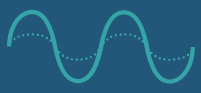
2024 STIFF PERSON SYNDROME SYMPOSIUM

Take-Away: Exploring the Fundamentals of SPS (Dr. Duarte Machado, MD)

Stiff Person Syndrome (SPS) is a rare immune-mediated neurological disorder.

Neurophysiological findings on electromyography (EMG) are supportive of a diagnosis.

SPS is due to an impairment of an inhibitory neurotransmitter called gamma-aminobutyric acid (GABA) by auto-antibodies, leading to a reduction of brain GABA.



There are several non-motor symptoms related to SPS that are also important to manage, including anxiety, constipation, and shortness of breath.

Anti-GAD antibody is the most frequently associated biomarker of SPS, though other antibodies can cause SPS including those to amphiphysin and the glycine receptor.

Treatment options for SPSD are divided into two main categories:

1. Symptomatic
2. Immunotherapeutic

The most used treatments are benzodiazepines such as diazepam, and intravenous immunoglobulin.

There are several clinical subtypes of SPS, with classic SPS being most common and resulting in episodic muscle spasms and truncal stiffness.

Additional symptomatic treatment options include baclofen, and other immunotherapies include immunosuppressants such as rituximab.