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Hydrocephalus Fact Sheet

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What is hydrocephalus?

The term hydrocephalus is derived from the Greek words "hydro" meaning water and "cephalus" meaning head. As the name implies, it is a condition in which the primary characteristic is excessive accumulation of fluid in the brain. Although hydrocephalus was once known as "water on the brain," the "water" is actually cerebrospinal fluid (CSF) — a clear fluid that surrounds the brain and spinal cord. The excessive accumulation of CSF results in an abnormal widening of spaces in the brain called ventricles. This widening creates potentially harmful pressure on the tissues of the brain.

The ventricular system is made up of four ventricles connected by narrow passages. Normally, CSF flows through the ventricles, exits into cisterns (closed spaces that serve as reservoirs) at the base of the brain, bathes the surfaces of the brain and spinal cord, and then reabsorbs into the bloodstream.

CSF has three important life-sustaining functions: 1) to keep the brain tissue buoyant, acting as a cushion or "shock absorber"; 2) to act as the vehicle for delivering nutrients to the brain and removing waste; and 3) to flow between the cranium and spine and compensate for changes in intracranial blood volume (the amount of blood within the brain).

The balance between production and absorption of CSF is critically important. Because CSF is made continuously, medical conditions that block its normal flow or absorption will result in an over-accumulation of CSF. The resulting pressure of the fluid against brain tissue is what causes hydrocephalus.

What are the different types of hydrocephalus?

Hydrocephalus may be congenital or acquired. Congenital hydrocephalus is present at birth and may be caused by either events or influences that occur during fetal development, or genetic abnormalities. Acquired hydrocephalus develops at the time of birth or at some point afterward. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease.

Hydrocephalus may also be communicating or non-communicating. Communicating hydrocephalus occurs when the flow of CSF is blocked after it exits the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open. Non-communicating hydrocephalus — also called "obstructive" hydrocephalus — occurs when the flow of CSF is blocked along one or more of the narrow passages connecting the ventricles. One of the most common causes of hydrocephalus is "aqueductal stenosis." In this case, hydrocephalus results from a narrowing of the *aqueduct of Sylvius*, a small passage between the third and fourth ventricles in the middle of the brain.

There are two other forms of hydrocephalus which do not fit exactly into the categories mentioned above and primarily affect adults: hydrocephalus ex-vacuo and Normal Pressure Hydrocephalus (NPH).

Hydrocephalus ex-vacuo occurs when stroke or traumatic injury cause damage to the brain. In these cases, brain tissue may actually shrink. NPH is an abnormal increase of cerebrospinal fluid in the brain's ventricles that may result from a subarachnoid hemorrhage, head trauma, infection, tumor, or complications of surgery. However, many people develop NPH when none of these factors are present. An estimated 375,000 older Americans have NPH.

Who gets this disorder?

The number of people who develop hydrocephalus or who are currently living with it is difficult to establish since the condition occurs in children and adults, and can develop later in life. A 2008 data review by the University of Utah found that, in 2003, hydrocephalus accounted for 0.6 percent of all pediatric hospital admissions in the United States. Some estimates report one to two of every 1,000 babies are born with hydrocephalus.

What causes hydrocephalus?

The causes of hydrocephalus are still not well understood. Hydrocephalus may result from inherited genetic abnormalities (such as the genetic defect that causes aqueductal stenosis) or developmental disorders (such as those associated with neural tube defects including spina bifida and encephalocele). Other possible causes include complications of premature birth such as intraventricular hemorrhage, diseases such as meningitis, tumors, traumatic head injury, or subarachnoid hemorrhage, which block the exit of CSF from the ventricles to the cisterns or eliminate the passageway for CSF within the cisterns.

What are the symptoms?

Symptoms of hydrocephalus vary with age, disease progression, and individual differences in tolerance to the condition. For example, an infant's ability to compensate for increased CSF pressure and enlargement of the ventricles differs from an adult's. The infant skull can expand to accommodate the buildup of CSF because the sutures (the fibrous joints that connect the bones of the skull) have not yet closed.

In infancy, the most obvious indication of hydrocephalus is often a rapid increase in head circumference or an unusually large head size. Other symptoms may include vomiting, sleepiness, irritability, downward deviation of the eyes (also called "sun setting"), and seizures.

Older children and adults may experience different symptoms because their skulls cannot expand to accommodate the buildup of CSF. Symptoms may include headache followed by vomiting, nausea, blurred or double vision, sun setting of the eyes, problems with balance, poor coordination, gait disturbance, urinary incontinence, slowing or loss of developmental progress, lethargy, drowsiness, irritability, or other changes in personality or cognition including memory loss.

Symptoms of normal pressure hydrocephalus include problems with walking, impaired bladder control leading to urinary frequency and/or incontinence, and progressive mental impairment and dementia. An individual with this type of hydrocephalus may have a general slowing of movements or may complain that his or her feet feel "stuck." Because some of these symptoms may also be experienced in other disorders such as Alzheimer's disease, Parkinson's disease, and Creutzfeldt-Jakob disease, normal pressure hydrocephalus is often incorrectly diagnosed and never properly treated. Doctors may use a variety of tests, including brain scans such as computed tomography (CT) and magnetic resonance imaging (MRI), a spinal tap or lumbar catheter, intracranial pressure monitoring, and neuropsychological tests, to help them accurately diagnose normal pressure hydrocephalus and rule out any other conditions.

The symptoms described in this section account for the most typical ways in which progressive hydrocephalus is noticeable, but it is important to remember that symptoms vary significantly from person to person.

How is hydrocephalus diagnosed?

Hydrocephalus is diagnosed through clinical neurological evaluation and by using cranial imaging techniques such as ultrasonography, CT, MRI, or pressure-monitoring techniques. A physician selects the appropriate diagnostic tool based on an individual's age, clinical presentation, and the presence of known or suspected abnormalities of the brain or spinal cord.

What is the current treatment?

Hydrocephalus is most often treated by surgically inserting a shunt system. This system diverts the flow of CSF from the CNS to another area of the body where it can be absorbed as part of the normal circulatory process.

A shunt is a flexible but sturdy plastic tube. A shunt system consists of the shunt, a catheter, and a valve. One end of the catheter is placed within a ventricle inside the brain or in the CSF outside the spinal cord. The other end of the catheter is commonly placed within the abdominal cavity, but may also be placed at other sites in the body such as a chamber of the heart or areas around the lung where the CSF can drain and be absorbed. A valve located along the catheter maintains one-way flow and regulates the rate of CSF flow.

A limited number of individuals can be treated with an alternative procedure called third ventriculostomy. In this procedure, a neuroendoscope — a small camera that uses fiber optic technology to visualize small and difficult to reach surgical areas — allows a doctor to view the ventricular surface. Once the scope is guided into position, a small tool makes a tiny hole in the floor of the third ventricle, which allows the CSF to bypass the obstruction and flow toward the site of resorption around the surface of the brain.

What are the possible complications of a shunt system?

Shunt systems are imperfect devices. Complications may include mechanical failure, infections, obstructions, and the need to lengthen or replace the catheter. Generally, shunt systems require monitoring and regular medical follow up. When complications occur, subsequent surgery to replace the failed part or the entire shunt system may be needed.

Some complications can lead to other problems such as overdrainage or underdrainage. Overdrainage occurs when the shunt allows CSF to drain from the ventricles more quickly than it is produced. Overdrainage can cause the ventricles to collapse, tearing blood vessels and causing headache, hemorrhage (subdural hematoma), or slit-like ventricles (slit ventricle syndrome). Underdrainage occurs when CSF is not removed quickly enough and the symptoms of hydrocephalus recur. Overdrainage and underdrainage of CSF are addressed by adjusting the drainage pressure of the shunt valve; if the shunt has an adjustable pressure valve these changes can be made by placing a special magnet on the scalp over the valve. In addition to the common symptoms of hydrocephalus, infections from a shunt may also produce symptoms such as a low-grade fever, soreness of the neck or shoulder muscles, and redness or tenderness along the shunt tract. When there is reason to suspect that a shunt system is not functioning properly (for example, if the symptoms of hydrocephalus return), medical attention should be sought immediately.

What is the prognosis?

The prognosis for individuals diagnosed with hydrocephalus is difficult to predict, although there is some correlation between the specific cause of the hydrocephalus and the outcome. Prognosis is further clouded by the presence of associated disorders, the timeliness of diagnosis, and the success of treatment. The degree to which relief of CSF pressure following shunt surgery can minimize or reverse damage to the brain is not well understood.

Affected individuals and their families should be aware that hydrocephalus poses risks to both cognitive and physical development. However, many children diagnosed with the disorder benefit from rehabilitation therapies and educational interventions and go on to lead normal lives with few limitations. Treatment by an interdisciplinary team of medical professionals, rehabilitation specialists, and educational experts is critical to a positive outcome. Left untreated, progressive hydrocephalus may be fatal.

The symptoms of normal pressure hydrocephalus usually get worse over time if the condition is not treated, although some people may experience temporary improvements. While the success of treatment with shunts varies from person to person, some people recover almost completely after treatment and have a good quality of life. Early diagnosis and treatment improves the chance of a good recovery.

What research is being done?

The National Institute of Neurological Disorders and Stroke (NINDS) and other institutes of the National Institutes of Health (NIH) conduct research related to hydrocephalus and support additional research through grants to major medical research institutions across the country. Much of this research focuses on finding better ways to prevent, treat, and ultimately cure disorders such as hydrocephalus. The NINDS also conducts and supports a wide range of fundamental studies that explore the complex mechanisms of normal and abnormal brain development.

The Hydrocephalus Clinical Research Network (HCRN, www.hcrn.org) is a multi-center collaborative research effort that was borne out of the first NIH workshop on hydrocephalus. NINDS supported the work of HCRN through the Challenge Grant process to advance their studies. HCRN consists of seven pediatric centers that pool their hydrocephalus patient population to more rapidly study the potential for improved treatments. HCRN conducts multiple, simultaneous studies at all of its centers and maintains a substantial registry of patients and procedures.

Where can I get more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN
P.O. Box 5801
Bethesda, MD 20824
(800) 352-9424
<http://www.ninds.nih.gov>

Information also is available from the following organizations:

[Hydrocephalus Association](http://www.hydroassoc.org)
4340 East West Highway
Suite 905
Bethesda, MD 20814
info@hydroassoc.org
<http://www.hydroassoc.org> 
Tel: 301-202-3811 888-598-3789
Fax: 301-202-3813

[National Hydrocephalus Foundation](http://www.nhfonline.org)
12413 Centralia Road
Lakewood, CA 90715-1653
debbifields@nhfonline.org
<http://nhfonline.org> 
Tel: 562-924-6666 888-857-3434

[Pediatric Hydrocephalus Foundation](http://www.hydrocephaluskids.org)
2004 Green Hollow Drive
Iselin, NJ 08830
info@hydrocephaluskids.org
<http://www.HydrocephalusKids.org> 
Tel: 732-634-1283

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