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Visual Diagnosis

Rhythmic Sawtooth Electroencephalograph Waveforms in Neonatal Hypoxic-Ischemic/Hypoglycemic Encephalopathy

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This baby was born at term to a primigravida group B streptococcus-positive mother. He had hypoxic-ischemic encephalopathy (cord pH 7.02) and hypoglycemia (blood glucose 0.8 mM/L). The evaluation for sepsis was negative. He had a Sarnat score of two and underwent total body cooling and rewarming on day four. Focal seizures were identified in the first days of life and required multiple medications to control. He was monitored with continuous electroencephalography, which on day six revealed previously unrecognized intermittent upbeat jerk nystagmus (Fig 1). Brain magnetic resonance imaging (on days one and nine) showed posterior-predominant diffuse brain injury, consistent with perinatal insults from both hypoglycemia and hypoxia/ischemia (Fig 2).

This child's abnormal eye movements were suspected to reflect injury to vestibular structures, but it is important to note that self-limited upbeat nystagmus may occur in healthy neonates due to immature vestibular pathways.[1] In individuals with pathological upbeat nystagmus, the underlying cause is often cerebellar/pontomedullary abnormalities or drug toxicity (e.g. organophosphates).[1] The differential diagnosis of vertical nystagmus also includes ophthalmologic pathologies. In an eight-year longitudinal study following 276 children presenting with infantile-onset nystagmus (birth to six months of age), 37 had vertical nystagmus; 11 of these children were found to have retinal disease, albinism or congenital idiopathic nystagmus.[1] In a separate study, three patients with congenital upbeat nystagmus that converted to a horizontal nystagmus before one year of age were subsequently diagnosed with Leber's amaurosis.[2]

We present a baby with widespread brain damage from hypoglycemic and hypoxic/ischemic insults who

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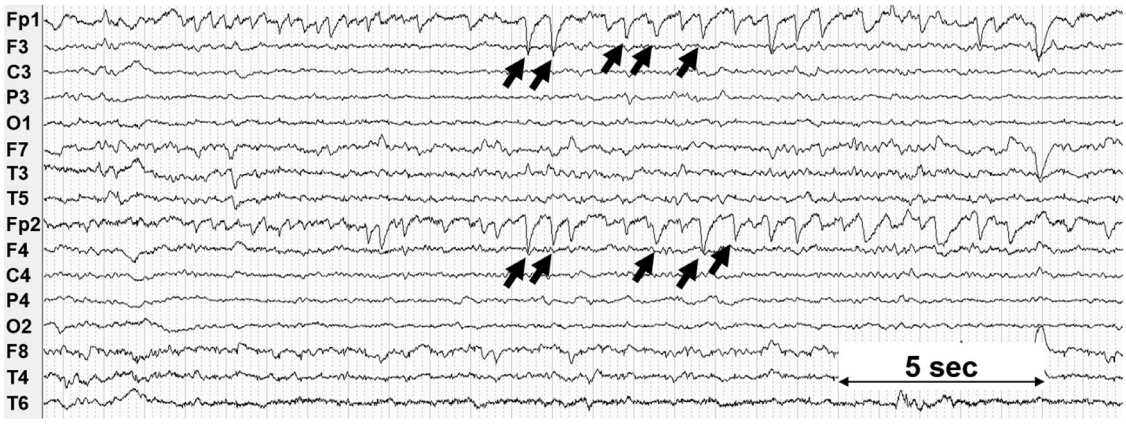

FIGURE 1. Upbeat nystagmus on EEG. EEG (average montage with sensitivity 10 μV/mm) shows symmetric sawtooth waveforms (arrows) over the frontal polar electrodes. The positive (downward) EEG deflection has steep incline, reflecting the fast phase upward of upbeat jerk nystagmus. Frequency is variable, typically from 1.5 to 2.5 Hz. EEG, electroencephalography.

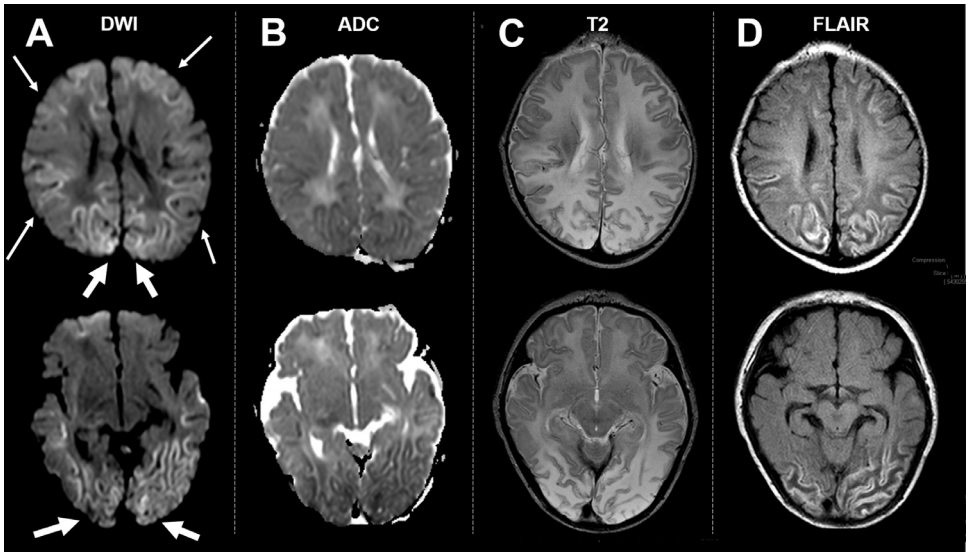

FIGURE 2. Combined effects of hypoxia-ischemia and hypoglycemia on MRI. (A, B) Day one: axial brain MRI (diffusion weighted imaging (DWI), apparent diffusion coefficient (ADC)) shows cytotoxic edema following (but not limited to) watershed distribution suggesting hypoxic-ischemic injury (thin arrows). Superimposed bilateral occipital injury suggests hypoglycemic injury as well (thick arrows). (C, D) Day nine: axial T2 and FLAIR show evolution of findings, consistent with cortical laminar necrosis.

exhibited intermittent upbeat nystagmus. This finding has a broad array of potential causes, including both brain and ophthalmologic disorders. Our child demonstrates that electroencephalography can identify ophthalmologic abnormalities in neonates whose diagnosis might otherwise be missed simply because young babies have their eyes closed the majority of the time.

References

[1] Shawkat FS, Kriss A, Thompson D, Russell-Eggitt I, Taylor D, Harris C. Vertical or asymmetric nystagmus need not imply neurological disease. *Br J Ophthalmol*. 2000;84:175–180.

[2] Hoyt CS, Gelbart SS. Vertical nystagmus in infants with congenital ocular abnormalities. *Ophthalmic Paediatr Genet*. 1984;4:155–161.