

Chronic Rheumatic Endocarditis an Echocardiographic Study

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Chronic rheumatic endocarditis (CRE) is a major problem especially in the under developed countries^{1,2}. The prevalence rate in India is about 6.4 per 1000 rural population and increases with age upto 25 years³. While it has been predominantly eradicated from the west it is a common complication of rheumatic fever (RF) and the leading cause of cardiac death in the young population in the underdeveloped countries^{4,5,6}. Bland and Jones showed that following acute rheumatic fever after 20 years 45% had evidence of CRE³. About 3-5% of patients with group-A beta hemolytic streptococci develop RF⁶ but the incidence increases to about 50% in the first year following the initial attack of RF and falls quite steeply reaching a plateau at 10% after 4 or 5 years⁷. Widespread use of echocardiography and Doppler flow studies have highlighted this problem and enabled an early and accurate anatomical diagnosis. The incidence of various valvular involvement in patients with CRE presenting with cardiac murmurs is presented.

Key words: Rheumatic, Endocarditis, Echocardiography.

All patients referred for evaluation of their cardiac murmurs to the Department of Medicine East Medical ward Mayo Hospital Lahore during the year 1996-1999 were studied echocardiographically. An echocardiographic and Doppler examination was carried out on GE RT-6800 machine using 2.5, 3.5 and 5.0 MHz transducer. Color flow mapping was also used to determine regurgitant flow. Continuous wave and pulsed Doppler was used to determine abnormal flow across the valves. Cine loop recordings were also reviewed to make the diagnosis. Standard views were obtained to visualize the valves and obtain Doppler samples. Correction angle was used where absolutely necessary. All patients with rheumatic valvular lesions were selected for the study and were categorized according to the anatomical site of involvement. The standard criteria laid down for the echocardiographic evaluation of various valves were used. Recordings were made on paper, video and entered in a register. The following criteria were used to diagnose different valvular involvement in CRE.

Criteria for rheumatic mitral valve involvement on echo^{8,9}

- Classical thickening of the valves on M-mode and 2-D echo had to be present.
- Diastolic doming of the anterior mitral leaflet with or without bulbous tips (hockey stick).
- Abnormal anterior diastolic motion of posterior mitral leaflet.
- Valvular stenosis on 2-D echo¹⁰, showing fish mouth or button hole orifice.
- Calcification of the mitral leaflets.
- Abnormal systolic or diastolic flow across MV on Doppler study^{11,12,13}.

Criteria for rheumatic aortic valve disease

- Thickened valves on 2-D with multiple echoes on M-Mode.

- Evidence of calcification (excluding congenital bicuspid AV).
- Diastolic fluttering of the anterior mitral leaflet in AR.
- Abnormal systolic or diastolic flow across the aortic valve on Doppler study.^{13,14,15}

Criteria for diagnosis of rheumatic tricuspid valve involvement

- They are similar to the corresponding left sided lesions¹⁶. Any case of secondary tricuspid valve involvement were not included.

Results :

The study includes 316 patients with 147 males (49.6%) and 169 females (50.4%). Their ages varied from 4 to 75 years with an average age of 30 years. The maximum incidence was found in the age group between 11-30 years comprising 69% of the total patients. The average age of patients with two valve involvement was 29 years while those with triple valve involvement (TVI) i.e. mitral valve (MV), aortic valve (AV) and tricuspid valve (TV) was 24 years. Mitral valve involvement (MVI) was seen in 304 out of 316 (96%) case. Pure mitral stenosis (MS) was the most common lesion seen in 127 patients (40%), with 47 males (37%) and 80 females (63%). This is followed by mixed mitral valve disease with either mitral valve thickening and predominant regurgitation 8.5% (27 patients) or significant MS with MR 22.5% (71 patients with 26 males and 45 females). Next frequent involvement was MV and AV involvement (AVI) in 20% {63 patients with 39 males (62%) and 24 females (38%)}. TVI was seen in 13 patients (4%) of which 9 were males and 4 female. Aortic valve involvement with minimal mitral valve thickening was seen in 12 patients (4%). While patients with a single valve involvement amounted to 75%, double valve involvement was seen in 20.7%, triple valve pathology in 4% and four valve involvement in 0.3%

Various Valvular involvements in our study.

Patients	No	%age	Males	Females
Total patients	316	147	169	
Mitral valve involvement (MVI)	304	96	140	164
Pure MS	127	40	47	80
MR with MV thickening	27	8.5	17	10
MS + MR	71	22.5	26	45
MS + TR	2	0.5	1	1
MS + AR	23	7	14	9
MS + AS	28	12	18	10
MR + AS	9	3	6	3
MR + AR	3	1	1	2
AS	5	1.5	3	2
AR	2	0.5	1	1
AS + AR	5	1.5	3	2
Single valve (MV or TV or AV or PV)	237	75		
Double valve (MV + AV or TV or PV)	65	20.7	40	25
Triple valve (TVI = MV + AV + TV or PV)	13	4	9	4
Four valve pathology (MV + AV + TV + PV)	1	0.3	1	0

There were 28 patients who had Doppler evidence of tricuspid regurgitation of whom only about half the number¹³ had actual tricuspid valve thickening. The incidence of pulmonary regurgitation on Doppler was seen in 8 patients.

Discussion :

MVI has been traditionally the commonest pathology in patients with CRE. This is well substantiated by our study. Among the 96% patients with MVI, 46% were males and 64% females. Mitral valve is involved alone, as mixed mitral valve disease or in combination with other valvular involvement. While pure MS remains the commonest lesion (40%) with 37% females and 63% males, it is closely followed by mixed mitral valve disease (31%) which is again more common in the females 56% vs. 44%. MR alone was less common in our study with a total of 27 patients (8.5%), but the total number of patients with MR were 119 (38%). It therefore occurred in combination with MS or other valvular involvement suggesting that MR may be a rather late feature of a more severe form of disease rather than an early sequel of CRE. The third commonest pathology was mixed MVI and AVI seen in 20% cases. The total number of patients with AVI were 63 with 39 males (62%) and 24 females (38%). In all 26 patients had AR while 37 had AV thickening with reduced opening compatible with AS. It was interesting to note that combined Mitral and aortic valve involvement was predominantly seen in males (62%). This represents more severe disease in males.

TVI was found in 13 of our patients (4%) of which 9 were males and 4 female. However combined MV and TV involvement was rare and seen in only 2 patients. It may

be noted that MVI is more common in females 64%. Triple valve pathology was seen in 4% with 69% males and 31% females. Tricuspid valve involvement occurred in 16 patients (5%) who had tricuspid valve thickening. However on Doppler study the total number of patients with tricuspid regurgitation was 28 suggesting the severity of disease process in our patients who due to back pressure had functional TR. TV involvement accompanies MVI^{17,18}, or more commonly as part of TVI. In one study 15% of the patients at autopsy had TS but it was clinically significant in only 5%¹⁹. TS is reported to be more common in females²⁰, but in our study we found TV involvement in 5% (16/131) patients and 68% (11/16) were males. The average age of patients with TV involvement was 24 years which reflects the severity of the rheumatic process which occurs in the underdeveloped countries. In three previous autopsy studies conducted by Cook and White in 1940 with 217 cases, second conducted by Lannigan²¹ in 1966 with 298 cases and a third conducted more recently by Kinare in 1983 with 500 cases the comparison of various valvular involvement is shown along with our present and previous echocardiographic study in the following table.

Comparison of the frequency of various valvular involvement in cre in different studies

Studies	Autopsy Cook & White	Autopsy Lannigan	Autopsy Kinnare	Echo Haroon	Echo Haroon
Year	1940	1966	1983	1995	2000
Number of patients	217	298	500	131	316
Mitral only	39.9	48.3	45.1	58	71.2
Mitral and Aortic	46.1	42.3	20.5	32	19.9
Aortic only	5.1	2.0	0.4	1.5	3.8
TVI	3.2	4.4	19.6	5.5	4.1
Mitral & Tricuspid	3.2	2.0	6.6	3.0	0.06
All four valves	2.5	0.6	5.7	0	0.03

Pure mitral valve disease remains the commonest lesion in all with a slightly higher incidence in our study (71%). This included patients with pure MS, MR and mixed mitral valve disease. Mixed mitral and aortic valve disease or two valvular involvement was the second commonest lesion (20%). It may be noted that all three studies are autopsy figures and represent terminal disease pattern while our study may represent a relatively early form of disease pattern.

The average age of our patients was 30 years while patients with two valve involvement was 29 years and with triple valve involvement (TVI) ie. mitral valve (MV), aortic valve (AV) and tricuspid valve (TV) was 24 years. In a study by Okada²² the mean age of patients with MS was 42 years, with MR was 32 years and with triple valve disease was 32 years. Multiple valvular involvement is not

uncommon (25%) being the result of a more fulminant rheumatic endocarditis. This may be explained on the basis of the following parameters²³.

1. Delay in recognition of disease.
2. Improper or insufficient treatment of acute RF.
3. Improper prophylaxis following acute RF.
4. Poor socioeconomic status.
5. Poor patient education.
6. Poor general resistance.
7. Overcrowding.

The reason for the males predominantly being affected in multiple valvular involvement i.e., 50 vs 29 with double, triple and four valve involvement firstly may be a failure to observe strict bed rest of 6-8 weeks in the early phase of acute carditis possibly due to prevalent male child labour to support large family size in our country. Secondly susceptibility to severe and recurrent soar throat may be increased due to close contact at places of work. Once RF has occurred the chances of recurrence with subsequent streptococcal soar throat rise to about 50% in the first year and remain high at about 10% for years to follow. There are various strains of streptococci recognized according to their M-proteins. Types 5, 14 and 24 are notorious in causing RF^{23,24,25}. It may be that these strains are more prevalent in Pakistan, but this remains to be studied. Furthermore appropriate medical care, improved sanitary conditions, primary and secondary prophylaxis are necessary to reduce the incidence of this devastating disease. This has been suggested in study by Longo-Mbenza²⁶ and Regmi²⁷. Widespread use of echocardiography is necessary to pick up early lesions and offer appropriate prophylaxis to prevent further damage through recurrent rheumatic fever.

The order of frequency of different valvular involvement in CRE correlates with the level of haemodynamic stress normally acting upon each valve (Wood 1954). Therefore the closed MV is exposed to the greatest pressure i.e., systemic arterial pressure, while the closed AV is exposed to systemic arterial diastolic pressure. After the initial injury the abnormal stress and strain acting through the abnormal flow continues to perpetuate the valvular injury. In general clinical manifestations produced by the more proximal of the two valvular lesions eg., MV in mitral plus aortic valve disease and TV in tricuspid and mitral valve disease are more prominent, than and tend to mask, those produced by the distal lesion. It is vital to recognize multiple valvular involvement if surgical treatment is contemplated, because failure to correct all significant valvular defects at operation results in increased mortality.

Conclusion :

Mitral valve disease traditionally remains the commonest valve to be involved. Chronic rheumatic endocarditis is more aggressive in males in our population as evident from multiple valvular involvement at a relatively younger age When involved alone, MS rather than MR seems to be

predominantly affecting the females. Echocardiography has revolutionized the diagnosis and needs to be done in all suspected cases. The study emphasizes the strong need at government level to encourage prophylactic measures and education programs especially in the rural areas.

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