

Intrathecal baclofen for Friedreich's ataxia

5 patients with spinocerebellar ataxia 3 or 7 or Friedreich's ataxia were included in this observational multicenter study. The patients were interviewed and completed outcome measures assessing pain (The Brief Pain Inventory), fatigue (Fatigue Severity Scale), and life satisfaction (LiSAT-9) before and 1 year after the treatment. Spasticity (Modified Ashworth Scale) and spasm frequency (SPFS) were measured objectively for each patient.

Results: The mean treatment time was 1.9 years. Evaluation of established standard forms revealed symptomatic relief from spasticity, spasms, pain, and fatigue in addition to improved body posture, sleep, and life satisfaction after ITB treatment.

They report the potential beneficial effects of ITB treatment in patients with inherited ataxia who also suffer from spasticity/spasms. ITB treatment indication in neurological disorders allows for extension to the treatment of spasticity/ spasms in patients with hereditary ataxia ¹⁾.

Case reports

A report describes the successful management of painful spasms in a 65-year-old woman with [Friedreich's ataxia](#) (FA) via [intrathecal baclofen](#) (ITB) therapy following unsuccessful medical treatments.

To Kalyvas et al., knowledge, this is the third reported case in the literature. Unfortunately, the pathophysiological characteristics of muscle spasms in FA are not well explored and understood while the therapeutic mechanisms of the different treatments are rather vague. Taking into consideration the suggested spinal atrophy in FA, the clinical resemblance of FA and chronic spinal injury muscle spasms, together with the rapid ITB therapy effectiveness in alleviating FA muscle spasms, they attempted to suggest a putative pathophysiological mechanism acting at the spinal level and possibly explained by the presence of independent spinal locomotor systems producing muscle spasms. Specifically, overexcitement of these centers, due to loss of normal regulation from upper CNS levels, may result in the uncontrolled firing of secondary [motor neurons](#) and may be the key to producing muscle spasms. However, further research under experimental and clinical settings seems to be necessary ²⁾.

A 50-year-old female patient with Friedreich ataxia (FA) was treated successfully with an intrathecal baclofen (ITB)-delivering pump for painful spasms. This is the second reported case of FA where ITB relieved painful and disabling spasms. Berntsson et al., suggest that ITB should be considered in the treatment of disabling spasms in patients with FA ³⁾.

Ben Smail et al., reported a patient suffering from Friedreich's ataxia (FA) with very painful and disabling spasms that were improved markedly by intrathecal baclofen infusion. This is the first report of an intrathecal baclofen-delivering pump implantation in an FA patient ⁴⁾.

¹⁾

Berntsson SG, Gauffin H, Melberg A, Holtz A, Landtblom AM. Inherited Ataxia and Intrathecal Baclofen for the Treatment of Spasticity and Painful Spasms. *Stereotact Funct Neurosurg*. 2019;97(1):18-23. doi: 10.1159/000497165. Epub 2019 Mar 14. PMID: 30870851.

2)

Kalyvas AV, Drosos E, Korfias S, Gatzonis S, Themistocleous M, Sakas DE. Intrathecal Baclofen Therapy for Painful Muscle Spasms in a Patient with Friedreich's Ataxia. *Stereotact Funct Neurosurg*. 2018 Jun 8:1-4. doi: 10.1159/000489220. [Epub ahead of print] PubMed PMID: 29886479.

3)

Berntsson SG, Holtz A, Melberg A. Does intrathecal baclofen have a place in the treatment of painful spasms in friedreich ataxia? *Case Rep Neurol*. 2013 Nov 21;5(3):201-3. doi: 10.1159/000356823. eCollection 2013. PubMed PMID: 24348400; PubMed Central PMCID: PMC3861848.

4)

Ben Smail D, Jacq C, Denys P, Bussel B. Intrathecal baclofen in the treatment of painful, disabling spasms in Friedreich's ataxia. *Mov Disord*. 2005 Jun;20(6):758-9. PubMed PMID: 15756654.

From:

<https://neurosurgerywiki.com/wiki/> - **Neurosurgery Wiki**

Permanent link:

https://neurosurgerywiki.com/wiki/doku.php?id=intrathecal_baclofen_for_friedreich_s_ataxia

Last update: **2024/06/07 02:54**

