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Original Article

Clinical Characteristics, Echocardiographic Features And Short-Term Outcomes of Children with Rheumatic Heart Disease

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ABSTRACT

Objective: To find out the clinical characteristics, echocardiographic features and short-term outcomes in children with rheumatic heart diseases (RHD).
Study Design: Prospective longitudinal study
Place and Duration of Study: The department of pediatric cardiology, "Children's Hospital & Institute of Child Health, Multan" from Sep 2021-Feb 2022.
Methodology: A total of 73 children of both genders aged 4 to 18 years with RHD were included. Clinical characteristics, presenting complaints and echocardiographic features were noted. Duration of stay among children admitted and managed as well as in-hospital outcomes in terms of mortality were recorded
Results: Out of 73 children with RHD, 44(60.3%) were boys. Mean age was 11.78±3.62 years. Breathlessness, feeling of rapid ponding of heart-beat and orthopnea were the most frequent presenting complaints in 50(68.5%), 46(63.0%) and 41(56.2%) children respectively. Primary prophylaxis for ARF/RHD was reported in 6(8.2%) children. Echocardiographic assessment revealed mitral regurgitation in 71(97.3%), mitral stenosis 27(37.0%), aortic regurgitation 68(93.2%), left ventricular systolic dysfunction 50(68.5%) and pulmonary hypertension in 51(69.9%) children. Mean duration of hospital stay among admitted children was 12.57±2.77 days. Mortality was noted in 2(2.7%) children.
Conclusion: Majority of the children with RHD are diagnosed late that exposes them to increased risk of undiagnosed valvular disorders. Palpitation, orthopnea and breathlessness were the most frequent presenting clinical features of RHD. 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 considered to be an important cause behind Valvular heart disease especially in developing countries.[1] Globally, 15 million people are estimated to have RHD while around 66% of those cases are between 5-15 years of age whereas nearly 80% of these patients are from under-developed regions.[2,3] Among pediatric patients with RHD, 3 out of 5 children are observed to develop carditis that can cause recurrent rheumatic fever which can further lead in to progressive and/or permanent vuvular heart disease.[4,5] Pakistan lies among countries which are considered to be high risk countries for RHD while recent local data suggests incidence of RHD to be between 5.7-22 per 1,000 population.[6-8]

Prophylaxis of acute rheumatic fever (ARF) is necessary as part of the preventive strategies against RHD while starting secondary prophylaxis in cases primary prophylaxis fails or does not provide appropriate cover is considered to be the way forward. Many of the cases of RHD in a developing country like Pakistan are diagnosed late as many patients are only brought to healthcare facilities when they have developed disabling issues like congestive heart failure.[5] Many of the ARF cases present with arthralgia and/or arthritis and many of these patients accompany mild symptoms that could be self-limiting or go away after administering pain killers so perception is that diagnosis of RHD is delayed in many cases in a resource constricted country like Pakistan. Common mode of presentation among RHD cases is the recurrence of ARF, infective endocarditis, dyspnea, anemia, infections, arrhythmias, cerebrovascular accident or cardiac failure.[9] but in Pakistan, not much is known about the typical presentation, clinical features, echocardiographic findings or outcomes of patients with RHD so the

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present study was planned. Objective of this study was to know the mode of presentation, echocardiographic features and short-term outcomes among patients of RHD at a leading tertiary childcare facility of South Punjab, Pakistan.

METHODOLOGY

This Prospective longitudinal study was done at the "department of pediatric cardiology, Children's Hospital & Institute of Child Health, Multan" from 1st September 2021 to 28th February 2022. Approval was acquired from "institutional ethical committee" (letter number 240/21, dated 25-08-20221). Written consents were sought from parents/caregivers of all study participants.

Inclusion Criteria: were children of both genders from 1 to 18 years of age with RHD were included.

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

Diagnosis of RHD: RHD was labeled clinically as per "Jone's Criteria".[10] The diagnosis of RHD was further confirmed echocardiographically if any of these were found: a) pathological mitral regurgitation and ≥2 morphological features of RHD of the mitral valve; b) mitral stenosis (MS) having mean gradient above or equal to 4 mmHg; c) pathological aortic regurgitation and ≥2 morphological features of RHD of aortic valve;d) borderline diseases of aortic as well as mitral valves.

All echocardiography examinations were performed by a consultant pediatric cardiologist. All children underwent 2-D M-mode, color Doppler, continuous wave and pulse wave doppler echocardiography employing standard echocardiographic views. Infective endocarditis was diagnosed as per "Duke's Criteria". During the study period, a total of 73 patients as per inclusion/exclusion criteria were included. Non-probability consecutive sampling technique was employed. Demographic variables including age and gender along with presenting complaints were recorded. Duration of stay among children admitted and managed as well as in-hospital outcomes in terms of mortality were recorded. Short-term outcome was noted among children who were admitted as per institutional criteria up till they were either discharged or died. A special proforma was designed to record all study information.

Data was analyzed using "Statistical Package for Social Sciences (SPSS)" version 26:00. Continuous variable were expressed as Mean±SD while categorical variables were represented as frequencies and percentages.

RESULTS

Out of 73 children with RHD, 44(60.3%) were boys and 29(39.7%) girls. Mean age was 11.78±3.62 years (ranging between 4 to 18 years). Majority of the children, 37(50.7%) were aged between 10 to 15 years. Residential status of 48(65.8%) children was rural. A total of 33(45.2%) children belonged to "New York Heart Association (NYHA)" Functional Class-IV while 21(28.8%) children belonged to NYHA functional class-III. Table-I is highlighting socio-demographic characteristics of all children.

Table-I: Socio-Demographic Characteristics of Children with Rheumatic Heart Disease (n=73)

Characteristics		Number (%)
Sex	Boys	44(60.3%)
	Girls	29(39.7%)
Age (years)	<5	2(2.7%)
	5-10	20(27.4%)
	10-15	37(50.7%)
	>15	14(19.2%)
Residential Status	Urban	25(34.2%)
	Rural	48(65.8%)
Socio-Economic Status	Low	52(71.2%)
	Middle	17(23.3%)
	High	4(5.5%)
NYHA Functional Class	I	1(1.4%)
	II	17(23.3%)
	III	21(28.8%)
	IV	33(45.2%)

Table-II is showing frequency of most common presenting complaints among children with RHD and breathlessness, feeling of rapid ponding of heart-beat, pedal edema and orthopnea were the most frequent presenting complaints noted among 50(68.5%), 46(63.0%) and 41(56.2%) children respectively.

Nineteen (26.0%) children were newly diagnosed while recurrent ARF was diagnosed in 17(23.3%) and progressive disease in 15(20.5%) children with RHD. RHD staging revealed mild, moderate and severe carditis in 7(9.6%), 42(57.5%) and 24(32.9%) children respectively. Table-III is showing diagnosis and disease severity among children with RHD. Primary prophylaxis for ARF/RHD was reported in 6(8.2%) children. ESR was raised in 50(68.5%) cases while C - reactive protein was raised in 55(75.3%) cases.

Prolonged PR internal on ECG was observed in 33(45.2%) children.

Table-II: Frequency of Presenting Complaint in Children with Rheumatic Heart Disease (n=73)

Frequency of Most Common Presenting Complaints	Number (%)
Breathlessness	50(68.5%)
Feeling of Rapid Pounding of Heart Beat	46(63.0%)
Orthopnea	41(56.2%)
Chest Pain	34(46.6%)
Fever	31(42.5%)
Pain Mitigating from One Joint to Another	24(32.9%)
Swollen/Red Joints	17(23.3%)
Fainting	15(20.5%)
Paroxysmal Nocturnal Dyspnea	41(56.2%)

Table-III: Diagnosis and Disease Severity among Children with Rheumatic Heart Disease (n=73)

Diagnosis and Disease Severity		Number (%)
Diagnosis	Newly Diagnosed RHD	29(39.7%)
	Recurrent Acute Rheumatic Fever	25(34.2%)
	Progressive Disease	19(26.0%)
Rheumatic Heart Disease Staging	Mild Carditis	7(9.6%)
	Moderate Carditis	42(57.5%)
	Severe Carditis	24(32.9%)

Echocardiographic assessment revealed mitral regurgitation (MR) in 71(97.3%), mitral stenosis 27(37.0%), aortic regurgitation (AR) 68(93.2%), left ventricular systolic dysfunction (LVDS) 50(68.5%) and pulmonary hypertension (PHTN) in 51(69.9%) children. Figure-I is elaborating frequency distribution of echocardiographic features revealed in children with RHD.

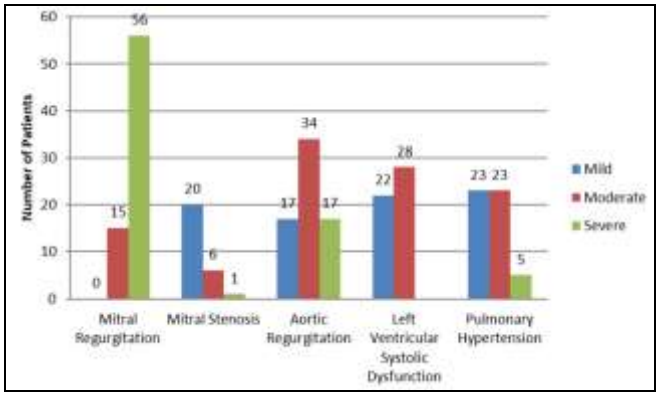

Figure-I: Frequency of Echocardiographic Findings among Children with Rheumatic Heart Disease (n=73)

A total of 49(67.1%) children were admitted and managed as per institutional management protocols of RHD. Mean duration of hospital stay among admitted children was noted as 12.57±2.77 days. Two (2.7%) children died during the hospital stay while all other 71(97.3%) children were discharged successfully and are now on follow ups. Among 2 children who died, both presented with congestive heart failure while during hospital stay, they developed arrhythmias and refractory heart disease.

DISCUSSION

Some of the major complications of RHD comprise of severe Valvular regurgitation, heart failure, stroke and infective endocarditis. The incidence of RHD has been minimizing in the developed countries while in developing regions, RHD still remains an important health concern inflicting considerable morbidity and mortality.

In this research, 60.3% of RHD cases were boys. A local research conducted in rural areas of Pakistan.[7] revealed that that majority of the RHD cases (53.3%) were boys which correlate well with the present findings. Regional data has also reported 64.8% cases of RHD to be boys.[10] Overall, mean age was 11.78±3.62 years in this study which is close to has been reported by Lilyasari O *et al.*[11] and Shrestha NR *et al.*[12] as 12.1+3.4 years and 9.6+2.9 years respectively. Literature highlights younger school going children as well as adults of child bearing age to be the most affected age groups for RHD.[13] but as we had only considered cases up to 18 years of age, this could be the reason why most the children in the present study were aged between 10 to 15 years.

In the present work, 39.7% children were newly diagnosed RHD cases while recurrent ARF and progressive disease were reported in 34.2% and 26.0% cases respectively. A study by Gray LA *et al.*[14] found 35.0% children with RHD to have recurrent ARF which again emphasizes that recurrent ARF is a common diagnosis at the time of presentation among patients with RHD. Breathlessness (68.5%) and feeling of rapid pounding of heart-beat (63.0%) were found to be the most frequently presenting complaints among children with RHD. Fever was noted in 42.5% children. Fever and arthralgia were the most common presenting complaints observed by Lilyasari O *et al* from Indonesia in 41.7% and 25.0% children with RHD respectively.[11] Abrar A *et al.*[15] in a local study found palpitation (54.0%), dyspnea (63.0%) and breathlessness (45%) to be the most common presenting complaints in patients with RHD.

MR was observed to be the most common echocardiographic finding in 97.3%, AR in 93.2%, LVDS in 68.5% while PHTN was seen in 69.9% RHD cases. A study from Indonesia showed isolated MR to be commonest echocardiographic findings observed in 61.9% RHD cases.[11] Regional population based data of echocardiographic evaluation among 2060 school going children revealed MR and AR to be the most common echocardiographic findings noted in 146 and 97 children respectively.[10] Literature suggests valvular disorders among RHD cases are largely attributed to genetic predisposition.[16] Researchers from Pakistan have alreay shown MS and MR to be the most frequent causes of valvular heart diseases.[7] Primary prophylaxis for ARF/RHD was noted in 8.2% children in this study. In a country like Pakistan, lack of primary as well as secondary prophylaxis of RHD and recurrence of ARF is exposing our children to raised risk for the development of vulvular disorders.[17,18] The findings of the present research are thought to add useful insights into most common patterns of presentation, clinical and echocardiographic features among children with RHD.

Limitations of the Study: As this was a single center study with a relatively small sample size, our findings cannot be generalized. As we only noted short-outcomes among children with RHD, further research is warranted to note most common complications and long-term outcomes among children with RHD.

CONCLUSION

Breathlessness and feeling of rapid pounding or heart-beat were the most common presenting complaints among children with RHD. Mitral regurgitation, aortic regurgitation and pulmonary hypertension were the most commonly noted echocardiographic abnormalities. Very few children with RHD had primary prophylaxis which highlights gaps regarding care of these children.

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Authors' Contribution

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

HMAUH & MSA: Data acquisition, data analysis, critical review, approval of the final version to be published.

MA & HA: Study design, data interpretation, drafting the manuscript, critical review, approval of the final version to be published.

ST: Conception, 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.

REFERENCES

1. Joseph N, Madi D, Kumar GS, Nelliyanil M, Saralaya V, Rai S. Clinical spectrum of rheumatic Fever and rheumatic heart disease: a 10 year experience in an urban area of South India. N Am J Med Sci. 2013; 5(11): 647-652.
2. Animasahun BA, Madise Wobo AD, Itiola AY, Adekunle MO, Kusimo OY, Thomas FB. The burden of rheumatic heart disease among children in Lagos: how are we fairing?. Pan Afr Med J. 2018; 29: 150.
3. Zuhlke LJ, Steer AC. Estimates of the Global Burden of Rheumatic Heart Disease. Global Heart. 2013; 8(3): 189–195. https://doi: 10.1016/j.gheart.2013.08.008
4. Carapetis JR. Rheumatic heart disease in developing countries. N Engl J Med. 2007; 357(5): 439-441.
5. Riaz A, Hanif MI, Khan IH, Hanif A, Mughal S, Anwer A. Quality of life in patients with rheumatic heart disease. J Pak Med Assoc. 2018; 68(3): 370-375.
6. Akhtar N, Sadiq M, Chagani H, Hafeez A, Rizvi FH, Mehboob M. Guidelines for prevention of Rheumatic fever and rheumatic heart disease. Pak J Cardiol 2004; 15: 136–48.
7. Rizvi SF, Khan MA, Kundi A, Marsh DR, Samad A, Pasha O. Status of Rheumatic heart disease in rural Pakistan. Heart 2004; 90(4): 394–399.
8. Ilyas M, Peracha MA, Ahmed R, Khan N, Ali N, Janjua M. Prevalence and pattern of rheumatic heart disease in the Frontier Province of Pakistan. J Pak Med Assoc 1979; 29(8): 165–168.
9. Makrexeni ZM, Pepeta L. Clinical presentation and outcomes of patients with acute rheumatic fever and rheumatic heart disease seen at a tertiary hospital setting in Port Elizabeth, South Africa. Cardiovasc J Afr. 2017; 28(4): 248-250. https://doi:10.5830/CVJA-2017-019
10. Mach DS. Revised Jones Criteria for acute rheumatic fever; Ten points to remember. https://www.acc.org/latest-in-cardiology/ten-points-to-remember/2015/05/08/15/22/revision-of-the-jones-criteria-for-the-diagnosis-of-acute-rheumatic-fever (accesses on 30th January 2022)
11. Lilyasari O, Prakoso R, Kurniawati Y, Roebiono PS, Rahajoe AU, Sakidjan I, et al. Clinical profile and management of rheumatic heart disease in children and young adults at a tertiary cardiac center in Indonesia. Front Surg. 2020; 7: 47.
12. Shrestha NR, Kalesan B, Karki P, Sherpa K, Basnet A, Urban P, et al. Rheumatic heart disease: pilot study for a population based evaluation of prevalence and cardiovascular outcomes among schoolchildren in Nepal. BMJ Open 2012; 2(5): e001616.
13. Cilliers AM. Rheumatic fever and rheumatic heart disease in Gauteng on the decline: Experience at Chris Hani Baragwanath Academic Hospital, Johannesburg, South Africa. S Afr Med J. 2014; 104(9): 632-4. https://doi: 10.7196/samj.8318.
14. Gray LA, D'Antoine HA, Tong SYC, McKinnon M, Bessarab D, Brown N, et al. Genome-wide analysis of genetic risk factors for Rheumatic Heart Disease in Aboriginal Australians provides support for pathogenic molecular mimicry. J Infect Dis. 2017; 216(11): 1460-1470.
15. Abrar A, Khan S, Rehman MU, Jan T, Faisal M, Khan N. Frequency of rheumatic heart disease in patients undergoing echocardiography in district dera ismail khan. Gomal J Med Sci. 2014; 12(3): 147-150.

16. Muhamed B, Shaboodien G, Engel ME. Genetic variants in rheumatic fever and rheumatic heart disease. Am J Med Genet C Semin Med Genet. 2020; 184(1): 159-177.
17. Antunes MJ. The global burden of rheumatic heart disease: Population-related differences (it is not all the same!). Braz J Cardiovasc Surg. 2020; 35(6): 958-963.
18. Dougherty S, Beaton A, Nascimento BR, Zühlke LJ, Khorsandi M, Wilson N. Prevention and control of rheumatic heart disease: Overcoming core challenges in resource-poor environments. Ann Pediatr Cardiol. 2018; 11(1): 68-78.