

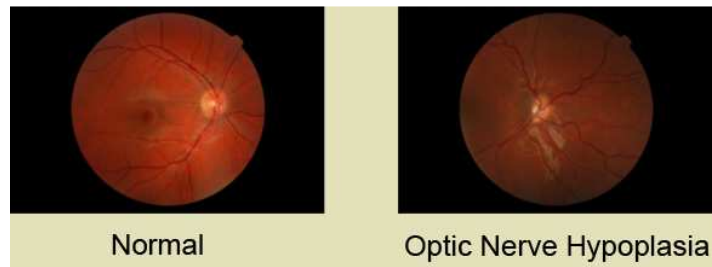
# Optic Nerve Hypoplasia

## What is the optic nerve?

The optic nerve is a collection of thousands of nerve fibers that transmit visual signals from the eye to the brain. The optic nerve develops and grows during intrauterine life.

## What is optic nerve hypoplasia?

Optic nerve hypoplasia (ONH) is a condition present at birth in which the optic nerve is underdeveloped (small) [See figure 1].



**Fig. 1** The optic nerve is underdeveloped in optic nerve hypoplasia.

## 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. An MRI scan is sometimes utilized to confirm the diagnosis. It is difficult to predict visual acuity potential on the basis of optic nerve appearance.

## What causes optic nerve hypoplasia?

Although optic nerve hypoplasia has been associated with maternal diabetes, maternal alcohol and drug abuse, maternal use of anti-epileptic drugs, and young maternal age (20 years of age or less), 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 noted when both eyes are involved. The incidence of strabismus is increased with ONH.

## **Is optic nerve hypoplasia associated with non-visual problems?**

Optic nerve hypoplasia can be associated with several developmental brain abnormalities, most commonly midline brain structures including the pituitary gland (septo-optic dysplasia or deMorsier syndrome). If the pituitary gland is abnormal, growth hormone deficiency may result in delayed growth and development of a child.

## **What tests should be done for children with optic nerve hypoplasia?**

An MRI scan is sometimes performed to evaluate brain and/or optic nerve structure. Evaluation by an endocrinologist (including blood hormone levels) may be indicated.

## **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 over the first few years of life as the result of brain maturation. 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.

## **Where can I find more information regarding septo-optic dysplasia?**

National Institute of Neurological Disorders and Stroke  
[http://www.ninds.nih.gov/disorders/septo\\_optic\\_dysplasia/septo\\_optic\\_dysplasia.htm](http://www.ninds.nih.gov/disorders/septo_optic_dysplasia/septo_optic_dysplasia.htm)

Copyright ©2009 AAPOS. All rights reserved.