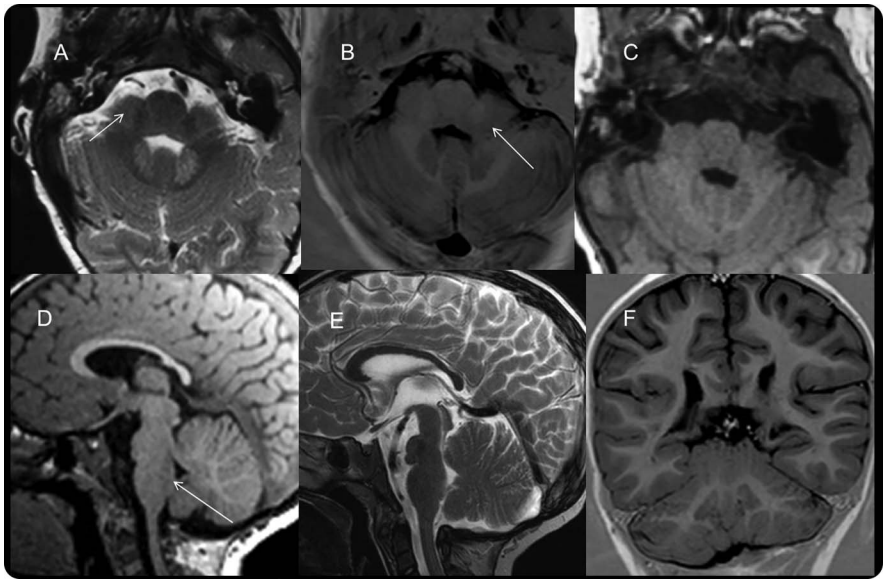
Medullary cap dysplasia

MRI and diffusion tensor imaging of a hindbrain malformation

Figure 1 MRI of the brain

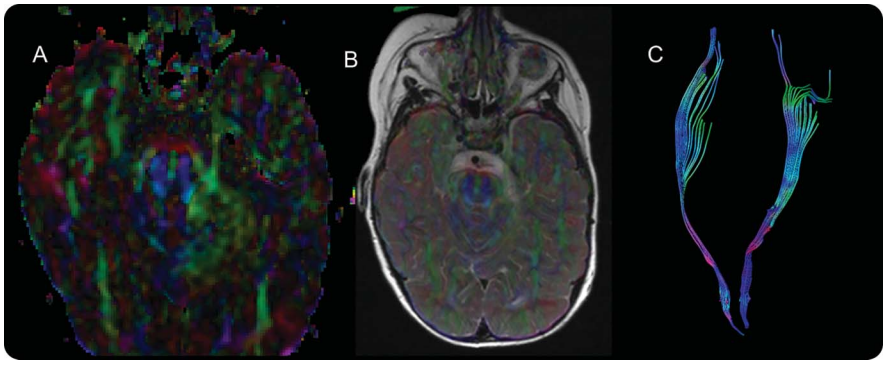


Abnormal rounded cap over the medulla oblongata (arrows) (A–D). Small pons with abnormal anterior outline, thickened medulla oblongata. Oversized and dysplastic cerebellar vermis. Hypogenesis of the splenium of corpus callosum (D, E). Dysplastic upper part of the cerebellar hemispheres (F).

A 3-year-old boy presented with respiratory distress and apnea, discrete dysmorphic features (facial asymmetry, large dysplastic earlobes, deep-set eyes), absence of the middle and distal phalanges of the hands and feet, gastroesophageal reflux, and hiatal hernia. Auditory brainstem response testing (brainstem auditory evoked response) was abnormal bilaterally with more severe changes on the right side.

MRI revealed hypoplastic pons and thickened medulla oblongata with anomalous mass of gray matter signal intensity around it and abnormal signal of the dentate nuclei (figure 1). Diffusion tensor imaging (figure 2) shows lack of pontine fiber crossing in this malformation.[1,2] Supratentorial anomalies included hippocampal

Figure 2 Diffusion tensor imaging and tractography



No decussating pontine fibers—no red spot (A, B) and absence of the crossing of the corticospinal tracts (C).

malrotations and corpus callosum hypogenesis. This malformation has been mentioned previously only by Barkovich and colleagues.[1,2]

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1. Barkovich AJ, Raybaud C. Pediatric Neuroimaging, 5th ed. Philadelphia: Lippincott Williams & Wilkins; 2011:491.
2. Barkovich AJ, Millen KJ, Dobyns WB. A developmental and genetic classification for midbrain-hindbrain malformations. Brain 2009;132:3199–3230.

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