

Atlanto-occipital membrane bulging without HBH in open spinal dysraphism may predict imminent herniation and guide prenatal therapy

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Introduction

Hindbrain herniation (HBH), a characteristic feature of Chiari II malformation, is present in most fetuses with open spinal dysraphism (OSD) and is a prerequisite for prenatal repair in many protocols. It is thought to arise early in gestation due to cerebrospinal fluid (CSF) leakage and impaired posterior fossa development. Protrusion of the atlanto-occipital membrane (AOM) is frequently observed alongside vermian displacement. After successful prenatal repair, reversal of HBH is reflected by upward migration of the cerebellum, while AOM bulging may persist as a CSF-filled structure. We report on two OSD cases without HBH on initial MRI but with isolated CSF-filled bulging AOM (Fig. A). In both cases, follow-up MRI after 2 weeks showed interval development of HBH to C1, leading to prenatal repair (Fig. B).

Case 1

A 22-year-old primigravida at 22.2 weeks' gestation presented with fetal myelomeningocele (L4), no ventriculomegaly, and preserved motor function (S1). Repeat MRI at 24.3 weeks showed HBH progression to C1. Prenatal repair was performed at 26.0 weeks. HBH resolved postoperatively, while AOM bulging remained. Delivery occurred at 31.5 weeks after PPRM. At 12 months, motor function is unimpaired and there is no need for CSF deviation.

Case 2

A 33-year-old primigravida at 20.3 weeks' gestation presented with fetal myelomeningocele (L5), normal ventricles, and unimpaired motor function (S1). Follow-up MRI confirmed HBH to C1. Prenatal repair was performed at 25.2 weeks. HBH resolved postoperatively, with persistent AOM bulging. Delivery occurred by Caesarean section at 33.1 weeks due to abnormal Doppler findings. At 3 months, motor function was intact and no shunt was required.

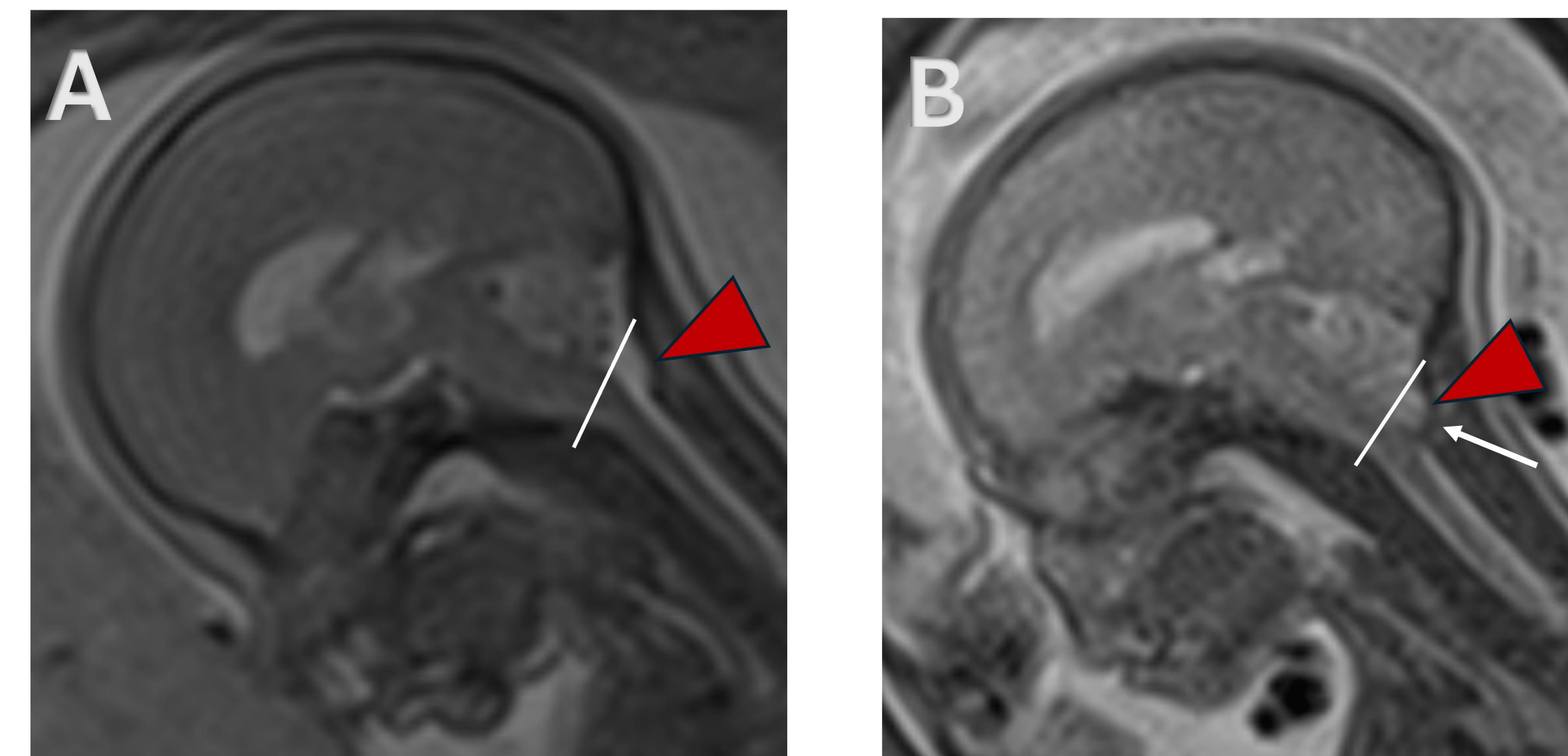


Fig A+B: CSF-filled bulging of the posterior atlanto-occipital membrane without hindbrain herniation at 22.3 weeks (A), followed by development of hindbrain herniation at 24.1 weeks (B). The line indicates the level of the foramen magnum, the arrowhead marks the AOM bulging, and the arrow indicates the hindbrain herniation

Conclusion

We postulate that AOM bulging in the absence of HBH represents an early sign of imminent herniation. Repeat MRI may help to identify cases with evolving HBH in the time window for fetal surgery. This hypothesis warrants prospective validation and may have direct implications for clinical management.