

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

Objective: The authors present a case series of patients with Chiari I malformations treated with distraction osteogenesis of the posterior cranial vault, utilizing a vertical distraction vector for appropriate cranial vault expansion while mitigating the risks of scaphocephaly and cerebellar ptosis.

Methods: Patients with syndromic and non-syndromic Chiari I malformations treated with vertical-vector distraction osteogenesis of the posterior cranial vault were identified from 2008 to 2014. Demographics, pre- and post-operative clinical symptoms, and perioperative details were assessed. Long-term aesthetic outcomes, complications, and symptomatic improvement were evaluated in conjunction 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, 2-3 internal distraction fixators were applied such that the vector of distraction was along a cephalad-caudad axis. Devices were activated on POD5 and distracted 1 mm/day. Three postoperative complications were found within the first three months which included a dislodged distraction arm, a device extrusion, and a local cellulitis. No complications affected the clinical outcome. Radiographic follow-up showed good bone formation, decompression of the posterior fossa, improved CSF flow, and no cerebellar ptosis. Neurological surveillance showed improvement in intracranial pressure, hydrocephalus, motor symptoms, and behavioral problems.

Conclusion: The authors have presented nine patients with Chiari I malformations treated with distraction osteogenesis, along with a novel technique to safely and effectively expand the posterior fossa while minimizing the risk of cerebellar ptosis.