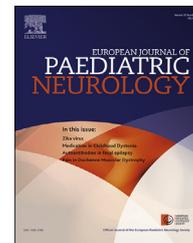




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Original article

Corticosteroid treatment in Sydenham's chorea

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A B S T R A C T

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Outcome

Sydenham's chorea (SC) is an immune-mediated hyperkinetic movement disorder, developing after group A Beta-hemolytic streptococcal (GABHS) infection. Aside from conventional symptomatic treatment (carbamazepine, valproate, neuroleptics), the use of steroids has also been advocated, mainly in severe, drug-resistant cases or if clinically disabling side effects develop with first line therapies. Based on the description of 5 cases followed in the Child Neurology Unit of Santa Maria Nuova Hospital in Reggio Emilia and on the available medical literature on this topic, we propose considering the use of corticosteroids therapy in children with SC, with the administration of IV methyl-prednisolone followed by oral deflazacort in severe cases and of oral deflazacort alone in mild and moderate degrees of involvement. In our experience this therapy is effective both in the short and long-term period, in different clinical presentations (chorea paralytica, distal chorea, hemichorea, "classic" chorea, association with mood disorder or dyspraxia) and very well tolerated (no significant side effects were recorded).

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1. Introduction

Sydenham's chorea (SC) is a hyperkinetic movement disorder characterized by clumsiness and rapid, uncoordinated and involuntary movements, secondary to group A Beta-hemolytic streptococcal (GABHS) infection and usually developing 4 to 8 weeks after an episode of GABHS pharyngitis.¹

1.1. Epidemiology

In spite of an incidence reduction, especially in high-income countries,² it remains a major manifestation in 20–40% of

cases of acute rheumatic fever,^{3,4} and it still represents the commonest form of acquired chorea.^{3,5}

1.2. Clinical findings

Chorea is defined as an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments.⁶ Generally, it involves the face and extremities, but any body district can be affected. In approximately 20% of cases SC remains confined to one body side (hemichorea).⁴ Motor impersistence is typical, especially during tongue protrusion or ocular fixation. Muscle tone is usually decreased, rarely leading to flaccid or paralytic

Abbreviations: D2R, dopamine-2 receptors; GABA, γ -amino-butyric acid; GABHS, group A Beta-hemolytic streptococcal; IV, intravenous; IVIG, intravenous immunoglobulins; SC, Sydenham's chorea; UFMG, Federal University of Minas Gerais; USCRS, Federal University of Minas Gerais Sydenham's Chorea Rating Scale.

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chorea in most severe cases (1.5% of overall cases,¹ 5% of patients with rheumatic chorea treated with neuroleptics).⁷ Oculogyric crises, dysarthria, hypometric saccades and vocal tics have also been reported. Emotional lability is frequent. In rare cases there is severe generalized hypotonia with absent or rare hyperkinetic movements, the so called chorea paralytica or chorea mollis. In severe cases, milk maids grip is appreciated as an alternating movement of the finger like a milking motion, originating spontaneously or when asked to maintain a constant, firm grip of examiner's fingers (the milker's sign).

1.3. Pathogenesis

SC is an autoimmune neuropsychiatric disorder probably caused by the presence of antineuronal antibodies rising from a cross-reaction of antibodies produced against GABHS infection with epitopes on neurons within the basal ganglia (molecular mimicry). From clinical and therapeutic reports there is a clear evidence of a dysimmune origin of SC. In particular, Dale et al. described the presence of antibodies to cell surface dopamine-2 receptors (D2R), supporting the autoimmune basis of SC, detecting the presence of D2R IgG antibody in the sera of 30% of patients with SC.⁸ D2R is an essential receptor that regulates dopaminergic neurotransmission and its involvement could explain the clinical findings of the disease and the good response to corticosteroid or immunoglobulin treatment. Moreover, the involvement of the basal ganglia was demonstrated by pathological studies, with perivenous inflammation predominantly affecting the striatum.⁹ A further recent study supports the hypothesis that in SC, antibodies-mediated D2R signaling on dopaminergic neurons could contribute to alterations of the central dopamine pathways and development of movement disorders.¹⁰

Neuroimaging is usually normal, but may show non-specific findings, whereas in severe cases white and grey matter involvement can occur.¹¹

1.4. Treatment

Considering aetiological, pathophysiological and clinical presentation, the treatment of SC has four main targets: elimination of the Streptococcus, symptomatic treatment (involuntary movements, uncoordination and psychiatric symptoms), treatment of the immune and inflammatory response, supportive measures.¹²

Primary treatment (elimination of the Streptococcus): long-term secondary prophylaxis with penicillin is mandatory and primarily directed against heart complications, although it is currently controversial whether it has any preventive effect on chorea relapses.¹³

Symptomatic treatments include dopamine receptor antagonists (haloperidol, pimozide, chlorpromazine, risperidone) or GABAergic drugs (benzodiazepines, sodium valproate, carbamazepine).

Immunological treatments: immunomodulatory therapies to shorten the course of the illness and prevent complications

are based on the use of corticosteroids, intravenous immunoglobulins and plasma exchange.¹²

It must be emphasized that, in spite of our progresses in the understanding of SC pathogenesis, treatment remains largely not evidence-based.

In this paper, we will first describe our own experience in the treatment of selected cases of SC and we will then compare our experience with the available literature on the use of corticosteroids in this condition.

2. Patients and methods

We describe patients with varying degrees of SC severity and different clinical presentations, undergoing steroid treatment with either intravenous (IV) methyl-prednisolone or oral deflazacort, who were evaluated and followed-up at the Child Neuropsychiatry Unit in Santa Maria Nuova Hospital, Reggio Emilia, Italy (Table 1).

Patients' presentation at symptoms onset, neurological examination, chorea severity, treatment, effectiveness and side effects and outcome during long-term follow-up are reported for all patients.

SC was diagnosed according to previously published modified Jones criteria in patients with acute onset of chorea (WHO).¹⁴

Treatment strategy in our unit is as follows: all patients are given prophylactic penicillin (penicillin 500 mg twice a day for 10 days together with rest, followed by secondary prophylaxis with intramuscular benzylpenicillin every 28 days or oral penicillin 250 mg twice daily), while symptomatic treatment is based on chlorpromazine 1 mg/kg/day or valproic acid 25 mg/kg/day as first-line and on steroids treatment as second line. IV methyl-prednisolone 25 mg/kg/day for 5 days followed by oral deflazacort 0.9 mg/kg/day for 3 months is reserved to severe cases,¹⁵ while mild and moderate cases undergo treatment with oral deflazacort at the dosages reported above.

In order to discuss our experience in light of the available scientific literature, we also performed a review of the literature on the use of corticosteroids in patients with SC. Only articles written in English were considered. Search terms used for this literature review comprise: Sydenham's chorea AND steroids, Sydenham's chorea AND corticosteroids. Additional relevant papers were retrieved by manually searching in the references list of each paper. Based on the accumulating evidence of an immunologic pathogenesis of SC, we only considered papers reporting on the use of steroids at immunosuppressive dosages. Articles reporting on the use of steroids at anti-inflammatory dosages were excluded.

3. Results

3.1. Chorea paralytica

At onset, this girl showed simple and complex motor tics, which were actually a fragmentary chorea. She progressively

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