

Rheumatic Chorea

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Rheumatic chorea is a major manifestation of acute rheumatic fever, characterized by involuntary movements, in coordination of voluntary movements, and emotional disturbances. The condition is usually seen in children, and infrequently after adolescence. Polyarthritides and chorea do not generally occur simultaneously. We report the clinical features of rheumatic chorea, its relationship to other manifestations of rheumatic fever, course of the disease and outcome.

Material and Methods

Patients with chorea, up to 14 years of age, admitted to the Pediatric Unit of General Hospital, Tata Tea Limited, Munnar during 1986-1992 were studied. Rheumatic chorea was diagnosed by clinical features and laboratory investigations after exclusion of other possible causes, including systemic lupus erythematosus, Wilson's disease, Huntington's chorea, and drug induced chorea. Investigations included complete blood count, antistreptolysin-O titre, C-reactive protein, and throat swab culture for the evidence of Group A streptococcal infection. X-ray film of the chest and electrocardiogram were taken whenever there was associated heart disease. The diagnosis of 'carditis' was based on physical findings and auscultation. There was no history of

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intake prior to admission. The treatment consisted of rest in bed, supervised feeding and drug therapy. All patients were treated with haloperidol, the dose varying from 0.5-1.5 mg in 2-3 divided doses for 4-8 weeks. Improvement was judged clinically and recovery was defined as complete absence of chorea as judged by parents, teacher and the treating physician.

Results

A total of 12 patients were studied. There were 6 girls and 6 boys. The age ranged from 7 to 13 years (mean 9.4 years). Eight patients (66.7%) had generalized chorea and 4 (33.3%) had hemichorea. *Table I* summarizes the clinical features. Ten patients (83.3%) had chorea for the first time, while 2 (16.7%) were admitted with relapse, the first attack occurring 2 years earlier. The patients with recurrence of chorea showed features of mitral regurgitation on auscultation, which were confirmed

TABLE I—Summary of Clinical Features

Clinical features	Number (n=12)	%
Symmetric chorea	8	67
Hemichorea	4	33
Gross hypotonia	2	17
Slurred speech	10	83
Aphonia (mute)	2	17
Relapse chorea	2	17
Arthralgia	5	42
Migratory polyarthritides	3	25
Simultaneous chorea and polyarthritides	2	17
Carditis	7	58
Mitral regurgitation	2	17
Clumsiness and change of behavior	6	50
Emotional lability	1	8

by echocardiography: Simultaneous presence of polyarthritis and chorea were seen in 2 patients. Clinical features of carditis were seen in both these cases. One patient showed emotional lability. Erythrocyte sedimentation rate was raised in 8 (66.7%) patients. ESR was normal when chorea was an isolated finding. Antistreptolysin-O titre and C-reactive protein were positive in 16.7% and 33.3% patients. Clinical features of carditis and polyarthritis were seen in these cases. Eleven patients (91.7%) responded satisfactorily to haloperidol. These patients showed a marked reduction in choreiform movements within a mean period of 5.3 days (range 3 to 8 days) of commencement of therapy. None of the patients showed side effects attributable to drug therapy.

Discussion

Rheumatic chorea is self limiting in most of the patients and attacks usually subside in 2 to 6 months (1,2). Although the incidence of rheumatic fever has declined during this century, recent outbreaks may account for regional increases in the incidence of chorea which has been seen in up to 40% of recent cases of rheumatic fever (3,4). Although girls are affected more often than boys, the incidence was equal in both sexes in our series. The onset was insidious in two-thirds of cases. Similar findings have been reported earlier (5,6). As the clinical diagnosis was suggestive of rheumatic chorea in all patients, slit lamp examination for the presence of Kayser-Fleisher ring to rule out Wilson's disease was not carried out.

Choreiform movements were marked on one side of the body in 4 (33.3%) cases. Whether a pure hemichorea of rheumatic origin ever occurs is doubtful, as close observation will usually reveal some choreiform movements on the other side as well (5). Speech was slurred and hesitating in most of the children (83.3%). Sanyal *et al.* in their

study have noted speech changes occurring in only 30% of their patients. Paralytic type of chorea described as the most severe variety accompanied by aphonia lasting for several days was observed in 2 (16.7%) cases. There was a significant association between chorea and carditis. A similar frequent association between chorea and rheumatic heart disease has been shown by Joshi *et al.* (7). Polyarthritis and chorea do not occur simultaneously (1,5). In our series also, polyarthritis and chorea occurred at the same time in only 2 (16.7%) patients. Although emotional disturbances presented as clumsiness and behavioral changes in 50% of patients, severe emotional lability was observed in only one child.

Erythrocyte sedimentation rate was significantly raised in chorea associated with carditis. Antistreptolysin-O titre (immunological evidence of recent streptococcal infection) and C-reactive protein were absent when chorea was an isolated finding. A recently identified B-cell alloantigen, D 8/17 is expressed more frequently in patients with rheumatic fever than in control subjects (8). Feldman *et al.* have postulated that the presence of this antigen in elevated levels might be used to differentiate patients with rheumatic chorea from those with systemic lupus erythematosus (9).

Several drugs are used in chorea with a variable degree of efficacy; haloperidol, valproic acid, tetrabenazine, chlorpromazine, barbiturates and corticosteroids. In our study, most of the patients showed signs of improvement in 3-8 days with haloperidol. The effectiveness of haloperidol in rheumatic chorea has been well documented (10). Relapse occurred in one patient, 2 months after stopping haloperidol. The drug was reinstated for a further 8 weeks with excellent results. A similar relapse after valproate therapy has also been documented (11).

None of the patients showed side effects to prolonged drug therapy. Associated arthritis and carditis were treated with aspirin and

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prednisolone. On admission, all the patients received intramuscular benzathine penicillin (weight <30 kg = 6,00,000 units, weight >30 kg = 12,00,000 units). Benzathine penicillin prophylaxis was continued at intervals of 3 weeks after discharge. A follow up of these patients at cardiac clinic is being carried out at regular intervals.

Finally, rheumatic chorea being a disease encountered in school children, close cooperation with the school medical service and educational authorities is essential in the management.

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