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Original Article

Clinical and Echocardiographic Features of Rheumatic Heart Disease at First Presentation at a Tertiary Care Setup

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ABSTRACT

Objective: To find out the mode of presentation and severity of cardiac involvement in children presented with established rheumatic heart diseases (RHD) at first presentation at a tertiary care setup.

Study Design: Cross-sectional study.

Place and Duration of the Study: Department of Pediatric Cardiology, National Institute of Cardiovascular Disease, Karachi Pakistan, from Jul 2020 to Jun 2021.

Methodology: One hundred seven children of genders aged 3 to 18 with a confirmed diagnosis of RHD were included. Demographic features, chief presenting complaints, possible reasons for late presentation, and echocardiographic findings were noted.

Results: In 107 patients with RHD, 63(58.9%) were male. The sensation of the rapid pounding of heartbeat (palpitation) was observed among 72(67.3%) children, while a similar number of cases, 72(67.3%), presented with breathlessness. There were 33(30.8%) children who were diagnosed for the first time with RHD. The most common reason for late diagnosis was socio-economic factors in children (18, 26.1%). Primary prophylaxis for acute rheumatic fever (ARF)/RHD was noted to be given in 11(10.3%) patients. Mitral regurgitation was the most common echocardiographic finding in 103(96.3%), followed by aortic regurgitation in 99(92.5%).

Conclusion: The majority of children with RHD are diagnosed late, which exposes them to an increased risk of undiagnosed valvular disorders. Palpitation, orthopnea and breathlessness were RHD most frequent presenting clinical features. Mitral regurgitation and aortic regurgitation were the most common echocardiographic findings.

Keywords: Echocardiography, Mitral regurgitation, Rheumatic fever, Rheumatic heart disease.

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INTRODUCTION

Rheumatic heart disease (RHD) is a major cause of valvular heart disease in underdeveloped countries.[1] Global estimates describe that around 12 million people have rheumatic fever or rheumatic heart disease (RHD). At the same time, 2/3rd of these cases are aged 5-15 years, while 79% of the cases hail from developing countries.[2] Around 60% of children with RHD are estimated to develop carditis, which can further influence the recurrence of rheumatic fever, leading to progressive and permanent valvular damage.[3,4] In Pakistan, RHD is one of the leading causes of premature death and disability.[5] Recent studies have reported that the frequency of RHD in Pakistan was around 22/1000 in inner Lahore and 5.7/1000 in rural Pakistan.[6,7] This concurs with the previously available data, putting Pakistan among the high-risk countries for RHD.[8] However, the fact is that in developing countries, medical consultation is often sought when the patient develops disabling symptoms such as congestive heart failure.[9]

Prevention of RHD depends mainly on primary prophylaxis of ARF, recognition of the first episode of ARF, and initiation of secondary prophylaxis if primary prophylaxis fails or is insufficient. Most of the first episodes are not diagnosed in countries with poor health facilities at primary health care centres because arthralgia and arthritis are self-limiting and, if mild, go unnoticed or are just treated by painkillers. Patients with RHD may present with recurrence of ARF, infective endocarditis, or cardiac failure due to decom-pensation, progressively increasing dyspnoea, anaemia, undue exertion and any infection, arrhyth-mias or CVA. This study was planned to find out the mode of presentation and severity of cardiac mani-festation in children presented for the first time with an established diagnosis of RHD at a tertiary care setup.

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METHODOLOGY

The cross-sectional study was conducted at The Department of Paediatric Cardiology, National Institute of Cardiovascular Disease, Karachi Pakistan, from July 2020 to June 2021, after approval from the Institutional Ethical Review Committee was taken (No. ERC-14/2021, Dated Feb 10th, 2021).

With the prevalence of 6% patients of RHD presenting with chorea as chief complaint, sample size was estimated using WHO calculator.[9]

Inclusion Criteria: Children of both genders aged 3-16 years with the diagnosis of rheumatic heart disease, were included.

Exclusion Criteria: Children having isolated aortic valve lesions in the absence of mitral valve abnormalities due to potential overlap with non-rheumatic aortic valve disease, were excluded.

Informed and written consent was acquired from all study participants or their guardians. Non-probability consecutive sampling technique was used. A clinical diagnosis of RHD was made using Jones's criteria.[10] Echocardiographic diagnosis of RHD was labelled if any of the following parameters were present, i) Pathologic MR and at least two morphologic features of RHD of the mitral valve, ii) Mitral stenosis with mean gradient ≥4 mmHg, iii) Pathologic AR and at least two morphologic features of RHD of the aortic valve, iv) Borderline disease of both the aortic and mitral valves. All patients underwent 2-D M-mode, colour Doppler, continuous wave and pulse wave Doppler echocardiography employing standard echocardiographic views. Demographic variables, including age and gender, along with chief complaints, were noted.

A special proforma was designed to record all study information. Statistical Package for Social Sciences (SPSS) version 26.0 was used for the data analysis. Continuous variables were expressed as Mean±SD, while categorical variables were represented as frequencies and percentages.

RESULTS

In a total of 107 patients with rheumatic heart disease, there were 63(58.9%) were male and 44(41.7%) female. Overall, the mean age at presentation was 12.6+3.2 years, while 54 (50.5%) children were between 10 and 15 years of age. Most children, 48(44.9%), belonged to the New York Heart Association (NYHA) Functional Class-IV (Table-I).

Table-I: Characteristics of Children with Rheumatic Heart Disease (n=107)

Characteristics		n(%)
Gender	Male	63(58.9%)
	Female	44(41.7%)
Age (years)	<5	0
	5-10	32(29.9%)
	10-15	54(50.5%)
	>15	21(19.6%)
Province	Punjab	20(18.7%)
	Sindh	60(56.1%)
	Khayber Pakhtunkhwa	9(8.4%)
	Baluchistan	15(14.0%)
	Kashmir/Gilgit Baltistan/Norther Areas	3(2.8%)
Residential Status	Urban	34(31.8%)
	Rural	73(68.2%)
Socio-Economic Status	Upper	2(1.9%)
	Upper Middle	3(2.8%)
	Middle	25(23.4%)
	Lower Middle	39(36.4%)
	Lower	38(35.5%)
Place of Presentation	Out-Patient Department	39(36.4%)
	Emergency	68(63.6%)
NYHA Functional Class	I	2(1.9%)
	II	27(25.2%)
	III	28(26.2%)
	IV	48(44.9%)

In terms of the most frequent chief presenting complaints, the sensation of the rapid pounding of the heartbeat was observed among 72(67.3%) children, breathlessness 72(67.3%), orthopnea 65(60.7%), pedal oedema 65(60.7%), and paroxysmal nocturnal dyspnea 61(57.0%), (Table-II)

Table-II: Frequency of Chief Presenting Complaint in Children with Rheumatic Heart Disease (n=107)

Frequency of Chief Presenting Complaints	n(%)
Fever	41(38.3%)
Swollen and Red Joints	26(24.3%)
Nodules over Swollen Joints	15(14.0%)
Skin Rashes	26(24.3%)
Breathlessness	72(67.3%)
Chest Pain	49(45.8%)
Sensation of Rapid Pounding of Heart Beat	72(67.3%)
Pedal Edema	65(60.7%)
Chorea	10(9.3%)
Syncope	16(15.0%)
Stroke	13(12.1%)
Orthopnea	65(60.7%)
Pain on one Joint that Migrates to Another Joint	34(31.8%)
Paroxysmal Nocturnal Dyspnoea	61(57.0%)

Table-III shows that 33(30.8%) children were diagnosed for the first time with RHD, while recurrent ARF and progressive disease were observed in 22(20.6%) children each. The presentation of the disease was timely in 38(35.5%) children, while

69(64.5%) children were found to have a late diagnosis. RHD disease staging was moderate in most children, as observed in 62(57.9%) cases.

Table-III: Diagnosis, Timing of Diagnosis and Possible Reasons of Delayed Diagnosis in Children with Rheumatic Heart Disease (n=107)

Variables Related to Diagnosis and Timing of Diagnosis		n(%)
Current Diagnosis	First Time Diagnosed as Rheumatic Heart Disease	33(30.8%)
	Recurrence Acute Rheumatic Fever	22(20.6%)
	Infective Endocarditis	16(15.0%)
	Arrhythmias	14(13.1%)
	Progressive Disease	22(20.6%)
Presentation of Disease	Timely	38(35.5%)
	Late	69(64.5%)
Rheumatic Heart Disease Stage	Mild Carditis	13(12.1%)
	Moderate Carditis	62(57.9%)
	Severe Carditis	32(29.9%)

A history of sore throat in the last 14 days was reported in 41(38.3%) children. In 69 cases with late diagnosis, the most common reason for late diagnosis as described by parents/guardians was socio-economic factors in 18(26.1%), the asymptomatic child in 15(21.7%), inadequate counselling by doctors 14(20.3%), lack of education or understanding of the disease on the part of the parents/guardians in 11(15.9%) and limited access to healthcare facilities in 11(15.9%).

Table IV shows echocardiographic findings among children with RHD, and it was seen that mitral regurgitation was the most common echocardio-graphic finding, seen in 103(96.3%), followed by aortic regurgitation in 99(92.5%).

Table-IV: Echocardiographic Findings among Children with RHD (n=107)

Echocardiographic Findings		n(%)
Mitral Regurgitation (n=103)	Mild	0
	Moderate	20(19.4%)
	Severe	83(80.6%)
Mitral Stenosis (n=45)	Mild	30(66.7%)
	Moderate	13(28.9%)
	Severe	2(4.4%)
Aortic Regurgitation (n=99)	Mild	26(26.3%)
	Moderate	47(47.5%)
	Severe	26(26.3%)
LVS Dysfunction (n=76)	Mild	32(42.1%)
	Moderate	44(57.9%)
	Severe	0
Pulmonary Hypertension (n=74)	Mild	36(48.6%)
	Moderate	32(43.2%)
	Severe	6(8.2%)

DISCUSSION

Rheumatic Heart Disease covers a range of various stages that are usually silent. At the same time, clinical manifestations are generally observed as valvular degeneration concluding in congestive heart failure, a rise in the chances of endocarditis and cerebrovascular events, which could further progress into death.[11] In the present study, we noted that 58.9% of the children with RHD were male. Similar to our findings, Nair *et al.* from India [12] noted that 64.8% of the children with RHD were male. A study from Indonesia [13] found 59.4% of cases with RHD to be male. Contrary to our findings, Boyarchuk *et al.* from Ukraine,[14] revealed that 48.5% of the children with RHD were female. We noted that the mean age of the children with RHD in this study was 12.6+3.2 years. Data from Nepal showed that the mean age of children with RHD was 9.6+2.9 years.[15]

In this study, 30.8% of children were newly diagnosed cases of RHD, while recurrent ARF and progressive disease were noted among 20.6% each. Diagnosis of almost 1/3rd of cases for the first time as RHD is alarming, considering 64.5% of the children in the present study had a late diagnosis of RHD. All this puts data from the developed world in perspective, where diagnosis and management of RHD are being made timely/earlier compared to data from the developing world. A study from Ukraine [14] analysing children with RHD revealed that 35.0% of children had recurrent ARF at the time of presentation.

In the present study, breathlessness (67.3%), palpitation (67.3%), followed by orthopnea (60.7%) and oedema (60.7%) were the most frequent clinical presentations among children with RHD, whereas CRP was elevated in 71.0% cases. Fever was seen in 38.3% of children. A study from Indonesia [13] found fever (41.2%). It elevated CRP (38.9%) to be the most frequent clinical presentation among children with RHD, emphasising the importance of thorough evaluation in these children as most clinical presenta-tions are non-specific. Differences are found in the most common types.

Regarding echocardiographic findings, mitral regurgitation was the most common finding observed in 96.3% of children, while aortic regurgitation was found in 92.5% and pulmonary hypertension in 72.9% of cases. Lilyasari *et al.*[13] described mitral regurgitation as the most frequent type of valvular affectation in RHD children. Data from Ukraine[14] suggested mitral regurgitation to be the most common echocardio-

graphic finding (43.8%) among children with RHD. Valvular disorders in patients with RHD are also credited to genetic predisposition.[15-17] In developing countries, lack of primary or secondary prophylaxis for RHD and frequent recurrent ARF might expose children to increased chances of valvular disorders.[18,19] Past local data suggested mitral stenosis followed by mitral regurgitation to be the commonest valvular defects, which shows that change in patterns of RHD in children is observed in the local population, which warrants further research.[7]

In recent years, the present study has emerged as the most prominent in describing the clinical presentation, diagnostic features, and echocardiographic findings among children with RHD from Pakistan. The present study also highlighted more pronounced clinical presentation, lack of rheumatic prophylaxis and occurrence of valvular disorders in children with RDH, which emphasises the need for cost-effective and easily accessible healthcare support.

LIMITATIONS OF STUDY

We could not gather follow-up data about possible complications and prognosis among the current children, and no data about managing children with RHD were analysed in the present study.

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CONCLUSION

The majority of children with RHD are diagnosed late, which exposes them to an increased risk of valvular disorders. Palpitation, orthopnea and breathlessness were RHD most frequent presenting clinical features. Mitral regurgitation and aortic regurgitation were the most common echocardiographic findings.

Conflict of Interest: None.

Authors' Contribution

Following authors have made substantial contributions to the manuscript as under:

FR & RC: Conception, study design, drafting the manuscript, approval of the final version to be published.

AA & SS: Data acquisition, data analysis, data interpretation, critical review, approval of the final version to be published.

ASS & MR & NP: Data acquisition, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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