# Hydrocephalus

What is Hydrocephalus?

Hydrocephalus is a neurological disorder most frequently seen in children, but it can also affect adults.

Hydrocephalus results from the excessive accumulation of fluid in the brain. The word hydrocephalus derives from the Greek words "hydro" meaning water and "cephalus" meaning head. Although the condition has often been called "water on the brain," the fluid is actually cerebrospinal fluid (CSF), which is chemically different than water.
This fluid is the liquid in which the brain floats.

There are several cavities in the brain known as ventricles. These ventricles are filled with CSF, which is constantly produced inside the ventricles and circulates around the brain and spinal cord. The ventricles are arranged like a series of rooms connected by narrow hallways. The CSF passes from one room to the next through the hallways. It carries nutrients and proteins to the brain and transports waste products away from surrounding brain tissue. The average adult produces about one pint of CSF daily.

When an injury or illness alters the circulation of the CSF, one or more of the ventricles becomes enlarged as the CSF accumulates. In infants and young children, whose skulls are not yet fused, the skull can enlarge to accommodate the excess fluid. The soft spot on an infant"s head (the fontanelle) can become firm as pressure develops. In an adult, however, the skull is rigid and cannot expand. In this case, the pressure in the brain may increase profoundly.

In infants and children, hydrocephalus usually results from a birth defect, viral infection, head injury, hemorrhage, meningitis or tumor. In adults, the causes include brain damage due to stroke or injury, Alzheimer"s Disease or obstruction of the ventricles. Often, the cause is unknown.

Hydrocephalus is a chronic condition. It can be controlled but usually not cured. With appropriate early treatment, however, many people with hydrocephalus lead normal lives with few limitations.

Two Types of Adult-Onset Hydrocephalus

The two most common forms of adult hydrocephalus are hydrocephalus ex-vacuo and normal pressure hydrocephalus (NPH).

Hydrocephalus ex-vacuo occurs when a stroke or injury damages the brain and brain matter actually shrinks. The brain may actually shrink in elderly patients or those with Alzheimer"s Disease, and the CSF volume increases to fill the extra space. In this instance, the ventricles are enlarged, but the pressure may or may not be elevated.

Normal pressure hydrocephalus results from the gradual blockage of the CSF draining pathways in the brain. The ventricles enlarge to handle the increased volume of the CSF, and the compression of the brain from within by the fluid-filled ventricles destroys or damages brain tissue. Normal pressure hydrocephalus owes its name to the fact that the ventricles inside the

brain become enlarged with little or no increase in pressure. Yet the name of the condition can be misleading. Some patients actually do show a fluctuation of CSF pressure from high to normal to low with pressure monitoring.

NPH can occur as the result of head injury, cranial surgery, hemorrhage, meningitis or tumor. Unfortunately, the majority of NPH cases are from an unknown cause, making it difficult to diagnose and understand. Compounding this difficulty is the fact that the symptoms of hydrocephalus are similar to the effects of the aging process. The majority of the NPH population is older than 60, and people sometimes live for months or even years with hydrocephalus before seeing a doctor.

Symptoms of Adult-Onset Hydrocephalus

Headaches and nausea are common symptoms of adult-onset hydrocephalus. Other signs of the condition are difficulty focusing the eyes, unsteady walking, weakness of the legs, sudden falls, and a distinctive inability to walk forward, as if the feet were stuck to the floor. As the condition progresses, those with hydrocephalus show decreased mental activity, reflected in withdrawn behavior, lethargy, apathy, impaired memory, and speech problems. Urinary and bowel incontinence can also occur. Dementia, involving loss of movement, sensory functions, and cognitive abilities, may result.

Diagnosing Hydrocephalus

Physicians have a wide variety of diagnostic tools to evaluate hydrocephalus.

* A Computed Tomography (CT) scan of the head uses an x-ray beam, which passes through the head allowing a computer to make a picture of the brain in slices. It is reliable, safe, painless, and quick (about 15 minutes). A CT will show if the ventricles are enlarged or if there is an obvious blockage.
* Magnetic Resonance Imaging (an MRI) is also safe and painless but takes a little longer (an hour). MRI uses radio signals and a very powerful magnet to create a picture of the brain that can reveal if the ventricles are enlarged and evaluate the CSF flow.
* Isotopic cisternography involves injecting a radioactive isotope into the lower back through a spinal tap. This allows the absorption of CSF to be monitored over a period of time (up to 4 days). Isotopic cisternography is considerably more involved than a CT or MRI but can aid the diagnosis of NPH.
* Lumbar puncture (spinal tap) can be used to measure CSF pressure and analyze the fluid. Sometimes the procedure helps indicate whether a shunt, the common treatment for hydrocephalus, would work. If lumbar puncture improves symptoms even temporarily, then the shunt may be successful.
* Intracranial pressure monitoring may be able to detect an abnormal pressure or pattern of pressure waves. Monitoring requires insertion of a catheter or small fiber optic cable through the skull into the brain. Both insertion techniques are safe and require admission to the hospital for 24 hours.

Treatment

The most common treatment of hydrocephalus involves diverting the buildup of CSF to somewhere else in the body through the use of a device called a shunt. A shunt is a small tube (the size of a spaghetti noodle) inserted into the ventricles to drain the fluid away from the brain. Usually, the tube is routed beneath the skin to the peritoneal cavity (the area surrounding the

abdominal organs). The CSF is eventually absorbed into the bloodstream. A valve is attached to the shunt to ensure that the CSF flows in a single direction and to regulate the pressure. The valve opens automatically when the CSF pressure in the brain exceeds a certain limit and closes when the pressure returns to an acceptable level.

Shunt operations are relatively common. An average of 75,000 shunt operations are performed annually in the United States, the majority in children. This surgical procedure controls hydrocephalus but does not cure it. Except in rare instances, hydrocephalus is a lifelong condition in which complications can arise. The shunt surgery is not particularly painful, and the hospital stay is typically short. After surgery, the shunt system is completely inside the body and often cannot be felt, even by experienced hands.

Over time, the shunt controls hydrocephalus by diverting the fluid before pressure can build up. The condition can also be treated directly. For example, a surgeon can remove a brain tumor that is causing obstruction of the ventricles or the connecting passages between the ventricles.

In certain cases of hydrocephalus, a neurosurgeon may recommend a surgical alternative to shunting. The procedure is called an endoscopic third ventriculostomy (ETV). The surgeon uses a special endoscope (small tube with a camera on the end) to create a small hole in the floor of the ventricles. This hole provides an alternative CSF passageway that bypasses an obstruction of the normal CSF flow through the ventricles.

Prognosis

The prognosis for hydrocephalus depends on the cause, the extent of symptoms and the timeliness of diagnosis and treatment. Some patients show a dramatic improvement with treatment while others do not. In some instances of normal pressure hydrocephalus, dementia can be reversed by shunt placement. Other symptoms such as headaches can disappear almost immediately if the symptoms are related to elevated pressure.

In general, the earlier hydrocephalus is diagnosed, the better the chance for successful treatment. The longer the symptoms have been present, the less likely it is that treatment will be successful. Unfortunately, there is no way to accurately predict in an individual case how successful surgery will be. Some patients will improve dramatically while others will reach a plateau or decline after a few months. Hydrocephalus caused by disorders other than infection carry the best prognosis. Hydrocephalus caused by tumors have the poorest outlook. When the cause of hydrocephalus is known, the success rate of shunting surgery nears 80 percent. In cases in which a cause is not known, the success rate can be much lower.

Shunt malfunction or failure can occur. The valve can become clogged or the pressure in the shunt may not match the needs of the patient, mandating additional surgery. In the event of an infection, antibiotic therapy is needed as well. A shunt malfunction may be indicated by headaches, vision problems, irritability, fatigue, personality change, loss of coordination, difficulty in waking up or staying awake, a return of walking problems, mild dementia and incontinence. Fortunately, most complications can be dealt with successfully. Neurosurgeons are the best medical professionals to treat hydrocephalus since they diagnose and treat disorders of the nervous system, the brain, and the spinal cord.