Nondecussating retinal-fugal fiber syndrome (achiasmatic syndrome)

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Figure Achiasmatic syndrome

Axial T2-weighted (A) and postcontrast T1-weighted (B) images through the optic nerves depict failure of optic nerve fibers to decussate at the chiasm before continuing as optic tract (black and white arrows), a characteristic of achiasmatic syndrome. Coronal (C) and sagittal (D) T2-weighted images through the sella demonstrate absence of optic chiasm (black arrow: optic tract, white arrowhead and arrow: pituitary gland).

A 9-week-old girl presented with poor visual responsiveness and congenital seesaw nystagmus. Congenital seesaw nystagmus usually implies a structural anomaly, such as achiasma, and warrants evaluation with MRI (figure).¹ Isolated absence of optic chiasm is extremely uncommon.² In nondecussating retinal-fugal fiber syndrome (achiasmatic syndrome), there is failure of decussation of nasal retinal fibers, with the misrouted nasal fibers projecting and misaligning with projections from the temporal retina of the same eye, leading to reversal of the left and right half of visual field coordinates (“mirror reversal”).¹ Mutation in a homeobox gene may underlie this aberration in pathfinding of retinal axons seen in achiasma.²

Author contributions

Rahul Nikam: critical revision for intellectual content. Vinay Kandula: supervision. Achala Donuru: concept and design.

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