



Information Sheet: Eye Problems Associated with Hydrocephalus

Some children and adults with hydrocephalus develop eye problems. Some of these may be “silent” at first, identifiable only through an ophthalmologic (eye) screening examination. It is thus important for all individuals with hydrocephalus to undergo a baseline ophthalmologic exam. In some cases, the appearance of the optic nerve may even help your neurologist and neurosurgeon determine the severity of your hydrocephalus.

If possible, children should see a pediatric ophthalmologist, one who gives special vision tests to children, using videos and automated toys to maximize their performance during testing. In any eye exam, the need for glasses is determined after the eyes have been dilated, using an objective test called “retinoscopy.” Glasses, if needed, can be prescribed even for very young children.

Visual Symptoms of Hydrocephalus

High cerebrospinal fluid (CSF) pressure can damage vision, the resulting problems ranging from mild deterioration to marked loss. In both children and adults, the following may be signs of hydrocephalus:

- Grayouts or fuzzouts of vision
- Double vision
- Reduced vision
- Misaligned eyes
- Unstable, shaking eyes
- The “sunsetting sign” (infants)

An ophthalmologist typically looks for the following when examining a patient with hydrocephalus:

- Swelling of the optic nerve (papilledema)
- Eye misalignment (strabismus)
- Reduced color vision
- Reduced side vision (visual field loss)
- Abnormal pupil reaction to light
- Less than 20/20 vision (with glasses if needed)

How Does Hydrocephalus Injure the Eye?

Visual information is transmitted to the brain by the optic nerve, a cord that runs from the eye to the brain. The meninges that surround the brain and spinal cord also surround the optic nerve. Thus, increases in CSF pressure around the brain can also produce pressure on the optic nerve.

This pressure chokes off the supply of food and oxygen to the optic nerve, causing it to swell. Swelling of the optic nerve due to hydrocephalus is called “papilledema.” The nerve can be damaged through high CSF pressure or because of lower pressure elevation that lasts for a prolonged time period. Damage to the optic nerve from papilledema can result in reduced vision, reduced color vision, and visual field loss.

In addition, some nerves travel from the brain to control the eye muscles. Three in particular (the third, fourth, and sixth nerves) control eye position and movement. With hydrocephalus, these nerves can be weakened, producing eye misalignment (strabismus). Adults may experience double vision as a result, while children may see double at first but, without treatment, will soon develop a lazy eye.

The sixth nerve controls the laterus rectus muscle, which pulls the eye away from the nose. Because of its long pathway in the brain, the sixth nerve is especially susceptible to injury in hydrocephalus. CSF pressure (or a change in pressure after shunt placement) can stretch the sixth nerve, causing the eyes to cross (esotropia). Sometimes a head turn develops to avoid the resulting double vision.

Visual Problems in Children

Children with hydrocephalus sometimes develop eye misalignment (strabismus) through nerve damage. Strabismus can force a child to assume an uncomfortable head posture, with a tilt or turn, causing pain. A child learning to walk will be hindered by a chin-down posture or large head turn. Strabismus in children with hydrocephalus should thus be carefully tracked by a pediatric ophthalmologist. Sometimes eye muscle surgery can improve eye alignment if the eyes do not straighten after management of the child’s hydrocephalus improves.

A lazy eye with poor vision (amblyopia) may occur in children under age eight when one eye is favored because of strabismus or another cause. Patching treats poor vision from amblyopia.

The “sunsetting sign,” sometimes observed in infants, is an eye misalignment that should be particularly emphasized. In this kind of misalignment, the eyes turn downward, the white showing above. This pattern occurs in infants before effective hydrocephalus treatment has been started, but a sudden reappearance of sunseting may indicate that an implanted shunt is not working properly.

The sudden development of a turn to the eye or a new head position, or the appearance or reappearance of sunseting eyes, should immediately be reported to a child’s physician. These conditions may be signs of a shunt malfunction.

Perceptual Problems

Some individuals with hydrocephalus also have perceptual problems. For example, a child may have trouble naming objects, reading, or calculating math. Such perceptual defects can add to a child’s frustrations at school and to an adult’s frustrations in the workplace. Both might find that such defects make social situations difficult.

However, perceptual difficulties can be assessed, and strategies can be implemented to counter them. Vision and hearing examinations should be early steps in assessing perceptual problems. Evaluation might also include a formal educational and developmental assessment through a child's school or a medical facility. Many hospitals offer developmental assessments and neuropsychological evaluations and testing through their departments of neurology or psychology.

Conclusion

Every individual with hydrocephalus, whether a child or a newly diagnosed adult, should have a baseline ophthalmologic examination. A child's examination should be performed by a pediatric ophthalmologist, while adults should find a qualified neuro-ophthalmologist. Your physician can help you determine how often follow-up examinations should be performed in your particular case.

Remember: Many eye problems, for both children and adults with hydrocephalus, represent the effects of pressure, both past and present. The eyes sometimes give us early warning signs of trouble. If you're in any doubt about visual symptoms you're experiencing, discuss your concerns with your physician.

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