

Hydrocephalus

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Introduction

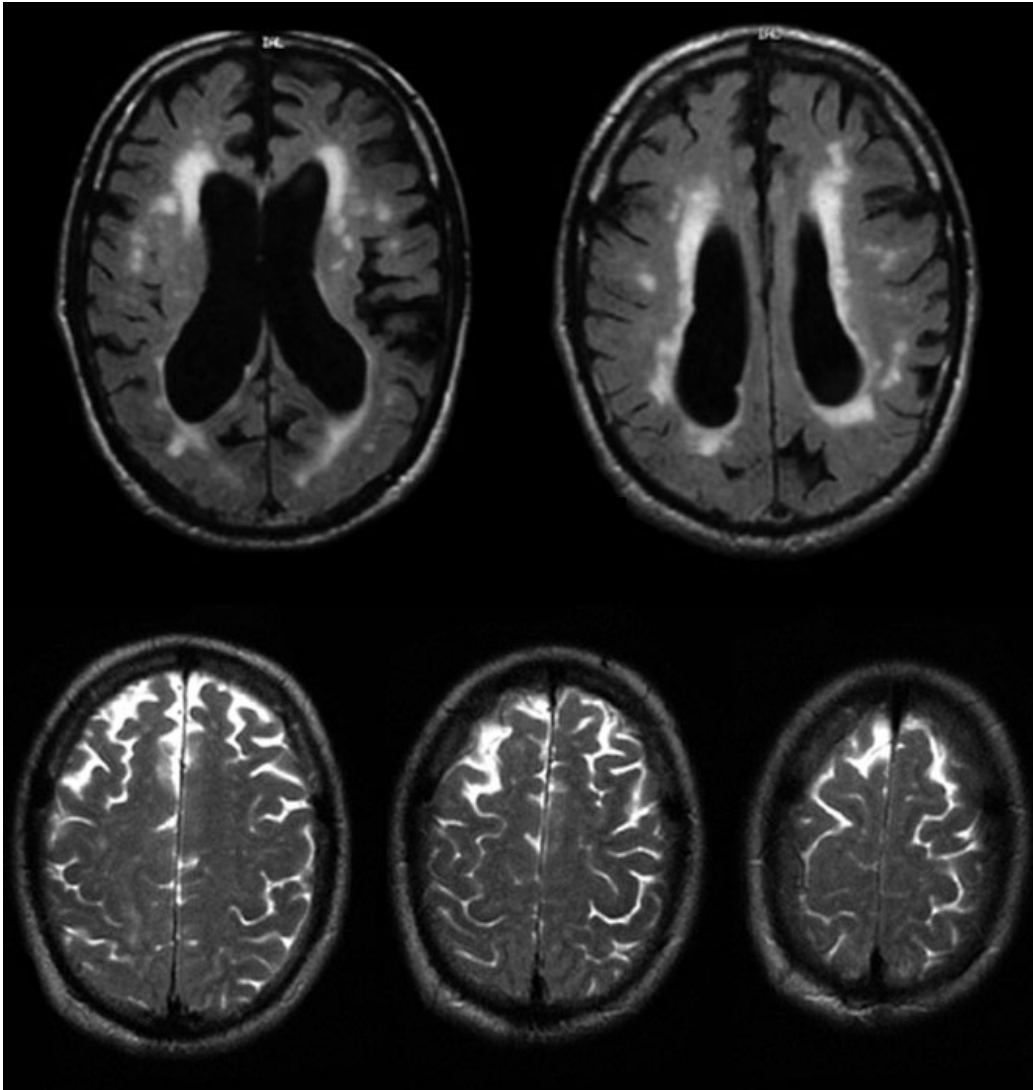
Background

Hydrocephalus can be defined broadly as a disturbance of formation, flow, or absorption of cerebrospinal fluid (CSF) that leads to an increase in volume occupied by this fluid in the CNS.^[1] This condition also could be termed a hydrodynamic disorder of CSF. Acute hydrocephalus occurs over days, subacute hydrocephalus occurs over weeks, and chronic hydrocephalus occurs over months or years. Conditions such as cerebral atrophy and focal destructive lesions also lead to an abnormal increase of CSF in CNS. In these situations, loss of cerebral tissue leaves a vacant space that is filled passively with CSF. Such conditions are not the result of a hydrodynamic disorder and therefore are not classified as hydrocephalus. An older misnomer used to describe these conditions was hydrocephalus ex vacuo.

Normal pressure hydrocephalus (NPH) describes a condition that rarely occurs in patients younger than 60 years.^[2] Enlarged ventricles and normal CSF pressure at lumbar puncture (LP) in the absence of papilledema led to the term NPH. However, intermittent intracranial hypertension has been noted during monitoring of patients in whom NPH is suspected, usually at night. The classic Hakim triad of symptoms includes gait apraxia, incontinence, and dementia. Headache is not a typical symptom in NPH.

Benign external hydrocephalus is a self-limiting absorption deficiency of infancy and early childhood with raised intracranial pressure (ICP) and enlarged subarachnoid spaces. The ventricles usually are not enlarged significantly, and resolution within 1 year is the rule.

Communicating hydrocephalus occurs when full communication occurs between the ventricles and subarachnoid space. It is caused by overproduction of CSF (rarely), defective absorption of CSF (most often), or venous drainage insufficiency (occasionally).

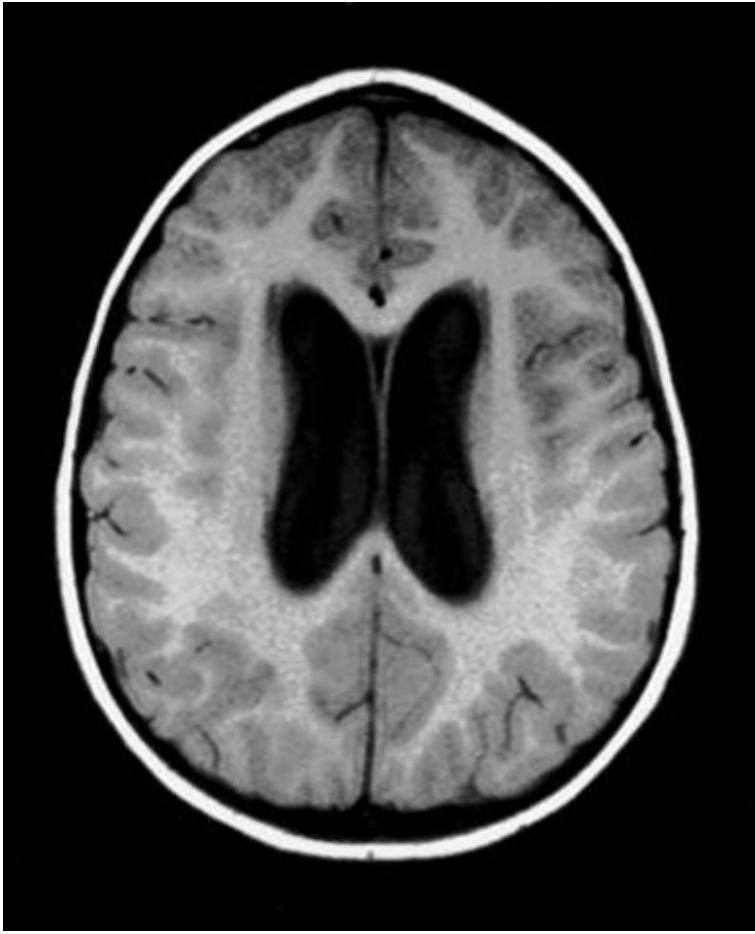


Communicating hydrocephalus with surrounding "atrophy" and increased periventricular and deep white matter signal on fluid-attenuated inversion recovery (FLAIR) sequences. Note that apical cuts (lower row) do not show enlargement of the sulci, as is expected in generalized atrophy. Pathological evaluation of this brain demonstrated hydrocephalus with no microvascular pathology corresponding with the signal abnormality (which likely reflects transependymal exudate) and normal brain weight (indicating that the sulci enlargement was due to increased subarachnoid cerebrospinal fluid [CSF] conveying a pseudoatrophic brain pattern).

Noncommunicating hydrocephalus occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space, resulting in impairment of the CSF from the ventricular to the subarachnoid space. The most common form of noncommunicating hydrocephalus is obstructive and is caused by intraventricular or extraventricular mass-occupying lesions that disrupt the ventricular anatomy.^[9]



Noncommunicating obstructive hydrocephalus caused by obstruction of the foramina of Luschka and Magendie. This MRI sagittal image demonstrates dilatation of lateral ventricles with stretching of corpus callosum and dilatation of the fourth ventricle.



Noncommunicating obstructive hydrocephalus caused by obstruction of foramina of Luschka and Magendie. This MRI axial image demonstrates dilatation of the lateral ventricles.



Noncommunicating obstructive hydrocephalus caused by obstruction of foramina of Luschka and Magendie. This MRI axial image demonstrates fourth ventricle dilatation.

Congenital hydrocephalus applies to the ventriculomegaly that develops in the fetal and infancy periods, often associated with macrocephaly.^[4]The most common causes of congenital hydrocephalus are obstruction of the cerebral aqueduct flow, Arnold-Chiari malformation or Dandy–Walker malformation.^[5]these patients may stabilize in later years due to compensatory mechanisms but may decompensate, especially following minor head injuries. During these decompensations, determining the extent to which any new neurological deficits may be due to the new acute event, compared with hydrocephalus that may have gone unnoticed for many years, is difficult.

Pathophysiology

Normal CSF production is 0.20-0.35 mL/min; most CSF is produced by the choroid plexus, which is located within the ventricular system, mainly the lateral and fourth ventricles. The capacity of the lateral and third ventricles in a healthy person is 20 mL. Total volume of CSF in an adult is 120 mL.

Normal route of CSF from production to clearance is the following: From the choroid plexus, the CSF flows to the lateral ventricle, then to the interventricular foramen of Monro, the third ventricle, the cerebral aqueduct of Sylvius, the fourth ventricle, the 2 lateral foramina of Luschka and 1 medial foramen of Magendie, the subarachnoid space, the arachnoid granulations, the dural sinus, and finally into the venous drainage.

ICP rises if production of CSF exceeds absorption. This occurs if CSF is overproduced, resistance to CSF flow is increased, or venous sinus pressure is increased. CSF production falls as ICP rises. Compensation may occur through transventricular absorption of CSF and also by absorption along nerve root sleeves. Temporal and frontal horns dilate first, often asymmetrically.

This may result in elevation of the corpus callosum, stretching or perforation of the septum pellucidum, thinning of the cerebral mantle, or enlargement of the third ventricle downward into the pituitary fossa (which may cause pituitary dysfunction).

The mechanism of NPH has not been elucidated completely. Current theories include increased resistance to flow of CSF within the ventricular system or subarachnoid villi; intermittently elevated CSF pressure, usually at night; and ventricular enlargement caused by an initial rise in CSF pressure; the enlargement is maintained despite normal pressure because of the Laplace law. Although pressure is normal, the enlarged ventricular area reflects increased force on the ventricular wall.

Frequency

United States

The incidence of congenital hydrocephalus is 3 per 1,000 live births; the incidence of acquired hydrocephalus is not known exactly due to the variety of disorders that may cause it.

International

Incidence of acquired hydrocephalus is unknown. About 100,000 shunts are implanted each year in the developed countries, but little information is available for other countries.

Mortality/Morbidity

In untreated hydrocephalus, death may occur by tonsillar herniation secondary to raised ICP with compression of the brain stem and subsequent respiratory arrest.

Shunt dependence occurs in 75% of all cases of treated hydrocephalus and in 50% of children with communicating hydrocephalus. Patients are hospitalized for scheduled shunt revisions or for treatment of shunt complications or shunt failure. Poor development of cognitive function in infants and children, or loss of cognitive function in adults, can complicate untreated hydrocephalus. It may persist after treatment. Visual loss can complicate untreated hydrocephalus and may persist after treatment.

Sex

Generally, incidence is equal in males and females. The exception is Bickers-Adams syndrome, an X-linked hydrocephalus transmitted by females and manifested in males. NPH has a slight male preponderance.

Age

Incidence of human hydrocephalus presents a bimodal age curve. One peak occurs in infancy and is related to the various forms of congenital malformations. Another peak occurs in adulthood, mostly resulting from NPH. Adult hydrocephalus represents approximately 40% of total cases of hydrocephalus.

Clinical

History

- Clinical features of hydrocephalus are influenced by the following:
 - Patient's age
 - Cause
 - Location of obstruction
 - Duration
 - Rapidity of onset
- Symptoms in infants
 - Poor feeding
 - Irritability
 - Reduced activity
 - Vomiting

- Symptoms in children
 - Slowing of mental capacity
 - Headaches (initially in the morning) that are more significant than in infants because of skull rigidity
 - Neck pain suggesting tonsillar herniation
 - Vomiting, more significant in the morning
 - Blurred vision: This is a consequence of papilledema and later of optic atrophy
 - Double vision: This is related to unilateral or bilateral sixth nerve palsy
 - Stunted growth and sexual maturation from third ventricle dilatation: This can lead to obesity and to precocious puberty or delayed onset of puberty.
 - Difficulty in walking secondary to spasticity: This affects the lower limbs preferentially because the periventricular pyramidal tract is stretched by the hydrocephalus.
 - Drowsiness
- Symptoms in adults
 - Cognitive deterioration: This can be confused with other types of dementia in the elderly.
 - Headaches: These are more prominent in the morning because cerebrospinal fluid (CSF) is resorbed less efficiently in the recumbent position. This can be relieved by sitting up. As the condition progresses, headaches become severe and continuous. Headache is rarely if ever present in normal pressure hydrocephalus (NPH).
 - Neck pain: If present, neck pain may indicate protrusion of cerebellar tonsils into the foramen magnum.
 - Nausea that is not exacerbated by head movements
 - Vomiting: Sometimes explosive, vomiting is more significant in the morning.
 - Blurred vision (and episodes of "graying out"): These may suggest serious optic nerve compromise, which should be treated as an emergency.
 - Double vision (horizontal diplopia) from sixth nerve palsy
 - Difficulty in walking
 - Drowsiness
 - Incontinence (urinary first, fecal later if condition remains untreated): This indicates significant destruction of frontal lobes and advanced disease.
- Symptoms of NPH
 - Gait disturbance is usually the first symptom and may precede other symptoms by months or years. Magnetic gait is used to emphasize the tendency of the feet to remain "stuck to the floor" despite patients' best efforts to move them.
 - Dementia should be a late finding in pure (shunt-responsive) NPH. It presents as an impairment of recent memory or as a "slowing of thinking." Spontaneity and initiative are decreased. The degree can vary from patient to patient.
 - Urinary incontinence may present as urgency, frequency, or a diminished awareness of the need to urinate.
 - Other symptoms that can occur include personality changes and Parkinsonism. Seizures are extremely rare and should prompt consideration for an alternative diagnosis.

Physical

- Infants
 - Head enlargement: Head circumference is at or above the 98th percentile for age.
 - Dysjunction of sutures: This can be seen or palpated.
 - Dilated scalp veins: The scalp is thin and shiny with easily visible veins.
 - Tense fontanelle: The anterior fontanelle in infants who are held erect and are not crying may be excessively tense.
 - Setting-sun sign: In infants, it is characteristic of increased intracranial pressure (ICP). Ocular globes are deviated downward, the upper lids are retracted, and the white sclerae may be visible above the iris.
 - Increased limb tone: Spasticity preferentially affects the lower limbs. The cause is stretching of the periventricular pyramidal tract fibers by hydrocephalus.
- Children
 - Papilledema: if the raised ICP is not treated, this can lead to optic atrophy and vision loss.

- Failure of upward gaze: This is due to pressure on the tectal plate through the suprapineal recess. The limitation of upward gaze is of supranuclear origin. When the pressure is severe, other elements of the dorsal midbrain syndrome (ie, Parinaud syndrome) may be observed, such as light-near dissociation, convergence-retraction nystagmus, and eyelid retraction (Collier sign).
- Macewen sign: A "cracked pot" sound is noted on percussion of the head.
- Unsteady gait: This is related to spasticity in the lower extremities.
- Large head: Sutures are closed, but chronic increased ICP will lead to progressive macrocephaly.
- Unilateral or bilateral sixth nerve palsy is secondary to increased ICP.
- Adults
 - Papilledema: If raised ICP is not treated, it leads to optic atrophy.
 - Failure of upward gaze and of accommodation indicates pressure on the tectal plate. The full Parinaud syndrome is rare.
 - Unsteady gait is related to truncal and limb ataxia. Spasticity in legs also causes gait difficulty.
 - Large head: The head may have been large since childhood.
 - Unilateral or bilateral sixth nerve palsy is secondary to increased ICP.
- NPH
 - Muscle strength is usually normal. No sensory loss is noted.
 - Reflexes may be increased, and the Babinski response may be found in one or both feet. These findings should prompt search for vascular risk factors (causing associated brain microangiopathy or vascular Parkinsonism), which are common in NPH patients.
 - Difficulty in walking varies from mild imbalance to inability to walk or to stand. The classic gait impairment consists of short steps, wide base, externally rotated feet, and lack of festination (hastening of cadence with progressively shortening stride length, a hallmark of the gait impairment of Parkinson disease). These abnormalities may progress to the point of apraxia. Patients may not know how to take steps despite preservation of other learned motor tasks.
 - Frontal release signs such as sucking and grasping reflexes appear in late stages.

Causes

- Congenital causes in infants and children^[4]
 - Brainstem malformation causing stenosis of the aqueduct of Sylvius: This is responsible for 10% of all cases of hydrocephalus in newborns.
 - Dandy-Walker malformation: This affects 2-4% of newborns with hydrocephalus.
 - Arnold-Chiari malformation type 1 and type 2
 - Agenesis of the foramen of Monro
 - Congenital toxoplasmosis
 - Bickers-Adams syndrome: This is an X-linked hydrocephalus accounting for 7% of cases in males. It is characterized by stenosis of the aqueduct of Sylvius, severe mental retardation, and in 50% by an adduction-flexion deformity of the thumb.
- Acquired causes in infants and children
 - Mass lesions: Mass lesions account for 20% of all cases of hydrocephalus in children. These are usually tumors (eg, medulloblastoma, astrocytoma), but cysts, abscesses, or hematoma also can be the cause.^[6]
 - Hemorrhage: Intraventricular hemorrhage can be related to prematurity, head injury, or rupture of a vascular malformation.
 - Infections: Meningitis (especially bacterial) and, in some geographic areas, cysticercosis can cause hydrocephalus.
 - Increased venous sinus pressure: This can be related to achondroplasia, some craniostenoses, or venous thrombosis.
 - Iatrogenic: Hypervitaminosis A, by increasing secretion of CSF or by increasing permeability of the blood-brain barrier, can lead to hydrocephalus. As a caveat, hypervitaminosis A is a more common cause of idiopathic intracranial hypertension, a disorder with increased CSF pressure but small rather than large ventricles.
 - Idiopathic
- Causes of hydrocephalus in adults

- Subarachnoid hemorrhage (SAH) causes one third of these cases by blocking the arachnoid villi and limiting resorption of CSF. However, communication between ventricles and subarachnoid space is preserved.^[7]
- Idiopathic hydrocephalus represents one third of cases of adult hydrocephalus.
- Head injury, through the same mechanism as SAH, can result in hydrocephalus.
- Tumors can cause blockage anywhere along the CSF pathways. The most frequent tumors associated with hydrocephalus are ependymoma, subependymal giant cell astrocytoma, choroid plexus papilloma, craniopharyngioma, pituitary adenoma, hypothalamic or optic nerve glioma, hamartoma, and metastatic tumors.
- Prior posterior fossa surgery may cause hydrocephalus by blocking normal pathways of CSF flow.
- Congenital aqueductal stenosis causes hydrocephalus but may not be symptomatic until adulthood. Special care should be taken when attributing new neurological deficits to congenital hydrocephalus, as its treatment by shunting may not correct these deficits.
- Meningitis, especially bacterial, may cause hydrocephalus in adults.
- All causes of hydrocephalus described in infants and children are present in adults who have had congenital or childhood-acquired hydrocephalus.
- Causes of NPH (Most cases are idiopathic and are probably related to a deficiency of arachnoid granulations.)
 - SAH
 - Head trauma
 - Meningitis

Differential Diagnoses

Brainstem Gliomas	Migraine Headache
Childhood Migraine Variants	Migraine Variants
Craniopharyngioma	Oligodendroglioma
Epidural Hematoma	Pituitary Tumors
Frontal and Temporal Lobe Dementia	Primary CNS Lymphoma
Frontal Lobe Epilepsy	Pseudotumor Cerebri
Frontal Lobe Syndromes	Pseudotumor Cerebri: Pediatric Perspective
Glioblastoma Multiforme	Subdural Empyema
Headache: Pediatric Perspective	Subdural Hematoma
Intracranial Epidural Abscess	Sudden Visual Loss
Intracranial Hemorrhage	
Meningioma	
Mental Retardation	

Other Problems to Be Considered

Brainstem syndromes
 Macrocephaly
 Hydranencephaly
 Chronic subdural hemorrhages
 Cerebral atrophy
 Cerebral tumors
 Periaqueductal glioma
 Agenesis of corpus callosum
 Septo-optic dysplasia
 Neuroimaging of vascular malformations and hematomas of the brain

Workup

Laboratory Studies

- No specific blood tests are recommended in the workup for hydrocephalus.
- Genetic testing and counseling might be recommended when X-linked hydrocephalus is suspected.
- Evaluate cerebrospinal fluid (CSF) in posthemorrhagic and postmeningitic hydrocephalus for protein concentration and to exclude residual infection.

Imaging Studies

- CT can assess the size of ventricles and other structures.
- MRI can evaluate for Chiari malformation or cerebellar or periaqueductal tumors. It affords better imaging of the posterior fossa than CT. MRI can differentiate normal pressure hydrocephalus (NPH) from cerebral atrophy although the distinctions may be challenging. Flow voids in the third ventricle and transependymal fluid exudates are helpful. However, numerous suitable patients have a brain pattern suggestive of atrophy and small vessel ischemic disease that may ultimately be NPH.^[8] Guidelines for imaging studies in suspected NPH have been established.^[9]
- CT/MRI criteria for acute hydrocephalus include the following:
 - Size of both temporal horns is greater than 2 mm, clearly visible. In the absence of hydrocephalus, the temporal horns should be barely visible.
 - Ratio of the largest width of the frontal horns to maximal biparietal diameter (ie, Evans ratio) is greater than 30% in hydrocephalus.
 - Transependymal exudate is translated on images as periventricular hypoattenuation (CT) or hyperintensity (MRI T2-weighted and fluid-attenuated inversion recovery [FLAIR] sequences).
 - Ballooning of frontal horns of lateral ventricles and third ventricle (ie, "Mickey mouse" ventricles) may indicate aqueductal obstruction.
 - Upward bowing of the corpus callosum on sagittal MRI suggests acute hydrocephalus.
- CT/MRI criteria for chronic hydrocephalus include the following:
 - Temporal horns may be less prominent than in acute hydrocephalus.
 - Third ventricle may herniate into the sella turcica.
 - Sella turcica may be eroded.
 - Macrocrania (ie, occipitofrontal circumference >98th percentile) may be present.
 - Corpus callosum may be atrophied (best appreciated on sagittal MRI). In this case, parenchymal atrophy and ex-vacuo (rather than true) hydrocephalus from a neurodegenerative disease should be considered.
- Ultrasonography through the anterior fontanelle in infants is useful for evaluating subependymal and intraventricular hemorrhage and in following infants for possible development of progressive hydrocephalus.
- Radionuclide cisternography can be done in NPH to evaluate the prognosis with regard to possible shunting. If a late scan (48-72 h) shows persistence of ventricular activity with a ventricular to total intracranial activity (V/T ratio) greater than 32%, the patient is more likely to benefit from shunting.^[10] Because of its poor sensitivity in predicting shunt response when the V/T ratio is less than 32%, this test is no longer commonly used.
- Skull radiographs may depict erosion of sella turcica, or "beaten copper cranium" (called by some authors "beaten silver cranium"). The latter can also be seen in craniosynostosis.
- MRI cine is an MRI technique to measure CSF stroke volume (SV) in the cerebral aqueduct. Cine phase-contrast MRI measurements of SV in the cerebral aqueduct does not appear to be useful in predicting response to shunting.^[11]
- Diffusion tensor imaging (DTI) is a novel imaging technique that detects differences in fractional anisotropy (FA) and mean diffusivity (MD) of the brain parenchyma surrounding the ventricles. Impairment of FA and MD through DTI allows the recognition of microstructural changes in periventricular white matter region that may be too subtle on conventional MRI.^[32]

Other Tests

- After shunt insertion, confirm correct positioning of installed hardware with a plain radiograph.
- EEG if seizure occurs

Procedures

- Lumbar puncture (LP) is a valuable test in evaluating NPH, but should be performed only after CT or MRI of the head. Normal LP opening pressure (OP) should be less than 180 mm H₂ O (ie, 18 cm H₂ O). Patients with initial OP greater than 100 mm H₂ O have a higher rate of response to CSF shunting than those with OPs less than 100 mm H₂ O. Improvement of symptoms after a single LP in which 40-50 mL of CSF is withdrawn appears to have some predictive value for success of CSF shunting.
- Continuous CSF drainage through external lumbar drainage (ELD) is a highly accurate test for predicting the outcome after ventricular shunting in NPH, although false negative results are not uncommon.^[12]
- Continuous CSF pressure monitoring can help in predicting a patient's response to CSF shunting in NPH. Some patients with normal OP on LP demonstrate pressure peaks of greater than 270 mm H₂ O or recurrent B waves. These patients tend to have higher rates of response to shunting than those who do not have these findings. This procedure also could differentiate NPH from atrophy.

Histologic Findings

- Thinning and stretching of the cortical mantle may be seen as a result of ventricular dilation.
- In the acute phase, edema of the periventricular white matter is observed. Relatively few neuronal lesions are present. Ventricular ependyma shows cellular flattening and loss of cilia.
- At a later stage, the edema disappears and is replaced by fibrosis, axonal degeneration, demyelination, focal loss of cerebral cortical neurons, cellular flattening, and further loss of cilia.

Treatment

Medical Care

- Medical treatment in hydrocephalus is used to delay surgical intervention. It may be tried in premature infants with posthemorrhagic hydrocephalus (in the absence of acute hydrocephalus). Normal CSF absorption may resume spontaneously during this interim period.
- Medical treatment is not effective in long-term treatment of chronic hydrocephalus. It may induce metabolic consequences and thus should be used only as a temporizing measure.
- Medications affect CSF dynamics by the following mechanisms:
 - Decreasing CSF secretion by the choroid plexus - Acetazolamide and furosemide
 - Increasing CSF reabsorption - Isosorbide (effectiveness is questionable)

Surgical Care

- Surgical treatment is the preferred therapeutic option.^[13]
- Repeat lumbar punctures (LPs) can be performed for cases of hydrocephalus after intraventricular hemorrhage, since this condition can resolve spontaneously. If reabsorption does not resume when the protein content of cerebrospinal fluid (CSF) is less than 100 mg/dL, spontaneous resorption is unlikely to occur. LPs can be performed only in cases of communicating hydrocephalus.
- Alternatives to shunting include the following:
 - Choroid plexectomy or choroid plexus coagulation may be effective.
 - Opening of a stenosed aqueduct has a higher morbidity rate and a lower success rate than shunting, except in the case of tumors. However, lately cerebral aqueductoplasty has gained popularity as an effective treatment for membranous and short-segment stenoses of the sylvian aqueduct. It can be performed through a coronal approach or endoscopically through suboccipital foramen magnum trans-fourth ventricle approach.
 - In these cases, tumor removal cures the hydrocephalus in 80%.
 - Endoscopic fenestration of the floor of the third ventricle establishes an alternative route for CSF toward the subarachnoid space. It is contraindicated in communicating hydrocephalus.
- Shunts eventually are performed in most patients. Only about 25% of patients with hydrocephalus are treated successfully without shunt placement. The principle of shunting is to establish a communication between the CSF (ventricular or lumbar) and a drainage cavity (peritoneum, right atrium, pleura). Remember that shunts are not perfect and that all alternatives to shunting should be considered first.

- A ventriculoperitoneal (VP) shunt is used most commonly. The lateral ventricle is the usual proximal location. The advantage of this shunt is that the need to lengthen the catheter with growth may be obviated by using a long peritoneal catheter.
- A ventriculoatrial (VA) shunt also is called a "vascular shunt." It shunts the cerebral ventricles through the jugular vein and superior vena cava into the right cardiac atrium. It is used when the patient has abdominal abnormalities (eg, peritonitis, morbid obesity, or after extensive abdominal surgery). This shunt requires repeated lengthening in a growing child.
- A lumboperitoneal shunt is used only for communicating hydrocephalus, CSF fistula, or pseudotumor cerebri.
- A Torkildsen shunt is used rarely. It shunts the ventricle to cisternal space and is effective only in acquired obstructive hydrocephalus.
- A ventriculopleural shunt is considered second line. It is used if other shunt types are contraindicated.
- Rapid-onset hydrocephalus with increased intracranial pressure (ICP) is an emergency. The following can be done, depending on each specific case:
 - Ventricular tap in infants
 - Open ventricular drainage in children and adults
 - LP in posthemorrhagic and postmeningitic hydrocephalus
 - VP or VA shunt

Consultations

- Neurosurgeon
- Neurologist
- Neurorehabilitation specialist
- Ophthalmologist

Diet

- Regular, as tolerated

Activity

- Most surgeons agree that, with the use of antisiphon devices, no special positioning is required after shunting. However, some surgeons used to leave patients in whom a standard shunt had been placed in a recumbent position for 1-2 days after surgery to minimize risk of subdural hematoma.
- In treatment of normal pressure hydrocephalus (NPH), gradual postoperative mobilization is recommended.

Medication

Acetazolamide (ACZ) and furosemide (FUR) treat posthemorrhagic hydrocephalus in neonates. Both are diuretics that also appear to decrease secretion of CSF at the level of the choroid plexus. ACZ can be used alone or in conjunction with FUR. The combination enhances efficacy of ACZ in decreasing CSF secretion by the choroid plexus. If ACZ is used alone, it appears to lower risk of nephrocalcinosis significantly.

Medication as treatment for hydrocephalus is controversial. It should be used only as a temporary measure for posthemorrhagic hydrocephalus in neonates.

Carbonic anhydrase inhibitors

These agents inhibit an enzyme found in many tissues of the body that catalyzes a reversible reaction in which carbon dioxide becomes hydrated and carbonic acid dehydrated. These changes may result in a decrease in CSF production by the choroid plexus.

Acetazolamide (Diamox)

Noncompetitive reversible inhibitor of enzyme carbonic anhydrase, which catalyzes the reaction between water and carbon dioxide, resulting in protons and carbonate. This contributes to decreasing CSF secretion by choroid plexus.

Dosing

Adult

Pediatric

25 mg/kg/d PO tid; not to exceed 100 mg/kg/d

Interactions

Alkalizes urine and may decrease excretion of amphetamines, procainamide, quinidine, flecainide, anticholinergics, and mecamlamine; may increase excretion and lower plasma levels of salicylate, phenobarbital, and lithium; can increase cyclosporine levels and decrease primidone levels; concurrent salicylates may increase accumulation and toxicity, including CNS depression and metabolic acidosis

Contraindications

Documented hypersensitivity; hepatic insufficiency, hyponatremia, hypokalemia, hyperchloremic acidosis, severe renal insufficiency, nephrocalcinosis, adrenal gland failure

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

Can cause hyperglycemia in diabetics; concurrent digoxin can increase susceptibility to ACZ-induced hypokalemia; in patients taking other diuretics, ACZ can aggravate hypokalemia; can aggravate preexisting acidosis, which can be prevented by initiating prophylactic electrolyte replacement; this may consist of sodium citrate starting at 8 mEq/kg/d titrated, keeping serum bicarbonate levels >18 mEq/L and sodium and potassium within reference ranges
Obtain baseline CBC prior to initiating therapy; recheck regularly during therapy

Loop diuretics

These agents increase excretion of water by interfering with the chloride-binding cotransport system, which results from inhibition of reabsorption of sodium and chloride in the ascending loop of Henle and distal renal tubule.

Furosemide (Lasix)

Mechanisms proposed for lowering ICP include lowering cerebral sodium uptake, affecting water transport into astroglial cells by inhibiting cellular membrane cation-chloride pump, and decreasing CSF production by inhibiting carbonic anhydrase. Used as adjunctive therapy with ACZ in temporary treatment of posthemorrhagic hydrocephalus in neonates.

Dosing

Adult

Pediatric

1 mg/kg/d IV

eractions

May increase ototoxic potential of aminoglycoside antibiotics; may increase salicylate toxicity if given with salicylate; may decrease arterial response to norepinephrine

Contraindications

Documented hypersensitivity to drug or sulfonyleureas, hepatic coma, anuria, severe electrolyte depletion, concurrent ethacrynic acid (may cause ototoxicity), or lithium (may cause lithium toxicity)

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

Excessive use can cause dehydration and circulatory collapse; can cause electrolyte imbalance as hypokalemia, hyponatremia, hypochloremic alkalosis, hypomagnesemia, and hypocalcemia; therefore, monitor serum electrolytes; may increase blood glucose in patients with diabetes; may cause photosensitivity

Follow-up

Further Inpatient Care

- Patients with shunt-dependent hydrocephalus should be admitted for consideration of shunt revision if shunt malfunction or infection is suspected.
- In children, shunt revisions are scheduled according to growth rate.

Further Outpatient Care

- Patients on acetazolamide (ACZ) or furosemide (FUR) should be followed for possible electrolyte imbalance and metabolic acidosis. Clinical signs that should prompt attention are lethargy, tachypnea, or diarrhea.
- Patients with shunts should be reevaluated periodically, including assessment of distal shunt length in growing children. The first follow-up examination usually is scheduled 3 months after surgery, and CT scan or MRI of the head should be done at that time. Follow-up is performed every 6-12 months in the first 2 years of life. In children aged 2 years and older, follow-up is performed every 2 years.

Inpatient & Outpatient Medications

- Medications include acetazolamide and furosemide. These are helpful for temporizing the hydrocephalus until compensation occurs. If compensation does not occur, then shunting is indicated.
- Medications should not be used in patients with functional shunts.
- Medication is not effective in long-term treatment of chronic hydrocephalus, and it may induce metabolic consequences.
- If seizures occur, antiepileptic drugs are recommended.

Transfer

- In cases of acute hydrocephalus or shunt complications, immediately transfer the patient to a center with a neurosurgery service.

Deterrence/Prevention

- Avoid trauma: The valve and tubing system are located superficially under the skin and can be damaged easily by trauma.

Complications

- Related to progression of hydrocephalus
 - Visual changes
 - Occlusion of posterior cerebral arteries secondary to downward transtentorial herniation
 - Chronic papilledema injuring the optic disc
 - Dilatation of the third ventricle with compression of optic chiasm
 - Cognitive dysfunction
 - Incontinence
 - Gait changes
- Related to medical treatment
 - Electrolyte imbalance
 - Metabolic acidosis
- Related to surgical treatment
 - Signs and symptoms of increased intracranial pressure (ICP) can be a consequence of undershunting or shunt obstruction or disconnection.
 - Subdural hematoma or hygroma is secondary to overshunting. Headache and focal neurological signs are common.
 - Treat seizures with antiepileptic drugs.
 - Shunt infection occasionally can be asymptomatic. In neonates, it manifests as alteration of feeding, irritability, vomiting, fever, lethargy, somnolence, and a bulging fontanelle. Older children and adults present with headache, fever, vomiting, and meningismus. With ventriculoperitoneal (VP) shunts, abdominal pain may occur.
 - Shunts can act as a conduit for extraneural metastases of certain tumors (eg, medulloblastoma).
 - Hardware erosion through the skin occurs in premature infants with enlarged heads and thin skin who lie on 1 side of the head.
 - VP shunt complications include peritonitis, inguinal hernia, perforation of abdominal organs, intestinal obstruction, volvulus, and CSF ascites.
 - Ventriculoatrial (VA) shunt complications include septicemia, shunt embolus, endocarditis, and pulmonary hypertension.
 - Lumboperitoneal shunt complications include radiculopathy and arachnoiditis.

Prognosis

- Long-term outcome is related directly to the cause of hydrocephalus.
- Up to 50% of patients with large intraventricular hemorrhage develop permanent hydrocephalus requiring shunt.
- Following removal of a posterior fossa tumor in children, 20% develop permanent hydrocephalus requiring a shunt. The overall prognosis is related to type, location, and extent of surgical resection of the tumor.
- Satisfactory control was reported for medical treatment in 50% of hydrocephalic patients younger than 1 year who had stable vital signs, normal renal function, and no symptoms of elevated ICP.
- Criteria exist for predicting improvement with shunting in NPH, but they are controversial.
 - If gait disturbance precedes mental deterioration, the chance of improvement is 77%. Patients with dementia and no gait disturbance rarely respond to shunting.
 - Focal impingement of corpus callosum on MRI indicates unstable ICP and is associated with a good response to shunting.
 - Initial OP of CSF greater than 100 mm H₂ O predicts better response.
 - Response to a single LP or to controlled CSF drainage via lumbar subarachnoid catheter (ELD) has some value in predicting outcome.
 - Cerebral blood flow of 32 mL/100 g per minute or greater predicts clinical improvement after shunt.

- CSF pressure of 180 mm H₂O with frequent Lundberg B waves on continuous CSF pressure monitoring is associated with good prognosis after shunting. Lundberg B waves represent an accentuation of physiological phenomena, reflecting arterial waves. They represent fluctuating ICP waves of 4-8 per minute frequency and 20-30 mm Hg (260-400 mm H₂O) amplitude. Occasionally they can occur in normal sleep.
- Large ventricles with flattened or invaginated sulci (entrapped sulci) suggest that hydrocephalus is not due to atrophy alone. These patients have good prognosis with shunting.
- If isotopic cisternography shows persistent ventricular activity on a late scan (42-72 h), the probability of improving with shunting is 75%.

Patient Education

- Knowledge of the signs and symptoms of shunt malfunction or infection and the necessity for emergent medical evaluation in these instances is mandatory in patients, family members, and caregivers.
- The patient, family, and caregivers should know that periodic re-evaluation is necessary.
- Pumping the shunt is contraindicated in most cases.
- Patients with vascular shunts, and some patients with other types of shunts, should receive prophylactic antibiotics before dental procedures or instrumentation of the bladder.

Miscellaneous

Medicolegal Pitfalls

- Failure to recognize signs and symptoms of new onset hydrocephalus
- Failure to recognize signs and symptoms of shunt malfunction or shunt infection, and failure to refer to a neurosurgeon immediately when these are suspected
- Failure to inform patients with shunts and family members concerning the lifelong possibility of shunt complications.

Special Concerns

- Patients with arrested hydrocephalus need close follow-up. They can decompensate at any time, often after minor head injuries or an infectious process. The patient and family should know the signs of acute and chronic progressive hydrocephalus.
- Occasionally in hydrocephalus due to a Chiari malformation, further herniation of cerebellar tonsils can occur after shunt placement. This can lead to quadriplegia or death.
- True normal pressure hydrocephalus (NPH) should be heralded by gait and not cognitive impairments; this hydrocephalus is disproportionate to the degree of atrophy (if any). Shunting ex-vacuo hydrocephalus (due to Alzheimer disease, for instance) can only be harmful.

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Keywords

normal pressure hydrocephalus, communicating hydrocephalus, noncommunicating hydrocephalus, obstructive hydrocephalus, arrested hydrocephalus, acute hydrocephalus, gait apraxia, incontinence, dementia, Arnold-Chiari malformation, papilledema, precocious puberty, Dandy–Walker malformation, obesity, delayed onset of puberty, urinary incontinence, Parkinsonism, seizures, toxoplasmosis, Bickers-Adams syndrome, mental retardation, medulloblastoma, astrocytoma, prematurity, achondroplasia, cysticercosis, treatment, diagnosis