

Stiff person syndrome: What psychiatrists need to know

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Stiff person syndrome (SPS) is a rare autoimmune condition characterized by stiffness and rigidity in the lower limb muscles. Because SPS often is misdiagnosed as a psychiatric illness and psychiatric comorbidities are common in patients with this disorder,¹ awareness and recognition of this unique condition is essential.

An insidious presentation

Patients with SPS present with:²

- axial muscle stiffness slowly progressing to proximal muscles
- unremarkable motor, sensory, and cranial nerve examinations with normal intellectual functioning
- normal muscle strength, although electromyography shows continuous motor activity
- spasms evoked by sudden movements, jarring noise, and emotional distress
- slow and cautious gait to avoid triggering spasms and falls.

Symptoms start slowly and insidiously. Axial muscle stiffness can result in spinal deformity. Involvement is asymmetrical, with a predilection for proximal lower limb and lumbar paraspinal muscles. Affected muscles reveal tight, hard, board-like rigidity. In later stages of SPS, mild atrophy and muscle weakness are likely.

Frequent misdiagnosis

Because facial muscle spasticity is prominent, SPS patients may be misdiagnosed with Parkinson's disease, primary lateral sclerosis, or multiple sclerosis. Spasms affecting respiratory and thoracic paraspinal muscles (status spasticus) may be misdiag-

nosed as an anxiety-related condition. These spasms can be life-threatening and require IV diazepam and supportive measures.

More than 60% of SPS patients have a comorbid psychiatric disorder.³ Anxiety disorders—generalized anxiety disorder, agoraphobia, and panic disorder—major depression, and alcohol abuse are the most frequent psychiatric comorbidities seen in SPS patients.³

SPS patients who panic when in public may be misdiagnosed with agoraphobia.³ Emotional stimuli may cause muscle spasms leading to falls. Treating muscle spasticity with γ -aminobutyric acid (GABA) agonists and narcotics can lead to drug abuse and dependence. Muscle spasticity can fluctuate from hour to hour, abate with sleep, and get worse with emotional distress. These findings are why approximately 70% of SPS patients are initially misdiagnosed; conversion disorder is a frequent misdiagnosis.⁴ Mood disorder in SPS patients may be resistant to antidepressants until these patients are treated with immunotherapy.⁴

Treating SPS patients

Although early intervention can reduce long-term disability, approximately 50% of SPS patients eventually have to use a wheelchair as a result of pain and immobility.⁵

Antibodies to glutamic acid decarboxylase, which is the rate-limiting enzyme for GABA synthesis, are present in 85% of SPS patients.⁵ Therefore, treatment usually includes GABA-enhancing drugs, including sedative anxiolytics (clonazepam and diazepam), antiepileptics (gabapentin,

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levetiracetam, tiagabine, and vigabatrin), antispasticity drugs (baclofen, dantrolene, and tizanidine), and immunotherapy (corticosteroids, IV immunoglobulins, and rituximab).⁵ Antidepressants, biofeedback, and relaxation training also can offer relief. Psychotherapy and substance dependency interventions may be needed.

To achieve optimum outcomes in SPS patients, a close collaborative relationship among all treating clinicians—including primary care physicians, neurologists, anesthesiologists, and psychiatrists—is necessary.

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