

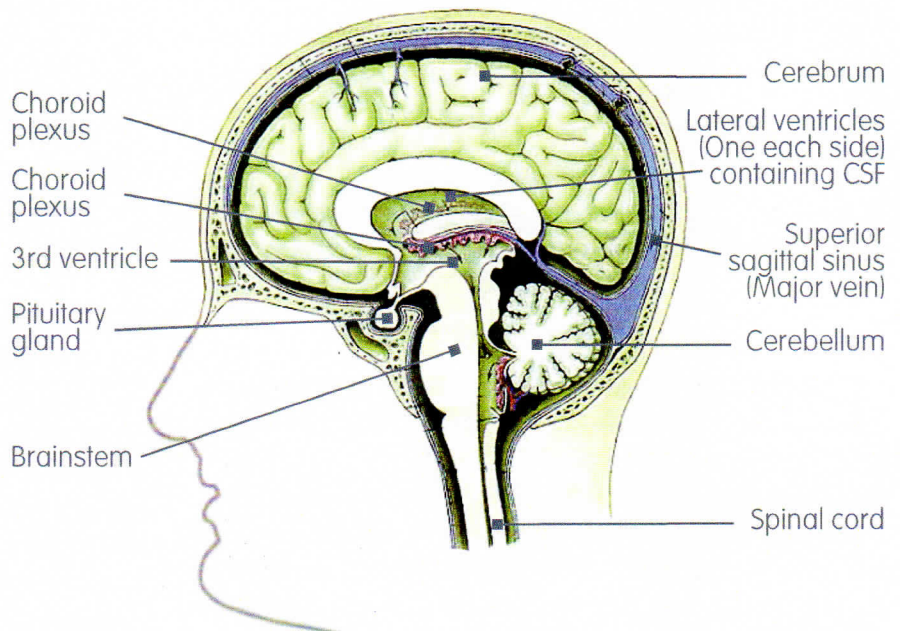
What is Hydrocephalus?

May 2011

Introduction

Hydrocephalus comes from the Greek 'hydro' meaning water and 'cephalie', meaning 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: between 400 and 600mls is produced each day. 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 the amount and pressure are normally kept within a fairly narrow range.

If the drainage of CSF is prevented 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.



Key

Cerebrum: The main part of the brain, in two halves one on each side (left and right hemispheres).

Cerebellum: The smaller part of the brain at the back of the head, involved in muscular coordination.

Ventricles: Connected spaces inside the brain, where CSF is produced. There are two lateral ventricles (one each side), a third ventricle and a fourth ventricle. CSF flows from here over the outside of

the brain before being absorbed back into the bloodstream.

Choroid plexus: The organ that makes CSF.

Superior sagittal sinus: The major vein that runs over the centre of the top of the brain and collects CSF, draining it back into the central bloodstream.

Pituitary gland: A gland situated in the brain cavity but just behind the face. It is very important in secreting

a large number of hormones that regulate body function and development. An example is growth hormone, and others are involved in sexual development.

Brainstem: A very important area connecting the spinal cord to the brain. It controls breathing and other vital functions.

Spinal cord: A complex trunk of nerves transmitting impulses to all parts of the body, allowing muscle movement, sensation and reflexes.

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. *Hydrocephalus can be congenital or acquired.*

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 but known causes can include:

Aqueduct Stenosis

One of the pathways (aqueduct) linking the ventricles in the brain is narrowed (stenosis) which prevents flow of CSF. Although always congenital, symptoms may not occur until sometime later.

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 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. For further information about spina bifida, see ASBAH's Information Sheet: What is spina bifida?

Cysts within the brain

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.

Genetics

In very rare circumstances, hydrocephalus is due to hereditary factors, which might affect future generations, such as x-linked hydrocephalus.

Maternal infections

These include group B streptococcus which can cause neonatal meningitis and toxoplasmosis in the pregnant woman. Both are rare but can cause hydrocephalus.

Acquired Hydrocephalus

This occurs after birth and causes will include:

Brain Haemorrhage

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

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.

(Brain haemorrhage and tumours may also occur congenitally).

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.

Prematurity

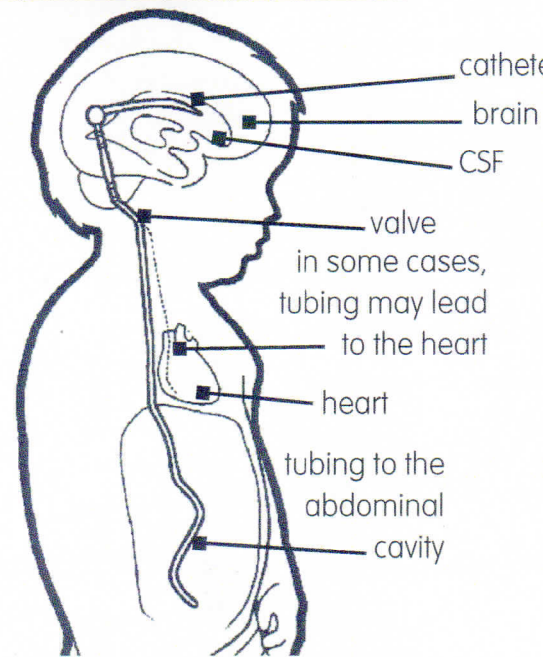
Babies born prematurely are at risk of developing hydrocephalus. The brain of a baby born early is far more vulnerable than one which goes to full term, since it is still developing. The area which lies just beneath the lining of the ventricles in the brain is particularly important – because of the activity in this area it has a 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 becomes severely ill from other causes.

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.

Normal Pressure Hydrocephalus

Seen in older adults and characterised by dementia, decreased mobility and urinary incontinence.

An example of a shunt in place



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.

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 a ventricle of the brain and the lower end leads either into the heart (ventriculo-atrial) or into the abdomen (ventriculo-peritoneal). The shunt may be a programmable (adjustable) type.

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 shunt) can also be used.

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 to lengthen it 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.

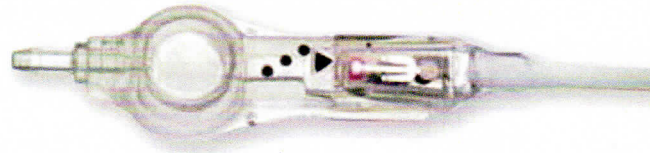
What symptoms should be looked for?

These vary enormously between individuals. Previous personal experience of a shunt problem is often 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 fits.*

Possible signs of chronic shunt blockage may include: *fatigue, general malaise, visual problems, behavioural changes, decline in academic performance, being just 'not right' from the carer's point of view.*

An example of a fixed pressure shunt



If a shunt blockage is suspected, medical advice should be sought from your neuro-surgical unit within four hours of acute symptoms starting.

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

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 then sealed off by tissue. There may be accompanying fever and abdominal pain or discomfort or redness over the shunt site.

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 when they were contracted.

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 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 can be expected to succeed, in most cases.

Endoscopic Third Ventriculostomy

An alternative treatment may be a third ventriculostomy, where a small hole is made in the floor of the ventricle to divert the flow of CSF. This treatment, if successful, avoids the need for a shunt. However, not all types of hydrocephalus can be treated by this method and it is not available in all neuro-surgical units.

For further information about ETV see ASBAH's info sheet Endoscopic Third Ventriculostomy.

Hydrocephalus and seizures

Some children and adults with hydrocephalus will develop seizures.

For information on epilepsy and treatment see the Epilepsy action web site. www.epilepsy.org.uk

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, giving problems with co-ordination, motivation, organisational skills and language. Physical effects such as visual problems, or early puberty in children, may also occur. Many of these effects can be reduced through teaching strategies or with 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.

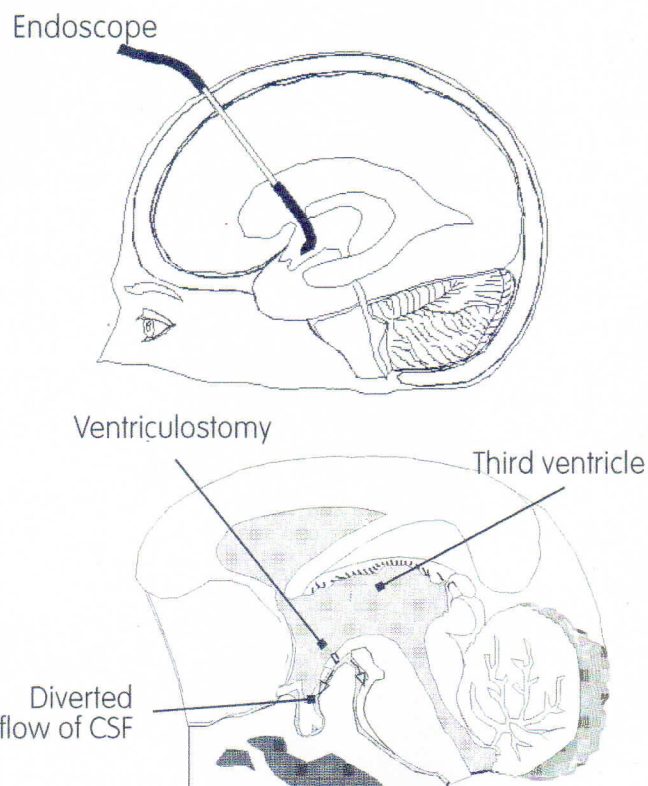
Support

ASBAH plays a leading role in providing support, through national and regional offices and local associations, it offers information and practical advice. Families who have a child or adult with a disability can sometimes feel very isolated and ASBAH can provide often much-needed contact with other families.

Information and publications are available on all aspects of care and management. Our national and regional staff, area and specialist advisers will give personal advice and support whenever possible.

Our services are available to individuals with the disabilities, parents, health professionals, schools, local authorities and other interested organisations. **Further details about our services can be obtained on request from our Helpline Telephone Services 0845 450 7755 between 10.00am & 4.00pm Monday to Friday. Alternatively, you can email your query to helpline@asbah.org**

Third ventriculostomy procedure



SHUNT ALERT CARDS

ASBAH has a series of Shunt Alert Cards, for adults and children. These should be carried at all times by people with hydrocephalus treated by a shunt (this will include some people with spina bifida).

The cards are available from ASBAH, Helpline & information, Telephone **0845 450 7755**.

The card emphasises that, if the cardholder is showing signs similar to those which occur when there is shunt blockage or infection, urgent assessment of shunt function should be carried out in a specialist neuro-surgical unit, in order to eliminate shunt failure as a cause.

People applying for a Shunt Alert Card can also request that an information sheet about hydrocephalus, specially written for doctors, is sent to their GP.

Teachers and employers can ask for "Shunt malfunctions: a guide to symptoms and suggested actions for schools/employers".



Help us

ASBAH relies on people's generosity and support so we can help our clients who depend on us for help and advice - people with hydrocephalus, spina bifida, their families and carers. To donate to ASBAH please visit www.asbah.org or call 01733 421329.