

Clonidine for the management of crises in Stiff-Person syndrome

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BACKGROUND AND AIMS

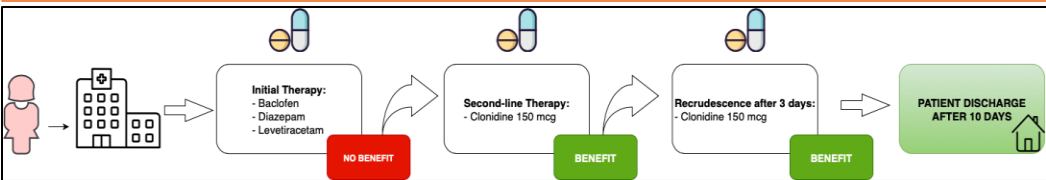
Stiff-person syndrome (SPS) is a rare autoimmune neurological disorder characterized by high titers of antibodies against glutamic acid decarboxylase (GAD) causing impaired GABAergic inhibitory neurotransmission. If not treated properly, SPS progresses to severe disability and respiratory insufficiency. To date, there are no defined guidelines for the treatment of SPS, several symptomatic drugs have been employed but a definite treatment is on demand. In this study, we report the use of clonidine in a woman affected by SPS to control disease exacerbations with immediate benefit and resolution of the acute episodes.

MATERIALS AND METHODS

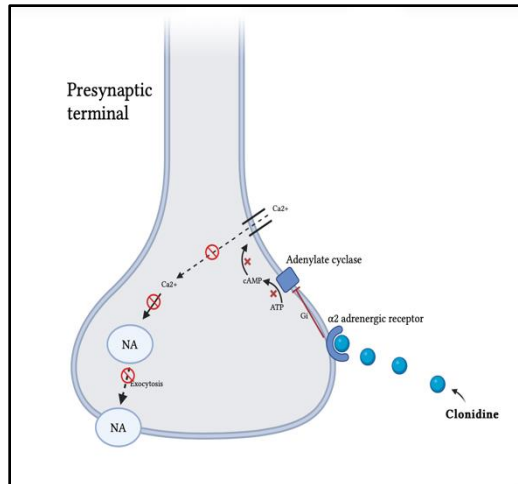
A 46-year-old woman affected by both Myasthenia Gravis (MG) and SPS came to our attention for exacerbation of SPS with a severe crisis with rigidity and spasms. In previous hospitalization to the neurology ward, the patient was treated with benzodiazepines, baclofen, antiepileptics, plasmapheresis and immunoglobulins, with unsatisfactory benefit. After several doses of Diazepam, Baclofen and Levetiracetam in association with immunoglobulins with no benefit, we administered Clonidine 150 mcg in two doses 3 days apart

RESULTS

We observed quick resolution of crises: initially the spasms disappeared after a few seconds, while the stiffness remained for a few minutes. After about 10 minutes an improvement in stiffness was evident especially to the upper limbs. After the second infusion of clonidine, with the same effects, the patient has not had any more serious relapses, but only some self-resolving recrudescences. The patient was discharged at home free of new crisis after 10 days from the last administration of Clonidine.



DISCUSSION. With this study we have reported a beneficial effect of clonidine in controlling exacerbations in Stiff-Person Syndrome, so far described in a single and pioneering case on humans in the study by Meinck et al. in 1984 (1). Based on animal studies, we know that Serotonin (5-HT) and Noradrenaline (NA) tend to increase the excitability of motor neurons and have been hypothesized to act as gain controllers for muscle activation at the spinal level (2). The mechanism of action of Clonidine justified its use as it stimulates central alpha-adreno-receptors resulting in feedback inhibition of noradrenaline release (2). Therefore, we used Clonidine in patient with SPS, resolving the crisis of rigidity and painful spasms.



(1) Meinck HM, Ricker K, Conrad B. The stiff-man syndrome: new pathophysiological aspects from an abnormal exteroceptive reflexes and the response to clomipramine, clonidine, and tizanidine. *J Neural Neurosurg Psychiatry*. 1984;47(3):280-287. doi:10.1136/jnnp.47.

(2) Johnson MD, Heckman CJ. Gain control mechanisms in spinal motoneurons. *Front Neural Circuits*. 2014;8:81. Published 2014 Jul 29. doi:10.3389/fnrc.2014.00081.