

What is Hydrocephalus?

Hydrocephalus was once known as 'water on the brain'. A watery fluid, known as cerebro-spinal fluid or CSF, is produced constantly inside each of the four spaces or ventricles inside the brain.

The CSF normally flows through narrow pathways from one ventricle to the next, then out over the outside of the brain and down the spinal cord. The CSF is absorbed into the bloodstream and re-circulates, and the amount and pressure is normally kept within a fairly narrow range.

If the drainage pathways are blocked at any point, the fluid accumulates in the ventricles inside the brain, causing them to swell and resulting in compression of the surrounding tissue. In babies and infants, the head will enlarge. In older children and adults, the head size cannot increase as the bones, which form the skull, are completely joined together.

What causes Hydrocephalus?

The condition is caused by the inability of CSF to drain away into the bloodstream. There are many reasons why this can happen:

- **Congenital Hydrocephalus**

This means that hydrocephalus is present at birth. It is important to remember that this term does not imply that it is hereditary. Often the exact cause of congenital hydrocephalus cannot be determined

- **Prematurity**

Babies born prematurely are at risk of developing hydrocephalus. The brain of a baby born early is far more vulnerable than one which goes to full term. This is because the premature baby is still developing. The area which lies just beneath the lining of the ventricles in the brain is particularly important – because of the activity in this area it has a very plentiful blood supply. Its blood vessels are very fragile and can easily burst if the baby suffers a large swing in blood pressure or becomes severely ill from other causes.

If these complications do occur, then the baby is at risk of developing a haemorrhage from a rupture of the fragile vessels. This can lead to a blood clot developing, which in some cases is big enough to break through the walls of the ventricle. Should the clot block the flow of CSF, the baby will develop hydrocephalus. The blockage may be temporary or permanent. Even if a blood clot does not develop, the blood cells from the haemorrhage can cause blockage.

- **Spina Bifida**

Most babies born with spina bifida have hydrocephalus.

In addition to the lesion in the spinal cord, there may be abnormalities in the structure of certain parts of the brain which develop before birth. This prevents proper drainage of the CSF.

The increase in pressure due to this can also compress the abnormal parts of the brain even further.

- **Brain Haemorrhage**

Other forms of brain haemorrhage, including those occurring in adults ('stroke'), can result in this type of post-haemorrhagic hydrocephalus.

- **Meningitis**

This is an infection of the membranes covering the brain. The inflammation and debris from this infection blocks the drainage pathways resulting in hydrocephalus. Meningitis can occur at any age, but it is more common in children. The Hib vaccine has drastically reduced the incidence of one form, haemophilus meningitis.

- **Dandy Walker Cysts**

There is a particular group of disorders involving the formation of fluid-filled cysts in the CSF system (for example Dandy Walker Cysts). In these cases, hydrocephalus is often due to pressure on the surrounding tissues by the enlarging cyst.

- **Tumours**

Tumours of the brain cause compression and swelling of surrounding tissues, resulting in poor drainage of CSF. In the treatment of brain tumours, it is often necessary to control hydrocephalus, which might only be temporary.

- **Genetics**

In very rare circumstances, hydrocephalus is due to hereditary factors, which might affect future generations.

- **Other Causes**

There are many other very rare causes of hydrocephalus.

How is Hydrocephalus Treated?

Some forms of hydrocephalus require no specific treatment. Other forms are temporary and do not require treatment on a long-term basis. However, most forms do require treatment, and this is usually surgical.

Drugs have been used for many years, but they may have unpleasant side-effects and are often unsuccessful.

The usual treatment is to insert a shunting device. It is important to note that this does not 'cure' the hydrocephalus, and the injury to the brain tissue remains. Shunting controls the pressure by draining excess CSF, so preventing the condition becoming worse.

Symptoms caused by raised pressure usually improve but other problems of brain injury will remain.

What is a Shunt?

A shunt is simply a device which diverts the accumulated CSF around the obstructed pathways and returns it to the bloodstream. It consists of a system of tubes with a valve to control the rate of drainage and prevent back-flow. It is inserted surgically so that the upper end is in the ventricle of the brain and the lower end leads either into the heart (ventriculo-atrial) or into the abdomen (ventriculo-peritoneal).

The device is completely enclosed so that all of it is inside the body. The fluid, which is drained into the abdomen, passes from there into the bloodstream. Other drainage sites such as the outer lining of the lungs (ventriculo-pleural) can also be used.

Third Ventriculostomy

An alternative treatment may be a third ventriculostomy. However, not all types of hydrocephalus can be treated by this method and it is not always available in all neuro-surgical units. This procedure sees a small hole made in the floor of the ventricle to divert the flow of CSF so that it can be absorbed in the usual way. It is not a new procedure but with the advent of non-invasive techniques (endoscopy) has become

more reliable and is helpful for some patients. This technique is not applicable to everybody with hydrocephalus and is only helpful in certain cases.

Possible Complications

In most cases, the shunts are intended to stay in place for life, although alterations or revisions might become necessary from time to time. The tube or catheter may become too short as the individual grows and an operation might be necessary.

Occasionally, as with any implant, there can be mechanical failure. Also, it is important to be aware that problems can occur with blockage or infection of the shunt.

What Symptoms Should be Looked for?

These vary enormously between individuals. Previous personal experience of a shunt problem is usually a reliable guide as to what to look for.

Possible signs of acute shunt blockage may include:

Vomiting, headache, dizziness, photophobia (sensitivity to light) and other visual disturbances, drowsiness, and seizures.

Possible signs of chronic shunt blockage may include:

Fatigue, general malaise, vision-perceptual problems, behavioural changes, decline in academic performance, being just 'not right' from the carer's point of view.

Medical advice should be sought from your neurosurgical unit if a shunt blockage is suspected, within four hours of acute symptoms occurring.

In shunt infections, symptoms vary with the route of drainage.

In babies, symptoms may be irritability, bulging fontanelle (soft spot), sunset eyes (downward facing eyes). In ventriculo-peritoneal shunts, the symptoms often resemble those of a blockage. This is because the shunt becomes infected and the lower catheter is very often sealed off by the tissue.

There may be accompanying fever and abdominal pain or discomfort.

In ventriculo-atrial shunt infections, fever is present in most cases though often intermittently. Anaemia is frequently present and sometimes skin rashes along with joint pains. In contrast to ventriculo-peritoneal shunts, such infections may not appear for months or years after the operation.

Various tests can be carried out for shunt infections and medical advice should always be sought if an infection is suspected.

How are Shunt Problems Treated?

Shunt blockages which are causing illness usually require an operation to replace or adjust the offending part of the shunt. Shunt infections are usually treated by the removal of the whole shunt and a course of antibiotics before insertion of a new system. Modern approaches to antibiotic therapy mean that such treatment can be expected to succeed, in most cases.

What are the Effects of Hydrocephalus?

There can be learning difficulties associated with hydrocephalus such as problems with concentration, reasoning, and short-term memory. Hydrocephalus can also result in subtle effects such as problems with co-ordination, motivation, and organisational skills. Physical effects such as visual problems, or early puberty in children, may also occur. Many of these effects can be overcome with teaching strategies or treatment where relevant. It must be stressed that the effects of hydrocephalus vary from one individual to another and some people will have very few, if any, problems.

Normal Pressure Hydrocephalus

This is a type of hydrocephalus which occurs in people who are 60 years old and older. It seems to be related to problems of cerebral spinal fluid absorption. The exact problem is unknown.

The condition is treated by the insertion of a shunt and many people with this condition are improved to some extent by the shunt.

Symptoms of normal pressure hydrocephalus are: (Signs and symptoms may take many months or years to develop.)

- **Changes in gait:** The person may feel as if they are frozen on the spot when taking their first step to start walking. They may appear to shuffle rather than walk.
- **Normal thinking process slows down:** The person may respond to questions more slowly than normal, there may be delayed reactions to situations. The individual's ability to process information slows down.
- **Urinary incontinence:** This usually comes after changes in gait.

Idiopathic Intracranial Hypertension

Intracranial hypertension (IH) is a build-up of pressure around the brain. It can happen suddenly, for example, as the result of a severe head injury, stroke or brain abscess. This is known as acute IH. It can also be a persistent, long-lasting problem, known as chronic IH. This is rare and sometimes it's not clear why it happens.

Symptoms: Blind spots, loss of peripheral (side) vision, double vision, temporary blindness, severe headaches.

Diagnosis: Physical exam, medical history, dilated eye exam, vision test, nervous system test, brain imaging, spinal tap.

Treatment: Weight loss through diet and exercise, medicine, surgery.

Hydrocephalus and Epilepsy

Some people with hydrocephalus may develop epilepsy. Epileptic seizures are not due to the hydrocephalus itself, but are usually associated with an underlying cause (meningitis, abnormal development of the brain, neonatal haemorrhage, etc.) As a general rule, seizures in persons with hydrocephalus should be treated in the same way as for those who do not have hydrocephalus.

Support

Spina Bifida Hydrocephalus Ireland (SBHI) offers a number of supports to individuals, families, carers, and professionals through teams such as our Family Support Service. Please see our Family Support Service leaflet for more information.

Contact details

01-4572329

info@sbhi.ie

www.sbhi.ie

Fundraising

SBHI relies on the generosity and support of the public so we can help our 1000 plus members who live with spina bifida and/or hydrocephalus, and thousands of other people who are affected by the conditions including family, friends, carers, and the education and medical professionals who interact with our members on a regular basis.

To donate to SBHI, please visit <http://www.sbhi.ie/donate>; call us on 01 457 23 29, or to donate €4 to SBHI via text message, please text GIVE to 50300.

100% of the text cost goes to Spina Bifida Hydrocephalus Ireland (SBHI) across most network providers. Some providers apply VAT which means that a minimum of €3.25 will go to SBHI.

Customer Service Contact Telephone Number: 076 680 5278. Customer Service Contact Email Address: hello@likecharity.com. Customer Service Website Address: www.likecharity.com

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Spina Bifida Hydrocephalus Ireland

National Resource Centre, Old Nangor Road, Clondalkin, Dublin D22 W5C1

T 01 457 2329 E info@sbhi.ie W www.sbhi.ie

Registered Charity (CHY) 5833 Registered Charity No. 20009366

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