Inferiorly-Directed Posterior Cranial Vault Distraction for Treatment of Chiari Malformations

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Purpose: Chiari I malformations involve a small posterior fossa and subsequent herniation of the hindbrain, causing altered cerebrospinal fluid flow and intracranial hypertension (Figure 1). Traditional decompressive management uses a posterior fossa craniectomy, which carries limitations and complication risks, including over-correction or the need for re-operation. A large single-stage craniectomy can lead to cerebellar ptosis and new or recurrent symptoms. Distraction of the posterior vault has been effective for relieving intracranial pressure in small case series.[1,2] The authors present the largest series of patients with Chiari I malformations treated with posterior cranial vault distraction osteogenesis. Also introduced is a previously unpublished technique utilizing a vertical distraction vector for vault expansion while mitigating the risks of scaphocephaly and cerebellar ptosis.

Methods: This multi-center study included patients with syndromic and non-syndromic Chiari I malformations treated with vertical-vector distraction osteogenesis of the posterior cranial vault from 2008 to 2014. All patients had follow-up of at least 1 year except one who recently completed the full protocol. Demographics, pre- and post-operative clinical symptoms, and perioperative details were assessed. Postoperative aesthetics, complications, and symptomatic improvement were evaluated with neurosurgery.

Results: Nine patients were identified. Five had known syndromes, two likely had unidentified syndromes, and two were non-syndromic. Seven had prior Chiari-related surgeries. Most presented with hydrocephalus, motor symptoms, and developmental delay. Operatively, internal distraction fixators were applied with the distraction vector following a cephalad-caudad axis. Devices were activated on POD5 and distracted 1 mm/day. Three post-operative complications included a dislodged distraction arm, device extrusion, and local cellulitis. No complications affected the clinical outcome. All patients completed the distraction protocol. Radiographic follow-up showed good bone formation, posterior fossa decompression, improved CSF flow, and no cerebellar ptosis (Figure 2). Neurological surveillance showed improvement in intracranial pressure, hydrocephalus, motor symptoms, and behavioral problems. Importantly, with average follow-up of 3.5 years, no patient required reoperation for persistent or recurrent symptoms.

Conclusion: The authors have presented the largest experience to date of Chiari I malformations treated with distraction osteogenesis, along with a novel technique to safely and effectively expand the posterior fossa while minimizing risk of cerebellar ptosis, scaphocephaly, and reoperations. With excellent functional, aesthetic, and clinical outcomes from this technique, it appears valuable as an initial or secondary operation for symptomatic Chiari I malformations.

References:

1. Steinbacher DM, Skirpan J, Puchała J & Bartlett SP. Expansion of the Posterior Cranial Vault Using Distraction Osteogenesis: Plast. Reconstr. Surg. 127, 792–801 (2011).
2. White N, Evans M, Dover MS, Noons P, Solanki G, Nishikawa H. Posterior calvarial vault expansion using distraction osteogenesis. Childs Nerv Syst. 25, 231–236 (2009).

Figure Legend:

Figure 1. Pre-operative MRI

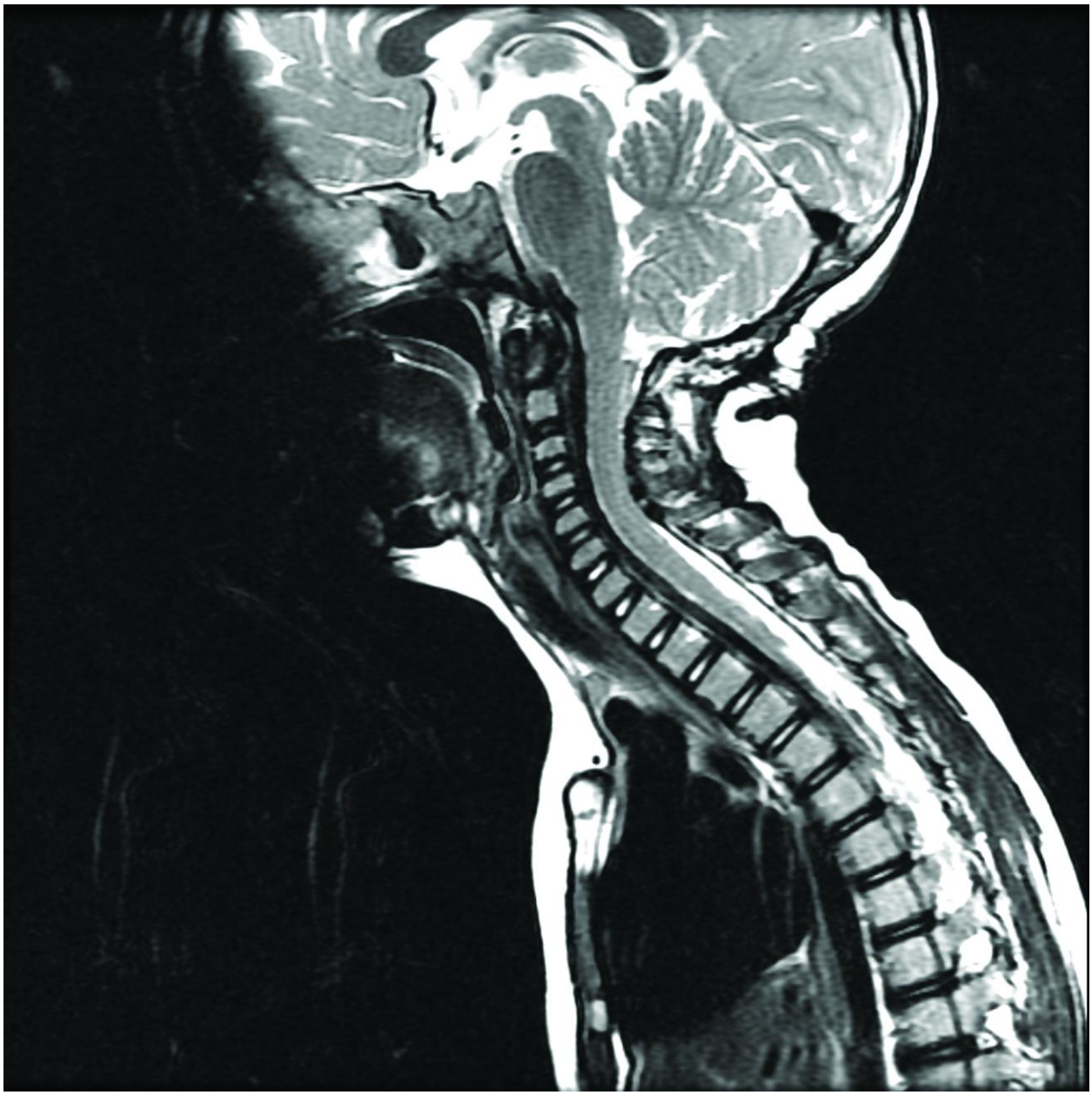

Figure 2. Post-Operative MRI