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# Physical Therapy Management for an Individual with Stiff Person Syndrome in the Acute Care Setting: A Case Report

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## Abstract

**Purpose:** Stiff Person Syndrome (SPS) is a rare neurological disease which is characterized by progressive rigidity and painful spasms of axial and limb muscles. We report here on the physical therapy management as well as medical complications which impacted the physical therapy plan of care for a patient with paraneoplastic SPS in the acute care setting. **Method:** This case report describes rehabilitation management of a 71-year-old woman who presented in the acute care setting for complaints of progressively worsening stiffness in the bilateral lower extremities leading to multiple falls as well as dysphagia. Primary interventions for this patient included passive range of motion for stretching, bed mobility such as rolling, supine to sitting transfers, and scooting at the edge of bed, and family and patient education. Education focused on the importance of maintaining range of motion and prevention of pressure wounds and contractures. **Results:** By the end of the hospitalization, the patient had a marked reduction in tone and spasms as well as improved bed mobility. Progress was limited due to multiple medical complications the patient faced during treatment. **Conclusion:** Physical therapist intervention for SPS in the acute care setting is challenging due to the paucity of literature present. However, physical therapy remains an important aspect of patient care in order to maximize independence and function from early on.



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**ABSTRACT**
**Purpose:** Stiff Person Syndrome (SPS) is a rare neurological disease which is characterized by progressive rigidity and painful spasms of axial and limb muscles. We report here on the physical therapy management as well as medical complications which impacted the physical therapy plan of care for a patient with paraneoplastic SPS in the acute care setting. **Method:** This case report describes rehabilitation management of a 71-year-old woman who presented in the acute care setting for complaints of progressively worsening stiffness in the bilateral lower extremities leading to multiple falls as well as dysphagia. Primary interventions for this patient included passive range of motion for stretching, bed mobility such as rolling, supine to sitting transfers, and scooting at the edge of bed, and family and patient education. Education focused on the importance of maintaining range of motion and prevention of pressure wounds and contractures. **Results:** By the end of the hospitalization, the patient had a marked reduction in tone and spasms as well as improved bed mobility. Progress was limited due to multiple medical complications the patient faced during treatment. **Conclusion:** Physical therapist intervention for SPS in the acute care setting is challenging due to the paucity of literature present. However, physical therapy remains an important aspect of patient care in order to maximize independence and function from early on.

**Key Words**: case report, stiff person syndrome, physical therapy, acute care

## INTRODUCTION

Stiff Person Syndrome (SPS) is an immune mediated disorder of the central nervous system with stimulus triggered muscle spasms of axial and limb muscles.[1] It is also known as Moersch-Woltz syndrome.[1] SPS may be idiopathic or associated with other disease processes, including manifestation as a paraneoplastic phenomenon with approximately 10% of all SPS cases presenting as a paraneoplastic variant. SPS is a rare condition, with an incidence estimated at about 1-2 cases per million.[2] The pathophysiology of SPS has been linked to high titers of anti-glutamic acid decarboxylase antibodies (GAD Ab).[3] This may lead to inhibition of glutamate decarboxylase and of GABA synthesis. This reduction in GABA may lead from muscle hyperfunction. Paraneoplastic SPS may also be associated with antiamphiphysin antibodies.[3] The cross-reactive binding of serum antibodies to malignant cells expressing neuronal antigens such as GAD and amphoteric fibrin may trigger an autoimmune response.[3]

SPS onset is typically insidious, usually with initial symptoms of stiffness in the trunk which eventually spreads to the bilateral lower extremities.[4] Some reports indicate bulbar involvement with symptoms of dysphagia and dyspnea.[4] The degree of stiffness experienced can vary from individual to individual and are superimposed by intermittent, painful spasms in the bilateral lower extremities and trunk. These painful spasms can be triggered by emotional and physical stimuli. Individuals may become diaphoretic and have elevated blood pressure with spasms.[4] SPS is progressive in nature and long-term complications can include extreme lumbar lordosis and gait impairments with limited hip and knee flexion during gait. These complications lead to significantly increased fall risk and make transfers and bed mobility progressively more difficult.

There is a paucity of literature regarding physical therapy management of SPS and its variants.[4-6] Furthermore, there is a significant gap in the literature regarding management of SPS in the acute care setting with much of the current literature comprised of case reports in the outpatient or inpatient rehabilitation settings. The purpose of this case study is to describe the acute care management of a patient with paraneoplastic SPS over a 40-day length of stay.

## CASE DESCRIPTION

The patient is a 71-year-old woman who presented for physical therapy evaluation in the acute care setting. She was retired and enjoyed living an independent, active lifestyle up until undergoing spinal surgery 2 months prior. At the time of her initial presentation to the emergency department, she presented with complaints of bilateral lower extremity spasticity, stiffness, and dysphagia. The patient was suspected of having complications from cervical myelopathy as a result of severe cervical spinal stenosis and subsequently underwent an operation for C4-C5 decompression with open C4-C6 laminoplasty. The patient was discharged to an acute rehabilitation facility for further rehabilitation after noting improvement in symptoms. Subsequently, she had worsening of her dysphagia and lower extremity stiffness and returned to the hospital for further workup. She was readmitted for new medical workup and her initial medical diagnosis was unclear as cervical myelopathy would not explain the bulbar symptoms present.

### Evaluation

At the time of evaluation, the patient presented with generalized weakness and increased extensor tone of the right upper extremity and bilateral lower extremities with each scoring a 3 on the Modified Ashworth Scale (MAS) noting significant difficulty with passive range of motion.[7,8] Full available range of motion was unable to be assessed as the patient would frequently have painful spasms leading to rigid hyperextension of the bilateral lower extremities and trunk. The patient required a maximum assistance of two in order to perform rolling for bed mobility and pressure relief and was unable to progress from supine to sitting due to the frequent spasms (Figure 1). The patient subsequently scored 0 on the Berg Balance Scale (BBS)[9] and was unable to complete a 10-meter walk test (10MWT)[10] and the 5 times sit to stand (5xSTS).[11] The evaluation was terminated as the patient became hypertensive with a systolic blood pressure of 181 and was diaphoretic. Physical therapy goals for this patient were within 2 weeks to be at a minimum assist for rolling and maximum assist for a supine to sit transfer so that the patient would be able to progress towards out of bed activity.

**Figure 1.** ICF Model of the Patient's Condition

| HEALTH CONDITION |
|---|
| Stiff Person Syndrome |

| BODY STRUCTURES / FUNCTION (IMPAIRMENTS) | ACTIVITY (TASKS) | | PARTICIPATION | |
|---|---|---|---|---|
| | Abilities | Limitations | Abilities | Restrictions |
| - Generalized weakness throughout body<br>- Increased tone and spasticity<br>- Hypersensitivity accompanied by painful spasms | - Able to assist with rolling<br>- Able to verbalize needs | - Limited bed mobility<br>- Unable to ambulate overground<br>- Unable to perform transfers | - Able to spend time with family in hospital setting for mental well-being | - Unable to travel to see grandchild and daughter in Seattle, Washington<br>- Unable to drive |

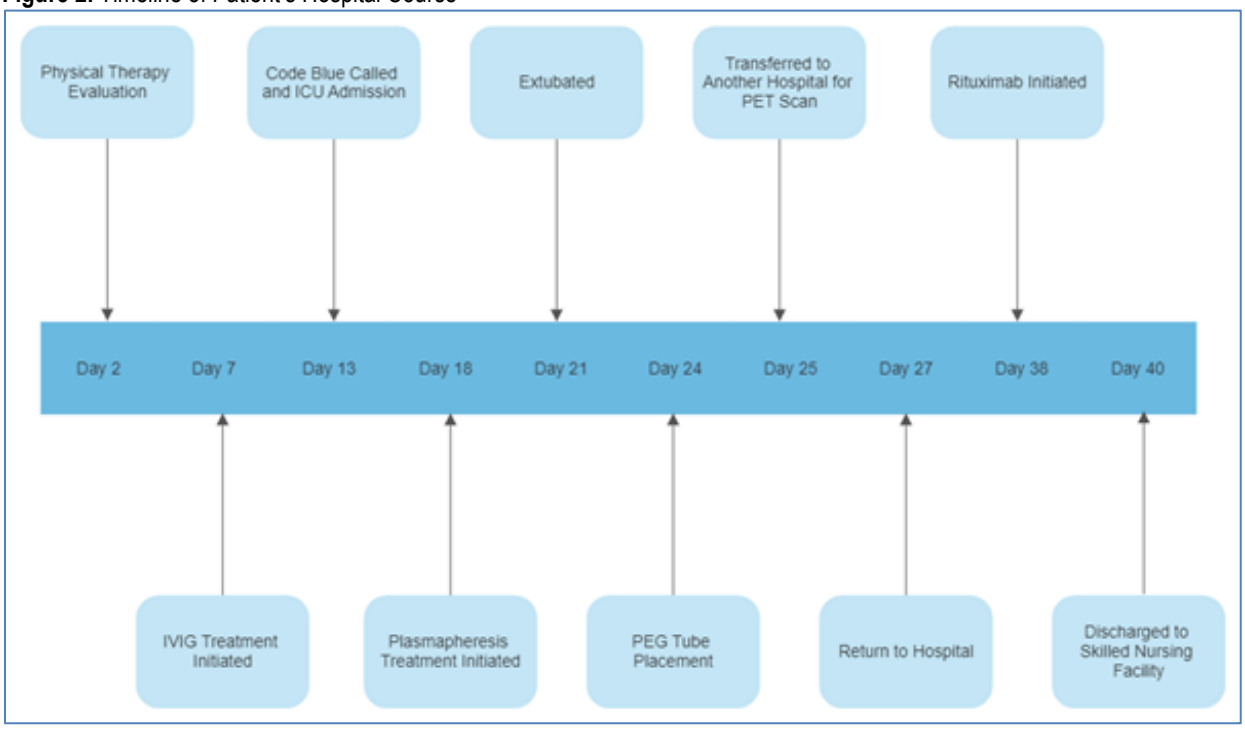
| ENVIRONMENTAL | | | |
|---|---|---|---|
| Internal | | External | |
| + | - | + | - |
| - Strong motivation to participate with therapy | - Multiple comorbidities which limited progress including but not limited to: cervical spinal stenosis, Waldenstrom's macroglobulinemia, Ogilvie's Syndrome | - Strong family support from sons and daughter who were agreeable to providing support as needed post discharge | - Concern about husband<br>- husband exhibiting signs of depression and expressing suicidal ideation |

**Hospitalization Course**

Throughout the patient's admission, the patient had multiple medical complications which significantly impacted the patient's physical therapy course of care. Seven days into the patient's hospitalization, the patient began intravenous immunoglobulin (IVIG) treatment for the management of suspected SPS which demonstrated a significant decrease in the patient's hypertonicity and spasms. However, physical therapy treatment was halted 11 days after admission due to the patient presenting with bloody sputum, increased work of breathing, and shortness of breath. 13 days after admission, the patient was transferred to the intensive care unit (ICU) due to a suspected aspiration event and was intubated and sedated for airway protection.

While in the ICU the patient began plasmapheresis treatment 5 days after being transferred and was extubated 8 days after being transferred. Eleven days after transferring to the ICU, the patient underwent a percutaneous endoscopic gastrostomy (PEG) tube placement due to continued dysarthria. Eighteen days after transferring to the ICU, a bone marrow biopsy was conducted due to concerns for Waldenstrom's macroglobulinemia,[12] a type of B-cell lymphoma; the diagnosis was confirmed through bone marrow biopsy. Rituximab therapy for the management of SPS was initiated 28 days after being transferred to the ICU and the patient was discharged to a skilled nursing facility for further rehabilitation shortly after (Figure 2).

**Figure 2.** Timeline of Patient's Hospital Course




**Intervention**

Initial intervention focused primarily on minimizing spasms to allow for further passive range of motion of the patient's bilateral lower extremities. The patient presented with significant risk for bilateral ankle plantarflexion contractures and a stretching program was put into place in order to increase range of motion and prevent contracture. However, due to the spasms, the patient was unable to tolerate significant stretching. Elastic wraps for compression onto the bilateral lower extremities were trialed to minimize spasticity and allow for increased range of motion. While there is some evidence to support the use of compression garments in children with cerebral palsy,[13] there is limited evidence to suggest benefits in individuals with SPS. Utilizing compression wrapping demonstrated significant benefits as the patient was able to demonstrate significant improvement in hip, knee, and ankle passive range of motion bilaterally without spasms. This allowed for the use of multipodus boots to maintain the patient's ankles in a dorsiflexed position to prevent further contracture, which the patient was unable to tolerate prior to the use of compression wrapping.

After IVIG treatment, the patient was able to initiate training for bed mobility as spasms were significantly decreased. The patient practiced rolling and moving from supine to sitting in the hospital bed with assistance as needed. While sitting, the patient worked on maintaining seated balance with reaching activity, weight shifting, and with trunk rotation.

After transferring to the ICU, intervention was limited due to increased fatigue and focused on increasing endurance in a seated position. The patient would move to sitting at the edge of bed with physical and occupational therapy and would be upright in a sling with the bed in chair mode with nursing staff to increase seated tolerance. The patient also worked on building upper extremity strength and weightbearing through the lower extremities with scooting at the edge of bed with assistance as needed. After PEG placement, the patient's activity tolerance again decreased due to significant abdominal distension accompanied by pain – of note, the patient was diagnosed with Ogilvie's syndrome during the course of acute rehabilitation prior to current admission. Due to these limitations, the patient was unable to progress to bed to chair transfers.

Throughout the course of hospitalization, the patient's family was instructed in appropriate passive range of motion for stretching and application of the multipodus boots to ensure the patient maintained available range of motion and to prevent contractures. The patient's family was dedicated and would assist the patient with passive range of motion daily despite COVID-19 protocols limiting visitation.

## RESULTS

Due to the multiple medical complications and the significantly longer than average length of stay, the patient was reevaluated by physical therapy 2 times at 2-week intervals. At the final reassessment, the patient's physical therapy goals were progressing from supine to sit at a supervised level and a bed to chair transfer using a transfer board at a moderate assist level. The patient was unable to meet these physical therapy goals prior to discharge to a skilled nursing facility.

At the time of discharge, the patient was at a standby assist level for rolling and a maximum assist for a supine to sit transfer. The patient was also a maximum assist for scooting at the edge of bed and was unable to progress to sit to stand or bed to chair transfers due to bilateral ankle contractures and deconditioning throughout the course of hospitalization (Table 1). The patient was able to progress to a BBS score of 3[9] but was unable to perform a 5xSTS[11] or 10MWT[10] by the time of discharge (Table 2). The patient had significantly reduced tone with a MAS[7] score of 2 in the bilateral lower extremities and right upper extremity.

**Table 1:** Functional Mobility Ability of Patient at Time of Evaluation and Discharge

| Functional Mobility | | |
|---|---|---|
| **Activity** | **Initial Evaluation** | **Discharge** |
| Rolling | Maximum Assist | Standby Assist |
| Supine to Sit | Unable | Maximum Assist |
| Sit to Stand | Unable | Unable |

**Table 2:** Outcome Measures Conducted at Time of Evaluation and Discharge

| Outcome Measures | | | |
|---|---|---|---|
| **Assessment** | | **Initial Evaluation** | **Discharge** |
| Berg Balance Scale | | 0/56 | 3/56 |
| 10-Meter Walk Test | | Unable | Unable |
| 5 Times Sit to Stand | | Unable | Unable |
| Modified Ashworth Scale (right upper extremity and bilateral lower extremities) | Right upper extremity | 3 | 2 |
| | Right lower extremity | 3 | 2 |
| | Left lower extremity | 3 | 2 |

## DISCUSSION

The management of SPS by physical therapy in the acute care setting is not well studied. The purpose of this case was to illustrate the efficacy of physical therapy treatment in the acute care setting and the barriers rehabilitation professionals may face during the course of care. This patient had a long and medically complex hospitalization which significantly complicated the plan of care initially outlined for physical therapy treatment.

Physical therapy interventions at this time were significantly limited due to the multiple medical complications which occurred during the stay and initially focused on spasticity and tone reduction techniques to facilitate mobility. As both medical management and range of motion exercises were unable to address her spasticity sufficiently, compression wrapping was trialed and successful, improving her ability to participate in rehabilitation activities. The patient's participation in rehabilitation varied significantly and required the rehabilitation team to collaborate with her medical team given frequent changes in patient's ability to tolerate mobility out of a supine position and multiple major complications interfered.

Early physical therapy examination in the acute care setting with appropriate interdisciplinary communication may assist with establishing the appropriate diagnosis and minimizing delays in treatment which can overall lead to improved outcomes.[3] Ongoing physical therapy is vital to assess changes in the patient's condition with medical management as response to pharmacologic intervention is variable. It should also be noted that with paraneoplastic SPS, significant improvement can be seen with cancer treatment and should be monitored closely by physical therapy to assess for functional change. With improving awareness of the condition amongst healthcare professionals, the role of physical therapists in the management of SPS will become increasingly defined.

Findings from this case report are not universally applicable to all individuals with SPS as they are based on a single case. However, this case study can still be used to inform higher methodological processes in the future with regard to physical therapy management of SPS in the acute care setting. It is paramount to continue to develop research around physical therapy management of SPS so that evidence-based intervention may be used in the future for improved outcomes for these patients.

**CONCLUSION**

Despite the patient being unable to reach all of her stated physical therapy goals within the context of the acute care setting and making slow progress with regards to functional mobility during her course of care, physical therapy management continues to be a critical service for individuals with SPS within the acute care setting. Physical therapy led to improvements with functional mobility and prevention of complications due to increased periods of immobility. Continuing to research the effects of physical therapy on SPS is crucial for future implementation of evidence-based care and increased awareness of the condition in all settings.

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