CONTINUED Professional Development

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OCULAR PROBLEMS ASSOCIATED WITH HYDROCEPHALUS

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INTRODUCTION

Hydrocephalus, loosely translated from Greek as "water on the brain" is the abnormal buildup of cerebrospinal fluid (CSF) in the ventricles or subarachnoid space of the brain. This may cause raised increased intracranial pressure (ICP) and progressive enlargement of the head, with accompanying convulsion, tunnel vision, and mental disability. In some instances, hydrocephalus can also cause death.

AETIOLOGY

CSF surrounds the brain and spinal cord, and helps cushion the brain. CSF normally moves through the brain and the spinal cord, and is soaked into the bloodstream. The pressure inside the skull is caused in part by the production of the CSF and in part by the pressure of the blood being pumped to the brain. As the CSF pressure increases, it will interfere with the brain's blood supply. This will deprive it of the oxygen and the glucose it uses to function effectively. The lack of these two elements will cause tiredness, irritability and drowsiness. If the pressure is not released, loss of consciousness will occur as the brain functions slowly shut down. CSF levels in the brain can rise if: the flow of CSF is blocked, it does not get absorbed into the blood properly or if the brain makes too much of it. Significant accumulation of CSF puts pressure on the brain. This pushes the brain up against the skull and damages the brain tissue.

Hydrocephalus may begin while the baby is growing in the womb. It is common in babies who have a myelomeningocele, a birth defect in which the spinal column does not close properly. Hydrocephalus may also be due to: genetic defects and certain infections during pregnancy.

In young children, hydrocephalus may be due to: infections that affect the central nervous system (such as meningitis or encephalitis), especially in infants, bleeding in the brain during or soon after delivery (especially in premature babies), injury before, during, or after childbirth, including subarachnoid hemorrhage, tumours of the central nervous system, including the brain or spinal cord and injury or trauma. Hydrocephalus can occur in adults.

DEFECTS PRODUCED BY HYDROCEPHALUS

As vital functions are controlled from the brain stem, a very high CSF pressure can lead to problems with the heart and breathing. In cases where damage to the brain has occurred, there can be a number of effects. Seizures are experienced by about one third of people with hydrocephalus at some point in their lives. This can be in relation to shunt revisions or due to a rise in ICP.

Learning disorders are common amongst those with hydrocephalus, but their exact effects vary considerably. There may be subtle problems of co-ordination of hand movements, with what the person sees, as well as a degree of clumsiness, which make it difficult to perform certain tasks or do specific jobs.

Visual perception may be a problem, leading to difficulties understanding position and relationship of objects in space. This manifests in problems with judging distance, direction, slopes, height of kerbs, width of doors, space in a room and speed. Obviously, problems with identifying difference between shapes impacts reading and writing.

The effects of hydrocephalus can include: problems with decision making, logical thinking, and organisation, inability to follow verbal instructions, short term memory difficulties and passive behaviour. Some people are very seriously distressed by everyday noises such as vacuum cleaners or washing machines. All of these have major implications for adult life. Symptoms of hydrocephalus depend on: age, amount of brain damage and the cause of the buildup of CSF fluid

In infants with hydrocephalus, it causes the fontanelle (soft spot) to bulge and the head to be larger than expected. Early symptoms may also include: eyes that appear to gaze downward, irritability, seizures, sleepiness and vomiting.

Symptoms that may occur in older children can include: brief, shrill, high-pitched cry, changes in personality, memory, or the ability to reason or think, changes in facial appearance, strabismus, nystagmus, difficulty feeding, excessive sleepiness, headache, irritability, poor temper control, urinary incontinence, loss of coordination and trouble walking, muscle spasticity, slow growth (child 0 - 5 years), slow or restricted movement and vomiting.

MECHANISM OF OCULAR PROBLEMS

Visual information is transmitted to the brain by the optic nerve. 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 creating papilloedema. Damage to the optic nerve from papilloedema can result in reduced vision, reduced colour vision, and visual field loss. Some nerves (three in particular, the third, fourth, and sixth nerves) travel from the brain to control the eye muscles by influencing eye position and movement. With hydrocephalus, these nerves can be weakened, producing strabismus. Adults may experience double vision as a result, while children may see double at first but, without treatment, will soon develop amblyopia.

Accreditation Number: ODO 002/02/08/2014

OCULAR PROBLEMS ASSOCIATED WITH HYDROCEPHALUS

The sixth nerve controls the lateral 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 causing esotropia. Sometimes a head turn, especially in children develops to avoid the resulting diplopia.

VISUAL SIGNS AND SYMPTOMS OF HYDROCEPHALUS

High cerebrospinal fluid (CSF) pressure can decrease vision, the resulting problems ranging from mild deterioration to marked loss. In both children and adults, the following may be signs of hydrocephalus: diplopia, reduced vision, strabismus, nystagmus, the "sunsetting sign" (infants), papilloedema, reduced colour vision, reduced visual field, refractive error and abnormal pupil reaction to light in some cases. Strabismus can force a child to assume a head posture, with a tilt or turn. A child learning to walk will be hindered by a chin-down posture or large head turn. The "sunsetting sign," sometimes observed in infants, is an eye misalignment where the eyes turn downward, with the superior bulbar conjunctiva showing above.

While most patients with hydrocephalus are able to move their eyes up and down quite well, in some cases these patients cannot raise their eyes to look up, and in doing so they develop a retraction of the upper eyelids. This syndrome is referred to as Parinaud's Oculogranular Syndrome.

Figure 1: "Sunsetting sign" in a hydrocephalus patient

CLINICAL RESEARCH

During 1999-2002, a population-based ophthalmologic study of all the children with hydrocephalus born in western Sweden was conducted. Aetiological, neurological and neuroimaging information was collected from the case records. In the study, the following were found: visual function deficits were identified in more than 80% of the children with hydrocephalus, low visual acuity, refractive errors, strabismus and difficulties with visual processing. A majority of the children had one or more neurological impairments.

MANAGEMENT

Regular tests are conducted to check the child's developmental milestones to evaluate physical, neurological and intellectual status.

The physician takes head circumference measurements, repeated over time. A head CT scan is one of the best tests for identifying hydrocephalus. Other tests that may be done include: cranial ultrasound, brain scan using radioisotopes arteriography, lumbar puncture, examination of the cerebrospinal fluid and skull x-rays.

The goal of treatment is to reduce or prevent brain damage by improving the flow of CSF. Surgery may be done to remove a blockage, if possible. If not, a flexible tube called a shunt may be placed in the brain to re-route the flow of CSF by placing of a ventricular catheter into the cerebral ventricles to bypass the impeding flow caused by obstruction/malfunctioning arachnoidal granulations and drain the excess fluid into other body cavities, from where it can be resorbed. Most shunts drain the fluid into the peritoneal cavity (ventriculo-peritoneal shunt), but alternative sites include the right atrium (ventriculo-atrial shunt), pleural cavity (ventriculo-pleural shunt), and gallbladder. A shunt system can also be placed in the lumbar space of the spine and have the CSF redirected to the peritoneal cavity (lumbar-peritoneal shunt). An alternative treatment for obstructive hydrocephalus in selected patients is the endoscopic third ventriculostomy (ETV), whereby a surgically created opening in the floor of the third ventricle allows the CSF to flow directly to the basal cisterns, thereby shortcutting any obstruction, as in aqueductal stenosis.

Other treatments may include: antibiotics if there are signs of infection, and removing or cauterising the parts of the brain that produce CSF. If epilepsy is developed it will be treated with anti-convulsive drugs, just like for people without hydrocephalus.

Several medical and surgical treatments that sometimes help people with nystagmus are available. Surgery usually reduces the null positions, lessening head tilt and improving cosmetic appearance. Drugs such as Botox or Baclofen can reduce some nystagmic movements, although results are usually temporary. The objective of the optometrist is to find a suitable lens that improves vision as well as reduce nystagmus. With this in mind, the final prescription is the maximum plus Rx that attains these two goals.

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OCULAR PROBLEMS ASSOCIATED WITH HYDROCEPHALUS

CLINICAL PEARLS

- Hydrocephalus most often occurs in children. Another type, called normal pressure hydrocephalus, may occur in adults.
- Without treatment, up to 6 in 10 people with hydrocephalus will die. Those who survive have different amounts of intellectual, physical, and neurological disabilities.
- Although a shunt generally works well, it may stop working if it disconnects, becomes blocked, infected, or it is outgrown. If this happens the cerebrospinal fluid will begin to accumulate again and a number of physical symptoms will develop (headaches, nausea, vomiting, photophobia, and seizures.) A sudden reappearance of the "sunsetting sign" or the sudden development of strabismus or a new head position may indicate that an implanted shunt is not working properly.
- Symptoms of increased intracranial pressure may include headaches, vomiting, nausea, papilledema, sleepiness or coma. Elevated intracranial pressure may result in uncal and/or cerebellar tonsill herniation, with resulting life-threatening brain stem compression.

CONCLUSION

Every individual with hydrocephalus, whether a child or a newly diagnosed adult, should have a baseline eye examination. Eyecare practitioners need to have a good understanding of the appropriate eyecare and overall management plan of the hydrocephalus patient.

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