

Hydrocephalus Patient Information Guide

Experience Life Without Boundaries




Aesculap Neurosurgery

AESCLAP.



Foreword

A vertical bar on the left side of the page, composed of a series of colored squares. From top to bottom, the colors are: dark blue, medium blue, light blue, very light blue, light green, medium green, dark green, and teal. The squares vary in size and are arranged in a descending staircase pattern.

The intention of this booklet is to provide information to patients, family members, caregivers and friends on the subject of hydrocephalus. The information provided is a general overview of the diagnosis and treatment of hydrocephalus and other conditions associated with hydrocephalus.

About us

History

Aesculap AG (Germany) was founded in 1867 by Gottfried Jetter, a master craftsman trained in surgical instrument and cutlery techniques. Jetter's original workshop, located in Tuttlingen, Germany, is the same location where Aesculap world headquarters resides today. The employee count has increased over the years (3,000+ employees), and the number of instrument patterns has grown from a select few to over 17,000; however, the German standard for quality and pattern consistency remains the same.

Prior to 1977, many of the instruments sold in America by competing companies were sourced from Aesculap in Tuttlingen. In response to the United States customer demand for Aesculap quality surgical instruments, Aesculap, Inc. (U.S.) was established in 1977. Headquartered in Center Valley, Pennsylvania, with over one hundred direct nationwide sales representatives, Aesculap, Inc. currently supports the marketing, sales and distribution of Aesculap surgical instrumentation in the U.S.

In order to support the company's continued growth and provide the service and quality which customers have come to expect, Aesculap relocated the corporate offices from San Francisco to Center Valley, Pennsylvania. This facility houses Aesculap's operations including sales, marketing, communications, customer service, quality assurance, regulatory affairs, human resources and administration. Aesculap's purchasing and distribution is centrally located in St. Louis, Missouri along with our technical services division for repairs.

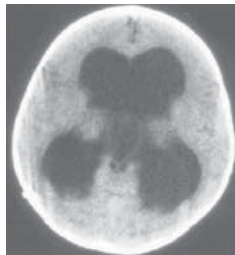
Aesculap has partnered with Christoph Miethke, a German engineer who is responsible for pioneering new technology in the field of hydrocephalus. Together, Aesculap-MIETHKE® offer a variety of hydrocephalus management products that address the different aspects of hydrocephalus.

Vision Statement

Through collaborative excellence we will improve the quality of a patient's life and meet the needs of the changing healthcare environment

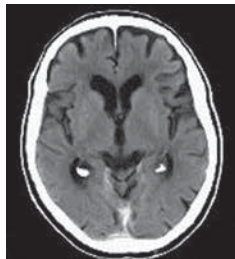
What is hydrocephalus? *

The term hydrocephalus is derived from the Greek words "hydro," meaning water, and "cephalus," meaning head. As its name implies, hydrocephalus 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.



enlarged ventricles

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 that bathes the surfaces of the brain and spinal cord, and then is absorbed into the bloodstream.



normal ventricles

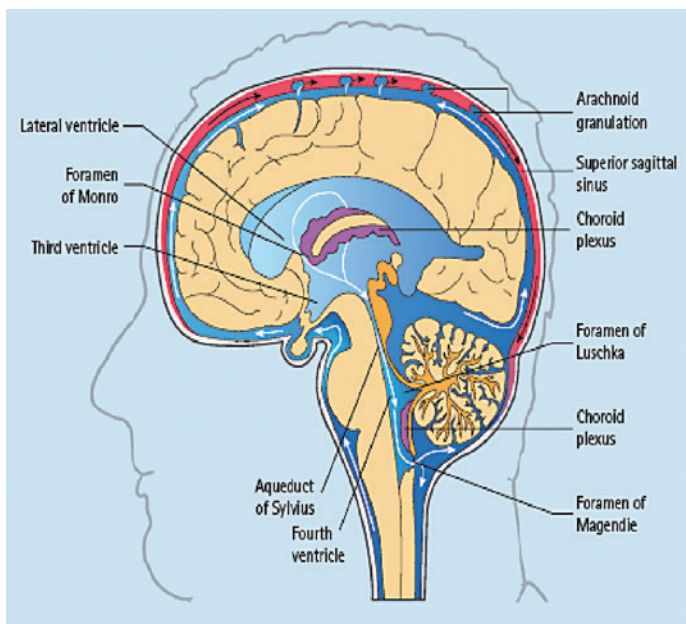
CSF has three important life-sustaining functions:

- Helps the brain tissue remain buoyant acting as a cushion or "shock absorber"
- Delivers nutrients to the brain and removes waste
- Flows between the head and spine to adjust with changes in intracranial blood flow 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 inhibit its normal flow. When this balance is disturbed, hydrocephalus is the result.

Types of Hydrocephalus

- Communicating
 - CSF circulation is intact, but the absorption into arachnoid granulations is blocked
- Non-communicating or Obstructive
 - Circulation of CSF is blocked somewhere between the lateral, third, and/or fourth ventricles
- Normal Pressure Hydrocephalus (NPH)
 - IVP (Intraventricular Pressure) is at “normal levels”
 - Typically affects elderly patients
 - Symptoms: Incontinence, Dementia, Gait Ataxia (loss of balance, shuffling walk)
 - Diagnosed: History, imaging, spinal tap



What you should know *

What causes hydrocephalus?

The causes of hydrocephalus are not all well understood. Hydrocephalus may result from 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 of hydrocephalus?

Symptoms 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 (fibrous joints that connect the bones of the skull) have not yet closed. In infancy, the most obvious indication of hydrocephalus is 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 "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

Vomiting and/or nausea

Papilledema (swelling of the optic disk which is part of the optic nerve)

visual disturbances: blurred vision, diplopia (double vision), and sun setting of the eyes

Difficulty with balance, poor coordination, and gait disturbance

Urinary incontinence

Slowing or loss of development

Lethargy, drowsiness, irritability and or other changes in personality or cognition including memory loss

Normal Pressure Hydrocephalus (NPH)

Symptoms of NPH include progressive mental impairment and dementia, problems with

walking, and impaired bladder control leading to urinary frequency and/or incontinence. They may have a general slowing of movements or may complain that their feet feel "stuck." Because these symptoms are similar to other disorders such as Alzheimer's disease, Parkinson's disease, and Creutzfeldt-Jakob disease, NPH is often misdiagnosed. Many cases go unrecognized and are never properly treated. Doctors may use a variety of tests, including brain scans (CT and/or MRI), a spinal tap or lumbar catheter, intracranial pressure monitoring, and neuropsychological tests to help them diagnose NPH and rule out other conditions.

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

When to talk with your doctor

If you notice any of the signs or symptoms of hydrocephalus, call your or your child's doctor. A high-pitched cry, problems with sucking or feeding, fever, acting lethargic, exhibiting an unwillingness to bend or move the neck or head, breathing difficulties, and seizures are all reasons to seek immediate medical care by calling 911 or other emergency help, or by going to the emergency room.

An older adult with walking difficulties that cannot be attributed to arthritis or injury should have a complete physical and neurological exam. In addition, seek emergency medical care if you or your child experiences a severe head injury.

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. In September 2005, an international team of scientists developed clinical guidelines to help physicians diagnose normal pressure hydrocephalus. The guidelines were published as a supplement to the journal *Neurosurgery* ["Diagnosing Idiopathic Normal-pressure Hydrocephalus," Vol. 57(3), Supplement S2-4--S2-16, 2005].

Treatment *

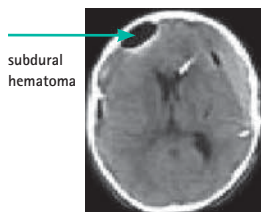
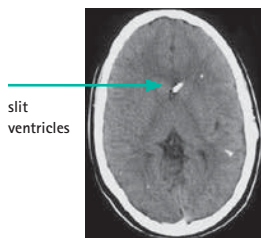
Treatment/Goal/Surgical Procedure

Implanting of a shunt system is a relatively short surgical procedure. The procedure is performed in an operating room using sterile techniques under general anesthesia. A neurosurgeon will perform the procedure where he or she will make an incision on the scalp and a small hole in the skull. This hole will allow access to the ventricle where the catheter will be placed. This catheter will be connected to the valve, which will allow the CSF to drain away from the brain. Lastly, another small incision is made in the abdomen to pass the end of the catheter into the abdominal cavity. Once all of the connections are made, the shunt system will automatically open to drain excess CSF whenever the pressure in the skull exceeds the opening pressure set on the valve.

Complications

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 follow up. When complications do occur, usually the shunt system will require some type of revision.

Some complications can lead to other problems, such as over-draining or under-draining. Over-draining occurs when the shunt allows CSF to drain from the ventricles more quickly than it is produced. This over draining can cause the ventricles to collapse, tearing blood vessels and causing headache, hemorrhage (subdural hematoma), or slit-like ventricles (slit ventricle syndrome). Under-draining occurs when CSF is not removed quickly enough and the symptoms of hydrocephalus recur. 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.



Prognosis *

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 build-up) 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 is, with rare exceptions, 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.

Helpful Websites

- www.aesculapusa.com
- www.ninds.nih.gov/disorders/hydrocephalus.com
- www.hydroassoc.org
- www.nhfonline.org

The Aesculap Shunts



Aesculap-MIETHKE Gravitational Valves Feature:

- Gravitational components which actively vary valve opening pressure to compensate for changing body positions—reducing drainage related discomfort
- Titanium construction allows the valves to be made very small, with thin walls to create large flow paths while retaining strength to protect against subcutaneous pressure
- Ball in cone valve design for superior performance

proGAV System – Adjustable Gravitational Valve System

- Adjustable and Gravitational valves in one system
- 21 precise pressure settings enables the surgeon to make subtle changes in valve pressure*
- *ActiveLock* mechanism prevents the valve's pressure setting from being changed by external fields, such as MRI's**

*Pressure settings from 0-20 cmH₂O in 1 cm increments

**Up to 3 Tesla

GAV and paediGAV—Fixed-Pressure Gravitational Valves

- Combined fixed-pressure valve and Gravitational valve in one system
- Slim, low profile design for improved aesthetics and easy implantation—paediGAV has an ultra-small design for the smallest patients
- Available in a number of different pressure levels to suit the needs of different patients

Shunt Assistant – Gravitational Unit

- Separate Gravitational valve—can be added to any existing shunt system to provide the benefits of an active, gravitational system
- Slim, low profile design for improved aesthetics and easy implantation
- Available in 6 different pressure levels to suit the needs of different patients

Medical Definitions *

Medical Definitions

- **Acquired Hydrocephalus:** Resulting from an outside factor; not inherited nor congenital
- **Aqueductal Stenosis:** A blockage or narrowing of the path from the third to the fourth ventricle
- **Arachnoid Granulations:** Protrusions of the arachnoid membrane of the brain. The arachnoid villi (or granulations) are very similar to a one-way valve, as they allow the CSF to drain from the subarachnoid space into the superior sagittal sinus, where it can be reabsorbed into the blood system
- **Benign External Hydrocephalus** (also referred to as External Hydrocephalus): An accumulation of CSF found on the outside of the brain, which usually presents itself at birth or soon thereafter. Head size will increase, but scans show no internal problems in the ventricles or pathways. Condition corrects itself within about 18 months of age (of the child)
- **Cerebrospinal Fluid (CSF):** The water-type fluid that bathes the brain and spinal cord
- **Cisterns:** Closed spaces that serve as reservoirs at the base of the brain that bathe the surfaces of the brain and spinal cord, and then is absorbed into the bloodstream
- **Communicating Hydrocephalus:** A type of hydrocephalus where the pathways and the ventricular system are not obstructed; however, the problem appears to be that of re-absorption of the used CSF
- **Congenital Hydrocephalus:** A child diagnosed in utero or at the time of birth as having hydrocephalus
- **Diagnosis:** Identification of a disease or disorder by a physician
- **Distal (catheter):** The catheter connected to the shunt valve and directed to the point where the CSF can drain and be re-absorbed

- **Encephalocele:** A hole in the bony covering of the spinal cord through which portions of the brain, spinal cord or meninges may herniate
- **Endoscopic Third Ventriculostomy:** A surgery in which a hole is punctured in the floor of the third ventricle to allow a new pathway for the CSF to be absorbed by the body
- **Hydrocephalus:** Derived from the Greek words "hydro" meaning water and "cephalus" meaning head, a condition in which the primary characteristic is excessive accumulation of fluid in the brain
- **Intraventricular Hemorrhage (IVH):** A bleed within the ventricular system of the brain (also referred to as a brain bleed)
- **Lateral Ventricles:** There are two- one on each side of the brain
- **Lethargic:** Drowsy inactive reluctance or inability to move or respond to stimuli
- **Lumbar Tap (also called Lumbar Puncture and/or a Spinal Tap):** The insertion of a hollow needle to remove some of the CSF, either for pathological testing (i.e. for infection) or to relieve some pressure and see if the patient improves
- **Meningitis:** An infection of the lining of the brain
- **Non-Communicating Hydrocephalus:** Hydrocephalus that is caused by an obstruction in the flow of the CSF
- **Normal Pressure Hydrocephalus:** Type of hydrocephalus where the ventricles are enlarged but there is no obstruction in the ventricular system. The cause appears to be that the CSF is not being absorbed
- **Papilledema:** Swelling of the optic disk, which is part of the optic nerve
- **Prognosis:** Probable outcome of a disease

- **Programmable Shunt:** Type of shunt where the physician can raise or lower the amount of CSF taken from the brain
- **Proximal (Catheter):** A catheter that is placed in the ventricle of the brain
- **Reservoir:** Part of the shunt where the CSF is stored and then released periodically
- **Revision:** When an operation is needed to replace part or all of the shunt
- **Shunt:** A tube implanted in the cranium to balance the flow of cerebrospinal fluid; used in the treatment of hydrocephalus
- **Spina Bifida:** A condition that is present at birth which can affect the development of the back bones, spinal cord, surrounding nerves, and the fluid-filled sac
- **Subarachnoid:** Space where CSF travels over the surface of the brain and the spinal cord
- **Tesla:** Unit of measurement to determine the strength of a magnet
- **Urinary incontinence:** Inability to hold urine in the bladder
- **Valve:** Controls how much CSF is taken from the brain
- **Ventricles:** Fluid filled regions inside the brain
- **Ventriculo-Artial (VA) Shunt:** Type of shunt where the distal catheter tip lies in the right atrium of the heart which allows the CSF to enter the bloodstream directly
- **Ventriculo-Pleural Shunt:** Type of shunt where the distal catheter is located in the pleural space that surrounds the lungs
- **Ventroperitoneal (VP) Shunt:** Type of shunt where the distal catheter is located in the lower part of the abdomen

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