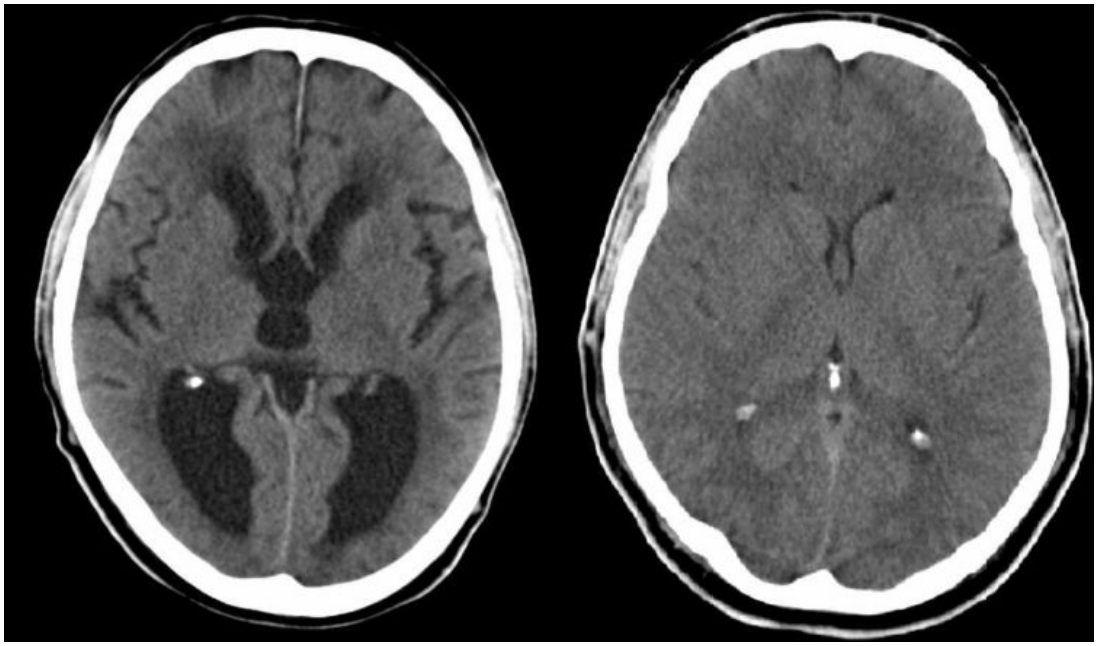
Hydrocephalus (Water in the Brain) — Symptoms and Surgery (Shunt and Ventriculostomy)

See online here

Hydrocephalus, commonly known as water in the brain, is the accumulation of cerebrospinal fluid in the subarachnoid space and cerebral ventricles leading to increased intracranial pressure.



Physiology of Cerebrospinal Fluid (CSF) and Circulation

CSF is produced mainly by the **choroid plexus** in the lateral ventricles. The choroid plexus consists of villous folds lined by epithelium with a central core of connective tissues.

CSF is produced by **a carbonic anhydrase–dependent active transport system** and can be blocked by carbonic anhydrase inhibitors such as **acetazolamide**. A smaller amount of CSF is produced through **diffusion from the cerebral tissue** and acts as brain lymph. The total amount of CSF is approximately 150 mL in adults and 50 mL in infants.

The CSF circulates through a pair of **lateral ventricles** that connect to the **midline third ventricle** via **the foramen of Monro**. The third ventricle connects with the fourth ventricle via the **aqueduct of Sylvius,** where the CSF circulates through two **foramina**

of Luschka and the **foramen of Magendie**.

Absorption of CSF occurs via the **arachnoid villi**, which drains into the systemic venous channels in the **sagittal venous sinus**. Arachnoid villi are protrusions from the subarachnoid space into the sinuses that allow passive absorption of CSF into the sinuses depending on the pressure gradient.

Classification of Hydrocephalus

Communicating hydrocephalus, in which the increased intracranial pressure is due to deficient absorption or increased production of the CSF in the subarachnoid space. There is no anatomical obstruction in normal CSF circulation; instead, there is uniform dilatation of the ventricles.

Non-communicating (obstructive) hydrocephalus, in which the increased intracranial pressure is due to the anatomical obstruction of CSF flow in the ventricular system. There is dilatation proximal to the obstruction, while the ventricular system distal to the region of obstruction is normal in size.

Normal-pressure hydrocephalus occurs mainly in adults. The ventricles are enlarged but there is no increase in intracranial pressure due to shrinking of the brain matter.

Pathology of Hydrocephalus

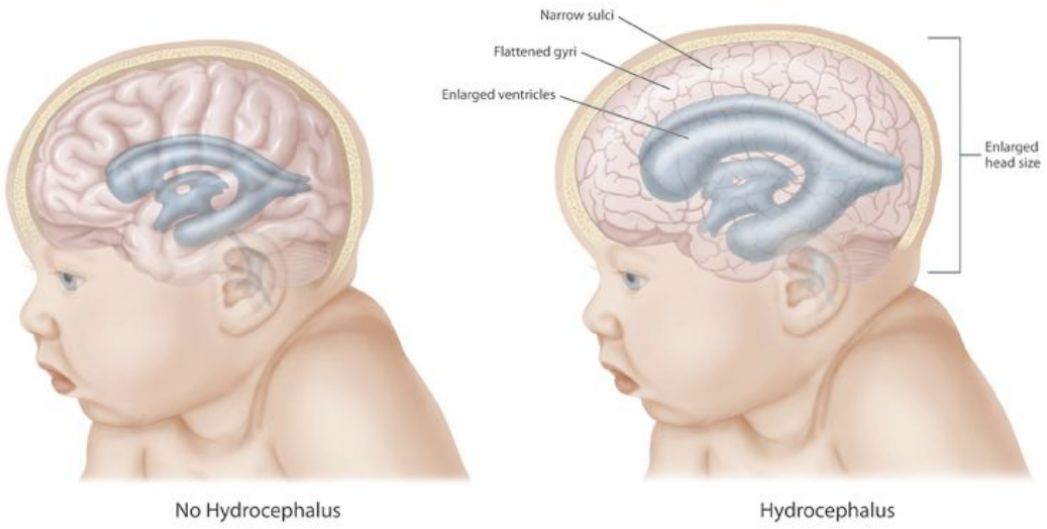


Image: “Hydrocephalus in Comparison to a Healthy Infant’s Brain.” by CDC. License: Public Domain

In obstructive hydrocephalus, anatomical obstruction of CSF flow leads to elevated pressure and subsequent dilatation of the ventricle proximal to the obstruction. The obstruction can occur at the level of the fourth ventricle, foramen of Monro, or aqueduct of Sylvius.

Excessive CSF production is seen in CSF-secreting choroid plexus papilloma. CSF levels exceed the rate of absorption and the pressure increases with dilatation of the ventricular system with no anatomical obstruction.

Impaired CSF absorption can occur due to inflammation of the arachnoid villi or elevated pressure in the venous sinuses.

Increased CSF pressure leads to enlargement of the head in infants before the fusion of the skull sutures. Further pressure on the brain parenchyma leads to compression of the brain tissue with **edema**, **ischemia,** and **disruption of the ependymal lining of the ventricles**.

This eventually leads to **atrophy of the white matter** with **flattening of brain gyri and narrowing of the sulci**, **demyelination,** and **axonal degeneration**. The pathology is more pronounced in adults owing to the fused skull sutures; consequently, acute hydrocephalus results with a rapid increase in intracranial pressure.

Etiology of Hydrocephalus

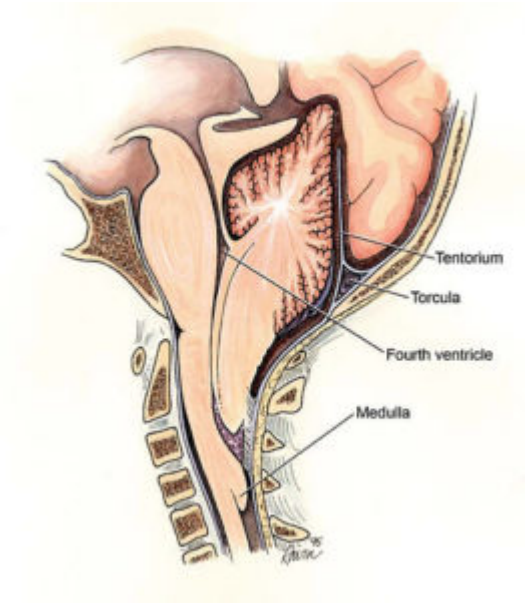


Image: "Chiari II Malformation Showing the Points of Potential Obstruction that Yield Different Subtypes of Hydrocephalus. With permission from Barrow Neurological Institute." by Rekate HL. License: CC BY 2.0

Congenital hydrocephalus

Neural tube defects and CNS malformations

Chiari II malformation is a disorder involving the posterior fossa and spine and is associated with myelomeningocele and encephalocele. The brain stem and cerebellum are displaced caudally leading to an obstruction in CSF flow into the spinal canal. These patients present with headaches, difficulty in swallowing, choking, vomiting, and dizziness. Chiari malformations are associated with syringomyelia.

Dandy-Walker malformation is characterized by a large cyst in the posterior fossa with maldevelopment of the cerebellum. CSF flow is obstructed by the atresia of the foramina of Luschka and Magendie.

Bickers-Adams syndrome is an X-linked disease with congenital stenosis of the aqueduct and mental retardation associated with adduction flexion of the thumb.

Rubella, syphilis, toxoplasma, the Zika virus, and cytomegalovirus can cause **intrauterine infections** and are responsible for congenital hydrocephalus. Inflammation, subsequent fibrosis, and calcification can lead to an obstruction in CSF flow

or decrease its absorption.

Aqueduct stenosis is responsible for isolated hydrocephalus due to congenital narrowing of the spinal canal or in the case of intrauterine infection or the use of antidepressants during pregnancy.

Choroid plexus papilloma can cause increased CSF production and communicating hydrocephalus.

Acquired hydrocephalus

During **subarachnoid hemorrhage**, bleeding in the ventricles or the subarachnoid space leads to impaired CSF absorption and obstruction in its flow. This condition can cause communicating and obstructive hydrocephalus in both adults and premature infants.

Infections, including meningitis and encephalitis, could lead to impaired CSF absorption or obstruction in its flow and may be accompanied by fibrosis and anatomical distortion.

Tumors, including those of the pineal body and posterior fossa, and ependymomas can lead to obstructive hydrocephalus.

Clinical Picture of Hydrocephalus

Symptoms

In infants with hydrocephalus, the first symptom is usually an **enlarged head. 'Failure to thrive'** and **developmental delays** are common. Older children present with complaints similar to those of adults, which include **persistent headache**, **nausea**, **vomiting**, **neck pain**, **blurring of vision**, **diplopia**, **gait disturbance**, and **focal neurological lesions** resulting from increased intracranial pressure. Enlargement of the third ventricle can lead to **precocious puberty** and **short stature**.

Adults usually present with a **headache**, **nausea**, **vomiting**, **vision abnormalities**, and **behavioral changes. Confusion** and **altered levels of consciousness** are observed in acute cases.

Signs

Manifestations of increased intracranial pressure in infants include an increase in the circumference of the head during serial measurements, indicating an **enlarged head. Prominent scalp veins** and **bulging anterior fontanelle** are other hallmark features. **Frontal bossing** is the abnormal contour of the skull that appears as an unusually prominent forehead in hydrocephalus. Abnormal cranial percussion notes can be heard across the skull sutures.

Enlarged head with bossing of the forehead makes the **sclera visible** above the iris and is known as the sunset sign. The upward gaze is also impaired due to increased pressure on the brain stem. Focal neurological signs include **spasticity of the lower limbs**. Eye examinations reveal **diplopia** and **ophthalmoplegia** that occur due to the stretching of the abducens or oculomotor nerve. **Papilledema** is more evident in adults than in children.

Hydrocephalus without an actual increase in CSF opening pressure but with an

enlargement in ventricular size is more common in adults > 60 years of age. Their clinical presentations include **dementia**, **gait disturbances,** and **urinary incontinence**.

Normal Pressure Hydrocephalus (NPH)

This condition may be idiopathic with no identifiable cause and could be secondary to subarachnoid hemorrhage, viral meningitis, or chronic inflammation and fibrosis. NPH leads to impaired absorption of the CSF, which consequently accumulates inside the ventricles and causes an increase in pressure and the stretching of the periventricular white matter.

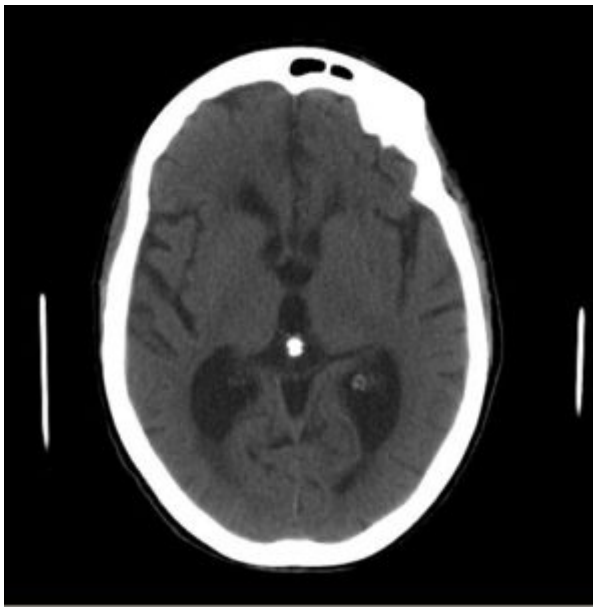


Image: "CT Scan of a Brain with Normal Pressure Hydrocephalus." by RadsWiki. License: CC BY-SA 3.0

Gait disturbances are characterized by slow and short steps. The patient's legs appear to be stuck to the ground and are externally rotated with little elevation and little forward progress. Falls are common and slow movement is evident. The gait usually improves dramatically after shunting or lowering the CSF pressure in these patients.

Urinary symptoms are evident in the form of urgency and incontinence.

Dementia, **depression,** and **apathy** are prominent early in the course of NPH. Cognitive impairment, psychomotor retardation, and diminished attention span are characteristics of NPH due to cortical ischemia. In late stages, patients may exhibit frontal release signs, e.g., grasping and suckling reflexes.

Differential diagnosis of NPH

Alzheimer's disease: Patients present with dementia, while motor involvement and gait abnormality appear later in the course of the disease.

Vascular dementia: Patients present with dementia and focal neurological lesions, but with a history of vascular disease and previous stroke. Upper motor neuron lesions can be seen with vascular dementia, while behavioral changes and urinary incontinence are more common in patients with NPH.

Parkinson's disease: Patients present with gait abnormality and depression in the case

of chronic illnesses; however, cognitive impairment is not common with Parkinson's disease as it is in NPH.

Imaging Studies in Hydrocephalus

Ultrasonography is an excellent imaging technique for antenatal assessment of neural tube defects and congenital anomalies. Measurement of the ventricular size can be achieved safely, even during early infancy, to detect any enlargement.

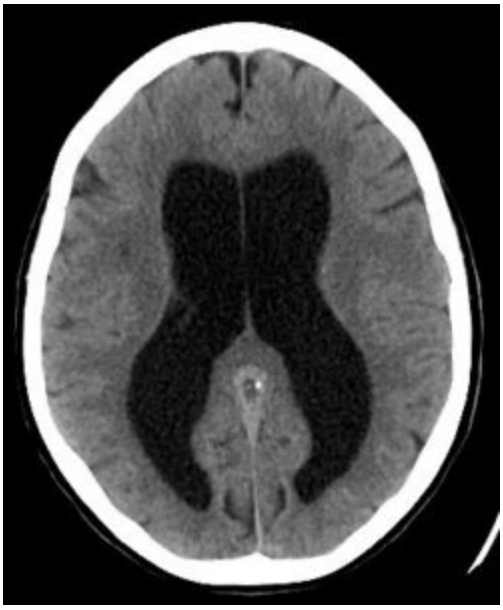


Image: "CT Scan of a Brain with Hydrocephalus." by Lucien Monfils – Own work. License: CC BY-SA 3.0

Computed tomography (CT) and **magnetic resonance imaging (MRI) studies** are used in older children and adults to assess brain anatomy and CSF flow. An MRI can detect small lesions in the brain, especially in the physiological CSF pathway.

Diffusion tensor imaging is used to assess changes in the periventricular white matter due to elevated CSF pressure.

Lumbar puncture and assessment of the opening pressure can be diagnostic in some cases but are contraindicated before brain imaging to exclude any space-occupying lesions and avoid herniation of the brain stem during the procedure. **CSF analysis** can be helpful in patients with infections. Lumbar puncture and drainage of approximately 50 mL of CSF are conducted preliminarily to test changes in gait and cognitive function. An improvement in these parameters within 60 minutes is considered to be indicative of the efficacy of shunting in NPH.

Radiological findings in hydrocephalus

It is important to know what to look for during imaging studies for early detection and to avoid late presentation and complications. Dilatation of the temporal and frontal horns of the lateral ventricles, elevation of the corpus callosum, stretching of the septum pellucidum with interstitial edema, and compression of the brain sulci are signs of increased CSF pressure.

Treatment of Hydrocephalus

Furosemide and **acetazolamide** are sometimes used to lower CSF production and pressure; however, drugs are only a temporary solution prior to surgery.

Medical therapy

Drug therapy is used when surgery is not the best option at the time such as in neonates and young children. Although drugs are usually not very effective in chronic hydrocephalus, the following options can be considered:

- **Acetazolamide with furosemide** are diuretics that work by eliminating excess body fluids and reducing the rate of CSF secretion
- **Isosorbide** increases the rate of cerebrospinal fluid absorption
- **Fibrinolytic therapy**
- **Serial lumbar punctures** are important to alleviate symptoms of increased intracranial pressure such as severe headaches and vomiting

As mentioned earlier, these are temporary solutions to lower CSF production and pressure before surgery.

Ventriculostomy

External ventricular drain (EVD) or **endoscopic third ventriculostomy (ETV)** can be performed in case of an emergency to lower high CSF pressure in acute hydrocephalus. Ventriculostomy can be life-saving in patients with an acute, rapid rise in intracranial pressure.

EVD allows the drainage of CSF via a catheter inserted through the skull, while ETV opens a hole in the third ventricular floor to allow CSF drainage into the subarachnoid space. ETV may be combined with **choroid plexus cauterization** for better outcomes. ETV is contraindicated in the case of communicating hydrocephalus as it has no beneficial outcome.

CSF shunt

CSF is allowed to drain from the ventricles into the systemic circulation or the peritoneal cavity where it is easily absorbed. A CSF shunt helps bypass the site of obstruction or reduce the increased CSF secretion. A catheter is placed in the frontal horn of one of the lateral ventricles and connected to a one-way valve that opens upon elevation of the pressure inside the ventricle. The valve is connected to a catheter that drains into the peritoneal space (**ventriculoperitoneal shunt**), which is the most common, right atrium (**ventriculoatrial shunt**), or pleural space (**ventriculopleural shunt**).

Lumpoperitoneal shunt can be used in communicating hydrocephalus by connecting the spinal canal with the peritoneal space, but it will not be effective in obstructive hydrocephalus.

Complications of surgical intervention

Overdrainage of CSF after shunting with constant headaches and subdural hemorrhage are frequent complications following shunting.

An **intracranial infection** may occur immediately after surgery or after several years.

Subclinical infections may lead to fibrosis and shunt malfunction. Infection following surgery or a malfunctioning shunt is a serious matter. The shunt should be removed and replaced after the infection resolves.

In the case of a shunt malfunction, the patient presents with worsening symptoms of increased intracranial pressure, i.e. headaches, nausea, vomiting, and papilledema. Shunt tapping demonstrates diminished flow, while the CSF pressure remains elevated. In such cases, a shunt replacement should be considered.

Seizures are common in children with shunt placement due to subarachnoid hemorrhage, infection, or trauma. Seizures can affect the cognitive outcome in affected patients.

Abdominal injury, **visceral perforation**, or **injury** may occur during shunt placement.

References

Normal pressure hydrocephalus via uptodate.com

Hydrocephalus in children: Physiology, pathogenesis, and etiology via uptodate.com

Hydrocephalus in children: Management and prognosis via uptodate.com

Hydrocephalus via medscape.com

Notes