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## MR and Diffusion Tensor Imaging of Isolated Tentorial Hypoplasia

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# MR and diffusion tensor imaging of isolated tentorial hypoplasia

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We report 3 patients (2 adults and 1 child) with isolated tentorial hypoplasia (ITH) from December 2015 to April 2016. Two patients did not have a visual field defect and ITH was an incidental finding on MRI. One patient was a 5-year-old boy with optic atrophy, who was referred by his ophthalmologist for brain MRI to rule out optic pathway mass. A focal defect in the right tentorium with herniation of the right temporal gyrus was found (figure 1, A and B). The second patient was a 43-year-old woman who was imaged for essential tremor. A focal right tentorial defect with herniation of the right cuneus was identified (figure 1C). In both patients, ITH was an incidental finding. The third patient was a 42-year-old woman who presented with blurry vision and found to have left-sided homonymous hemianopia on formal visual field testing (figure 2), with more prominent left homonymous inferior quadrantanopia (figure 3). The optic disc and central vision were normal. Direct ophthalmoscopy did not detect optic nerve hemihypoplasia. Brain MRI demonstrated a moderate-sized defect in the right tentorium and associated herniation of the medial right occipital gyri (figure 1D). Magnetic resonance DTI/tractography, an extension of diffusion-weighted imaging based on the measurement of Brownian motion of water molecules, demonstrated that at least a portion of the white matter extending into the herniated occipital lobe represents some of the visual pathway tracts (figures 1, E and F). The herniated and displaced white matter tracts correspond to the patient's symptomatology based on visual field testing.

## PRACTICAL IMPLICATIONS

Consider isolated tentorial hypoplasia incidentally noted on imaging evaluation as a cause for any associated clinical signs, including visual field defect, and use of diffusion tensor imaging (DTI)/tractography to delineate the optic pathway in patients with isolated tentorial hypoplasia incidentally noted on imaging.

## Discussion

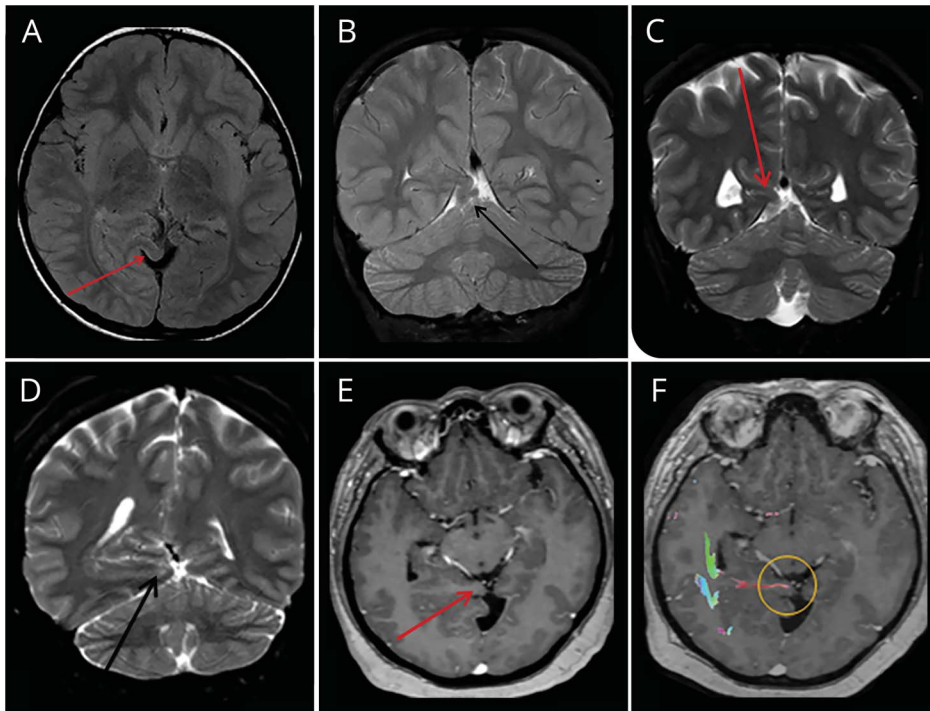
The tentorium cerebelli is the second largest dural reflection in the human body after the falx cerebri, and its main structural function is to separate the supratentorial from infratentorial contents within the cranial vault. A partial or complete defect of the tentorium is often associated with congenital abnormalities, such as Dandy-Walker or Arnold-Chiari II malformations, which were not present in any of these 3 patients. ITH has been widely regarded as a normal developmental variant and a benign or incidental finding on neuroimaging. There is little information on MR imaging findings of ITH. The association of ITH with a resultant concordant neurologic visual field defect is rare.

In 1985, Gund<sup>1</sup> reported the first case of ITH, an 8-year-old girl who was evaluated for an occipital mass. Since then, there have been few isolated case reports of ITH. One report described 3 middle-aged adults with ITH.<sup>2</sup> Cross-sectional imaging showed hypoplasia of the right tentorial leaf in all 3 patients, but the patients' symptoms were not related to the imaging findings. Another study reported a middle-aged woman who presented with vertigo and headache. A partial right occipital lobe herniation secondary to a congenital focal hypoplasia of the tentorium was noted on MRI.<sup>3</sup>

The most accepted hypothesis for the development of ITH is abnormal fusion of the medial and lateral aspects of the tentorium during embryogenesis.<sup>4</sup> It has been suggested that birth trauma

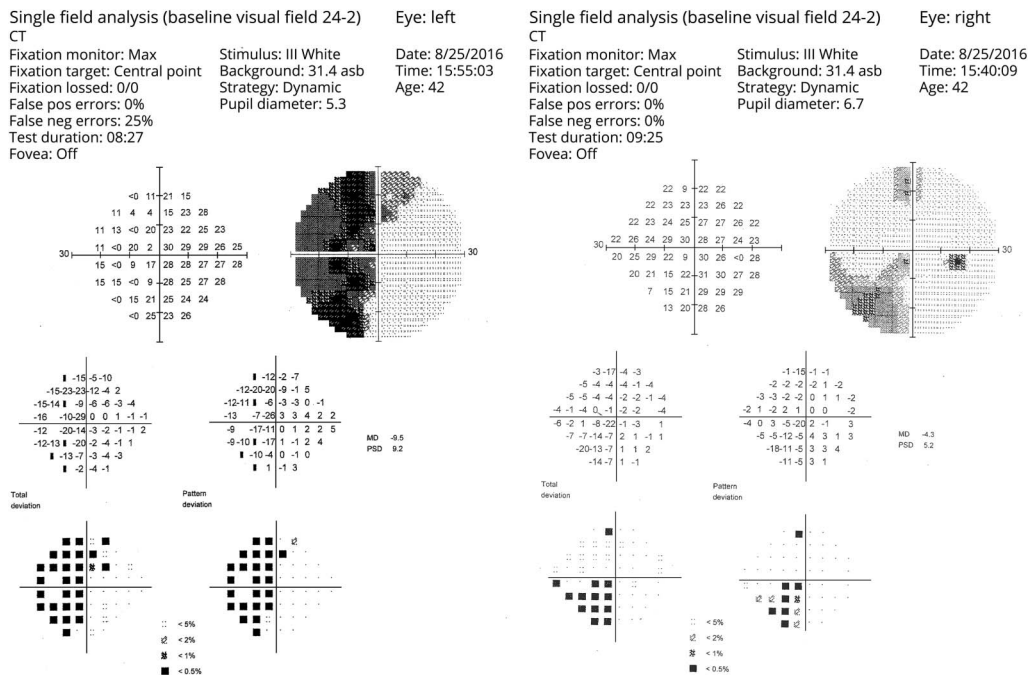
Section of Neuroradiology, Department of Radiology (YS, SB, HM), and Department of Neurology (CK, BA-S, AT), Westchester Medical Center, New York Medical College, Valhalla, NY. Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the authors is available with the full text of this article at [Neurology.org/cp](http://Neurology.org/cp).

**Figure 1** Imaging



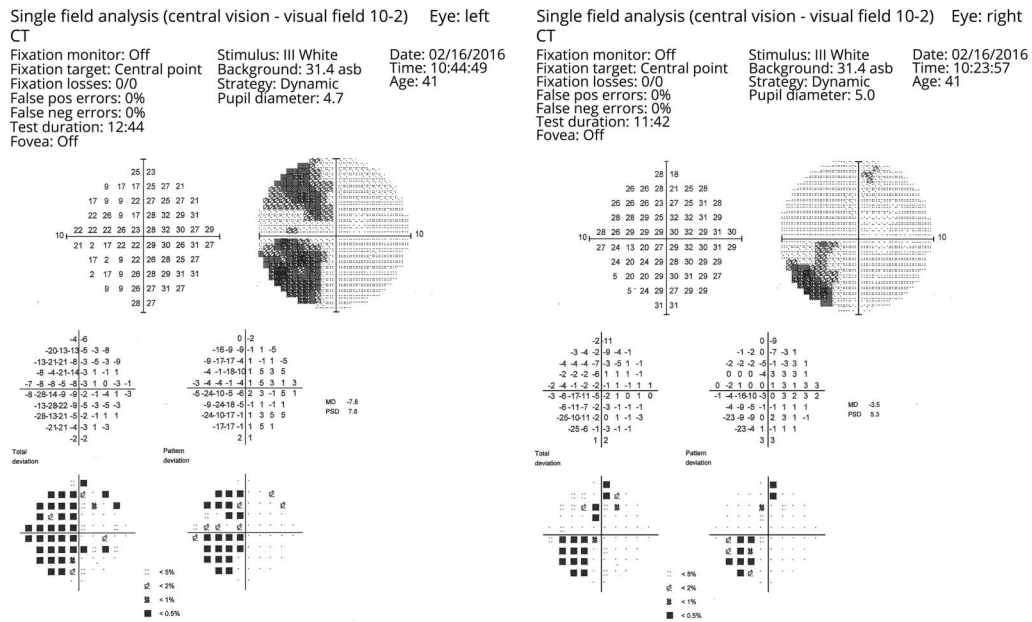
(A) Axial fluid-attenuated inversion recovery sequence of the brain and (B) coronal T2-weighted sequence of the brain of the 5-year-old boy demonstrate a focal defect in the right tentorium with herniation of the right temporal gyrus (red and black arrows, respectively). (C) Coronal T2-weighted sequence of the brain of the 43-year-old woman demonstrates focal defect in the right tentorium (red arrow). (D) Coronal T2-weighted MRI of the brain of the 42-year-old woman demonstrates a moderate-sized defect in the right tentorium and herniation of the right occipital lobe gyri (black arrow). (E) Axial T1-weighted postcontrast sequence and (F) axial diffusion tensor imaging (DTI) sequence of the MRI of the brain in the same patient reveals the right cuneus herniation through the right tentorial defect as indicated by the red arrow with corresponding optic pathway fibers within the herniated white matter on DTI sequence (yellow circle).

**Figure 2** 24-2 Visual field testing



24-2 Visual field testing of the left and right eyes demonstrate left-sided homonymous hemianopia.

**Figure 3** 10-2 Visual field testing



10-2 Visual field testing of the left and right eyes, which is better at assessing central vision more accurately, demonstrates more prominent left homonymous inferior quadrantanopia.

and other perinatal insults may play a role in the process, but the exact mechanism is unclear. Of interest, our 3 patients, and all the patients described here in other case reports, demonstrate right-sided ITH, an observation of unclear etiology. However, this finding may not be purely coincidental. Random events such as trauma or infection, as suggested by other authors, may be less likely to be the cause, although the number of reported cases is small.

ITH is often viewed as a benign or incidental finding on neuroimaging without known associated clinical symptoms, which is supported by the few case reports cited earlier and 2 of our 3 patients. However, our third patient demonstrated clinical findings of left-sided homonymous hemianopia, which corresponds anatomically to the MRI findings of ITH. DTI/tractography provides MRI evidence that a portion of the optic pathway white matter tracts is included in the right occipital lobe herniation. This case suggests that ITH may not be an incidental finding in this patient. Although there is no consensus on direct management of ITH, it is important to raise awareness of ITH as being potentially symptomatic. ITH can be easily detected on MRI or CT, as the tentorium is visible on 99% of contrast-enhanced cerebral CT studies<sup>5</sup> and virtually 100% of brain MRI examinations.

### Author contributions

Y. Sun: study concept and design, acquisition of data, analysis and interpretation of data, generation of manuscript. S. Bobra:

study concept and design, analysis and interpretation of data, critical revision of manuscript. C. Kurian: acquisition of data, analysis and interpretation of data. B. Ahluwalia-Singh: interpretation of data. Anila Thomas: interpretation of data. Hasit Mehta: corresponding author, study concept and design, critical revision of manuscript.

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### Disclosure

Y. Sun, S. Bobra, C. Kurian, B. Ahluwalia-Singh, and A. Thomas report no disclosures. H. Mehta performs neuroimaging as primary clinical practice (Westchester Medical Center MRI Brain with DTI [5% effort]). Full disclosure form information provided by the authors is available with the full text of this article at [Neurology.org/cp](http://Neurology.org/cp).

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