

Background

Stiff person syndrome (SPS)

- A rare autoimmune neurological disorder of the central nervous system that is characterized by painful rigidity and spasticity, commonly affecting the truncal and proximal limb muscles.¹
- A multidisciplinary approach to care with pain management, physical therapy, and occupational therapy is recommended (Figure).²
- Symptomatic treatment consists of GABAergic medications, baclofen, and disease-modifying therapy of intravenous immunoglobulin (IVIG).¹
- If refractory to above, Intrathecal baclofen (ITB) has shown to significantly reduce spasticity in SPS patients.^{3,4}

Purpose

- The overall aim of our study was to bring awareness to the complex pain management in patients with Stiff Person Syndrome and present a case of significant improvement in refractory spasticity in SPS following ITB trial.

Case Description

- **Chief Complaint:** 38-year-old male diagnosed with SPS with positive anti-glutamic acid decarboxylase antibodies (anti-GAD) in 2021 presents to chronic pain clinic with **severe muscle spasticity, skin hypersensitivity, and hyperacusis**.
- **Prior Treatments:** monotherapy with diazepam, combined pharmacotherapies, immunotherapy with IVIG and botulinum toxin injections (Table).
- **Intervention:** ITB trial with administration of **50 mcg baclofen** into intrathecal space of L2-3.
- **Result:** **Reported reduction in frequency of spasms and severity of pain** for three hours after administration.
- **Further Plan:** deemed an adequate candidate for and currently pending ITB pump implantation.

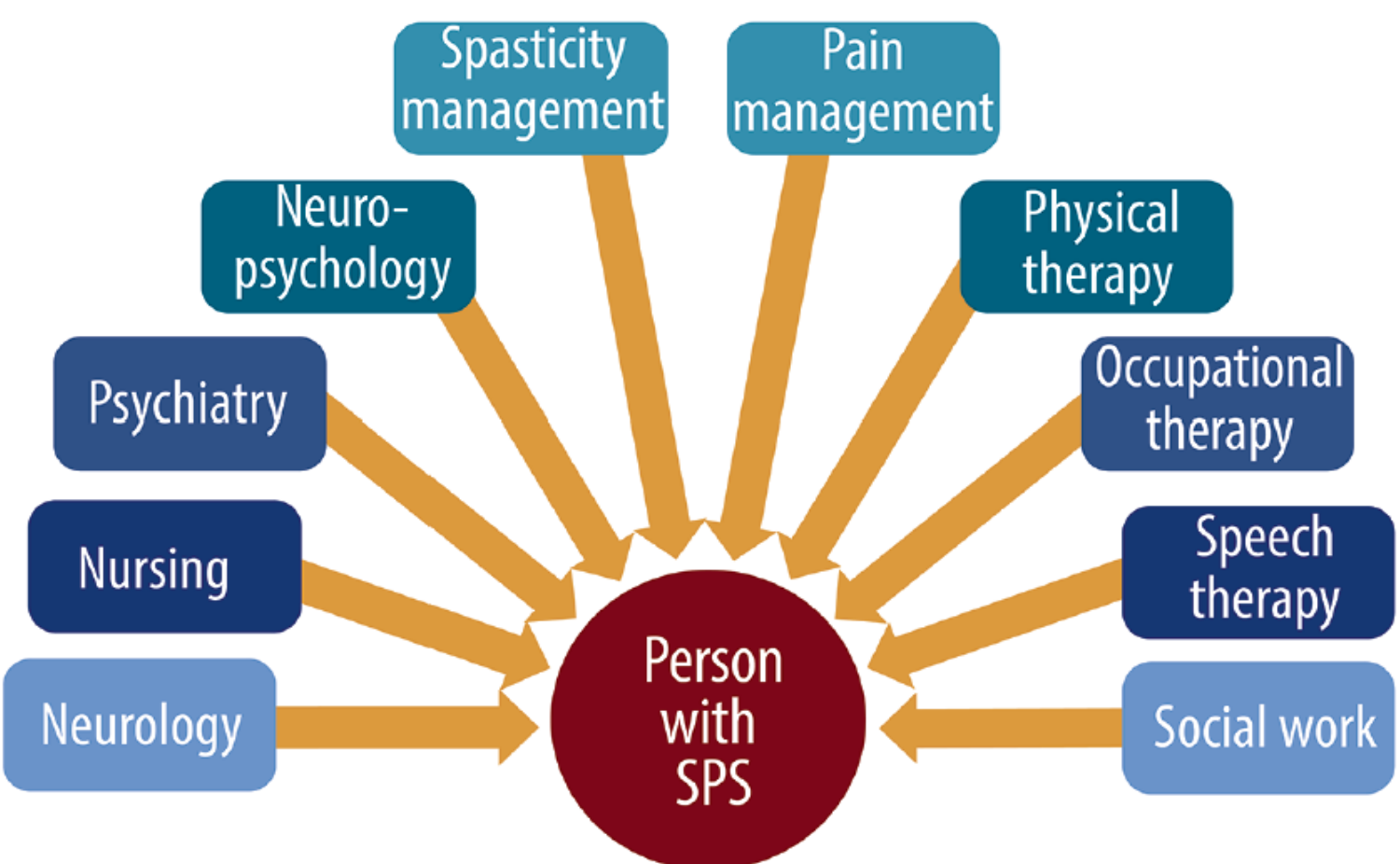


Figure. Multidisciplinary care for Stiff Person Syndrome (SPS).²

Table. List of treatment modalities the patient tried and failed prior to intrathecal baclofen trial.

Drug	Dose/day	Mechanism
Benzodiazepine		
Diazepam	25 mg daily	GABA _A agonist
Antispasmodics		
Baclofen	10 mg BID and 40 mg TID	GABA _B agonist
Tizanidine	4 mg QID	Alpha-2 adrenergic receptor agonist
Cyclobenzaprine	10 mg nightly	Reduction of tonic somatic motor activity and Serotonin 5-HT2 receptor antagonist
Anticonvulsants		
Oxcarbazepine	750 mg daily	Sodium channel blocker
Gabapentin	600 mg TID	Inhibits release of presynaptic excitatory neurotransmitters
Antidepressants/Anxiolytics		
Bupropion	300 mg daily	Norepinephrine-dopamine reuptake inhibitor
Venlafaxine	300 mg daily	Serotonin-norepinephrine reuptake inhibitor
Buspirone	15 mg QID	Serotonin 5-HT1a receptor partial agonist
Anesthetics/Analgesics		
Low-dose naltrexone	4.5 mg daily	Opioid antagonist
Ketamine nasal spray	10 mg QID PRN	NMDA glutamate receptor antagonist
Ketamine infusions	Per outside infusion center	NMDA glutamate receptor antagonist
Immunotherapy		
IVIG	1g/kg every 4 weeks	Neutralize anti-GAD antibodies
Procedural Intervention		
Botulinum Toxin injections	200 units	Presynaptic inhibition of acetylcholine release

mg: milligram, GABA: gamma-aminobutyric acid, BID: twice a day, TID: three times a day, QID: four times a day, PRN: pro re nata, NMDA: N-methyl-D-aspartate, IVIG: intravenous immunoglobulin.

SPS refractory to multiple oral medications and immunotherapies should prompt early consideration for ITB trial and implantation.

Discussion

- SPS is a debilitating autoimmune neurological disorder affecting one’s functionality and quality of life.
- If oral antispasmodics and immunotherapy fails, ITB has shown to be a successful adjunct for SPS with intractable muscle spasms.^{3,4}
- Following implantation, patients have reported improved functionality including ambulation without assisted devices; decreased pain and severity of spastic hypertonia.^{4,5}
- Initial ITB dosing ranging from 32 to 120 mcg/day and maintenance dosing from 48 to 320 mcg/day have been shown to be successful.⁵
- Advantages of ITB:⁵
 - Less systemic side effects compared to oral therapy
 - Ability to titrate to therapeutic cerebrospinal fluid levels
 - Maximized efficacy with smaller drug doses
- Common potential post-procedural risks include infection, hardware malfunction, pump failure, and catheter leakage leading to acute baclofen withdrawal.⁶

Conclusion

- Further research is needed to establish standard dosing protocol for ITB trial and post-implantation doses.
- Treating chronic pain from SPS can be complex; however, SPS refractory to multiple oral medications should prompt early consideration for ITB trial and implantation.

References

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