

Types of Hydrocephalus



Hydrocephalus can be identified in one of two ways - Congenital and Acquired.

Congenital

Congenital Hydrocephalus results from a complex interaction of genetic and environmental factors and is present at birth. It is important to remember that the term genetic does not imply that it is hereditary. Often the exact cause of congenital Hydrocephalus cannot be determined. Though it might not be recognised and diagnosed immediately, congenital Hydrocephalus is often diagnosed before birth through routine ultrasound. Hydrocephalus diagnosed in adulthood may have existed since birth and can still be considered congenital and may be referred to as compensated Hydrocephalus.

Acquired

Acquired Hydrocephalus develops after birth as a result of neurological conditions. This type of Hydrocephalus can affect individuals of all ages and may be caused by head trauma, brain tumour, cyst, intraventricular haemorrhage or infection of the central nervous system.

Within both of these areas, congenial and acquired, Hydrocephalus can be described as communicating or non-communicating. Communicating Hydrocephalus occurs when the flow of CSF is blocked after it exits the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open. Non-communicating Hydrocephalus - also called "obstructive" Hydrocephalus occurs when the flow of CSF is blocked along one or more of the narrow passages connecting the ventricles.

There are two other forms of Hydrocephalus which do not fit exactly into the categories mentioned above and primarily affect adults: Benign External Hydrocephalus and Normal Pressure Hydrocephalus.

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:

Aqueductal Stenosis (*non-communicating*)

The most common cause of congenital Hydrocephalus is an obstruction called

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aqueductal stenosis. When the long, narrow passageway between the third and fourth ventricles (see 'What is Hydrocephalus' diagram) is narrowed or blocked, perhaps because of infection, haemorrhage or a tumour. Fluid accumulates "upstream" from the obstruction, producing Hydrocephalus.

Neural Tube Defect or NTD (*communicating*)

An open NTD, where the spinal cord is exposed at birth and is often leaking CSF, is often referred to as Spina Bifida (see 'What is Spina Bifida'). This kind of NTD causes part of the cerebellum and the fourth ventricle to push downward through the opening at the base of the skull into the spinal cord area, blocking CSF's flow out of the fourth ventricle and producing Hydrocephalus.

Arachnoid Cysts (*non-communicating*)

Arachnoid Cysts may occur anywhere in the brain. In children, they're often located at the back of the brain and in the area of the third ventricle. These cysts are filled with CSF and lined with the arachnoid membrane. Some arachnoid cysts are self-contained, while others are connected with the ventricles or the subarachnoid space. The fluid trapped by the cysts may block the CSF pathways, producing Hydrocephalus.

Dandy-Walker Syndrome (*non-communicating*)

In Dandy-Walker syndrome the fourth ventricle becomes enlarged because its outlets are partly or completely closed, and part of the cerebellum fails to develop. Dandy-Walker syndrome may also be associated with irregular development in other parts of the brain and sometimes leads to aqueductal stenosis. In some instances, two shunts are placed in the child's ventricles - one in the lateral ventricle and another in the fourth ventricle to manage the Hydrocephalus.

Arnold-Chiari Malformation (*communicating*)

There are two types of Arnold-Chiari malformation. Both types occur in the bottom of the brain stem where the brain and spinal cord join. The lowest portion of the brain is displaced and is lower than normal pushing down into the spinal column. (For further information see "What is Chiari Malformation")

Acquired Hydrocephalus

This means that Hydrocephalus has occurred after birth and can be caused by:

Intraventricular Hemorrhage (*communicating*)

An intraventricular haemorrhage, which most frequently affects premature newborns, may cause an acquired form of Hydrocephalus. When small blood vessels alongside the

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ventricular lining rupture, blood may block or scar the ventricles or plug the arachnoid villi. The arachnoid villi is located in the second layer covering the brain which allows CSF to be absorbed. When the CSF can't be absorbed, Hydrocephalus results.

Meningitis (*communicating*)

Meningitis is an inflammation of the membranes of the brain and spinal cord. Caused by a bacterial or (less frequently) viral infection, meningitis can scar the delicate membranes (meninges) that line the CSF pathway. An acquired form of Hydrocephalus may develop if this scarring obstructs the flow of CSF as it passes through the narrow ventricles or over the surfaces of the brain in the subarachnoid space.

Head Injury (*communicating*)

A head injury can damage the brain's tissues, nerves, or blood vessels. Blood from ruptured vessels may enter the CSF pathway, causing inflammation. Sites of CSF absorption might then be blocked by scarred membranes (meninges) or by blood cells. The CSF flow is restricted and Hydrocephalus develops.

Brain Tumours (*non-communicating*)

In children, brain tumours most commonly occur in the back of the brain (posterior fossa). As a tumour grows, it may fill or compress the fourth ventricle, blocking the flow of CSF and causing Hydrocephalus. A tumour somewhere else in the brain might also block or compress the ventricular system.

Ventriculitis (*non-communicating*)

Ventriculitis is a disease causing inflammation and or infection of the ventricles. It is most common in infants and is often an extension of meningitis.

Choroid plexus papilloma (*communicating*)

A Choroid plexus papilloma (CPP) is a rare, slow-growing, tumour that is commonly located in the ventricular system of the choroid plexus (see 'What is Hydrocephalus' diagram). It may obstruct the cerebrospinal fluid flow, causing increased intracranial pressure and Hydrocephalus.

Prematurity (*communicating*)

Babies born prematurely are at risk of Hydrocephalus as their brain is still developing. The area which lies just beneath the lining of the ventricles in the brain is particularly important in this development as the activity in this area has a plentiful blood supply. The blood vessels are very fragile at this time 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 occur, then the baby may be at risk of developing a haemorrhage. This can lead to a blood clot developing, which in some cases is big enough to break

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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 and Hydrocephalus can occur.

Benign External Hydrocephalus (*communicating*)

Benign External Hydrocephalus (also referred to as External Hydrocephalus) occurs when an accumulation of CSF is found outside the brain, which usually presents itself at birth or soon thereafter. The infants head size will increase, but scans show no international difficulties in the ventricles or pathways. This condition usually corrects itself within 18 months of age.

Normal Pressure Hydrocephalus (*Non-communicating*)

Normal Pressure Hydrocephalus can happen to people at any age, but it is most common among the elderly. It may result from a subarachnoid haemorrhage, head trauma, infection, tumour, or complications of surgery. However, many people develop Normal Pressure Hydrocephalus even when none of these factors are present for reasons that are unknown. (For further information see 'What is Normal Pressure Hydrocephalus')