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British Journal of Ophthalmology 2008;92:236-240; doi:10.1136/bjo.2007.126342

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ORIGINAL ARTICLES

Nystagmus characteristics in congenital stationary night blindness (CSNB)

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Aim: To analyse nystagmus characteristics in patients with congenital stationary night blindness (CSNB) for differentiation from other forms of early childhood nystagmus.

Methods: Horizontal and vertical eye movements of 10 patients (6–46 years, mean 17.1 years, median 12.5 years) with CSNB (eight with CSNB1, two with CSNB2) were recorded with the scleral magnetic search coil technique or by electro-oculography. Nystagmus characteristics such as the amplitude, frequency, conjugacy and intermittency were analysed.

Results: All patients had continuous, pendular, oblique and mostly dysconjugate nystagmus of high frequency and low amplitude. In seven cases, a large horizontal or vertical jerk nystagmus with increasing, decreasing or constant velocity was superimposed. Jerk nystagmus was mostly intermittent and conjugate. Head nodding was found not to be compensatory.

Conclusions: Eye-movement recordings of CSNB patients disclosed specific nystagmus characteristics, such as an oblique direction, superimposed waveforms and dysconjugate eye movements. These features may help to distinct nystagmus in CSNB from other forms of early infancy nystagmus, such as congenital idiopathic nystagmus, latent nystagmus and spasmus nutans. We found nystagmus in CSNB to be similar to the nystagmus reported in blue-cone monochromatism and rod monochromatism, and in patients with a severe sensory defect. The nystagmus characteristics described should prompt electroretinographic investigation in cases of uncertain diagnosis.

Competing interests: None.

Funding: The study was performed at the Department of Strabismus and Neuroophthalmology, Kantonsspital St. Gallen, Switzerland and supported by the Swiss National Science Foundation (no. 32-52503.97) and by the OPOS Foundation, with no propriety interests.

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