Hydrocephalus

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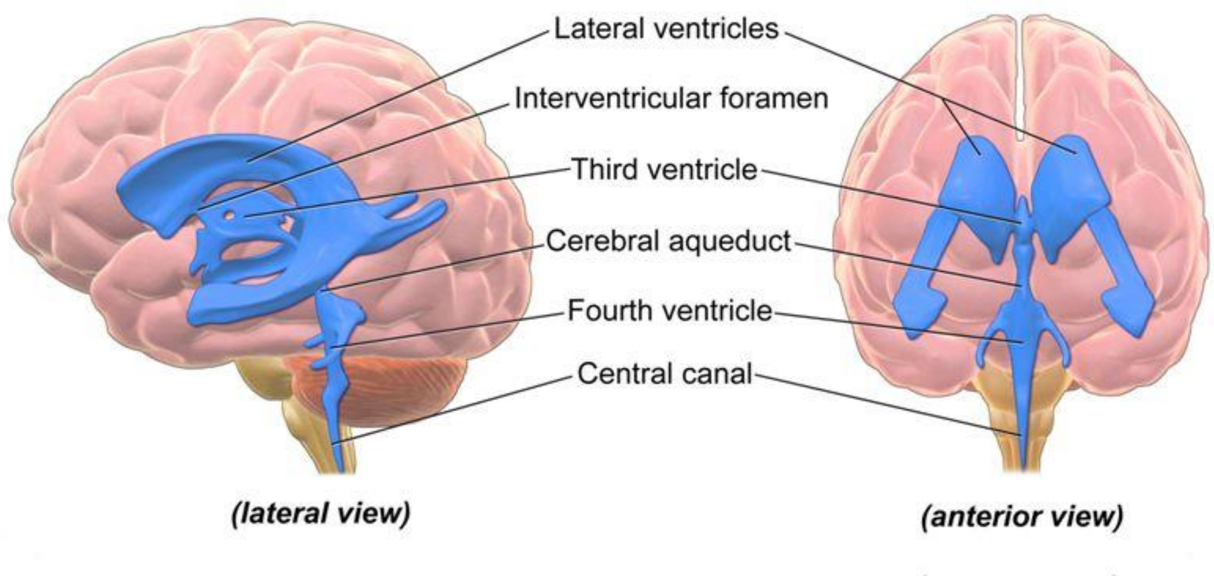
Hydrocephalus comes from the Greek words **hydro** meaning water and **cephalus** meaning head.

Hydrocephalus is an abnormal amount of cerebrospinal fluid (CSF) within venricle cavities in the brain. Cerebrospinal fluid is produced in the ventricles and in the choroid plexus. The CSF circulates through the ventricular system in the brain and is absorbed into the bloodstream. This fluid is in constant circulation. It has many functions: 1.) surround the brain and spinal cord and act as a protective cushion against injury. 2.) It contains nutrients and proteins necessary for the nourishment and normal function of the brain, and 3.) carries waste products away from surrounding tissues.

Hydrocephalus occurs when there is an imbalance between the amount of CSF that is produced and the rate at which it is absorbed. As the CSF builds up, it causes the ventricles to enlarge and the pressure inside the head to increase.

Definition



3D rendering of ventricles (lateral and anterior views).

Who develops hydrocephalus?

Hydrocephalus affects a wide range of people, from infants and older children to young, middle-aged and older adults.

- Over 1,000,000 people in the United States currently live with hydrocephalus.
- For every 1,000 babies born in this country, 1 to two will have hydrocephalus.
- Hydrocephalus is the most common reason for brain surgery in children.
- It is estimated that more than 700,000 Americans have Normal Pressure Hydrocephalus or NPH, but less than 20% receive an appropriate diagnosis.

Classifications and Causes

Hydrocephalus is a condition, not a disease. It can develop for a variety of reasons, sometimes as part of another condition.

Congenital hydrocephalus means the condition is present at birth, that may be caused by a complex interaction of genetic and environmental factors during fetal development. Congenital hydrocephalus is now often diagnosed before birth through routine ultrasound.

Acquired hydrocephalus develops after birth as a result of neurological conditions such as head trauma, brain tumor, cyst, intraventricular hemorrhage or infection of the central nervous system.

Normal pressure hydrocephalus occurs in older adults when the ventricles of the brain are enlarged, but there is little or no increase in the pressure within the ventricles. Sometimes the cause of NPH is known – but most often it is idiopathic, which means the cause is not known.

Common Causes of Congenital Hydrocephalus

Neural tube defect – A common cause of hydrocephalus is a neural tube defect (NTD). An open NTD, where the spinal cord is exposed at birth and is often leaking CSF, is called a myelomeningocele, and is often referred to as spina bifida. This kind of NTD usually leads to the Chiari II malformation, which causes part of the cerebellum and the fourth ventricle to push downward through the opening at the base of the skull into the spinal cord area, blocking CSF flow out of the fourth ventricle and causing hydrocephalus. Please see the National Institute of Neurological Disorders and Stroke (NINDS) for more information on neural tube defects.

Aqueductal stenosis – The most common cause of congenital hydrocephalus is an obstruction called aqueductal stenosis. This occurs when the long, narrow passageway between the third and fourth ventricles (the aqueduct of Sylvius) is narrowed or blocked, perhaps because of infection, hemorrhage, or a tumor. Fluid accumulates "upstream" from the obstruction, producing hydrocephalus.

Common Causes of Congenital Hydrocephalus (continued)

Arachnoid cysts – Congenital hydrocephalus can also be caused by arachnoid cysts, which may occur anywhere in the brain. In children, they're often located at the back of the brain (posterior fossa) and in the area of the third ventricle. These cysts are filled with CSF and lined with the arachnoid membrane, one of the three meningeal coverings. Some arachnoid cysts are self-contained, while others are connected with the ventricles or the subarachnoid space. The fluid trapped by the cysts may block the CSF pathways, causing hydrocephalus.

Dandy-Walker syndrome – In Dandy-Walker syndrome, another cause of congenital hydrocephalus, the fourth ventricle becomes enlarged because its outlets are partly or completely closed and part of the cerebellum fails to develop. Dandy-Walker syndrome may also be associated with abnormal development in other parts of the brain and sometimes leads to aqueductal stenosis. In some instances, two shunts are placed in the child's ventricles — one in the lateral ventricle and another in the fourth ventricle to manage the hydrocephalus. Please see the National Institute of Neurological Disorders and Stroke (NINDS) for more information on Dandy-Walker syndrome.

Chiari malformation – There are two types of Chiari malformation. Both types occur in the bottom of the brain stem where the brain and spinal cord join. The lowest portion of the brain is displaced and is lower than normal pushing down into the spinal column. Please see the National Institute of Neurological Disorders and Stroke (NINDS) for more information on Chiari malformation.

Common Causes of Acquired Hydrocephalus

Intraventricular hemorrhage – An intraventricular hemorrhage, which most frequently affects premature newborns, may cause an acquired form of hydrocephalus. When small blood vessels alongside the ventricular lining rupture, blood may block or scar the ventricles or plug the arachnoid villi, which allow CSF to be absorbed. When the CSF can't be absorbed, hydrocephalus results.

Meningitis – Meningitis is an inflammation of the membranes of the brain and spinal cord. Caused by a bacterial or (less frequently) viral infection, meningitis can scar the delicate membranes called meninges that line the CSF pathway. An acquired form of hydrocephalus may develop if this scarring obstructs the flow of CSF as it passes through the narrow ventricles or over the surfaces of the brain in the subarachnoid space.

Head injury – A head injury can damage the brain's tissues, nerves, or blood vessels. Blood from ruptured vessels may enter the CSF pathway, causing inflammation. Sites of CSF absorption might then be blocked by scarred membranes – meninges – or by blood cells. The CSF flow is restricted, and hydrocephalus develops.

Brain tumors – In children, brain tumors most commonly occur in the back of the brain which is referred to as the posterior fossa. As a tumor grows, it may fill or compress the fourth ventricle, blocking the flow of CSF and causing hydrocephalus. A tumor somewhere else in the brain might also block or compress the ventricular system. Please see the National Institute of Neurological Disorders and Stroke (NINDS) for more information on brain tumors.

Treatment

Shunt System

A shunt is a flexible tube placed into the ventricular system of the brain that diverts the flow of cerebrospinal fluid (CSF) into another region of the body, most often the abdominal cavity, where it can be absorbed. A valve within the shunt maintains CSF at normal levels and pressure within the ventricles.

Shunt Systems

Overview

The management of hydrocephalus has challenged neurosurgeons, neurologists, engineers and medical device developers alike because of the unique nature of cerebrospinal fluid (CSF) dynamics in each patient.

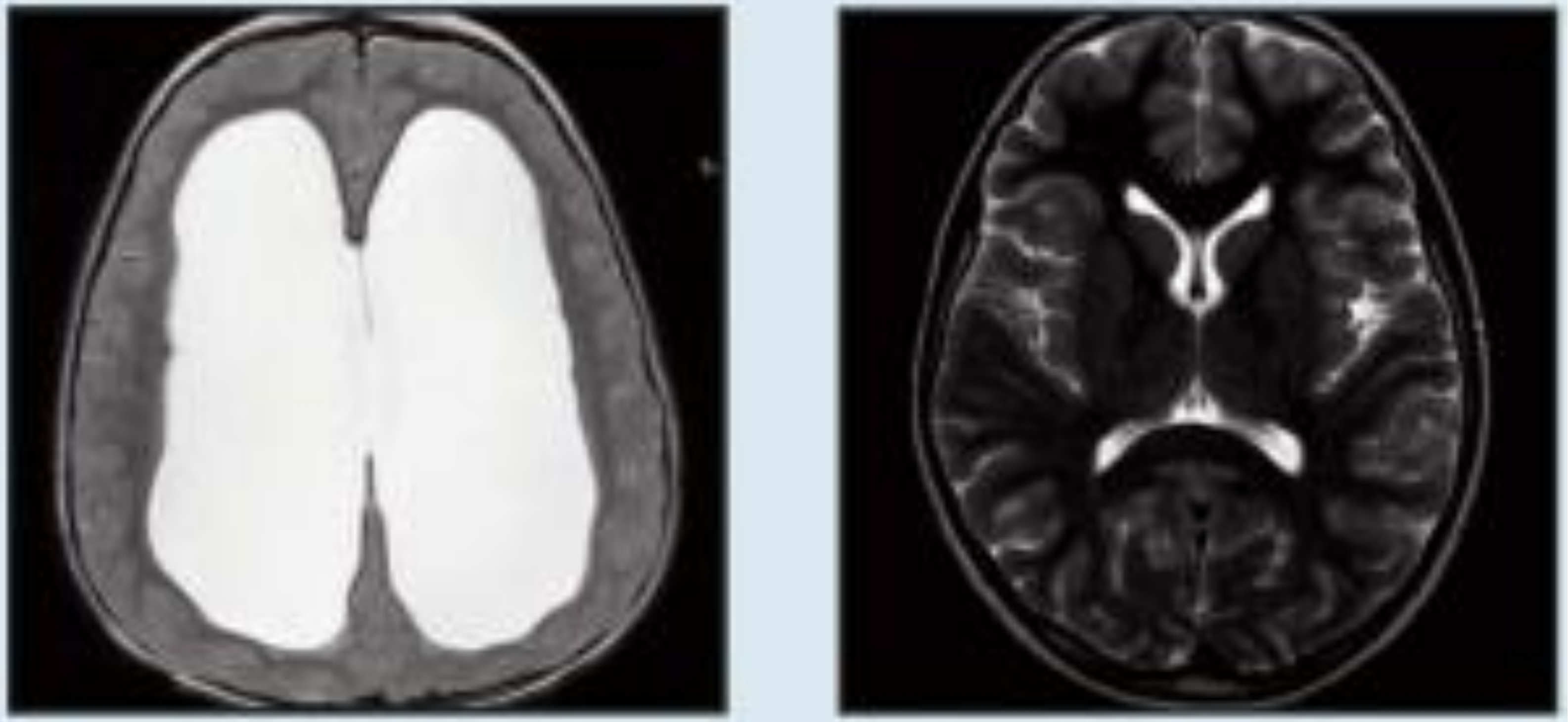
CSF diversion devices or shunts have been used successfully and have become the primary therapy for hydrocephalus treatment for nearly 60 years.

An implanted shunt diverts CSF from the ventricles within the brain or the subarachnoid spaces around the brain and spinal cord to another body region where it will be absorbed.

This creates an alternative route for removal of CSF which is constantly produced within the brain and usually restores the physiological balance between CSF production, flow, and absorption when one or more of these functions has been impaired.

Valves contained within the shunt pathway act like on-off switches, opening when the differential pressure (DP) – i.e., the pressure difference across the valve – exceeds the valve’s opening pressure.

Valves are either set to a fixed pressure or they can be adjustable from outside the body.



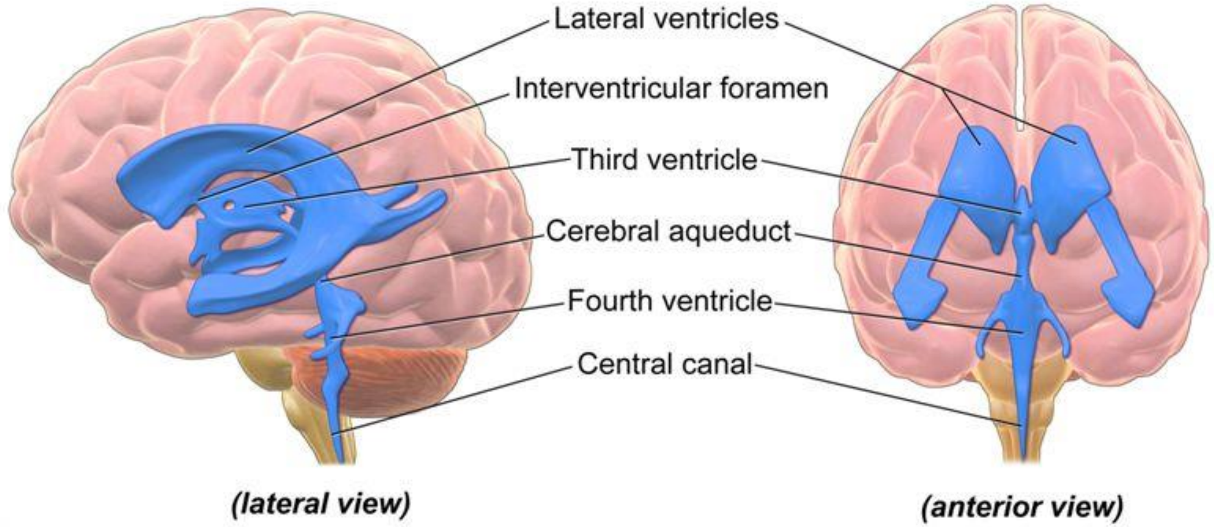
Enlarged ventricles **Normal ventricles**

MRI scans showing the ventricles from the top view.

Shunt System Components

A shunt provides an alternative fluid pathway through which CSF bypasses an obstruction(s) in the fluid compartments of the brain, and acts when CSF absorption is otherwise impaired. Such a bypass relieves the excess fluid buildup that is responsible for hydrocephalus. When CSF production and absorption are in balance, hydrocephalus is considered “compensated”; when out of balance, complications associated with elevated pressure or over drainage occur— causing the signs of a malfunctioning shunt.

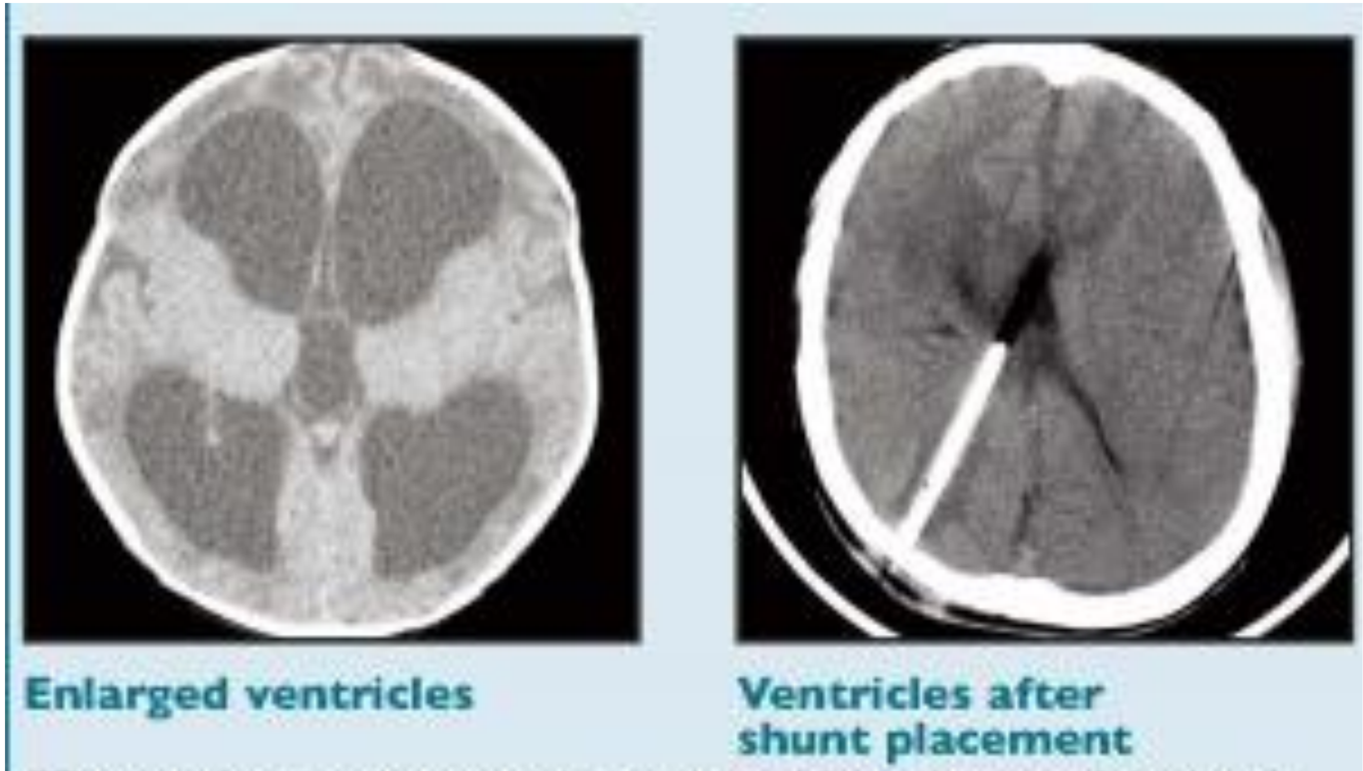
Definition



3D rendering of ventricles (lateral and anterior views).

Symptoms of Shunt Malfunction:

Infants	Toddlers	Children and Adults
•Enlargement of the baby's head •Fontanel is full and tense when the infant is upright and quiet •Prominent scalp veins •Swelling along the shunt tract •Vomiting •Irritability •Sleepiness •Downward deviation of the eyes •Less interest in feeding	•Head enlargement •Vomiting •Headache •Irritability and/or sleepiness •Swelling along the shunt tract •Loss of previous abilities (sensory or motor function)	•Vomiting •Headache •Vision problems •Irritability and/or tiredness •Personality change •Loss of coordination or balance •Swelling along the shunt tract •Difficulty in waking up or staying awake •Decline in academic performance



CT scans showing the ventricles as viewed from the top of the head.

Shunts typically consist of three major components:

An inflow (proximal or closer to the inflow site) catheter:

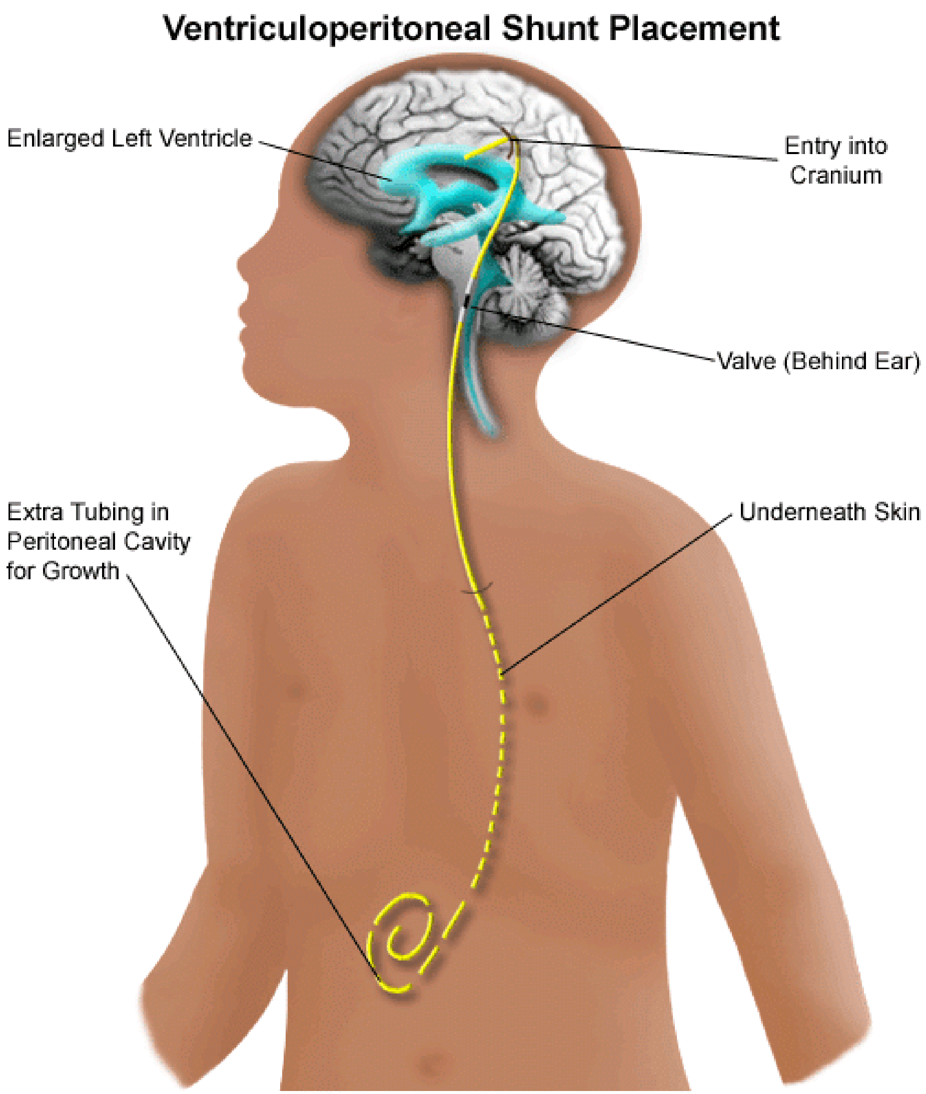
which drains CSF from the ventricles or the subarachnoid space; this tube leaves the brain through a small hole in the skull and then runs for a short distance under the skin.

A valve mechanism:

Which regulates differential pressure or controls flow through the shunt tubing; this device is connected to the proximal catheter and lies between the skin and the skull, usually on top of the head or just behind the ear.

An outflow (distal or farther away from the inflow site) catheter:

which runs under the skin and directs CSF from the valve to the abdominal (or peritoneal) cavity, heart or other suitable drainage site.


Ventriculoperitoneal Shunt Placement
Enlarged Left Ventricle
Entry into Cranium
Valve (Behind Ear)
Extra Tubing in Peritoneal Cavity for Growth
Underneath Skin

Catheters and Tubing

Catheters and tubing are placed at the site where there is an excess volume of CSF is located in the brain. The tubing is routed to a location within the body where the CSF will be absorbed. The Distal end of the catheters are typically placed in the abdominal (or peritoneal) cavity. They may also be placed in the heart, pleural cavity (lungs) and other suitable locations where CSF is drained into the bloodstream.

The type of shunt system is named by the inflow and outflow locations, e.g. if the proximal catheter is in the ventricle and the distal catheter is in the peritoneal cavity it is called a ventriculo-peritoneal (VP) shunt (Table 1).

Shunt tubing is made of flexible silicone, with short plastic tubes used at times as connectors to the valve mechanisms.

Some shunt tubing is impregnated with antibiotics to reduce the incidence of infection during the post-operative period; examples include the Codman Bactiseal™ catheter and the Medtronic Ares™ catheter.

Valve Mechanisms

To assure that the rate of flow through the shunt is controlled, a valve is placed in the tubing system. Some shunt systems are a one-piece design from the manufacturer. Others are “custom-made” by the surgeon in the OR using connectors to add components to the system.

Most valves operate on the principles of change in differential pressure (DP)—the difference between the pressure at the proximal catheter tip and the pressure at the drainage end.

Neurosurgeons select a DP valve based upon the age of the patient, the size of the ventricles, the amount of pressure, and other important clinical factors.

A number of newer shunts can be adjusted non-invasively (i.e. the DP is changed magnetically from outside the body), while others have self-adjusting flow-regulating mechanisms. When such valves are used, a second surgical operation is avoided as the valve’s operating characteristics may be changed non-invasively (programmable valves) or adjusted automatically (flow-regulated valves).

Shunt Malfunction

Shunt malfunction is a partial or complete blockage of the shunt that causes it to function intermittently or not at all. When a blockage occurs, cerebrospinal fluid (CSF) accumulates and can result in symptoms of untreated normal pressure hydrocephalus.

A shunt obstruction from blood cells, tissue or bacteria can occur in any part of the shunt. Both the ventricular catheter – the portion of the tubing placed in the brain – and the distal part of the catheter – the tubing that drains fluid to another part of the body – can become blocked by tissue from the choroid plexus or ventricles. The distal part of the catheter is more often blocked in adults.

Shunts are very durable, but their components can become disengaged or fractured as a result of wear or as a child grows, and occasionally they dislodge from where they were originally placed. More rarely, a valve will fail because of a mechanical malfunction.

Signs and Symptoms of a Complication

Symptoms of shunt malfunction can vary from person to person. It’s important to be aware of what your symptoms were prior to shunt implantation, so that you or your caregiver is able to alert your physician if symptoms return after shunt implantation. Returning symptoms may be a sign of an obstructed or malfunctioning shunt. Symptoms of shunt failure in normal pressure hydrocephalus:

- Difficulty walking/gait disturbances
- Nausea and vomiting
- Cognitive challenges/mild dementia
- Urinary urgency or incontinence
- Swelling along shunt track
- Fever (sign of shunt failure or infection)
- Redness along the shunt tract (sign of shunt failure or infection)

Conclusions

- CSF shunts are commonly used to treat hydrocephalus.
- If left unchecked, the CSF imbalance can lead to elevated intracranial pressure (ICP) which can lead to a variety of complications.
- CSF shunts can be used to alleviate these problems in patients who suffer from hydrocephalus.
- Shunts can come in a variety of forms but all of them consist of an inflow catheter communicating with an outflow catheter under the control of a valve which regulates pressure (differential pressure valves) or controls flow (flow-regulated valves).
- The main differences between shunts are usually in: the materials used to construct them, the types of valves mechanism used, the source of CSF (ventricular, lumbar, etc.) and location of the drainage end-point (peritoneal, atrial, pleural, etc.), andwhether or not the valve is externally adjustable.
- Routine treatment complications are infection, obstruction, and over drainage to name a few.
- Although some (regrettably, the minority) of the patients with shunts can go for years without complications, even those lucky few are potentially one shunt malfunction away from a major crisis. At any time, and without warning, a shunt complication can require emergency intervention.