

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

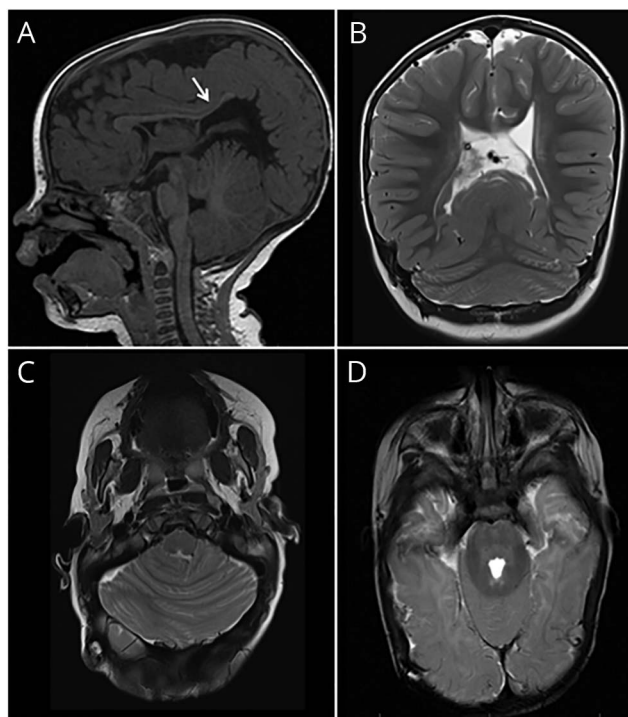
Andrea Accogli, MD, and Myriam Srour, MD, PhD

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**Correspondence**

Dr. Srour  
myriam.sroure@mcgill.ca

## 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 vermian 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).<sup>1</sup> 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.<sup>2</sup> Its presence should alert clinicians of possible underlying rhombencephalosynapsis or other posterior fossa malformations.

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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

From the Departments of Pediatrics (A.A., M.S.) and Neurology and Neurosurgery (M.S.), McGill University, Montreal, Canada; Istituto Giannina Gaslini (A.A.), Genova; and Università degli Studi di Genova (A.A.), Italy.

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supervision. Myriam Srour: drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, study supervision.

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### Disclosure

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