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2.1 Hydrocephalus

Hydrocephalus is a condition resulting from an imbalance between the production and absorption of cerebral spinal fluid (CSF). This imbalance results in an increased volume of spinal fluid, dilation of the ventricular system, and often increased intracranial pressure. Hydrocephalus onset can be acute and occur over hours or days. It may also be chronic and occur over months or years. Hydrocephalus can occur as an isolated condition or one associated with numerous other neurological conditions and diseases.

2.2 History of Hydrocephalus

The term hydrocephalus is derived from the Greek words “hydro” meaning water and “cephalus” meaning head. The description and treatment of hydrocephalus dates back to the eras of Hippocrates and Galen. Galen (130–200 AD) identified the ventricles. He believed that the soul was purified through the pituitary gland. Waste was discharged via the nose as “pituita.” During the renaissance, Vesalius (1514–1564) described the ventricular system in his original text on human anatomy. A

century later, Franciscus Sylvius (1614–1672) described the cerebral aqueduct. Morgagni (1682–1771) described the pathology of hydrocephalus, and Monro (1733–1817) named the intraventricular foramen. In 1768, Whytt distinguished internal and external hydrocephalus.

Early treatment included bleeding, purging, surgical release of the fluid, puncturing the ventricles to drain the fluid, injection of iodine or potassium hydriodate into the ventricles, binding of the head, application of a plaster of herbs to the head, application of cold wraps to the head, lumbar puncture, and diuretics. Confusion about hydrocephalus persisted into the 1800s. It was thought to be caused by fevers, rheumatism, pulmonary consumption, and worms; however, treatment did not change.

The earliest attempts at surgery occurred during the late 1800s. The first shunts diverted spinal fluid from the ventricles to the subcutaneous or subdural spaces. During the early 1900s, other surgical procedures were attempted to treat the condition. These procedures included surgical removal of the choroid plexus, diversion of spinal fluid through a third ventriculostomy, and continued attempts at shunting, including attempts to shunt into the vascular space. Most of these patients did poorly, and either suffered the consequences of prolonged increased intracranial pressure or died. Many institutions cared for and housed these disabled

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children with very large heads, small bodies, and severe mental retardation.

Modern shunting procedures began in the 1950s with the introduction of the antireflux valve. The first valves, developed by Nulson and Spitz in 1952, used a spring and steel ball valve. Holter then developed the first slit valve. He was particularly interested in shunt development, as he had a son with a myelomeningocele and hydrocephalus. These first modernized shunts diverted CSF from the ventricles to the right atrium of the heart. The ventricular to peritoneal shunt became the preferred shunt in the 1970s because it allowed for the child to grow and not outgrow the length of the shunt tubing. This has remained the preferred shunt procedure among modern neurosurgeons. Neurosurgeons have also placed shunts leading from the ventricle to the pleural space, gall bladder, ureter, or fallopian tube if the abdominal cavity is not a suitable place to terminate the shunt. Numerous improvements of shunt hardware have occurred in the last four decades.

A genetic understanding of hydrocephalus and diseases associated with hydrocephalus has occurred in the last decade. Such knowledge of genetics has allowed for improved prenatal diagnosis and genetic counseling.

2.3 Incidence of Hydrocephalus

Hydrocephalus is the most common neurosurgical problem encountered by pediatric neurosurgeons. The overall incidence is difficult to determine, as hydrocephalus can occur as an isolated condition or in conjunction with many other neurological diseases and conditions. The overall incidence of hydrocephalus at birth is 0.5–4 per 1,000 live births. As an isolated congenital disorder, the incidence of hydrocephalus is 0.5–1.5 per 1,000 live births. Hydrocephalus occurs in about 80–85 % of infants born with a myelomeningocele. Because hydrocephalus is associated with so many other diseases and conditions, it is impossible to know how many such children actually exist in the general population. A recent study revealed that pediatric hydrocephalus results in 38,200–39,900 annual hospital admissions, with

total hospital charges of \$1.4–2.0 billion dollars. Hydrocephalus accounts for 3.1 % of all pediatric hospital charges (Simon et al. 2008). Surgeries to place and revise shunts comprise approximately half of a pediatric neurosurgeon's annual operative cases (McLone 2001).

2.4 Prognosis

The prognosis for children with hydrocephalus has markedly improved with modern shunting. The natural history of unshunted hydrocephalus was studied, and it revealed a 46 % survival rate for 10 years (Laurance and Coates 1962). Of the surviving population, 62 % suffered from intellectual impairment (Laurance and Coates 1962). Children who are adequately treated for hydrocephalus have a considerably better outcome. Their survival rate after 10 years is 95 %, and only 30 % have impaired intellectual function (Shurtleff et al. 1973).

The prognosis of an infant or child with hydrocephalus is mostly dependent on the underlying cause of the hydrocephalus. Prognosis may also be related to the complications that occur, such as shunt malfunctions and infections. The best predictors of a good outcome are the prompt treatment of the hydrocephalus and the ability of the brain to grow normally in the newborn once a functioning shunt is placed. Shunt dependency is associated with a 1 % mortality rate per year (Ditmyer 2004).

2.5 Classifications of Types of Hydrocephalus

Hydrocephalus is subdivided into several different categories. Communicating and noncommunicating are the most common categories. These terms were previously used interchangeably with obstructive and nonobstructive. The latter terms have fallen from use, as it is believed that in almost all cases of hydrocephalus, there is some obstruction of CSF reabsorption; the exception is the rare state of overproduction of CSF. Hydrocephalus is also subdivided into

Table 2.1 Classifications of hydrocephalus

<i>Communicating</i>	
Congenital	
Achondroplasia	
Associated with craniofacial syndromes	
Acquired	
Posthemorrhagic: intraventricular or subarachnoid	
Choroid plexus papilloma or choroid plexus carcinoma	
Venous obstruction as in superior vena cava syndrome	
Postinfectious	
<i>Noncommunicating</i>	
Congenital	
Aqueductal stenosis	
Congenital lesions (vein of Galen malformation, congenital tumors)	
Arachnoid cyst	
Chiari malformations either with or without myelomeningocele	
X-linked hydrocephalus	
Dandy-Walker malformation	
Acquired	
Aqueductal gliosis (posthemorrhagic or postinfectious)	
Space-occupying lesions such as tumors or cysts	
Head injuries	

congenital versus acquired and internal versus external (Table 2.1). Other categories include normal pressure hydrocephalus and ex vacuo hydrocephalus.

2.5.1 Communicating Hydrocephalus

Communicating hydrocephalus is a condition that results when the arachnoid villi are unable to adequately reabsorb cerebral spinal fluid. Intraventricular or subarachnoid hemorrhage may cause the arachnoid villi to become unable to function adequately, either temporarily or permanently. This is a consequence of the effect of the end products of red blood cell breakdown on the arachnoid villi. Infectious processes such as meningitis may also render the arachnoid villi to be nonfunctional (due to, e.g., toxins or scarring). Communicating hydrocephalus may also be due to the overproduction of CSF. This is rare and is

usually associated with a choroid plexus papilloma or a choroid plexus carcinoma.

2.5.2 Noncommunicating Hydrocephalus

Noncommunicating hydrocephalus is a condition that results when the ventricular system does not communicate with the arachnoid villi due to some obstruction in the normal pathways of CSF flow. Consequently, CSF is produced in the ventricular system but cannot flow to the arachnoid villa to be reabsorbed. Such obstruction can occur when pathways are blocked by a tumor, congenital abnormalities of the brain, cysts, inflammation from infection, or any other condition that interferes with the patency of these pathways. Some consider the failure of the arachnoid villi to reabsorb CSF to be an obstruction at the level of the arachnoid villi.

2.5.3 Congenital Hydrocephalus

Congenital hydrocephalus is caused by any condition that existed before birth. The hydrocephalus may or may not be present at birth. Examples include aqueductal stenosis, Dandy-Walker malformation, and X-linked hydrocephalus. Congenital hydrocephalus is also associated with myelomeningocele, Chiari malformations, encephalocele, and with prenatal infections such as cytomegalo inclusion virus (CMV) or rubella.

2.5.4 Acquired Hydrocephalus

Acquired hydrocephalus is hydrocephalus resulting from a condition that did not previously exist in the patient. The condition either obstructs normal spinal fluid flow, causes overproduction of CSF, or prevents reabsorption of CSF. Examples include tumors that obstruct CSF flow and other space-occupying lesions that were not congenital. Infection in the brain may also occlude small passageways. Overproduction of spinal fluid may be caused by a choroid plexus tumor. Acquired

conditions that interfere with reabsorption of CSF include intraventricular hemorrhage (IVH) and subarachnoid hemorrhage.

2.5.5 Internal Hydrocephalus

Internal hydrocephalus refers to ventricular dilation and the associated pathophysiology. The term hydrocephalus is used most commonly to refer to internal hydrocephalus.

2.5.6 External Hydrocephalus

External hydrocephalus refers to the accumulation of spinal fluid in either the subarachnoid or subdural spaces. CSF collection in the subarachnoid space may be a benign condition in infancy, which is called benign subdural hygromas of infancy or idiopathic external hydrocephalus of infancy. The neuroimaging characteristics of this condition typically show enlarged frontal subarachnoid spaces and moderately enlarged ventricles. Infants present with rapidly increasing head circumference and sometimes developmental delays. Some children may show temporary or permanent psychomotor delays (Zahl et al. 2011). This is usually a self-limiting condition. The child is usually treated conservatively, although some may require a shunt.

CSF mixed with blood in the subdural space may not be benign and usually requires further investigation and treatment, as it may be related to trauma (possibly nonaccidental trauma). If these fluid collections exert pressure on the brain and cause symptoms or cause very accelerated head growth, surgical treatment may be necessary.

2.5.7 Ex Vacuo Hydrocephalus

Ex vacuo hydrocephalus refers to a condition of brain volume loss. The condition may be present at birth. It may be the result of failure of the fetal development of the brain as in schizencephaly (abnormal development of the brain, leading to

the characteristic appearance of abnormal clefts in either one or both cerebral hemispheres) or hydranencephaly (abnormal development of the brain, leading to absence of the cerebral hemispheres of the brain). The brain may also undergo destruction or atrophy from infections, very poor nutrition, or unknown causes. The ventricles become large to “fill the space” where there is an absence of brain tissue and may or may not be under increased pressure. There is technically not an imbalance of CSF production and absorption but rather the actual loss of brain matter.

2.5.8 Idiopathic Normal Pressure Hydrocephalus

Idiopathic normal pressure hydrocephalus is primarily a condition of the elderly. It is a condition that occurs with normal intracranial pressure and ventricular dilation. These patients develop symptoms slowly over time. The classic symptoms include dementia, gait difficulties, and urinary incontinence. A shunt may be helpful if the symptoms improve after a lumbar puncture or lumbar drain.

2.6 Pathophysiology of Hydrocephalus

2.6.1 Overview of CSF Production and Flow Dynamics

Most of the CSF (approximately 60 %) is produced in the choroid plexus; the rest is produced in the ependymal of the cerebral ventricles, the aqueduct of Sylvius, and the subarachnoid space. Studies by Milhorat looking at CSF production after choroid plexectomy demonstrated that the total amount of produced CSF was reduced by only one-third, thus suggesting that other sites can produce larger amount of CSF (Milhorat 1982). He proposed that CSF is also produced as the result of cellular metabolism of periventricular cortical gray matter. These other areas account for 20–50 % of CSF production. CSF production requires the expenditure of energy (Albright et al. 2007).

2.6.2 CSF Pathways

CSF flows from the ventricles, passes through a series of channels, and exits the ventricular system via the fourth ventricle. There are two lateral foraminae on the lateral aspect of the fourth ventricle, named the foramen of Luschka, and medially located opening called the foramen of Magendie. After exiting the fourth ventricle, the CSF flows into the subarachnoid space and up over the convexities of the brain, to be absorbed into the large intracranial sinuses (Albright et al. 2007). Alternative pathways for CSF have been scientifically supported and include lymphatic drainage into the cervical lymphatic chain and paranasal sinuses. After being absorbed, the CSF is returned to the right atrium via the superior vena cava (Albright et al. 2007).

2.6.3 Intracranial Pressure

A study of rabbits by Dr. McComb found that CSF flows passively, and absorption of CSF does not require the expenditure of energy (Albright et al. 2007). For each drop of CSF that is produced, the same amount should be absorbed. Several factors affect the flow of CSF, including resistance, which may result from an obstruction or restriction of a pathway. Other considerations include the plasticity of the brain itself, as well as the flexibility of the intracranial venous structures.

Plasticity refers to the brain inability to change shape. For example, an increase in the intraventricular volume will enlarge the ventricles, causing distortion of the cerebral cortex. As we age, our brain may become stiffer. Neonatal brains are very elastic. Anoxic injury can change the brain's ability to maintain its normal stiffness and can also be hydroplastic. The intracranial venous system includes the dural sinuses which are more rigid than the cortical veins. Cortical veins join the dural sinuses at such an angle that a valvular mechanism is created and a pressure gradient is maintained. The jugular veins, returning the blood to the heart, have no valves. When we stand, negative pressure produced in the jugular

veins causes them to collapse and assist humans in maintaining normal intracranial pressure. Shunting systems are used when the CSF pathways are somehow obstructed. The valves that are used to regulate the flow attempt to mimic normal flow.

The normal rate of CSF production in infants and children is about 0.33 ml/kg/h. Normal newborns have about 5 ml total volume of CSF. Adults have about 125 ml of total CSF, with about 20 ml located within the ventricles. CSF is produced continually by the choroid plexus, which is located within the ventricles. It is continually reabsorbed by the arachnoid villi (Figs. 2.1 and 2.2).

The pathophysiology of hydrocephalus is much more complex than the radiographic picture. The computed tomography (CT) or magnetic resonance imaging (MRI) scan may reveal many structural changes including enlarged ventricles, thinning of the cortical mantle, distortion of structures, and possible transependymal flow of CSF. These visible changes may also affect the biochemistry, metabolism, and maturation of the brain. Adequate treatment and resolution of the dilated ventricles does not always reverse the other injuries that have occurred to the brain.

Three factors are critical in determining the severity of injury caused by hydrocephalus: age at onset, underlying cause (etiology), and duration of the hydrocephalus. Age is a salient factor because hydrocephalus may affect the normal maturation processes of the brain in addition to the other expected effects of increased intracranial pressure. Furthermore, the underlying disease process responsible for the hydrocephalus may have its own destructive effects on maturation and brain function. Examples of such diseases are encephalitis, meningitis, tuberous sclerosis, and tumors. Treatment of these diseases may also have destructive effects on the brain and brain maturation. For example, radiation treatment of brain tumors in very young children can interrupt normal maturation permanently and development does not always proceed normally, even after the resolution of the hydrocephalus. The duration of the hydrocephalus has a critical

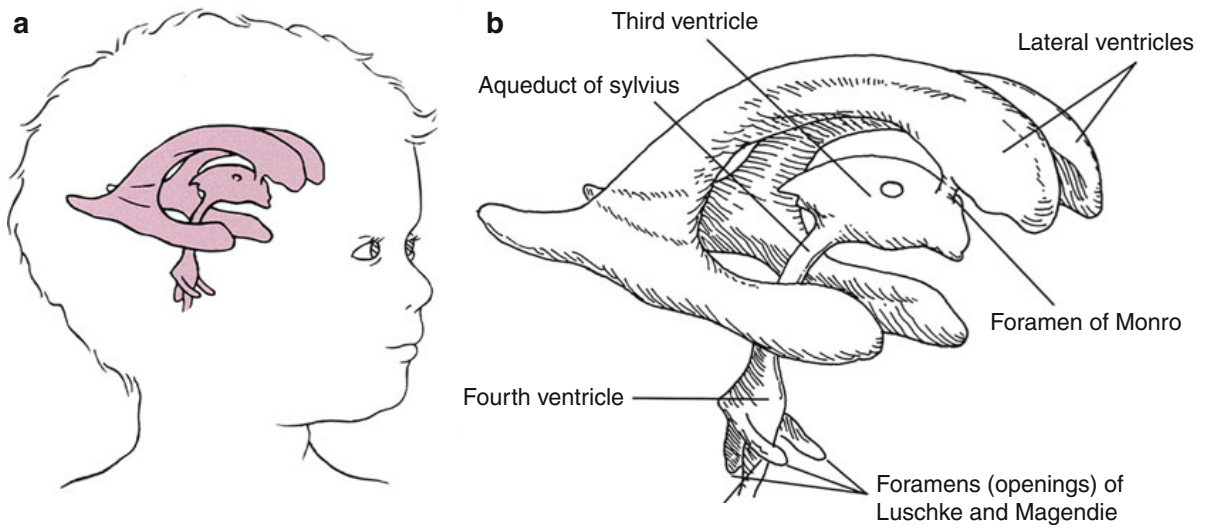


Fig. 2.1 Illustration of position and configuration of intracranial ventricles

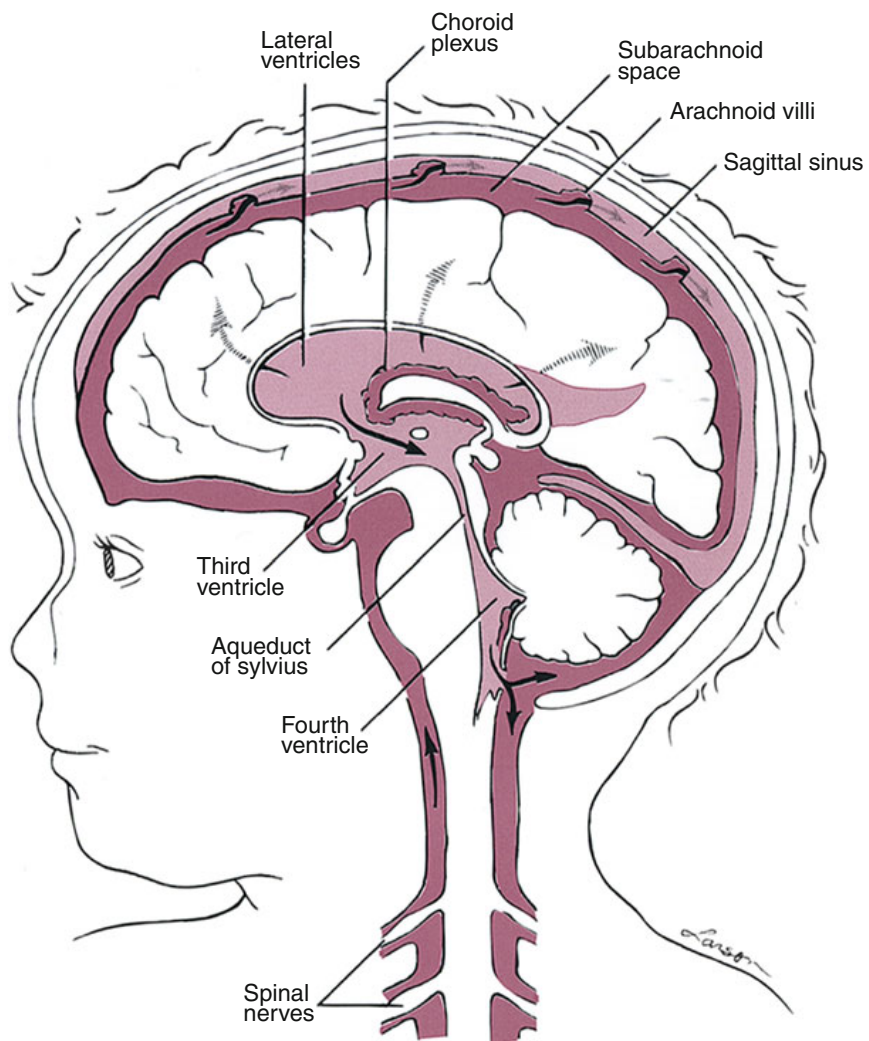


Fig. 2.2 Illustration of cross section of brain and ventricles shows pathways of CSF flow

role in determining the long-term recovery. Long-standing ventricular dilation and increased intracranial pressure tend to lead to poor recovery of function, even after ventricular size normalizes.

2.6.4 Structural Changes

Ventricular dilation seen on the CT or MRI is the hallmark of hydrocephalus. The temporal and frontal horns of the lateral ventricles usually dilate first and are sometimes asymmetrical. This is due to the accumulation of spinal fluid and leads to distortion of the adjacent structures, compression of the nearby white matter, reduction of cerebral cortex, and thinning of the cortical mantle. The ependymal cells lining the ventricles may become damaged and allow transependymal flow of CSF. The septum pellucidum may become damaged, leading to its disappearance and the formation of one large ventricular cavity. In some situations, ventricular size may not change even though pressure is elevated and symptoms are present.

2.6.5 Vascular Changes

The distortion of the brain tissue that occurs with hydrocephalus also affects the arteries, veins, and capillaries. Deep vessels are affected the most as they may be directly compressed from the increased ventricular size. Peripheral vessels are also affected as they try to supply the brain tissue that is suffering from the insult of increased intracranial pressure. Blood flow has been shown to be globally decreased to the brain in acute hydrocephalus (Da Silva et al. 1995). Blood flow is primarily decreased to the periventricular white matter in chronic hydrocephalus (Da Silva et al. 1995). Hypoperfusion may cause damage to neurons and glia and interfere with normal maturation of all brain structures.

2.6.6 Metabolic Changes

The brain of a child consumes about 50 % of total body oxygen, and an infant's brain consumes

more than 50 %. The adult brain consumes only about 20 % (Sokoloff 1989). The brain uses glucose as its primary source of energy with few exceptions. Therefore, any decrease in cerebral blood flow that decreases the amount of oxygen and glucose available can markedly alter metabolism. This impairment of metabolism may lead to damage to the brain. Furthermore, during infancy and childhood, a significant portion of the energy used by the brain is used for maturational activities such as myelination, neuronal maturation, and protein production. Normal maturation may be disturbed and possibly permanently altered, due to these metabolic alterations.

2.6.7 Cerebral Spinal Fluid Changes

Abnormal amounts of spinal fluid in the brain may lead to changes in the CSF itself. Metabolites may accumulate in the CSF during hydrocephalus. Protein levels in the CSF may be altered by the underlying cause of the hydrocephalus. For example, after an IVH, protein levels may be very elevated. If the hydrocephalus damages the ependymal cells lining the ventricles, the CSF may flow out of the ventricles into the periventricular white matter. CSF production may or may not decrease as intracranial pressure increases. As intracranial pressure increases, reabsorption of CSF may increase assuming that the arachnoid villi are functional.

2.6.8 Brain Tissue Changes

The white matter surrounding the enlarged ventricles is called the periventricular white matter. As the ventricles dilate, the white matter may become compressed, saturated with CSF, and possibly damaged. Periventricular leukomalacia may result from ischemia to affected white matter. The corpus callosum may also become thinned.

The myelination process may also be delayed in children with hydrocephalus. Myelination occurs in a stepwise fashion during development. If one step is interrupted, it cannot occur at a later

time, and this prevents subsequent steps in the overall process.

The cerebral cortex is also markedly affected by hydrocephalus. The cortex is thinned as it is pushed out by the ventricles and restricted by the skull. Histological changes within the cortex are usually subtle, but damage to cells occurs and results in a change in function.

The goal of treatment of hydrocephalus is to prevent further damage and to restore function. Treatment usually reverses symptoms of acute hydrocephalus. However, timing is critical and treatment should occur before vascular, metabolic, and other changes described interfere with normal maturation and brain function. Without prompt treatment, acute hydrocephalus and increased intracranial pressure can lead to brainstem herniation and death.

2.7 Etiologies of Hydrocephalus

Hydrocephalus is primarily a condition of obstructed CSF circulation or absorption. In infants and children, it may be congenital or associated with other congenital abnormalities. It may also be associated with central nervous system (CNS) infection, hemorrhage, tumors, or cysts.

2.7.1 Aqueductal Stenosis

In 1900, Bourneville and Noir noted an association between hydrocephalus and stenosis of the aqueduct of Sylvius (Cinalli et al. 2004). The aqueduct of Sylvius is a narrow passageway connecting the third and fourth ventricles. The most common cause of hydrocephalus in children is aqueductal stenosis, and it accounts for 70 % of cases (Greenberg 2010). Hydrocephalus due to aqueductal stenosis is characterized by enlargement of the lateral and third ventricles with a normal fourth ventricle. This constriction of the aqueduct of Sylvius is best seen on MRI scan (sagittal view).

Stenosis of this passageway may be congenital or acquired, although in 50–75 % of cases, the cause may be unknown. It may be associated with

Chiari I malformation, vein of Galen malformation, or Dandy-Walker malformation. Aqueductal stenosis may also be due to an X-linked recessive gene, *L1CAM* mutation, (Langingham et al. 2009) occurring only in males. This is rare and is associated with characteristic adducted thumbs, spastic paraparesis, and mental retardation. Acquired cases of aqueductal stenosis may be the result of hemorrhage, inflammation from infection, or obstruction from a nearby tumor or cyst.

2.7.2 Myelomeningocele and Chiari II Malformation

Myelomeningocele is a neural tube defect that occurs during embryonic development resulting in failure of the neural tube to close. This malformation involves the entire CNS. At the level of the spinal defect, there is a midline lesion containing meninges, spinal cord, nerves, and CSF. The bony structures of the spine may be abnormal or absent. Associated abnormalities in the brain include Chiari II malformation, hydrocephalus, and possibly other structural abnormalities.

Chiari II malformation occurs in almost all infants born with myelomeningocele.

It is a malformation of the hindbrain, fourth ventricle, and brainstem and includes herniation of these structures into the cervical spinal canal. Herniation of the brain stem and fourth ventricle may result in obstruction of CSF flow. The development of the hydrocephalus is related to the Chiari II malformation, aqueductal stenosis, venous hypertension in the posterior fossa, and closure of the myelomeningocele (Sgouros 2004a, b).

Hydrocephalus develops in about 85 % of children with myelomeningocele. Approximately 50 % have significant hydrocephalus at birth (Wang and Avellino 2005). About 80–90 % will eventually require a CSF shunt (Dias 2005) or an endoscopic third ventriculostomy. Before modern shunting of these infants in the 1960s, only about 20 % of non-shunted children lived into adulthood. Today, the hydrocephalus can usually be adequately treated. Infants and children who die from this complex condition usually die from the Chiari II malformation and brainstem dysfunction. New

research shows that patients who have undergone prenatal closure of the myelomeningocele have lower rates of hydrocephalus, and in some patients, there was no formation of a Chiari II malformation (Adzick et al. 2011).

Myelomeningocele is discussed in detail in Chap. 4.

2.7.3 Chiari I Malformation

Chiari I malformation is one of the four types of Chiari malformations. In Chiari I, the cerebellar tonsils are elongated and herniated into the cervical spinal canal. Chiari I is not associated with myelomeningocele and may be acquired from increased intracranial pressure or occur as an isolated condition.

Hydrocephalus occurs in 10 % of children with Chiari I malformation, most likely due to blockage of CSF flow at the craniovertebral junction. A small posterior fossa may also alter CSF flow. Treatment for patients with symptomatic Chiari I malformation is often a posterior fossa decompression. A small percentage of children develop hydrocephalus after the decompression.

Chiari malformations are discussed in detail in Chap. 5.

2.7.4 Dandy-Walker Malformation

Dandy-Walker malformation is a continuum of posterior fossa abnormalities including Dandy-Walker malformation and Dandy-Walker variants. The abnormalities associated with these conditions include cystic dilation of the fourth ventricle, partial or complete absence of the cerebellar vermis, upward displacement of the tentorium, and usually hydrocephalus. Dandy-Walker may be differentiated from a posterior fossa cyst by the atrophy or agenesis of the vermis seen on MRI scan. Dandy-Walker malformation/variant may also be associated with other intracranial abnormalities in 70 % of patients. These abnormalities include agenesis of the corpus callosum, aqueductal stenosis, schizencephaly, holoprosencephaly (failure of the prosencephalon, the embryonic

forebrain, to sufficiently divide into the double lobes of the cerebral hemispheres, resulting in a single-lobed brain and severe craniofacial defects), neural tube defect, and occipital encephalocele. Dandy-Walker is found in 2–4 % of all children with hydrocephalus (Greenberg 2010). Other abnormalities associated with Dandy-Walker malformation/variant include congenital heart defects, renal malformations, polydactyly/syndactyly, cleft palate, perineal malformations, Klippel-Feil malformation, and facial hemangiomas.

Hydrocephalus occurs in 90 % of children with Dandy-Walker malformation/variant (Greenberg 2010). Initially, it was believed that the hydrocephalus was caused by obstruction of the foramina of Luschka and Magendie. Dandy and Blackfan (1914) believed that the foramina failed to develop or were obstructed due to a prenatal inflammatory process. However, in some cases, the foramina are found to be patent. Also, about 80 % of infants with Dandy-Walker malformation do not have hydrocephalus at birth (Cinalli et al. 2004). The pathophysiology of the hydrocephalus is now felt to be multifactorial. Contributing factors include aqueductal stenosis, basal arachnoiditis from an inflammatory process, abnormally developed subarachnoid space, and venous hypertension from direct pressure from the posterior fossa cyst (Cinalli et al. 2004) (Fig. 2.3).

2.7.5 Vein of Galen Malformation

A vein of Galen malformation is a rare vascular malformation. It is a venous aneurysm of the vein of Galen fed by numerous aberrant branches of the carotid or vertebrobasilar vessels. In addition, arteriovenous malformations may occur within the feeding vessels.

Infants with a vein of Galen malformation often present at birth with congestive heart failure and hydrocephalus. They may also develop hydrocephalus later. Hydrocephalus may be caused by the venous malformation causing obstruction of the cerebral aqueduct. Elevated intracranial venous pressure may also decrease CSF reabsorption and cause hydrocephalus (Fig. 2.4a–b).



Fig. 2.3 A 2-month-old female with Dandy-Walker malformation. MRI shows a posterior fossa cyst of the fourth ventricle and subsequent development of severe hydrocephalus

Cerebrovascular diseases are discussed in detail in Chap. 9.

2.7.6 Arachnoid Cysts

An arachnoid cyst is a benign congenital cyst occurring within the brain. The cyst forms during fetal development with the splitting of the arachnoid membrane (Raffel and McComb 1994), creating an intra-arachnoid space and the resultant cyst. Most of these cysts do not change or cause any other problems. Such cysts are often found incidentally when a child has a scan for some other reason (i.e., head injury). If the cyst enlarges, it may compress the surrounding structures and cause symptoms from mass effect. Depending on the location, as the cyst expands, it may compress nearby CSF pathways and cause hydrocephalus. A suprasellar cyst may expand upward pressing on the floor of the third ventricle and obstruct the foramen of Monro or aqueduct of Sylvius. A cyst in the quadrigeminal cistern or supracollicular

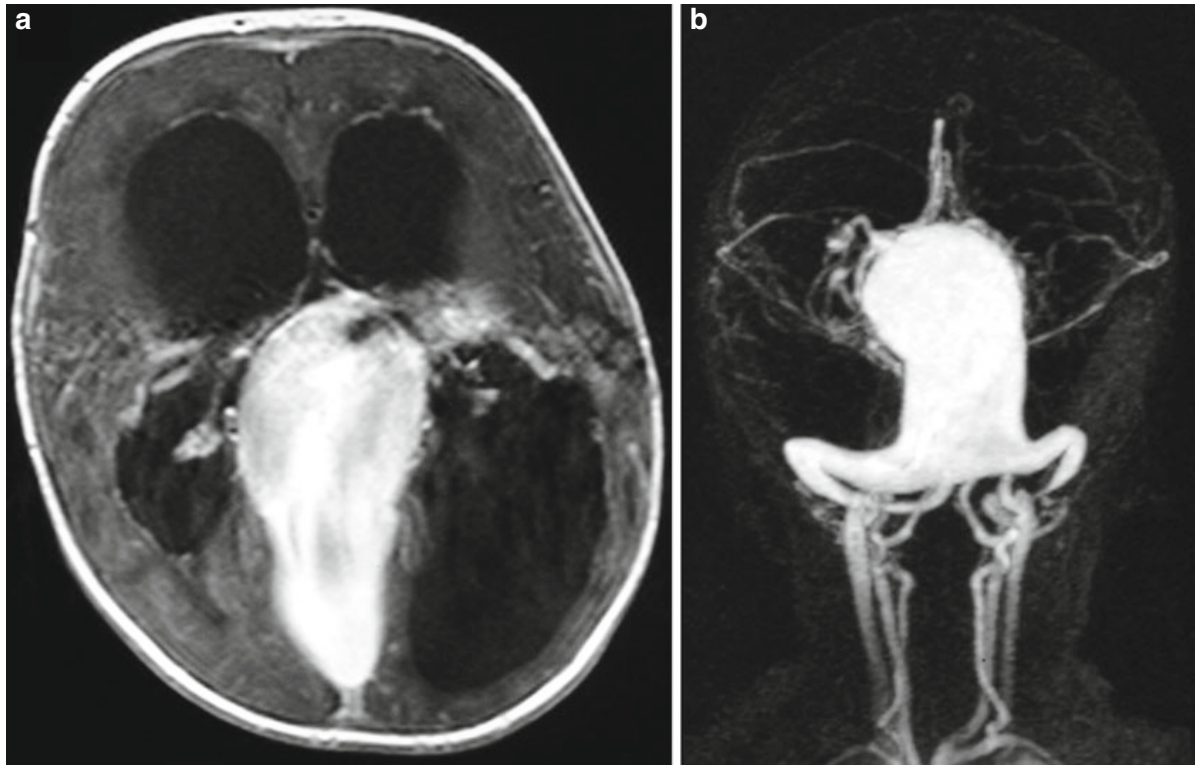


Fig. 2.4 A 4-month-old male with vein of Galen malformation: (a) MRI shows the dilated vein of Galen; (b) cerebral angiogram shows the dilated vein of Galen and the surrounding vasculature

region may cause obstruction of the aqueduct of Sylvius. A posterior fossa arachnoid cyst can cause obstruction at the level of the fourth ventricle. A posterior fossa cyst can be differentiated from a Dandy-Walker malformation by the presence of the cerebellar vermis and a normal appearing fourth ventricle on an MRI. The etiology of expansion of the cyst is unknown.

Surgical intervention is required if hydrocephalus occurs or there are symptoms of mass effect from the cyst. What appears to be an arachnoid cyst may also be associated with a brain tumor. Therefore patients with a newly found cyst require a full MRI of the brain, possibly with contrast, to rule out a tumor. An arachnoid cyst identified before the age of two typically presents with macrocephaly. These cysts are thought to be affected by CSF dynamics, requiring a greater rate of treatment. In Zada's (2007) study, after fenestration, 57 % of patients still required shunt placement (Zada et al. 2007) (Fig. 2.5).

2.7.7 Posthemorrhagic Hydrocephalus of Prematurity

The most common cause of hydrocephalus in the premature infant is a germinal matrix hemorrhage. The germinal matrix is a very vascular



Fig. 2.5 MRI shows a large posterior fossa cyst which effaces the aqueduct of Sylvius and fourth ventricle causing severe hydrocephalus

area in the fetal brain, in the subependymal region located at the level of the foramen of Monro. It is from the very thin-walled germinal matrix vessels that the bleeding is thought to occur in preterm infants. Bleeding can spread, most often to the adjacent ventricles and into the surrounding parenchyma. The germinal matrix gradually involutes after 34 weeks' gestation and nearly disappears by 40 weeks. A grading system has been devised to describe the severity of the bleeding—grades I–IV (Wang and Avellino 2005) (Table 2.2).

Premature infants less than 34 weeks' gestation with very low birth weight (<1,500 g) are at greatest risk for developing intraventricular hemorrhage (IVH). With current management, 20 % of these preterm infants will develop an IVH. The risk of developing posthemorrhagic hydrocephalus (PHH) is directly related to the extent of the hemorrhage. Hydrocephalus develops in 20–74 % of infants with IVH (Boop 2004). Infants with a Grade I or II bleed do not have hydrocephalus by definition; 55 % of infants with a Grade III hemorrhage and 80 % of those with a Grade IV bleed develop hydrocephalus (Boop 2004). PHH may develop as a result of the accumulation of blood and hemorrhagic debris within the ventricles and subarachnoid spaces (Fig. 2.6). Obstruction of the aqueduct of Sylvius or foramen of Monro may occur. The breakdown of blood may also render the arachnoid villi unable to reabsorb the CSF. Multiloculated hydrocephalus may occur after IVH due to ventriculitis. Ventricular septations may develop causing isolated compartments of fluid within the ventricles.

Many premature infants require surgical intervention to treat the hydrocephalus until it is resolved. About 20–30 % will require permanent

Table 2.2 Grading of intraventricular hemorrhage

Grade	Extent of hemorrhage (IVH)
I	Subependymal germinal matrix hemorrhage
II	IVH without ventriculomegaly
III	IVH with ventriculomegaly
IV	IVH with parenchymal hemorrhage

Wang and Avellino (2005)

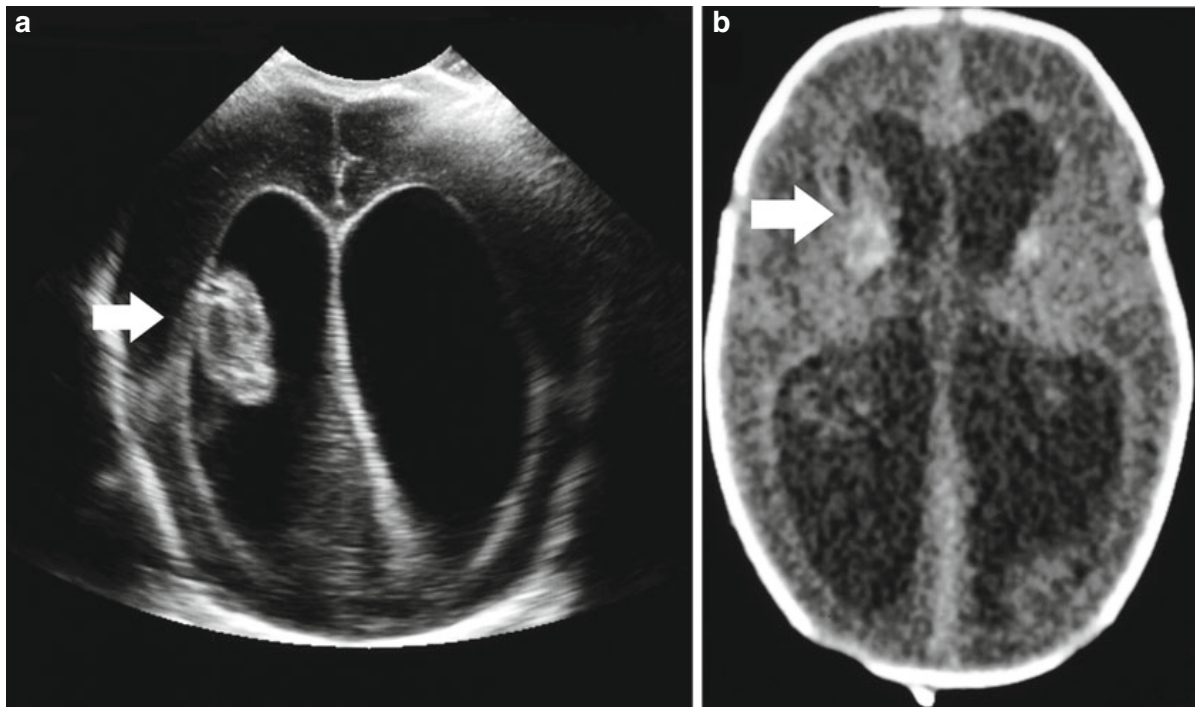


Fig. 2.6 A 25-week premature male with an intraventricular hemorrhage and subsequent development of hydrocephalus: (a) CUS shows the right-sided intraventricular hemorrhage; (b) CT also shows parenchymal hemorrhage

shunting (Boop 2004). Figure 2.6 illustrates IVH and PHH of prematurity.

2.7.8 Postinfectious Hydrocephalus

Intracranial infection at any age may cause hydrocephalus. Hydrocephalus may follow bacterial, fungal, viral, and parasitic infections of the CNS. *In utero*, CNS infections may cause intracranial injury leading to obstruction of CSF flow. Toxoplasmosis may cause inflammation and blockage of the CSF pathways and blockage within the subarachnoid spaces (Ciurea et al. 2004). During the neonatal period, gram-negative bacteria are the leading cause of bacterial meningitis (Ciurea et al. 2004). Gram-negative bacteria may also cause ventriculitis (Ciurea et al. 2004) leading to hydrocephalus.

After the neonatal period, gram-positive bacteria are the leading cause of meningitis. Meningitis and ventriculitis may lead to multiloculated hydrocephalus, a condition where noncommunicating pockets of CSF occur within the ventricles. Viral infections, including CMV, parainfluenza, and

influenza A, can affect ependymal cells leading to acquired aqueductal stenosis and hydrocephalus (Ciurea et al. 2004). Tuberculosis meningitis may cause obstructive hydrocephalus from mass effect of a tuberculoma or cause a communicating type of hydrocephalus by affecting the basal cisterns. Hydrocephalus may develop in conjunction with the intracranial infection or much later after recovery.

Cysticercosis occurs throughout the world. It is rare in the United States but is found throughout Latin America. Humans can acquire the pork tapeworm, *Taenia solium*, by eating undercooked pork or by consuming the tapeworm eggs from food contaminated with human feces. The tapeworm larva enters the body and form cysticerci. Neurocysticercosis results when the cysts enter the brain. The cysts can implant in the parenchyma, ventricles, subarachnoid space, or cisterns. Hydrocephalus can occur when cysts are in the ventricles, subarachnoid space, cisterns, or cause arachnoiditis. In areas such as southern California, Arizona, and New Mexico, neurocysticercosis must be considered as an etiology of hydrocephalus.

2.7.9 CNS Tumors

Hydrocephalus is a complicating factor of pediatric brain tumors. Hydrocephalus can be present at the time of diagnosis of the tumor, may occur during or after tumor treatment, or may develop if the tumor reoccurs. Most of the time, hydrocephalus associated with tumors is due to the obstruction of CSF pathways.

About 60 % of brain tumors in children are located infratentorially or in the posterior fossa, occurring in the cerebellum, fourth ventricle, or brainstem. The most common tumors of this region include medulloblastoma, astrocytoma, and ependymoma. Hydrocephalus is common with tumors in this area. It results from obstruction of CSF flow, particularly if the tumor is in the fourth ventricle or exerting pressure on the fourth ventricle. A tectal plate tumor is an indolent tumor of the midbrain and results in hydrocephalus. In all of these tumors, hydrocephalus is often a major contributor to symptoms at the time of diagnosis. If the hydrocephalus is severe, urgent treatment is needed to relieve increased intracranial pressure. Hydrocephalus may also occur from blood and debris in the CSF after tumor resection. Approximately 25–50 % of children will require placement of a permanent shunt (Wang and Avellino 2005) or endoscopic third ventriculostomy after the tumor resection. Certain factors are associated with the need for permanent CSF diversion including age less than 10 years, midline tumors, incomplete tumor resection, CSF infection, and persistent pseudomeningocele (Sainte 2004).

About 40 % of pediatric brain tumors occur in the supratentorial area. The most common site is the suprasellar region, followed by the cerebral hemispheres, thalamus and basal ganglia, pineal region, intraventricular spaces, and meninges. Hydrocephalus is associated with some of these tumors and is usually due to obstruction of CSF flow at the aqueduct of Sylvius. Tumors in the suprasellar region most commonly associated with hydrocephalus are craniopharyngioma and optic pathway glioma. Craniopharyngiomas can also form cysts that exert mass effect that causes symptoms and/or hydrocephalus. Pineal region

tumors are commonly associated with hydrocephalus. Tumors that grow within the ventricles may cause hydrocephalus as a result of overproduction of CSF. There are two types of choroid plexus tumors: choroid plexus papilloma and choroid plexus carcinoma. They arise from the choroid plexus, located within the lateral, third, and fourth ventricles. Occasionally, germ cell tumors and pituitary adenomas may cause hydrocephalus. Hydrocephalus may also occur in patients with neurofibromatosis or tuberous sclerosis secondary to obstruction of CSF flow.

Spinal cord tumors are rare in children. They may be associated with hydrocephalus due to arachnoiditis and elevated protein in the CSF (Fig. 2.7).

Brain and spinal cord tumors are discussed in detail in Chap. 6.

2.7.10 Head Trauma

Hydrocephalus may occur after head injury if there is intracranial blood. This is particularly true if there is subarachnoid hemorrhage or IVH. The breakdown of blood may alter the ability of the arachnoid villi to absorb CSF. Debris and blood may also obstruct normal CSF pathways and cause obstructive hydrocephalus.

2.8 Signs and Symptoms of Hydrocephalus

The signs and symptoms of hydrocephalus in infants and children vary depending on their age, the degree of hydrocephalus at presentation, the primary etiology, and the time over which the hydrocephalus develops. Because of the plasticity of the infant brain and the ability of the cranium to expand, ventriculomegaly can progress without obvious signs of increased intracranial pressure. In premature infants, in which hydrocephalus is caused predominately by IVH, there is a general correlation between the severity of hemorrhage and the degree of hydrocephalus (Table 2.3). Infants with PHH may have minimal symptoms or may exhibit

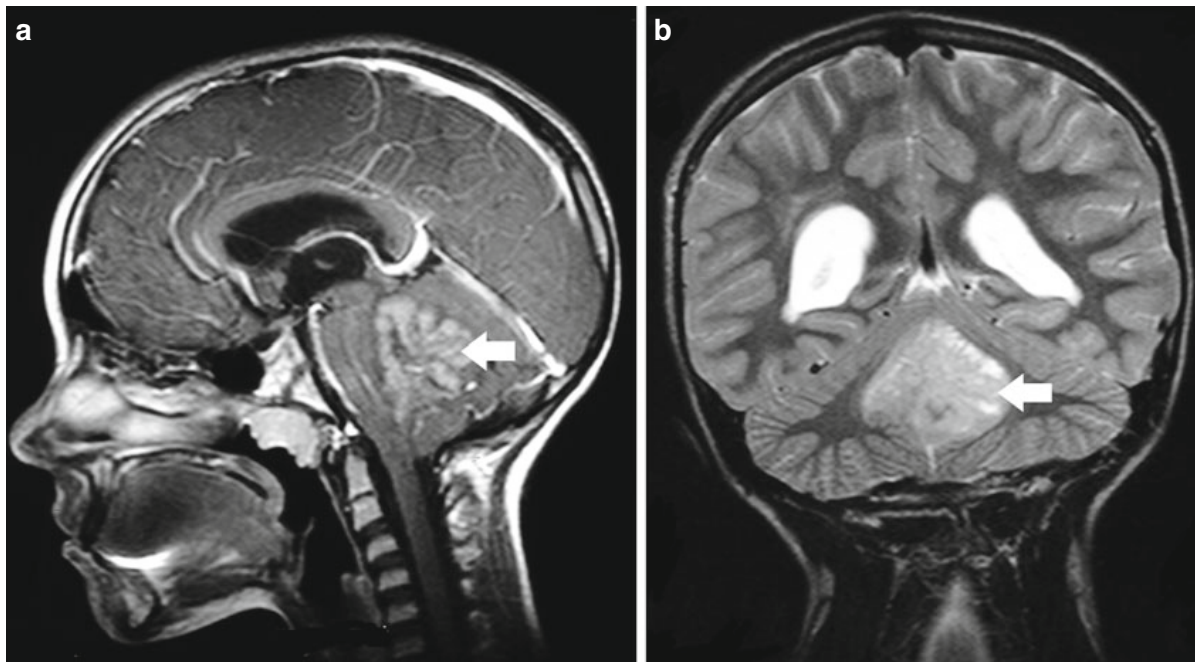


Fig. 2.7 (a–b) A 8-year-old female with a posterior fossa brain tumor and hydrocephalus

Table 2.3 Signs and symptoms of hydrocephalus in children

Premature infants	Full-term infants	Toddlers and older
Apnea	Macrocephaly	Headache
Bradycardia	Rapid head growth	Nausea
Hypotonia	Decreased feeding	Vomiting
Acidosis	Increased drowsiness	Irritability
Seizures	Tense fontanelle	Lethargy
Rapid head growth	Vomiting	Delayed development
Tense fontanelle	Distended scalp veins	Decreased school performance
Splayed cranial sutures	Splayed cranial sutures	Behavioral disturbance
Vomiting	Poor head control	Papilledema
Sunsetting eyes	Parinaud's sign	Parinaud's sign
	Sunsetting eyes	Sunsetting eyes
	Frontal bossing	Bradycardia
		Hypertension
		Irregular breathing patterns

increasing spells of apnea and bradycardia. They may also have hypotonia, sunsetting eyes, ophthalmoplegia, and seizures. As the ventriculomegaly progresses, the fontanel will bulge, become tense and nonpulsatile, and the cranial sutures become splayed. In a healthy premature infant, the head circumference generally increases about 1 cm a week. In premature infants with progressive ventriculomegaly, the head circumference may increase more rapidly

than normal (when charted on the head growth chart) but may not accurately reflect the rate of increase in ventricular size.

In full-term infants, signs often include macrocephaly and progressively increasing occipital frontal head circumference, crossing percentile curves. Normal head circumference for a full-term infant is 33–36 cm at birth. A normal head circumference increases by approximately 2 cm/month during the first 3 months, by 1.5 cm/month

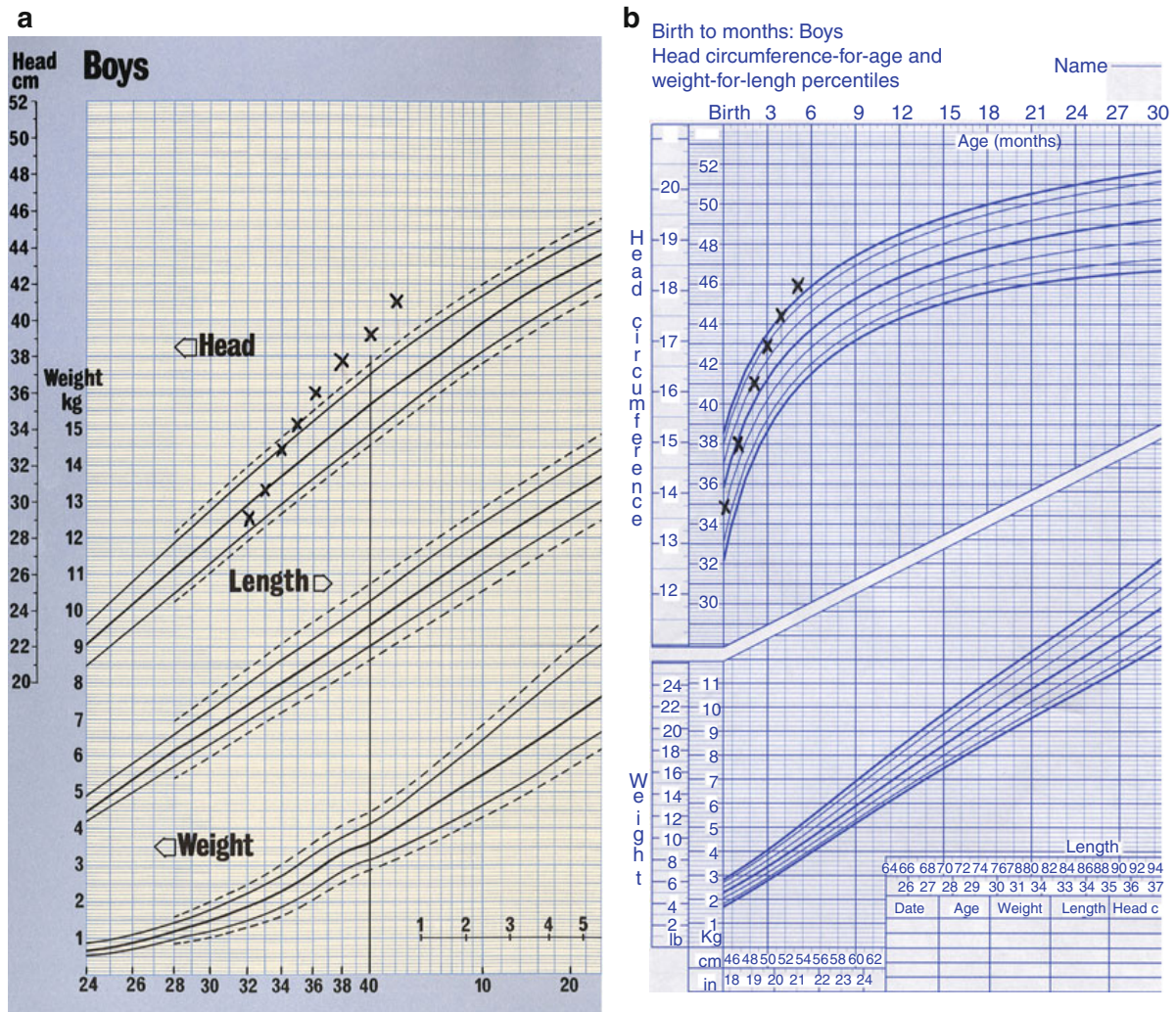


Fig. 2.8 Growth charts show the head circumference rapidly crossing percentile curves: (a) premature male infant growth chart; (b) full-term male infant growth chart

during the fourth and fifth months, and by about 0.5 cm/month from months 6–12 (Fig. 2.8).

Other common signs in full-term infants include a bulging, tense anterior fontanel; splayed cranial sutures; irritability; poor feeding; episodes of spitting up or vomiting; increased sleeping; distended scalp veins; and if the head is large relative to size, poor head control. Visual changes may also be noted and include paralysis of upward gaze (Parinaud’s sign) and sunsetting eyes.

Children older than 2 or 3 years may have a more acute presentation of symptoms since the cranial fontanels and sutures are closed, and the skull is no longer able to compensate for the increasing ventricular size. The predominant

symptom is usually a persistent headache that typically occurs upon wakening and is often associated with nausea, vomiting, and lethargy. The child is often irritable. A child who has a gradual onset of hydrocephalus may have more subtle signs, such as delayed development in both motor and cognitive function. Older children often present with decreased school performance and behavioral disturbance. Other less common signs may include papilledema and visual complaints. If hydrocephalus is severe, Cushing’s triad of bradycardia, systemic hypertension, and irregular breathing patterns may occur. This triad denotes a severe case of increased intracranial pressure and requires emergent treatment.

2.9 Diagnosis of Hydrocephalus by Imaging Studies

The three major techniques used for diagnosis and evaluation of hydrocephalus are ultrasonography (US), CT, and MRI.

2.9.1 Ultrasonography

Prenatal US can be highly reliable and accurate in diagnosing hydrocephalus. Hydrocephalus can be detected in a fetus as early as the later part of the first trimester of pregnancy, although abnormal dilation of the fetus's ventricles are more clearly detectable after 20–24 weeks' gestation (University of California, San Francisco 2000). Although prenatal US can detect an abnormal CSF collection, it may not show the precise site or cause of obstruction. Amniocentesis can often detect the presence of open neural tube defects, such as myelomeningocele, chromosome abnormalities, and *in utero* infections, and may also help indicate the overall health of the fetus. In general, the first trimester development of significant hydrocephalus can be a poor prognostic sign for infant mortality and developmental progress. In some cases, mild ventricular dilation identified by US will resolve by the third trimester (University of California, San Francisco 2000).

Cranial US (CUS) is useful in infants and young children while the anterior fontanel is still open, usually under the age of 18 months (Fig. 2.9). Through the open fontanel, CUS can demonstrate lateral ventricular morphology and intraventricular clots. It is less accurate in its ability to look at the third and fourth ventricles and subarachnoid spaces. For this reason, the precise diagnosis and cause of hydrocephalus is rarely made by CUS alone. It is particularly useful, however, for follow-up screening of infants with untreated and treated hydrocephalus. The equipment is portable, involves no radiation, does not require sedation, and is considerably less expensive than CT/MRI.

2.9.2 Computed Tomography (CT)

Since the advent of CT scanning in 1976, it is a common radiologic technique for the diagnosis and follow-up of hydrocephalus. CT images can accurately demonstrate the ventricular size and shape, the presence of blood and calcifications, cysts and shunt hardware. With hydrocephalus, an enlarged ventricular system is usually seen and is typically first seen in the lateral ventricles (Fig. 2.10). CT images can also accurately reflect signs of increased intracranial pressure, such as compressed cerebral sulci, absent subarachnoid spaces over the convexity, and transependymal

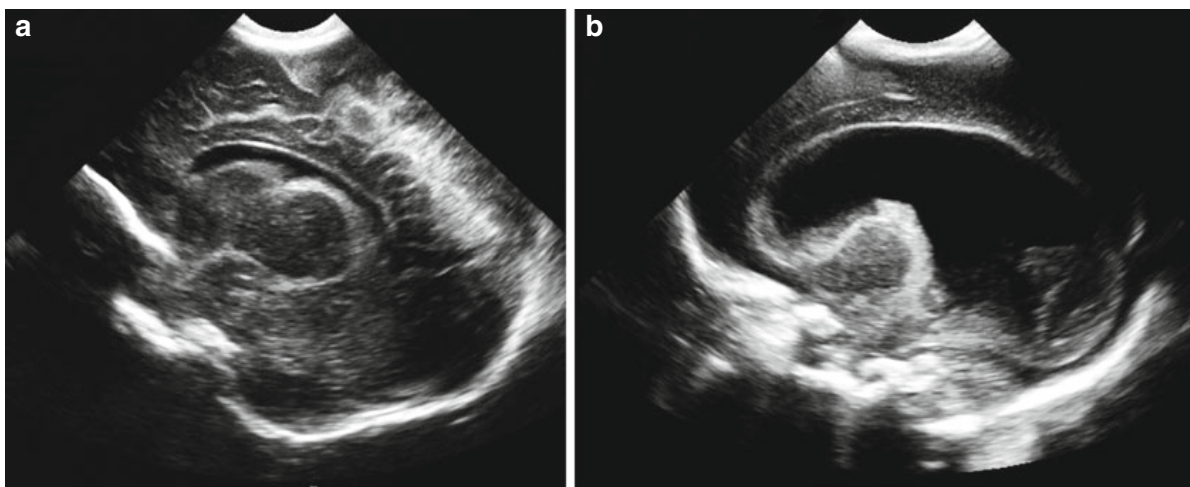


Fig. 2.9 (a) Normal CUS in a 1-month-old female; (b) hydrocephalus in a 3-week-old male

reabsorption of CSF into the white matter. When contrast enhancement is used, tumors, abscesses, and some vascular malformations can be visualized. It is a diagnostic screening tool, taking only a few minutes, and few children need to be sedated for the procedure. CT does expose the patient to radiation, and little is known about the long-term effects of multiple scans. Many providers have concern about the effects of repeated CT scan on the growing brain. Alternatives include low-dose CT or HASTE (limited T2) MR scans. These are acceptable methods for following ventricular size. The Alliance for Radiation Safety has created the “Image Gently” campaign (www.imagegently.com) to assist providers in obtaining the images they need while decreasing the radiation exposure to the patient.

2.9.3 Magnetic Resonance Imaging (MRI)

Commercially available MRI was introduced in 1986 and is the examination of choice for revealing the underlying cause of hydrocephalus. It

allows anatomical visualization in the axial, coronal, and sagittal planes, providing detailed information regarding the anatomy and the position and extent of lesions. Subtle findings, such as white matter pathology, dysmorphic anatomy, and characteristics of lesions, can be readily demonstrated. In addition, the aqueduct of Sylvius can be visualized, as well as membranes and loculated ventricular systems. With the addition of gadolinium (an intravenous contrast medium), some neoplasms, infectious and vascular lesions, can be better visualized. CSF flow dynamics can be visualized through the use of phase-contrast cine MRI. This sequence takes only a few extra seconds and allows for real-time flow measurements that are demonstrated on the sagittal plane of the MRI. Furthermore, constructive interference in the steady state (CISS) sequence MRI may be used. This sequence provides great detail of the ventricular system and basal cisterns and may show membranes not otherwise seen on conventional MRI. Both phase-contrast cine MRI and CISS sequence MRI can be very helpful in determining the underlying cause of hydrocephalus. They can also provide valuable preoperative

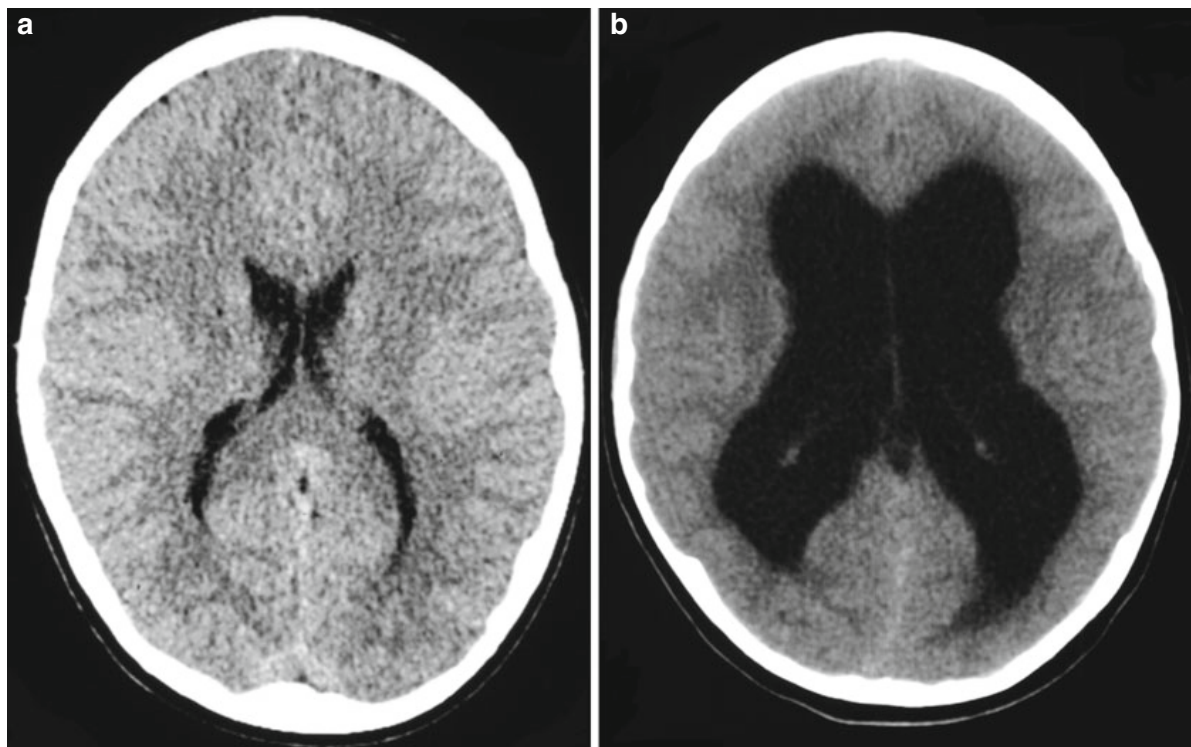


Fig. 2.10 (a) Normal CT in a 9-year-old male; (b) hydrocephalus in a 2-week-old male

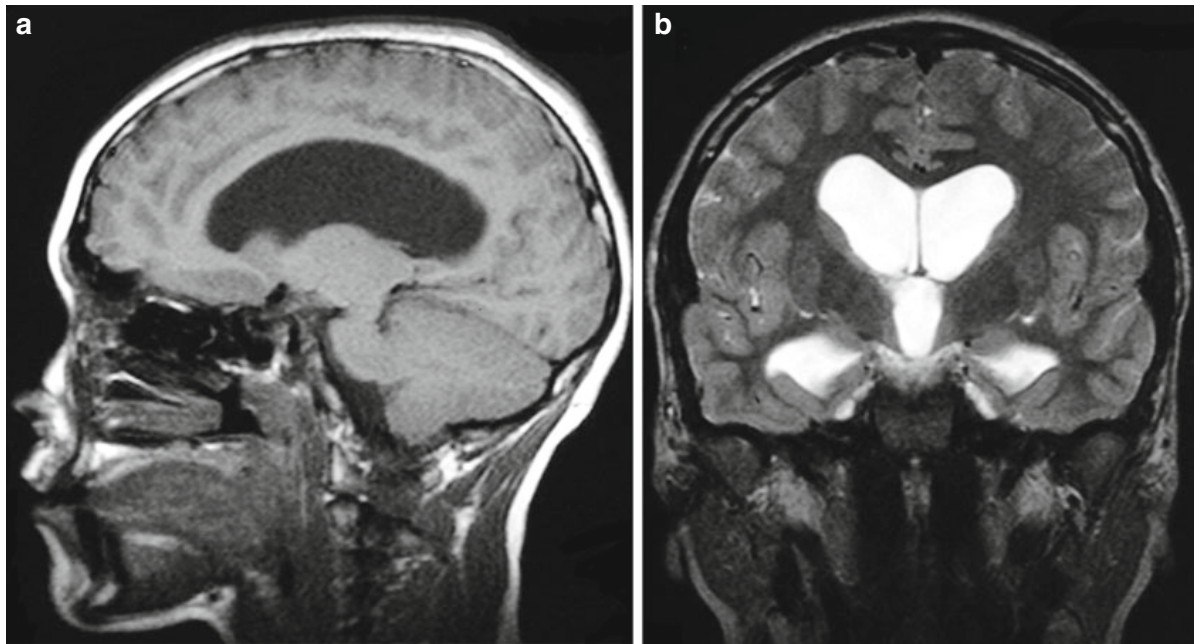


Fig. 2.11 A 13-year-old male with aqueductal stenosis and hydrocephalus: (a) Sagittal T1 MRI shows enlarged lateral ventricles (CSF appears black); (b) coronal T2 MRI shows enlarged lateral and third ventricles (CSF appears white)

information related to the potential success of endoscopic third ventriculostomy, as well as postoperative information by being able to visualize the CSF flow pattern. MRI takes approximately 45 min or longer, and, therefore, young children need to be sedated for the exam. Typically developing children over the age of 5 or 6 can often do the exam without sedation.

The half-Fourier single-shot turbo spin-echo (HASTE) MRI is a limited T2 image that shows ventricular size. It is an alternative to CT, does not expose the child to radiation, and requires no sedation as it is a short study (Penzkofer et al. 2002).

Aqueductal stenosis and hydrocephalus are shown in Figs. 2.11, 2.12, and 2.13, along with cine and CISS MRI scans.

2.10 Treatment of Hydrocephalus

2.10.1 Medical Therapy

There is currently no medical therapy that definitively treats hydrocephalus effectively. Occasionally, in borderline cases of progressive hydrocephalus and in PHH, diuretics may be

useful as a temporizing measure to try to avoid the need for a permanent shunt. Acetazolamide, a carbonic anhydrase inhibitor, has been shown to decrease CSF production. The dose may be as high as 100 mg/kg/day, and in order for it to be effective, more than 99 % of carbonic anhydrase must be blocked before CSF production decreases significantly. Furosemide, 1 mg/kg/day, has also been used. The mechanism of action is unknown, but it is thought to decrease brain extracellular fluid. Although these have been used historically as temporizing measures, comprehensive analysis of data from clinical trials on diuretic therapy for PHH by the Cochrane Collaboration concluded that acetazolamide and furosemide were neither effective nor safe for the treatment of PHH (Whitelaw et al. 2001).

Serial lumbar or ventricular punctures to evacuate CSF are also used as a temporizing measure. The efficacy of these punctures is controversial, but some centers routinely use them in infants until they are stable enough to tolerate a surgical intervention. The goals are to decrease the ICP and help clear the CSF of toxic chemicals produced by the breakdown of blood. If the infant continues to have inadequate CSF reabsorption, a more permanent shunt may be implanted.

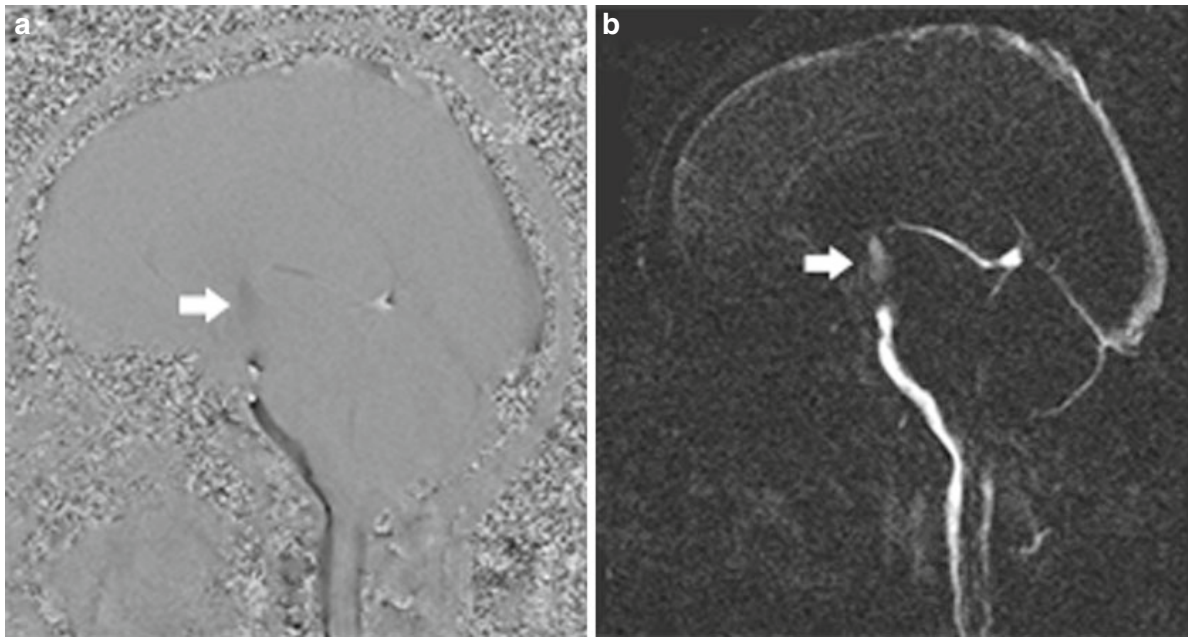


Fig. 2.12 A 13-year-old male with hydrocephalus secondary to aqueductal stenosis, status post endoscopic third ventriculostomy (ETV). CSF cine flow study demonstrates

CSF flow across the fenestration in the anterior third ventricle: (a) phase-contrast magnitude cine MRI; (b) phase-contrast directional cine MRI

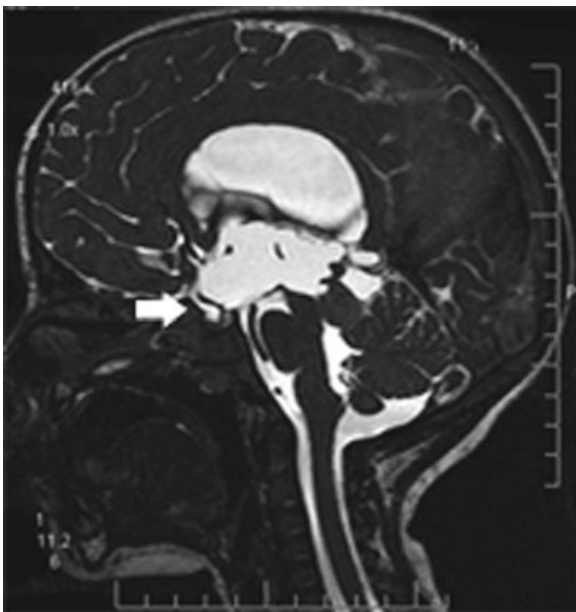


Fig. 2.13 A 13-year-old male with hydrocephalus secondary to aqueductal stenosis. Preoperative CISS (constructive interference in the steady state) MRI demonstrates the floor of the third ventricle and the position of the basilar artery

2.10.2 Surgical Intervention

2.10.2.1 Shunts

CSF shunting is the most common standard treatment in the long-term management of hydrocephalus. It involves the placement of a ventricular catheter to divert CSF to another body cavity, where it can be absorbed. There are many different shunting devices with different components, all having similar features. The three main components of a shunt are a proximal (ventricular) catheter, a valve, and a distal catheter (Fig. 2.14). The ventricular catheter is a silastic tube that is placed either through a frontal or parieto-occipital approach, usually in the right nondominant cerebral hemisphere, as shown in Fig. 2.15. A burr hole is made in the skull, and the catheter tip is generally placed in the frontal horn of the lateral ventricle. This placement is advantageous because there is less choroid plexus in this area and, therefore, less chance for the holes in the catheter to become occluded.

There are many different valves made by many different manufacturers. They all regulate the flow of CSF by means of a one-way valve. The valves most commonly used in the pediatric setting today

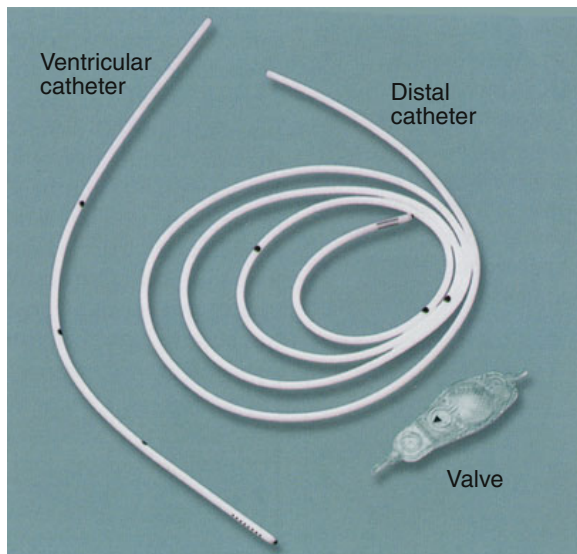


Fig. 2.14 Shunt components (Courtesy of Medtronic Neurologic Technologies)

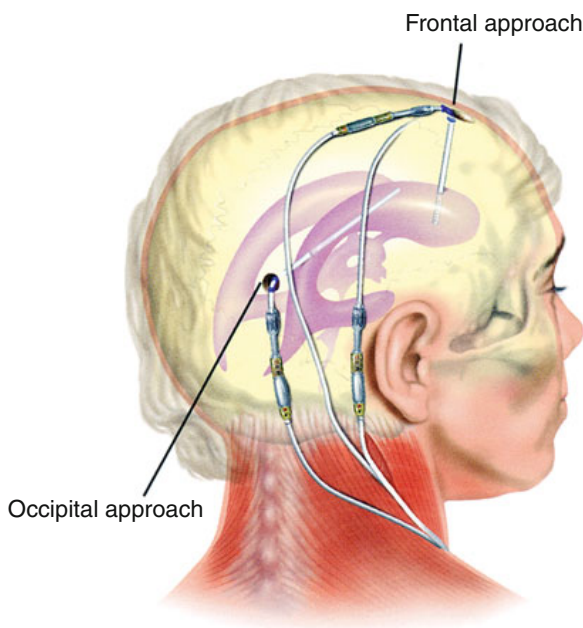


Fig. 2.15 Illustration demonstrates that proximal catheter placement is generally through a frontal or occipital approach

are differential pressure valves, flow-regulating valves, and siphon-resisting valves. The pressure at which the valves open is termed the opening pressure. Typically there are low, medium, and high-pressure valves in each category, referring to opening pressures of approximately 5, 10, and

15 cm H₂O, respectively. Most valves are differential pressure valves and are designed to open and allow drainage of CSF as the intraventricular pressure rises above the valve's opening pressure. Once the pressure falls below the closing pressure, the valve closes and the flow of CSF ceases. Flow-regulating valves attempt to keep the CSF flow constant despite changing pressure differentials and patient position. Siphon-resisting valves are used to avoid siphoning of CSF and the complication of over-drainage. Siphoning is a phenomenon that occurs in some patients in whom there is gravity-enhanced flow of CSF when the patient is in an upright position. The choice of which valve to use is based on the personal preference of the neurosurgeon and is usually based on training and personal experience. No data exists to support a recommendation of one particular shunt design or valve over another. Fixed pressure valves are shown in Fig. 2.16.

A recent advance in shunt valve technology has been the introduction of programmable valves (Figs. 2.17, 2.18, 2.19, and 2.20).

Programmable valves allow the opening pressure of the valve to be adjusted externally with the use of a special magnetic device. This avoids the need for an operative procedure should the patient need a valve with a different pressure. This type of valve tends to be well suited for the management of difficult cases of over-drainage or under-drainage or in children whose pressure needs are expected to change over time. It is not clear that the benefits outweigh the increased cost in all patients. Since the programmable valve contains a magnet, most valves need to be reprogrammed immediately after all MRIs. Several programmable valves that are not altered by a magnetic field are also available. In these valves, the setting is locked and can only be changed with the manufacturer's specific magnetic programmer. Common everyday household equipment like mobile telephones and computers are not strong enough to affect programmable valves, although special precautions should be taken when the patients are around other strong magnetic sources.

Distal catheters are also made of silastic material. The peritoneal cavity is the preferred and

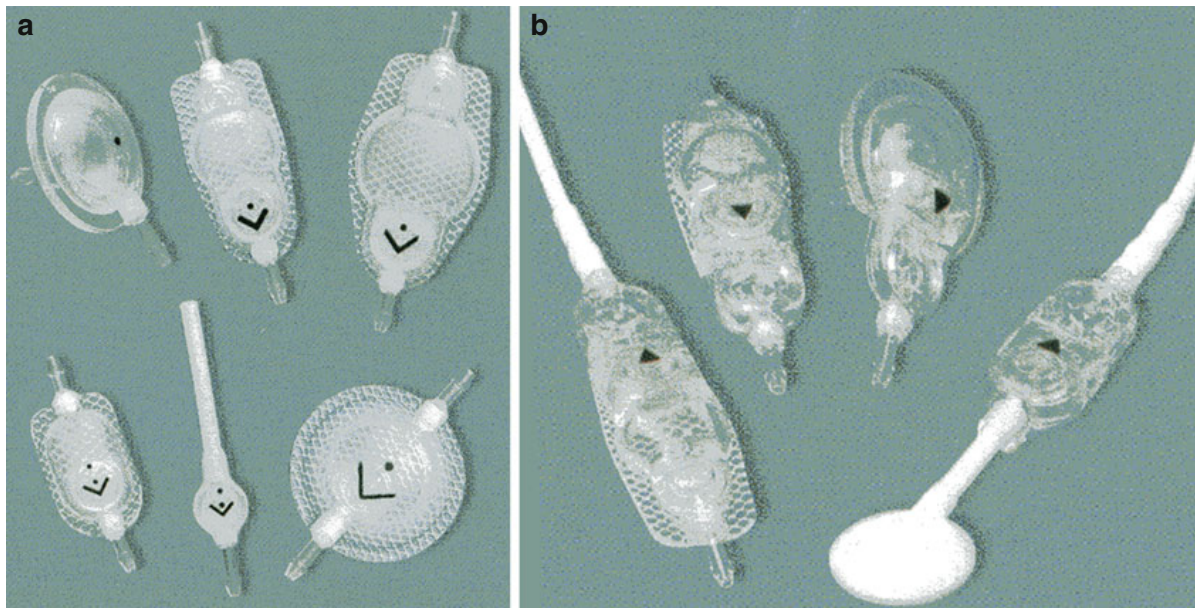


Fig. 2.16 (a) Fixed pressure valves; (b) delta valves with siphon control (Courtesy of Medtronic Neurologic Technologies)

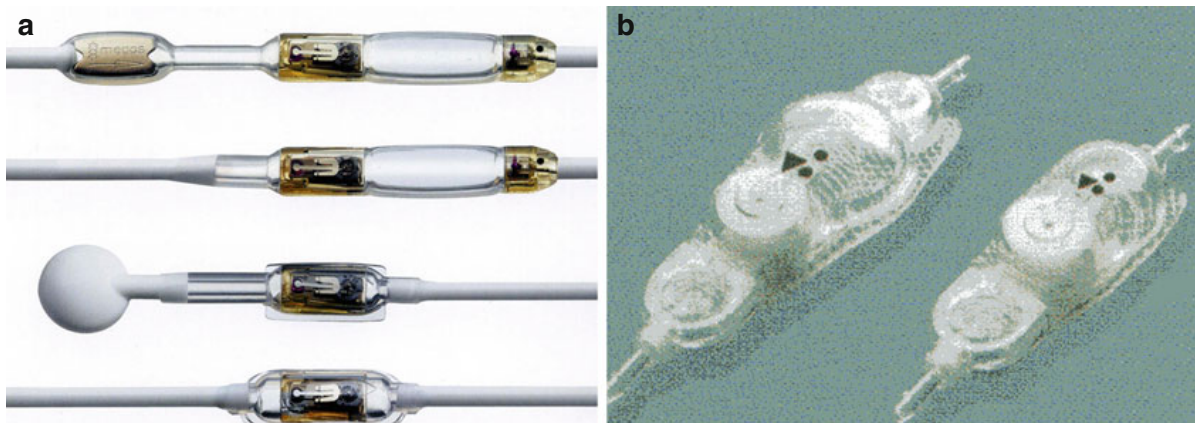


Fig. 2.17 (a) Codman Hakim programmable valves; (b) strata programmable valves (Courtesy of Medtronic Neurologic Technologies)

most commonly utilized location for the shunt to terminate. There are two main advantages to placing the distal tubing in the peritoneum. First, if an infection develops, it usually stays localized rather than disseminating, as can happen with shunts placed in the heart. Second, a large amount of tubing can be placed in the peritoneal cavity to allow for growth of the child and minimize the need for revisions during expected childhood growth. In addition, the peritoneal cavity is an extremely efficient site of absorption and is also easily accessible to the surgeons. If the peritoneal cavity is not appropriate for placement of the

distal tubing, either due to an abdominal malformation, post surgical adhesions, infection, or inadequate reabsorption, the second and third choices for the distal catheter placement are the right atrium of the heart or the pleural cavity.

Ventriculoatrial shunts are placed through the neck, into the jugular vein to the superior vena cava and into the right atrium. The shunt tip should lie just above the tricuspid valve, and on plain chest radiograph should be at the superior vena cava/right atrial interface. The tip of the catheter can also be evaluated by looking for it at the level of the sixth/seventh thoracic vertebrae.



Fig. 2.18 Codman Hakim valve programmer

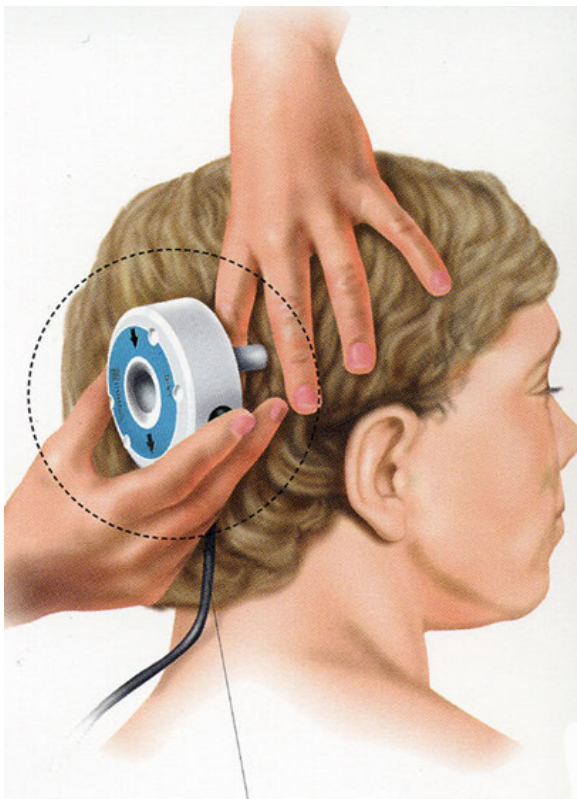


Fig. 2.19 Illustration demonstrates programming a Codman Hakim programmable valve

If it is above this level, a shunt-lengthening procedure may be indicated. Infants should have a chest x-ray every 6 months and older children every year to make sure the distal placement is adequate.

Ventriculopleural shunts are guided subcutaneously to an area just below the nipple, where an incision is made, and the tube is inserted into the pleural space. There is concern that pleural shunting may be poorly tolerated in the young child due to lack of adequate absorptive pleural surface. In addition, the length of time the pleural cavity retains its absorptive capacity varies from individual to individual. Complications may include respiratory compromise secondary to hydrothorax. Other less common distal placements include the gallbladder and ureter.

Less frequently used in the pediatric population, lumboperitoneal shunts are sometimes used in patients with communicating hydrocephalus, slit ventricle syndrome, or benign intracranial hypertension (pseudotumor cerebri). Although classically performed using a limited laminectomy, percutaneous placement using a Tuohy

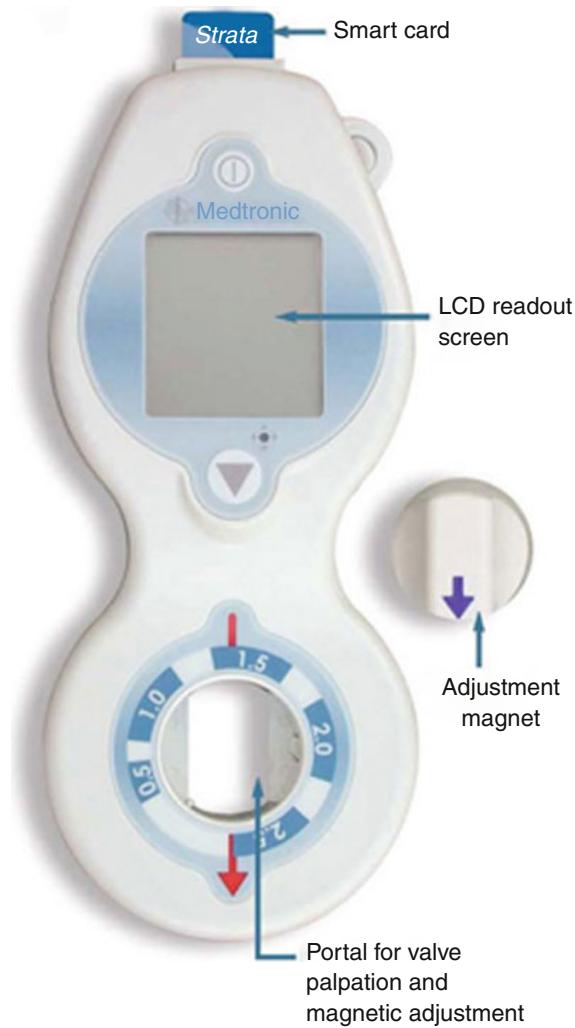


Fig. 2.20 Strata valve programmer (Courtesy of Medtronic Neurologic Technologies)

needle in children over the age of 2 years is now the preferred method of insertion of the catheter into the intradural space (Greenberg 2010) (Figs. 2.21 and 2.22).

2.10.2.2 Surgical Endoscopy

A significant development in pediatric neurosurgery has been the evolution of neuroendoscopy and its application in the management of hydrocephalus. It has been used for endoscopic third ventriculostomy, cyst fenestration and septostomy, and shunt placement and retrieval. The pioneering stage of neuroendoscopy began in the early 1900s but quickly fell out of favor due to poor equipment and a high rate of associated morbidity and mortality. In the 1970s, there was

renewed enthusiasm for its use because of improvement in endoscopes, light sources, camera equipment, and instrumentation. In the past decade, there has been a marked increase in the use of endoscopy.

2.10.2.3 Endoscopic Third Ventriculostomy

Endoscopic third ventriculostomy (ETV) is used as an alternative to shunting in selected patients with noncommunicating hydrocephalus. The success of the procedure depends largely on proper patient selection. The patients most likely to benefit from the procedure are those with significant obstruction of CSF flow between the ventricles and the subarachnoid space and those with normal CSF absorption between the subarachnoid space and the venous system. An MRI should be done as part of the work-up to confirm that the basilar artery does not lie below the floor of the third ventricle. Such placement of the artery is usually a contraindication for a third ventriculostomy due to the added risk of hemorrhage at the time of surgery. In the past, third ventriculostomy has been controversial, and patients under the age of 6 months have not had uniformly good results (Cinalli et al. 1999). Correct patient selection for an ETV has been shown in multiple studies to be vital in the success of the ETV. Many neurosurgeons now consider the factors identified in the ETV Success Score to identify patients based on the criteria of age, etiology, and previous exposure to shunt hardware (Kulkarni et al. 2009; Oertel et al. 2009). Based on these factors, a score is easily calculated and estimates the percentage probability of a successful ETV. Patients with aqueductal stenosis are, in general, excellent candidates for the procedure. It has also been used successfully in patients with posterior fossa tumors.

The goal of ETV is to bypass the obstruction of CSF by diverting it through the floor of the third ventricle and returning it to the normal subarachnoid space. An endoscope is introduced into the lateral ventricle via a coronal burr hole and advanced through the foramen of Monro and into the third ventricle. Once the thin translucent floor of the third ventricle is visualized, a rigid probe is

Fig. 2.21 Illustration shows placement of ventriculoperitoneal (VP) and ventriculoatrial (VA) shunts

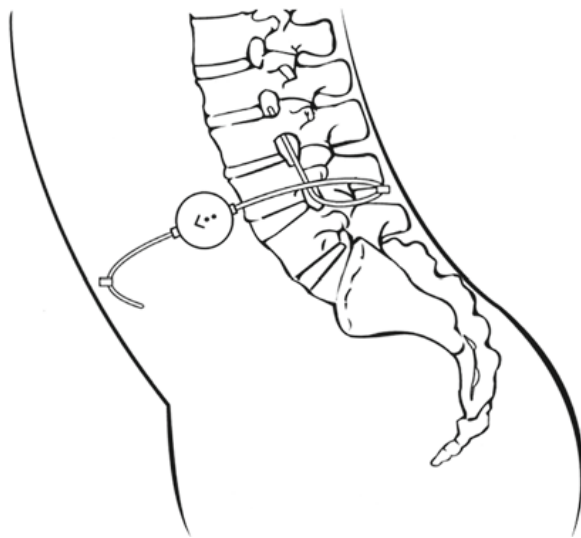
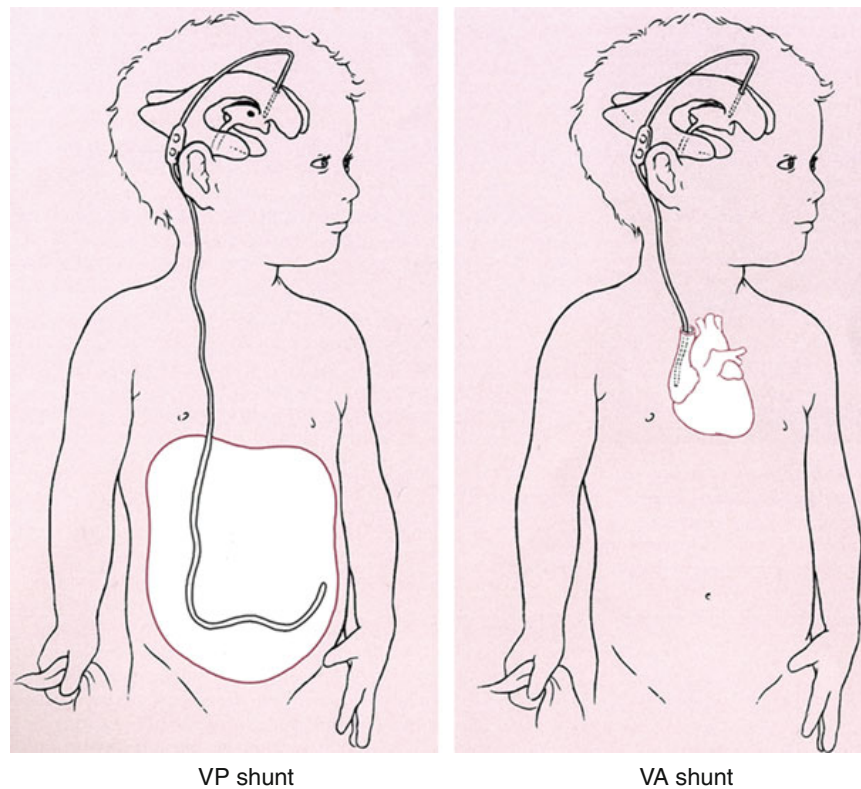


Fig. 2.22 Illustration shows lumboperitoneal shunt (LPS) placement

used to puncture the membrane, and the fenestration is enlarged by balloon catheter dilatation. A laser may also be used to fenestrate the floor of the third ventricle. An external ventricular drain with an intracranial pressure monitor may be placed after the procedure and is usually kept clamped (to drainage). This allows monitoring of

the ICP and possible diversion of CSF should the ETV be unsuccessful.

Patency of the third ventriculostomy can be confirmed noninvasively using phase-contrast cine MRI, or CISS sequence MRI, to identify the CSF flow through the fenestration. However, the finding of a patent fenestration does not guarantee that the procedure has been successful. If there is an obstruction to the circulation of CSF further downstream or inadequate absorption within the subarachnoid space, it is possible for the procedure to fail, even in the setting of a patent fenestration. Ventricular size often does not change after an ETV, even though the pressure is normalized in the ventricles.

ETV has an overall success rate of approximately 75 % after 3 years (Teo and Mobbs 2005). With improved selection, this success rate may be as high as 83 % at 5 years (Kulkarni 2009). Failure of ETV can occur early or late. Early failure may be the result of factors including bleeding around the fenestration site, unnoticed additional arachnoid membranes occluding the flow of CSF, or an inadequate size of the fenestration. Late failure can be caused by subsequent

closure of the fenestration by gliotic tissue or arachnoid membrane. Tumor progression and inadequate CSF absorption at the level of the arachnoid villi may result in either early or late failure.

Patients with an ETV require ongoing neurosurgical follow-up as they can exhibit deterioration after months or years of successful ETV. The problem can be potentially serious because failure can develop over a short period of time and may be unpredictable. The patient develops signs and symptoms of increased intracranial pressure and requires further intervention, either another ETV or a shunt.

Choroid plexus cauterization (CPC) may be coupled with ETV. CPC is a process by which the choroid plexus is cauterized bilaterally within the lateral ventricles through an endoscope. This helps to reduce the rate of CSF production. It has been found that combining the CPC with ETV was more successful than ETV alone for treating hydrocephalus in infants less than 1 year of age (Warf and Campbell 2008).

2.10.3 Treatment of Hydrocephalus in Specific Malformations/Diseases

2.10.3.1 Posthemorrhagic Hydrocephalus

Infants who develop increasing PHH or become symptomatic need temporizing treatment initially. Some centers will treat these small infants with serial lumbar or ventricular punctures; however, infection is a risk. Repeated ventricular taps may lead to porencephaly (cyst or cavity in the cerebral hemisphere).

In infants who have PHH and are able to tolerate a surgical intervention, a ventricular access device (VAD) can be implanted. This is a catheter that is surgically placed into the ventricle with an attached subcutaneous reservoir. The reservoir can be tapped as often as needed through the skin using a 23-gauge butterfly needle. Most infants with a VAD require a tap every 2–3 days, but some may require taps as often as twice daily.

Another procedure that may be used is the placement of a subgaleal shunt. This is similar to a VAD, but the distal limb of the catheter is left to

drain into a subgaleal pocket, which is created at the time of the placement of the device. The CSF under pressure drains through the catheter and distends the scalp, which allows absorption of the CSF by the galea. In some cases, CSF production may exceed the absorptive capability of the subgaleal space, and intermittent taps of the pocket may be required. When repeated taps are needed through either the VAD or the subgaleal shunt, there is a risk of infection. Alternatively, some providers use a temporary external ventricular drainage device in the management of PHH. This has the advantage of maintaining a constant intraventricular pressure, whereas with the other aforementioned treatments, intraventricular pressure can alternate between being very high and very low. The disadvantage of all of these devices is that the catheters can become clogged from the blood or its by-products or, as previously mentioned, infected.

Eventually, over time, it will become clear whether the PHH is resolving or if the infant will need a permanent shunt placed. It has been demonstrated by studies on temporary measures to treat PHH that lumbar punctures and ventricular taps showed no reduction in ongoing need for VP shunt placement. Despite temporizing measures, only 10–35 % of infants will show resolution of their hydrocephalus (Boop 2004). About 20–30 % will require a permanent shunt. In the majority of these infants, the shunt dependency is usually lifelong. There is currently some debate as to the most appropriate timing for a permanent shunt placement. A shunt should be considered when the CSF is cleared of posthemorrhagic debris and blood and the CSF protein is <500 mg/dl (Rekate 1999). The infant should weigh >1.5 kg (variable), have progressive hydrocephalus (Wang and Avellino 2005), and be otherwise stable (Box 2.1).

2.10.3.2 Myelomeningocele

About 80–90 % of children with myelomeningocele will eventually require surgical CSF diversion (Dias 2005). The treatment of hydrocephalus in the infant with a myelomeningocele usually involves placement of a ventriculoperitoneal shunt. The timing of the shunt placement depends

Box 2.1. Case Study: A 27-Week Premie with IVH

Baby P was born at 27 weeks gestation. The patient was born via an emergency C-section due to maternal hypertension and preeclampsia. The patient required CPR at birth and was intubated. An initial cranial ultrasound (CUS) revealed bilateral IVH, grade III on the right and grade IV on the left.

Baby P was initially watched closely by the NICU team with daily OFC and twice weekly CUS. The patient was extubated and started on high-flow nasal cannula. The patient was noted to have a gradual increase in head size. The patient was also noted to have an increase in bradycardic and apneic events. CUS revealed slight increase in ventricles.

On day 24 of life, the patient weighed 1.1 kg. Baby P was taken back to the operating room for a placement of a left frontal subgaleal shunt with a Rickham reservoir, which entailed creation of a subgaleal pocket. The patient initially had a stable subgaleal pocket that was fluid filled and soft. The OFC and US remained stable.

However, over the next several weeks, the subgaleal pocket lessened. By 6–7 weeks of life, Baby P was noted to have a rising OFC and fuller fontanel, with splayed sutures. A CUS, done on week 8 of life, revealed the ventricles to be larger. The patient also demonstrated episodic desaturations. The subgaleal reservoir was tapped using sterile technique. 10 cc/kg of CSF was withdrawn per this hospital's policy. The patient tolerated procedure well and his/her clinical status improved.

Baby P's weight was now 2.2 kg. Therefore, the decision was made to take the patient back to the operating room. The patient underwent placement of a ventricular-peritoneal shunt with a programmable valve. The patient tolerated the procedure well and continued to be followed while inpatient with daily OFCs and weekly CUSs. The patient's shunt valve resistance was increased (resulting in decreased flow of CSF) once due to sunken fontanel and overriding sutures. Baby P did well and was discharged home at 3 months of age.

on the severity of the hydrocephalus but historically has been deferred until after the myelomeningocele repair. Waiting until the hydrocephalus clearly progresses allows for the proper selection of infants who need permanent shunting. Placing a shunt at the time of the back closure may also be more difficult, since the infant should not be positioned on the newly closed myelomeningocele repair site. The advantages to doing it simultaneously with the myelomeningocele repair include a decreased risk of a CSF leak from the repair site and a decreased risk of CSF infection. Some neurosurgeons perform an endoscopic third ventriculostomy instead of a shunt, although performing an ETV in infants is debated.

Placement of an external ventricular drain (EVD) at the time of closure is another option; this allows for the decision of a permanent shunt to be deferred and minimizes the risk of leaking from the back closure.

The *in utero* surgical repair of myelomeningocele was studied in a multicenter study funded by the National Institutes of Health. The purpose of the study was to determine efficacy, safety, and benefit of *in utero* closure of the back (Sutton 2005). Prenatal surgery for the myelomeningocele repair performed before 26 weeks' gestation was found to be associated with decreased risk of death, decreased need for a shunt by 12 months of age, improved scores on mental and motor function, and decreased degree of hindbrain herniation associated with Chiari II malformation (Adzick et al. 2011).

2.10.3.3 Vein of Galen Malformation

Initial treatment for the neonate is supportive and may include immediate resuscitative efforts with ventilatory support. The goal is stabilization until a transvenous and/or transarterial endovascular approach for embolization can reduce blood flow

through the malformation and feeding vessels. The infant often presents with, or develops, hydrocephalus. However, the placement of a cerebrospinal shunt in an infant or child with a vein of Galen malformation has a very high risk of associated intracranial hemorrhage (Schneider et al. 1992). Many neurosurgeons will try to avoid placing a shunt by initially treating the malformation with embolization. If the malformation can be successfully embolized, it may shrink and the hydrocephalus may resolve. Although treatment has greatly improved outcome, mortality and morbidity of these malformations remains high. These patients can have a wide range of cardiac symptoms related to the increase in venous return to the right atrium and chronically increased preload. Patient's cardiac symptoms can range from mild volume overload to congestive heart failure or cardiogenic shock (Hoang et al. 2009).

2.10.3.4 Intracranial Cysts

Many types of intracranial cysts may occur including arachnoid cysts, choroid plexus cysts, neoplastic cysts, and multiloculated cysts associated with infection, tumors, hemorrhage, or trauma. Arachnoid cysts are often diagnosed as an incidental finding when a CT is done for another reason. At the time of the initial diagnosis of an arachnoid cyst, an MRI is often done to rule out a tumor. An intracranial cyst may cause no mass effect or symptoms. Conservative treatment of such a cyst, including follow-up scans to verify that there is no change, may be adequate. The cyst can cause mass effect with symptoms and noncommunicating hydrocephalus from obstruction of normal CSF pathways. In these cases, surgical intervention is required. Rarely, patients present with hemorrhage into the cyst that may also require surgical intervention.

Neuroendoscopic fenestration of the cyst wall may eliminate the need for a shunt. The surgeon breaks the cyst wall with an endoscope, and the fluid in the cyst is allowed to drain into normal CSF passageways. The goal is to reduce the cyst size and avoid placement of a shunt or, if a shunt is necessary, to avoid placing multiple shunts into noncommunicating spaces. Some surgeons may decide to shunt the cyst first because of the high

rate of cyst recurrence after fenestration (Abtin and Walker 1999). If a ventricular shunt is needed, a shunt catheter can be placed into the cyst(s) and another into the ventricle. These catheters can be "y'ed" together into a distal catheter terminating in the peritoneal cavity. The failure rate of multiple shunt catheters is high, and it is difficult to determine which catheters are functional and which are not at the time of malfunction. If the lateral ventricles are loculated (isolated) by membranes or cysts, the surgeon may fenestrate the septum pellucidum (septostomy) to eliminate the need for more than one shunt catheter.

2.10.3.5 Brain Tumors

Approximately two-thirds of children who present with a posterior fossa tumor will have hydrocephalus. A smaller number of children with supratentorial tumors will have associated hydrocephalus at the time of diagnosis. If the hydrocephalus is severe and the child is very symptomatic, an emergent external ventricular drain or shunt may need to be placed. When the surgeon anticipates a resection of the tumor, an external ventricular drain is usually the most appropriate choice because of the risk of shunt failure after tumor surgery due to blood and debris in the CSF from the surgery. If hydrocephalus is present, most neurosurgeons will place an external ventricular drain immediately before a posterior fossa tumor resection. The CSF is allowed to drain for 48–96 h postoperatively, and then the child is gradually weaned from the drainage device over several days. Approximately 25–50 % of these children will be unable to tolerate weaning or removal of the external ventricular drain and will need a permanent shunt (Wang and Avellino 2005) or endoscopic third ventriculostomy (Box 2.2).

2.11 Complications of Shunts and Treatment

Complications of cerebrospinal fluid (CSF) shunts include mechanical failure of the shunt, infection, and over-shunting. Depending on the location of the distal catheter of the CSF shunt, risks of other complications are possible.

Box 2.2. Case Study: An 8-Year-Old Boy with Fourth Ventricular Mass

J is an 8-year-old boy who presented with headaches and nausea/vomiting for 1 month. The patient had CT scan that revealed a fourth ventricular mass, measuring 3×3×2.5 cm with significant ventriculomegaly and transependymal flow. J was immediately flown to a pediatric neurosurgery center. An MRI of the brain and spine was subsequently done on arrival which showed a homogenous and uniformly enhancing fourth ventricular mass with restricted diffusion and severe hydrocephalus. Spine MRI was normal.

On arrival at this center, J was emergently taken to the operating room for placement of a right frontal external ventricular drain (EVD). J tolerated the procedure well and his symptoms improved. The EVD was drained at a level of 15 cm H₂O cm above the external auditory meatus. The patient was started on intravenous antibiotics for prophylactic coverage of the drain and high-dose steroids (dexamethasone).

The following day, J was taken back to the operating room for a suboccipital craniotomy and resection of a posterior fossa tumor. Postoperatively, the EVD was set at a level of 10 cm H₂O. The CSF output initially was bloody and pinkish in color. He had daily sodiums checked which remained stable.

Over the next 3 days, the patient's EVD continued to be monitored. The CSF had less blood products and became straw colored. The patient continued to recover and did remarkably well. Therefore, on day 4 postoperatively, his EVD was raised to 15 cm H₂O. The patient continued to have no headaches or other symptoms, and the following day the drain was raised to 20 cm H₂O. While at 20 cm H₂O, he started to develop headaches and became sleepy. A pseudomeningocele developed at the site of his incision. A limited MRI (HASTE MRI) revealed enlarged ventricles. The patient's EVD was lowered to 15 cm H₂O since he had failed the attempted wean.

The patient was taken to the operating room the following morning for a placement of a right ventricular-peritoneal shunt which he tolerated well. He was able to be discharged home 4 days later after receiving further therapies.

Pathology revealed the tumor was an ependymoma, grade 2. The patient was enrolled in a study involving treatment with chemotherapy and radiation; he continued to be followed by the neurosurgery team.

2.11.1 Shunt Malfunction

Mechanical failure of the shunt can be due to improper placement, obstruction, disconnection, fracture, and migration of the hardware. Malfunctions may occur in the operating room, soon after surgery or years later. However, the most common time for a malfunction is within the first 6 months after placement or revision (McLone 2001).

Obstruction of the shunt hardware comprises 50 % of all CSF complications (Choux et al. 1999). Most often, the obstruction occurs in the proximal portion of the shunt. Total proximal obstruction of the shunt is frequently associated

with rapidly increasing intracranial pressure and requires emergent intervention. The proximal catheter may become obstructed in the operating room or shortly thereafter with blood or air. Proximal occlusion may also be the result of the choroid plexus growing around the proximal portion of the catheter or from blood or other proteinaceous material within the catheter or valve. Occlusion may also be from the catheter being improperly placed during surgery or slipping out of the ventricle later.

If the proximal catheter is obstructed or partially occluded, there may be swelling along the shunt tract over the skull and neck, with minimal signs of shunt failure. Such swelling may also

occur with a functional or partially occluded shunt if there is a large hole in the dura around the shunt catheter, resulting in a CSF leak around the shunt. Obstruction of the distal catheter may be the result of distal infection, scarring, adhesions, or fat occluding the shunt. As the child grows, the distal end of the catheter may slip out of the abdominal cavity. A tract may form allowing CSF to flow beyond the shunt tip, usually failing slowly over time.

Disconnections happen most often at connection sites between components of the shunt. A disconnection may occur between the proximal catheter and valve or between the valve and distal catheter. Tubing that has been in place for a long time may become fixed by the development of scar tissue around the catheter. Over time, calcification may also develop around the catheter and the catheter itself may degrade. Both the fixation and calcification may lead to catheter breakage with growth, particularly in the neck where there is constant motion (Fig. 2.23). The patient may also develop pain along an old calcified shunt tract.

Migration of the distal catheter may occur to a number of sites, including the scrotum, umbilicus, stomach, mouth, intestine, chest, anus, uterus, internal jugular vein, and coronary sinus. When migration occurs, malfunction of shunt often results from blockage of CSF flow and/or lack of reabsorption of CSF (Greenberg 2010). Infection may also occur in conjunction with migration to another site. Other complications may occur depending on where the distal shunt is located.

2.11.1.1 Evaluation and Treatment of Shunt Malfunctions

The initial work-up of the patient with a suspected shunt malfunction includes a thorough history and physical exam. The radiological exam includes shunt series x-rays of the shunt hardware and a CT (without contrast) of the head or limited T2 (HASTE) MRI.

Shunt series x-rays include anterior/posterior and lateral films of the skull, neck, abdomen, and pelvis. These are done to evaluate the continuity of the shunt hardware, location of the hardware,

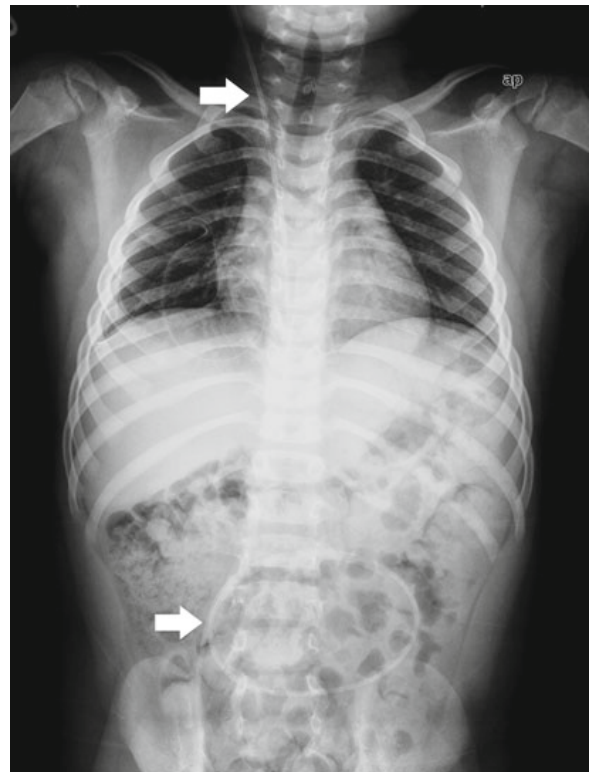


Fig. 2.23 Radiograph shows a broken ventriculoperitoneal shunt catheter near the clavicle. The most distal portion of the catheter can be seen in the *bottom* of the peritoneal cavity

valve type, and any other abnormalities. There may be areas of the shunt system that seem translucent on plain films, particularly over the valve and connectors. Comparing the films to previous postoperative films may provide further information about how the shunt hardware appears at baseline. Comparing the films to x-rays of the most common shunt valves and connectors may also be helpful.

The head CT, MRI, or US can also be compared with previous scans. If the patient is symptomatic and the ventricles have increased in size, a shunt failure is usually confirmed. The scan may also reveal other intracranial complications such as improper placement of the proximal catheter or extra-axial fluid or blood.

Ventriculomegaly does not always occur with shunt malfunctions. In some patients, the ventricles remain small due to decreased ventricular compliance. The child who does not demonstrate increased ventricle size with shunt failure needs further testing to evaluate shunt

function. Additional information about shunt function may be obtained by tapping the shunt and measuring the intracranial pressure. This can be done by accessing the shunt via the reservoir or valve with a 23-gauge butterfly needle connected to a manometer. When the shunt is working, there is usually spontaneous flow of CSF through the catheter into the manometer with good respiratory variation; sometimes pulsation will be seen. The lack of CSF flow from the shunt in the presence of normal or dilated ventricles suggests proximal obstruction. If the ventricles are slit-like, no flow may be normal.

Intracranial pressure is measured manually with the manometer. The pressure is measured with the “0” on the manometer at the level of the external auditory meatus (see Sect. 2.13.5 “External Ventricular Drains”). Pressure will vary depending on age and activity of the child (Box 2.1). To obtain a true pressure, the child must be calm. Intracranial pressure measured with a manometer is measured in cm of water (cmH₂O) (Table 2.4).

A nuclear medicine study (shuntogram) is another test that may be useful in the evaluation of shunt function. A 23-gauge butterfly needle is inserted into the shunt reservoir or valve and the opening pressure is measured. A radioisotope is then injected into the shunt, and gamma camera images are taken of the head, neck, and abdomen to evaluate movement of the tracer. Normal findings of a shuntogram include an opening pressure between 0 and 20 cm of H₂O (dependent on the age and activity of the child), and radioisotope flows out of the needle hub, clears out of the reservoir and shunt, and disperses freely into the peritoneal cavity (the half-time should be less than 5 min). Both a shunt tap and a nuclear medicine study can sometimes provide confusing results.

If a shunt malfunction is confirmed, the child is taken to the operating room for a shunt revision. Sometimes, a very symptomatic patient without clear diagnostic findings during the work-up may be taken to the operating room to explore the shunt. During a shunt revision, all parts of the shunt are evaluated. Shunt hardware parts that are malfunctioning are replaced (Box 2.3).

Table 2.4 Normal intracranial pressure for infants and children

Age	Pressure-cm of H ₂ O	Pressure-mm of Hg
Neonate	<3	<2
Newborn	2–8	1.5–6
Young child	4–9 1.36 cm of H ₂ O = 1 mm of Hg	3–7

Adapted from Wang and Avellino (2005)

2.11.2 Shunt Infection

Infection is the second most common complication of cerebrospinal fluid shunts. The incidence is greatest in the first year after placement, with 80 % appearing in the first 6 months. Rates of infection range from 4.1 to 20.5 % per patient and 2.5 to 12.3 % per procedure (Simon et al. 2009a). Patient characteristics can influence risk. Infants and younger children, those with concurrent infection, those who have had a recent shunt revision, those with previous shunt infection, and those with postoperative disruption of the incision exposing the shunt hardware, have a higher risk of shunt infection.

The most common infecting organism is Staphylococci. *Staphylococcus epidermis* (coagulase-negative staphylococcus) is seen in 50–75 % of all shunt infections (Haines 1999). *Staphylococcus aureus* (coagulase-positive staphylococcus), gram-negative organisms (usually *Escherichia coli*, *Klebsiella*, *Proteus*, and *Pseudomonas*), streptococcal species, *Neisseria*, *Haemophilus influenzae*, and fungi make up the remainder of most infections. Infections with gram-positive organisms correlate with a better outcome than those with gram-negative organisms. The infection usually occurs in one of three ways: (1) via intraoperative contamination, (2) via the bloodstream, or (3) via retrograde travel from a contaminated distal catheter.

2.11.2.1 Evaluation and Treatment of a Cerebrospinal Shunt Infection

If the child has had a recent shunt procedure or a history of shunt infections and presents with symptoms of infection, one should have a high index of suspicion of a shunt infection. If the child

Box 2.3. Case Study: A 6-Year-Old with Shunt Mal Function

CS is a 6-year-old male who has a shunt in place due to a history of congenital hydrocephalus caused by aqueductal stenosis. He was shunted at birth. He underwent an endoscopic third ventriculostomy after shunt failure at the age of 3. He did well and then developed symptoms of hydrocephalus again at the age of 5. An MRI was done and revealed that the ETV had closed. He then underwent another shunt placement.

CS presented to the ED with an 18-h history of headache, vomiting, and 6 h of increasing lethargy. He was seen in the ED by the neurosurgical nurse practitioner. Upon exam, he was arousable but sleepy. He was not sure where he was but was asking for his mother. The mother stated that he is a typically developing child and in the first grade. He was complaining of headache and vomited in the ED. His heart rate was in the 60s, and his other VS were normal for age. He could follow some directions such as moving an extremity, but he was not consistently following directions and at times just cried or whimpered. An urgent low-dose CT was done that showed enlarged ventricular size. A shunt series showed that the shunt system was intact, and his Strata valve was set at 1.0, as it had been previously. When he returned from CT, he seemed slightly more somnolent to the mother. His heart rate was occasionally dipping into the 50s. A shunt tap was done, and there was no flow from the shunt. The pressure was measured and found to be -10 cm of water.

Due to the enlarged ventricles, no flow from the shunt tap and negative pressure in the shunt system, a proximal shunt malfunction was presumed. He was taken urgently to the operating room for a shunt revision. He was found to have occlusion of the proximal catheter, and the intracranial pressure was found to be 30 cm of water. A new catheter was placed and attached to the existing valve and distal catheter. Postoperatively, CS was awake and alert. He denied headache and had complaints of being hungry later in the evening. He also denied any memory of the events in the ED. He stayed in the hospital overnight and was discharged the following morning in good condition.

is stable and has not had a shunt procedure in the last several months, the most common diseases of childhood should be ruled out. A thorough physical exam and laboratory work (including complete blood count, C-reactive protein, erythrocyte sedimentation rate, blood cultures, urinalysis, and chest films) may help locate the source of the infection. Most neurosurgeons are reluctant to tap a shunt unless there is clearly no other source of infection, due to the risk of infecting the shunt with the tap.

The child presenting with a shunt infection may range from minimally to gravely ill. The child may have one or more signs and symptoms of infection: fever, irritability, redness and/or swelling over the shunt tract, or redness and/or drainage from shunt incisions. The infected shunt may or may not fail. Therefore, the child may or may not have signs and symptoms of a

shunt malfunction, including headaches, nausea, vomiting, and lethargy. If the distal portion of the shunt is infected, the child may have abdominal symptoms, including pain, tenderness to palpation, and distension. An abdominal CT or US may reveal an intra-abdominal loculated CSF collection.

Diagnosis of a shunt infection is confirmed by a positive CSF culture from the shunt (or a positive culture from explanted hardware). The shunt reservoir is aspirated via a shunt tap for CSF and sent to the lab for glucose, protein, cell count, Gram stain, and culture. The CSF with infection usually shows mild to marked elevation of WBCs. The Gram stain may show the presence of polymorphonuclear leukocytes and bacteria. The glucose may be decreased, and the protein elevated. Infection in the tissues surrounding the reservoir is usually a contraindication to tapping the shunt.

Even presumed sterile aspiration of the shunt in this setting could lead to contamination and subsequent infection of the shunt.

The treatment of a shunt infection varies, but in general principles of treatment of infection in the setting of a foreign body are followed. Cultures are obtained, and intravenous broad-spectrum antibiotics are started to cover the most likely organisms. The shunt hardware is either externalized or totally removed and replaced with an external ventricular drain. Some neurosurgeons may not remove the shunt if it is functional and treat the patient only with intravenous antibiotics. However, this is somewhat controversial.

Once the infecting organism's sensitivities are known, the antibiotics may be altered. The child is treated with intravenous antibiotics until the CSF has been sterile for several days. There is no consistent agreement about the number of days that the CSF should be sterile before the shunt hardware can be reimplanted. Many neurosurgeons also prefer for the CSF to have less than 50/mm³ white blood cells and the protein to be less than 500 mg/dl before replacing the shunt. Most commonly, the child will receive 5–10 days of treatment before the shunt is replaced. There is also no consistent agreement as to the length of antibiotic treatment after the shunt has been replaced. Factors such as the specific organism, the severity of the infection, and previous history of infections may all affect the length of antibiotic treatment.

Preventing infection is the best way to improve outcome. At present, adherence to meticulous intraoperative aseptic technique and perioperative prophylactic systemic antibiotics are accepted procedures. There is no proven drug of choice or length of treatment after a shunt revision. Most neurosurgeons use intravenous cefazolin, nafcillin, vancomycin, ceftriaxone, or methicillin. Length of treatment varies from one dose to multiple doses (up to 72 h). Antibiotic-impregnated shunt material has been shown to decrease infection rates (Kestle et al. 2011). These catheters may be impregnated with vancomycin, rifampin, clindamycin, or iodine. More studies are currently being done on standardizing protocols to reduce CSF infections. These studies standardize

how the room is set up, number of personnel in the room, double gloving, preparation of skin, antibiotics (Vancomycin and Gentamicin) injected into the shunt intraoperatively, and post-operative care. These protocols have been proven to decrease overall infection rates (Kestle et al. 2011) (Box 2.4).

2.11.3 Complications Related to Distal Catheter Location: Ventriculoperitoneal Shunts

The abdominal cavity is the preferred area to place the distal end of a cerebral spinal fluid shunt in most cases. However, the abdomen may be the site of other surgical procedures or diseases. This is particularly an issue in young children with chronic medical conditions. These children may need frequent urological procedures, gastrostomy tube placement and revisions, or other bowel surgeries. Intra-abdominal adhesions, scarring from old procedures, or previous shunt infections may decrease the absorptive capability of the peritoneum. Pseudocysts may develop around the tip of the catheter, with or without infection. The presence of an intra-abdominal infection, such as appendicitis, may or may not infect the shunt and make the abdominal cavity unsuitable for another shunt. The distal catheter may erode into the bowel leading to shunt infection and peritonitis. Other complications that may occur from intra-abdominal shunts include a 17 % risk of inguinal hernia development (if the shunt is placed in young infants whose process vaginalis is still patent) or the development of a hydrocele.

2.11.3.1 Ventriculoatrial Shunts

If the abdomen is not an appropriate site for the distal shunt catheter, it may be placed in the right atrium of the heart. Ventriculoatrial catheters potentially have more serious complications than ventriculoperitoneal catheters. Complications include migration of the shunt into the superior vena cava (usually with failure of the shunt), pneumothorax, endocarditis, shunt nephritis, pulmonary embolism, septicemia, septic emboli, cardiac arrhythmias, cardiac tamponade, detachment

Box 2.4. Case Study: A 4-Year-Old Child with Shunt Infection

AB is a 4-year-old female who was born with congenital hydrocephalus. A shunt was placed shortly after birth and revised once in the first year of life and again 5 weeks ago at 4 years of age. She did well after the most recent revision and was discharged on the first postoperative day. She returned for a wound check 2 weeks later. The wound appeared to be healing. She had vague complaints of abdominal pain. Her mother also reported some issues with constipation. The neurosurgery nurse practitioner counseled the mother on constipation management.

She returned 1 week later, still complaining of abdominal pain. On exam, the incisions were healing normally. Palpation of her abdomen revealed that it was soft, slightly distended, and mildly tender. She was afebrile. Labs were ordered, and her WBC was normal; CRP was elevated at 5.4. A low-dose head CT and shunt series were done that showed resolution of her increased ventricle size. An abdominal CT was done showing a loculated fluid collection around the tip of the catheter. A shunt tap was done: glucose 28, protein 140, RBC 2 and WBC 190. Gram stain: moderate polymorphonuclear leukocytes and gram-positive cocci. A diagnosis of a shunt infection was made. She was taken to the operating room where all the shunt hardware was removed and an external ventricular drain was placed. She was started on vancomycin and ceftriaxone. The original culture eventually grew out staph epidermis, and sensitivities showed it was sensitive to nafcillin. The antibiotics were changed to nafcillin. CSF cultures were drawn daily and were negative after the shunt was removed. She received a total of 8 days of IV vancomycin/nafcillin. She was then taken back to the OR for placement of a new ventricular to peritoneal shunt. She received the standard postop dose of antibiotics. She was discharged home the following day.

of the catheter with migration into the coronary sinus, and obstruction of the vena cava system. Shunt nephritis is an unusual complication that can cause proteinuria, hematuria, and decreased kidney function and is often caused by a low-grade infection of the shunt. If shunt infection is suspected, blood cultures and 24-h urine for protein are obtained in addition to the other labs to rule out shunt nephritis. If the patient with a ventriculoatrial shunt presents with fever, blood cultures are always performed to rule out systemic bacteremia.

Another problem with ventriculoatrial shunts is that extra tubing cannot be placed into the right atrium to allow for growth. Therefore, infants and young children may outgrow these shunts in months resulting in shunt failure. A ventriculoatrial shunt may require frequent revisions to allow for growth. By examining the chest x-ray, one can diagnose the distal catheter being dislodged from the right atrium. The catheter tip should be visible on plain x-ray at the level of the sixth/seventh thoracic vertebrae.

When examined at autopsy, multiple complications may have occurred around a

ventriculoatrial catheter: fibrinous material may surround the catheter, vegetation may be seen within the wall of the right atrium, and there may be evidence of pulmonary emboli (Marlin and Gaskill 1994).

2.11.4 Lumboperitoneal Catheter Complications

Historically, a number of complications have been associated with lumboperitoneal shunts, including frequent shunt failure, scoliosis, arachnoiditis, back stiffness, back pain, sciatica, neurological changes in the lower limbs, and hindbrain herniation (Olson 2004). These complications have been reduced with changes in shunt hardware and careful preoperative screening. First, with the introduction of percutaneously implanted shunts and improved shunt catheters, the need to perform a laminectomy for shunt placement is now rare. This has reduced the rate of scoliosis and arachnoiditis. Second, preoperative evaluation of patients for hindbrain

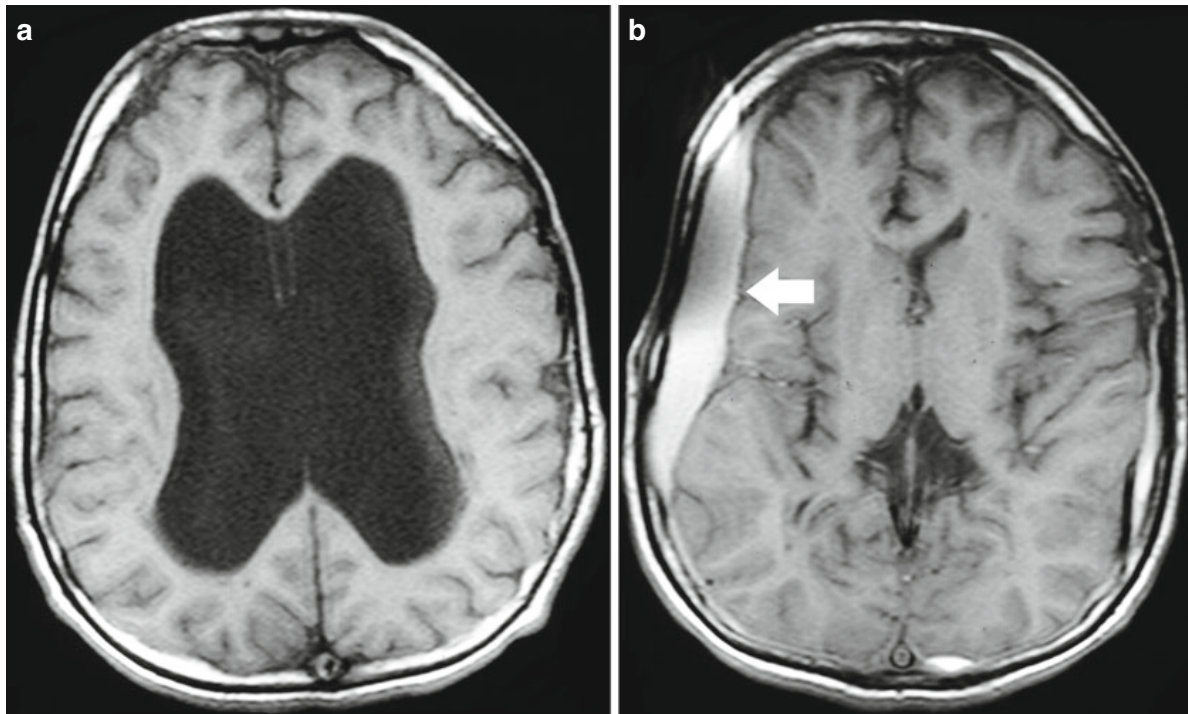


Fig. 2.24 A 15-year-old male with achondroplasia and chronic hydrocephalus: (a) preoperative MRI shows enlarged ventricles; (b) postoperative MRI shows collapsed

ventricles with subsequent development of bilateral (*right* greater than *left*) subdural hematomas

herniation, including a CSF flow study, can help determine when posterior fossa decompression prior to placement of the LP shunt is appropriate. Thorough preoperative evaluation and the use of a higher-pressure valve may decrease the risk and incidence of hindbrain herniation and decrease lumboperitoneal shunt complications (Rekate and Wallace 2003).

2.11.5 Over-Drainage Causing Extra-axial Fluid Collection

After a CSF shunt is placed, if the ventricles decompress excessively or too rapidly, extra-axial fluid collections and/or a subdural hematoma may occur. Fragile bridging veins on the brain's surface may tear as the brain falls away from the dura and bleeding can occur creating a subdural hematoma (Fig. 2.24). This is a risk when shunts are placed for the first time in older children. Treatment depends on severity, symptoms, and the type of valve used. If the valve is programmable, the pressure may be temporarily

increased. If not, the valve may need to be replaced with a higher-pressure valve. By allowing the ventricles to remain more dilated, the brain will resume its normal position against the dura and skull. If a subdural hematoma is present, it may need to be drained.

2.11.6 Special Diagnostic and Treatment Challenge: Slit Ventricle Syndrome

Approximately 75 % of patients with slit ventricles on scan have no symptoms. Slit ventricle syndrome usually occurs after the shunt has been in place for many years, making it more common in the older child and adolescent; however, younger children and infants may also be affected. Slit ventricle syndrome has been used in the literature to describe a number of different situations that include symptomatic small ventricles. This has led to confusion in choosing the most effective treatment option and evaluating the outcome.

In a review of the literature, Olson (2004) found at least 5 different clinical scenarios in which children have radiologically slit ventricles and headaches. Patterns include an on-off (intermittent) symptom complex, over-drainage and siphoning with negative intracranial pressure (particularly when the patient is upright), recurring proximal ventricular catheter dysfunction/obstruction, chronic subdural collections due to shunt over-drainage, and headaches unrelated to shunt function. Most authors applied the syndrome to an on-off (intermittent) symptom complex. This has further been defined as “severe intermittent headaches lasting 10–90 min associated with smaller than normal ventricles on imaging studies, and a slow refill on valve pumping devices” (Rekate 1999). The pathophysiology supporting these symptoms is that with slit ventricles, the catheter becomes intermittently obstructed with surrounding tissue, the pressure rises, and when it is high enough, the ventricles minimally dilate, allowing the catheter to function again. To avoid confusion, using the term noncompliant ventricle syndrome instead of slit ventricle syndrome has been recommended. Of children with radiologically slit ventricles and headaches, 6–22 % have noncompliant ventricle syndrome (Olson 2004).

The exact mechanism underlying the syndrome is not known and may be a combination of proposed theories. First, because there is a relationship between ventricular pressure and intracranial pressure, when CSF pressure drops, there is an increase in venous congestion and brain elasticity. Second, an increased pressure with subependymal flow can cause subependymal gliosis and periventricular gliosis with increased ventricular wall stiffness. Consequently, a higher than normal intracranial pressure is needed to dilate the ventricles. Third, in newborns, over-shunting leads to radiologically slit ventricles, as well as the development of microcephaly and suture synostosis. Because of the small ventricles, catheters become easily plugged. With a small cranial compartment, ventricular dilatation is restricted and can lead to increased intracranial pressure.

Symptoms are those associated with shunt malfunction (intermittent headaches, nausea,

vomiting, and other signs of increased intracranial pressure). Headaches are the most common complaint. In some patients, being upright worsens the symptoms, and lying down improves them.

Evaluation initially involves the typical work-up for shunt malfunction. When the CT is normal, but significant symptoms persist, further studies are warranted. A shuntogram may confirm CSF patency and flow but can be misleading due to the intermittent nature of the problem. Continuous invasive intracranial pressure monitoring may correlate symptoms with pressure. This may be done via a fiberoptic intracranial monitoring device or via an external ventricular drainage catheter attached to an intracranial monitoring device.

Some patients may benefit from antimigraine therapy using propranolol, dihydroergotamine, or amitriptyline. The mechanism by which these drugs work is probably by reducing venous congestion. Propranolol, a beta blocker, cannot be used in children with asthma, as it will render asthma medications (beta-adrenergic agonist bronchodilators such as albuterol) ineffective.

A revision of the shunt is the most common treatment for noncompliant ventricle syndrome. If headaches occur with low pressure, an anti-siphon device may be added, and the valve may be changed to a higher-pressure or programmable valve. These shunt changes may decrease over-drainage and promote slightly larger ventricles, allowing for more consistent shunt function. Multiple changes in ventricular valve pressure are often needed during the evaluation and treatment of noncompliant syndrome. Programmable valves have made this possible without repeated surgical procedures. Before any such changes in an infant, it is important to rule out craniosynostosis, as increasing the valve pressure in this situation could cause a pathological increase in intracranial pressure. Shunt replacement in the setting of small ventricles can be difficult and may require the use of endoscopy.

Success has recently been reported with the placement of a lumboperitoneal shunt in addition

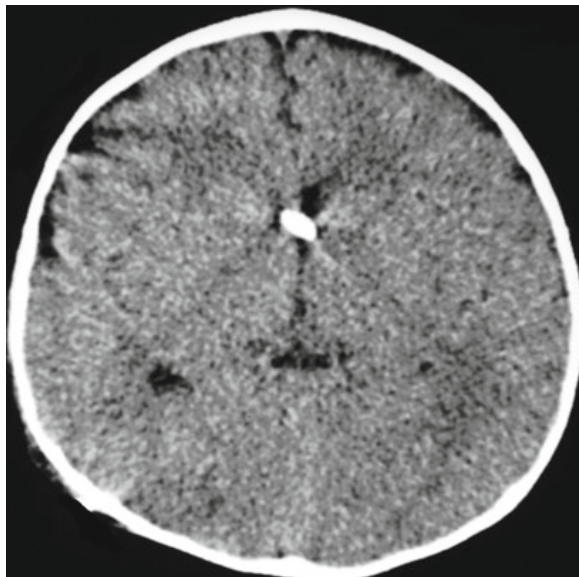


Fig. 2.25 A 11-month-old male with a ventriculoperitoneal shunt and slit ventricle syndrome. He had many months of irritability and trouble feeding and had several shunt revisions, including the placement of programmable valves and the placement of a lumboperitoneal shunt. He eventually underwent a cranial expansion procedure, and his symptoms improved

to a ventriculoperitoneal shunt (Olson 2004). This may occur due to an added increase in CSF drainage when an increase in intracranial pressure occurs. A potential risk factor with a lumbar shunt in addition to a VP shunt is that if the ventricular catheter fails, hindbrain herniation may occur (acquired Chiari I).

Other surgical procedures have been utilized, including subtemporal decompression, calvarial expansion, and third ventriculostomy. Subtemporal decompression reduces intracranial pressure by removing a portion of the skull. The procedure can be accomplished with low risks, and some recommend this as a first-line treatment for noncompliant ventricle syndrome in patients with synostosis and small calvarium. Calvarial expansion is a much more extensive procedure, and bleeding is a significant risk (Fig. 2.25).

2.12 Pseudotumor Cerebri in the Pediatric Population

Another type of CSF absorption problem is a condition known as pseudotumor cerebri (PTC).

It is described as elevated intracranial pressure without hydrocephalus, mass lesion, infection, or hypertensive encephalopathy (Cinalli et al. 1999). PTC is the result of CSF malabsorption or obstruction in the intracranial venous system. Sometimes an exact cause can be found, such as thrombosis, which may be the source of the obstruction. Many times, however, a cause is not found. Thus, PTC is usually a diagnosis of exclusion (Brack et al. 1994).

In the general population, PTC is found in 0.9 per 100,000, but the incidence increases to 90 per 100,000 in obese adult females. There is a female to male predominance of 8:1–2 in adults, but there is no gender predominance in children. Obesity is less frequently a cause of PTC in children. There is a peak incidence in the third decade (range 1–55 years). About 37 % of PTC cases are in children. Ninety percent of children are diagnosed between 5 and 15 years of age, and PTC is rarely seen in infants (Boop 2004).

There are three types or classifications of PTC: primary, secondary, and atypical pseudotumor. The most common form, primary PTC, is idiopathic, and thus the cause is unknown. Secondary PTC presents as the results of another illness or cause. Secondary PTC may be associated with a known neurological disease, the result of a systemic illness (i.e., clotting disorder), or caused by the ingestion or withdrawal of exogenous agents (i.e., vitaminosis A, antibiotics, and others). Finally, atypical PTC presents without papilledema or may be seen in infants. The most common causes of PTC in children include venous thrombosis, steroid withdrawal, malnutrition, or exogenous substances. Obesity, as in the general population, is beginning to be seen more frequently as a cause of PTC in children.

2.12.1 Pathophysiology

CSF is absorbed into the venous system after traveling passively through the arachnoid villi. A failure of CSF absorption may be caused by a blockage somewhere in these veins, such as by a thrombus. Right heart failure in infants sometimes leads to PTC by causing a retrograde

elevated intracranial pressure, and thus a CSF absorption problem.

Several exogenous agents have been known to cause PTC. These include vitamin A, chemotherapy agents like vincristine, and some antibiotics. Although there are theories as to how these agents cause changes in CSF absorption pathways at the cellular level, the exact cause of PTC from the ingestion of vitamin A or other medicines remains unclear (Abtin and Walker 1999; Cinalli et al. 1999, 2004).

2.12.1.1 Clinical Evaluation

The examiner should obtain a thorough history and perform a complete physical examination, including an age-appropriate neurological examination (please refer to Chap. 1 regarding neurological examination). CT and plain MRI will not aid in diagnosis but are performed to exclude other causes of increased intracranial pressure such as mass lesions (Said and Rosman 2004). Magnetic resonance venograms may show an obstruction or occlusion. A lumbar puncture can demonstrate an elevated opening pressure and is performed with the patient lying in the lateral position. It is important to position patient on their side, as patients placed on their abdomens may have an artificially elevated opening pressure. Retrograde venography is the measurement of the intracranial venous systems via a catheter threaded upward from the femoral vein. Abnormal readings, including elevation of right-sided heart measurements, demonstrate the exact location of an obstruction. This is known as a “gradient.” Ophthalmologic examinations are performed to note any changes in visual fields, papilledema, and other tests of visual acuity. Intracranial pressure monitoring may be done with an intraparenchymal transducer, or similar device. This is helpful to note changes in pressure throughout the day while awake and asleep. Changes in position may also cause elevations in pressures, such as turning the head from side to side, because of venous obstruction on one side. Psychological evaluations may be performed, as there can be complicated comorbid involvement in some patients who receive secondary gain from having headaches that cannot always be diagnosed.

2.12.1.2 Treatment

Sometimes PTC may resolve spontaneously. Serial lumbar punctures to remove CSF have been shown to be beneficial in the resolution of PTC. More permanent CSF diversion may be necessary. This includes the implantation of a lumbar shunt with the valve system being the choice of the neurosurgeon. The lumbar shunt communicates with the CSF outside the ventricular system at the level of the subarachnoid space (28). The implantation of a ventricular access device such as a Rickham Reservoir (Codman, Raynham, MA, USA) allows for rapid and accurate measurement of intracranial pressure manometrically with little discomfort to the child. Because the ventricles are small in this condition, the placement of the reservoir is best performed using stereotactic guidance. Shunts from the ventricles to the peritoneum are difficult to maintain because of the small size of the ventricle, and thus the ventricular shunt cannot provide adequate drainage of CSF from the subarachnoid spaces.

Optic nerve sheath fenestrations are performed to reduce pressure on the optic nerve, as well as drain CSF, because the CSF may communicate with the subarachnoid space.

Medication may be helpful by reducing the production of CSF (e.g., acetazolamide, furosemide). This is usually a temporary measure until either a more permanent solution is found or the PTC resolves. Medications are used with caution, as there are side effects that include electrolyte abnormalities.

Obese patients with PTC and stable visual symptoms are best treated with weight loss to avoid shunt placement or optic nerve sheath fenestration (24). Even a small amount of weight loss can reduce intracranial pressure. Bariatric surgery may be considered for a morbidly obese patient who is in their late teens.

2.12.1.3 Nursing Care

Sometimes, children are placed in the pediatric intensive care unit for several days, while their intracranial pressure is monitored via an intraparenchymal wire. It is usually inserted in the operating room, under anesthesia, to maintain

strict asepsis and to reduce anxiety. The nurse plays an important role in monitoring elevations in pressure, and assisting the family to keep a “headache diary” during their stay. Monitoring of visual changes is also very important.

Patient and family education is needed so that lifestyle changes can be made to prevent loss of vision and the adaptation to the possible shunt systems. There are several Web sites that provide education to families and allow patients to communicate with one another. Patient and family support are provided by nursing, social service, and psychological intervention as needed.

Finally, there is ongoing research for future cures and diagnosis of PTC, including new ways to measure intravenous pressure. The placement of intracranial stenting devices that bypass a venous obstruction has been done in a small number of adults with some success.

2.13 Nursing Care of the Hydrocephalus Patient After Surgery

The most common operations that children with hydrocephalus undergo are shunt placement, shunt revision, and endoscopic third ventriculostomy. Because these children frequently have other diseases related to the hydrocephalus, they often undergo other surgeries to treat a multitude of other problems.

2.13.1 Neurological Assessment

Neurological assessment of the child after surgery to treat the hydrocephalus needs to be done frequently to detect any changes. The surgeon will usually specify the frequency, but assessment should occur every 1–4 h, depending on the condition of the child. An exam that is changing subtly over time may be an indication of failure of the surgical treatment or postoperative complication. The first signs of increasing intracranial pressure are usually subtle and related to mildly increasing somnolence, lack of interest in activities (feeding) or play, and subtle behavioral

changes. Level of consciousness is the most important single indicator of neurological status. Altered level of consciousness may progress to confusion, disorientation, somnolence, lethargy, obtundation, stupor, and coma.

Parents and families are an excellent resource to provide information about their particular child’s developmental level. The signs and symptoms of increasing intracranial pressure may initially be very subtle. Hence, the child’s caretaker is a valuable resource in such assessment and may notice subtle changes before nursing and medical staff.

A thorough neurological assessment starts with watching the child play and interact with those around him. Assessment also includes asking the child if he has a headache. Nonverbal infants and children may exhibit behavioral signs of headache. The child should be examined for his ability to answer questions appropriately and follow directions. Asking a child to move his arms and legs will also allow the examiner to assess muscle strength, tone, and movement. Vital signs should also be assessed. Bradycardia is a sign of increased intracranial pressure and should be closely monitored in the presence of other symptoms. Increased blood pressure is usually not a common finding in children until late in the process of increasing intracranial pressure.

It is important to carefully examine the eyes; checking pupils without further exam is never an adequate exam. The pupils are checked for equality, roundness, and reactivity to light. Dilated and nonreactive pupils are a very late sign of increased intracranial pressure. A “sunsetting” appearance to the eyes or the loss of the upward gaze is an abnormal finding and indicative of increased intracranial pressure. The extraocular movements should be intact.

The infant’s head should be examined. The occipital frontal circumference should be measured and documented on a daily basis to determine appropriate head growth. The fontanelles should be palpated with the child upright and calm. The anterior fontanelle should feel soft and pulsatile. A tense or bulging fontanel is suspicious for increased intracranial pressure. The suture lines of the skull should also be examined.

Normal suture lines are palpable and apposed. If they are overriding, the infant may have over-drainage of the shunt. If the sutures are splayed, there is likely increased intracranial pressure.

2.13.2 Wound and Dressing Care

The child will usually come from the operating room with a dressing over the incision. The dressing is normally removed or changed during the first few postoperative days. If a dressing is soiled or saturated with blood, most surgeons agree that it should be replaced. If the child is likely to pick at the incision, a dressing may be kept over the incision to prevent infection. Before a child goes home, most surgeons agree the dressing should be changed and the wound inspected for any erythema, drainage, swelling, or infection.

2.13.3 Medications

The majority of neurosurgeons will order intravenous postoperative antibiotics to prevent shunt infection. Cefazolin and nafcillin are the most commonly used antibiotics, as gram-positive organisms demonstrate sensitivity to them. Vancomycin may also be used. Length of treatment is variable.

Pain management starts with good pain assessment. Age-appropriate pain assessment scales such as the CRIES (crying, requires increased oxygen administration, increased vital signs, expression, sleeplessness), Objective Pain Scale, and Oucher scale may be used. There is a wide variety of pain experienced by children after surgery for hydrocephalus. Pain may be related to the cranial incision(s), the abdominal incision, the amount of intra-abdominal manipulation, and the tunneling of the distal catheter through the subcutaneous tissue. Other factors influencing pain may include the age of the child, the child and/or family's experience with pain, and the child and family's anxiety. Pain is usually managed with medications although other techniques may be helpful. The first drug of choice is usually acetaminophen. It should be adequately dosed at

10–15 mg/kg/dose, maximum dose of 75 mg/kg/day not to exceed 4,000 mg/day, and can be given orally or rectally. Nonsteroidal anti-inflammatory drugs (NSAIDs) may be used, but they can inhibit platelet aggregation and prolong bleeding time. For this reason, some neurosurgeons do not use NSAIDs during the immediate postoperative period.

If the child needs additional medication for pain, the surgeon's beliefs about pain control in neurosurgical patients may be a factor. Some neurosurgeons will order opiates such as morphine sulfate, oxycodone, or codeine, while others do not want to alter the patient's neurological exam with these drugs. The nurse should not administer these drugs if she is concerned that the pain is due to increasing intracranial pressure or the neurological exam is changing. Other modalities to relieve pain may include age-appropriate relaxation techniques, play therapy, music therapy, massage, distraction, and acupuncture or acupressure.

Some children will experience nausea and vomiting from anesthesia that may be worsened by extensive intra-abdominal manipulation during the surgery. Medications to treat this include metoclopramide and ondansetron. The nurse should not administer these drugs repeatedly if there is a possibility that the nausea and vomiting are due to increasing intracranial pressure. Treating the symptoms and ignoring the underlying cause may result in further increased pressure and delay of needed treatment.

Children who are on antiseizure medications preoperatively should have these resumed as soon as possible. Often, because of vomiting before or after the surgery, doses are missed, so it is helpful to check blood levels of the drugs to ensure they are therapeutic. If the levels are subtherapeutic, extra doses may be ordered. Children with low levels of their antiseizure medications are at increased risk for seizures.

Infants and children require some intravenous fluids after surgery until they can take adequate fluids orally. Fluid loss from vomiting should be replaced. Electrolytes should also be monitored during periods of vomiting. The nurse should assess the child for symptoms of adequate

hydration. Usually the child will receive maintenance fluids postoperatively for at least 12 h. The child with a shunt in place should never have an intravenous line placed into the scalp because of the risk of introducing bacteria to the area around the shunt hardware.

2.13.4 Other Nursing Care

The surgeon will usually specify the position that the child should assume. Elevating the head of the bed 30–45° will enhance shunt function by gravity aiding the flow of the CSF through the shunt. The surgeon may specify that the infant or child be placed flat if he is concerned about over-drainage of the ventricles. If the ventricles are allowed to drain too quickly, the outside of cerebral cortex may pull away from the dura. This may cause tearing of the fragile bridging veins and result in a subdural hematoma. Infants with overriding sutures are usually placed flat to minimize over-drainage. If overriding sutures are allowed to occur for a long period of time, the sutures may fuse prematurely. The nurse should also position the infant or child off of the incision and shunt hardware. Young infants who are allowed to lie on the hardware may experience skin breakdown and shunt infection within hours. X-rays of the shunt system are also done during the postoperative period to assure correct placement of the shunt, that the system is intact, and for any other potential complications (i.e., pneumothorax associated with a ventriculoatrial shunt).

These children have all the other usual postoperative needs of pediatric surgery patients. Nurses should be concerned with adequate diet, good pulmonary care, mobilization issues, skin care, adequate rest, and emotional care.

2.13.5 Extraventricular Drainage

CSF can be temporally diverted outside the body using an extraventricular drain (ventriculostomy or EVD) and may be used with or without intracranial pressure monitoring. It is commonly used

in the treatment of shunt infections, in which the colonized shunt tubing, as well as the infected CSF, needs to be removed in order to completely eradicate the infection. Usually the entire shunt system is removed, although occasionally just the distal portion of the shunt is externalized. An EVD is also commonly used after posterior fossa tumor resections, to help drain blood and surgical debris out of the ventricular system. Other uses may include the administration of intrathecal antibiotics, emergent diversion of CSF in acute hydrocephalus, and intracranial pressure monitoring after endoscopic third ventriculostomy and in association with head injury.

Several different drainage systems are available for an EVD, but all have similar features (Fig. 2.26). The ventricular catheter is usually placed in the operating room, which allows for maximal aseptic technique at placement and tunneling of the catheter under the skin before it exits the skin. These two factors may decrease the infection rate with external drains. The ventricular catheter is generally inserted into the frontal horn of the lateral ventricle (on the non-dominant side of the brain) and is connected to the CSF collection chamber via a closed sterile setup.

The surgeon should specify the level that the chamber needs to be placed, as well as the level of the head of bed, in the postoperative orders. The chamber is generally placed in reference to the external auditory meatus, which is at the level of the foramen of Monro. Careful attention needs to be made to assure that the system is set up and measured properly, and that the catheter does not become kinked, dislodged, or disconnected. In addition, it is important to clamp the system before the patient changes position (Fig. 2.26).

The details of CSF drainage need to be monitored regularly and the amount, color, and presence of blood or sediment must be recorded on a frequent basis. The normal amount of CSF that is produced daily is approximately 350–700 ml in adults (Brack et al. 1994). Infants and children produce less, about 0.33 ml/kg/h. Excessive or insufficient CSF drainage is a common complication that may occur. Excessive drainage results when the pressure at which the drainage occurs is



Fig. 2.26 External ventricular drain (EVD) (Courtesy of Medtronic Neurologic Technologies)

too low. This may be caused by the movement of the child above the predetermined ordered level or increased intracranial pressure secondary to coughing, crying, sneezing, or the Valsalva maneuver. Excessive draining may cause the ventricles to rapidly collapse, leading to a subdural or subarachnoid hemorrhage.

Insufficient CSF drainage may cause increased intracranial pressure, with associated symptoms. Inadequate drainage may be caused by the child moving to a position that is lower than the ordered level, by kinks in the catheter, or by occlusion of the catheter from blood or cellular debris. Occlusion should be suspected if there is no fluctuation of CSF in the catheter with respirations

or with lowering of the chamber. If there is no drainage or fluctuation within the tubing, the neurosurgery team should be notified immediately. If the occlusion in the catheter cannot be dislodged by flushing it, the patient may need to return to the operating room and have a new drain placed. If the system is accidentally pulled apart or broken, the proximal catheter should be clamped immediately, the open tip placed in a sterile covering, and the neurosurgery team notified.

CSF is generally clear and has the same consistency as water. In the presence of infection, it may become cloudy, darker in color, and more viscous. CSF samples are often drawn on a regular basis to monitor the treatment of infection or to rule out infection. The studies completed normally consist of a cell count, levels of glucose and protein, Gram stain, and culture. In the presence of infection, protein and white blood cells are usually elevated, and glucose is usually decreased. Blood is often present after posterior fossa tumor resections. The amount of blood present should be noted as well as any new bleeding.

CSF contains approximately 120 mEq/l of sodium. Therefore, the child's electrolytes should be monitored closely. Some surgeons may want to replace CSF with intravenous normal saline, particularly in younger children.

Other nursing considerations include keeping the head dressing clean, dry, and intact, as well as pain control. Distraction techniques or restraining the child may be necessary to keep the patient from moving or dislodging the catheter.

In patients with posterior fossa tumor resections, there is a chance that the ventricular drain may be weaned and removed completely. This is often done once the cellular debris and blood have cleared. The chamber is raised slowly over 24–96 h and may eventually be clamped. If the patient does not develop signs and symptoms of increased intracranial pressure, and a CT or HASTE MRI does not show enlargement of the ventricular system, the drain may be removed. If there are signs and symptoms of increased pressure and enlargement of the ventricles, the patient will most likely need a permanent shunt or may be a candidate for endoscopic third ventriculostomy.

2.13.6 Discharge

Most infants and children can be discharged 24–48 h after a shunt placement or revision. The child undergoing a third ventriculostomy may be in the hospital longer while evaluation of the efficacy of the procedure is carried out. In preparation for discharge, the nurse should discuss with the parents, or other caregiver, wound care, pain management, signs and symptoms of shunt failure and infection, and other issues that may be relevant. The caregiver should be instructed on how to take care of the dressing and/or wound. The surgeon will usually specify recommendations regarding dressings, bathing, and suture removal. The nurse should know what those specifications are and relay them to the family verbally and in writing. Often, families need to go home with dressing supplies if a dressing is to be kept in place.

Most children can be discharged with acetaminophen or ibuprofen for pain. The nurse should give the care provider the appropriate dose for the child. Some surgeons will prescribe a stronger pain medication such as oxycodone or codeine if it is necessary. Parents should be instructed to use any medication cautiously. If the child's pain is increasing over time, the child may be experiencing another shunt failure or infection. Pain is usually expected to be incisional or related to distal catheter implantation by the time of discharge.

The family needs to be instructed on the signs and symptoms of shunt failure and infection. The signs and symptoms may be subtle and confusing in a child already recovering from surgery. The caregivers should be told that an infected shunt may or may not function. Parents should also be advised that any fever during the first month after shunt placement could be related to a shunt infection. The patient's family also needs instruction on what to do if they suspect shunt failure, infection, or have other concerns. If the shunt fails at night or on the weekend, the child still needs immediate evaluation. Each surgeon handles this somewhat differently, and the nurse should know the expectations of the particular

surgeon. Follow-up appointments should also be scheduled.

2.13.7 Family Support

The family of a child with hydrocephalus is often quite anxious because the child may need to undergo repeated surgeries, and there are often other major illnesses or conditions associated with hydrocephalus. They may also be concerned about the lifelong implications of hydrocephalus, whether the infant or young child will be normal, have delays, cerebral palsy, or other conditions related to the hydrocephalus (or its etiology). The nurse or physician may not be able to adequately answer such questions and that only further increases parental anxiety.

Families need anticipatory guidance and teaching about hydrocephalus, including what the signs and symptoms are, why they occur, and what needs to be done. They need to understand what tests are done to diagnose hydrocephalus and shunt failure. Sometimes these tests are confusing or inconclusive for the provider. This further confuses families and causes more anxiety. Families also need to know the importance of prompt treatment of suspected shunt failure and have a plan as to how that will occur.

A child with a shunt should be encouraged to live as normal of a life as is possible. If the child does not have other associated conditions or delays, there will usually be no restrictions. If the child is delayed, in a wheelchair, blind, or otherwise disabled, many restrictions will be needed because of the underlying issues. Parents should still be encouraged to treat the child as normally as possible. Some surgeons do not want children with shunts to play rough contact sports such as football or wrestling because of the possibility of damaging the shunt hardware. There is no contraindication to flying in commercial pressurized aircraft.

Children with shunts need good primary care. The primary care provider as well as the parent can follow head growth in infants. A primary provider or nurse may help the family with all

the standard issues that parents face including discipline, toileting, sleep issues, child care, and schooling. These children need all the regular immunizations. The diphtheria-tetanus-pertussis vaccine should be administered to an infant or child with stable neurological conditions, including controlled seizures (Committee on Infectious Diseases 2000). They need good dental care to avoid the possibility of dental carries seeding a shunt infection during a shunt revision. Current American Dental Association guidelines do not recommend any prophylactic antibiotics for any patients containing neurosurgical hardware (including ventriculoatrial shunts) (Lockhart et al. 2007). Some surgeons may still recommend antibiotics before dental work in children who have had repeated shunt infections. Children also need routine vision screening because of the associated visual abnormalities.

Families who have children with hydrocephalus may benefit from a support group. Families may also benefit from information from national organizations such as the following:

2.13.7.1 Organizations and Web sites

Hydrocephalus Association
870 Market Street, Suite 705
San Francisco, CA 94102
415-732-7040; 888-598-3789
www.hydroassoc.org

Hydrocephalus Foundation
910 Rear Broadway, Rt. 1
Saugus, MA 01906
781-942-1161
www.hydrocephalus.org

National Hydrocephalus Foundation
12413 Centralia Road
Lakewood, CA 90715
562-402-3532; 888-857-3434
www.nhfonline.org

National Information Center for Children
and Youth with Disabilities
PO Box 1492
Washington, DC 20013
www.nichcy.org

Spina Bifida Association of America
4590 MacArthur Blvd. NW, Suite 250
Washington, DC 20007-4226
202-944-3285; 800-621-3141
www.sbaa.org

United Cerebral Palsy Association, Inc.
1660L Street, Suite 700
Washington, DC 20036
www.ucpa.org

2.14 Living with Hydrocephalus

Hydrocephalus is a chronic, lifelong condition. Untreated hydrocephalus has a mortality rate of 50–60 %. Surgically treated hydrocephalus in children with minimal or no evidence of irreversible brain damage is associated with a mortality rate of 10 % (Milhorat 1982). Some infants have a shunt placed at birth, require few revisions, and grow to be cognitively and physically normal. Others have a shunt placed and require many revisions, experience complications, and are mildly or markedly developmentally delayed. These ongoing issues with the shunt are not usually the only cause of the developmental abnormalities, but they may contribute to them. Some children with hydrocephalus have other chronic diseases that are associated with or are the cause of the hydrocephalus. Such illnesses include brain tumors, neurofibromatosis, myelomeningocele, craniofacial abnormalities, cerebral palsy, and various brain malformations. The treatment of hydrocephalus is just one aspect of the complicated care that these children require. Many factors affect the outcome of children with hydrocephalus, such as the age at onset, the underlying cause, the timing of the surgical intervention, complications such as repeated shunt failures and infections, and the associated comorbidities of other diseases.

All children with hydrocephalus are at risk for certain associated problems, including cognitive delays, learning disabilities, motor delays, behavioral abnormalities, visual abnormalities, seizures, precocious puberty, and diabetes insipidus. A French study (Hoppe-Hirsch et al. 1998)

evaluated 129 children with shunts. These children were shunted before age 2 years and followed for 10 years. The study found that 60 % had motor disabilities, 25 % had visual or auditory abnormalities, and 30 % had seizures. Sixty percent were in school, some with special services, and their IQs were highly variable. Thirty percent had IQs that were in the normal range above 90, 30–60 % had mild to moderate mental retardation, and 7–20 % had severe mental retardation (16). Many were 1–2 years behind their peers. Behavioral disorders were common.

2.14.1 Cognitive Abnormalities

Intellectual function is difficult to predict in the infant and young child. Abnormalities of, and insults to, the CNS may both contribute greatly to impaired function. The younger the child at the age of the onset of hydrocephalus, the greater the risk for intellectual abnormalities. Cognitive difficulties can also be caused by the underlying condition and associated treatment. Shunt infections, especially with gram-negative organisms, can further impair cognitive function. Some infants have extreme hydrocephalus at birth, but once shunted, the brain may grow into the existing space. Some of these children can develop normally. A cortical mantle of less than 5 mm in thickness seems to be predictive of a poor outcome. Cognitive function is also impossible to predict from radiographic studies. Some children are remarkably functional despite markedly abnormal appearing brains on CT and/or MRI scans. Other children have severe intellectual impairment with relatively normal appearing scans.

Sgouros et al. (1995) followed 70 patients with shunts for 16 years. He found that children with IVH and meningitis as the underlying cause of their hydrocephalus did the worst cognitively. Thirty to forty percent of these children had cognitive delays. He also found that two-thirds of these patients were socially independent but living with their parents.

Among children with myelomeningocele and hydrocephalus, cognitive abnormalities are more

pronounced in those with a higher-level defect, as compared to those with a sacral defect. Those that require a shunt (80–85 %) have overall lower IQ scores than those who do not require shunting (Mapstone et al. 1984).

Children with hydrocephalus also have a higher risk of learning disabilities. These children have difficulty with encoding and retrieval in both verbal and nonverbal tasks (Scott 1998). In addition, such children may have difficulty with reading comprehension (Yamada 2002). They also have difficulties with concentration, nonverbal learning, processing complex language, short-term memory, and poor spatial relations. These children are at a higher risk of attention deficit hyperactivity disorder than the general population. Dysmorphology of the cerebellum may be associated with oral and motor speech deficits (Huber-Okrainec et al. 2002).

Neuropsychological testing of the child will help to better define where the deficits exist. With such knowledge, learning and activities of daily living can be modified to fit the needs of the individual child. Special therapy, such as speech or occupational therapy, may help the child to become more functional.

The importance of social factors must also be considered when evaluating the intellectual function of these children. Those who have access to the most state-of-the-art medical care, therapy services, and educational services may do better functionally than those who do not. Some of these children also need complex care from their families on a daily basis. While some families are well equipped to deal with these demands, other families seem to be in a continual state of crisis, have several children with special needs, have one or both parents absent, or many other problems that make it difficult to care for these children.

2.14.2 Motor Disabilities

Sixty percent of children with hydrocephalus have varying degrees of motor abnormalities. The motor deficits are often related to the underlying etiology of the hydrocephalus. Children

with hydrocephalus may have global motor delays and achieve milestones, such as sitting and walking late, or not at all. Hydrocephalus may also affect fine motor control. Such fine motor difficulty may be exacerbated by visual impairments. These children may have trouble learning to write, so keyboards and communication boards may be useful. Premature infants with IVH may develop hydrocephalus and cerebral palsy. The cerebral palsy may be mild and affect only the lower extremities (spastic diplegia), or it may be severe and affect the entire body (spastic quadriplegia).

2.14.3 Ocular Abnormalities

Optic atrophy from chronic papilledema was the leading cause of blindness from congenital malformations, before the successful treatment of hydrocephalus. Increased intracranial pressure from hydrocephalus causes pressure on the cranial nerves. The cranial nerves that are involved in eye function are II (Optic), III (Oculomotor), IV (Trochlear), and VI (Abducens). As intracranial pressure increases, signs and symptoms become evident as these nerves are affected. Common findings include limited upward gaze, extraocular paresis, decreased vision, and diplopia (Table 2.5).

Papilledema is a less common finding and is difficult to diagnose in young children. The child is often referred to an ophthalmologist for a complete eye exam, including dilation of the pupil, to correctly diagnose papilledema. Papilledema is not a common finding in children with increased intracranial pressure unless it is chronic.

Ocular abnormalities are a common finding in infants and children with untreated hydrocephalus and during periods of shunt malfunction. If treatment is not prompt, visual damage is a risk, including blindness. Visual deficits are common in children with hydrocephalus. Refractive and accommodative errors are found in 25–33 % of these children. Gaze and movement disorders, such as nystagmus, astigmatism, strabismus, and amblyopia, are found in 25–33 % (Rosen 1998). Abnormalities in vision may be associated with

Table 2.5 Cranial nerves and eye symptoms

<i>II-Optic nerve</i>
Responsible for transmitting visual images from the eye to the brain
Test: check for light perception, visual acuity, peripheral vision, and normal appearance of the optic disc
<i>III-Oculomotor nerve</i>
Responsible for controlling four of the six muscle groups that move the eye
Medial rectus: moves eye inward
Superior rectus: moves eye upward and in
Inferior rectus: moves eye downward and in
Inferior oblique: moves eye upward and out
Responsible for constriction and accommodation of the pupil and closing of eyelid
Test: have child follow object in six cardinal positions of gaze, check for pupil reaction to light, check for closing of eyelid
<i>IV-Trochlear nerve</i>
Responsible for controlling superior oblique muscle that moves eye inward and down
Test: have child look down and in
<i>VI-Abducens nerve</i>
Responsible for lateral rectus muscle that moves eye temporally
Test: have child look temporally

lower IQ scores. Correctable vision issues should be identified and treated as early as possible so that they do not add to developmental and learning difficulties.

2.14.4 Seizures

Hydrocephalus alone is not commonly recognized as a cause of seizures. However, seizures are associated in children with hydrocephalus who have shunts implanted. The incidence of epilepsy in the general population is 1 % in children. The incidence of seizures in children with shunted hydrocephalus is 20–50 % (Sato et al. 2001). Since modern shunting became the standard treatment for hydrocephalus, controversy has existed about shunt procedures and complications of shunts leading to seizures. Children with hydrocephalus have numerous risk factors that increase the risk of seizures. These include age at the original shunt placement, the location of the shunt catheter in the

brain, the actual placement of the shunt catheter, repeated revisions of the proximal catheter, the presence of the hardware itself in the brain, the location of the burr hole, shunt infections, intracranial hemorrhage at the time of shunt placement or revision, repeated episodes of increased intracranial pressure, the presence of tumors or cysts, the underlying etiology of the hydrocephalus, and any associated developmental delay. When a child has seizures, a work-up is indicated including an electroencephalogram. Seizures are not usually a symptom of shunt malfunction although a very small percentage of patients will present with seizures as the main symptom of shunt malfunction.

2.14.5 Precocious Puberty

Precocious puberty is defined as the onset of puberty 1–2 years before the expected age. Normal onset of puberty may occur as young as eight in girls and nine in boys. Precocious puberty is fairly common in children with hydrocephalus, myelomeningocele, cerebral palsy, and microcephaly. It can start as early as 5 years of age.

Precocious puberty is presumed to be caused by chronic or intermittent increased intracranial pressure that affects the hypothalamus and pituitary gland. These two areas deep in the brain are responsible for timing the release of gonadotropins and sex hormones. Children who have had many shunt revisions when they are very young are at greater risk for precocious puberty.

Precocious puberty causes the growth plates to close early in the long bones leading to short stature. Additionally, precocious puberty is associated with risk of pregnancy in young children with associated cognitive and behavioral difficulties.

2.14.6 Transition to Adulthood

Hydrocephalus is a chronic disorder that will follow children into adulthood. Specifically in a shunted patient, it is a condition that requires regular follow-up and established care in case of a medical emergency, such as a shunt malfunction. A study by Simon et al. (2009a, b) showed that the number of young adults aged 18–35 with hydro-

cephalus that need treatment in the United States is predicted to exceed 40,000 annually within the next two decades. These young adults need access to both expert surgical and medical providers. As pediatric providers, it is vital to prepare patients for transition to adulthood. Studies show that one-half to two-thirds of patients failed to maintain adequate shunt follow-up after transitioning to adult care (Simon 2009). It is recommended to start discussions with families as young as 16 years of age, to establish who their adult provider will be and how transition of care will happen. The hope is to have the young adult establish care while in a healthy state if possible and not have first contact with their new provider in an emergency. It is also important to discuss with families the issue of private insurance versus qualifying for a government-funded program, to ensure adequate accessibility to the healthcare system.

Conclusion

The Nurse's Dictionary of Medical Terms and Nursing Treatment (Morton, circa 1898) stated the following about hydrocephalus: "water on the brain, a disease most common in children, and causing the head to swell to an enormous size. The victim is always idiotic. Nourishing diet – cod liver oil; as little fluid as possible." As this chapter has shown, nurses today need to know far more about the condition and its treatment. Most importantly, they need to know that many children with hydrocephalus can lead normal lives because of available surgical treatment. Despite advances in technology and surgical technique, nurses have and will continue to play major roles in achieving the best possible outcomes for these patients.

Pediatric Practice Pearls

- If the mother thinks that the child is acting abnormally or that the shunt is not working, there is a high probability that she is right.
- Small ventricles do not assure adequate shunt function.
- Altered mental status is the first and most salient symptom of increased

intracranial pressure, thus the child needs to be awakened for assessment.

- Mental status should be assessed over time for changes. Never give patients with altered mental status, or an unstable exam, medications that could mask the exam and symptoms.

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