

What is Hydrocephalus?

One of a series of fact sheets produced by the Family Support Service

Introduction

A clear, saltwater-like liquid called cerebro-spinal fluid (CSF) surrounds the brain. This fluid protects and nourishes the brain, carries away wastes from brain cells and contains important chemicals and nutrients. Each day the brain produces about a pint of fluid which flows in a continuous circuit through the brain cavities (ventricles) and over the surface of the brain and spinal cord until it is absorbed by the body.

If the drainage pathways are obstructed 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 that form the skull are completely fused together.

What causes Hydrocephalus?

The condition is caused by the inability of CSF to drain away into the bloodstream.

Hydrocephalus may be communicating or non-communicating:

Communicating hydrocephalus occurs when the flow of CSF is blocked after it exits from the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open.

Non-communicating hydrocephalus – also called obstructive hydrocephalus – occurs when the flow of CSF is blocked along one or more of the narrow pathways connecting the ventricles.

There are many reasons why this can happen:

- **Congenital Hydrocephalus**

This means hydrocephalus is present at birth. It is important to remember that this term does not imply

that it is hereditary. The exact cause of congenital hydrocephalus cannot always be determined.

- **Spina Bifida**

Most babies born with spina bifida have hydrocephalus. In addition to the lesion in the spinal cord, there are abnormalities in the structure of certain parts of the brain that 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.

- **Prematurity**

Babies born prematurely are at risk of developing hydrocephalus. The brain of a baby born early is far more vulnerable than one that goes to full term since it 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 too large a swing in blood pressure or in the amount of fluid in the system. If these complications do occur, then the baby is at risk of developing a haemorrhage from rupture of the fragile vessels. This can lead to a blood clot developing, which in some cases is big enough to break through the wall 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.

- **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 block the drainage pathways resulting in hydrocephalus. Meningitis can occur at any age but it is more common in children. The incidence of one form, haemophilus meningitis, has been drastically reduced by Hib vaccine.

- **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 can be benign or malignant. 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.

- **Genetic**

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 not often successful.

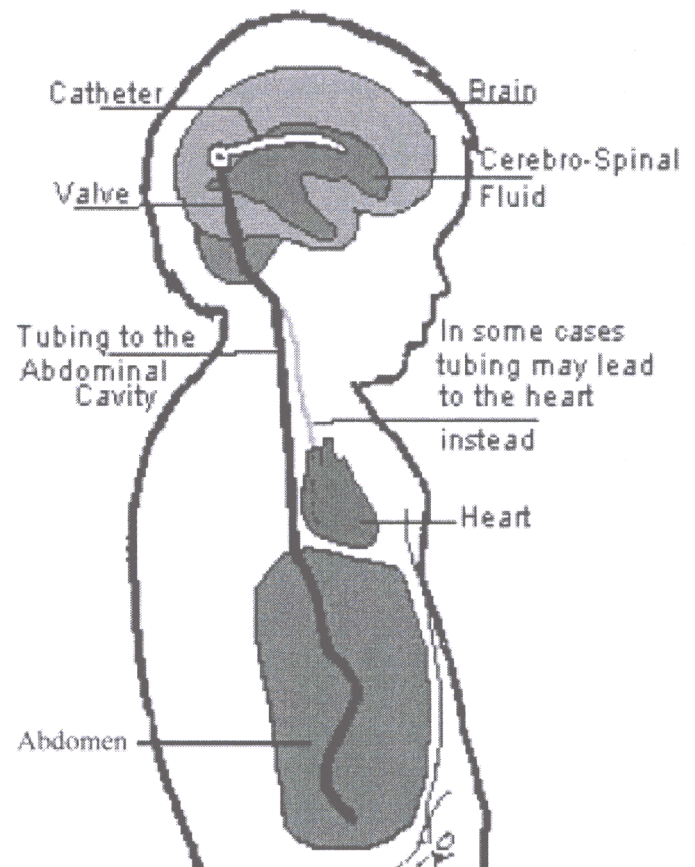
The usual treatment is to insert a shunting device. It is important to note that this does not 'cure' the hydrocephalus and damage to the brain tissue remains. Shunting controls the pressure by draining excess CSF, so preventing the condition becoming worse. Symptoms caused by raised pressure usually improve but other problems of brain damage will remain.

Recently, the technique of third ventriculostomy has been improved by the use of neuro-endoscopes. This treatment, if successful, avoids the need for a shunt. However, not all types of hydrocephalus can be treated by this method.

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



An example of a valve in place

the rate of drainage and prevent back-flow. It is inserted surgically so that the upper end is in a ventricle of the brain and the lower end leads into the abdomen (ventriculo-peritoneal or V.P. Shunt).

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 or the heart can also be used.

In most cases the shunts are intended to stay in place for life, although alterations or changing the shunt (revisions) might become necessary from time to time.

Are there any complications with this form of treatment?

Complications are usually caused either by blockage of the system or by infection. They are only occasionally due to mechanical failure of the valve.

The tube or catheter may become too short as the individual grows and an operation to lengthen it might be necessary.

Shunt Blockage

Symptoms usually develop gradually. In some cases blockage shows itself in a gradual deterioration in overall performance. Occasionally, symptoms are quite sudden and severe and may include headaches and vomiting. Various tests can be carried out to confirm the diagnosis. Medical advice should be sought if a shunt blockage is suspected.

If symptoms worsen rapidly, specialist attention (preferably at your neurosurgical unit) should be obtained.

Shunt Infection

Symptoms vary with the route of drainage. In ventriculo-peritoneal shunts the symptoms will often resemble those of blockage. This is because the shunt becomes infected and the lower catheter is very often then sealed off by tissue. There may be accompanying fever and abdominal pain or discomfort. The most usual timespan for infection to appear is soon after the operation to insert the shunt.

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

Overdrainage

Symptoms of overdrainage are also similar to those of blockage. A severe headache, which is reduced when lying down, is a common symptom.

What symptoms should be looked for?

Whenever there is a possibility that hydrocephalus is causing problems, it is important to seek the correct help immediately.

Possible signs of **ACUTE** shunt blockage or infection may include:

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

Possible signs of **CHRONIC** shunt blockage may include:

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

How are Shunt problems treated?

Shunt blockages that are causing illness usually require an operation to replace or adjust the affected part of the shunt. Shunt infections are usually treated by 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 in most cases can be expected to succeed.

Hydrocephalus and fits

Although people with hydrocephalus sometimes have fits, the fits 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, fits in those with hydrocephalus should be treated in the same way as those that occur in children who do not have hydrocephalus. An epileptic fit is not usually a sign of a blocked shunt.