Ventriculomegaly

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of ventriculomegaly and hydrocephalus and where to get help.

Ventricles are cavities within the brain filled with cerebro-spinal fluid (CSF) acting as a ‘cushion’. CSF also supplies nutrients to the brain. The brain has four ventricles: two lateral ventricles, the third ventricle and the fourth ventricle. CSF is created within the brain and flows from the lateral ventricles into the third ventricle. It then flows through a narrow tube (the cerebral aqueduct) into the fourth ventricle which lies towards the base of the brain. From the fourth ventricle, it flows around the spinal cord and over the surface of the brain before being re-absorbed.

Ventriculomegaly is the medical term used to describe enlargement of the ventricles of the brain. Hydrocephalus is the term used when enlargement of the ventricles has been caused by an increase in the pressure of the CSF within them.

Without signs of increased pressure in the brain (hydrocephalus), ventriculomegaly most likely will not cause any problems. However, it can be linked with hydrocephalus and other problems. Ventriculomegaly can be diagnosed during pregnancy and occurs in around two per cent of all pregnancies.

What causes ventriculomegaly?

In many cases, we do not know what causes ventriculomegaly (in the absence of any raised CSF pressure) but it can occur if there has been brain damage for any reason leading to loss of brain tissue. Often however it is a “chance” finding and when the ventricles are only a little enlarged of little significance.

What causes hydrocephalus?

Hydrocephalus can be caused by obstruction of the flow of CSF either inside or over the surface of the brain. Structural abnormalities can interfere with the flow of CSF through the brain as the passages between the ventricles are very small and easily blocked. Infections (like meningitis) or bleeding within the head may also block the CSF pathways over the surface of the brain.

Hydrocephalus can sometimes be associated with other congenital (present at birth) conditions, such as agenesis of the corpus callosum, spina bifida and heart defects. The doctors will examine your child closely to check if this is the case.
What are the symptoms of hydrocephalus?
Symptoms and signs of hydrocephalus (one of the causes of raised pressure in the head) include (in babies) a bulging fontanelle (the “soft spot” on the top of the head), increasing head size (its circumference as measured with a tape measure), vomiting, crying and irritability and lack of responsiveness.

How are ventriculomegaly and hydrocephalus diagnosed?
Enlargement of the ventricles may be discovered during pregnancy on routine ultrasound scans. As well as regular ultrasound scans to monitor the ventricle size, magnetic resonance imaging (MRI) scans may also be suggested as these are generally safe in pregnancy (no x-rays). Additional tests may be suggested to try to discover the cause of the ventriculomegaly although this may not always be identified. These may include blood tests and amniocentesis to check for genetic problems. Babies with ventriculomegaly can often be born normally at term, with a caesarean section only required if there are other problems or baby has a very large head.

After birth, babies have a full review including further imaging scans such as ultrasound scans, computerised tomography (CT) and MRI scans to determine whether the enlargement of the ventricles is due to increased CSF pressure within them (hydrocephalus) in which case some form of surgical treatment may be needed.

What is ventriculomegaly treated?
Treatment is usually only required if a baby shows signs of developing hydrocephalus. The aim of treatment is to reduce the pressure on the brain by draining away the CSF.

In most cases, the fluid is drained away using a ‘shunt’, which is a long tube made of silicone. It is inserted into the ventricles in an operation and drains the CSF away to either the abdomen or, more rarely, the heart. The shunt contains a valve so that the fluid can only drain away from the head at the desired rate. A different operation called ventriculostomy may be performed in specific cases, which drains the CSF away from the ventricles without the need for a shunt.

What happens next?
If there are no clinical or radiological (scans) signs of raised pressure or increase in the ventriculomegaly children may not require any further medical intervention. Children with hydrocephalus will need life-long monitoring to make sure that the pressure on the brain remains normal. Shunts can become infected or blocked so any symptoms of raised pressure such as headaches or vomiting should be monitored immediately. Often a shunt will need to be replaced several times during a person’s lifetime. Only a small proportion with ventriculomegaly will have any level of developmental delay and this is usually owing to an associated condition rather than the ventriculomegaly. If a genetic cause for the ventriculomegaly is suspected, genetic counselling may be suggested for future pregnancies.

Further information and support
SHINE can offer advice and support for parents of children with hydrocephalus. Call them on 01733 555 988 or visit their website at www.shinecharity.org.uk

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