

## Stiff-person Syndrome.

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### Abstract

Stiff-person syndrome (SPS) is a progressive neurologic disorder characterized by 1) stiffness that is prominent in axial muscles, with co-contraction of agonist and antagonist muscles; 2) sudden episodic spasms; and 3) absence of another disease that causes similar symptoms. The diagnosis of SPS is based on clinical grounds and requires a high degree of suspicion. The diagnosis is, however, aided by electromyography, which demonstrates motor unit firing at rest simultaneously from the agonist and antagonist muscles, and by high serum titers of antibodies against glutamic acid decarboxylase (GAD), the rate-limiting enzyme for the synthesis of gamma-aminobutyric acid (GABA), which is the brain's main inhibitory neurotransmitter. The reduced GABA level in the brain and cerebrospinal fluid explains the patients' stiffness and justifies the clinical improvement observed by drugs enhancing GABAergic transmission. The association of SPS with other autoimmune disorders or autoantibodies, the intrathecal GAD-specific immunoglobulin G antibody synthesis, and the suppression of GABA by the patient's antibodies supports the autoimmune nature of SPS and justifies the use of immunotherapies. At present, GABA-enhancing agents, such as benzodiazepines, valproate, vigabatrin, tiagabine, gabapentin, and baclofen, provide symptomatic relief. Plasmapheresis, steroids, and periodic intravenous immunoglobulin infusions provide additional and lasting benefit. In this article, the treatment options for patients with SPS are discussed based on the authors' experience and that of others. The beneficial effects from the first controlled study conducted in SPS using intravenous immunoglobulin are presented.

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