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Neuro-Ophthalmological Findings in Children and Adolescents with Chronic Ataxia.

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Abstract

Chronic ataxia is a challenging problem in paediatric neurology. It is caused by a multitude of disorders that at least initially have similar or non-specific phenotype. Some of these disorders have associated neuro-ophthalmological signs (N-OS). The aims of this study are to describe the N-OS and their frequencies in general and by disease aetiology in paediatric patients with chronic ataxia. The authors identified 184 patients under age 17 years with chronic ataxia (>2 months duration or recurrent) during 1991-2008 from multiple sources. Diagnoses and N-OS were ascertained following charts review. Mean age (SD) was 15 (7.7) years. Median duration of follow-up was 6.4 years. There were 214 N-OS in 115 patients (median = 2, range = 1-5 N-OS/patient). Strabismus was present in 29.3% of patients, nystagmus 27.7%, impaired smooth pursuit 23.4%, hypometric saccades 10.3%, decreased visual acuity 9.2%, abnormal optic discs 8.7%, abnormal pupillary examination 2.7%, hypermetric saccades 2.2%, impaired ductions 1.6%, and abnormal visual fields in 1.1% of patients. N-OS were reported most commonly among patients with the following disorders (commonest N-OS): hypoxic-ischaemic encephalopathy following birth (strabismus), episodic ataxia (nystagmus), neuronal ceroid lipofuscinosis (abnormal optic discs), neuronal migration disorder (strabismus), ischaemic stroke (nystagmus), Joubert syndrome-related disorders (strabismus), leukodystrophy (nystagmus), Friedreich ataxia (hypometric saccades, impaired smooth pursuit, nystagmus), mitochondrial disease (strabismus, nystagmus), ataxia telangiectasia (impaired smooth pursuit), and Angelman syndrome (strabismus). N-OS occur commonly in children with chronic ataxia. Although non-specific, they vary with disease aetiology, potentially aiding in the assessment of these patients.

KEYWORDS:
Chronic ataxia; neuro-ophthalmology; paediatrics