



"Nuestros niños, nuestros pacientes, nuestra razón de ser"

## TÍTULO: A SYMPTOMATIC CHIARI II MALFORMATION TREATED BY SUPRATENTORIAL POSTERIOR CRANIAL VAULT EXTERNAL DISTRACTION OSTEOGENESIS: THE FIRST CASE REPORT

**IMPORTANCIA DEL CASO:** Although posterior cranial vault distraction osteogenesis has been described as a treatment for complex craniosynostosis, its use as a treatment for Chiari II malformation has never been reported. Surgical options for Chiari II malformation are limited and controversial.

**DESCRIPCIÓN DEL CASO CLÍNICO:** A 7-year-old boy with a lumbar myelomeningocele (MMC) repaired in the first 72 hours of life and with a ventriculo-peritoneal shunt placed few days later was followed since then in the pediatric neurosurgery outpatient clinic. An asymptomatic Chiari II malformation was present since diagnosis. He grew up with a mild global developmental delay, on tailored therapies. When he turned 6 years-old, he started complaining of transient severe occipital headaches associated to vomits. A brain MRI ruled out hydrocephalus and highlighted the Chiari II malformation secondary to a small volume posterior fossa and a low-lying tentorium causing cranio-vertebral junction compression. The surgery was planned after a brain venous CT scan was performed.

A bi-parietal zig zag skin incision was made on a prone position. A circumferential bi-parieto-occipital craniotomy above the torcula was performed and 3 external distractors were uneventfully placed. Three days later, we started a 0.9mm distraction/day. He was discharged on day 5, maintaining the daily distraction at home. On day 21 post op the distractors were removed and 3 titanium plates were implanted while re-ossification of the gap was taking place. He went home the day after. The CT scan showed an increase of 80 cm<sup>3</sup> and of 6.6 cm<sup>3</sup> of total cranial volume and of infratentorial volume, respectively, with cisterns and sulci re-permeabilization, tonsils' ascension and ventricles expansion. On follow-up, 3 and 6 months later, he presented full symptoms resolution.

**LECCIONES APRENDIDAS O IDEAS CLAVE:** A supratentorial posterior cranial vault distraction osteogenesis worked successfully in promoting (1) indirect cranio-vertebral junction decompression and (2) matching in cranio-cerebral proportions in a patient with symptomatic Chiari II malformation.