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3.01 The usefulness of visual electrophysiology in pendular nystagmus in infancy

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Purpose: Strictly pendular nystagmus in infancy, partially or completely overlapping with so-called spasmus nutans-type nystagmus, has long been considered a benign entity. It is now a well-recognised symptom of various neurological or retinal diseases. In this study we looked at the place and usefulness of visual electrophysiology in the etiological work-up of pendular nystagmus in infancy.

Methods: All cases of infants presenting to our institution nystagmus clinic with a strictly pendular nystagmus between 2010 and 2016 were included. Cerebral imaging and visual electrophysiology were performed. Infants with a causal diagnosis known before the first nystagmus clinic appointment were excluded.

Results: Fifty infants (34 boys, median age at nystagmus onset: 6 months) were included. MRI showed a large chiasmal glioma in 10 cases (20%), a leukoencephalopathy in five cases (10%), and a significant malformation in three cases (6%). Visual electrophysiology was obtained in 44 cases (88%). ISCEV full-field ERG allowed diagnosis of 19 cases (38%) with retinal dysfunction: early-onset severe retinal dystrophy (nine cases, two of which also exhibited dysmyelination), stationary cone dysfunction (eight cases), and congenital stationary night blindness (two cases).

Conclusions: In 70% of cases, pendular nystagmus was associated with a retinal or neurological disorder. Visual electrophysiology was the key exam for diagnosing a retinal disorder. Unless a chiasmal glioma is found on MRI, a full-field ERG should be part of the work-up of pendular nystagmus in an infant, including in the presence of leukoencephalopathy.