

## **SYDENHAM'S CHOREA**

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**Objective:** To study the clinical profile in patients with Sydenham's chorea. **Design:** Prospective. **Setting:** Medical college and hospital. **Subjects:** Sixty cases with Sydenham's chorea, between 1988 and 1994, were studied. Of these 36 were girls; the mean age at presentation was 11.1 yr (range 7-16 yr). **Results:** Female predominance was apparent only after 9 years of age. There was a high familial incidence for both chorea and rheumatic fever. Generalized chorea was seen in 40 and hemichorea in 20 patients. The chorea lasted from 5 to 40 days; 13 patients had recurrent episodes. Gait disturbances, dysarthria and weakness were common. Six patients had co-existing heart disease, 4 had arthritis and one had subcutaneous nodules. A raised ESR and positive ASO titer were seen in 37 and 20 patients respectively. **Conclusions:** There was an increased incidence of Sydenham's chorea after 9 years of age in girls possibly suggesting the influence of female sex hormones. A high incidence of neurologic manifestations was noted. Acute phase reactants were raised in almost half the patients. Sodium valproate, whenever used, was effective in controlling the chorea.

**Key words:** Rheumatic chorea, Sodium valproate.

**S**YDENHAM'S chorea (SC) is a major manifestation of rheumatic fever and with the 1992 modification of the Jone's criteria(1), is sufficient alone to make the diagnosis of acute rheumatic fever. SC is manifested predominantly by involuntary movements and infrequently by other neurologic symptoms(2,3).

Large series of patients with SC were described previously(2,4), but there are no recent reports from India. As the clinical spectrum seems to be changing(2), we conducted a prospective study to evaluate the clinical features of this condition.

### **Subjects and Methods**

Sixty patients with SC admitted to the C.G. Hospital, Davangere, during 1988 to 1994 were included. The diagnosis of SC was made on presence of semi-purposeful choreiform movements which disappeared during sleep, impaired coordination and

muscular weakness. Other causes of choreiform movements like Wilson's disease, systemic lupus erythematosus, hypocalcemia, encephalitis, cerebrovascular accidents, degenerative disorders, collagen vascular disease, oral contraceptives use, familial choreas, familial calcification of basal ganglia, hyperthyroidism, and hypoparathyroidism were excluded by detailed clinical evaluation, history and relevant biochemical investigations.

Investigations like ESR, ASO titer, chest radiographs and throat cultures were done in all patients. Electrocardiograms and echocardiograms were done in all patients and electroencephalogram (EEG) in 12 patients. Slit lamp examination was done to look for the presence of Kayser-Fleischer ring. Rheumatoid factor and LE cell phenomena were done in all cases.

The treatment for SC included phenobarbitone 3 mg/kg/day, chlorpromazine 2 mg/kg/day, diazepam 0.2 mg/kg/day, haloperidol 0.05 mg/kg/day and sodium valproate 20 mg/kg/day used either singly or in combination. All drugs were given orally. The effect of the treatment was assessed by cessation of abnormal movements.

All cases were treated with intramuscular benzathine penicillin once every 3 weeks. During the follow up, detailed clinical examination was done; further investigations were done when there were clinical findings of rheumatic activity or cardiac involvement.

### Results

Of 60 patients, 36 were girls; the mean age was 11.1 (range 7-18 yr). The onset of choreiform movements was gradual, over 10-14 days in 58 and abrupt, over 1-2 days in 2 patients. Chorea was generalized in 40 cases (66.7%), and involved the right and left side in 10 cases each (16.7% each). The duration of the illness ranged from 5 to 40 days.

Thirteen cases (21.6%) had repeat attacks; 8 had only one recurrence, 3 had two recurrences and one each had three and five recurrences. The period between recurrence ranged from 6 months to 4 yr (mean 1.7 yr). In 10 cases when the initial attack was hemichoreic, the subsequent attack was also hemichoreic and involved the same side. Three patients had variation in the recurring attack. One had generalized chorea at first episode and the second attack was left hemichoreic. Two patients had left hemichorea at first attack and subsequently had right hemichorea.

Family history was present in 5% patients. This included 2 mothers and 1 father. A family history of rheumatic fever was obtained in 4 patients (6.7%). Previous history of sore throat was present in 11 patients (18.3%). Neurological symptoms included gait disturbances in 22 cases (36.7%), speech impairment in 17 cases

(28.3%), weakness in 17 cases (28.3%), reflex changes in 13 cases (21.7%), headache in 8 cases (13.3%) and cranial neuropathy in one case. Cardiac involvement was seen in 6 cases. Five had valvular involvement; mitral insufficiency in 2 patients and aortic insufficiency in one, combined mitral insufficiency and aortic insufficiency in one and combined mitral insufficiency and stenosis in one. One patient had carditis. Arthritis was seen in four patients and subcutaneous nodules in one. Raised ESR was seen in 37 patients (62.0%), positive ASO titer in 20 (33%) and all throat cultures were negative. EEG was done in 12 patients and all were within normal limits.

The mean duration in which response was noted varied with different drug regimens. It was 17.9 days with chlorpromazine (9 cases), 21.8 days with chlorpromazine and phenobarbitone (13 cases), 13.7 days with phenobarbitone (10 cases), 15 days with diazepam (3 cases), 12.6 days with haloperidol (17 cases) and 9.7 days with sodium valproate (8 cases). No residual neurologic deficits were present at the time of discharge. No deaths were reported. Twenty patients were followed for a period of 1-5 yr; 13 of these had recurrence of chorea. The remaining 7 patients came for regular penicillin prophylaxis.

### Discussion

Most cases of SC are in the age range of 5 to 15 yr(5,6). The youngest in our study was 7 yr old. The mean age of 11.1 yr in this study is slightly higher as compared to previous reports(5,7). The disease is rare in infancy and infrequent in young adults. Female predominance is noted in all large series of SC occurring at a ratio of approximately 2:1. In our study, this sex disproportion was evident only after 9 yr of age, as observed by others(2,5) and it is suggested that sex hormones may play a role for the sex difference(2).

In previous studies a high familial incidence has been noted, but the mode of inheritance is unknown(8). Earlier reported studies found chorea in 3.5% of parents and 2.1% siblings of choreic patients and a family history of rheumatic fever in 26%(5). In our study, the familial incidence of SC and rheumatic fever was 5% and 6.7% respectively.

Most patients in our study had only chorea as the neurological symptom. Speech disturbances occurred in 28.3%. This is thought to be extrapyramidal in origin due to dyskinesias of the muscles of articulation rather than disordered pyramidal or cerebellar function. Gait disturbances including ataxia are known in SC and were seen in 36.7% of our cases. The clinical features of SC observed in this study are similar to those reported earlier(2).

Recurrent attacks of chorea are not uncommon. In our series, 13 patients(21.8%) had repeat attacks. Most of them had only one recurrence, at a mean interval of 1.7 yr after the first attack. Recurrences many years after the initial attack are uncommon and suggest that late chorea may be due to reactivation by another mechanism, such as pregnancy or drugs(9). Patients with SC may have chorea during pregnancy (chorea gravidarum) and are at higher risk for rare types of chorea induced by phenytoin or oral contraceptives(9,10).

Without documentation of an antecedent streptococcal infection, the diagnosis of Sydenham's chorea must be made by excluding other causes of chorea in childhood. A relationship to previous infection with group A streptococci has been assumed, but often difficult to demonstrate. Raised ESR and ASO titre are unusual in SC(11). We found a raised ESR in 62% and a positive ASO titer in 33% at the time of first episode. Whether these values could predict later development of heart disease is unclear.

Recent reports indicate that sodium valproate is an effective drug in the management of abnormal movements in SC(12,13). Sodium valproate is known to raise the level of "gamma aminobutyric acid (GABA), particularly in the striatum and substantia nigra(14).

This increase may exert its effect through modification of the GABA-nergic synaptic transmission and hence control the abnormal movements. This effect on the basal ganglia is totally different from the anticonvulsant effect of the drug(13). Eight of our cases treated with sodium valproate recovered faster when compared to those treated with other drugs. This observation should initiate randomised controlled trials to evaluate the superiority of sodium valproate over other drugs.

Newer modes of treatment, aimed at clearing antineuronal antibodies, including plasma exchange and intravenous immunoglobulin therapy(15) are still investigational. The role of corticosteroids is also being studied(15).

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## NOTES AND NEWS

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### PEDIATRIC NEPHROLOGY UPDATE

The Department of Pediatrics, Sir Ganga Ram Hospital and IAP Delhi Branch are organizing an Update in Pediatric Nephrology on 10th march 1996, at Sir Ganga Ram Hospital, New Delhi. The registration fee for the Update is Rs. 100/- (inclusive of lunch and tea). For further details please contact Dr. P.K. Pruthi, Organizing Secretary, Department of Pediatrics, Sir Ganga Ram Hospital, New Delhi 110 060.