Utilization of Dantrolene in Stiff-Person Syndrome: A Case Report

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ABSTRACT

Background: Stiff-Person Syndrome (SPS) is a rare, idiopathic, neurological disorder characterized by axial and limb rigidity, and sudden spasms. It is often misdiagnosed and underrecognized. Dantrolene, a peripherally-acting agent, is used to treat spasticity and rigidity. We describe a patient with SPS treated with dantrolene during acute rehabilitation to reduce rigidity and improve function.

目的: 本研究は、スプリントマンシンドローム(SPS)患者に対するダンタルネンの使用についての一例を報告し、急性リハビリテーション中の筋緊張の治療について考察するものである。

結果: 75歳の女性で、緊張と不安の既往歴があり、右コレス骨折を治療して非手術的である。個々のアーチナセリドに神経衰弱と緊張を伴っていた。GABA誘導体であるDannyヶ月目には筋緊張が治まり、機能が改善した。

結論: ダンタルネンは、SPSに対する有用な追加治療として機能する。

CASE REPORT

Setting: University hospital-based acute rehabilitation.

Patient: 75-year-old woman with Stiff-Person Syndrome (SPS) with a recent fall and Colles fracture.

Case Description: Four months prior to admission, the patient was diagnosed with SPS, negative for anti-GAD antibodies. Diagnosis was based on a 3-year history of progressive rigidity leading to frequent falls and fractures. Her gait was unsteady, and she sustained a sacral pressure ulcer during acute hospitalization. On admission, her history was remarkable for undiary gait and muscle cramps exacerbated when startled or excited. Examination was remarkable for rigidity in her axial and limb muscles. She presented at the maximal assist level for transfers and toileting and moderate assist level for grooming and ambulation using a platform walker. She was unable to tolerate titration of dantrolene due to dizziness and syncope.

Results: During acute rehabilitation, rigidity was treated with titration of dantrolene (25 to 50 mg four times daily) in addition to maximal tolerated doses of diazepam (1 mg qAM and 2mg qPM) and baclofen (20mg TID). The addition of dantrolene reduced rigidity and improved range of motion, both subjectively and objectively.

Discussion: SPS results in significant activity of daily life and ambulatory dysfunction as exemplified by her pressure ulcer and multiple falls. Although GABA agonists are the preferred treatment for SPS, the adverse effects of high doses can increase the risk of falls. Dantrolene reduced muscle rigidity and improved function without sedative or hypertensive effects.

Conclusion: Dantrolene was a useful additional treatment for SPS rigidity.

TREATMENT

Physical Therapy: strength training, balance, endurance, range of motion exercises.

Occupational Therapy: daily activities, compensatory skills, range of motion exercises.

Rehabilitation Psychology: education and counseling focused on accommodating the physical and psychosocial impacts of SPS, including management of anxiety and depression.

RESULTS

- She progressed to supervision for grooming, moderate assistance for transfers and toileting, and minimal assistance for ambulation.
- Anxiety and depression were improved with psychological counseling, buspirone, and paroxetine.
- Improvement of Modified Ashworth Score from 3 to 1-2 in all limbs with dantrolene.

CONCLUSION

- SPS is a rare, progressive disorder characterized by rigidity, caused by hyperactivity of spinal cord projection neurons.
- GABA agonists are the current primary treatment, but central side effects (sedation, hypotension) limit dosing, especially in the elderly who have a high falls risk.
- Dantrolene, a peripherally-acting agent, lacks significant central side effects.
- Although its potential use is considered in SPS literature, there are no clinical trials.
- Recommended dosage at 200-400mg/day; side effects include diaphoresis and hepatotoxicity.

FIGURE 2: PATHOGENESIS OF STIFF-PERSON SYNDROME

- GABA agonists are GABA enhancing drugs and considered the first-line treatment;
- Benzodiazepines and baclofen (GABA agonists) are the preferred treatment for SPS, but central side effects (sedation, hypotension) limit dosing, especially in the elderly who have a high falls risk.
- Dantrolene, a peripherally-acting agent, lacks significant central side effects.
- Although its potential use is considered in SPS literature, there are no clinical trials.
- Recommended dosage at 200-400mg/day; side effects include diaphoresis and hepatotoxicity.