WHAT IS OPTIC NERVE HYPOPLASIA (ONH)?

Optic Nerve Hypoplasia (ONH) refers to a congenital condition in which the optic nerve does not fully develop during pregnancy. The optic nerve is made of about one million optic nerve fibers that carry information from the eye to the brain. The various fibers are responsible for different parts of the field of vision (i.e. central versus peripheral). For people with ONH, optic fibers may be under-developed or completely non-existent. The term “hypoplasia” itself means under-development. As a result of these missing fibers, adequate information does not reach the brain for processing.

CHARACTERISTICS OF ONH

ONH has variable characteristics that are dependent on the size and abnormality of the hypoplastic (under-developed) nerve. Patients with ONH, for example, may have lack of visual fibers in areas causing some mild losses in visual acuity (the ability to distinguish object details and shape) or, in more severe cases, may have no light perception at all (total blindness).

ONH may affect the visual field in varying degrees as well, ranging from a general loss in both central (main) and peripheral (side) fields to subtle or slightly detectable peripheral loss.

Optic Nerve Hypoplasia may affect either one or both eyes. In cases where both eyes are affected, the degree may vary, again, dependent on the severity of the nerve fibers’ under-development.

ONH is a stable condition. Visual function does not deteriorate over the course of time. It also does not affect perception of color.

ASSOCIATED CONDITIONS

Many people with Optic Nerve Hypoplasia have abnormalities of the brain. These abnormalities may include how the brain is formed (brain structure) and how the brain works (brain function). The areas are those along the “midline” and are physically near the optic nerve.

When ONH patients have malformed or nonexistent brain structures, a host of medical issues may ensue. The major areas which can be involved are the use of large muscle (gross motor) and small muscle (fine motor), intelligence and learning, speech, and interacting with people. Hormone-producing areas of the brain may be affected such as the Pituitary gland, which is known as the “master control gland” due to its role in producing important hormones (such as human growth hormone) and its direction to other glands (such as the thyroid) to produce hormones.

Other affects may include difficulty regulating body temperature and blood sugar.
A high percentage of people with ONH have associated involuntary rhythmic movements of the eye called nystagmus. This usually results in a significant reduced visual acuity in both eyes.

Some mild photophobia (light sensitivity) may occur in people with ONH.

Additionally, ONH has been associated with amblyopia (commonly known as “lazy eye”). This condition can further reduce vision if one eye has better acuity than the other.

Many with ONH have a sensory integration problem. This means their bodies do not process information they receive from their senses properly. These individuals may demonstrate Austistic behaviors and characteristics, including sensitivity to textures, sights and sounds. Other stereo-typed behaviors may also exist, including physical movements and impaired social interaction.

TREATMENT AND IMPLICATIONS

While there are no current treatments for the condition of ONH itself, there are several common strategies for teachers working for students with ONH and its related conditions.

Early intervention to provide stimulation and adaptive learning may minimize the impact of the vision loss on overall development.

Increasing size, contrast and lighting of materials will assist with visual loss as well as nystagmus.

A child with ONH will need the chance to learn the aspects of depth perception through fine and gross motor activities.

Children with ONH should have regular evaluations with an Endocrinologist to address issues with hormone levels.