What is the optic nerve?
The optic nerve is a collection of over one million nerve fibres that transmit visual signals from the eye to the brain. The optic nerve develops and grows as the baby develops in the womb.

What is optic nerve hypoplasia (ONH)?
Optic nerve hypoplasia (ONH) is present from birth; the optic nerve in one or both eyes is too small and does not have enough nerve fibres to give normal vision.

How is optic nerve hypoplasia diagnosed?
The diagnosis of ONH is made in clinic by looking into the eyes with an ophthalmoscope. An MRI (brain scan) is sometimes used to confirm the diagnosis.

What causes optic nerve hypoplasia?
Most times there is no reason why ONH develops. It is thought to be more common when the mother is young. It is also thought to be more common when the mother has diabetes, alcohol or drug use during pregnancy. The drugs might be “recreational” (such as cocaine) or medical (such as medicines for epilepsy). In the past, there have been various theories about the cause, such as exposure to pesticides, but these are no longer thought to be correct. It is extremely unusual for ONH to be passed on by genes (inherited) from one member of the family to another.

What is the effect on vision?
The vision can be very different from one child to another. There can be near-normal vision through to severe loss of sight in one or both eyes. Nystagmus (wobbling of the eyes) is often the first sign the vision is not perfect. The reduced vision can lead to a squint (one eye turning in or out).

Are there other problems?
Sometimes children with optic nerve hypoplasia (ONH) can have other health problems that might affect their general development including brain and hormone changes. The condition is called septo- optic dysplasia or deMorsier syndrome. If the pituitary gland is underdeveloped then children often need to take medicines to boost reduced hormones, and prevent effects on their growth and development.

What tests should be done for children with optic nerve hypoplasia?
An appointment is made to meet a paediatrician who specialises in hormone problems (a paediatric endocrinologist).

Does optic nerve hypoplasia get worse over time?
No. ONH is a stable condition which does not worsen with time. Children’s vision seems to improve slightly over the first few years of life as the brain matures. The nystagmus may become less obvious over time.

Is there any treatment for optic nerve hypoplasia?
There is no medical or surgical treatment for ONH. However, patching treatment of the better seeing eye is used sometimes to help the vision in the eye with ONH. Children with significant vision loss in both eyes will be helped by careful supervision and adjustment of their education. Stem cell treatment does not help children with optic nerve hypoplasia.
Where can I find more information about 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:


OPTIC NERVE HYPOPLASIA (ONH)

This leaflet has been produced for parents and doctors by the Eye Department, December 2013.

Further Information
We hope this leaflet will help you to understand the treatment offered to your child. If you feel you need more information or have any concerns please contact the Eye Department on 0121 333 9462.
Further information and internet access is available in the Child and Family Information Centre. This is on the Ground Floor of the hospital near the main Reception Desk.