# Intrathecal Baclofen Therapy for GAD (+) Stiff-Person Syndrome: A Case Series of 3 Patients

Lior Ungar MD PhD; Takaomi Taira MD, PhD; Zion Zibly MD
Department of Neurological Surgery, The Charles Clore Hospitalization Tower, West Wing Sheba Medical Center, Ramat Gan, Israel
The 2015 Dr. Pinchas Borenstein Talpiot Medical Leadership Program, The Chaim Sheba Medical Center, 52621 Tel HaShomer, Israel.
Department of Neurosurgery, Tokyo Women's Medical University, Tokyo, Japan

assfn 2016 BIENNIAL MEETING
June 18-21 | Chicago, IL

Sheba - Academic Medical Center Hospital

## Objective

To report three cases of a rare disorder, GAD (+) Stiff Person Syndrome (SPS), treated with Intrathecal baclofen therapy

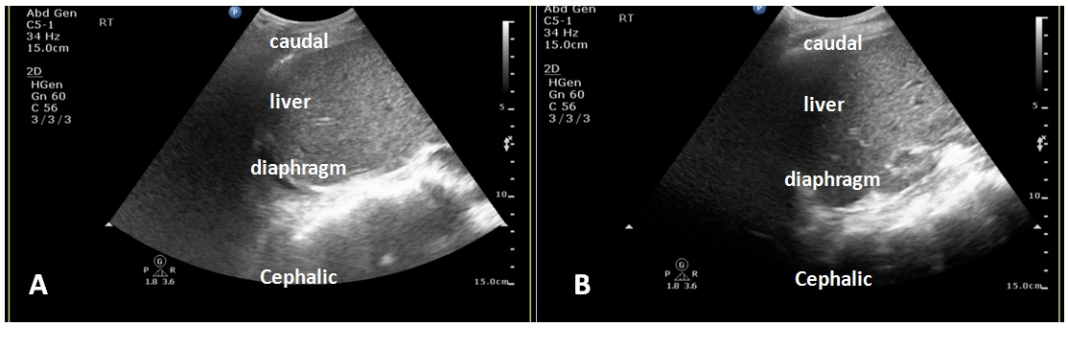
## Introduction

Stiff person syndrome is a rare disorder (the prevalence is estimated at about 1/1,000,000) of unclear etiology, characterized by progressive rigidity and stiffness. The stiffness primarily affects the truncal muscles and is superimposed by spasms, resulting in postural deformities. Stiff-Person syndrome is an autoimmune disorder and highly correlates with the presence of anti–glutamic acid decarboxylase (GAD) antibodies. Baclofen and benzodiazepines are the primary treatments for SPS. Autoimmune suppressive treatments such as steroids, rituximab and plasma exchange have been also used in SPS patients; however, when the above treatments are inadequate, intrathecal baclofen may be used.

## Methods

We reported 3 patients with GAD (+) SPS which presented severe refractory neurological manifestations of the disease and greatly benefited from intrathecal baclofen (ITB) therapy.

**Figure 1**




Diaphragm movement and contractility Eevaluation by thoracic ultrasound. A.Diaphragm view in the right subcostal area before ITB pump implantation. B. Diaphragm view in the right subcostal area after ITB pump implantation.

## Results

1. A 34 year old man diagnosed with SPS presented with refractory progressive spasms of the lower trunk. Unresponsive to oral baclofen, diazepam, steroids and eventually IVIG, the patient had lost his ability to walk. Treated with implantation of programmable baclofen pump, the patient's spasm had decreased dramatically, and he had regained his ability to walk.

2. The second patient was a 33 year old woman diagnosed with acute SPS developed to a life threatening disease. Starting with increased muscular tonus and limb hyperextensions; the patients had developed progressive diaphragmatic spasm, leading to respiratory failure. While hospitalized at the intensive care unit, the patient underwent a promising intrathecal baclofen administration trial following an implantation of intrathecal baclofen pump. The diaphragm contractility was regained (as demonstrated in ultrasound study - figure 1). Soon after, the patient had withdrawal from mechanical ventilation.

3. The third patient was a 17 year old girl presented with truncal muscle spasm that was induced by voluntary movements, resulting in flexion of the arms and extension of the legs. Initially she was treated with oral diazepam and baclofen, but the symptoms progressed to involve the truncal muscles. She underwent plasma exchange without benefit. Muscle spasm gradually got worse and was associated with severe spasm related pains. Two months after the patient's admission, the patient demonstrated progressive breathing difficulties due to muscle spasm, and she had to be managed with mechanical ventilation. After an ITB pump implantation, the patient's condition rapidly improved, and after withdrawing from mechanical ventilation and a short period of rehabilitation, the patient had been discharged from the hospital fully functional, with a substantial reduction in her symptoms.

## Conclusions

We described three cases of GAD (+) SPS patients, presented with severe debilitating and even life threatening manifestations of their condition. After poor response to conventional SPS therapy, including oral baclofen, benzodiazepines, steroids and plasma exchange, all 3 patients described highly benefited from intrathecal baclofen (ITB) therapy. We conclude that intrathecal baclofen (ITB) therapy is a highly recommended therapy in the early stages of SPS.