Brain and Spine 3 (2023) 101785

Contents lists available at ScienceDirect

Brain and Spine

journal homepage: www.journals.elsevier.com/brain-and-spine

# Remarkable improvement of neurological deficits after surgery in patients with Idiopathic spinal cord herniations. The impact of peroperative neuromonitoring. Case reports

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ARTICLE INFO

Handling Editor: Prof F Kandziora

*Keywords:*
Idiopathic spinal cord herniation
Brown-Sequard syndrome
Case report
Myelopathy
Spastic paraparesis

ABSTRACT

*Introduction:* Chronic Idiopathic Spinal Cord Herniation (ISCH) is a very rare spinal cord deformation occurring predominantly in thoracic levels. ISCH lead to progressive myelopathy, spastic paraparesis and Brown Séquard syndrome.
*Research question:* We want to hypothesize that a) the herniated segment can regain its function after untethering despite long-term and complete neurologic dysfunction. b) Intraoperative Electrophysiologic Monitoring (IOEPM) may identify intraoperative changes by monitoring specific neural pathways confirming the efficacy of the intervention in the forthcoming cases.
*Material & method:* It is a retrospective review of data of two cases prospectively collected showing improvement of neurological deficit in cases of ISCH in thoracic levels. We describe two patients with progressive neurological deficits due to ISCH who underwent surgery using electrophysiologic monitoring and have been followed to reach remarkable clinical improvement.
*Results:* The spastic paraparesis of the first case improved remarkably after surgery. Complete foot drop of the other case, persistent for 7 months before intervention, improved after the release of the herniated segment of the cord. Peroperative electrophysiological monitoring did not show changes during surgery.
*Conclusion:* We want to hypothesize that the herniated segment can regain its function after untethering despite long-term and complete neurologic dysfunction. Intraoperative Electrophysiologic Monitoring (IOEPM) may confirm the efficacy of the intervention in the forthcoming cases.

## 1. Introduction

Idiopathic Spinal Cord Herniation (ISCH) is a rare disorder usually involving the thoracic spine except for one case, which occurred at the C7 level (Rajapakse et al., 2016). The first case of ISCH was reported by Wortzman et al. in 1974 (Wortzman et al., 1974). It occurs predominantly in the female sex. A congenital dural layer disorder with or without an acquired event is needed to cause the herniation of the spinal cord. Trauma, inflammation, and disk disease have been proposed as predisposing factors for ISCH. Pulsation of cerebrospinal fluid (CSF) is the other theory for cord herniation into the split dural pouch. ISCH appears in the ventral and ventrolateral side of spinal cord and therefore, the posterior subarachnoid space enlarges in the same level. ISCH can lead to progressive myelopathy, spastic paraparesis and Brown-Séquard syndrome. Brown-Sequard syndrome is the most common clinical presentation and has the best prognosis while complete and long–standing preoperative neurological deficits are usually not reversible (Ellgera et al., 2006). We present two patients with ISCH, with good recovery after surgery despite preoperative long-term and advanced neurological deficit.

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https://doi.org/10.1016/j.bas.2023.101785
Received 19 March 2023; Received in revised form 10 May 2023; Accepted 20 July 2023
Available online 21 July 2023

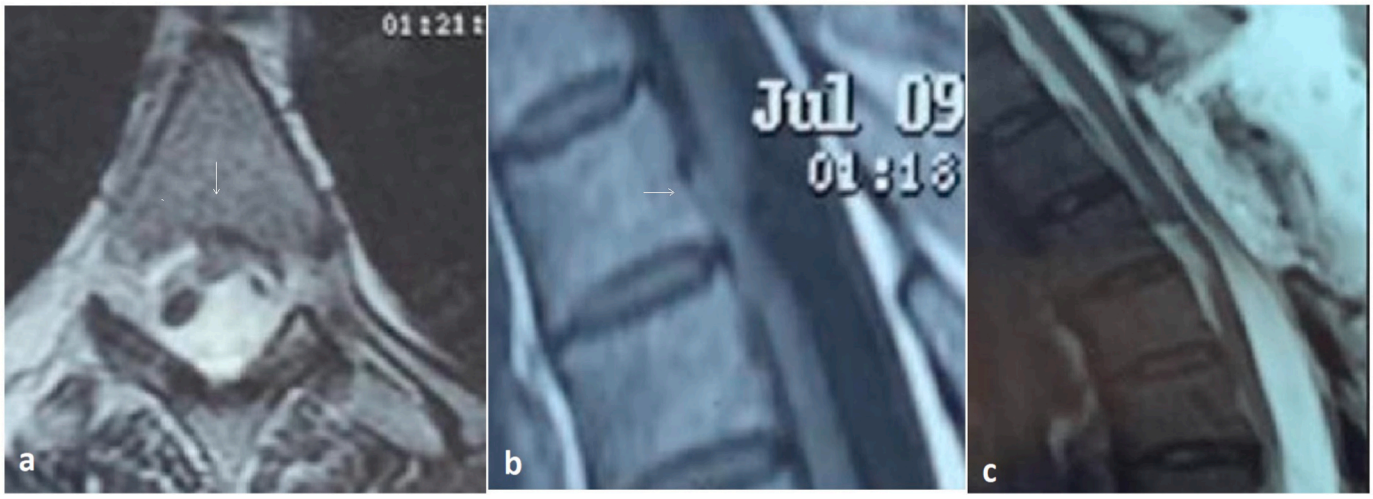


**Fig. 1.** Preoperative MRI in case 1. a: axial T2 MRI and b: sagittal T1 MRI showing the typical 'scalpel sign'. c: postoperative sagittal T2 MRI presenting the release in the ventral herniated part.

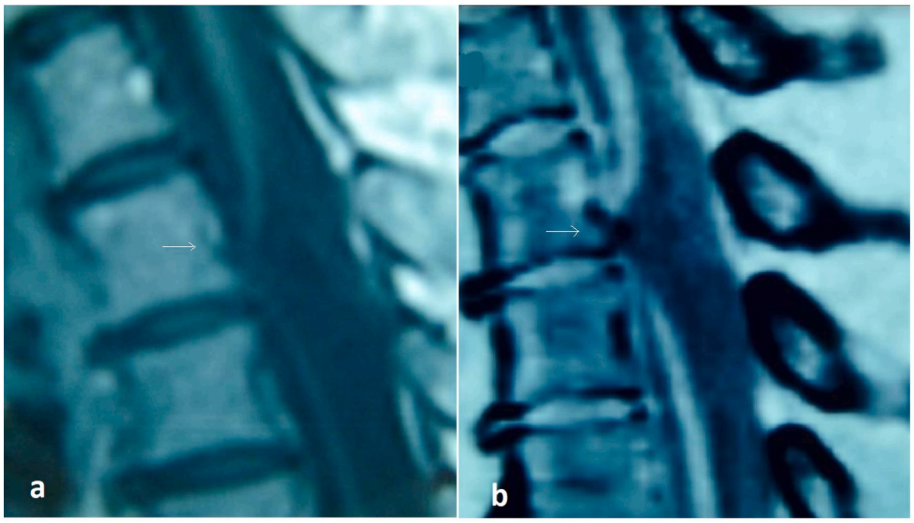


**Fig. 2.** Preoperative MRI in a female (case 2) A: axial T2 and B: sagittal T1 MRI.

## 2. Case report

Case 1- A 55-year-old female was referred to us due to progressive weakness of both legs and occasional overflow incontinence of more than 2 years duration. She was spastic in both legs and her pin prick sensation was diminished bilaterally up to the T10-11 level. The prominent findings in imaging were ventral kinking of the spinal cord with close adherence of the cord to the dura and scalloping of the adjacent T7 vertebral body. No enhancement was visible after contrast material injection (Fig-1a, b). Different aspects of the disease and surgical treatment, with the intention to release the adherence of the cord at that level were discussed with patient and her family, and informed consent was taken for surgical intervention.

Case 2- A 47-year-old woman presented with progressive weakness of the left leg, for a duration of 1 year. She had a motorcycle accident two years prior and progressive left foot drop of 7 months duration. Neurological examination revealed Brown-Séquard syndrome with weakness of the left leg and an impaired sense of temperature and pain below the T-4 dermatome on the right side without any sphincter problems. MRI revealed a left ventrolateral spinal cord displacement at the T2–3 vertebral level, with tethering and incarceration of the cord without changes in signal intensity in the spinal cord. MRI demonstrated a ventral C-shaped kinking of the thoracic cord in the sagittal views (Fig. 2a and b). Surgery was recommended, and written consent was obtained from the patient.

## 3. Surgical technique in both cases

Under general anesthesia and without using steroid or neural protection medications perioperatively, each patient was placed in prone position and neuromonitoring system was applied. After appropriate laminectomy, the dura was opened in the midline and careful microdissection and detachment of the dentate ligaments was performed. The rostral, caudal, and lateral edges of the ventral dural defect were defined using microscope. Using sickle knife, the dural defect was enlarged and the herniated part of spinal cord was released into the dural sac. The dura was duplicated ventrally in both cases. Discolored ischemic spinal cord appeared like a tumor in the first glance in both cases, but it became normal in appearance after the release of the dural attachments. Visceral dural layer could be enlarged and then the space was filled with fat and gelfoam (Fig. 3a–d). Somatosensory evoked potentials and motor evoked potentials (using NIM Vital™ Medtronic system) did not change remarkably neither in the upper nor in the lower limbs in both cases during surgery.

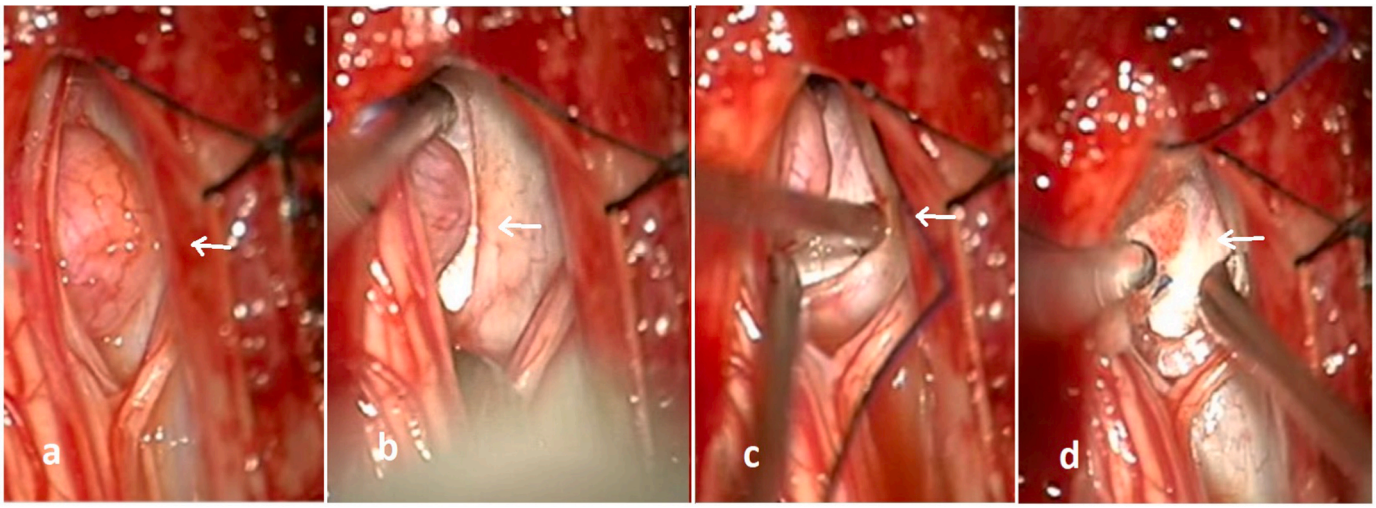


**Fig. 3.** Intraoperative photography in case 1. A: released the dentate ligament and arachnoid band to reposition the herniated part into the spinal sac. B and C: releasing upper, lower, and lateral edge of the dural defect. D: enlargement of the ventral dural defect with sickle knife. E: using gelfoam to fill the dural split space. F: postoperative sagittal T2 MRI presents the release in the ventral herniated part.

## 4. Postoperative course

**Case 1**. Spasticity was improved remarkably by reduction of leg muscle tone and better quality of walking after the operation but there was no change in dysesthesia in the legs after the 2 years follow up. In the post-operative MRI, no reattachment or tethering was detectable (Fig. 1c).

**Case 2.** Six months after surgery, left ankle dorsiflexion was improved to 3/5 in power and the pain and temperature sensations in right leg recovered remarkably. The control MRI did not show any retethering and abnormal findings after one year.

## 5. Discussion

Several mechanisms have been suggested for ISCH, including congenital dural defect, disk disease with dural erosion, and trauma. The direction of CSF flow in the spine is from the posterior subarachnoid space to the anterior and is more prominent in the cervical and thoracic regions. It seems that CSF pulsation and a concomitant defect in the dural layers may lead to progressive spinal cord herniation (Ellgera et al., 2006). Based on MRI findings, some authors have classified ISCH into three types: Type K (Kink), Type D (Discontinuous), and Type P (Protrusion) (Imagama et al., 2009). These types may represent *different stages* in the development of ISCH. Intraoperative findings have been variable in the reported cases and even after biopsy specimens were taken (Bartels et al., 2017).

Both nonsurgical and surgical treatments have been recommended in the management of ISCH (Imagama et al., 2009; Hostettle et al., 2021). To reach the dural defect in the surgical management, either anterior or posterior approaches have been suggested (Imagama et al., 2009). Posterior approach was more feasible and less morbid in our hands.

In both cases, we widened the dural defect, released the herniated cord and tried to remove the dural fold to prevent spinal cord re-herniation. In case 1, improvement of the function of the corticospinal tract was obvious but the function of the dorsal and lateral columns did not change during the follow up period. In case 2, we did not expect the foot drop to recover due to the long-term complete deficit but not only the pain and temperature sensation improved on the right side but also the left ankle dorsiflexion improved to 3/5 in power even though still incomplete. *To the best of our knowledge, this is the first case series of ISCH reporting complete improvement of neurological deficit after surgery*. Regarding the risk of recurrence or retethering, we will follow up with our cases regularly (Selviaridis et al., 2009).

The main purpose of surgery is to release the incarcerated spinal cord and to prevent re-lodgment of the cord into the dural sac. To prevent re-herniation of spinal cord, different techniques have been suggested: widening the dural defect and refilling the space with fat or gelfoam, closing the dural defect with suture, micro staples with and without graft (Delgado-López et al., 2017), using artificial graft with dentate ligament hitch stitches, and spinal cord suspension (Lui et al., 2018).

According to our findings and a brief review of the relevant literature (Delgado-López et al., 2017; Lui et al., 2018), we suggest that it is better to release the incarcerated spinal cord without being concerned about the severity and chronicity of neurological deficits and provide the chance of improvement for the patients even though no obvious changes in peroperative neuromonitoring electrophysiologic findings would appear.

## 6. Limitations

Two cases are very few to conclude an exceptionally good outcome for a procedure. The availability of a good neuromonitoring system during surgery could explain our results in a scientific way even though we did not encounter remarkable changes in the waves during the operations. Long term follow up would be necessary to conclude a persistent and good functional outcome and not an incidental occurrence.

## 7. Conclusion

By these clinical observations, we would like to suggest that, in cord herniation syndrome, as we like to coin it, the herniated segment can be alive and regain function after cord untethering despite of long-term and complete neurological deficit.

## Declaration of competing interest

The authors declare that they have no conflict of interest. Ethical approval: “All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (NIMHANS ethics committee) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.”

Patient’s consent not required as patients’ identity are not disclosed or compromised.

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