

Rheumatic Heart Disease Occurrence, Patterns and Clinical Correlates in Children Aged Less than Five Years

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Background and aim of the study: Rheumatic fever (RF) and chronic rheumatic heart disease (RHD) are common in developing countries. Two-thirds of RHD patients are school-children aged between 5 and 15 years. Pre-schoolers aged <5 years are not immune to RF however, and to date RHD patterns in this very young age group have not been studied systematically.

Methods: Records of all RHD patients seen at the authors' institution between January 1999 and December 2000 were retrospectively reviewed. A special analysis was conducted among pre-school children aged <5 years.

Results: Thirty-eight (6.8%) of the RF/RHD admissions were aged <5 years, and 28 of these patients (20 males, 8 females) presented with acute RF. The mean age of acute RF diagnosis was 4 years. All RF/RHD patients aged <5 years were in normal sinus rhythm. Joint pain and swelling (25 cases; characteristic migratory polyarthritis in six, monoarthritis in five) and fever (24 cases) were the most frequent symptoms. Arthritis, carditis and chorea occurred in 75%,

Today, acute rheumatic fever (RF) and its sequelae, chronic rheumatic heart disease (RHD), account for about one-third of cases in the cardiology services of developing countries (1-3). Currently, two-thirds of RHD patients are of school age, from 5 to 15 years (4). Due to the high incidence of an accelerated course of RF and progressive multivalvular disease among the developing nations, overall life expectancy in these patients may be as little as 20-25 years (5,6).

Low socioeconomic status, poor hygiene, overcrowding, and close person-to-person contact account

50% and 4% respectively, with no instances of erythema marginatum or subcutaneous nodules. Effort intolerance, chest discomfort and palpitations were reported by nine, five and three cases, respectively. Mitral regurgitation was the most common valvular lesion in RF. The youngest case of confirmed acute RF was an 18-month-old male. The only patient with mitral stenosis in the present series was a 4-year-old girl. None of the patients required surgical intervention, and there were no deaths.

Conclusion: RHD is common in very young age groups of <5 years. Pre-schoolers account for a significant proportion of acute RF and chronic RHD admissions among children. Mitral regurgitation is the most common cardiac manifestation, but obstructive valve disease is distinctly rare in this age group. Aortic regurgitation, left ventricular dysfunction and pulmonary hypertension may complicate the course of RF in these very young children.

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for the high RHD incidence of four per 1000 seen among school children (7-9). This is in stark contrast to the very low incidence of RHD reported over the past three decades from Western populations (10).

The prevalence and patterns of RF/RHD in school-going children aged between 5 and 15 years has been well studied. Children aged <5 years are not immune to RF, though as yet patterns of RHD in this very young age group have not been studied systematically. The present study represents an analysis of the authors' experience of RHD among preschool children aged <5 years, the aim being to gain a better understanding of the condition in this age group.

Materials and methods

The records of all RHD patients attending the Pediatric Cardiology Department of the authors' institution between January 1999 and December 2000 were

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analyzed retrospectively. All acute RF and chronic RHD patients aged <5 years were eligible for the study. Acute RF was diagnosed according to updated Jones' criteria (11). Chronic RHD was evaluated clinically, and the diagnosis confirmed echocardiographically. Inclusion criteria for the study were hospital admission age <5 years, and a definite diagnosis of RF conforming to Jones' criteria, or of chronic RHD by echocardiography. Exclusion criteria were age >5 years, failure to meet Jones' criteria for acute RF, a lack of echocardiographic confirmation of chronic RHD, and incomplete evaluation or case records.

Results

A total of 556 consecutive RF/RHD admissions to the pediatric cardiology service was screened, after having excluded readmissions. Among these patients, 38 (6.8%) were aged <5 years. Acute RF accounted for 28 admissions, and recurrence of RF in known RHD accounted for the remaining.

The mean age of the 28 patients (20 males, eight females) diagnosed with acute RF was 4 years. All RF/RHD patients aged <5 years were in normal sinus rhythm. The most frequent presenting symptoms were joint pain and swelling (25 cases, with characteristic migratory polyarthritis in six cases and monoarthritis in five) and fever (24 cases). Effort intolerance, chest discomfort and palpitations were reported by nine, five and three cases, respectively. The incidence of Jones' criteria features among the acute RF subjects (n = 28) are listed in Table I

Carditis was initially diagnosed by auscultation of systolic murmur in 10 of the 14 patients who subsequently had echocardiographic confirmation of the condition. Electrocardiography showed inappropriate age-corrected sinus tachycardia in 20 cases, and PR interval prolongation in five. Chest X-radiography revealed an increased cardiothoracic ratio in eight cases, and pulmonary venous hypertension (grade I-II) in four.

The echocardiographic features of acute rheumatic carditis included mitral and sometimes aortic regurgitation, mitral valve prolapse, annular dilatation, myocardial dysfunction and pericardial effusion. Chronic RHD typically produces characteristic mitral valve thickening, reduced posterior mitral leaflet mobility and, commonly, diastolic doming of the mitral leaflets. Regurgitation and stenosis occur based on the degree of deformity, fibrosis and commissural fusion. Rheumatic reactivation can result in reduced myocardial contractility and pericardial effusion, apart from worsening of the regurgitant lesions.

Echocardiographically confirmed mitral regurgitation (mild in nine cases, moderate in five) was the most

Table I: Jones' criteria among acute rheumatic fever patients (n = 28) aged <5 years.

Jones' criteria	n
Major criteria	
Arthritis	21 (75)
Carditis	14 (50)
Chorea	1 (4)
Subcutaneous nodules	0 (0)
Erythema marginatum	0 (0)
Minor criteria	
Arthralgia	25 (89)
Fever	24 (85)
ESR >40 mm at 1 h	28 (100)
C-reactive protein	20 (71)
PR interval prolongation	5 (18)
Essential criteria	
Positive throat culture	2 (7)
ASO titer >330 Todd units	15 (53)

Values in parentheses are percentages.

ASO: Anti-streptolysin O; ESR: Erythrocyte sedimentation rate.

common valvular lesion in RF cases. All recurrent RF cases had mitral valve thickening and reduced mobility. Mild (n = 2) and moderate (n = 1) aortic regurgitation was detected in three cases with mitral involvement.

Myocarditis with severe left ventricular dysfunction (ejection fraction 22%) due to the first episode of RF occurred in one patient, who presented with fever and swollen tender joints and mild mitral regurgitation and minimal pericardial effusion. Cardiac function was normalized over a three-week period with steroid treatment, antifailure medication and supportive care.

Pulmonary hypertension related to tricuspid regurgitation (Doppler-quantified) was identified in four cases (mean age 4 years), all of whom had a prior history suggestive of RF. Moderate mitral regurgitation was detected with mild tricuspid regurgitation. Rheumatic tricuspid valve involvement was not detected in any of these cases.

The youngest case of confirmed acute RF was an 18-month-old male child who presented with a two-week history of fever and joint swelling. Cardiac auscultation was normal. The erythrocyte sedimentation rate (ESR) was raised at 55 mm/h, and the anti-streptolysin O (ASO) titer was elevated to 440 Todd units. Electrocardiography and chest radiography were normal, except for sinus tachycardia. Echocardiography identified the presence of mild mitral regurgitation and mild pericardial effusion. The patient responded

well to salicylates and steroids, and was discharged on oral prophylactic penicillin to treat the RF. At 28 months of age, the patient was well and free of RF recurrence.

The only patient with mitral stenosis in the present series was a 4-year-old female child who presented with RF recurrence. She had had two previous episodes of RF from the age of 2.5 years, and was not compliant with penicillin prophylaxis which had been prescribed. Echocardiography demonstrated the presence of moderate mitral stenosis with a mitral valve area 1.4 cm² (1.34 cm²/m²) measured using planimetry and pressure half-time methods. Left atrial enlargement, mild pulmonary hypertension and mild aortic regurgitation were also identified.

Infective endocarditis was suspected in three cases, but subsequently confirmed in none. All cases were receiving medical management and none of the patients required surgical intervention. There were no RHD-related deaths in this age group.

Discussion

Almost 7% of RF/RHD cases in this pediatric study occurred in the <5-year age group. Only one other study has been reported in very young children, and this was conducted in a Mexican RF/RHD series with patients aged <6 years (12). The steep rise in the incidence of RF after the age of 3 years was also noted in the latter report, with 12 patients (33%) aged 3 years and 19 (52%) aged 4 years. On occasion, patients present with polyarthritis and fever, but fail to meet the criteria for RF. Although initially normal, these patients have a higher than baseline risk for later development of RHD. Thus, close long-term follow up and penicillin prophylaxis seems prudent, especially in regions where this condition is endemic (8).

The occurrence of many of the Jones' criteria in non-specific intercurrent illness reduces its specificity. Of note, further testing beyond throat swab and ASO titer are not usually carried out due to cost issues. In the absence of migratory large-joint arthritis, rapid response to salicylates, remission without residue and an absence of any other evidence of connective tissue disorders can aid in diagnosing rheumatic carditis.

In the present study, the clinical presentation and echocardiographic findings of acute RF were compared among various age groups. Polyarthritis was identified more, and carditis less, in subjects aged <5 years, but this difference may have been due to earlier unrecognized occult RF episodes among older age groups. Clinical suspicion is vital in very young RF patients, as non-specific intercurrent infection-related arthritis is commonly encountered, and polyarthritis may not demonstrate a typical migratory nature. The

limitation of physical activity, together with cardiac symptoms may take longer to recognize. In addition, the systolic murmur of mitral regurgitation in RF may be mistaken for innocent pediatric murmurs and excess flow related to fever.

Thus, even in countries where RHD is endemic and physicians routinely diagnose carditis by auscultation, echocardiography may still play a significant role (13). Diagnosing carditis in this age group may involve very subtle two-dimensional echocardiographic changes such as mild leaflet thickening, a slight reduction in mobility, leaflet doming without obstruction, and insignificant regurgitation. A pansystolic, eccentric color Doppler jet extending beyond the mitral leaflets differentiates carditis from physiological mitral regurgitation. In the present series, echocardiography offered a definitive diagnosis of RF in 14 of the 28 RF cases. Serial echocardiography with documentation of interval changes may help to clinch the diagnosis in more cases, but these longitudinal follow up data were not available in the present investigation.

The low incidence of obstructive lesions is due to the longer time course required for the development of fibrosis and scarring of valvulitis, which results in commissural fusion. Regurgitation on the other hand can manifest early due to incorrect coaptation of the valve margins caused by small verrucae of RF, and reduced pliability of the mitral apparatus in RHD.

The natural history and overall disease progression has been observed to be more rapid and fulminant with an earlier age of RHD onset. Early diagnosis and effective antibiotic prophylaxis may thus assume more importance in this setting (14).

In conclusion, RHD is common in the very young, namely those aged <5 years, and pre-schoolers account for a significant proportion (6.8%) of acute RF and chronic RHD admissions among children. Mitral regurgitation is the most common cardiac manifestation, while obstructive valve disease is distinctly rare in this age group. Aortic regurgitation, left ventricular dysfunction and pulmonary hypertension may complicate the course of RF in this age group.

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