Hydrocephalus

U.S. DEPARTMENT OF HEALTH
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Hydrocephalus

What is hydrocephalus?

Hydrocephalus is an abnormal buildup of fluid in the ventricles (cavities) deep within the brain. This excess fluid causes the ventricles to widen, putting pressure on the brain's tissues.

Cerebrospinal fluid (CSF) is the clear, colorless fluid that protects and cushions the brain and spine. Normally, cerebrospinal fluid flows through the ventricles and bathes the brain and spinal cord before being reabsorbed into the bloodstream. The body typically produces enough CSF each day and absorbs the same amount. However, when the normal flow or absorption of CSF is blocked it can result in a buildup of CSF. The pressure from too much CSF can keep the brain from functioning properly and cause brain damage and even death.

Fortunately, there are treatment options that can restore normal levels of CSF. Though treatment is often helpful, it may take multiple surgeries to treat hydrocephalus. (Hydrocephalus is the most common reason for brain surgery in young children.) With treatment many people lead normal and productive lives.

What are the different types of hydrocephalus?

Hydrocephalus can affect anyone at any age but is most common in infants and older adults. Some of these cases can be associated with abnormalities in the brain and spinal cord during pregnancy.

The two major types of hydrocephalus are called communicating hydrocephalus and non-communicating hydrocephalus.

- **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, the passages between which remain open. Reduced flow and absorption of CSF into specialized blood vessels called arachnoid villi can also result in a buildup of CSF in the ventricles and communicating hydrocephalus.
- **Non-communicating hydrocephalus** happens when the flow of CSF is blocked along one or more of the narrow passages connecting the ventricles.

Two additional types of hydrocephalus include:

- **Hydrocephalus ex-vacuo** results from brain damage caused by stroke or injury. In these cases, brain tissues around the ventricles shrink, and the ventricles are bigger than normal because of this. Strictly speaking, this is not a true hydrocephalus, but rather, a "hydrocephalus look-alike" condition.
- **Normal Pressure Hydrocephalus (NPH)** can be the result of bleeding in the brain's CSF (subarachnoid or intraventricular

hemorrhage), head trauma, infection, tumor, or a complication of surgery. However, many people develop NPH when none of these factors are present. The increase in cerebrospinal fluid in NPH occurs slowly enough that the tissues around the ventricles compensate and the fluid pressure inside the head does not increase. NPH causes problems with walking, bladder control, and difficulties thinking and reasoning. Sometimes NPH can be mistaken for Alzheimer's disease.

What causes hydrocephalus?

Hydrocephalus may be present at birth (congenital) or may develop over time as a result of injury or disease (acquired). Except for hydrocephalus secondary to physical obstruction of CSF passages within the brain or skull by blood or tumor, the exact causes of hydrocephalus are still not well understood.

Congenital hydrocephalus

Babies may be born with hydrocephalus or develop the condition shortly after delivery. In these cases, hydrocephalus may be caused by:

- inherited genetic abnormalities that block the flow of CSF
- developmental disorders such as those associated with birth defects in the brain, spine, or spinal cord
- complications of premature birth such as bleeding within the ventricles
- infection during pregnancy such as rubella that can cause inflammation in the fetal brain tissue.

Acquired hydrocephalus

Certain factors can increase the risk of developing hydrocephalus at any age, including:

- brain or spinal cord tumors
- infections of the central nervous system such as bacterial meningitis
- injury or stroke that causes bleeding in the brain.

What are the symptoms?

The symptoms of hydrocephalus can vary significantly from person to person and mostly depend on age. Conditions other than hydrocephalus can cause similar symptoms so it is important to see a doctor to receive proper diagnosis and treatment.

Infants

Signs and symptomzs of hydrocephalus in infants include:

- a rapid increase in head size
- an unusually large head
- a bulge on the soft spot (fontanel) on the top of the head
- vomiting
- problems sucking or feeding
- sleepiness
- irritability
- eyes that are fixed downward (also called “sun setting”) or are not able to turn outward
- seizures.

Older children, young adults, and middle-aged adults

Symptoms in older children and adults may include:

- headache
- blurred or double vision
- nausea or vomiting
- problems with balance
- slowing or loss of developmental progress like walking or talking
- vision problems
- decline in school or job performance
- poor coordination
- loss of bladder control and/or frequent urination
- difficulty remaining awake or waking up
- sleepiness
- irritability
- changes in personality or cognition including memory loss.

Older adults

Symptoms in older adults may include:

- problems walking, often described as feet feeling "stuck"
- progressive mental impairment and dementia
- general slowing of movements
- loss of bladder control and/or frequent urination
- poor coordination and balance.

How is hydrocephalus diagnosed?

Hydrocephalus is diagnosed through a clinical neurological exam and by using brain imaging techniques and other tests based on

- age
- symptoms
- known or suspected abnormalities in the brain or spinal cord.

Neurological exam

The neurological exam may involve tests to determine:

- muscle strength and reflexes
- coordination and balance
- vision, eye movement, and hearing
- mental functioning and mood.

Brain imaging and other tests

Tests to accurately diagnose hydrocephalus and rule out other conditions may include:

- **Ultrasound** is often the first test a doctor uses to diagnose infants because it is relatively simple and low risk. When used during routine prenatal exams, ultrasound may also detect hydrocephalus in unborn babies.
- **Magnetic resonance imaging (MRI)** can determine if the ventricles have enlarged, assess the CSF flow, and provide information about the brain tissue surrounding the ventricles. MRI is usually the initial test used to diagnose adults.
- **Computed tomography (CT)** can show doctors if the ventricles are enlarged or if there is an obstruction.

- **Spinal tap (lumbar puncture)** allows doctors to estimate CSF pressure and analyze the fluid by inserting a needle in the lower back and removing and testing some of the fluid.
- **Intracranial pressure monitoring (ICP)** uses a small pressure monitor inserted into the brain or ventricles to measure the pressure and detect the amount of swelling that may have occurred in the brain. If the pressure is too high, a doctor may drain the CSF to maintain the flow of oxygenated blood to the brain.
- **Fundoscopic examination** uses a special device to view the optic nerve at the back of the eye. It can show evidence of swelling that suggests elevated intracranial pressure, which can be a result of hydrocephalus.

What treatments are available?

Surgical treatments

Hydrocephalus is treated with one of two surgical options:

- **A shunt** (tube) is surgically inserted into the brain and connected to a flexible tube placed under the skin to drain the excess fluid into either the chest cavity or the abdomen so it can be absorbed by the body.
- **Endoscopic Third Ventriculostomy (ETV)** improves the flow of CSF out of the brain. A tiny hole is made at the bottom of the third ventricle and the CSF is diverted there to relieve pressure. Sometimes this is done in conjunction with choroid plexus cauterization to try and decrease the production of CSF. Choroid plexus cauterization uses electric current to burn the CSF-producing tissue (i.e., the choroid plexus) in the lateral ventricles in the brain, so it produces less CSF.

Shunt systems generally function well but they can fail to properly drain the CSF due to mechanical failure or infection. When this happens the CSF once again begins to build up in the brain and earlier symptoms may recur. To reduce the buildup of CSF, the clogged shunt system is replaced to restore drainage of CSF.

Shunts require monitoring and regular medical checkups. Multiple surgeries may be needed to repair or replace a shunt throughout a person's lifetime. Seek medical help immediately if symptoms develop that suggest the shunt system is not working properly.

Signs and symptoms of shunt malfunction may include:

- headache
- double vision or sensitivity to light
- nausea or vomiting
- soreness of the neck or shoulder muscles
- seizures
- redness or tenderness along the shunt tract
- low-grade fever
- sleepiness or exhaustion
- reoccurrence of hydrocephalus symptoms.

Other treatments

Many people diagnosed with hydrocephalus benefit from rehabilitation therapies and educational interventions. Treatment by an interdisciplinary team of medical professionals, rehabilitation specialists, and educational experts is critical to a positive outcome.

Supportive therapies for children may include:

- occupational and developmental therapists who can help children learn life skills and develop social behaviors
- special education teachers who can help tackle learning disabilities
- mental health providers or social workers who can provide emotional support and help families find services.

Adults may also require similar support, including social workers, occupational therapists, and specialists in dementia care.

What is the prognosis?

If left untreated, hydrocephalus can be fatal. Early diagnosis and successful treatment improve the chance for a good recovery.

With the benefits of surgery, rehabilitative therapies, and educational interventions, many people with hydrocephalus live relatively normal lives.

The symptoms of NPH 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.

What research is being done?

The mission of the National Institute of Neurological Disorders and Stroke (NINDS) is to seek fundamental knowledge about the brain and nervous system and to use that knowledge to reduce the burden of neurological disease. NINDS is a

component of the National Institutes of Health (NIH), the leading supporter of biomedical research in the world. NINDS conducts research and clinical studies to find better ways to prevent, treat, and ultimately cure disorders such as hydrocephalus.

The NINDS helps support the Hydrocephalus Clinical Research Network (HCRN), a collaboration of pediatric neurosurgery centers working together to improve the lives of children with hydrocephalus. The HCRN centers pool their hydrocephalus patient populations to more rapidly study the potential for improved treatments. The HCRN conducts multiple, simultaneous studies at its centers and maintains a substantial registry of patients and procedures.

Cellular mechanisms

Hydrocephalus is a relatively common developmental abnormality, but its underlying mechanisms are not well understood. NINDS supports a wide range of studies that explore the complex mechanisms of normal and abnormal brain development.

For example, NINDS-funded researchers are trying to find gene mutations associated with congenital hydrocephalus. NINDS also supports research exploring how hydrocephalus affects brain nerve networks and brain function.

The early stages of brain development play an important but understudied role in the development of hydrocephalus. NINDS-funded researchers are investigating how signals in the amniotic fluid and early CSF instruct neural stem cell behavior during the pivotal early stages of brain development. This research will

help scientists better understand and diagnose disorders like congenital hydrocephalus.

Other NINDS-funded researchers are examining the cellular mechanisms involved in hydrocephalus in order to help identify hydrocephalus risk factors. The results will potentially improve diagnosis, genetic risk assessment, and treatment.

Diagnosis and treatment

Normal Pressure Hydrocephalus may account for more than 5 percent of all cases of dementia. Unlike most other causes of dementia, NPH can sometimes be reversed with treatment. However, not all surgeries are successful, and researchers do not know why some people respond to treatment and others do not. NINDS-funded researchers are developing new imaging methods to help physicians determine whether shunt surgery is likely to improve the cognitive and motor difficulties that often accompany NPH.

Shunts are the standard treatment of hydrocephalus, but shunts can malfunction, and repeated surgery may be needed. The rate of malfunction of shunts in children is estimated to be 40 percent in the first year after placement, and 10 percent per year after that. Current methods for diagnosing shunt malfunctions are often invasive and expensive. To help solve this problem, NINDS-funded researchers are working to develop a safe, cost-effective method for diagnosing shunt malfunctions using ultrasound.

Not only do shunts malfunction, they also carry a high risk of infection. NINDS-funded scientists are attempting to identify and describe all the microorganisms present in CSF when shunts are

placed, revised, and infected in order to improve prevention and treatment of CSF shunt infection.

More information about hydrocephalus research supported by the NINDS and other NIH Institutes and Centers can be found using NIH RePORTER (projectreporter.nih.gov), a searchable database of current and past research projects supported by NIH and other federal agencies. RePORTER also includes links to publications and resources from these projects.

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
www.ninds.nih.gov

Information also is available from the organizations including:

Hydrocephalus Association
4340 East West Highway, Suite 905
Bethesda, MD 20814
301-202-3811
888-598-3789
www.hydroassoc.org

National Hydrocephalus Foundation
12413 Centralia Road
Lakewood, CA 90715
562-924-6666
www.nhfonline.org

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