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# A case report on rheumatic chorea – neurological disorder

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#### **ABSTRACT:**

Acute Rheumatic Fever is generally seen in academy-aged children and presents with migrant polyarthritis, Sydenham chorea, carditis, erythema marginatum, and subcutaneous nodes in various combinations. Sydenham's chorea mainly involves jerky, uncontrollable, and purposeless movements of the hands, arms, shoulder, face, legs, and trunk. These movements look like twitches and disappear during sleep. The diagnosis of SC is clinical and can be confirmed by laboratory tests: an elevation of erythrocyte sedimentation rate, or the antistreptolysin-O and anti-DNAse B titers. Electroencephalography and neuroimaging are often normal, but they can be useful for differential diagnosis, especially in patients with unusual features. Penicillin is extensively used as an antistreptococcal antibiotic both as acute treatment and as long-term secondary prophylaxis.

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## **INTRODUCTION:**

Acute Rheumatic Fever (ARF) is a multisystem seditious complaint that occurs as a delayed autoimmune response to group A beta-hemolytic streptococcal sore throat infection. ARF is generally seen in academy-aged children and presents with migrant polyarthritis, Sydenham chorea (SC), carditis, erythema marginatum, and subcutaneous nodes in various combinations<sup>[1]</sup>. Sydenham chorea (SC), or rheumatic chorea, is one of the major clinical instantiations of acute rheumatic fever (ARF) and is the most common form of acquired chorea in nonage. It's a movement complaint characterized by chorea (involuntary detail, arbitrary, and irregular movements of the branches and face), emotional lability,

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#### J Pharm Adv Res, 2023; 6(11): 1961-1963.

and hypotonia <sup>[2]</sup>. Pathophysiology of SC is easily related group A streptococcal (GAS) infection, to its pathogenesis isn't fully understood. Molecular belittlement, in which antibodies directed against part of the GAS bacterium cross react with host antigens in susceptible subjects, is allowed to play an important part<sup>[3]</sup>. Sydenham's chorea mainly involves jerky, uncontrollable, and purposeless movements of the hands, arms, shoulder, face, legs, and trunk. These movements look like twitches and disappear during sleep. Other symptoms may include changes in handwriting, loss of fine motor control, especially of the fingers and hands, and loss of emotional control, with bouts of inappropriate crying or laughing.

Symptoms of RF may be present. These may include high fever, heart problems, joint pain or swelling, skin lumps or skin rashes, and Nosebleeds<sup>[4]</sup>.

The diagnosis of SC is clinical and can be confirmed by laboratory tests: an elevation of erythrocyte sedimentation rate, or the antistreptolysin-O (ASLO) and anti-DNAse B titers. Electroencephalography (EEG) and neuroimaging are often normal <sup>[3,6]</sup>, but they can be useful for differential diagnosis, especially in patients with unusual features (e.g., hemi-chorea) <sup>[5]</sup>.

In clinical practice, the operation of SC is grounded on three main keystones <sup>[6]</sup>: antibiotics, characteristic medicines, and immunomodulatory treatment. Penicillin is extensively used as an anti-streptococcal antibiotic both as acute treatment and as long-term secondary prophylaxis. Characteristic treatments have long been used to control the motor and NP instantiations of SC dopamine antagonists, they're either similar to haloperidol and pimozide, or antiseizure specifics (ASMs), like valproic acid (VPA) and carbamazepine (CBZ). Immunomodulatory treatments with corticosteroids (CS), intravenous immunoglobulins (IVIG), or plasmapheresis have surfaced in more recent times <sup>[7]</sup>. Although supported by the autoimmune nature of this complaint and despite being extensively used in clinical practice, their use as a first-line remedy in SC cases is still controversial<sup>[8,9]</sup>.

# **CASE PRESENTATION:**

An 11-year-old female child was brought by her father to the outpatient department (OPD) of a tertiary care hospital, in virudhunagar, with a two weeks history of numbness of right hand on & off. The involuntary movements first began in the lower jaw and lower limbs which recovered on their own and finally, both lower limbs were involved. Her mother reported that she has a history of inability to walk before 1 ½ months. She had a history of fever one month back, which was treated on an outpatient basis.

The general physical examination shows that the child was alert, active, with mild pallor, and oriented to place and time. Her vitals were taken which showed a heart rate of 96 beats/min, respiratory rate of 24 cycles/min, afebrile, and blood pressure of 110/70 mmHg. She was mildly anemic. Pertinent examination findings included involuntary movements of upper limbs, unclear speech, and difficulty in walking. There were irregular contractions and relaxation of muscles showing pronation of forearm on outstretched hand above her head called "Milkmaid's sign."

Her laboratory investigations showed hemoglobin (Hb) levels: 10.5 mg/dL (normal range: 12.0 to 15.5), hematocrit (HCT) level: 34.8 % (normal range: 37 to 48 %), total count (TC): 8600 cells/mm (normal range: 50,000-10,000 cells /mm), erythrocyte sedimentation rate (ESR): 55 mm 1st hour (normal: <20), antistreptolysin O (ASO titer): 400 (normal: 166 Todd units), electrocardiogram (ECG) and echocardiogram were with normal sinus rhythm thus; rheumatic carditis was ruled out.

Other investigations include;

> Peripheral Smear Report: The report suggests that the child is in Mild hypochromic microcytic anemia

Echocardiographic Report: LVID (d): 5.1, LVID(s):
4, LVEF (%): 60

Impression: Rheumatic heart disease with moderate mitral regurgitation.

### **TREATMENT:**

The patient was initially managed with oral Penicillin 250 mg BD and IV cefotaxime 700 mg. After 24 h of hospital admission, she was started with Tablet Haloperidol 1.5 mg  $\frac{1}{4}$  BD as per the neurologist and psychiatrist's opinion. The child also receives Tablet FST and Tablet BCT from day 2 of admission. On day 3 and day 4, the frequency of Tablet Penicillin is modified to QID. On the fifth day of admission, Joint pain was observed which is treated with Tablet Paracetamol 500 mg  $\frac{1}{2}$  BD. Injection of Benz penicillin 1.2 mil IU every three weeks (till the age of 21) was also advised.

#### **DISCUSSION:**

The most prevalent type of autoimmune chorea is Sydenham's chorea (SC), which is one of the main

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Drug	Dose (mg)	Route	Frequency	Day 1	Day 2	Day 3	Day 4	Day 5
Tablet Penicillin	250	Oral	BD	G	G	G		
Tablet Haloperidol	1.5	Oral	1⁄4 BD		G	G	G	G
Tablet FST	333.5	Oral	OD			G	G	G
Tablet BCT	30.5	Oral	OD			G	G	G
Tablet Penicillin	250	Oral	QID				G	G

indicators for the diagnosis of rheumatic fever. Females have greater impacts than males by SC, which often **Table 1. Drug therapy of the patient.** 

manifests between the ages of 5 and 15 years. Chorea typically appears 4 to 8 weeks following pharyngitis triggered by group A beta-hemolytic streptococcal (GABHS); this is later than the onset of other rheumatic fever symptoms such as carditis or arthritis, which typically appear 2 to 3 weeks after infection. In SC, acute and preventive penicillin therapy, symptomatic drugs, and maybe immunomodulatory therapy are currently being considered as chorea therapy. The list of recommended symptomatic treatments is lengthy and includes the use of neuroleptics such as pimozide, haloperidol, risperidone, and olanzapine as well as anticonvulsants like valproate and carbamazepine off-label<sup>[10]</sup>.

# **CONCLUSION:**

Rheumatic chorea is one of the major clinical instantiations of acute rheumatic fever (ARF) characterized by chorea (involuntary detail, arbitrary, and irregular movements of the branches and face), emotional lability, and hypotonia. This condition was treated and prevented with penicillin therapy and symptomatic drugs. In this case, the patient was treated with Tablet Penicillin and Tablet Haloperidol. The patient is advised to review every follow-up appointment. Recognition of RC and its timely management is crucial in preventing rheumatic heart disease.

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