

STIFF PERSON SYNDROME (SPS): EMERGENCY INFORMATION

Patient Name: _____
Date of Birth: _____
Emergency Contact _____ Phone: _____
Treating Physician _____ Institution: _____
Physician/Clinic Contact Information: _____

1. OVERVIEW

SPS is a rare autoimmune neurological disorder causing muscle stiffness and painful muscle spasms (spontaneous or triggered by noise, light, touch, stress, temperature changes, procedures). Symptoms may also include impaired mobility, exaggerated startle, and may result in **respiratory involvement in severe cases**.

Pathophysiology involves impaired GABA-mediated inhibition, leading to continuous muscle activation. Episodes are not psychiatric. Common SPS therapies used are benzodiazepines, muscle relaxants, and immunotherapies.

2. LIFE-THREATENING OR HIGH RISK COMPLICATIONS

Respiratory Compromise

Severe rigidity and spasms may impair chest wall movement → **hypoventilation or respiratory failure**

Severe Generalized Spasm Episodes (“SPS Crisis”)

Sustained, whole-body rigidity and spasms; may resemble seizures but are non-epileptic. Patients are typically conscious but unable to move or speak

Autonomic Instability

Fluctuations in blood pressure, heart rate, and temperature; more prominent in severe variants (e.g., Progressive Encephalomyelitis with Rigidity and Myoclonus [PERM])

Rhabdomyolysis

Prolonged muscle contraction may lead to muscle breakdown and potential renal complications

3. IMPORTANT EMERGENCY CONSIDERATIONS

Musculoskeletal Injury Risk

Severe spasms and falls may cause fractures, tendon injuries, or joint damage → Evaluate carefully if focal pain is present

Respiratory Failure Risk with Opioid Use

Patients are often on chronic benzodiazepines; Avoid or use with extreme caution with opioids → Combination may cause respiratory depression or failure

Medication side effects or complications

Certain medications can cause side effects, such as benzodiazepines and baclofen, which can cause sedation and confusion. If these medications are abruptly stopped without a taper, especially from a high dose, there is a risk of medication withdrawal, including seizures. IVIg can cause infusion reactions (such as aseptic meningitis, hypertension) and has a risk of thromboembolism. Rituximab can cause infusion reactions (including allergy-type reactions and serum sickness) and is an immunosuppressant that can be associated with a higher risk of infections.

Use of Steroids

Steroids are not typically helpful in the management of acute SPS. Patients often have uncontrolled diabetes → Use cautiously due to risk of hyperglycemia or diabetic ketoacidosis

References:

1. Recurrent Acute on Chronic Respiratory Failure in Stiff Person Syndrome. Case Reports in Neurology. 2023. PMC10601654. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10601654/>
2. Acute Respiratory Failure in a Patient with Stiff-Person Syndrome. Neurocritical Care. PMID 27430873. <https://pubmed.ncbi.nlm.nih.gov/27430873/>
3. Stiff Person Syndrome. StatPearls. <https://www.ncbi.nlm.nih.gov/books/NBK573078/>

MEDICATIONS

