What is **Hydrocephalus?**

Shine
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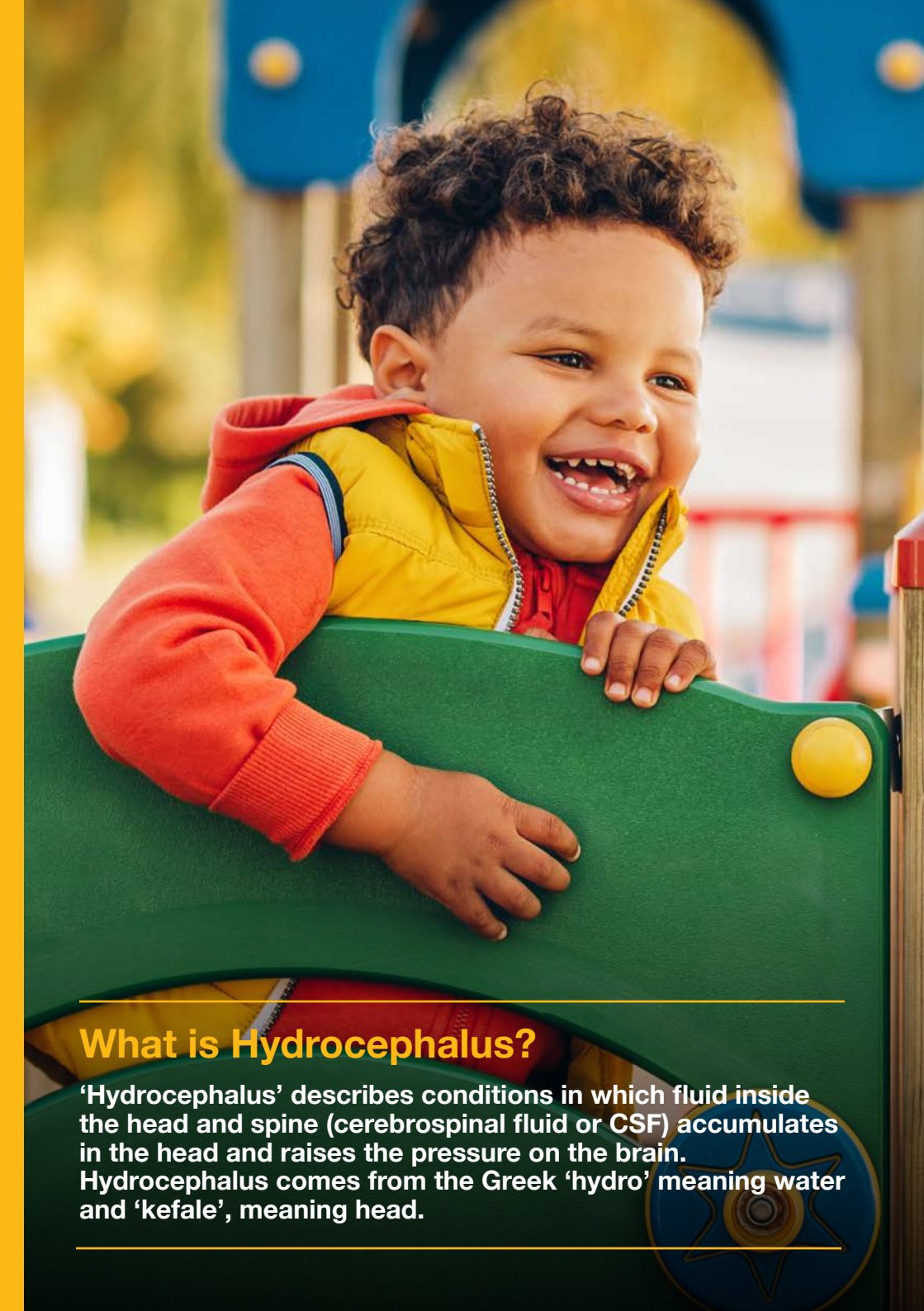


What is Hydrocephalus?

‘Hydrocephalus’ describes conditions in which fluid inside the head and spine (cerebrospinal fluid or CSF) accumulates in the head and raises the pressure on the brain. Hydrocephalus comes from the Greek ‘hydro’ meaning water and ‘kefale’, meaning head.

What causes hydrocephalus?

Hydrocephalus is caused by an inability of CSF to drain away into the bloodstream.

There are many reasons why this can happen, from differences in the way the brain develops, to failure of fluid absorption in an otherwise typical brain, or damage to brain tissue through head injury, haemorrhage or infection.

To help understand hydrocephalus, it is useful to explain what CSF is...

CSF is produced constantly inside each of the four spaces (ventricles) inside the brain – between 400 and 600ml is produced each day. CSF normally flows through narrow pathways from one ventricle to the next, over the outside of the brain and down the spinal cord, before being absorbed into the bloodstream.

From around the middle of pregnancy, CSF begins to flow through holes to the outer surface of the brain and spinal cord, between the membranes which cover the brain (meninges).

If the drainage of CSF is prevented at any point, the fluid accumulates in the ventricles, causing them to swell and compress the surrounding tissue.

In babies and infants, this will cause the head to enlarge, as the bones which form their skull are not yet fully fused.

Once the bones of the skull begin to join together, the head can no longer enlarge, so the pressure inside the head rises. This pressure begins to stretch brain tissue and reduce the blood flow to the brain.

What does CSF do?

Research into the exact functions of CSF is ongoing, but it's believed to support the brain and allow it to 'float' inside the head, preventing blood vessels from being squashed.

CSF also helps supply the brain with vitamins, and removes toxic waste chemicals like beta-amyloid (associated with normal pressure hydrocephalus and Alzheimer's).

As the brain develops in the womb, CSF carries chemical messages to guide brain cells to their correct positions. Without these chemicals, the brain development may differ from the norm, and some learning or behavioural effects may arise during childhood.

Types of hydrocephalus?

Hydrocephalus can be congenital (you are born with it) or acquired (occurring at any point in your lifetime).

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
- Spina bifida and hydrocephalus
- Genetics
- Maternal infections
- Cysts within the brain

Other developmental disorders of the brain can cause severe hydrocephalus, with a very poor outlook for the baby.

Holoprosencephaly is a condition in which the embryonic brain fails to divide into the usual two hemispheres.

Porencephaly is a rare condition in which a cyst or cavity within the brain fills with fluid.

It can damage or destroy brain tissue, create pressure on the brain tissue and cause hydrocephalus.

Acquired hydrocephalus

This occurs after birth and causes can include:

- Brain haemorrhage
- Prematurity
- Meningitis

Normal pressure hydrocephalus

Seen in older adults, normal pressure hydrocephalus, or NPH, is characterised by dementia, decreased mobility and urinary incontinence.

Recommended reading...

You can read more about different types of hydrocephalus on our website: ***www.shinecharity.org.uk/typesofhydrocephalus*** or, for an in-depth look at NPH, ask for a copy of our leaflet titled "What is Normal Pressure Hydrocephalus"

What are the effects of hydrocephalus?

The effects of hydrocephalus can vary greatly from one individual to another and some people may have very few, if any, problems.

However, it can be associated with learning difficulties, affecting:

- Concentration
- Reasoning
- Short-term memory
- Co-ordination
- Motivation
- Organisational skills
- Language

Physical effects may include visual problems, or early puberty in children.

Important to know:

Many of these effects can be reduced through teaching strategies or with treatment where relevant.

Recommended reading...

For more information about learning and behaviour, visit:

www.shinecharity.org.uk/learningandbehaviour

How is hydrocephalus treated?

Hydrocephalus is usually treated by diverting CSF to a place in the body where it can be absorbed.

Some forms of hydrocephalus require no specific treatment or are temporary and do not require treatment on a long term basis.

However, most forms do require treatment and this usually involves surgery to insert a shunt.

A shunt will generally improve symptoms caused by raised pressure, but other problems relating to brain damage may remain.

Important to know:

Fortunately, hydrocephalus is treatable for most people, but if the increase in pressure is sudden, damage to the brain may occur. In some cases the brain may be pressed against or even through the hole in the base of the skull (foramen magnum), which can crush important cranial nerves and can be life threatening.

An introduction to the shunt

What is a shunt?

A shunt is a thin tube-like device that prevents a build-up of pressure by draining excess CSF from the ventricles (or other CSF filled spaces) and diverting it to be absorbed in another area of the body.

The most common is ventriculopertioneal (VP), from the ventricles to the abdomen.

Less commonly used are:

- **Ventriculoatrial** (VA) from the ventricals to the heart
- **Ventriculopleural** (VPLS) from the ventricles to the outside of the lung
- **Ventriculosubgaleal** (VSGS) shunts in babies, from the ventricles to just under the scalp.

They are made from silicon, a strong, long-lasting and inert substance that very few people are allergic to.

Types of shunt

Shunts can be fixed pressure, adjustable pressure (programmable) or dual pressure (gravitational). Gravitational shunts can also be programmable.

Antimicrobial shunts can help reduce infection after surgery.

Programmable

Some shunts have valves that can be adjusted to open at varying pressures, by using a special device placed on the skin.

This way, the neurosurgeon can alter the way the shunt works without having to replace it. This is particularly helpful in children, as the drainage pattern changes as the child grows, and also for people with NPH.

Currently the most commonly used programmable shunts are adjusted with a magnetic device.

Gravitational

Shunts drain at different rates depending on whether we are standing or lying, owing to the effects of gravity.

They tend to drain more when we are upright, owing to a ‘siphon effect’, and the greater the height between the head and the bottom end of the shunt, the more drainage tends to occur.

To counteract this effect, some operate with two settings, one for standing, and another for lying down, to help reduce over drainage.

Recommended reading...

Antimicrobial

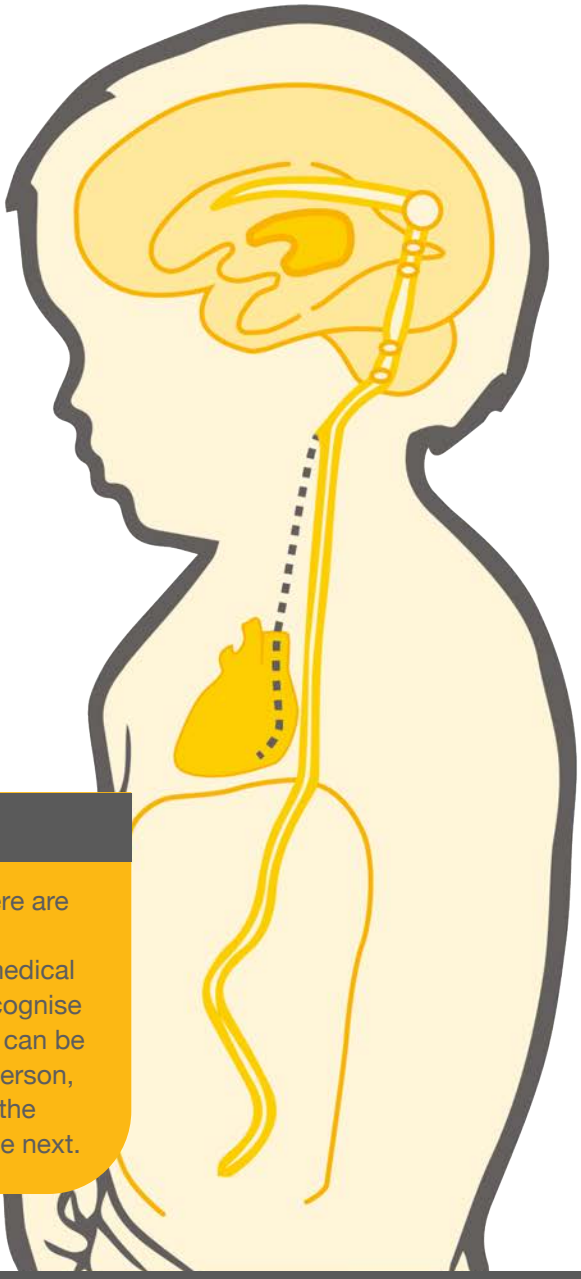
Bactiseal shunts are impregnated with two antibiotics to kill bacteria that are introduced at the time of the shunt surgery.

The antibiotics are embedded throughout the shunt's material, rather than being coated onto the inside of the shunt.

This means the antibiotics aren't washed off by the flow of the CSF, so continue to kill bacteria for several weeks after the shunt is inserted.

Antisiphon devices

Some shunts have devices included or added, to reduce or prevent over drainage owing to gravity when standing up (siphoning).



Important to know:

Shunts sometimes go wrong. There are various reasons why a shunt may malfunction and need specialist medical intervention, and being able to recognise the possible signs and symptoms can be vital. These vary from person to person, from infancy to adulthood, and in the same person from one shunt to the next.

Shunt complications

Infections

Occasionally, bacteria from the skin can infect the shunt at the time of surgery. Bacteria live within layers of the skin, no matter how clean it is or how thoroughly the skin is prepared for surgery.

The bacteria are released into the surgical cut onto the underlying tissue. Although harmless when on unbroken skin, they can cause infections in other parts of the body.

It is not possible for a shunt to become infected through day-to-day illnesses, such as colds, flu or dental problems, however, VP shunts can become infected following abdominal infections, like a burst appendix, or bladder or bowel surgery.

Blockages

If shunts block, they are unable to drain the excess CSF and the pressure within the head (intracranial pressure, or ICP) increases.

Shunts can block for a number of reasons:

The tiny drainage holes in the proximal catheter may become clogged with healthy brain cells as the catheter is passed through the brain on its way to the ventricle. This may be a repeated problem for some people, leading to a shunt blocking within a few days of being inserted, even with no infection present.

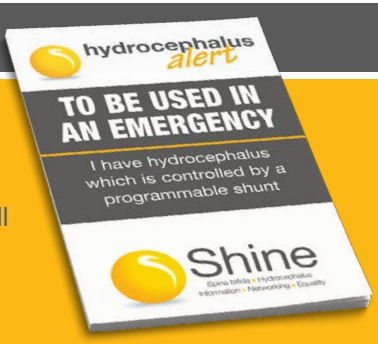
Some people are more prone to this, and at the moment we don't know why that is. The drainage holes can also become blocked with choroid plexus, a tissue in the ventricles that produces CSF.

The shunt can move slightly so the tip is no longer in the ventricle but in the brain tissue itself. The valve can also become blocked with protein or blood from haemorrhage, although this is rare.

The bottom end of a VP shunt can become blocked by the tissue covering the bowel, especially if there is a lot of scar tissue (adhesions) in the abdominal cavity, or infections or blood in the abdomen.

Important to know:

Shine offer a range of **Shunt Alert Cards** which advise what action needs to be taken in the event of a shunt complication. They should be carried at all times by anyone with hydrocephalus who is treated by a shunt. **Order yours online at:** www.shinecharity.org.uk/shuntalert



Shunt Revisions

If it is necessary to change part or all of your shunt, your neurosurgeon will aim to achieve this with the smallest risk of complications possible.

Shunts may, over time, become attached to the surrounding tissues, and it can be difficult to remove them without causing complications. However, if the shunt is infected it will usually be necessary to remove the whole shunt so that the infection can be treated effectively.

If the shunt needs changing for another reason, it may be possible to leave a non-functioning shunt in place and just add another shunt system, in another part of the head.

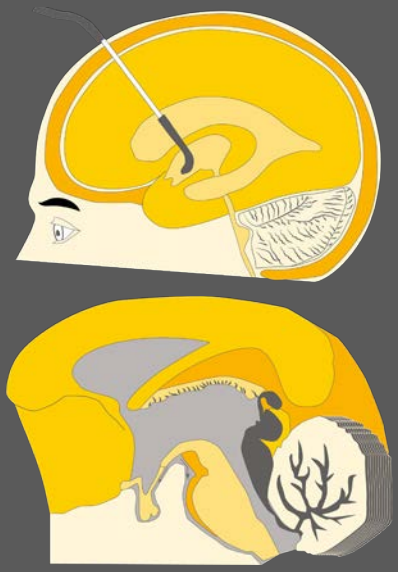
If the proximal catheter is blocked, but the distal catheter is working, it may be possible to just change the blocked component.

Shunts which are under or over draining may be 'tied off' when another shunt is inserted, to stop them working.

Endoscopic third ventriculostomy

An endoscopic third ventriculostomy (ETV) is a alternative treatment to fitting a shunt. It involves an operation where a small hole is made in the floor of the third ventricle using an endoscope to divert the flow of CSF so that it can be absorbed in the usual way.

Very occassionally, an ETV is combined with removal of some of the tissue which produces CSF (choroid plexus) in order to reduce the amount of CSF produced.



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 neurosurgical units.

For more information about ETV's visit: *www.shinecharity.org.uk/ETV*

Shine's specialist services

If you have hydrocephalus, live with or care for someone who does, are a professional providing support or are expecting a baby with hydrocephalus - we're here to help.

Support and Development Workers (SDWs)

Shine employs a team of SDWs across England, Wales and Northern Ireland. They are on hand to help you make important choices

Can advise on:

- Health conditions and treatment
- Staying healthy
- Living independently

...or just provide a listening ear

Your SDW can also refer you to one of the following:

Health Team

Shine's Health Team understand the worries, concerns and frustrations you are dealing with. They provide one to one support for individuals, parents, pregnant women, relatives, carers and fellow professionals.

Can advise on:

- Shunts and ETVs
- Pain
- Tests and treatments
- Depression or anxiety
- Living with the effects of hydrocephalus

Educational Advisers

Our Educational Advisers know all about the challenges of getting a good education and can support you to ensure the very best experience for your child.

Can advise on:

- Finding the right school
- Ensuring good support
- School transitions

...and more.

Benefits Advisory Service

Offering one to one advice and support, we are here to support Shine members to access key disability benefits.

Can advise on:

- Specific benefit issues
- Applications and claims
- Appealing a decision

...and more.

Join Shine FREE today to access all these services, and lots more!

Members also enjoy the following free benefits:

- Access to expert specialist support
- Regional face to face groups and online communities
- Invitations to regional and national events
- A network of friends and peers to help you
- A copy of our twice yearly magazine ‘Together’

For more information or to join today, visit us online at www.shinecharity.org.uk or call 01733 555988

Who are Shine?

With around 13,000 members across England, Wales and Northern Ireland, Shine is Europe's leading charity for people affected by spina bifida and hydrocephalus.

For over 50 years, we've been at the centre of developments which have improved the lives of thousands of people, enabling and empowering our members to lead the lives they want to live.

Get in touch and join today for FREE!

Many of Shine's services are entirely funded by the generosity of the public

Without your support, we could not offer vital services that help improve the lives of children, families and adults affected by hydrocephalus.

Please continue to support us in giving our members the best quality of life.

Donate today at:
shinecharity.org.uk

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