RESIDENT & FELLOW SECTION

Teaching Video NeuroImages: Figure 8 head-shaking stereotypy in rhombencephalosynapsis

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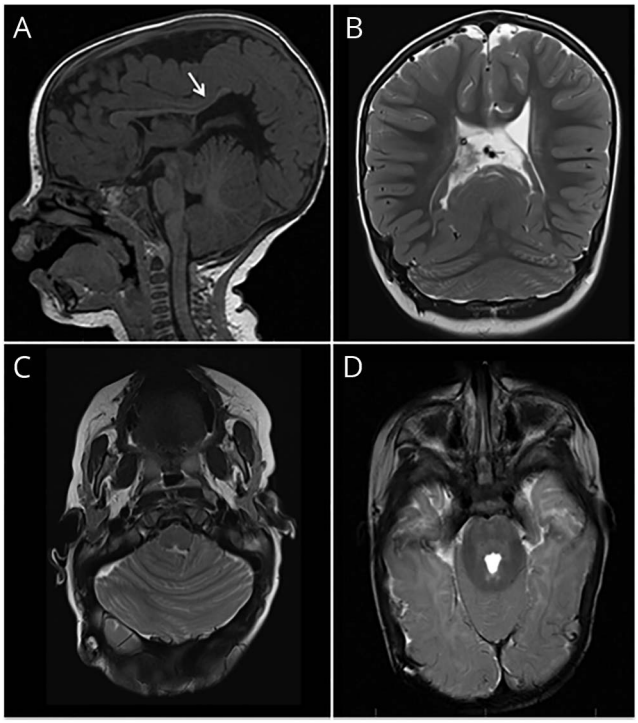
Figure Imaging



Midline sagittal T1-weighted image (A) reveals hemispheric cerebellar lobulation pattern and outstretched posterior corpus callosum (arrow) due to interhemispheric cyst. Coronal (B) and axial (C) T2-weighted images show vermis agenesis and fusion of cerebellar hemispheres with continuity of folia across the midline. Axial T2-weighted image (D) demonstrates a keyhole-shaped 4th ventricle.

A 4-year-old boy had congenital hydrocephalus due to aqueductal stenosis and was treated with a ventriculoperitoneal shunt at birth. Brain MRI revealed absence of the cerebellar vermis with continuity of the cerebellar hemispheres across the midline, consistent with rhombencephalosynapsis (figure).[1] He has mild global developmental delay. Neurologic examination revealed absent nystagmus, oculomotor apraxia, dysmetria, or ataxia. Since the age of 1, he has had stereotyped head movements consisting of rhythmic figure 8 and side-to-side shaking (video). This distinctive stereotypy is reported in 85% of individuals with rhombencephalosynapsis.[2] Its presence should alert clinicians of possible underlying rhombencephalosynapsis or other posterior fossa malformations.

MORE ONLINE

Video

Teaching slides:
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Author contributions

Andrea Accogli: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval, acquisition of data, study

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Disclosure

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