

Neuro-Ophthalmic Signs in Children with Ataxia

Causes of Ataxia

- Hypoxic-Ischemia encephalopathy
- Episodic ataxia
- Neuronal ceroid lipofuscinosis
- [Joubert syndrome](#)
- Neuronal migration disorder
- Ischemic stroke
- Friedreich ataxia
- Mitochondrial Disease
- Severe epilepsy
- Ataxia telangiectasia
- [Angelman syndrome](#)

Common Associations

Neuronal-ophthalmic Finding	Frequency
Afferent	
Decreased visual acuity	9%
Abnormal optic discs	9%
Abnormal pupils	3%
Abnormal Visual Fields	1%
Efferent	
Strabismus	30%
Pathologic nysagmus	28%
Gaze evoked nystagmus	15%
Impaired smooth pursuit	23%
Hypometric saccades	10%
Hypermetric saccades	2%
Impaired Ductions	2%
Ataxia Disorder	Most Common Association
Hypoxic-Ischemic encephalopathy	Strabismus
Episodic ataxia	Nystagmus
Neuronal ceroid lipofuscinosis	Abnormal Optic discs
Joubert Syndrome	Strabismus
Neuronal Migration Disorder	Strabismus
Ischemic Stroke	Nystagmus
Friedreich Ataxia	Impaired Saccades

Ataxia Disorder	Most Common Association
Mitochondrial Disease	Strabismus/Nystagmus
Severe epilepsy	Decreased Visual acuity
Ataxia Telangiectasia	Impaired smooth pursuit
Angelman Syndrome	Strabismus

- Adapted from Salman et. al. Neuroophthalmology. 2015;39(3):125-131

References

- Neuro-Ophthalmological Findings in Children and Adolescents with Chronic Ataxia. Salman MS. and Chodirker BN. Neuroophthalmology. 2015;39(3):125-131

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