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SYDENHAM'S CHOREA (SC) LATE PRESENTATION OF ACUTE RHEUMATIC FEVER (RF)

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ABSTRACT

OBJECTIVE: *To report the clinical profile of sydenham's Chorea and its relationship to other manifestation of rheumatic fever.*
DESIGN: *Cross Sectional*
SETTING: *Department of Paediatrics and Neurology Chandka Medical College Larkana, Sindh Pakistan.*
SUBJECT: *Twenty four cases with sydenham's chorea between 2005 to 2007 were studied of these 12 were girls mean age of presentation was mean 9.4 years.*
RESULTS: *Sydenham.s Chorea seen equally in the both sexes, generalized chorea was seen in 16 (66.6%) and hemi chorea in 8 (33.3%) patients 20 (83.3%) patients had chorea for the first time while 4 (16.6%) with relapse. 4 patients had Co-existing heart disease and arthritis. None of patients showed subcutaneous nodule and erythema marginatum. A raised Erythrocyte Sedimentation Rate (ESR) and positive ASO titer were seen in 16 and 8 patients respectively, all the patients were treated with haloperidol.*
CONCLUSION: *A high incidence of carditis and arthritis observed with Sydenham's Chorea. Acute phase treatment were raised in almost half the patients, haloperidol is effective in controlling of chorea.*

KEY WORDS: Rehumatic Chorea, Clinical profile, Haloperidol.

INTRODUCTION:
Rehumatic fever (R.F) is amongst the most significant health problem faced by third world countries. It is a multi systemic disorder typically following an episode of streptococcal phyringitis [1, 2]. As saying goes RF. Bites the heart and licks the joints in children.

HISTORICAL BACKGROUND:
Individual manifestation were first described in 17th and 18th centuries. Arthritis by de Baillous, chorea by sydenhams and vavular disease by lancisi and morgagni, later observers, notably chreadle in 1889 brought together the various manifestations of the reheumatic fever syndrome much as we know it today[3,4]. Over all annual incidence is 100/100000. **Sydenham's Chorea (SC)** is major late manifestation of rheumatic fever and with the 1992 modifications of the jone's criteria, is sufficient alone to make, the diagnosis of acute rheumatic fever. SC is manifested predominantly by involuntary movements and infrequently by other neurologic symptoms. SC may occur by it self or following poly arthritis it is a self limiting condition and heals without residual.
Treatment of SC when isolated neither salicylates nor steroids should be used sedative may be helpful, like, phenobarbitone, sodium valproica and chlorpromazine or haloperidol may be tried. Patient with SC ever in he absence of other manifestations requires prolong anti-sterptococal prophylaxis.
Objective of the present study was to report the clinical features of Syderhaum's Chorea and its relationship to other manifestation of rheumatic fever.

SUBJECTS & METHODS.
Patients with SC up to 18 years of age admitted in paediatrics and neurology unit of CMC Larkana during May 2005 to May 2007 were studied. SC was diagnosed by clinical features and laboratory investigations after the exclusion of other possible causes, like Wilson diseases, Huntington's Chorea and drug induced Chorea investigations included complete blood count, antistreptolysinotitre (ASOT), X-Ray of Chest, electrocardiogram

(ECG) and echo-cardiography were taken when ever there was associated heart disease. The treatment consists of bed rest and drug therapy.

All the patients were treated with halopriedol with the Dose of 0.5mg to 1.5mg/day 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 and treating paediatrician and neurologist.

RESULTS:

A total of 24 patients were studied, there were 12 girls and 12 were boys. The age range from 5 – 18 years (Mean 9.4 years). Sixteen patients (66.6%) had generalized Chorea and 8 (33.3%) had hemi Chorea Table 1. Summarized the clinical features. Twenty patients (83.3%) had Chorea for the first time, while 4 (16.6%) were admitted with relapse, the first attack occurred 2 years earlier.

The patient with recurrence of chorea showed features of mitral regurgitation on auscultation, which were confirmed by echocardiography, simultaneous presence of polyarthritis and chorea were seen in 4 patients (16.4%) clinical features of carditis were seen in these cases, two patients (8.2%) showed emotional liability.

Erythrocyte sedimentation rate (ESR) was raised in 16 (66.6%) table 2 Summarized the investigations, ESR was normal when chorea was isolated finding.

Antistreptolysinotiter was positive in 8 cases (33.3), X-Ray chest showed cardiomegally in 4 cases (16.6) and Echocardiograply showed mitral regurgitation in 4 case (16.6%).

Twenty three patients (95.8%) responded satisfactory to haloperidol these patients showed a marked reduction in choreiforam movements with in 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.

TABLE 1
SUMMARY OF CLINICAL FEATURES:

S #	Clinical Features	Number (n=24)	%
1	Symmetric Chorea	16	66.6
2	Hemi Chorea	8	33.3
3	Gross Hypotomia	4	16.6
4	Slurred Speech	20	83.3
5	Relapse Chorea	4	16.6
6	Arthralgia	10	41.6
7	Migratory Polyarthritis	6	25.1
8	Simultaneous Chorea & Polyarthritis	4	16.6
9	Corditis	7	29
10	Mitral regurgitation	4	16.6
11	Clumsiness & Change of Behavior	12	50
12	Emotional Liability	2	8.3

TABLE 2
SUMMARY OF INVESTIGATION:

S #	Investigation	Number (n=24)	%
1	ESR raised	16	66.6
2	ASO Titer increased	8	33.3
3	X-Ray Chest showed Cardiomegaly	4	16.6
4	Echocardiography showed Mitral regurgitation	4	16.6

DISCUSSION:

Majority (70%) of our SC patients were usually age range of 5 to 18 years, the mean age of onset of Chorea was 9.4 years. SC is self limiting in most of the patients and attacks usually subsided in 2 to 6 months. [1,2]. Although girls are affected more often than boys, the incidence was equal in both sexes in our series the onset was insidious in two third of cases. Similar findings have been reported earlier [5-6]. As the clinical diagnosis was suggestive of SC in all patients, out even then slit lamp examination was carried out for the presence of Kayser Fleisher (KF) ring to rule out Wilson disease, none of the SC patient found suffering rigosture choreiform movements were marked on one side of the body in 8 (33.3%) cases. Whether a pure hemi chorea of rheumatic origin ever occurs in doubtful, as close observations will usually reveal some courier form movements on the other side as well [5].

Speech was slurred and hesitating in the most of children (83.3%) Sanyaletal, in their studies speech changes found 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 4 (16.6%) cases. There was a significant association between chorea and corditis. A similar frequent association between chorea and rheumatic heart disease has been showed by joshietal. Poly arthritis and chorea do not occur simultaneously but in our series also poly arthritis and chorea occurred at the same time in 4 (16.6%) patient. Although emotional disturbances presented as clumssness and behavioral changes found in 50% of patients. Erythrocyte sedimentation rate was significantly raised in Chorea associated with cordites. ASOTiter was within normal limit when chorea was an isolated finding. Several drugs are used in chorea with a variable degree of efficacy.

Haloperidol valproic chlorpromazine, barbiturates and corficosteroids in our study most of the patient showed signs of improvement in 3-8 days with haloperidol. The effectiveness of haloperidol in rheumatic chorea has been well documented [10]. non of the patients showed side effects to prolonged drug therapy. SC associated arthritis and carditis were treated with asprin and prednisolone on the admission and all the patients received intramuscular benzathene panicillin (weight <27 kg 600000 units, weith >27 kg 12,00,000 units) Benzathine penicillin prophylaxis was continued at intervals of 3 weeks of the discharge. A follow up of these patients at Paediatric and Neurological outdoor patients department (OPD) was carried out at regular intervals.

CONCLUSION:

Finally Sydenham's chorea is late and sole manifestation of acute rheumatic fever, encountered in school children, close co-operation with the school medical service and educated authorities is essential in the management.

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