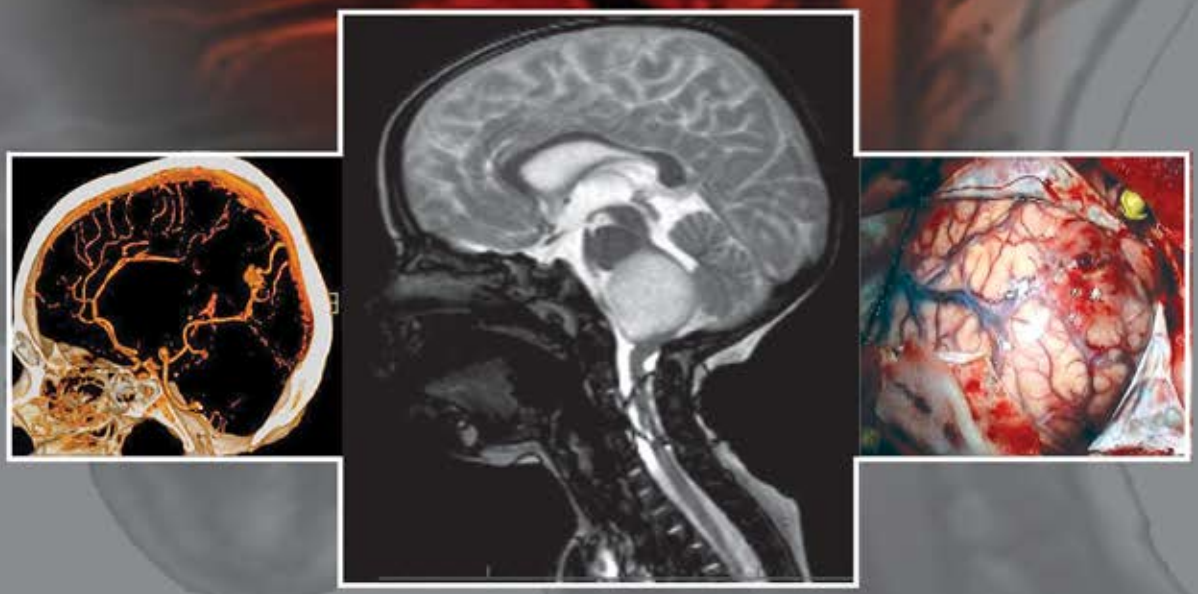
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Textbook of Focused Neurosurgery



Or Cohen-Inbar

Foreword
H Richard Winn

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Textbook of
FOCUSED NEUROSURGERY

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Foreword

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CHAPTER 10

Hydrocephalus

CLASSIFICATION AND ETIOLOGY

Communicating Hydrocephalus

- Panventricular dilation secondary to obstruction to CSF flow in the subarachnoid space, distal to Luschka and Magendie.
- This is commonly caused by infection or hemorrhage or is idiopathic. Lumbar puncture is generally safe (Table 10.1).

Noncommunicating or Obstructive Hydrocephalus

- Characterized by a pattern of ventricular dilation that reflects the site of obstruction.
- Obstruction within the ventricular system or at the level of the outlets of the fourth ventricle.
- Noncommunicating hydrocephalus may develop after multiple shunt revisions. Lumbar puncture may be dangerous.

Long-standing Overt Ventriculomegaly in Adults

- This form of hydrocephalus develops during childhood, with symptoms manifested during adulthood.
- Associated with macrocephalus (>2 SD in head circumference).
- Aqueduct stenosis is present but may be secondary because it may develop after third ventriculostomy.
- A spectrum of patients ranging from those with symptoms and signs of increased ICP to those with dementia, gait disturbance, and urinary incontinence may be encountered.
- There is a major risk with surgical intervention for subdural hematomas.

Normal-pressure Hydrocephalus

- NPH is characterized by gait apraxia, dementia, and incontinence with normal CSF pressure and dilated ventricles.
- The designation "normal" is misleading because continuous intracranial monitoring has demonstrated the presence of waves of increased ICP, particularly during rapid eye movement (REM) sleep.
- It has been suggested that these abnormal CSF pressure spikes, called B waves, slowly increase ventricular size by exerting intermittent high pressure on the brain parenchyma that results in ischemic damage. The exact pathogenesis is still unknown.

Table 10.1: Communicating hydrocephalus types

Disruption to flow in the subarachnoid space	Leptomeningeal inflammation, infections (meningitis), hemorrhage (SAH), carcinomatosis, foreign matter, tonsillar descent/basilar impression, massess (neoplastic or other)
Descreased absorption of CSF at arachnoid granulations	Congenital deficiency of arachnoid granulations, raised cerebral venous pressure
Abnormal CSF	Abnormally high CSF volume, raised intraventricular CSF pulse pressure (choroid plexus papilloma), increased CSF viscosity (high protein or cellular content, like in the presence of spinal neurofibromas)
Idiopathic	

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Arrested Hydrocephalus

- Some patients with hydrocephalus reach a state in which ventricular size remains unchanged in the absence of a shunt or in the presence of a nonfunctioning one. It is well accepted that careful follow-up is still required, particularly in children, because of reported cases of sudden death, sometimes years after the initial diagnosis.

Isolated Fourth Ventricle Syndrome

- The term "entrapped fourth ventricle" has also been used to describe the situation in which the fourth ventricle no longer communicates with the third ventricle, as well as the basal cisterns.
- Patients with prolonged infections or multiple shunt operations are particularly at risk for this syndrome.
- It is thought that secondary aqueduct stenosis from adhesions, obstruction of Luschka or Magendie, or infective debris pooling in the basal cisterns may be responsible for this condition.

Slit Ventricle Syndrome

- The lateral ventricles may collapse second to overshunting or remain at a fixed size because of subependymal gliosis. This may lead to intermittent or complete shunt malfunction. Patients may experience raised ICP without ventricular enlargement, and therefore imaging findings may be falsely reassuring in such cases (unresponsive ventricles).
- Symptomatic patients may respond to a change in valve setting if a programmable valve is in situ or to revision surgery.

INFANTILE POSTHEMORRHAGIC HYDROCEPHALUS

- Intraventricular hemorrhage (IVH) is a leading cause of infantile hydrocephalus. The management of hydrocephalus in these infants is critical because the initial neonatal treatment can have lifelong implications for both shunt survival and the child's neurological outcome. IVH and subsequent posthemorrhagic hydrocephalus (PHH) arise from different pathophysiologic mechanisms, pose different clinical and management issues, and suggest different prognoses in preterm versus term infants.

Intraventricular Hemorrhage in Preterm Infants

Definitions—SEH (subependymal hemorrhage), GMH (germinal matrix hemorrhage), IVH (intraventricular hemorrhage) (Table 10.2).

Risk factors for SEH

Increased CPP and increased CBF

A. Asphyxia: Including hypercapnia.
B. Volume expansion
C. Seizures
D. Pneumothorax
E. Cyanotic heart disease (including PDA)
F. Mechanical ventilation/RDS.

Extracorporeal membrane oxygenation (ECMO).
Maternal cocaine abuse.

Epidemiology: The survival of all preterm cohorts has risen dramatically over the past few decades owing to improvements in perinatal medicine, but many of these infants remain at risk for neurodevelopmental deficits.

- Preterm infants account for 64–75% of infant mortality, 42–47% of children with cerebral palsy, 27% of children with cognitive deficits, 37% of children with visual impairments, 23% of those with hearing impairments.
- The prevalence of IVH varies, depending on availability and quality of neonatal intensive care.
- The incidence of IVH increases inversely with decreasing birth weight or EGA (estimated gestational age).

***Incidence*:** Many are asymptomatic.

Realistic—40–45% of preemies <1500g or <35 weeks gestation. Most common <32 weeks gestation.

- Many occur within 6 hours of birth, 50% occur within 24 hours, >90% occur in the first 72 hours of life.

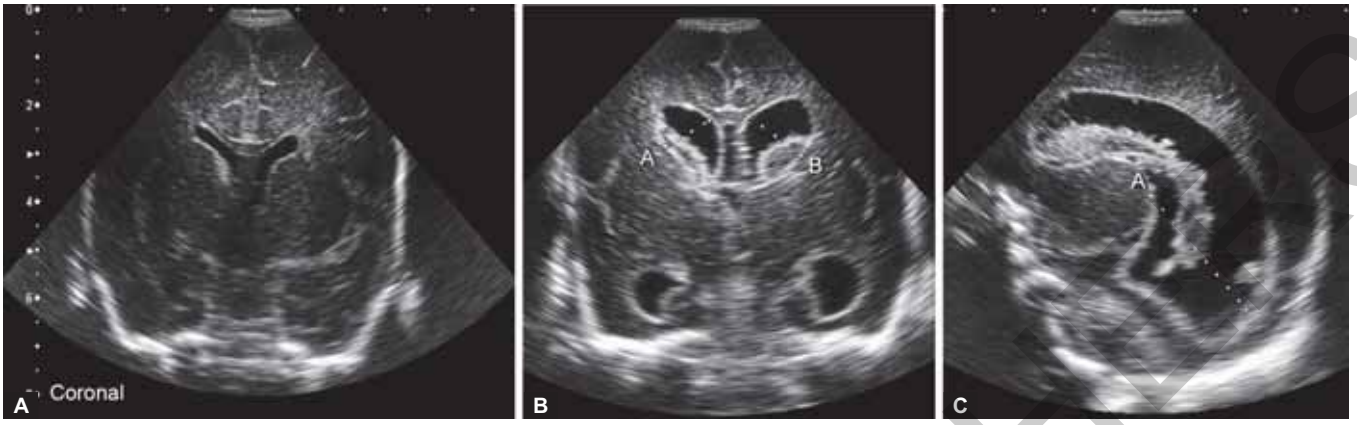
Progression—Documented in 10–20%.

- Because less than 10% of infants with IVH develop PHH and require a shunt, a strong collaboration between

Table 10.2: Papille's classification of preterm IVH on ultrasonography and related prognosis and complications

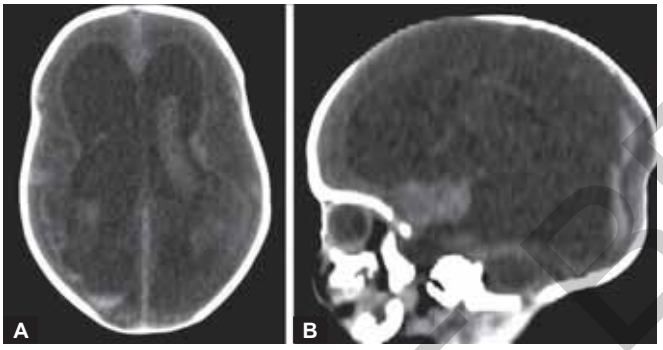
Grade	*Definition*	*Progressive hydrocephalus (%)*	*Deaths (%)*
I	Isolated GMH	0–10	0
II	IVH without enlarged ventricles	15–25	5–15
III	IVH with enlarged ventricles	65–100	50–65
IV	IVH with parenchymal component		

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Figs 10.1A to C: (A) Grade-1 GMH on the right; (B + C) GMH grade-III, ventriculomegally. Note the echogenic material anterior to the caudothalamic groove

Source: Gaillard F, Singh G. www.radiopaedia.org



Figs 10.2A and B: GMH Grade-III with communicating hypdrocephalus and periventricular leukomalacia. Non contrast CT axial (A) sagittal (B)

Source: Rabou AA. www.radiopaedia.org

the neurosurgical team and the neonatology team is required to obtain neurosurgical consultation in a timely manner and to avoid unnecessary nonsurgical consults.

- Intravenous access in cranial veins should be avoided in any preterm infants who might develop hydrocephalus.

Pathophysiology of Germinal Matrix Hemorrhage and Posthemorrhagic Infarction

- The germinal matrix is a transient structure in the subependyma of the ventricular walls that generates neural cell progenitors for the overlying cortex, primarily from 8 to 28 weeks' EGA.
- The germinal matrix typically completes involution by 32 weeks' EGA.
- The germinal matrix is a metabolically active area with friable, immature vessels prone to hemorrhage.
- Hemorrhage can be limited to within the germinal matrix (grade I), but about 80% of germinal matrix hemorrhages rupture through the ventricular wall into the ventricle (grade II, III, or IV) (Figs 10.1 and 10.2).
- Grade IV is also called periventricular hemorrhagic infarction.
- Because of their immaturity, preterm infants have impaired cerebral autoregulation. Systemic fluctuations in blood volume, flow, and pressure that commonly occur in preterm infants are risk factors for GMH.
- After IVH occurs, the absorption of cerebrospinal fluid (CSF) is impaired, likely caused by a combination of decreased absorption across the arachnoid villi into veins and from ependyma into parenchyma.
- IVH can occur in association with periventricular leukomalacia (PVL), injury to the developing white matter. Cystic PVL is visible on cranial ultrasonography (US).
- MRI and autopsy studies have shown that injury to the developing central nervous system affects both white and gray matter. The combined injury has been designated encephalopathy of prematurity, which more accurately reflects the widespread nature of the injury not only to the cerebral gray and white matter but also to the diencephalon, brainstem, etc.

Clinical Presentation and Diagnostic Evaluation

- Most IVH occurs within the first 72 hours of life, when most preterm newborns are quite unstable. IVH is readily diagnosed by bedside US. CT should be reserved

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for evaluation of diffuse, life-threatening hemorrhages when MRI is not feasible (Table 10.3).

- If possible, all preterm newborns weighing less than 1500 g should have a cranial US within the first 48 hours of life, and then weekly or biweekly as needed. PVL can also be assessed by US.
- Infans with IVH should be observed closely with daily measurements of the occipitofrontal circumference. An increase of 0.5 to 1 cm/day for 2 to 3 consecutive days often suggests symptomatic hydrocephalus.
- The anterior fontanelle in very small infants is often quite large.
- Splayed sutures is often a reliable finding on daily examinations to assess symptomatic hydrocephalus.
- Preterm infants with symptomatic hydrocephalus can have episodes of spontaneous apnea or bradycardia.
- Other symptoms and signs include refractory seizures, lethargy, and impaired upward gaze ("sunset" phenomenon).

***Hydrocephalus ex vacuo*:** Ventricular dilation without increased ICP. Reflects encephalomalacia, or a lack of adequate parenchymal brain growth.

Treatment: Because grades III + IV IVH tend to occur in the smallest and sickest infants, they are more likely to have other confounding illnesses. Interventions for symptomatic hydrocephalus are offered in a stepwise progression to identify patients in whom permanent CSF diversion is not required (Table 10.4).

Nonsurgical Treatment

Serial lumbar punctures: A temporizing measure. Reduce need for permanent shunt. As long as there is no large posterior fossa hemorrhage, most preterm infants can safely undergo lumbar puncture.

Temporary CSF diversion device: Used to delay the need for definitive surgical treatment.

- Currently, medical therapy is not recommended for preterm infants with symptomatic hydrocephalus.
- A meta-analysis of repeated lumbar or ventricular punctures failed to show that CSF removal improved outcomes compared with no intervention. This study, however, included ventricular taps (more complications), and also included patients without progressive ventricular dilation.

Temporary Surgical Interventions: Preterm infants have immature immune systems, fragile skin, and impaired wound healing. In addition, infants weighing less than 1500 g often have insufficient peritoneal absorptive capacity to handle the CSF volume.

- Temporary CSF diversion can be achieved with a ventricular access device or a ventriculosubgaleal shunt.
- A ventricular access device consists of a reservoir that caps a ventricular catheter, and the reservoir is accessed via a needle through the scalp every 12 to 48 hours to withdraw CSF.
- Ventriculosubgaleal shunts are also composed of a ventricular catheter and reservoir, with a 4- to 8-cm

Table 10.3: Clinical presentation in GMH, IVH

Acute	A sudden neurologic deterioration, stupor, changes in muscle tone, seizures, tense fontanelle, hypotension
Subacute	Irritability, reduced activity, abnormal eye movements
Asymptomatic	Many ICHs will be unsuspected. 78% 6-month survival

Table 10.4: Treatment concepts and principles in GMH, IVH

Optimize CPP*	Normalizing pCO_2, mean arterial (carotid) pressure	
Ventriculomegaly	Transient	In the first few days after SEH. Self limited
	Progressive	20–50% of cases
	"Hydrocephalus ex vacuo"	Due to loss of brain tissue
Medical treatment	Osmotic agents	Isosorbide, glycerol
	Acetazolamide	Carbonic anhydraze inhibitor, slows CSF production
	Furosemide	Decreases ICP by increasing serum osmolarity
Surgical/interventional treatment	Evacuation of an intracerebral hemorrhage	Not indicated with exception of posterior fossa
	Serial ventricular taps	
	External ventricular drainage (EVD)/ventricular access device, ventriculosubgaleal shunt	
	Early VP shunting	High infection rate, peritoneal cavity not suitable

*CPP: Cerebral perfusion pressure

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piece of shunt tubing exiting the reservoir and ending in a subgaleal pocket. CSF collects in the subgaleal scalp pocket and is slowly reabsorbed.
- No randomized multicenter trial comparing the two methods has been performed.
- Temporary shunts can be used for several months if necessary.
- Direct ventricular taps with a needle through the fontanelle should generally be avoided as a routine treatment modality, reserved for life-threatening emergencies.

Permanent CSF Diversion: VP-shunts are required in only a small minority of preterm infants who suffer IVH. These children are prone to cerebral palsy, epilepsy, cognitive delay, behavioral abnormalities regardless of whether the CSF diversion is effective.

Endoscopic Third Ventriculostomy: A very effective treatment for non-communicating hydrocephalus in children older than 2 years. Its use in communicating hydrocephalus remains controversial.
- Several studies have shown that endoscopic third ventriculostomy is not effective in more than half of infants with nonobstructive hydrocephalus.

Complications

Infection: Unlike in older children, shunt infections can often be treated initially with antibiotics without removal of the hardware. As part of their immature immune systems, some infants produce insufficient immunoglobulins and benefit from immunoglobulin G infusions.

Other Complications: Rarely, epidural, subdural, or intraparenchymal hematomas are observed on postoperative US. The hematomas arise from a combination of factors, including friable vessels and a relatively soft, deformable cerebral cortex.

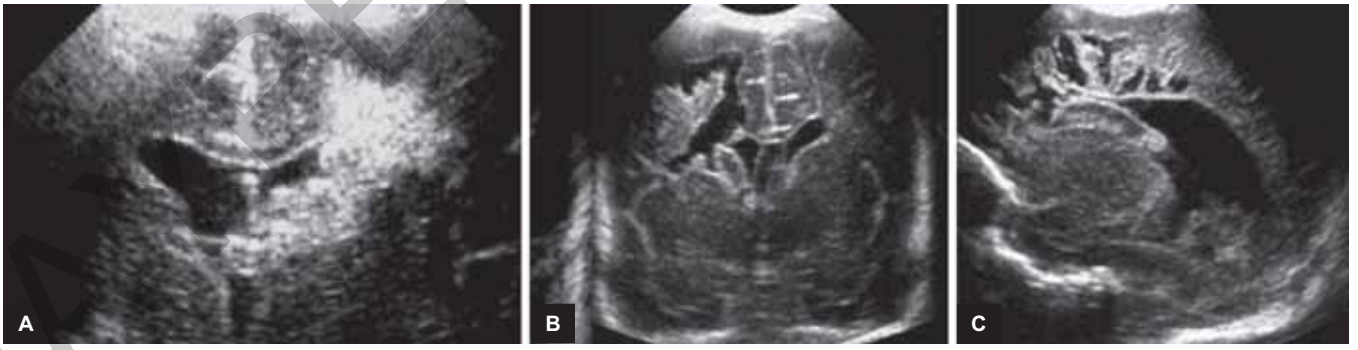
Neurological Outcome and Comorbidities: Preterm infants with IVH who require a shunt are more likely to have neurological deficits than those with IVH without PHH, suggesting that the need for a shunt is an independent risk factor for poor outcome.
- An inherent part of using standardized techniques to assess outcomes in young children is that studies of preterm infants tend to focus on severe, well-defined deficits present at about 2 years of age.
- Thus, the more subtle deficits that can significantly impact a child's achievement are not well documented.

Intraventricular Hemorrhage in Term Infants

Epidemiology: IVH in term infants is rare. In contrast to the minority of preterm infants who have low-grade IVH, 71.7% of term infants have a grade I lesion on cranial US, and 27.7% have grade II.

Pathophysiology: Similar to preterm infants, IVH appears to arise from both genetic and environmental factors. Male gender, poor Apgar score, RDS, and hyperbilirubinemia increase the risk of IVH.
- Term infants are more likely to develop IVH from venous infarction due to hypercoagulopathy or from hemorrhage of the choroid plexus (Fig. 10.3).

Clinical Presentation: Term infants with IVH typically present with lethargy or seizures. Seizures are often difficult to control. Most term infants with IVH benefit from a coagulation evaluation.



Figs 10.3A to C: Grade IV IVH in a term infant. (A) A large subependymal and a large area with increased echogenicity in the brain parenchyma lateral to the ventricle. This is probably the result of a venous infarct; (B and C) These venous infarctions resolve with cyst formation. A different patient with a grade 4 hemorrhage at a later stage with extensive cyst formation. These cysts can merge with the lateral ventricle, finally resulting into a porencephalic cyst

Source: http://www.radiologyassistant.nl

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- Many may eventually develop symptomatic hydrocephalus and require a shunt, usually during the first year.

Treatment: Term infants can undergo ventriculoperitoneal shunt insertion similar to other full-term neonates and infants.

NORMAL-PRESSURE HYDROCEPHALUS

Clinical Findings and Differential Diagnoses

- The concept of a "symptomatic occult hydrocephalus with 'normal' CSF pressure" was described by Hakim and Adams in 1965. Current guidelines deal primarily with idiopathic as opposed to secondary NPH, which occurs after trauma, subarachnoid hemorrhage, intracranial surgery, or meningitis.
- The classic finding in NPH is a clinical triad of symptoms—gait disturbance, dementia, and urinary incontinence.

Gait Disturbance: The most common initial symptom and occurs in almost 90% of patients.

- Common initial symptoms include unsteadiness, recurrent falls, shuffling, and reduced walking speed.
- More advanced symptoms include difficulty initiating gait and imbalance on turning.
- Gait in patients with NPH is described as being "magnetic" in nature, characterized by broad based, slow small steps with reduced height clearance as though the feet are "stuck to the floor."
- The gait disturbance may be difficult to distinguish from other types of frontotemporal pathologies.
- Unlike patients with Parkinson's disease, who are able to increase their stride length and walking speed with the aid of external cueing such as counting, patients with NPH have a gait apraxia unresponsive to such aids.
- In addition, patients with NPH tend to mobilize with a relatively preserved arm swing (unlike in Parkinson's).
- The presence of upper motor neuron signs or lower limb weakness may be indicative of cervical myelopathy and lumbar canal stenosis, respectively.
- The anatomic basis for gait disturbance in patients with NPH remains controversial.

Urinary Incontinence: May be a separate symptom or may be a consequence of gait disturbance or cognitive impairment. Some patients have urinary frequency rather than true incontinence.

- Thought to be due to involvement of the sacral fibers of the corticospinal tract (to Onuf's nucleus).
- Because urgency of micturition and incontinence are both common problems in older age, the clinical finding may be that of a change or worsening of urinary symptoms rather than a new problem.

Cognitive Impairment: NPH is estimated to account for <5% of all cases of dementia.

- The pattern of NPH appears to be a subcortical frontal dysexecutive syndrome.
- Cognitive deficits typically include memory loss, reduced attention, difficulty planning, slowness in thought, apathy.
- This differs from the cortical deficits of aphasia, apraxia, and agnosia seen in patients with Alzheimer's disease.
- The most difficult DD in the context of NPH is Binswanger's disease, a form of subcortical vascular encephalopathy. The pathologic changes are the result of small-vessel ischemia.

Neuroradiologic Features

Guidelines: CT or MRI is required to establish the diagnosis of NPH. Ventricular enlargement not entirely attributable to cerebral atrophy or congenital enlargement (Evans' index >0.3 or comparable measure) should be present (Figs 10.4 and 10.5).

***Evans' index*:** Defined as the maximal width of the anterior ventricular horns divided by the maximal width of the calvaria at the level of the foramen of Monro. Defined as hydrocephalus when >0.3 (Fig. 10.6).

***Bicaudate ratio*:** An alternative measurement, has excellent interobserver agreement and is more sensitive to changes in ventricular size. This is the minimal intercaudate distance divided by the brain width along the same line. Significant ventriculomegaly is defined as a ratio of ≥0.25.

Supportive features required

Enlargement of the temporal horns—not attributable to hippocampus atrophy

Callosal angle of 40 degrees or greater

Evidence of altered brain water content (periventricular changes)—not attributable to vascular ischemic

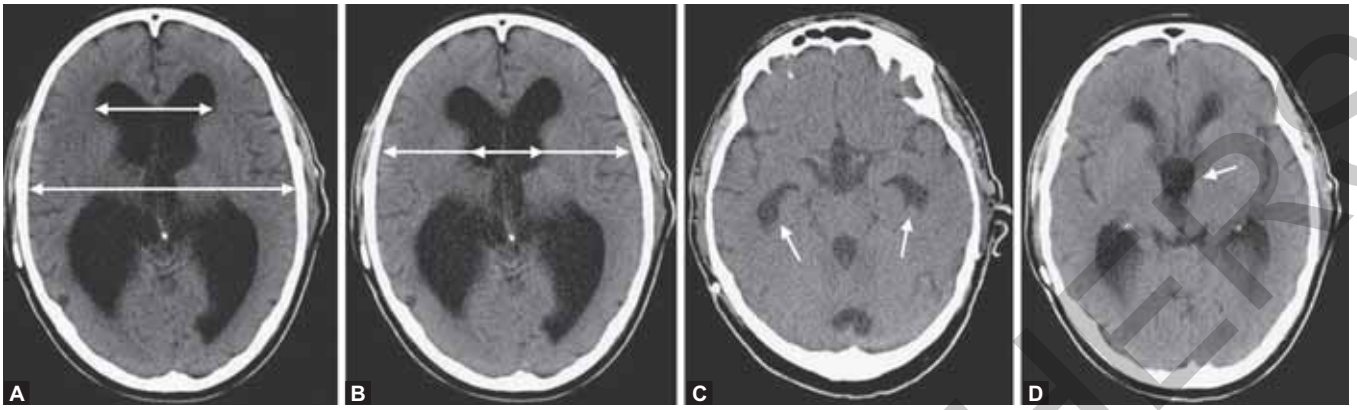
Aqueductal/fourth ventricular flow void on MRI.

- The imaging finding of deep white matter hyperintensities in a patient with NPH has been shown to be inversely correlated with shunt responsiveness. Such white matter hyperintensities are probably a marker of comorbidity.

Supplementary Prognostic Testing

- A single standard for the prognostic evaluation of patients with idiopathic NPH is lacking.

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Figs 10.4A to D: Hydrocephalus. (A) Evans' index = 0.46. (ventriculomegaly >0.3); (B) Bicaudate ratio is 0.35. (ventriculomegaly >0.25); (C) Dilation of the temporal horns; (D) Dilation of third ventricle

Source: http://www.radiologyassistant.nl



Fig. 10.5: NPH. MRI demonstrates prominent ventricles, without an obstructive lesion. The ventricles are large compared to the sulci and there is some crowding of the sulci superiorly with somewhat prominent sylvian fissures. Additionally the flow void in the aqueduct is more pronounced than usual, indicating rapid flow

Source: Gaillard F. www.radiopaedia.org

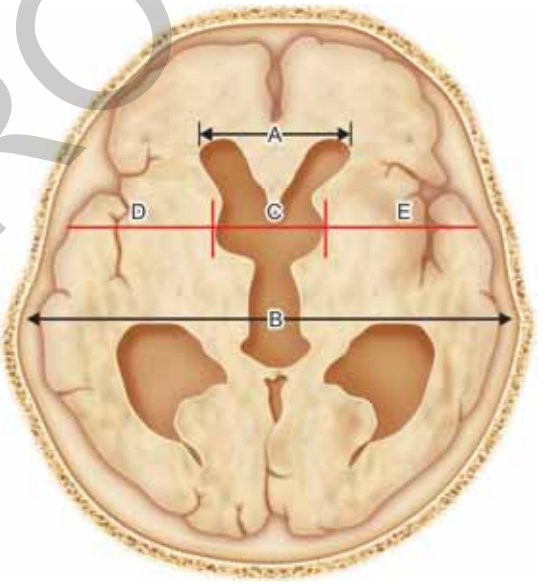


Fig. 10.6: Evans index: A/B, Bicaudate indexL C/ (C+D+E)

- Radionuclide cisternography is no longer a favored option because this technique does not improve the diagnostic accuracy of combined clinical and CT criteria in patients with presumed NPH.

Lumbar puncture (LP): Shown to produce a specificity of 100% with a sensitivity of 26%, provided that it is performed at a high volume (withdrawal of 40 to 50 mL of CSF). Symptomatic improvement after removal of CSF has a high PPV (73–100%).

- Remember that improvement after a shunt is often delayed in many patients.
- Its low sensitivity precludes using this method as a diagnostic tool for exclusion, used as a first-line tool.

Prolonged lumbar drainage: >300 mL, associated with highest sensitivity (50–80%), specificity (80%), PPV (80–100%).

- This method requires inpatient stay and carries a risk for nerve root irritation, hemorrhage, infection.

Measurement of CSF outflow resistance: Thought to reflect the CSF absorption pathways. Fluid is injected into a CSF space either by bolus or infusion. CSF outflow resistance can then be calculated. The advantage of this technique is that it requires only day attendance and can also be performed through a preimplanted ventricular reservoir device.

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NEURORADIOLOGIC FEATURES OF HYDROCEPHALUS

SHUNTING

Valve Design and Terminology

- During the past 50 years, the predominant theme in the evolution of valve design has been the goal of preventing CSF overdrainage—anti-siphon devices, flow-restricting elements, multistage valves, adjustable valves.
- Studies demonstrate that the in vivo behavior of even the simplest ventriculoperitoneal shunt with a standard pressure valve, is poorly predicted by the first-order, steady-flow equations that are the basis of many designs.
- There is no single valve mechanism, design, or arrangement that is the "best". Shunt management is a trial-and-error process.

Differential pressure valve: A common misconception is that the valve opening pressure must be lower than the ventricular pressure (as measured at the time of surgery) for CSF to flow down the shunt. This is clearly an invalid assumption (Fig. 10.7).

- An intracranial pressure reduction occurs even with a very high valve opening pressure.

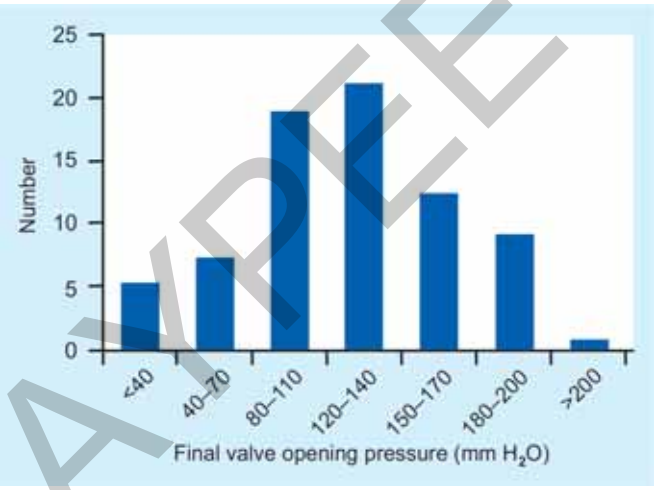


Fig. 10.7: Range of valve opening pressures in the treatment of idiopathic NPH. Histogram of final differential valve opening pressure values shows a gaussian distribution centered at approximately 140 mm H_2O. The wide range of valve opening pressures required indicates that no single valve opening pressure is appropriate for the treatment of idiopathic NPH

- The misconception arises from oversimplification of intracranial pressure and CSF flow hydrodynamics.
- In reality, the intracranial pressure waveform is pulsatile, with significant elevations of intracranial pressure occurring because of coughing and valsalva maneuvers as well as intrinsic vasomotor changes.
- The unidirectional flow valve results in the shunt's draining CSF even with opening pressures exceeding the mean intracranial pressure. This demonstrates that use of a low-pressure valve setting is not necessary and results in excessive CSF drainage in many patients.

Adjustable ("Programable") Valves: A "programable" or adjustable valve is created by adding a mechanism. That enables precise changes of the spring tension of a differential pressure valve (Fig. 10.8).

- There are several competing designs enabling this—all incorporating a magnetic actuation of a rotor.
- Adjustable valves arose from the realization that fixed-pressure differential pressure valves result in either

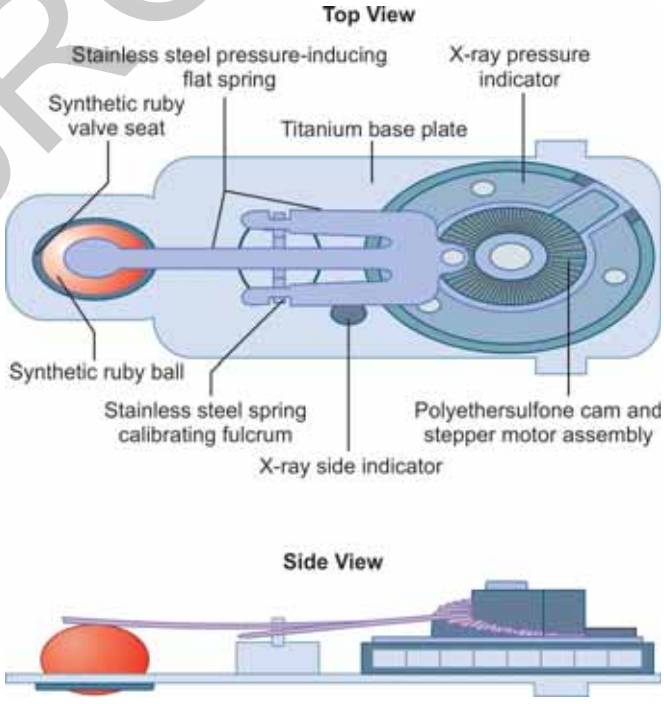


Fig. 10.10.8: Schematic diagram of the Codman Medos programable valve. The valve has an adjustable ball-spring mechanism. A cam and stepper motor assembly (akin to a spiral staircase) is used to adjust the tension on the spring. The motor assembly is adjusted with an externally applied magnetic field from the valve programer

Source: Drake JM, Sainte-Rose C. The Shunt Book. New York: Blackwell Science; 1995:94-95.

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overdrainage or underdrainage in a significant number of adult patients.
- Some remain reluctant to use adjustable valves. On their side are the results of a prospective, randomized trial comparing the adjustable valve and a standard differential pressure valve that failed to demonstrate any difference.
- Another drawback of adjustable valves has been MRI compatibility. To date, two manufacturers (Sophysa Polaris and Aesculap proGAV) have designed a locking mechanism that in theory prevents resetting of the valve.

Siphon-control and Anti-siphon Devices

- We refer to these collectively as anti-siphon devices (ASDs), although there are differences. ASDs are add-on devices, meaning that they are used in conjunction with a differential pressure valve mechanism.
- In general, the device is based on a membrane that is mechanically coupled to the subcutaneous tissue overlying it. The pressure differential between the internal valve lumen and the atmosphere, transmitted across the skin and ASD membrane, determines the flow-pressure characteristics of the ASD device.
- ASDs were developed on the basis of the premise that "siphoning" is the etiology of shunt-related CSF overdrainage. Better termed gravity-dependent drainage, occurs as the result of gravity-driven CSF flow down the distal catheter when the patient is in the upright position.
- Early studies documented significantly negative intracranial pressures in shunted patients in the upright position. Thus, it was natural to assume that overdrainage complications (subdural hematomas) were due to this gravity-dependent drainage.
- Current opinion is that gravity-dependent overdrainage plays a lesser role in the etiology of complications.
- A shunt with a differential pressure valve essentially lowers the intracranial pressure nearly equally across the head-of-bed angulation range. The degree of intracranial pressure reduction is largely a function of the valve opening pressure.

Key Point: A patient who fails to improve clinically (or deteriorates), a patient who remains with significant ventriculomegaly that did not change, a patient who has low measured intracranial pressure, or a patient with an ASD should not be written off as a nonresponder. The diagnosis of low-pressure hydrocephalus should be considered.

Flow Restriction Devices

- Another approach taken to counteract shunt overdrainage is the incorporation of a CSF flow restriction mechanism. Thus, by limiting the maximum CSF flow rate, overdrainage should be averted.
- A large, prospective, randomized study comparing a standard differential pressure valve, the Medtronic Delta valve, and the Orbis-Sigma valve (the original design, predating the Orbis-Sigma II) found no statistical difference in the rate of ventricular reduction, the final ventricle size, or the incidence of clinical shunt failure.
- Another approach to flow restriction is the incorporation of a high-resistance element. The Codman SiphonGuard is a coiled helical device that is placed immediately distal to a differential pressure valve (adjustable or fixed pressure).
- Unlike some ASDs, the SiphonGuard device is unaffected by scar tissue encapsulation or external pressure.
- Medtronic manufactures a peritoneal catheter with a smaller internal diameter, which also achieves a fixed added flow resistance. This catheter is intended to be used in conjunction with a valve.

Gravitational Devices

- Gravity-induced CSF flow (siphoning) is considered by many to be the primary cause of overdrainage. To offset the negative pressures generated by the long hydrostatic column, the gravitational (also termed hydrostatic) device interposes a very high differential pressure valve while the patient is in the upright position. When the patient is in the recumbent position, a low (lower) opening pressure is operational. CSF is diverted to the high-pressure valve in the upright position.
- If used alone without a series adjustable differential pressure valve, gravitational devices do not prevent overdrainage or underdrainage clinical conditions.

Other Valve Characteristics

- There are other practical considerations to valve selection. One is the physical profile of the valve.
- A high-profile (prominent) valve housing can have significantly negative cosmetic consequences, likely to cause overlying skin breakdown in susceptible patients.

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RECOMMENDATIONS FOR SPECIFIC CHALLENGING SCENARIOS

High Protein Concentration or Cell Count in the Cerebrospinal Fluid

- High CSF protein concentration alone does not appear to increase the incidence of shunt obstruction.
- It is the cell count that is more problematic.

Patient Undergoing Anticoagulation or Antiplatelet Therapy

- Aspirin is stopped 8 days before surgery; clopidogrel (Plavix) is stopped 14 days before surgery. Warfarin therapy is reversed and, if necessary, enoxaparin is temporarily prescribed and discontinued 24 hours prior to surgery.
- After surgery, warfarin can be restarted as early as 3 to 5 days. Aspirin or clopidogrel is restarted 7 to 10 days after surgery.
- Current view is that combined aspirin-clopidogrel is at least relatively contraindicated after shunt surgery because of high-risk of CSDH.

The Hemicraniectomy Patient

- Management of hydrocephalus in a patient with a hemicraniectomy is challenging. This state is prone to both shunt underdrainage and overdrainage situations in the same patient, depending on body position.
- If possible, perform the cranioplasty before the shunt placement or as soon as possible if a shunt is already present.

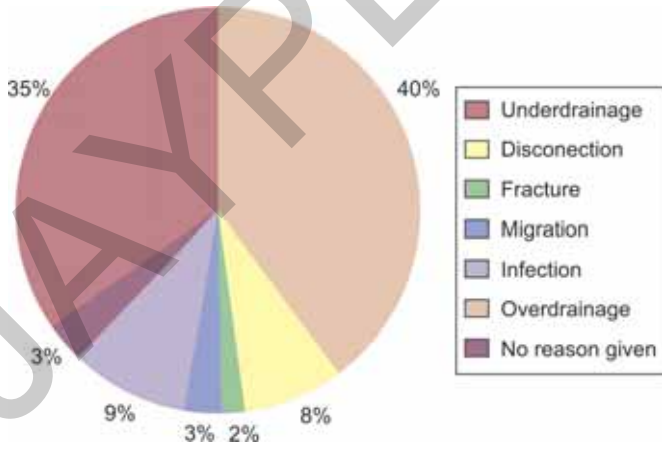


Fig. 10.9: Reasons given for shunt revision in adults (17 years and older)

- During this cranioplasty, consider placing multiple central dural tack-up sutures ("Poppen" sutures) every 2 cm. Failure to do so greatly increases the risk of an epidural hematoma once the shunt is placed.

Shunt Operations Associated with Other Procedures

- For patients undergoing a lengthy craniotomy (such as for tumor resection), avoid placing a shunt at the same operation. There is typically a higher CSF cell count at the time, and the longer operative time increases the risk of shunt infection.

SHUNT COMPLICATIONS

Figures 10.9 and 10.10 overview the reasons given for shunt revision in adults, and the estimated risk of subdural hygroma formation.

Evolution of Chronic Subdural Hematomas

The presence of defective neovasculature certainly potentiates the process of rebleeding and absorption of plasma proteins into the subdural space, but this does not fully explain the evolution of CSDHs (Fig. 10.11).

- Normal hemostatic mechanisms should halt the process long before the CSDH reaches a clinically significant size.

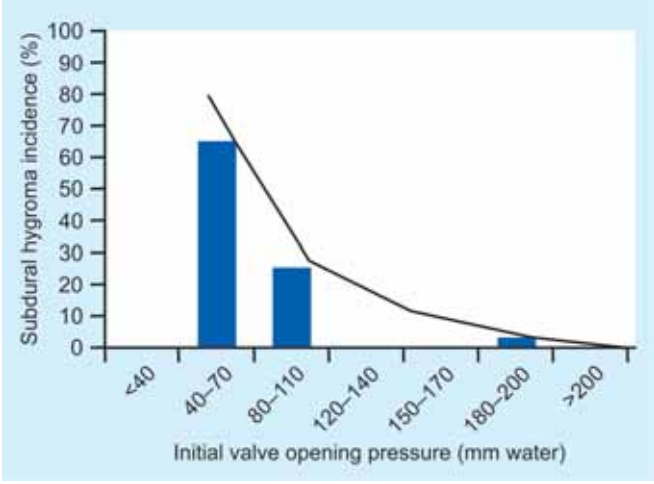


Fig. 10.10: Estimated risk of subdural hygroma formation with idiopathic NPH. The Dutch NPH study documented a subdural hygroma (effusion) incidence of ~70% and ~30% with low- and medium-pressure differential pressure valves, respectively. Thus, a direct relationship between valve opening pressure and subdural hygroma incidence

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- Instead, evidence supports presence of a localized coagulopathy within the CSDH.
- There is accelerated fibrinolytic activity after the rapid and defective clot formation. The end result is a milieu of anticoagulant proteins (chiefly FDPs) and depleted coagulation factors.
- Localized coagulopathy and defective clot formation within the CSDH completes the cycle of bleeding, membrane formation, defective vascularization, fibrinolysis, coagulopathy, and rebleeding.

Recurrence:

- Reported recurrence rates of CSDH range from 2–37%, largely because the mechanisms have not been fully elucidated.
- Theories range from recurrent outer membrane bleeding, to brain stiffness preventing reexpansion, to altered cerebral blood flow and its effects on reexpansion.

Shunt Infections and Their Treatment

General: Shunt infection rates differ among reported studies (Table 10.5).

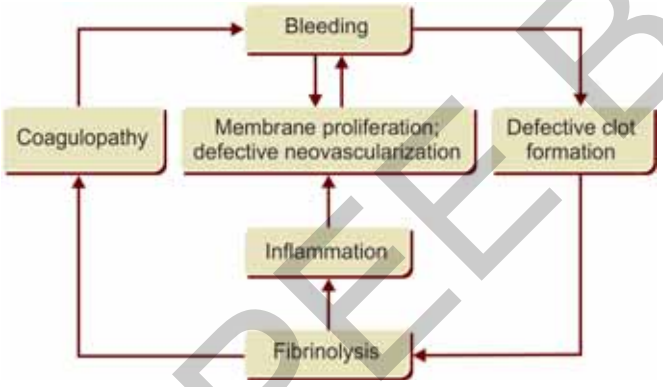


Fig. 10.11: Defective clot formation and increased fibrinolysis contribute to rebleeding into the chronic acute subdural hematoma

Studies

***Borgbjerg 884 individuals (1958-1989)*—6.2%** for the first postoperative month, overall rate of 7.4%.

Casey

A 344 patients—Nearly 40% of newly inserted shunts failed in the first year, with a shunt infection rate of 8.1% over the follow-up period of 1 to 3 years.

- These data reflect the shunt infection rate following a single shunt procedure.
- If patients are followed for longer periods, the cumulative likelihood of shunt infection related to multiple shunt procedures ranges from 19% to 38%.

The hydrocephalus association database: Nearly 40% of patients who had hydrocephalus for at least 10 years experienced at least one shunt infection.

A reasonable conclusion: Overall infection rate for new shunts is between 3–8%.

Timing of infection: Among pediatric patients, the majority of shunt infections occur relatively soon after operation.

***Casey and colleagues*: 92%** of infections occurred within 3 months of initial shunt placement.

Infants: Appear to have higher shunt infection rates, the majority occurr in the first 3 months.

***Delayed shunt infections*:** Occur 2–3 years after shunt insertion.

Risk Factors:

Most Important

Patient age—Neonates and very young children at greatest risk.

Children 6 months or younger—19%.

Older children—7%.

Explanations—Increased susceptibility to pathogens due to the relative deficiency of the neonatal immune system.

- Some data suggest that more highly adherent strains of coagulase-negative *Staphylococcus*, the most prevalent organism in shunt infections, occur in neonates.

Table 10.5: CSF shunt infections—presenting features in children

Feature	*VP Shunt (%)*	*VA Shunt (%)*	*Most Common Organisms*	*Percent*
Fever	95	100	*S. epidermidis* (SE)	32–57
Shunt malfunction	57	14	*S. aureus* (SA)	4–38
Abdominal pain	48	0	SA + *S. viridans*	4–15
Meningismus	29	0	Gram negatives ± SE	15 (3)
Headache	14	14	SE + *Enterococcus*	7
Irritability	19	43	SE + *S. Pyogenes*	4
Nephritis	0	14	*Enterococcus/Candida*	4

VA, ventriculoatrial; VP, ventriculoperitoneal

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- Two studies reported that patients with congenital hydrocephalus had lower rates of infection than did patients with either postinfectious or posthemorrhagic hydrocephalus.
- With respect to surgical factors, there is a paucity of convincing data.

Outcome

Reinfection rate—26%, two thirds of the reinfections caused by the same organism.

- Surprisingly, the reinfection rate was not affected by the duration of antibiotic therapy.
- Shunt infection is associated with poor functional results - mean "late" postoperative IQ is 40 among children who had at least one shunt infection, versus an IQ of 60 among children without infection.

Prevention—Data regarding the use of prophylactic antibiotics are strongly suggestive, but confirmation is lacking.

***Antibiotic-impregnated shunt*—**Rifampin, clindamycin, minocycline. A few studies show a reduction in infection rates. A particular benefit was noted in the neonatal subgroup receiving first-time shunts.

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