgitation. The progression of the milder valve lesion is variable. However, in one Israeli follow-up study (mean 13 ± 7 years) of rheumatic valvular disease patients (mean age 61±10 years) with mild aortic valve disease who had required mitral valve surgery showed that in the vast majority of cases, the aortic valve disease remained stable without disease progression [19,20].

In younger patients, the degree of adherence to antibiotic prophylaxis would be the major determinant of the progression of the non-operated valve disease. The combination of significant mitral and aortic regurgitation is a surgical challenge and carries a high risk of ventricular dysfunction. Surgical intervention is indicated at the onset of symptoms, or if LV dysfunction is identified.

Till date, no primary preventive strategies for acute ARF are available in developing countries like India. The only proven cost effective intervention is secondary prophylaxis i.e., the long-term administration of antibiotics to people with a history of acute ARF or RHD, to prevent ARF recurrences and the development or deterioration of RHD. The drug that we use for this purpose is intramuscular benzathine penicillin G administered once every 3 weeks. Decisions about duration of secondary prophylaxis relate to the balance between the risk of recurrent ARF and the risk to the patient, should a recurrence occur (higher with increasingly severe heart disease) [21].

#### **Conflict of interest**

The authors do not have any conflict of interest including financial in publication of this article.

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