

VP shunt

Hydrocephalus and Spina Bifida

Disclaimer: This fact sheet is for education purposes only. Please consult with your doctor or other health professional to make sure this information is right for your child.

Many people living with Spina Bifida also have a condition called Hydrocephalus. Hydrocephalus means “water on the brain.” Hydrocephalus occurs due to a defect at the base of the brain, known as the “Arnold Chiari (Chiari 2) malformation.” This is where parts of the brain push into the top part of the spinal canal. This blocks the normal flow of cerebrospinal fluid (CSF), forcing the fluid to cause pressure in the brain. In babies, the pressure from the fluid can cause the head size to increase. In older children and adults, the head cannot increase as the bones forming the skull have already fused.

What are the effects of Hydrocephalus?

Learning

People living with Spina Bifida associated Hydrocephalus may have difficulty learning. Some of these problems may include difficulties such as:

- Organising
- Planning
- Initiating
- Problem solving and
- Dealing with novel tasks.

These difficulties can impact all kinds of learning and development, including school work, activities at home, and self-care and employment opportunities.

Strategies to support learning include:

- A structured environment with lots of repetition
- Assistance in the classroom or workplace to stay on task.

The Spina Bifida team can assist with individual strategies.

Eyesight

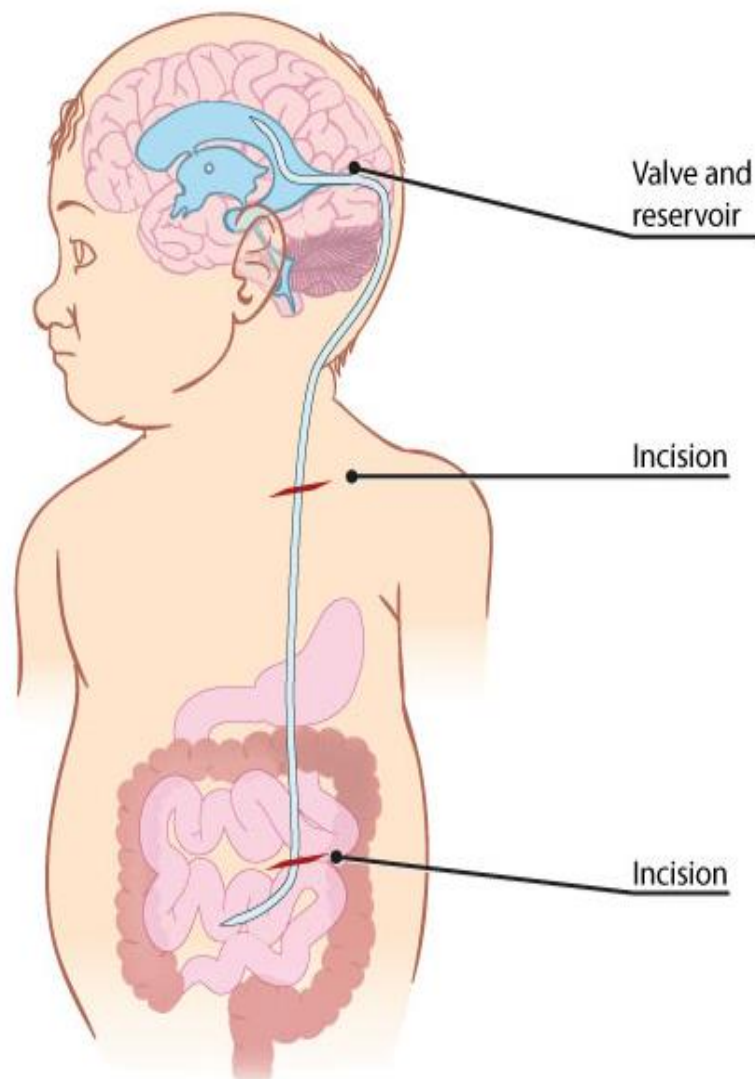
Eye problems can be associated with Hydrocephalus. These might include nystagmus (involuntary rapid movements of the eyes), optic atrophy (degeneration of the optic nerve), and squints (cross-eyed) and reduced eyesight. The development of a squint can also suggest dysfunction in the shunt used to treat Hydrocephalus. Having regular eye tests is also good for monitoring pressure in the head.

Seizures (fits)

People living with Hydrocephalus are at greater risk of having a seizure (or a fit). This can happen at any time throughout their lifetime. If the fits occur regularly, it may be necessary to take medication. An infected ventricular peritoneal shunt can cause a seizure.

How is Hydrocephalus treated?

The increased pressure on the brain caused by Hydrocephalus can be relieved by an operation to insert a Ventricular peritoneal (VP) shunt (common type of shunt). The shunt drains the fluid from the brain to another part of the body. The shunt is a long, thin piece of tubing with a one way valve that takes the fluid from the ventricles in the brain, under the skin to the abdominal cavity. It is completely inside the body, but the outline of the tubing can be seen as it passes down the side of the neck, below the ear. A long coil of tubing is normally placed in the abdomen to allow the shunt to lengthen as the person grows. The shunt does not cure Hydrocephalus, but by draining the excess fluid it can allow the head and brain to grow normally.



What problems can occur with shunts?

A shunt won't work properly if the tubing is blocked, or kinked, breaks and becomes disconnected or if the valve stops working properly. Infection can also cause shunt dysfunction. Shunt infection can occur at any stage, but is more common just after a new shunt has been inserted or after revision of the shunt.

Hydrocephalus is monitored in the outpatient clinic by the paediatrician, rehabilitation specialist and/or the neurosurgeon. A review of the signs and symptoms of shunt dysfunction is carried out at each clinic visit. Having a paediatrician is vital for monitoring of milestones, and a good general practitioner is required into adult life.

The warning signs of shunt dysfunction or infection can include some of the following:

In infants:

- Rapid head growth (in babies).
- Full, bulging or tense soft spot (fontanel) at the top of the head (present until the skull closes at about 18 months of age).
- Swelling and/or pain along the shunt, especially at the side of the neck.
- Unusual irritability
- Nausea and vomiting
- Crossed eyes/sun setting eye
- Periods when the baby stops breathing (apnea) or drowsiness
- Difficulty drinking/swallowing/crying.

In older children:

- Headache
- Visual disturbance (blurred vision)
- Drowsiness
- Loss or decrease of consciousness
- Lethargy.

Less common signs can include:

- Difficulty in walking and changed mobility (due to weakness, balance problems, deterioration in sensation worsening of orthopaedic problems)
- Memory problems or significant change in intellectual functioning
- Decline in milestones
- Seizures (new or worsening or increased frequency of existing seizures)
- Back pain or worsening scoliosis
- Changes in incontinence
- Hearing sensitivities.

Signs and symptoms of an infected shunt:

- Fever
- Neck stiffness
- Redness
- Leakage from the shunt wound area or tenderness along the tract
- Abdominal pain.

Please be aware that shunt malfunction can be life threatening. It is important to know what kind of shunt you have, symptoms of shunt malfunction and where to go for help.

Things to remember:

- There will be a lot of new information.
- Try and keep a list of questions to ask the doctor when you see them.
- Ask your Spina Bifida team if you have any concerns.