

Fact Sheet

Shunt Systems for the Management of Hydrocephalus

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. Accessory devices may be added to the shunt to modify valve function; for example, to counter gravitational forces an anti-siphon device may be attached in-line with the valve to minimize over-drainage of CSF when a patient stands up. In addition, a bubble-like reservoir can provide external access to the shunt system for evaluation of CSF or measurements of pressure.

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 overdrainage occur— causing the signs of a malfunctioning shunt.

Symptoms of Shunt Malfunction:

Infants	Toddlers	Children and Adults	Adults with NPH
<ul style="list-style-type: none"> • 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 	<ul style="list-style-type: none"> • Head enlargement • Vomiting • Headache • Irritability and/or sleepiness • Swelling along the shunt tract • Loss of previous abilities (sensory or motor function) 	<ul style="list-style-type: none"> • 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 	<ul style="list-style-type: none"> • Return of symptoms that were present before shunt was placed

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.

Other shunt components may include reservoirs and/or chambers for CSF sampling or injecting medications or dyes, on/off devices, anti-siphon or other flow-compensating devices, or auxiliary catheters to modify performance or adapt the basic system to the patient's specialized needs. In selected cases (such as when cysts or subarachnoid fluid collections are drained), a shunt may not contain a valve or a very low resistance valve may be used.

Catheters and tubing

Catheters and tubing divert CSF from the site where there is an excess volume of CSF to a location within the body where the CSF will be absorbed. The proximal catheter (ventricular or lumbar catheter) drains excess CSF from the ventricles or the spinal lumbar sac through rows of small holes at its origin. Distal catheters are typically placed in the abdominal (or peritoneal) cavity, but 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). At times temporary CSF drainage is performed before a full shunt system is implanted; these short-term drainage systems are called external drains (ventricular or spinal) because the distal catheter is open or drains into a bag outside the body. 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.

Table 1: Most common shunt systems

Shunt Pathway	Shunt Type	CSF Inflow Location	CSF Drainage Location
Ventriculo-peritoneal	VP	Ventricle	Peritoneal cavity
Ventriculo-atrial	VA	Ventricle	Right atrium of the heart
Ventriculo-pleural	VPL	Ventricle	Pleural cavity
Lumbo-peritoneal	LP	Lumbar spine	Peritoneal cavity

Valve Mechanisms

To assure that the rate of flow through the shunt is controlled, a valve is placed in the tubing system. In addition, the function of the valve may be modified with the addition of accessories such as siphon control, gravity compensating devices or flow regulating devices. 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. Sometimes the selected DP range does not adequately address the patient's requirements and a valve with a higher or lower DP range may need to be implanted; these are referred to as "high" or "low" pressure valves, respectively, based on the opening pressure of the valve. 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).

Most commercially available fixed DP shunts are provided in three to five ranges: low, medium or high pressures (and very low and very high), depending on their response to the pressure differential between the shunt's upper and lower ends. Voluntary industry standards on how to measure pressure/flow characteristics, such as those developed by independent or government standards, have not been adopted universally, nor are there industry-defined values for each of the nominal DP ranges. In addition, due to limitations in the technology related to quality control during the manufacturing process, it is not possible to make all shunts of a specific variety (e.g., medium pressure) function within the same range. These factors make the choice of the valve very complicated, and clinicians who are not familiar with the specific characteristics of different valves may have difficulty selecting the best one.

The following shows examples of Shunt Valves:



ProGAV Adjustable shunt

Aesculap Inc.
Toll free: 1-800-282-9000
Website: www.aesculapusa.com



Codman® Certas™ Programmable Valve

Codman, a Johnson and Johnson Company
Toll free: 1-800-225-0460
Website: www.codman.com



OSV II® Flow Regulating Valve

Integra Life Sciences:
Toll free: 1-800-654-2873
Email: custsvcnj@integralife.com
Website: www.integralife.com



Strata Adjustable Valve

Medtronic Neurologic Technologies:
Toll free: 1-800-468-9710
Website: www.medtronic.com



Diamond Valve

Sophysa
Telephone: 219-663-7723
Website: www.sophysa.com



Polaris Adjustable Valve

Currently, there are three basic valve designs:

Slit valves: These relatively old valves are characterized by a cut (slit) in the wall of the end of the distal catheter. Fluid pressure within the lumen of the catheter, if sufficient, will open the slit and allow CSF to flow out of the catheter. The operating pressure characteristics depend on the mechanical properties of the slit, i.e. the stiffness of the silicone, the thickness of the wall, and the length of the slit. Aging of silicone rubber materials and mishandling during surgery may significantly alter the performance of this type of valve.

Diaphragm valves: For these valves, a flexible membrane moves in response to pressure differences. Pressure differentials cause the diaphragm to move, allowing CSF to flow around the diaphragm. When the pressure is reduced (after some CSF flows through the valve), the diaphragm returns to its original position and seals the mechanism.

Spring-loaded, ball-in-cone valves: These valves incorporate a metallic coiled or flat spring that applies a calibrated force to a ball manufactured from a synthetic ruby located in a cone-shaped orifice. When the force (pressure) of CSF gets high enough it pushes the ball against the spring and opens the valve to allow flow. When the fluid pressure is reduced the ball returns to its original position and closes the valve. Ball-in-cone valves are less prone to the effects of the aging of materials than are slit valves, and have been demonstrated to not clog with higher CSF protein levels.

Fixed vs. adjustable valves

Adjustable valves incorporate an external adjustment tool, applied outside the body, to select a DP setting from a range of available settings. The number of adjustable settings depends on the manufacturer. Once adjusted to a specific setting, these valves operate like a fixed pressure valve until re-adjusted by a clinician. Since they are designed to be adjusted by a strong magnetic field, some of these valves may be susceptible to adjustment by strong environmental magnetic fields and **care must be taken to keep toys with magnets and other sources of magnetic fields away from patients with these implanted valves.** Some adjustable shunts incorporate mechanisms that cannot be adjusted by magnetic fields other than those produced by the programmer.

Adjustable valves incorporate a ball-in-cone mechanism regulated by a spring that can be adjusted noninvasively using a magnet. So-called “programmable” valves are DP valves whose opening pressure can be altered using a magnetic field transmitted through the skin. These programmable valves, which can be changed during a routine office visit, are not self-adjusting; DP pressure settings must be reprogrammed until an acceptable setting is reached, which may take several trials. Because siphoning occurs with these systems, some gravity-compensating or anti-siphon mechanism may be needed in selected cases. These mechanisms do not compensate for acute changes brought about by the patient’s daily activities. Also, because they contain an amount of metal, some programmable valves produce artifacts on scans. They also may be reprogrammed in strong magnetic fields such as an MRI and must be checked after being exposed to a strong magnetic field.



Enlarged ventricles



Ventricles after shunt placement

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

Multi-stage flow-regulating valves: These valves maintain the drainage flow rate close to the rate of CSF secretion, regardless of patient position and other conditions that normally promote overdrainage. These valves combine the features of a DP valve with the benefits of a variable flow restrictor.

Antechambers/Reservoirs

An antechamber is a sampling chamber or reservoir located under the skin between the inflow catheter and the valve. They can be used to sample CSF in the shunt, inject drugs into the brain through the proximal catheter, and measure pressure. In addition, these “bubbles” can be felt through the skin and pumped manually to help keep the shunt open. In general, if one pushes on the reservoir and it does not spring back, then there might be an obstruction in the proximal catheter because the reservoir is not filling with CSF. On the other hand, if the reservoir feels rather stiff and more force is needed to depress it, then the distal catheter may be clogged. Some clinicians recommend pumping the reservoir periodically to help keep the shunt open.

Connectors

Short plastic tubes are used to connect catheters with tubing, tubing with valves or accessories, etc. Typically, such connectors require the surgeon to secure them with a suture tie during surgery and are weak points that can become loose and disconnect the catheter.

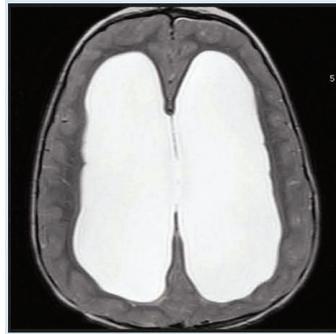
Overdrainage Control Devices

Ideal shunt pressure-flow characteristics must match the patient’s specific needs. However, postural (standing, sitting, or lying down) and vasogenic (blood flow) influences modify shunt function. Standing posture causes a siphon or sucking effect, essentially “pulling” fluid out of the brain or lumbar region when the patient stands. During the night, there are periodic, small increases in blood volume in the head. This nocturnal cerebral vasogenic wave activity occurs several times a night during sleep. Similar blood volume increases occur when the patient strains, as during a cough or a bowel movement. These volume increases, in turn, “push” CSF out of the ventricles through the shunt. The terms “siphoning”, “overdrainage” and “overshunting”, (often used interchangeably) are not the same. Siphoning is a purely postural phenomenon. Overdrainage and overshunting refer to shunting CSF in excess of that required and may be caused by postural and vasogenic effects. A variety of siphon and overdrainage regulating devices have been developed to mitigate the effects of CSF overdrainage. Many major shunting complications are due to shunt overdrainage, including, but not limited to:

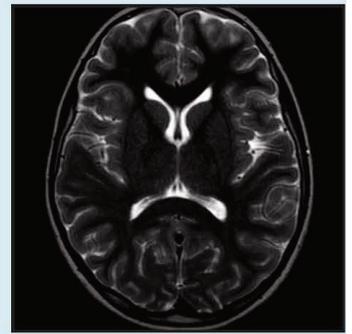
- proximal shunt obstruction, in which tissue (i.e. choroid plexus) can get sucked into the holes of the proximal catheter and reduce inflow,
- headache and dizziness,
- slit (or small) ventricle syndrome, a condition in which there is an absence of CSF within the ventricles combined with a growing brain. This can lead to a situation in which the intracranial pressure or brain pressure can rise to dangerous levels before the ventricles have time to expand.
- subdural collections, or fluid accumulations between the external coverings of the brain (the arachnoid and dura mater membranes).
- extradural collections, or fluid accumulations between the dura mater and the inner surface of the skull
- secondary craniosynostosis, a cranial defect in which the bony sutures (the fibrous joints between the bones of the skull) of an infant close too early, causing problems with normal brain and skull growth.

Siphon-resistive devices (SRD)

These so-called anti-siphon or siphon-control devices incorporate a silicone membrane that closes when conditions favoring postural overdrainage are present. Siphon-resistive devices react to hydrostatic pressure across the two ends of the catheter and close the valve (by increasing the valves opening pressure) when the patient assumes a vertical posture. Such devices tend to create a positive intracranial pressure in the standing position. Communication with atmospheric pressure is the basis for the function of such devices and thus their function can be impaired by the development of scar tissue over the devices diaphragm membrane. Some children may be too short to generate sufficient negative hydrostatic pressure to keep the SRDs closed; positive pressures above the SRD will keep the device open and allow excessive flow. Similarly, if either device is placed in the subcutaneous tissue of the upper neck, then tilting or turning of the neck can change the subcutaneous pressure over the device, causing it to malfunction.



Enlarged ventricles



Normal ventricles

MRI scans showing the ventricles from the top view.

Hydrostatic mechanisms (gravitational devices) typically involve the use of metal balls that fall down into a cone-shaped seat when the patient is standing or in an upright position. When the mechanism is positioned vertically, this adds resistance (depending on the number of balls) to the flow resistance in the shunt flow pathway approximately equal to the height of the hydrostatic column. When the mechanism is horizontal, the balls rest away from the fluid pathway, adding little resistance to the CSF flow pathway.

The difficulty in using such devices lies in choosing the proper valve (available in several resistances) since, in the case of children, growth requires replacement of the gravity compensating mechanism with one with a higher setting, necessitating another operative intervention. Such devices may create issues relative to the ability to evaluate the patients using MRI and creating artifacts on imaging scans.

Flow-regulating devices (FRD) attempt to maintain a constant flow through the valve at different pressures. Valve resistance is increased as differential pressure across the valve is increased. Such valves have mechanisms that vary the size of the opening and associated resistance to CSF flow depending on the differential pressure across the valve. These valves have a lower incidence of early obstruction; the time to revision appears to be longer than that with conventional differential pressure valves (*Sainte-Rose C. Shunt obstruction: a preventable complication? *Pediatr Neurosurg.* 1993 May-Jun; 19(3):156-64.*



Enlarged ventricles



Normal ventricles

Ultrasonounds showing the ventricles as viewed from the top of the head.

What are the complications of shunting?

This important topic is covered in more detail on our website, www.hydroassoc.org, nevertheless, a simple introduction is offered here.

Malfunction

As mechanical devices, shunts may break or become disconnected, migrate (move) or, more commonly, become blocked. Breakage causes a total or partial interruption in the shunt pathway, which may obstruct fluid flow and add resistance to the system. Sometimes a disconnection may occur, but the formation of a scar tissue tunnel around the subcutaneous catheter still allows fluid to flow. Migration may also alter shunt function, causing catheters to move to locations that may restrict flow.

Infection

Shunts may be colonized with bacteria or, more rarely, fungi, most often at the time of shunt implantation. Most of these infections are caused by skin bacteria that enter or surround the shunt during surgical implantation. Infection rates vary from about 1 percent to 15 percent of shunt operations. Recently, some manufacturers have added anti-microbial coatings to the shunt (i.e., the Codman Bactiseal™ or Medtronic Ares™ catheters) which appear to lower the rate of infection during the immediate postoperative period.

Material degradation

Originally, barium sulfate was mixed with silicone to allow shunt catheters to be visible on x-ray. These barium sulfate crystals eventually dissolved, making the tubing surface rough. Tissue in-growth to the roughened surface caused binding of the tubing at specific locations which promoted breakage or deterioration of the tubing. Shunt tubing design has been changed and clear silicone elastomer now covers the surface greatly reducing the possibility of this to occur.

Other complications

The development of seizures has been associated with ventricular catheter implantation. Ventriculoatrial (VA) shunts have been associated with pulmonary hypertension, pulmonary tree embolization and shunt nephritis (an inflammation of the small round filters (glomeruli) located in the kidney).

Rare Complications

Rare complications include intestinal volvulus (twisting) around the shunt catheter, formation of encapsulated intra-peritoneal CSF compartments or development of reactions to the implanted materials.

Shunt revisions

Surgical revisions may be required at any time to correct one or more of the above problems or to compensate for the growth of individuals. Shunt malfunction should be suspected when any symptoms of hydrocephalus return. A neurosurgeon must evaluate the implanted shunt system to determine whether a complete or partial blockage has occurred in the system, the possibility of a disconnection or fracture, or whether the present shunt does not match the patient's physiological requirements. In some patients, **shunt malfunction** is a medical emergency and **must be treated promptly to avoid complications**.

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.), and
- whether or not the valve is externally adjustable.

Routine treatment complications are infection, obstruction, and overdrainage 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.

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Integra Life Sciences

Medtronic Neurologic Technologies

Sophysa

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