

Optic Nerve Hypoplasia¹

What is the optic nerve? The optic nerve is a collection of more than a million nerve fibers that transmit visual signals from the eye to the brain [See Figure 1]. The optic nerve develops the first trimester of intrauterine life.



Fig. 1: Normal optic nerve.

What is optic nerve hypoplasia? Optic nerve hypoplasia (ONH) is a congenital condition in which the optic nerve is underdeveloped (small) [See figure 2].

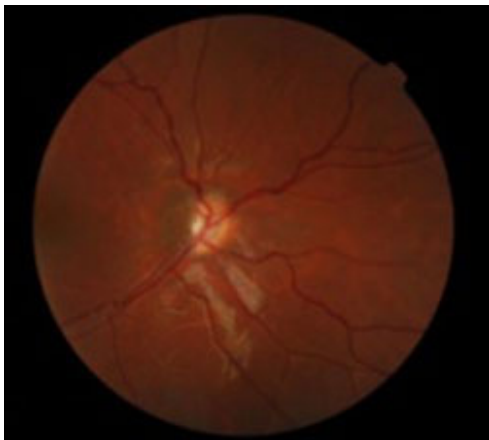


Fig. 2: Optic nerve hypoplasia.

¹ <https://aapos.org/browse/glossary/entry?GlossaryKey=56af0bdd-d0b6-4108-a9d3-5b06b3c26335>

How is optic nerve hypoplasia diagnosed? The diagnosis of ONH is typically made by the appearance of small/pale optic nerve on ophthalmoscopic examination of the eye. It is difficult to predict visual acuity potential on the basis of optic nerve appearance.

What causes optic nerve hypoplasia? Most cases of ONH have no clearly identifiable cause. There are no known racial or socioeconomic factors in the development of ONH, nor is there a known association with exposure to pesticides.

What visual problems are associated with optic nerve hypoplasia? Vision impairment from ONH ranges from mild to severe and may affect one or both eyes. [Nystagmus](#) (shaking of the eyes) may be seen with both unilateral and bilateral cases. The incidence of [strabismus](#) is increased with ONH.

Is optic nerve hypoplasia associated with non-visual problems? Optic nerve hypoplasia can be associated with central nervous system (CNS) malformations which put the patient at risk for other problems, including seizure disorder and developmental delay. Hormone deficiencies occur in most children, regardless of associated midline brain abnormalities or pituitary gland abnormalities on MRI. In fact, most children with growth hormone deficiency have a normal MRI.

What tests should be done for children with optic nerve hypoplasia? An MRI scan is indicated for all children with optic nerve hypoplasia. Evaluation by an endocrinologist should be mandatory for all patients under five years of age.

Does optic nerve hypoplasia get worse over time? In general, ONH is a stable and nonprogressive condition which does not deteriorate. Vision may improve slightly and nystagmus may decrease over time.

Is there any treatment for optic nerve hypoplasia? There is no medical or surgical treatment for ONH. However, occlusion of the better seeing eye may improve vision in the other eye. Children with significant vision loss in both eyes may benefit from early supportive attention by low vision specialists. Stem cell treatment has not been shown to be effective for ONH.

More information regarding septo-optic dysplasia

- [National Institute of Neurological Disorders and Stroke](#)

Links to Articles on Stem Cell Therapy for Optic Nerve Hypoplasia and Other Brain Disorders

- Fink C, Garcia-Filion P, Borchert M. "Failure of stem cell therapy to improve visual acuity in children with optic nerve hypoplasia." J AAPOS. 2013 Oct; 17 (5):490-3. [St. Louis](#)



[Children's Hospital Press Release](#), February 18, 2008, "Stem cell treatment for children with eye nerve disease called 'medical hoax'".

- Ophthalmology Times, September 1, 2008 issue, Industry News, "[Parents warned about 'medical hoax' surrounding optic nerve hypoplasia](#)". (Ophthalmology Times E-News, August 1, 2008).
- EyeWorld, January 2009, [Sounding the alarm, stem cell treatment scam](#). Maxine Lipner, Senior EyeWorld Contributing Editor.
- 60 Minutes, September 15, 2010, "[21st Century Snake Oil](#)". (Consultant)
- Tychsen L, Lueder G, Stahl, E, Giangiacoimo J: [Stem cell therapy does not improve visual function or structure in children with optic nerve hypoplasia \(septo-optic dysplasia\)](#). 2010 AAPOS Abstract #034. J AAPOS 2010;14(1) e9

