Fact Sheet: Cerebrospinal Fluid Shunt Systems for the Management of Hydrocephalus

What Is a Shunt?
A shunt is a surgically implanted device that diverts cerebrospinal fluid (CSF) in a controlled manner away from the central nervous system (CNS) fluid compartments (the ventricles or fluid space near the spine) to an internal delivery site, such as the abdomen or heart. It thus provides an alternative pathway when CSF absorption is impaired, because of obstruction(s) in the CNS fluid compartments or other factors. This relieves the excess fluid buildup that is responsible for hydrocephalus, reducing CSF volume and intracranial pressure (ICP)—the pressure fluid can cause within the skull. Shunting has been used to treat hydrocephalus for nearly half a century.

When CSF production and absorption are in balance, hydrocephalus is considered “compensated.” When production and absorption are out of balance, complications associated with elevated pressure or overdrainage occur—the signs of a malfunctioning shunt.

Shunts typically consist of three components:
• An inflow (proximal) catheter, which drains CSF from the ventricles or the lumbar subarachnoid space, using a tube positioned to transmit CSF to a valve
• A valve mechanism, which regulates differential pressure or controls flow through the shunt tubing;
• An outflow (distal) catheter, which directs CSF from the valve to the peritoneum, heart, or other suitable drainage site

Shunts may also include reservoirs and/or antechambers for sampling or injecting medications or dyes, on/off devices, anti-siphon or other flow-compensating devices, auxiliary catheters to modify shunt performance or adapt the basic system to the patient’s specialized needs, and so on. In selected cases (e.g., when extraventricular fluid collections are drained), a shunt may not contain a valve.

What Is a Valve?
A valve is a mechanical device that either regulates pressure (a differential pressure [DP] valve) or restricts flow (a flow-regulating valve). A valve typically functions as follows:

When the difference between the inlet pressure and the outlet pressure exceeds the opening threshold, the valve opens.
The pressure difference across the valve when it opens is called the “opening pressure”; the pressure difference when the valve closes is called the “closing pressure.” The “operating pressure” is the pressure difference across the valve at a specific flow rate, as tested and specified by the
manufacturer.
Valves’ opening and closing pressures may differ because of the nature of the materials used in valve design. For instance, silicone elastomer (used in slit, diaphragm, and miter valves) behaves differently when opening than it does when closing. Or slit and miter valves may stick, causing large deviations among opening, operating, and closing pressures.
The difference between the pressure/flow curves at a valve’s opening and at its closing is called “hysteresis.” Miter and diaphragm valves demonstrate a larger degree of hysteresis compared to ball-in-cone valves. Depending on individual circumstances, a valve with more or less hysteresis might be more appropriate.

What Is Siphoning/Overdrainage?
Siphoning is the gravity effect of the hydrostatic (fluid) column on valve function when an individual is vertical. When the individual stands or sits, the fluid column height in the outflow catheter changes, and a negative pressure equal to the vertical height of the fluid column in the catheter is produced. (Intracranial pressure [ICP], therefore, can drop into the negative range, but even in persons without hydrocephalus and a shunt, upright ICP is slightly negative.) This gravity effect increases the differential pressure across the valve, keeping it open and allowing more CSF drainage. The effects of siphoning depend on multiple factors, occurring to a greater or lesser degree depending upon the type of valve implanted and present conditions.

Ventricular overdrainage occurs when siphoning happens chronically. Overdrainage may in turn cause the brain to pull away from the inner surface of the skull, tearing the bridging scalp veins and causing associated bleeding and brain compression, debilitating headaches, and slit ventricles.

What Types of Valves are Used to Treat Hydrocephalus?
Most valves used in treating hydrocephalus are DP valves, operating on the principle of change in differential pressure—the difference between the pressure at the proximal catheter tip (inlet?) and the pressure at the drainage end (outlet?). Neurosurgeons select a specific DP valve based upon an individual’s age, the size of his or her ventricles, and other 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 be implanted. A number of newer shunts can be programmed noninvasively (the DP is changed magnetically), while others have self-adjusting flow-regulating mechanisms.

Most commercially available DP shunts are provided in three to five ranges: low, medium, and high pressure (and sometimes very low and very high), depending on a shunt’s response to the pressure differential between its upper and lower ends. The actual values of these arbitrary ranges and the way valves are tested vary from manufacturer to manufacturer. Voluntary industry standards on measuring pressure/flow characteristics, such as those developed by ASTM and ISO, have not been adopted universally, nor are there industry-defined values for each of the nominal DP ranges. In addition, because of limitations in the technology related to quality control during the manufacturing process, it is not possible to make all shunts of a particular variety (e.g., medium pressure) function within the same range.
Types of Differential Pressure (DP) Valves

Slit valves. These valves, made of curved, silicone rubber material, are characterized by a cut, or slit. Flow on the concave side of the valve, if sufficient, will open the slit. The hydrodynamic properties of the valve depend on the thickness and stiffness of the elastomer and the number of slits. Some slit valves incorporate an internal spring to prevent slit inversion, which would lead to reflux (backward flow). These valves’ operating characteristics depend on slit integrity. The aging of silicone rubber materials and mishandling during surgery may significantly alter the performance of slit valves. The Codman Holter Valve, the Uni-shunt, the Phoenix Diamond, the Cruciform, and the CRx are all slit valves.

Duck-bill and miter valves. These valves are characterized by a round orifice that converges into two flat, horizontally opposed leaflets made of silicone. The valve leaflets open when the DP across them increases. These valves operating characteristics depend on the size, shape, thickness, and length of the leaflets. The Integra Lifesciences (Heyer-Schulte) Mischler Valve is a miter valve.

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. Such valves’ opening pressure is defined by the properties of the spring. The size of the valve module opening changes according to DP across the mechanism: the higher the pressure differential, the larger the size of the opening. The ball moves away from the seat as DP increases, under control of the spring, thereby increasing the cross-sectional area through which CSF flows. When DP across the valve decreases, the ball moves toward the seat, reducing the cross-sectional area through which CSF flows.

NMT Neuroscience produces a gravity-compensating lumboperitoneal valve (the HV Lumbar Valve) and a separate Gravity-Compensating Accessory that add resistance when an individual stands through the weight of several stainless steel balls on a ball-in-cone mechanism. Flow is not restricted when an individual is lying down.

Ball-in-cone valves are less prone to the effects of the aging of materials than are miter or slit valves, and they have been demonstrated to handle higher CSF protein levels. Examples of ball-in-cone valves include the Hakim Valve, available from NMT Neuroscience and Codman, a Johnson & Johnson Company, and the Phoenix Accura valves, available from Phoenix Biomedical Corp.

Diaphragm valves. In these valves, a mobile flexible membrane moves in response to pressure differences. The membrane may be held by a central piston that moves in a sleeve, or it may surround the piston and act as an occluder. The membrane can also be a dome that deforms under pressure. These valves’ operating characteristics are based on the stiffness of the silicone rubber diaphragm, which is mounted beneath an integral pumping reservoir and has no metal parts. Pressure differentials cause the diaphragm to move, allowing CSF to flow around it. When pressure abates, the diaphragm seals the mechanism. The Radionics Contour Flex Valve and the Medtronic PS Medical Delta Valve are examples of pressure-regulated diaphragm valves.

Some DP valves include integral Anti-Siphon Devices (ASD) or Siphon Control Devices (SCD). Examples include the Medtronic PS Delta and the Integra Lifesciences Novus valves. Although
their mechanisms vary, the goal is the same: to counteract gravity’s effects on the fluid column below the valve when the patient sits or stands. For example, when pressure within the distal fluid column drops below atmospheric pressure, a diaphragm in the ASD might close the fluid passage, blocking CSF flow through the system, until pressure within the ventricles is again above atmospheric pressure. When this occurs, the diaphragm is pushed back, and flow resumes. The dependence on atmospheric reference, however, may cause such a mechanism to malfunction when tissue fibrosis occurs around it or when an individual’s head presses on it during sleep.

Adjustable and programmable valves. Noninvasively adjustable valves incorporate a ball-in-cone mechanism regulated by a horseshoe-shaped spring that can be adjusted using a magnet.

The resistance of programmable valves, such as those manufactured by Codman and Sophysa (not currently available in the U.S.), can be altered using a magnetic field transmitted through the skin. These programmable valves, whose settings can be changed during a routine office visit, are not self-adjusting; their pressure settings must be reprogrammed until an acceptable level is reached. Because siphoning occurs with these systems, a gravity-compensating or anti-siphon mechanism may sometimes be needed.

The Hakim Programmable Valve includes a programmer and a transmitter. Depending on spring position on a series of steps within the valve, more or less tension is placed on the mechanism’s ball, adjusting DP to one of 18 settings between 30 and 200 mmH2O in 10 mmH2O increments. This mechanism, however, does not compensate for acute changes brought about by an individual’s daily activities. Also, because programmable valves contain metal, they produce artifacts on scans. They also may be reprogrammed in strong magnetic fields such as an MRI.

Multi-stage flow-regulating valves. These valves maintain the drainage flow rate at close to the rate of CSF secretion, regardless of patient position and other conditions that normally promote overdrainage. They combine the features of a DP valve with the benefits of a variable flow restrictor. Examples include the NMT Orbis-Sigma Valve (OSV II) and the Phoenix Diamond Valve. Such valves typically are not used for drainage of extraventricular structures that require a valve with very low operating pressure—for example, a check valve with very little resistance—or no valve at all.

What Are the Complications?
First, some statistics: In the pediatric population, approximately 60 percent of shunts are still functioning one year after implantation. One pediatric study found that the average shunt functions for approximately 5.5 years; some shunts require revision much sooner, others later. For an infant under one year of age, the likelihood of shunt revision during the first year is relatively high—approximately 40 percent. Often, several revisions are required during childhood if hydrocephalus is treated from infancy.

Generally, shunt malfunctions are caused by suboptimal surgical technique, shunt inadequacy, or the unique characteristics of an individual patient’s body. When shunts fail, the highest failure rate is during the acute postoperative period, sometimes because of complications of the surgical procedure itself, such as the presence of blood, debris, etc. As mechanical devices, shunts may
break, migrate (move), or, more commonly, become blocked (occluded). Breakage causes a total or partial interruption in the shunt pathway, which completely or partially obstructs fluid flow and adds resistance to the system. Migration alters shunt function, causing catheters to move to locations that may restrict flow.

As foreign bodies, shunts may become infected—colonized with bacteria or, more rarely, fungi, most often at the time of implantation. Infection occurs in about 1 to 15 percent of shunt operations. More rare complications include intestinal volvulus (twisting) around the shunt catheter, the formation of encapsulated intraperitoneal CSF compartments, and adverse reactions to the implanted materials. Seizures have also been associated with ventricular catheter implantation. Ventriculoatrial (VA) shunts have been associated with pulmonary hypertension, pulmonary tree embolization, and shunt nephritis.

Shunt Revisions
Shunt malfunction should be suspected when symptoms of hydrocephalus return (x-ref here). When a malfunction is suspected, a neurosurgeon must evaluate the implanted shunt system to determine its cause: complete or partial system blockage, the possibility of shunt disconnection or fracture, a mismatch between the individual’s requirements and shunt capabilities. Surgical shunt revisions may be required at any time to correct one or more of these problems or to compensate for individual growth.

Which Companies Manufacture Shunts?
During the past decade, the U.S. shunt industry has been consolidating. Presently, six companies serve the U.S. market:

Codman, a Johnson and Johnson Company, sells several shunt lines: Holter, Medos, Hakim, Accuflow Unishunt, and Denver.
Integra Neurocare produces Heyer Schulte and Novus.
Medtronics PS Medical’s products include the Delta Shunt.
NMT Neuroscience, which now includes Cordis, produces the Orbis-Sigma Valve.
Phoenix Biomedical Corp., which now includes Holter-Hausner, Phoenix Bioengineering, and Theramedics, features the Diamond Valve.
Radionics.

Are There Alternatives for the Management of Hydrocephalus?
Within this decade, neurosurgeons have made advancements in the development of surgical alternatives to implanted shunts for the treatment of hydrocephalus. These alternatives include endoscopic third ventriculostomy for aqueductal stenosis (narrowing of the passageway between the third and fourth ventricles); fenestration (cutting of the walls) of loculated ventricles; and microsurgical removal of tumors to try to restore normal CSF passageways. Despite great improvements in surgical technique, however, the long-term success of endoscopic third ventriculostomy has not been determined. For most people with hydrocephalus, shunt implantation is the only viable therapeutic alternative.
Conclusion
In the medical device industry, rarely has any other product had such a profound effect on patient care. Our understanding of shunts—how they function, how they might function better, which components best suit an individual’s given clinical condition—is constantly improving. However, the goal of most neurosurgeons involved in the treatment of hydrocephalus is to someday be able to provide an alternative to shunting, one without shunting’s relatively high rate of complications. Still, although shunting may not give us a perfect way to manage hydrocephalus, for now, for the majority of patients, it is the best option. And it is improving; neurosurgeons are constantly evaluating the success of various techniques to improve our ability to safely manage hydrocephalus, while shunt manufacturers continue to work on new and improved shunts to address the limitations of existing designs.

References

Patient Information Materials

Hydrocephalus Association

“About Hydrocephalus: A Book for Parents” (in English or Spanish).
“About Normal Pressure Hydrocephalus: A Book for Adults and Their Families.”
“Prenatal Hydrocephalus: A Book for Parents.”

NMT Neurosciences, Inc.

“Just Like Any Other Beagle.” A coloring book on hydrocephalus for children with hydrocephalus and their families. Contact the Hydrocephalus Association or NMT Neurosciences, Inc.: 3450 Corporate Way, Duluth GA, 30096.

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For additional resources about hydrocephalus and information about the services provided by the Hydrocephalus Association, please contact:

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