Hydrocephalus

U.S. DEPARTMENT OF HEALTH
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What is hydrocephalus?

The term hydrocephalus is derived from the Greek words "hydro" meaning water and "cephalus" meaning head. As its 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 surrounding the brain and spinal cord. The excessive accumulation of CSF results in an abnormal dilation of the spaces in the brain called ventricles. This dilation causes potentially harmful pressure on the tissues of the brain.

The ventricular system is made up of four ventricles connected by narrow pathways. 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 is absorbed 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 to 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. Ideally, the fluid is almost completely absorbed into the bloodstream as it circulates; however, there are circumstances which, when present, will prevent or disturb the production or absorption of CSF, or which will inhibit its normal flow. When this balance is disturbed, hydrocephalus is the result.

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 environmental

influences during fetal development or genetic predisposition. 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 from 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 pathways 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 passageway between the third and fourth ventricles in the middle of the brain.

There are two other forms of hydrocephalus which do not fit distinctly into the categories mentioned above and primarily affect adults: ***hydrocephalus ex-vacuo*** and ***normal pressure hydrocephalus.***

Hydrocephalus ex-vacuo occurs when there is damage to the brain caused by stroke or traumatic injury. In these cases, there may be actual shrinkage (atrophy or wasting) of brain tissue. Normal pressure hydrocephalus commonly occurs in the elderly and is characterized by many of the same symptoms associated with other conditions that occur more often in the elderly, such as memory loss, dementia, gait disorder, urinary incontinence, and a general slowing of activity.

Who gets this disorder?

Incidence and prevalence data are difficult to establish as there is no existing national registry or database of people with hydrocephalus and closely associated disorders; however, hydrocephalus is believed to affect approximately 1 in every 500 children. At present, most of these cases are diagnosed prenatally, at the time of delivery, or in early childhood. Advances in diagnostic imaging technology allow more accurate diagnoses in individuals with atypical presentations, including adults with conditions such as normal pressure hydrocephalus.

What causes hydrocephalus?

The causes of hydrocephalus are not all well understood. Hydrocephalus may result from genetic inheritance (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 blocking the exit from the ventricles to the cisterns and eliminating the cisterns themselves.

What are the symptoms?

Symptoms of hydrocephalus vary with age, disease progression, and individual differences in tolerance to CSF. For example, an infant's ability to tolerate CSF pressure 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 the 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 "sunsetting"), and seizures.

Older children and adults may experience different symptoms because their skulls cannot expand to accommodate the buildup of CSF. In older children or adults, symptoms may

include headache followed by vomiting, nausea, papilledema (swelling of the optic disk which is part of the optic nerve), blurred vision, diplopia (double vision), sunsetting of the eyes, problems with balance, poor coordination, gait disturbance, urinary incontinence, slowing or loss of development, lethargy, drowsiness, irritability, or other changes in personality or cognition including memory loss.

The symptoms described in this section account for the most typical ways in which progressive hydrocephalus manifests itself; it is, however, important to remember that symptoms vary significantly from individual to individual.

How is hydrocephalus diagnosed?

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

What are the current treatments?

Hydrocephalus is most often treated with the surgical placement of a shunt system. This system diverts the flow of CSF from a site within the central nervous system (CNS) to another area of the body where it can be absorbed as part of the circulatory process.

A shunt is a flexible but sturdy silastic tube. A shunt system consists of the shunt, a catheter, and a valve. One end of the catheter is placed in the CNS—most usually within a ventricle inside the brain, but also potentially within a cyst or in a site close to the spinal cord. The other end of the catheter is commonly placed within the peritoneal (abdominal) cavity, but may also be placed at other sites within the body such as a chamber of the heart or a cavity in 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 patients can be treated with an alternative procedure called third ventriculostomy. In this procedure, a neuroendoscope—a small camera designed to visualize small and difficult to reach surgical areas—allows a doctor to view the ventricular surface using fiber optic technology. The scope is guided into position so that a small hole can be made in the floor of the third ventricle, allowing 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 not perfect 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 followup. When complications do occur, usually the shunt system will require some type of revision.

Some complications can lead to other problems such as overdraining or underdraining. Overdraining occurs when the shunt allows CSF to drain from the ventricles more quickly than it is produced. This overdraining can cause the ventricles to collapse, tearing blood vessels and causing headache, hemorrhage (subdural hematoma), or slit-like ventricles (slit ventricle syndrome). Underdraining occurs when CSF is not removed quickly enough and the symptoms of hydrocephalus recur (*see* "What are the symptoms of hydrocephalus?"). 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 patients diagnosed with hydrocephalus is difficult to predict, although there is some correlation between the specific cause of the hydrocephalus and the patient's outcome. Prognosis is further complicated by the presence of associated disorders, the timeliness of diagnosis, and the success of treatment. The degree to which decompression (relief of CSF pressure or buildup) 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.

Treatment of patients with hydrocephalus is life-saving and life-sustaining. Left untreated, progressive hydrocephalus is, with rare exceptions, fatal.

What research is being done?

Within the Federal government, the leading supporter of research on hydrocephalus is the National Institute of Neurological Disorders and Stroke (NINDS). The NINDS, a part of the National Institutes of Health (NIH), is responsible for supporting and conducting research on the brain and the central nervous system. NINDS conducts research in its laboratories at NIH and also supports studies through grants to major medical institutions across the country.

One NINDS-supported study is examining cognitive development, academic achievement, and behavioral adjustment in children with hydrocephalus. Researchers hope this study will shed new light on the influence of hydrocephalus on development as well as the more general issue of the effect of early brain injury.

The NINDS also conducts and supports a wide range of fundamental studies that explore the complex mechanisms of normal brain development. The knowledge gained from these studies provides the foundation for understanding how this process can go awry and, thus, offers hope for new means to treat and prevent developmental brain disorders such as hydrocephalus.

Where can I find more information?

Private, voluntary organizations that offer information and services to those affected by hydrocephalus include the following:

Guardians of Hydrocephalus Research Foundation, Inc.
2618 Avenue Z
Brooklyn, New York 11235-2023
718/743-GHRF (4473)
800/458-8655

Hydrocephalus Association, Inc.
870 Market Street
Suite 955
San Francisco, California 94102
415/732-7040

Hydrocephalus Support Group
PO Box 4236
Chesterfield, Missouri 63006-4236
314/532-8228

National Hydrocephalus Foundation
12413 Centralia Road
Lakewood, CA 90715-1623
562-402-3523/888-260-1789
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http://nhfonline.org

For more information on the research programs of the NINDS, contact:

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