

ECHOCARDIOGRAPHIC PROFILE OF VALVULAR LESIONS IN CHILDREN WITH ACUTE RHEUMATIC FEVER / RHEUMATIC HEART DISEASE IN A TERTIARY CARE HOSPITALP. Ramu¹, A. Deepak Kumar²**HOW TO CITE THIS ARTICLE:**

P. Ramu, A. Deepak Kumar. "Echocardiographic Profile of Valvular Lesions in Children with Acute Rheumatic Fever/Rheumatic Heart Disease in a Tertiary Care Hospital". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 26, March 30; Page: 4429-4437, DOI: 10.14260/jemds/2015/640

ABSTRACT: CONTEXT (BACKGROUND): Rheumatic Heart disease is still a leading cause of valvular disease in developing countries like India and constitutes 10 to 50% of the cardiac patients in Indian hospitals. Echocardiography is a very sensitive investigation for the diagnosis of Rheumatic Carditis and its sequelae like Mitral, Aortic and Tricuspid valve disease as well as sub clinical Carditis. **AIMS & OBJECTIVES:** To study the profile, severity and gender based differences of valvular lesions on the Trans Thoracic Echocardiographic records of children diagnosed as Acute Rheumatic fever/Rheumatic Heart disease. **MATERIALS & METHODS:** This study was conducted during December'2014 by reviewing the records of trans thoracic Echocardiographic reports of our 36 cases (childhood Acute Rheumatic fever/ Rheumatic Heart disease cases) for type and degree of valvular involvement according to AHA guidelines. **RESULTS:** Mitral valve was involved most often (23 cases – 63.89%). Severe Mitral regurgitation is the common type of valvular lesions both in Boys and Girls (19.44% and 25% respectively). Isolated Aortic valve/ Tricuspid valve involvement was not found in our study. Mixed lesions were seen most often than pure isolated valvular lesions. **CONCLUSION:** Mitral valve was most commonly affected, while Regurgitant lesions were more common than Stenotic lesions and most severe in children less than 15 years age group. Aortic Regurgitation (AR) was found more commonly in Girls than Boys in our study. But all these cases of AR had some other valvular involvement as Mixed lesions. Therefore Echocardiography should be done routinely for patients with Rheumatic Heart Disease, focusing on younger population to facilitate diagnosis and definitive treatment before complications set in.

KEYWORDS: Acute Rheumatic fever, Rheumatic Heart disease, Valvular lesions Mitral regurgitation, Mitral stenosis, Aortic regurgitation, Tricuspid regurgitation, Echocardiography.

INTRODUCTION: In 400 B.C. Hippocrates,¹ a Greek Physician gave the first written description of Acute polyarthritis in man and during the Galenic era of medicine all joint deformities were thought to be due to Gout. In the year 1812, Wells presented the clinico-pathological study in 'Rheumatism of the heart and he described the subcutaneous nodules of rheumatic origin. Later definitive studies were made by Bouillaud and he published his famous treatise in 1836 - 'Nouvelles Recherchessur le Rheumatism Articulaire'. He is known as the Father of Rheumatic Heart disease. In the next 250 years, all the clinical manifestations were recognized as an individual entity and in 1889 Cheadle in his famous classic lecture brought together all the diverse manifestations under one common name 'The Cheadle cycle' or Acute Rheumatic fever as it is known today.

A survey conducted by the Indian council of Medical Research² (ICMR) involving 133,000 children 6 to 16 years in age showed the incidence to be 5.3/1000. Whereas incidence of Rheumatic fever following streptococcal throat infection in the western countries is 0.3 percent in the general

ORIGINAL ARTICLE

population and 1 to 3 percent in crowded communities like army barracks. This shows the magnitude of the problem in the country.

Host factors are also known to play an important role in incidence and also clinical manifestation of Rheumatic fever. Monozygotic twins have higher concordance rate for Rheumatic fever than dizygotic twins suggesting a genetic factor. Sex plays no part in the incidence of Rheumatic fever but has a role in the manifestations of Rheumatic fever. The sexes are nearly equally affected, but Mitral valve disease and Chorea are more common in females whereas Aortic valve, involvement is seen more often in males. Age is an important variable. The commonest age group involved is 5 to 15 years. Although Rheumatic fever is believed to be less common below the age of 5 years, it is unlikely to be so in India, since established juvenile rheumatic Mitral stenosis is often seen in the pediatric age group (below 12 years). Rheumatic fever has been reported in patients in sixties but disease manifestations are altered by increasing age. The recurrence rate is less as the age advances. This is because of the loss of the acquired hyper reactivity to Streptococcal pharyngitis with advancing age.

Rheumatic fever is a multisystemic disease affecting primarily the heart, joints, brain, cutaneous and subcutaneous tissue secondary to an immune reaction to Group A Beta hemolytic streptococcal infection by rheumatogenic strains 1, 3, 5, 6, 18 and 24.³ The frequency of manifestation varies from study to study depending on patient selection whether initial attacks or recurrent attacks were included and on the changing pattern of Rheumatic fever. There has been a declining trend in developed countries which has not been observed in developing countries. Mitral valve involvement is commonest resulting in regurgitation/ stenosis. Aortic valve involvement resulting in aortic regurgitation/ stenosis. Paediatric Mitral stenosis constitutes 10% of all Rheumatic Mitral stenosis patients. But Rheumatic Aortic stenosis is not seen in children. Clinically pure Aortic regurgitation without associated Mitral valve disease is rare and occurs in 5 -8% of patients. Pathologically pure Rheumatic Aortic valve disease is almost unknown. Features indicative of Tricuspid regurgitation are seen in almost 22-50% patients with Rheumatic Heart disease in our country. In individual patients it is difficult to decide whether the Tricuspid regurgitation is organic or functional. Clinical evidence of pulmonary valve involvement in Acute Rheumatic Fever is never seen.

Paucity of criteria to diagnose Acute Rheumatic fever led to dilemma which resulted in under diagnosis or over diagnosis and this prompted Duckett Jones from Boston to propose a set of criteria to diagnose Acute Rheumatic fever. He subgrouped these criteria into major and minor, neither of them having any prognostic connotation in the year 1944.⁴ These original criteria did not differentiate between arthritis and arthralgia and erythema marginatum was classified as minor criteria. Subsequently, the original Jones criteria have been modified four times.⁵ The latest revision by American Heart association⁶ was made in 1992 and according to this, the guidelines are as follows:

MAJOR CRITERIA: Carditis, polyarthritis, chorea, erythema marginatum and subcutaneous nodules.

MINOR CRITERIA: These include clinical findings i.e. fever and arthralgia laboratory findings i.e. elevated acute phase reactants (namely erythrocyte sedimentation rate and C-reactive protein) and prolonged PR interval.

Supportive evidence of antecedent group A streptococcal infection in the form of positive throat culture or rapid streptococcal antigen test or elevated/ rising streptococcal antibody titer.

ORIGINAL ARTICLE

For making a diagnosis of Acute Rheumatic fever (Initial attack), two major or one major and two minor criteria should be present along with supporting evidence of antecedent group A streptococcal infection. The original Jones criteria was modified by American Heart Association after recognizing that Acute Rheumatic fever follows upper respiratory tract infection with Group A Beta hemolytic streptococcus, 2 major or one major and 2 minor criteria are required to diagnose Acute Rheumatic fever.

In 1988 W.H.O has made certain recommendations wherein three special categories in which diagnosis of Rheumatic fever is accepted without 2 major or one major and 2 minor criteria. These are (1) chorea (2) insidious/ late onset carditis with no other explanation (3) Rheumatic recurrence. For (1) and (2) evidence of Streptococcal infection is not required with 'chorea' being a diagnosis of exclusion. In patients with documented Rheumatic heart disease, the presence of one criterion as of fever, arthralgia or elevated acute phase reactants suggests presumptive diagnosis of recurrence with evidence of previous Streptococcal infection.

ECHOCARDIOGRAPHY (2D ECHO)^{7,8,9,10,11}: For the first time, the role of echocardiography was discussed in the latest revision of Jones criteria by American Heart Association (1992) for the diagnosis of Acute Rheumatic fever. A short paragraph was included about the two-dimensional and Doppler echocardiography in evaluation of carditis, however the conclusion was that "at present there is insufficient information to allow the use of echocardiography, including Doppler, to document valvular regurgitation without accompanying auscultatory findings as the sole criterion for valvulitis in Acute Rheumatic fever". These guide lines also highlighted a sub group of "exception to Jones criteria" in which a diagnosis of Rheumatic fever can be made without strictly following the Jones criteria. These were patients with chorea, indolent carditis and those with a previous history of Rheumatic fever or Rheumatic heart disease.

The diagnosis of carditis in Rheumatic fever is usually based on the presence of significant murmurs. But such clinical diagnosis may be difficult due to tachycardia associated with fever. Similarly diastolic murmur of mild Aortic regurgitation may be missed. It is well established that Doppler Echo is more sensitive in picking up minor degrees of valvular regurgitation than clinical examination. Thus there is case of use of Doppler Echo cardiography in diagnosing subclinical carditis or mild forms of carditis, which do not produce audible murmurs. It must be remembered however that trivial regurgitations on color Doppler may occur in normal subjects also. These are common across Mitral and Tricuspid valves but very rarely observed in Aortic valve. To differentiate physiological from pathological leaks, certain guidelines have been given. Physiological leaks usually do not extend more than one cm beyond the valve coaptation point. Also the regurgitant signal does not last throughout the phase of cardiac cycle i.e. a physiological Mitral regurgitation signal is not holosystolic. On the other hand, pathological regurgitation is seen as a substantial color jet in two planes extending well beyond the valve leaflets and the signal lasts all through the phase of cardiac cycle with a well-defined high velocity spectral envelope.

There are several studies on the use of Echocardiography to enhance the yield of cases with carditis. The initial report came from Steinfeld in 1986 where out of 14 children with Acute Rheumatic fever, 5 cases had only Doppler evidence of mild mitral regurgitation; clinically there was no murmurs. The wildly quoted out-break of Acute Rheumatic fever in Utah, USA also utilized echo-Doppler in all 74 children. 14 of these had Doppler, but not clinical evidence of Mitral regurgitation.

ORIGINAL ARTICLE

In another study from Utah, USA published in 1994, echo Doppler identified 15 of 32 patients with polyarthritis and 30 of 53 patients with chorea, has having carditis by demonstrating mitral regurgitation. Vasan et al,¹² however, did not detect any additional case of valvular regurgitation by echo Doppler in 28 patients with Acute Rheumatic fever. Echocardiography with color flow imaging has there-fore provided an important advance in the ability to recognize carditis which is a major criteria. The diagnosis of sub clinical carditis in a given case is likely to influence management. Echocardiography is also helpful to eliminate false positive diagnosis as some of the children of Acute Rheumatic fever may have innocent systolic murmur due to anemia, fever etc.

However, according to the proceedings of the Jones criteria workshop published in 1992, there are insufficient data to support a revision of the Jones criteria and reaffirmed the guidelines iterated in the 1992 statement. In the absence of a “gold standard” for the diagnosis of Rheumatic fever, no single specific laboratory test exists that is pathognomic of Acute Rheumatic fever or its recurrences. At present, Doppler echocardiography should be used as an adjunctive technique to confirm clinical findings and to evaluate chamber sizes, ventricular function, degree of valvular regurgitation, and morphological features of the valves. It should not be used as a major criterion for establishing the diagnosis of Carditis associated with Acute Rheumatic fever in the absence of clinical findings. It is hoped that future refinements of Doppler echocardiography and prospective studies of its predictive value will prompt reassessment of its role in the diagnosis of Acute Rheumatic fever.

OTHER ABNORMALITIES ON ECHOCARDIOGRAPHY: Rheumatic fever produces inflammation, deformation of valves and hence can result in prolapse following elongation and/or rupture of the chordae. The incidence of prolapse has been variously reported. Vasan et al demonstrated valve prolapse in nine of 71 patients with Mitral regurgitation and Acute Rheumatic carditis, a much higher incidence has been reported from China. It is important to remember that Acute Rheumatic carditis can be a cause of valve prolapse.

Vasan et al have described focal nodular thickening of leaflets in 25% of patients with acute carditis. These nodules are present at the tips and bodies of leaflets, measure about 3-5 mm, are more echogenic but do not exhibit the bizarre motion of vegetation. These nodules tend to disappear on follow up. The most common site is Mitral valve, rarely nodules occur on Aortic and Tricuspid valves.

CONCLUSION: There is no single symptom, sign or laboratory test that is diagnostic of Acute Rheumatic fever and carditis. Revised, edited and updated Jones criteria are guidelines to assist practitioners and are not a substitute for clinical judgment. The main concern in liberalizing these criteria in developed countries may be over diagnosis of Acute Rheumatic fever. In developing countries, however, following Jones criteria strictly may result in under diagnosis of this disease.

AIM OF THE STUDY: To study the profile, severity and gender based differences of valvular lesions on the Trans Thoracic Echocardiographic records of children diagnosed as Acute Rheumatic fever/ Rheumatic Heart disease.

METERIAL AND METHODS: This study was conducted during Dec-2014 by reviewing the Trans thoracic Echocardiographic records of 36 Rheumatic Heart disease patients presenting to the Paediatric department of King George Hospital over 2 years period i.e. July 2004 to June 2006. The types and degrees of valvular involvement were defined according to AHA guidelines.

ORIGINAL ARTICLE

In this study we classified a valvular lesion with combination of any degree of stenosis and regurgitation as a mixed lesion. We examined the Echocardiographic reports of 36 children less than 15 years age group (8 cases of 1st attack Acute Rheumatic fever and 28 reactivation cases of established Rheumatic Heart Disease) and included them in our study.

RESULTS: A total of 36 patients 18(50%) Boys, 18(50%) Girls, all less than 15 years age group with Rheumatic Heart Disease were analyzed in this record based study.

Valve condition	Mitral Valve		Aortic Valve		Tricuspid valve	
	Number of cases	%	Number of cases	%	Number of cases	%
Normal valves	2	5.56	2	5.56	2	5.56
Stenosis	1	2.78	0	0	0	0
Regurgitation	23	63.89	15	41.67	19	52.78
Mixed Disease	9	25	0	0	0	0

Table I (n=36): Distribution of 2D Echo - valvular lesions in total study population

In this study only 2 patients had normal valves and both of them were 1st attack cases of Acute Rheumatic fever.

Majority of patients had MR 23(63.89%) followed by TR 19(52.78%) and AR in the 15(41.67%) patients. MS (severe) was found in 1(2.78%) 11 years old female patient and none of our patients had AS/ mixed Aortic valve disease/ TS/ mixed TV diseases.

Overall valvular regurgitations remained commoner than valvular stenoses. Mixed valvular disease was most prevalent in the Mitral valve of 9(25%) patients.

LESION	BOYS	%	GIRLS	%
Mitral Stenosis (MS)	0	0	1	2.78
Mitral Regurgitation (MR)	1	2.78	1	2.78
Aortic Regurgitation (AR)	0	0	0	0
Tricuspid Regurgitation (TR)	0	0	0	0
MS+MR	3	8.33	6	16.67
MS+AR	0	0	0	0
MS+TR	0	0	0	0
MR+AR	1	2.78	2	5.56
MR+TR	6	16.67	3	8.33
AR+TR	0	0	0	0
MS+MR+AR	0	0	0	0
MS+MR+TR	1	2.78	2	5.56
MR+AR+TR	5	13.89	3	8.33
MS+AR+TR	0	0	1	2.78
MS+MR+AR+TR	1	2.78	2	5.56

Table II (n=36): Distribution of the various valvular lesions and their various combinations among Boys & Girls with 2D Echo

ORIGINAL ARTICLE

The data was analyzed with respect to sex (Boys/ Girls) as frequency 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-II. In our study we found mixed Mitral valve disease in 6 Girls (16.67%) and 3 Boys (8.33%). Mitral stenosis was founding a single Girl patient accounting for 2.78% of total cases. Isolated MR was found in 01 Boy (2.78%) and 01 Girl (2.78%). None of our cases had isolated AR or TR or TS. Among the patients with multi valvular involvement MR+AR+TR was common pattern found in our study (includes 5 Boys and 3 Girls) in 8 patients (22.22%) followed by MS+AR+TR (1 Boy and 2 girls) each found in 8.33% of total cases.

All three valves involved in 8 (22.22%) of our patients, combination of Mitral and Aortic valves in 3(8.33%) of patients. Mitral and Tricuspid valve involvement in 9 (25%) and only mitral valve involvement in (includes isolated MS, isolated MR and MS + MR) 12 (33.33%) of our patients.

Valvular lesions	BOYS		GIRLS	
	Number of cases	%	Number of cases	%
Mild MS	0	0	0	0
Mod MS	0	0	0	0
Severe MS	0	0	1	2.78
Mild MR	4	11.11	1	2.78
Mod MR	1	2.78	1	2.78
Severe MR	7	19.44	9	25
Mild AS	1	2.78	0	0
Mod AS	0	0	0	0
Severe AS	0	0	0	0
Mild AR	4	11.11	7	19.44
Mod AR	2	5.56	0	0
Severe AR	1	2.78	1	2.78
Mild TR	2	5.56	2	5.56
Moderate TR	6	16.67	5	13.89
Severe TR	2	5.56	2	5.56

Table III: Frequency distribution of valvular lesions as per their degree of severities (mild, moderate, severe) among Boys and Girls with 2D Echo

Severe Mitral Regurgitation is the common type of valvular lesion both in Boys (7 cases- 19.44%) and Girls (9 cases - 25%).

LESION	Dr. Aswin Reddy study ¹³ (March'2001 to August '2001) n=50 5 to 16 years	%	Present study (July'2004 to June'2006) n=36 <15 years age group	%
MS	1	2	1	2.78
MR	10	20	2	5.56
AR	1	2	0	0

ORIGINAL ARTICLE

TR	0	0	0	0
MS+MR	5	10	9	25
MS+AR	0	0	0	0
MS+TR	4	8	0	0
MR+AR	5	10	3	8.33
MR+TR	5	10	9	25
AR+TR	0	0	0	0
MS+MR+AR	0	0	0	0
MS+MR+TR	11	22	3	8.33
MR+AR+TR	4	8	8	22.22
MS+AR+TR	2	4	1	2.78
MS+MR+AR+TR	2	4	3	8.33

Table IV: Comparative study of various Valvular lesions with 2D Echo in children

Among isolated valvular lesions MR was the most common in the both these studies MS + MR was found in 25% (9) of cases in our study. Where as in Dr. Aswin Reddy study¹³ 10% (5) of cases had MS +MR. MS+MR and MR + TR are the most common combination of valvular lesions found in our study (25% of cases each). Whereas MS+MR+TR in the most common combination of valvular lesions found in Dr. Aswin Reddy study. Isolated Tricuspid valve involvement was not seen in both the studies as in agreement with standard literature and previous studies.

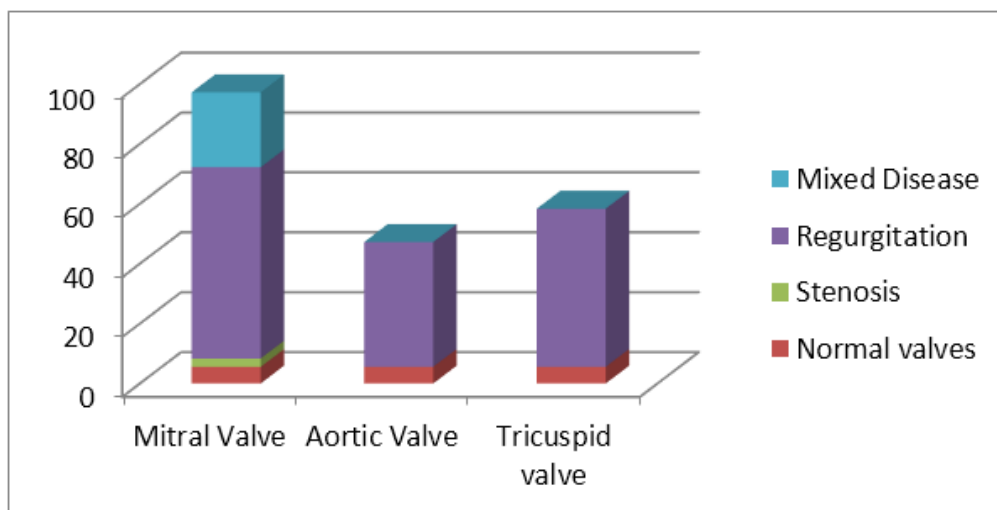


Figure 1

ORIGINAL ARTICLE

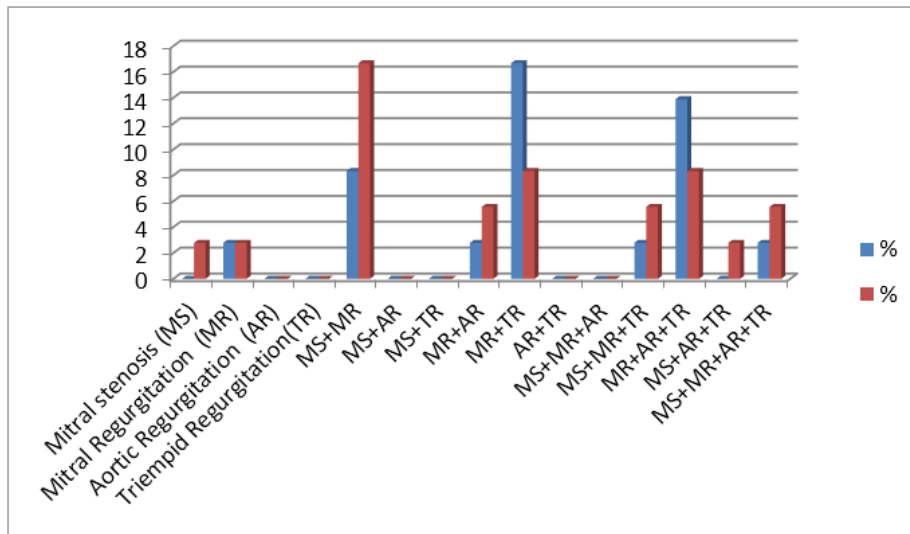


Figure 2

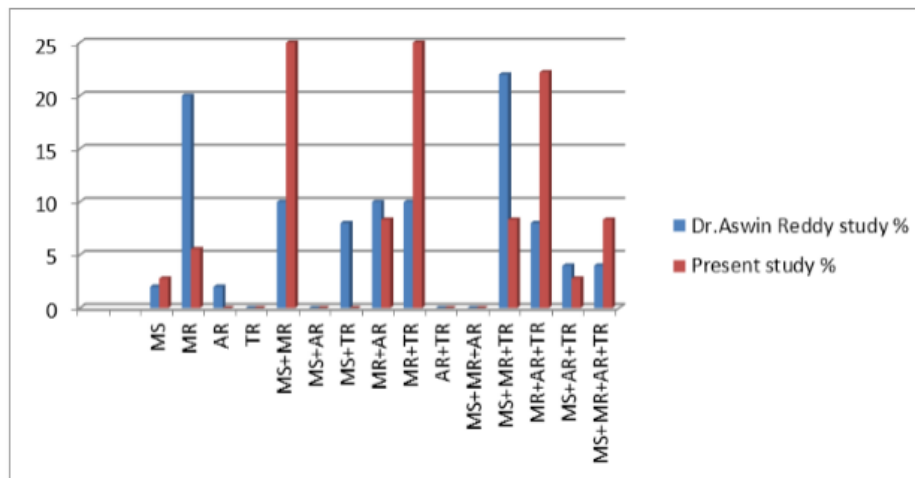


Figure 3

BIBLIOGRAPHY:

1. Hippocrates The genuine works of Hippocrates (translated form Greek by Francis Adams) New York. Wood 1986; 192-273.
2. O. P. Ghi's Essential paediatrics 6th ed-2004; 15.2 & 15.3: 375 – 391.
3. Nelson text book of Paediatrics Daniel ernstein 17th ed-2004; 168 & 430: 874-879 & 1570-1571.
4. Jones TD: Diagnosis of Rheumatic fever. JAMA 1944; 126: 481-484.
5. Committee on Rheumatic fever and bacterial endocarditis of the American Heart association. Jones criteria (revised) for guidance in the diagnosis of Rheumatic fever. Circulation 1984; 69: 203A-208A.

ORIGINAL ARTICLE

6. Guidelines for the diagnosis of Rheumatic fever: Jones criteria, updated 1992, JAMA 1992; 268-2069-73.
7. Sahn DJ, Maciel BC. Physiological valvular regurgitation: Doppler echocardiography and the potential for iatrogenic heart disease. Circulation 1988; 78: 1075-1077.
8. Folger GM Jr., HAJAR R. Doppler echocardiographic findings of mitral and aortic valvular regurgitation in children manifesting only Rheumatic arthritis. Am J Cardiol 1989; 63: 1278-1280.
9. Folger GM Jr., Hajar R, Robida A, Hajar HA. Occurrence of valvular heart disease in Acute Rheumatic fever without evident carditis: colour-flow Doppler identification. Br Heart J 1992; 67: 434-438.
10. Abernethy M, Bass N, Sharpe N et al, Doppler echocardiography and the early diagnosis of carditis in Acute Rheumatic fever. Aust NZ J Med 1994; 24: 530-535.
11. Zhou L, Lu K. Inflammatory Valvular prolapse produced by Acute Rheumatic carditis; echocardiographic analysis of 66 cases of Acute Rheumatic carditis. Int J Cardiol 1997; 58: 175-178.
12. Vasan RS, Shrivastava S, Vijayakumar M, Narang R, Lister BC, Narula J. Echocardiographic evaluation of patients with Acute Rheumatic fever and Rheumatic carditis. Circulation 1996; 94: 73-82.
13. Dr. Ashwin Reddy, Lt Col SK Jatana, Col MNG Nair, MJAFI 2004; Vol. 60.No.3;: 255-258.

AUTHORS:

1. P. Ramu
2. A. Deepak Kumar

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Paediatrics, Andhra Medical College, King George Hospital, Visakhapatnam.
2. Assistant Professor, Department of Paediatrics, Andhra Medical College, King George Hospital, Visakhapatnam.

FINANCIAL OR OTHER

COMPETING INTERESTS: None

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. P. Ramu,
14-1-122/3, FF-3, Rajasagi Residency,
Nowroji Road, Maharani-peta,
Visakhapatnam, Andhra Pradesh-530002.
E-mail: drpramu73@gmail.com

Date of Submission: 03/03/2015.
Date of Peer Review: 04/03/2015.
Date of Acceptance: 19/03/2015.
Date of Publishing: 27/03/2015.