Spinal cord stimulation in the treatment of neuropathic pain in chronic inflammatory demyelinating polyneuropathy: a case report

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# ABSTRACT

We describe a case of a 70-year old man with sensorimotor chronic inflammatory demyelinating polyneuropathy (CIDP) with small-fibre involvement resulting in severe diffuse neuropathic pain which was refractory to immunotherapy and anti-neuropathic medication. His pain was successfully treated with implantation of a spinal cord stimulation (SCS) system comprising bilateral cervical and lumbar epidural leads. Following SCS programming, he experienced a 50% reduction in average pain severity with substantial improvement in quality of life, persisting at 18 months after surgery. SCS has been employed to treat a variety of neuropathic pain syndromes. However, this is the first report to our knowledge of SCS utilised effectively for pain in CIDP. This therapy should be considered in painful CIDP for neuropathic pain refractory to medical management, though further studies are required to evaluate its efficacy.

## Key words

Spinal cord stimulation; chronic inflammatory demyelinating polyradiculoneuropathy; pain

# Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is the most common autoimmune neuropathy with a prevalence of 0.8–8.9 cases per 100,000 individuals. It presents as a progressive symmetrical sensorimotor polyneuropathy with a clinical progression lasting, by definition, more than 8 weeks [1]. Symptomatically, motor deficits in proximal and distal muscle groups predominate, though up to 90% can display sensory manifestations during the course of the disease [2, 3] and around 10% exhibit a pure sensory variant [4]. Although not a classical sensory feature, chronic neuropathic pain is well-recognised in CIDP with one study reporting that 39% of patients experienced moderate to severe pain [5]. The mainstay of CIDP treatment is regular and repeated immunomodulatory therapy such as intravenous immunoglobulin (IVIG), plasma exchange or corticosteroids. Here we describe a case of sensorimotor CIDP with small-fibre involvement causing severe neuropathic pain which was refractory to medical management but treated successfully with spinal cord stimulation (SCS).

# Case description

A retired 70-year old Caucasian man was diagnosed with painful sensorimotor relapsing-remitting CIDP approximately twenty years prior, with symptoms present for at least 6 years before initial diagnosis. Peripheral pain had been a prominent feature from onset and had initially responded to immunotherapy. Latterly, in view of continuous pain no longer responsive to immunotherapy, skin biopsy was performed and demonstrated very profound intra-epithelial small-fibre loss. The pain was neuropathic in character (expressed as “sharp” and “shooting”), localized to all four extremities in a glove-and-stocking distribution and was exerting a considerable detrimental effect on quality of life. He had initially been treated with corticosteroids before commencing IVIG therapy ten years prior. He received a course of 160 g IVIG every six weeks which improved his neuromuscular function with no effect on the pain. Prior to treatment with SCS he had been taking Pregabalin 300 mg twice a day with marginal pain reduction despite side effects including erectile dysfunction. He had tried alternative neuropathic pain agents previously, including gabapentin, duloxetine and amitriptyline, with intolerable adverse effects including mood disturbance, acute closed-angle glaucoma and weight gain. His past medical history included well-controlled steroid-induced diabetes mellitus and a recent diagnosis of prostatic carcinoma managed expectantly.

He was referred by his treating neurologist to the Pain Neuromodulation service. Following multi-disciplinary assessment, he was offered SCS as a therapy for chronic neuropathic pain. Pre-operatively, pain scores were 8/10 on average and 10/10 at worst on the numeric rating scale (NRS). After informed consent, he underwent full implantation of a SCS system. Four eight-contact dorsal epidural leads were placed percutaneously at cervical and thoracolumbar levels bilaterally (Figure 1). Paraesthesia coverage of all four extremities was achieved during on-table testing. The leads were connected to a 32-channel rechargeable pulse generator (Precision Spectra, Boston Scientific, USA) which was implanted in the left buttock. Post-operative recovery was uneventful.

Following programming (baseline settings: continuous stimulation to all four leads at 1 kHz, 2.4 mA, pulse width 130 µs), the patient reported an improvement in average pain score to 4/10 on the NRS, which he deemed to be an excellent result. 18 months following implantation his average pain remained at 4/10 with substantial ongoing improvement in overall quality of life. Formal assessments of pain and quality of life at baseline and 18-month follow-up are summarised in Table 1. This benefit was accompanied by reduction in the Pregabalin dose to 150 mg twice daily. His patient programmer was configured with additional programmes to direct increased stimulation to each of his extremities on demand, a facility he employed sporadically as required. The rest of his CIDP treatment remained unchanged. Despite electrophysiological evidence of underlying disease progression 12 months following implantation, the response of his pain to SCS was stable.

# Discussion

SCS is an increasingly utilized neuromodulatory therapy in chronic pain. It is applied in various intractable neuropathic pain syndromes but class I evidence is confined to its use in the spectrum of ‘failed back surgery’ and complex regional pain syndromes [6, 7]. Case series exist for other neuropathic indications including painful diabetic polyneuropathy, postherpetic neuralgia and phantom limb pain [8-10]. The case described here is, to our knowledge, the first description of SCS used successfully for treating pain in CIDP.

Pain in CIDP has been reported to improve following immunotherapy aimed at treating the underlying inflammatory process and to respond poorly to symptomatic drug treatments (Barohn et al 1989). In this case, pain had become refractory to IVIG despite its beneficial effect on neuromuscular function, probably reflecting the extent of the underlying small-fibre neuropathy. The neuropathic nature and severity of the pain, its major impact on quality of life and failure to achieve satisfactory analgesia with medical management warranted consideration of SCS. The diffuse distribution demanded a four-lead implantation strategy with cervical and thoracolumbar leads providing upper and lower extremity coverage, respectively. The 50% improvement in average pain severity is comparable to the benefit seen in painful diabetic polyneuropathy [8] and failed back surgery syndrome [11, 12] and has been sustained for up to 18 months post-implantation despite electrophysiological evidence of disease progression.

This case demonstrates that SCS has the potential to be an effective treatment option for refractory neuropathic pain in CIDP. Practitioners caring for patients with painful CIDP should consider referral for neuromodulation in those who fail to achieve adequate pain control with immunotherapy and analgesic medication. Further studies are required to evaluate the efficacy of SCS for this indication.

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# Table 1

Summary of clinical assessments prior to SCS and at 18-month follow-up. NRS pain scores are on a scale 0 (no pain) to 10 (worst imaginable pain). Brief Pain Inventory (BPI) severity score is the mean score for BPI items 1–4, and interference score the mean score for BPI items 5A–5G, both on a scale from 0 (no pain/no interference) to 10 (worst imaginable pain/interferes completely). EQ-5D-5L quality of life index score is calculated using an online tool with the United Kingdom value set (https:\\euroqol.org) and is on a scale from 0 (health state equivalent to death) to 1 (full health).

|  |  |  |
| --- | --- | --- |
|  | **Baseline** | **18-months** |
|  |  |  |
| Average pain score (NRS) | 8 | 4 |
| Worst pain score (NRS) | 10 | 8 |
| BPI severity score | 9.0 | 5.0 |
| BPI interference score | 8.1 | 5.7 |
|  |  |  |
| EQ-5D-5L health state | 31352 | 21132 |
| EQ-5D-5L index score | 0.103 | 0.683 |

# Figure 1 - caption

Post-implantation anteroposterior (AP) radiographs of the bilateral thoracolumbar (A) and cervical (B) dorsal epidural eight-contact leads. Both thoracolumbar leads span the T11/12 to L1 vertebrae, the left cervical lead C5/6 to C7/T1, and the right cervical lead C6/7 to T1/2.