Siphon regulatory devices: their role in the treatment of hydrocephalus

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Object. The management of hydrocephalus can be challenging because of the unique cerebrospinal fluid (CSF) dynamics in each patient. Various shunt systems have been developed for the treatment of hydrocephalus. One of the main issues surrounding these systems is overdraining due to siphoning. In this paper the authors discuss the pathophysiology of CSF siphoning as well as the various devices used to treat this problem. The pros and cons of each device are discussed, as are the key differences among them. Future concepts are also introduced with an emphasis on upcoming device designs.

Methods. The authors performed a literature review of articles addressing CSF dynamics, shunting, and regulatory devices. The literature consisted of original research articles, company literature on each device, and patent information. A number of siphon regulatory devices have been developed over the past two decades. Each device has a distinct design, requiring specific techniques of implantation for optimal function.

Conclusions. For the past two decades, a variety of siphon regulatory devices have been used to help deal with CSF siphoning. With the increasing mobility of the population, every neurosurgeon will be seeing patients with older and newer devices. Familiarity with the various devices will assist in the evaluation and care of these patients.

KEY WORDS • hydrocephalus • siphon regulatory device • shunt • antisiphoning device • shunt assistant • siphon control device

CEREBROSPINAL FLUID diversionary devices have been successfully used in the treatment of hydrocephalus for more than 50 years. Many of the major complications (proximal shunt obstruction, headache, dizziness, slit ventricle syndrome, subdural or extradural collections, secondary craniosynostosis, cranial nerve palsies, and so forth) are caused by the overdrainage of CSF.

Whereas there are numerous factors that can lead to excessive CSF flow through a shunt, siphoning is a major contributing factor to overdrainage. A shunt diverts CSF from the CNS to another body cavity. Whenever the distal catheter opening is at a level lower than the top of the CNS fluid level, CSF will flow through the shunt, or “siphon,” until the fluid level in the absorbing cavity equilibrates with the level in the CNS. Every positional change is associated with a change in hydrostatic pressure.

Numerous devices have been designed to limit or regulate siphoning (Table 1). Although some of these devices have been beneficial, none has proven to be universally able to prevent siphoning in all patients or to increase the duration of shunt success. One of the earliest descriptions of a device to prevent siphoning was provided by Rudolf R. Schulte, who in 1973 invented a valve that closes to downstream suction. In 1976 Portnoy described a shunt system with an ASD reference to atmospheric pressure, an early ASD. In 1975 Harris and Hakim described the use of weights in a valve to compensate for the pressure drop when vertical. This feature was used for lumboperitoneal shunts as an early H/V valve and was subsequently modified to form the GCA. In 1989, Portnoy described using balls as a downward force to compensate for the hydrostatic force increasing the differential pressure when the patient was in vertical. Schulte and colleagues described an SCD with the improvements of a ring and a composition of dissimilar materials to prevent the silicone membrane from sticking. In 2006 Saul described a method of using a computer to control CSF drainage in the shunt. A battery supplies the energy so that a pressure sensor can send a signal to a microprocessor, which in turn is used to control a solenoid valve.
In this article we discuss both currently available and future SRDs.

CEREBROSPINAL FLUID SIPHONING PATHOPHYSIOLOGIES

If two containers are filled with liquid and the containers are connected with a tube, because of gravity the liquid will flow from the container with the higher fluid level into the container with the lower level until the levels are equal in the two containers. This phenomenon will occur regardless of the shapes of the containers. To maintain two different levels in the connected containers, a valve must be inserted into the connecting tube to balance the hydrostatic difference between the two containers.

Normally, CSF circulates within the craniospinal compartment, being both produced and absorbed within this space. Regardless of the body’s position, CSF and blood are at the same hydrostatic level at any site of absorption within the craniospinal compartment. When CSF is diverted into a body cavity outside this compartment and the body is horizontal, the pleural cavity, peritoneal cavity, or right atrium have approximately the same hydrostatic levels; and when the body is vertical, the absorbing cavity has a lower hydrostatic level than that within the craniospinal compartment and CSF is siphoned down to this lower level.2,14

Normally, when moving between horizontal and vertical body positions, both the CSF and blood are transferred from the cranial compartment to the spinal compartment in a balanced fashion. Hydrocephalus can interfere with the transfer of CSF by obstructing flow to the spinal compartment. The site and degree of CSF flow obstruction can be unique in every patient with hydrocephalus. This variety makes the balanced cotransfer of CSF and blood more complicated and harder to predict. Hence, two individuals with different levels of CSF obstruction can require different shunting systems to accommodate the differential drainage of CSF from the cranial compartment during a change in body position.

The physiological features of any individual point to even more complications when one considers the difficulties in determining not only the vertical level of CSF in the craniospinal compartment but also the fluid level in the absorbing cavity. Thus, it is difficult to calculate the needed vertical hydrostatic difference between the two compartments. As a child grows, these vertical hydrostatic differences change, and in any given individual a change in body weight can result in a change in the vertical hydrostatic difference. Changes in the compliance of either the craniospinal compartment or the absorbing cavity will lead to changes in the vertical hydrostatic difference.

In general, the

<table>
<thead>
<tr>
<th>Manufacturer</th>
<th>SRD</th>
<th>Combination Valve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aesculap</td>
<td>Miehlke Shunt Assistant</td>
<td>ProGAV, GAV, Dual Switch valve</td>
</tr>
<tr>
<td>Codman</td>
<td>Siphonguard</td>
<td>Codman Hakim Programmable valve with Siphonguard</td>
</tr>
<tr>
<td>Integra Lifesciences</td>
<td>Hakim GCA</td>
<td>H-V valve</td>
</tr>
<tr>
<td>Rehr-Schulte Antisiphon Device</td>
<td>Novus valve</td>
<td>Delta valve</td>
</tr>
<tr>
<td>SLD</td>
<td>Equi-Flow valve</td>
<td>Delta valve</td>
</tr>
<tr>
<td>OSV II Smart Valve System</td>
<td>Delta chamber</td>
<td>Accura Elite shunt system</td>
</tr>
<tr>
<td>Medtronic Neurologic Technologies</td>
<td>Strata valve</td>
<td></td>
</tr>
<tr>
<td>Vygon Neuro</td>
<td>Phoenix GCR</td>
<td></td>
</tr>
<tr>
<td>CRx Diamond valve</td>
<td></td>
<td></td>
</tr>
</tbody>
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description of some of these devices, chronologically organized, with a description of each device mechanism, applications for use, device limitations, and some of the studies whose data support the effectiveness of each device. This list of SRDs is by no means complete but serves merely as an introduction to the most widely known devices.

Antisiphoning Device

The Heyer-Schulte Anti-Siphon Device is distributed by Integra Neurosciences. It has two chambers with a pressure-responsive membrane separating the compartments. It is configured so that the area of pressure sensitivity in one of the compartments is greater than that in the other compartment. In the relaxed state, the device has an open pathway between the chambers. When the distal catheter is dependent, the negative siphoning action causes suction on the pressure-sensitive membrane, obstructing the flow or limiting the siphoning. If the pressure in the proximal chamber builds up and is greater than atmospheric pressure, then the proximal fluid pressure can lift up the membrane, restoring the flow of CSF. Early configurations of the ASD were very prone to occlusion by applying gentle pressure on the scalp overlying the device. However, the device was improved by adding an elevated ring around it to minimize the effect of scalp pressure.

Given that the device is referenced to atmospheric pressure, it will easily open when the proximal pressure is greater than atmospheric pressure. While the patient is in a vertical position and if the device is placed at the same level as the opening into the ventricular catheter, then there will be no difference in hydrostatic pressure between the device and the inlet to the shunt. If the device is placed too cephalad and therefore above the opening into the ventricular catheter and the patient is in a vertical position, there will be a hydrostatic pressure difference between the device and the inlet to the shunt so that the ICP will have to be additionally elevated to overcome the added negative pressure distal to the device. If the device is placed too caudal, then when the body is vertical the added hydrostatic column of fluid proximal to the device will keep it open and prevent it from having an antisiphoning effect. When the patient is horizontal and if the device is placed posterior to the ventricular catheter openings, then there is also a hydrostatic column of fluid keeping the device open; given that the body is horizontal, however, this situation is of no consequence.

Siphon Control Device

Medtronic Neurologic Technologies distributes the Strata valve and the Delta valve (Figs. 1 and 2). The major difference between the ASD and the SCD is that the latter is normally closed and remains closed with negative or siphoning distal pressures and opens when the proximal pressure is greater than atmospheric pressure. If the patient is positioned so that the SCD is above the inlet openings of the shunt, then the intraventricular pressure must increase to overcome the hydrostatic difference between the ventricular catheter openings and the level of the SCD.

There are a few limitations with the ASD and SCD. Many children are too short to generate sufficient negative hydrostatic pressure to keep the valves closed, and therefore the positive pressure above the ASD or SCD will keep the device open and allow excessive flow. The original ASD can easily become encapsulated with scar tissue, whereas the SCD has a surrounding elevated ring to prevent encapsulation. Note that encapsulation alters the transference of atmospheric pressure. Both devices are sensitive to pressure over the device; for example, a hematoma in the subgaleal space over the device can lead to a malfunction. 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.

To date, despite the theoretical advantage of these devices, no randomized study data have demonstrated that patients with these devices undergo fewer shunt revisions. Major limitations of many of the previously performed studies have to do with patient age and shunt implantation technique. Findings in pediatric patients may not be applicable to adult patients. Studies in which the authors did not control for the implantation technique—to ensure that the ASD or SCD is placed at the optimal level—might have had a different result if the level of ASD or SCD implantation was considered.

Orbis Sigma Valve

The OSV II Smart Valve System is distributed by Integra Lifesciences. This valve has a pressure-responsive membrane that, in combination with a specially designed piston, varies the size of the opening and therefore resistance to CSF flow depending on the differential pressure across the valve. When a patient’s body is in the vertical position, the valve slows the rate of fluid flow. The very small opening in these valves can make them very susceptible to obstruction by small amounts of blood or de-
bris. This susceptibility is especially problematic in patients undergoing proximal shunt revisions, who frequently can have a small amount of blood and debris released into their ventricles. In contrast, if these valves do not get an early obstruction, then the time to revision seems to be longer than that with conventional differential pressure valves.6

Diamond Valve

Phoenix manufactures the CRx Diamond valve (Fig. 3), which has a diamond-shaped opening that narrows, limiting the flow with increasing differential pressure (such as might result with siphoning). It is very difficult to manufacture the silicone slit precisely so that a reproducible response is achieved with all valves.

Cerebrospinal fluid production increases as the brain grows, and therefore shunt flow requirements increase as a patient matures. A valve with a fixed flow rate must have a rate above 20 ml/hour to accommodate CSF production in the adult. When implanted in infants, both the CRx Diamond valve and OSV allow flow rates in excess of CSF production.

Both the CRx Diamond valve and the OSV follow an S-shaped pressure flow curve. This curve is designed so that, at very low differential pressures, the valves open easily and allow flow. As the differential pressure builds, the valves narrow the opening and thus restrict flow. As a result, the flow rate remains fairly constant over an increasing differential pressure—as the differential pressure exceeds a certain limit, the increase in flow is directly proportional to the increased differential pressure. Examining the idealized flow curves of both of these valves, one sees that both devices allow excessive flow of CSF when a patient is vertical and the hydrostatic difference exceeds 20 cm H₂O (such as when an individual taller than 4 feet is vertical). This finding might explain the inability of these valves to prevent the long-term consequences of siphoning in many patients.

Siphon Guards

Codman & Shurtleff, Inc., distributes the Siphonguard Anti-Siphon Device (Fig. 4). This valve has features similar to those of the OSV and CRx Diamond valve.20 It has two pathways: when the flow increases beyond a certain limit, the low-resistance pathway closes and the high-resistance pathway is open, significantly lowering the flow rate. However, this remaining high-resistance pathway still permits flow in excess of 20 ml/hour, which in most patients is greater than their CSF production rate; hence, overdrainage is still possible.

Gravity Compensating Accessory

The Hakim GCA is distributed by Integra Lifesciences.9 This device has three different settings: low, medium, and high. The ranges are not precise. The valves must be oriented so that they are vertical when the patient is vertical. The valve mechanism involves the use of two sets of three

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Fig. 3. Diagram showing the CRx Diamond Valve, which is manufactured by Vygon Neuro. Image courtesy of Vygon Neuro.

Fig. 4. Schematic demonstrating the Siphonguard, which is distributed by Codman. Image courtesy of Codman.
or four stainless steel balls that fall down, occluding an opening when the valve is vertical. The balls roll out of the way of the opening when the valve (patient) is horizontal. When vertical, the fluid pathway is such that the pressure across the opening must be greater than the weight of the balls to move the balls up, allowing for flow through the valve. These valves can be very effective in increasing the differential pressure across the shunt to compensate for an increased differential pressure change when vertical. The difficulty lies in choosing the proper valve, keeping in mind that as a child grows, a gravity compensating valve with a higher setting may be needed, entailing another operative intervention.

**Shunt Assistant**

Miethke improved on the GCA design by using a tantalum ball and making a miniature valve. Aesculap, Inc., distributes the Shunt Assistant (Fig. 5). It has the advantages of weights that are much more precise and valves that come in settings of 10, 15, 20, 25, 30, 35, and 40 cm H$_2$O. The smaller size allows for easier placement of the valve, and the various settings allow more options to adjust the valve to each patient’s requirements. Still it is difficult to determine before surgery the optimal setting needed in a particular patient, and like most other valves, the setting must be readjusted after MR imaging.

These valves are referenced as follows (cm H$_2$O): opening pressure in the horizontal position/opening pressure in the vertical position. A general guideline is to use 00/10 in a premature or small infant, 00/15 in most infants and small children, and 00/20 in an older child or a small adult. A 00/25 valve is appropriate in most adults, and some thin or tall adults might require an even higher valve setting. When the patient is vertical, a normal ICP can be anywhere between the level of the external auditory meatus to 15 cm below it, and the pressure in the abdomen can be near the top of the abdominal cavity to closer to the level of the umbilicus. Therefore, it is difficult to establish a precise protocol for measuring the optimal shunt assistant for a particular patient. In general, obese patients have higher vertical ICPs and intraperitoneal pressure levels. As a rough guideline, we measure the distance between the angle of the jaw and the tip of the xiphoid process and use the shunt assistant close to or slightly longer than this distance for a thin person and a shunt assistant up to this length for an obese person.

A patient who has never had a shunt seems to tolerate the presence of a shunt assistant better than a patient with a longstanding conventional differential pressure valve. The latter patients can become accustomed to having a valve. These valves are referenced as follows (cm H$_2$O): opening pressure in the horizontal position/opening pressure in the vertical position. A general guideline is to use 00/10 in a premature or small infant, 00/15 in most infants and small children, and 00/20 in an older child or a small adult. A 00/25 valve is appropriate in most adults, and some thin or tall adults might require an even higher valve setting. When the patient is vertical, a normal ICP can be anywhere between the level of the external auditory meatus to 15 cm below it, and the pressure in the abdomen can be near the top of the abdominal cavity to closer to the level of the umbilicus. Therefore, it is difficult to establish a precise protocol for measuring the optimal shunt assistant for a particular patient. In general, obese patients have higher vertical ICPs and intraperitoneal pressure levels. As a rough guideline, we measure the distance between the angle of the jaw and the tip of the xiphoid process and use the shunt assistant close to or slightly longer than this distance for a thin person and a shunt assistant up to this length for an obese person.

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**Gravity Compensating Reservoir**

The Phoenix GCR is distributed by Vygon Neuro. The GCR uses a tantalum ball and comes in five differential pressures: 120, 170, 225, 300, and 370 mm H$_2$O. The ball falls down inside a bur hole reservoir and increases the opening pressure when the patient is oriented in a vertical position. The GCR has the advantage of being incorporated in the tapping reservoir. It requires a frontally inserted ventricular catheter and must be properly oriented on the top of the head.

Ball-type SRDs (GCA, shunt assistant, and GCR) that are placed cranially will be activated when the body is supine and the head is flexed. In contrast, these devices will be deactivated when the patient is vertical when the head is either flexed or extended. Although theoretically this phenomenon might appear to be problematic, in our experience it does not seem to be an issue clinically.

**Combination Valve Therapy**

Theoretically, any differential valve can be used in combination with an SRD. All of the major shunt manufacturers and distributors offer combination valves using their SRD. The Delta valve (Medtronic) is a standard fixed opening-pressure differential valve with an SCD or a Delta chamber. The Strata valve is Medtronic’s programmable valve with the Delta chamber. Codman makes their Codman Hakim programmable valve with or without a siphon guard. Integra Lifesciences distributes an Equi-Flow valve, which is a silicone differential-pressure valve with an SLD. Integra also distributes the Novus valve, which is also a silicone differential-pressure valve with the Heyer-Schulte ASD. Vygon Neuro distributes the Accura Elite shunt system, which is a ball valve with their Phoenix GCR. Aesculap distributes a GAV and Dual.
Switch valve, which are ball valves with shunt assist mechanisms. Soon to be released in the US is the Aesculap Miethke proGAV shunt, a programmable valve with a shunt assistant. The advantage of this programmable valve will be its locking mechanism, which eliminates the need to reprogram the valve after MR imaging. The Sophysa Polaris valve also has a locking mechanism, which prevents the setting from changing during MR imaging but does not yet come preassembled with an SRD (although it is compatible with any stand-alone SRD).

We adjust a patient's horizontal differential pressure by using a programmable valve. For the vertical pressure, a 00/10 shunt assistant is added in a premature or very small infant, 00/15 in infants or small children, 00/20 in larger children or small adults, 00/25 in larger adults, and a 00/30 in taller adults of less than average weight.

The OSV II Smart Valve distributed by Integra Lifeiences minimizes the flow rate between 10 and 35 cm H2O differential pressure. The valve allows a flow rate of close to 20 ml/hour when the differential pressure is less than 20 cm H2O; however, when the differential pressure exceeds 30 cm H2O (as it does in most vertically oriented patients), the flow rate exceeds 25 ml/hour, allowing for overdrainage. Theoretically, adding a 00/10 or even a 00/15 shunt assistant to the OSV II Smart Valve should keep the flow rate close to 20 ml/hour when a patient is vertical. We have tried this combination without success. The problem is that eventually such patients become symptomatic from elevated ICP and not from proximal obstruction. With the GCA and the Shunt Assist there is a graduated opening pressure that varies with the angle of inclination.

There are advantages to using the OSV II, the Diamond CRx, and the Siphonguard. These three valves are not subject to encapsulation, can be placed at any level (unlike the GCA, H/V valve, shunt assistant, and GCR).

FUTURE DEVELOPMENTS

Numerous programmable siphon regulatory valves are being developed. The simplest has two parallel pathways: one with a heavy tantalum ball that can fall down to occlude one of the pathways and the other with a programmable valve that can go from 10 to 45 cm H2O in 5–cm H2O intervals. When an individual is horizontal, CSF flows preferentially through the open pathway as the tantalum ball rolls to the side of the outlet opening. When a patient is vertical, the low-resistance pathway is obstructed and the programmable valve determines the differential pressure. A potential concern with this type of device is that there are only two fixed pressures—one for the horizontal position and the other for the vertical position. If the patient is reclining, the low-resistant pathway can become obstructed and the programmable valve setting could be excessive; it is unknown whether this situation will be a clinical problem. With the GCA and the Shunt Assist there is a graduated opening pressure that varies with the angle of inclination.

Numerous new technologies are being worked on to better control shunt overdrainage or siphoning. Magram has described the incorporation of a starling resistor into a shunt. A starling resistor is a tube that collapses when the distal pressure is less than the external pressure. When the tube is collapsed, flow will flutter and the rate of flow will depend on the rate of fluid production, independent of the differential pressure between the proximal and distal openings. The superficial cervical veins are an example of a starling resistor. When vertical, the veins collapse, preventing overdrainage of blood from the head; therefore, the flow through the veins is dependent on the rate of blood inflow and independent of the differential pressure between the heart and the head.

Computerized shunts are another technology in the making. These would allow flow to be titrated to an individual's needs. With the miniaturization of computers and batteries, these devices will be available in the near future. The computerized shunts will involve a control system, that is, sensors that will detect variables, such as ICP, CSF flow through the tubing, or body position, and send this signal to a computer. The computer will then send a signal either to regulate a valve or to regulate a pump. The ICP signal can be analyzed. As the intracranial compliance decreases, the amplitude of the ICP signal increases. And when the amplitude of the signal reaches a programmed threshold, the computer opens a valve. In essence, the computer will regulate the flow by compliance criteria rather than by mean ICP. If a patient goes from a horizontal to a vertical position and the mean ICP falls or the differential pressure increases across the shunt, the control system can limit the flow until the amplitude of the pressure wave increases to a certain threshold. In this manner, the use of compliance can serve as a method of limiting siphoning. By sensing the pulse pressure through the ventricular catheter, the computer can detect a partial proximal obstruction.

There are many challenges to be overcome before using a computerized shunt. First is the cost. Although any shunt that can prevent a future revision will, in the long term, prove to be cost effective, the initial cost might be excessive in many parts of the world. Furthermore, cost savings to the insurance company that are associated with a decreased incidence of shunt malfunction do not get passed on to the hospital. Therefore, the hospital might be reluctant to approve a more expensive device unless the insurance company covers it.

Computerized systems will need to be individualized and reprogrammed as the patient changes. One of the lessons concerning programmable valves and the addition of various SRDs is that although they can help, they can also be problematic. If a valve is programmed to a setting that is too high or too low, under- or overdrainage can occur. With a computerized shunt, any fixed setting may not be optimal for a particular patient or situation. Intracranial pressure and compliance vary with position; therefore, a computerized shunt will need to have a position sensor to adjust the control pressure or compliance setting, that is, increasing the pressure setting and decreasing the compliance setting when horizontal and visa versa when vertical. How a computerized shunt should change when a patient exercises (heart rate and pulse pressure change) needs to be determined. An algorithm will need to take into account not only the ICP waveform but also various cardiopulmonary factors. Computerized shunts that sense
ICP only might overdrain with physiological increases in cerebral blood flow.

**CONCLUSIONS**

Positional changes are responsible for wide variations in the differential pressure across a CSF shunt valve. Whereas programmable valves allow for an adjustment of the opening pressure, any setting is fixed. For most patients, the range of differential pressure changes that result when moving between a horizontal and a vertical position makes it difficult to select a programmable valve setting that will be acceptable at both positions. Therefore, an SRD can be a useful addition to a programmable valve. Numerous SRDs are currently available. They all require proper positioning to be effective. A familiarity with the various devices will help the neurosurgeon optimize the function of a shunt in a particular patient.

**References**


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