SPINA BIFIDA ASSOCIATION

National Resource Center
jpanlener@sbaa.org
1-800-621-3141 x800
Hablamos español

# Hydrocephalus and Shunts in the Person with Spina Bifida

September 2024

**What is Hydrocephalus?**

Our brains continuously produce a specialized fluid, called *cerebrospinal fluid* (CSF), every minute of every day. Most CSF is produced from blood by specialized cells, called *choroid plexus*, located within fluid-filled pockets of fluid, called *ventricles*, located inside the brain. The brain contains four such ventricles – two lateral ventricles (one located on each side of the brain), and a third and fourth ventricle, both located in the midline. Like a chain of lakes, the CSF travels from one ventricle to another through channels, eventually finding its way to the outside of the brain from the fourth ventricle where it surrounds and cushions the brain and spinal cord. The CSF is eventually returned to the blood through other specialized cells, called *arachnoid granulations*, located within the coverings of the brain.

Under normal circumstances, the amount of CSF that is produced from the choroid plexus is balanced by the amount that is returned to the blood stream at the arachnoid granulations, keeping the amount of CSF within the ventricles steady. *Hydrocephalus* occurs when the brain can't drain the CSF fast enough because there is either a blockage between one ventricle and the next, or the arachnoid granulations are clogged. The CSF builds up, enlarging the ventricles and potentially increasing the pressure in the brain. The enlarged ventricles can be seen on a Computerized Tomographic (CT) or Magnetic Resonance Imaging (MRI) scan.

How much ventricular enlargement is too much? Unfortunately, the answer isn't always clear. In some cases, the answer is obvious – there is a large amount of excess CSF accompanied by symptoms such as headaches and/or vomiting. In others, the amount of excess CSF is small and doesn't appear to be causing any problems. Some people also appear to handle this increased fluid better than others. Because of this, there isn't a specific degree of ventricular enlargement that experts all agree should be treated. Under these circumstances, some neurosurgeons might treat the hydrocephalus whereas others would not.

## Common Treatments

The most common treatment for hydrocephalus is to insert a *shunt* to drain the excess CSF from the ventricles to another place where the body can absorb it back into the blood stream naturally. Shunts have three parts: 1) a short catheter, called a ventricular catheter, that is inserted into a ventricle (most commonly one of the two lateral ventricles), and is attached to 2) a valve that controls the flow of CSF (there are many types) and 3) a longer tube that transports the CSF to another location in the body where it can be reabsorbed back into the blood stream. All shunts are inserted through small incisions and tunneled under the skin; all are therefore internal (meaning they have no external parts on the outside of the body)

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## Types of shunts

The most common type of shunt is the ventriculo-peritoneal (VP) shunt. This shunt drains fluid from the ventricle to the body's abdominal cavity (the cavity in which sit the stomach, intestines, liver, and other organs). Note that the shunt is not inserted into the stomach itself. The CSF within the abdominal cavity is reabsorbed back into the blood stream.

Other, less common, types of shunts include:

- Ventriculo-atrial (VA) shunts that transport CSF directly into a vein, usually in the neck (jugular vein) or under the collarbone (subclavian vein).
- Ventriculo-pleural shunts that move CSF to the chest cavity that surrounds the lungs, from where it is reabsorbed back into the blood stream.

There are several types of shunt valves, all having two purposes: 1) they ensure that the CSF moves only in one direction – from the head to another part of the body; and 2) they help to control how much CSF is allowed to drain. Most shunt valves are constructed to work automatically; the valve opens whenever the pressure in the head reaches a certain level. The pressure at which the valve opens is called its '*opening pressure*'; valves come in a variety of opening pressures that surgeons may choose for various circumstances. Some valves are 'fixed', meaning their opening pressure cannot be changed, whereas others are 'variable' or 'programmable', meaning their opening pressures can be changed (usually using an external magnetic device placed over the valve). Some valves also contain additional devices, called *siphon control devices*, that restrict the CSF that drains when a person is upright (sitting or standing).

It is important to understand that all shunts have been approved by the federal government to have met the same standards, and it's not clear from studies that one shunt clearly works better than another. Surgeons are therefore free to pick shunts that they think are best for their individual patients.

The ventricular catheter is inserted through the brain and into the ventricle through a small incision and burr hole in the skull. The catheter is most commonly placed through one of three sites on the head:

- The top of the head (in an infant this is near the edge of the soft spot)
- Above and behind the ear
- The back of the head

Studies have not determined whether one place works better than another, so where to place the ventricular catheter is also up to what the surgeon thinks is best in each circumstance.

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About 60-80% of people with Spina Bifida require a shunt. Almost all shunts are put in during the first days or weeks after birth; if hydrocephalus is severe at birth, the shunt may be inserted at the time of the initial back closure. A child who doesn't need a shunt by the time they are five months old probably will never need one.

## Signs of shunt problems

The two most common problems encountered with shunts are *malfunction* and *infection*.

Most people with Spina Bifida and shunted hydrocephalus will need the shunt for life. The most common problem with shunts is that they can malfunction, either getting blocked up, breaking, or coming apart. About 30-40 percent of shunts will fail within one year and will need to be revised. Within 5 years, 50-60 percent will fail, and within 10 years, 80-85 percent will fail. About 20 percent of people with Spina Bifida will need more than one shunt revision. Importantly, if a shunt is revised, the odds that it will fail thereafter return to 30-40% in the year following the revision.

Signs of shunt malfunction most commonly include a headache, sometimes, but not always, associated with nausea and/or vomiting. Less common signs of a shunt problem in somebody with Spina Bifida may include:

- Seizures (either the onset of new seizures or an increase in the frequency of existing seizures);
- A significant change in intellect, school performance or personality;
- Back pain at the Spina Bifida closure site;
- Worsening arm or leg function (increasing weakness or loss of sensation, worsening coordination or balance and/or worsening orthopedic deformities);
- Increasing scoliosis;
- Worsening speech or swallowing difficulties.

Infants may in addition have:

- Rapid head growth;
- Full (bulging) or hard (tense) soft spot (also called the fontanelle);
- Unusual irritability;
- Repeated vomiting;
- Newly crossed eyes or an inability to look up;
- Periods in which the baby stops breathing (called apnea);
- Difficulty swallowing;
- A hoarse or weak cry.

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A head ultrasound (only available in infants when the fontanelle is still open), CT or MRI scan will show the CSF build-up as an increase in the size of the ventricles. If available, a baseline scan performed when the shunt was working well (if available) should be compared with the present scan, looking for changes in ventricular size. However, 5-15% of people with Spinal Bifida may have a shunt malfunction with no changes in ventricular size on CT or MRI scan, so vigilance is key; families and health care providers should pay close attention to a person's symptoms, especially if they are similar to those that were present with previous shunt problems. As the symptoms or signs of shunt malfunction can look similar to those of a Chiari malformation or spinal cord tethering, it is important to consider a shunt malfunction as a potential cause of any deterioration in brain or spinal cord function without another clear cause. Finally, some people with shunted hydrocephalus develop a slit (or stiff) ventricle syndrome in which too much CSF drainage leads to very small (or slit) ventricles, the sides of which can temporarily block the ventricular catheter. This can lead to repeated temporary shunt malfunctions without any visible increase in the size of the ventricles.

Shunt infection is a serious complication, complicating between 5 and 10 percent of shunt operations. Shunt infections are higher in babies than in older children and adults. Seventy percent of shunt infections happen within the first two months after a shunt operation, and eighty percent develop within the first six months. Skin bacteria (Staphylococcus epidermidis) are the most common causes. Half of people with shunt infections show signs of a shunt malfunction. Additional signs of an infection may include:

- Fever;
- Neck stiffness;
- Pain, tenderness, or redness along the shunt tract;
- Drainage from the shunt incisions or tract;
- Abdominal pain.

The diagnosis can be confirmed by tapping the shunt - putting a small needle into the shunt and collecting CSF for study. An increase in the CSF white blood cell count suggests an infection; however, some infections may have few or no CSF white blood cells. Sometimes bacteria in the CSF can be seen under the microscope; CSF cultures will usually grow the bacteria within 1-7 days.

Infections are commonly treated with antibiotics *plus* removal and replacement of the entire shunt system. There are two ways of doing this. The first is to remove the entire shunt system and place a temporary external ventricular drainage catheter (EVD) at the beginning of antibiotic treatment; the EVD drains to an external bag. When the course of antibiotic treatment is complete (usually 7-10 days later) the EVD is removed, and a new shunt is placed. This most frequently cures the infection but requires two operations. The second option (if the shunt is working) is to keep the infected shunt in place until

the end of the antibiotic treatment. The old shunt is then removed, and a new shunt inserted. This requires only one operation, but in some studies has been slightly less successful in curing the infection.

## Making decisions

The opinion and experience of the health care provider is very important when working with someone with Spina Bifida and shunted hydrocephalus. When making decisions, here is some helpful advice to families and health care providers:

- Pay attention to parent's (and particularly mom's) perspective about shunt problems — particularly if the symptoms are similar to those that occurred with prior shunt problems.
- Be aware that shunt problems can cause many symptoms that may not, at first glance, appear to be shunt-related.
- Be on the lookout for shunt problems, and make sure the shunt is working before performing other neurosurgical treatments, such as those for Chiari or spinal cord tethering.

*By Mark S. Dias, MD, FAANS*

*Revised by Marks S. Dias, MD, FAANS and Brandon Rocque, MD*

*This information does not constitute medical advice for any individual. As specific cases may vary from the general information presented here, SBA advises readers to consult a qualified medical or other professional on an individual basis.*