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Hydrocephalus





What is hydrocephalus?

The brain and spinal cord are surrounded by a clear fluid called cerebrospinal fluid (CSF). This fluid is produced and stored in cavities (ventricles) in the brain. Each day the brain produces about a pint of CSF which protects and nourishes the brain, supplies important chemicals and nutrients and carries away waste from the brain cells. Any excess fluid drains away and is absorbed by the body.

Hydrocephalus is a condition where the CSF is unable to drain away through the drainage pathways and fluid accumulates. The ventricles then enlarge to accommodate the extra fluid which causes pressure on different parts of the brain and compression of the surrounding tissue. There are two types of hydrocephalus - "Communicating" which occurs when the flow of CSF is blocked after it exits from the ventricles. It's known as communicating because the CSF can still flow between the ventricles which remain open. "Non-communicating" is also known as obstructive hydrocephalus and occurs when the flow of CSF is blocked after of the narrow pathways connecting the ventricles.



What causes hydrocephalus?

Congenital hydrocephalus

This is where hydrocephalus is present at birth and it is important to know that this does not mean that it is hereditary. Often the exact cause is not known and may not be detected on ante natal scans.

Premature births

The brain of a baby born prematurely is more vulnerable than one at full term as it is still developing. The area that lies just beneath the lining of the brain is particularly important as it has a plentiful blood supply. Its blood vessels are very fragile and can easily burst if the baby has an extreme change in blood pressure or in the amount of fluid in the system. If these occur, the baby is at risk of developing a haemorrhage from rupture of the fragile vessels. This can lead to a clot forming which in some cases is big enough to break through the wall of the ventricle and block the flow of CSF. This blockage can be temporary or permanent and even if a blood clot does not develop, the blood cells from the haemorrhage can cause blockage.

Spina bifida

Around 80% of babies born with spina bifida have hydrocephalus due to a malformation of the brain structure called the Arnold Chiari Malformation.

This develops before birth and prevents proper drainage of the CSF.



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Meningitis

This is an infection of the membranes covering the brain and the inflammation and "debris" from this infection block the drainage pathways resulting in hydrocephalus. Meningitis can occur at any age but is most common in children.

Tumours

Tumours can be benign or malignant and can cause compression and swelling of surrounding tissues resulting in poor drainage of CSF, which may result in hydrocephalus.

Genetic causes

In some rare circumstances, hydrocephalus can be inherited and if there is a history of hydrocephalus in the family it is advisable to discuss this with a Geneticist to determine the risk of it recurring.

Dandy Walker Syndrome

This is a congenital brain malformation involving an area at the back of the brain which controls movement (cerebellum) and the fluid filled spaces surrounding it. Common features of this syndrome are:

- An enlargement of the fourth cavity (ventricle) in the brain which allows fluid to flow freely between the upper and lower areas of the brain and spinal cord.
- A partial or complete absence of the area of the brain responsible for coordination of voluntary muscle movements.
- Fluid-filled cysts near the internal base of the skull which can cause pressure on the surrounding tissues.



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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 the most common form of treatment is by the insertion of a shunt or valve into the brain (diagram on the next page) which is carried out under a general anaesthetic. This is a small device consisting of a one-way valve inserted in the brain. This is then attached to a thin silicone tube (catheter) which is placed just under the skin and ends in the abdominal cavity. This system diverts the accumulated CSF around the blockage and allows it to drain via the end of the catheter into the abdominal cavity where it is absorbed into the bloodstream. It is important to note that this does not "cure" hydrocephalus but prevents the condition becoming worse and any symptoms caused by the raised pressure usually improve.

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Ventriculoperitoneal Shunt Placement

Endoscopic Third Ventriculostomy

A procedure called Endoscopic Third Ventriculostomy (ETV) may avoid the need for a shunt. This is carried out under a general anaesthetic and involves a small perforation made in the floor of the third ventricle using a small instrument called an endoscope, allowing the CSF to escape and therefore avoiding the need for a shunt. However not all types of hydrocephalus can be treated by this method and should be discussed with your Neurosurgeon.



Shunt Alert Cards

If you do have a shunt operation, Spina Bifida Hydrocephalus Scotland can provide you with a free medical alert card to keep with you at all times. This contains useful information for both you, your doctor and any medical staff who may need to care for you. Call SBH Scotland on 03455 211 300 to request your card.



If you have questions or would like further information, please call the **SBH Scotland Helpline** on **03455 211 300** or email **support@sbhscotland.org.uk** For general enquiries call **03455 211 811** or visit **www.sbhscotland.org.uk**

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