A CLINICAL, LABORATORY AND ECHOCARDIOGRAPHIC PROFILE OF CHILDREN WITH ACUTE RHEUMATIC FEVER

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ABSTRACT

A retrospective study of 42 children with acute rheumatic fever admitted to Hospital Universiti Sains Malaysia from April 1985 to March 1989 was undertaken to assess the clinical, laboratory, echocardiographic aspects and outcome. The ages of the children ranged from 5 years 9 months to 11 years 11 months. There was no significant sex difference. 69.4% were admitted between November and April with a seasonal low between May and August. Sixteen children (38.1%) were hospitalised for recurrence of rheumatic fever. Carditis was the commonest manifestation and was seen in 28 (66.6%) children, followed by arthritis in 24 (57.1%), and chorea in 3 (7.1%). Echocardiography detected abnormalities in 24 out of 35 cases and the most common echocardiographic findings were poor coaptation of mitral valve (ten)left ventricular dilatation (ten), thickened mitral valve cusps (seven) and pericardial effusion (seven). In those children followed up, there were 2 recurrences while on secondary prophylaxis and complete recovery was seen only in 11 (26.9%).

Keywords : Acute rheumatic fever, Malaysia, echocardiography.

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INTRODUCTION

Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) continue to be major health problems amongst children from various countries across the world [1]. Studies published have demonstrated conflicting views and facts about the true geographic difference in clinical profile and outcome [1]. However, we were unable to find recent Malaysian literature describing the incidence of ARF or prevalence of RHD and the clinical profile of the illness in the local population. This is surprising since ARF and RHD have been shown to be the commonest aetiology of acquired heart disease in children in many Asian countries [1-4]. In the present study, we describe the clinical profile, results of laboratory investigations, echocardiographic features in the acute phase of ARF and the outcome in Kelantanese children in our hospital.

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MATERIALS AND METHODS

The materials for the study consisted of a consecutive series of 42 hospitalised children aged less than 12 years old, admitted to Hospital Universiti Sains Malaysia (HUSM) between April 1985 and March 1989. HUSM is a teaching hospital and a tertiary referral centre for the state of Kelantan and Nothern Terengganu, ARF is also treated in the district hospitals in Kelantan. All these children were diagnosed to have ARF based on the revised Jones Criteria, 1984 [5]. Of the children presenting with arthralgia and fever, only those who met the Jones Criteria by the presence of carditis or subsequently developing carditis or arithritis were included in the study. Sydenham's chorea was accepted as ARF in the absence of other criteria or supporting evidence after excluding other possible causes of chorea. Children with established RHD without evidence of recurrence of carditis or ARF were excluded from the study. Similarly, one child suspected of suffering from ARF was excluded because he had a positive blood culture for group A betahaemolytic streptococci. Carditis was diagnosed on the basis of the presence of congestive cardiac failure, cardiomegaly, new significant murmurs and pericarditis with or without first degree heart block in the electrocardiogram. All the cases were evaluated by a history, physical examination and laboratory investigations. Echocardiography was performed on 35 children. Thickened mitral valve cusps and M-mode appearance suggestive of mitral stenosis or aortic valve lesions indicated a chronic rheumatic heart disease but if clinically present with congestive heart failure, strongly suggested a rheumatic recurrence. All children were treated conventionally with aspirin and/or steroids and added antifailure therapy if indicated. Secondary prophylaxis with oral or intramuscular penicillin was given to all children and they were followed up at

the Paediatric Clinic for a variable period of up to 4 years.

RESULTS

A total of 42 children with ARF were hospitalised during the period of 4 years. This constituted about 0.002% of total paediatric admissions. Twenty were males and 22 females. The age of presentation varied from 5 years 9 months to 11 years 11 months with 31 (73.5%) children in the age group of 8 to 12 years. All the children were Malays. 69.4% were admitted between November and April with a seasonal low between May and August, (see Table I). The majority of the children admitted were from Kota Bharu district. History of preceding sore throat was elicited only in 3 (7.1%) children. Past history of rheumatic fever or RHD prior to admission to HUSM was present in 11 children. In this latter group there was no data available regarding regular prophylaxis prior to hospitalisation. There was insufficient information regarding the living conditions and socio-economic status of the children concerned.

Table I
Monthly Admission of Kelantanese Children with Acute Rheumatic Fever (n = 42)

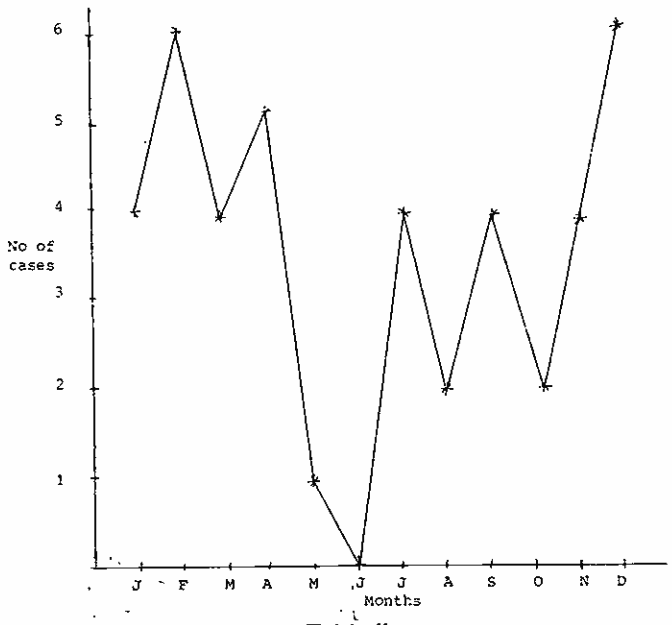


Table II
Presenting Symptoms in Kelantanese Children with Acute Rheumatic Fever (n = 42)

Symptoms	No. of children (%)
Fever	39 (92.9)
Joint pain	28 (66.7)
Joint swelling	24 (57.1)
Breathlessness	12 (28.6)
Palpitation	9 (21.4)
Cough	8 (19)
Precordial pain	7 (16.7)
Abdominal pain	6 (14.3)
Skin rash	5*(11.9)

* 2 suggestive of erythema marginatum

The main presenting symptoms are shown in Table II. Non-migratory polyarthritis was present in 4 children; in the rest, a history of migratory polyarthritis was obtained. Small joints such as metatarsophalangeal and proximal interphalangeal joints were involved in 2 children. The commonest joints affected were knees and ankles. Abnormalities in the 28 children with carditis are shown in Table III. Congestive cardiac failure was a common complication as seen in 20 out of 28 children with carditis. Pericarditis was not clinically obvious in most of these children although echocardiography demonstrated pericardial effusion in 7. The clinical profiles of these children when compared to other countries are listed in Table IV. The results of relevant investigations are listed in Table V. Table VI shows the echocardiographic findings in 35 children who underwent

Table III
The Cardiac Abnormalities in Kelantanese Children with Carditis (n = 28)

Abnormality	No. of children (%)
Congestive heart failure	20 (71.4)
Mitral incompetence	25 (89.3)
Mitral stenosis	8 (28.6)
Aortic incompetence	5 (17.9)
Aortic stenosis	2 (7.1)
Pericardial effusion (clinical)	2 (7.1)

Table IV
Comparison of Clinical Profiles of Acute Rheumatic Fever (%)

Major Manifestation	Kelantan 1985-89 %	Thailand (3) 1987 %	Philippines (2) 1974 %	Indonesia (2) 1969-72 %	India (6) 1980 %	Pakistan * (7) 1982-85 %
Carditis	66.6	90	90	57	33.7	61
Arthritis	57.1	24	66	39.8	66.6	56.5
Chorea	7.1	8	0.9	3.5	20.5	19.5
Subcutaneous nodules	4	8	5	0.1	1.9	3.4
Erythema marginatum	4	5	0.5	1.7	1.9	–

* Figures for Pakistan: Average calculated on the initial episode and recurrence

Table V
Results of Relevant Investigations in Acute Rheumatic Fever in Kelantanese Children (n = 42)

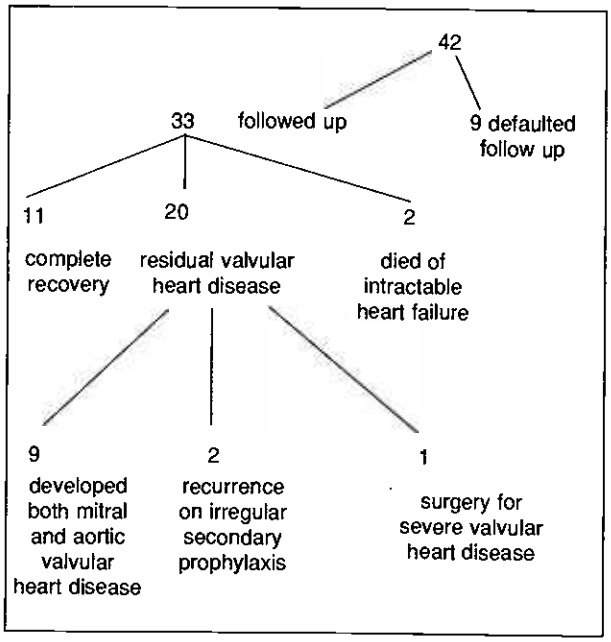
Investigation	Positive in No. of patients	%	No. with no data
ASOT > 400 Todd units	26	70	5
Throat culture positive for group A beta haemolytic streptococci	2	5.7	7
ESR > 20mm/hr	39	95.1	1
Abnormal ECG			
First degree heart block	13	31	
Second degree Wenkebach's	1	2.4	0
Others	11	26.2	
Abnormal Chest X-ray			
Cardiomegaly	16	44.4	6
Pulmonary congestion	9	2.5	
Abnormal echocardiography	24	68.6	7

Table VI
Echocardiographic Findings in Kelantanese Children with Acute Rheumatic Fever (n = 35)

Echocardiographic Findings	No.	(%)
Poor coaptation of mitral valves	10	28.6
Left ventricular dilatation	10	28.6
Thickened mitral valve cusps	7	20
Pericardial effusion	7	20
Mitral regurgitation*+	2	5.7
Mitral stenosis*	2	5.7
Normal echocardiography	11	31.4

* Based on M-mode findings
+ Doppler studies not performed

Table VII
Outcome in Kelantanese Children with Acute Rheumatic Fever (n = 42)



the investigation. Secondary prophylaxis was given to 39 children. The outcome in 42 children are shown in Table VII.

DISCUSSION

In developed countries, ARF was often described as a 'vanishing disease' until early eighties [5,8-11]. However, in the developing countries, the disease continues to be the commonest cause of acquired heart disease in children resulting in fatalities or in progressive crippling valvular heart disease [1]. It is difficult to comment on the probable incidence of ARF in Kelantan, since milder cases of ARF and RHD from the periphery tend to be treated in district hospitals rather than referred to a tertiary centre. However, Kelantan is a rural state bordering South Thailand and the incidence of ARF is probably comparable to that in Southern Thailand where the prevalence rate of RHD is about 1.9-2.7 per 1000 school children [2]. The 0.002% frequency of total paediatric admissions of rheumatic fever in our hospital is low when compared to a figure of 22-50% reported by several states in India [12-14]. Comments could not also be made on the seasonal variation and racial distribution of ARF because of small number of cases (forty two) and hospital based cohort.

It could be possible that in Kelantan as in other developing countries,families tend to pay attention to those children with severe illness and minor ailments tend to be ignored [1,7,15,16]. Hence children with carditis tend to be hospitalised and not those with arthritis or erythema marginatum. The incidence of carditis in our study was comparable to that from Indonesia and Pakistan [2,7]. Involvement of small joints in addition to the larger ones has also been observed in the Indian subcontinent [17]. Joint involvement in rheumatic arthritis is typically migratory; however, 4 children had non-migratory polyarthritis when it occurs alone, non migratory polyarthritis is likely to be post-streptococcal reactive arthritis [18]. The value of polyarthralgia or migratory polyarthralgia in the diagnosis of ARF is uncertain. In one study, a 5 year follow up of 81 patients with polyarthralgia (with no other manifestation of rheumatic fever or RHD) revealed the development of mitral stenosis in 2 cases [19]. However, inclusion of polyarthralgia would lead to over diagnosis and is not favoured [20]. As in other developing countries chorea, erythema marginatum and subcutaneous nodules were not common.

There are no pathognomonic tests for confirming the diagnosis of ARF and diagnosis in these children was based on clinical criteria. However, we have found elevated erythrocyte sedimentation rate (ESR) and an antistreptolysin O titre (ASOT) of ≥ 400 units useful. The unreliability and low specificity of ESR is well known. ASOT, on the other hand, can be elevated in up to 20% of normal school age population [20]. C-reactive protein estimation has a high sensitivity but we were unable to do it on all the children. Positive throat swabs found in only 2 children were possibly because of (a) administration of antibiotics prior to throat culture (b) long patent period between antecedent streptococcal infection and the development of symptoms of rheumatic fever and (c) delay in diagnosis of rheumatic fever and thus in obtaining throat cultures [21]. Combined use of echocardiography and Doppler ultrasonography have high sensitivity and specificity in the assessment of RHD,thus providing information of valvular and functional abnormalities [21]. Poor coaptation of mitral valves and left ventricular dilatation found in 10 children each could be due to acute carditis and congestive cardiac failure. We have found chest X-ray and electrocardiography less useful investigations on ARF.

The problems of high default rate, poor compliance and irregular medications are not peculiar to Kelantan, but are common to most developing countries, and are linked to poor economic status, superstitious belief regarding the cause of illness, poor transport facilities, poor medical facilities and lack of health education.

In conclusion, the clinical profile of ARF in Kelantan is similar to those seen in the other developing countries. Universiti Sains Malaysia is currently undertaking a prospective study to map out the epidemiological pattern of ARF and RHD in Kelantanese school going children with a view for prevention.

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