CONDITION

Congenital hydrocephalus

Congenital hydrocephalus is a buildup of fluid in the brain that begins before birth.

WHAT IT IS
Excess brain fluid present from birth

BODY SYSTEM
Nervous system

MAIN SYMPTOMS
Rapid head growth, bulging soft spot, vomiting, unusual sleepiness

WHAT CAUSES IT
Developmental differences, inherited changes, infection, or bleeding

HOW IT IS DIAGNOSED
Prenatal ultrasound; after birth, brain ultrasound or MRI

MAIN TREATMENT
Surgery to divert or reroute brain fluid

CAN IT BE CURED?
No - treatment controls fluid buildup

IS IT AN EMERGENCY?
Sometimes - severe sleepiness, seizure, or rapid decline

LONG-TERM OUTLOOK
Many live full lives; development and surgery needs differ

Overview

Congenital hydrocephalus is a buildup of cerebrospinal fluid, or CSF, within spaces in the brain called ventricles. CSF normally cushions the brain and spinal cord. It flows through the ventricles before being absorbed into the bloodstream.

Hydrocephalus develops when this fluid cannot flow normally, is not absorbed properly, or, very rarely, is produced in excess. The ventricles become enlarged. This may place pressure on the developing brain or stretch nearby brain tissue.

"Congenital" means the condition began before birth. It may be found during a pregnancy ultrasound, at birth, or later in infancy. Congenital does not necessarily mean inherited.

A baby's skull bones have not yet joined firmly, so increasing pressure can make the head grow faster than expected. This gives the brain some room, but it does not prevent harm from

ongoing pressure.

Enlarged ventricles are also called ventriculomegaly. Ventriculomegaly does not always mean a baby has active hydrocephalus. Some mild cases remain stable and do not require surgery. Doctors consider the imaging findings, head growth, symptoms, and whether the ventricles are continuing to enlarge.

Hydrocephalus matters because untreated pressure can affect vision, movement, learning, and development. Early monitoring and treatment can limit harm, although outcomes are also shaped by the underlying cause and any other brain or spinal differences.

Symptoms

Congenital hydrocephalus may first appear as an unexpected finding on a prenatal ultrasound. After birth, symptoms depend on the child's age and how quickly pressure is increasing.

Possible signs in babies include:

A large head by itself does not prove that a baby has hydrocephalus. Head size can run in families, and some children have a larger head without harmful pressure. The pattern of growth and brain imaging are more informative than one measurement.

In an older baby or child whose skull bones have joined more firmly, increasing pressure may cause headache, nausea, vomiting, blurred or double vision, balance problems, unusual tiredness, or changes in school performance or behaviour.

Symptoms can develop gradually or suddenly. A sudden change is especially important in a child with a shunt or a previous endoscopic procedure.

- A head that grows faster than expected
- A tense or bulging soft spot on top of the head
- Prominent veins on the scalp
- Eyes that appear driven downward, sometimes called "sunsetting"
- Repeated vomiting, poor feeding, or irritability
- Unusual sleepiness or difficulty waking
- A high-pitched cry
- Seizures
- Delayed movement or loss of previously gained skills

Causes and risk factors

Congenital hydrocephalus has several possible causes. Sometimes no single cause is found.

Fluid may be blocked by a narrow or closed passage inside the brain. Aqueductal stenosis is one example. It affects a small channel connecting two of the ventricles.

Hydrocephalus can occur with differences in how the brain, skull, or spine developed. These include myelomeningocele, a severe form of spina bifida, and conditions affecting the back of the brain, such as a Dandy-Walker malformation.

Other possible causes include:

Some inherited forms can affect more than one family member, but most babies with congenital hydrocephalus do not have a parent with the condition. Genetic counselling may be offered when imaging, family history, or other physical findings suggest an inherited cause.

Nothing a parent did during an ordinary pregnancy causes most cases. Finding the cause can help guide testing and explain the outlook, but it is not always possible.

- Bleeding in or around the brain before or near birth
- Infections during pregnancy that affect the developing brain
- Cysts, tumours, or other tissue blocking fluid flow
- Changes in genes involved in brain development
- Chromosome conditions or broader genetic syndromes

Diagnosis

Congenital hydrocephalus may be suspected when a prenatal ultrasound shows enlarged ventricles. Follow-up ultrasound examinations can show whether they are stable or getting larger. Fetal MRI may provide more detailed pictures of the brain and help identify an underlying developmental difference.

After birth, a healthcare professional measures the baby's head and plots it on a growth chart. The rate of growth matters more than a single measurement. The soft spot, eye movements, alertness, feeding, muscle tone, and development are also assessed.

Brain imaging confirms enlarged ventricles and helps show the cause. Tests may include:

Imaging alone does not always show whether pressure is harmful. Specialists combine the scan with symptoms, head growth, examination findings, and changes over time.

Additional tests may look for congenital infection, bleeding, or a genetic condition. These tests

investigate the cause rather than confirm hydrocephalus itself.

- Cranial ultrasound, which uses the open soft spot as a window
- MRI, which gives detailed images without ionising radiation
- CT, which is fast but uses radiation and is usually reserved for situations where it is needed

Treatment and management

A specialist team usually includes a paediatric neurosurgeon, who performs brain and spinal surgery. Treatment is considered when fluid buildup is causing symptoms, raising pressure, or continuing to progress.

The main treatment is surgery. There are two common approaches.

A shunt is a thin flexible tube that carries CSF away from the brain. Most shunts drain fluid into the abdomen, where the body absorbs it. A valve controls the direction and, in some designs, the amount of flow. The tubing sits under the skin.

Shunts often control hydrocephalus effectively, but they may become blocked, infected, disconnected, or outgrown. Many children need one or more shunt operations during childhood or later life. A shunt should be treated as a lifelong medical device unless a specialist confirms it is no longer needed.

Endoscopic third ventriculostomy, or ETV, creates a small opening inside the brain so fluid can bypass a blockage. It avoids an implanted shunt but is not suitable for every child. Its chance of working is influenced by the child's age, the cause of hydrocephalus, and the brain's anatomy.

In selected babies, ETV may be combined with choroid plexus cauterisation, or CPC. CPC treats tissue that produces CSF. Availability and suitability differ between specialist centres.

Medicines do not provide a reliable long-term cure. They may occasionally be used for short-term management in specific hospital situations.

A baby with mild, stable ventriculomegaly and no signs of increasing pressure may be monitored instead of having immediate surgery. Follow-up may include head measurements, developmental checks, eye examinations, and repeat imaging.

Support may also include physiotherapy, occupational therapy, speech and language therapy, vision care, or educational help. These services address the child's individual development rather than the fluid buildup itself.

Complications

Ongoing pressure can injure developing brain tissue. Possible effects include difficulties with movement, coordination, vision, learning, speech, attention, or seizures. The risk is affected by how severe and prolonged the pressure was and by the condition that caused the hydrocephalus.

Treatment reduces pressure but brings its own risks. A shunt can become blocked or break, allowing fluid to build up again. Infection may develop, particularly around the time of surgery. A shunt can also drain too much fluid, which may cause headaches or bleeding around the brain.

An ETV opening can close. This may happen soon after surgery or years later. Closure can cause pressure to rise quickly, so families need to know the warning signs even when the procedure has worked for a long time.

Some complications come from an associated condition rather than hydrocephalus itself. For example, a child with spina bifida may also need care for mobility, bladder, bowel, or spinal problems.

Prevention

Most congenital hydrocephalus cannot be prevented. This is especially true when it results from an early developmental difference or an inherited genetic change.

Taking the recommended folic acid before pregnancy and during early pregnancy reduces the risk of neural tube defects such as spina bifida. It does not prevent every neural tube defect or every case of hydrocephalus. People with certain medical or pregnancy histories may be advised to take a different amount, so local pre-pregnancy guidance is important.

Recommended vaccinations and infection-prevention measures before and during pregnancy can reduce the risk of some infections that may affect the fetal brain. They cannot remove all risk.

When an inherited cause is identified, genetic counselling can explain the chance of the condition occurring in another pregnancy and discuss available testing. This does not mean that either parent caused the condition.

Living with it and what to expect

The course of congenital hydrocephalus differs from child to child. Some children have

hydrocephalus alone and make steady developmental progress after treatment. Others have ongoing physical, learning, vision, or seizure-related needs, often because of the underlying brain or spinal condition.

Treatment does not usually make an enlarged head immediately become smaller. Instead, head growth may slow and return to a more expected pattern. The ventricles may remain larger than average even when pressure is controlled.

Children with shunts need long-term follow-up because a shunt can fail at any age. Families are often given information about the device and the symptoms that have previously occurred when it malfunctioned. These details are useful during travel, school, and emergency care.

Children treated with ETV also require ongoing awareness. A successful ETV may avoid a shunt, but its opening can close later.

Routine follow-up may include neurological examinations, head measurements in infancy, vision checks, developmental assessment, and imaging when clinically needed. Regular imaging is not necessarily required at every visit.

Many children attend school, play, travel, and take part in family life. Some need classroom adjustments, mobility support, extra time, or therapies. Activities are usually based on the child's abilities and advice about protecting any shunt tubing or other associated condition.

Changes in learning, coordination, continence, headaches, or vision should be discussed during routine or earlier follow-up. They may have several causes and do not always mean that pressure has returned.

When to get medical help

Contact the child's hydrocephalus or neurosurgical team promptly if there is a new pattern of headache, vomiting, irritability, poor feeding, unusual tiredness, downward-looking eyes, a bulging soft spot, or rapid head growth. A child with redness, swelling, tenderness, or fluid along a shunt also needs urgent assessment.

Fever with new neurological symptoms or changes around a shunt may indicate infection and should be assessed urgently. Do not wait for a routine appointment if symptoms are progressing.

Seek emergency help for:

For non-urgent concerns about development, school progress, vision, coordination, or recurring mild headaches, arrange routine follow-up. Keep scheduled appointments even when the child seems well because hydrocephalus and its treatments need long-term monitoring.

- A child who is very difficult to wake or rapidly becoming less responsive
- A seizure, especially a first seizure or one lasting several minutes
- New weakness, collapse, serious breathing difficulty, or loss of consciousness
- Severe or rapidly worsening headache with repeated vomiting
- Sudden symptoms suggesting shunt failure or ETV closure

Common questions

Can a baby with congenital hydrocephalus live a typical life?

Many can take part fully in family, school, and community life. Some have few long-term difficulties, while others need continuing support. The outlook is influenced by the cause, associated brain differences, response to treatment, infections, seizures, and developmental progress.

Is congenital hydrocephalus genetic?

Sometimes. Certain gene changes and genetic syndromes can cause it, but many cases are not inherited. A genetics assessment may be helpful when there is a family history, a suspected syndrome, or other developmental differences.

Does a large head always mean hydrocephalus?

No. Head size may be inherited, and some healthy children have larger heads. Hydrocephalus is assessed using the rate of head growth, symptoms, examination, and brain imaging.

Will a shunt stay in for life?

It often does. Even if a child appears well, a shunt should not be assumed to be unnecessary. Removing or changing it is a specialist decision. Shunts can need revision during childhood or adulthood.

Can hydrocephalus return after an ETV?

Yes. The opening created by ETV can close, sometimes years later. New headache, vomiting, marked sleepiness, or other previous pressure symptoms need urgent assessment.

Can congenital hydrocephalus be seen before birth?

It is often detected on prenatal ultrasound when the ventricles appear enlarged. Further ultrasound or fetal MRI may help show the cause and whether enlargement is progressing. The full effect on development cannot always be predicted before birth.

Sources

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