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

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References
Nondecussating retinal-fugal fiber syndrome (achiasmatic syndrome)
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