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50 Years Ago in *The Journal of Pediatrics*

The Cause of Spasmus Nutans and Congenital Nystagmus: Frozen in Time

Jayalakshmi P, McNair Scott T, Tucker H, Schaffer D. Infantile nystagmus: a prospective study of spasmus nutans, congenital nystagmus, and unclassified nystagmus of infancy. J Pediatr 1970;77:177-87.

Jayalakshmi et al presented the largest prospective study of infants with nystagmus grouped by the contemporaneous criteria of spasmus nutans and congenital nystagmus. After years of study of wide-reaching environmental, maternal health, maternal–child relationship, prenatal and perinatal complications, and other neurologic associations, the authors determined that the cause was likely of neurologic basis from "organic cerebral dysfunction."

Now 50 years later, the type of nystagmus and associated features are better defined, but the causes of these disorders remain frozen in time. Spasmus nutans is typically a benign idiopathic self-limiting disorder characterized by a triad of asymmetric shimmering nystagmus with compensatory head bobbing and torticollis. Rare associations have been recognized with suprasellar chiasmal lesions, metabolic disorders, and retinal dystrophies, with current recommendations for a thorough eye exam and neuroimaging for all infants with spasmus nutans. The broad term, congenital nystagmus, is now termed infantile nystagmus syndrome (INS), classified as either with visual sensory deficits (INS secondary to the cause of the sensory deficit, previously congenital sensory nystagmus) or without visual sensory deficits (infantile idiopathic nystagmus [IIN], previously congenital motor nystagmus). INS is a bilateral, conjugate jerk or pendular nystagmus that remains horizontal in up-and-down gaze. Although both INS and spasmus nutans share features of head nodding and torticollis, INS has greater consistency and symmetry in movement than spasmus nutans.

The dysfunctional neurologic pathways for both conditions remain speculative, however, interesting environmental associations (eg, nonwhite ethnicity, low home luminance, low socioeconomic status) have been better defined in spasmus nutans,[1] and a pathological genetic variant in *FRMD7* has been identified in familial cases of INS. There is no effective treatment for nystagmus in infants with spasmus nutans or INS, and any advancements in therapeutics likely will only be possible by answering the question posed more than 50 years ago: what is the "organic cerebral dysfunction"?

Shannon Beres, MD
Department of Neurology & Ophthalmology
Lucile Packard Children's Hospital at Stanford University
Stanford, California

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