Teaching Video Neurolmage: Acquired or congenital gaze-evoked nystagmus?

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A 40-year-old female migraineur reported intermittent unsteadiness with headache. On examination, she had gaze-evoked horizontal-torsional nystagmus that was also evident during pursuit (video). Nystagmus recordings showed increasing-velocity slow phases typical for congenital nystagmus (CN) (figure 1). A relative subsequently confirmed that the nystagmus was long-standing.

While usually isolated, CN can be associated with visual system abnormalities or strabismus.¹ It is usually conjugate, horizontal with a subtle torsional component,² and minimized in a certain “null” position. Eye movement recordings help differentiate it from other nystagmus types (figure 2).¹ Confirmation that it is long-standing supports the diagnosis, helping to avoid unnecessary investigations.

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Figure 2 Typical nystagmus waveforms

(A) Increasing-velocity slow phase waveform typical of congenital nystagmus. (B) Decreasing-velocity slow phase waveform typical of cerebellar gaze-evoked nystagmus. (C) Constant-velocity slow phase waveform typical of vestibular nystagmus. Position of fixation target (Fix) is indicated by arrowheads. While the search-coil technique is the current gold standard, satisfactory eye movement recordings can be obtained using electro-oculography, infrared-, or video-based techniques, often through a neuro-otology or otolaryngology service.

Figure 1 Three-dimensional search-coil recordings of the patient’s nystagmus

The patient is attempting to fixate leftward (A), central (B), and rightward (C) targets (Fix), indicated by arrowheads next to the horizontal traces. The traces show leftward-upward-counterclockwise jerk nystagmus (A), alternating-direction horizontal-torsional jerk nystagmus with extended foveation (B), and rightward-upward-clockwise jerk nystagmus with extended foveation (C). Each nystagmus slow phase increases in velocity with time. Positive directions are rightward, upward, and clockwise, as indicated.

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