CME

Recognizing and managing hydrocephalus in children

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ABSTRACT

Hydrocephalus is one of the most common indications for pediatric neurosurgical intervention and is associated with the need for lifelong monitoring. All clinicians should be familiar with the complications that may arise throughout life in these patients so that they can provide timely intervention. This article focuses on the assessment of hydrocephalus, the appropriate diagnostic workup and differential diagnoses, and evidence-based surgical treatments and associated outcomes.

Keywords: hydrocephalus, pediatric, complications, cerebrospinal fluid, macrocephaly, VP shunt

Learning objectives

- Recognize the signs and symptoms of hydrocephalus in infants and children.
- Accurately assess and diagnose hydrocephalus in infants and children.
- Understand the most common management modalities for hydrocephalus in children and their corresponding indications and complications.

In patients with hydrocephalus, an alteration in the production, circulation, or absorption of cerebrospinal fluid (CSF) results in an overaccumulation of volume in the central nervous system (CNS) and, subsequently, abnormal enlargement of the cerebral ventricles.[1] The Congress of Neurological Surgeons estimates that hydrocephalus occurs in as many as 1 in every 500 children in the United States, making it one of the most common indications for neurosurgical intervention in children.[2]

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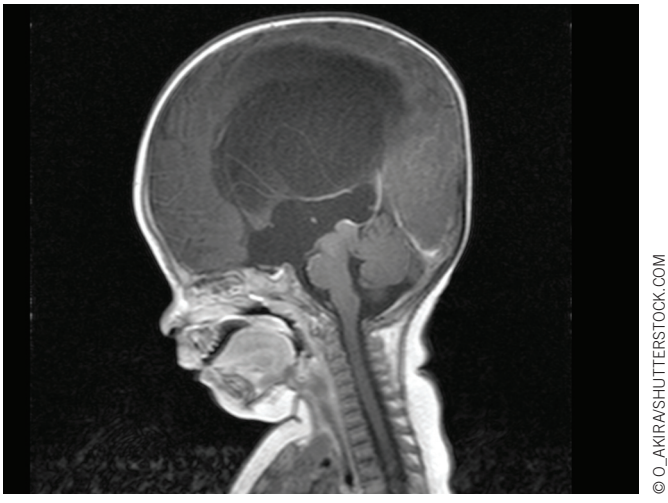


Potential risk factors for hydrocephalus include the infant's male sex and the mother's young or advanced age (under 20 years or 35 years and older), poor access to healthcare, and multifetal pregnancies.[3,4]

Pediatric-onset hydrocephalus is associated with lifelong implications that must be managed throughout the child's lifespan. Untreated hydrocephalus is associated with high morbidity and mortality. Delays in treatment can lead to lifelong intellectual, sensory, communication, and physical disabilities.[5,6] Clinicians in all settings must be able to identify the clinical manifestation of hydrocephalus in order to promptly implement appropriate treatment.

CAUSES

Pediatric-onset hydrocephalus may arise from a wide array of congenital and acquired processes affecting various points in CSF circulation that result in blockage of the CSF outflow, reduced CSF absorption, or excess CSF production (**Figure 1**). These pathophysiologic processes can alternatively be classified into different forms, including obstructive and communicating, both of which describe the disturbance to CSF circulation. Obstructive processes involve blockage of one or more of the channels connecting the ventricles. Communicating processes develop in the setting of disproportionate absorption and production of CSF, without underlying ventricular obstruction. Congenital causes include primary aqueductal stenosis, Dandy-Walker malformation, spinal dysraphism

Key points

- Hydrocephalus is one of the most common indications for pediatric neurosurgical intervention and is associated with the need for lifelong monitoring.
- Delays in treatment can lead to lifelong intellectual, sensory, communication, and physical disabilities.
- Children with hydrocephalus often present with complaints consistent with increased ICP. This presentation may differ based on age, cause, and comorbidities.
- Surgical diversion of CSF is the mainstay of treatment for children with hydrocephalus.

(such as myelomeningocele or encephalocele), Chiari malformations, space-occupying lesions, and genetic disorders.[7] Acquired causes include infection, intracranial hemorrhage, and benign and neoplastic lesions.[7]

CLINICAL MANIFESTATIONS

Children with hydrocephalus often present with complaints consistent with increased intracranial pressure (ICP).[7] This presentation may differ based on age, cause, and comorbidities. Promptly recognizing findings associated with increased ICP is critical to preventing irreversible brain injury.

The earliest complaint in patients with ICP elevation is severe headache. In infants and young children, headaches may present as inconsolable irritability. Other general findings of elevated ICP include profuse vomiting, changes in vision (diplopia or blurred vision), agitation, fatigue or lethargy, gait abnormalities, anorexia, seizures, weakness, strabismus (most often esotropia), and the *setting sun* sign, which is characterized by the downward gaze of pupils (**Figure 2**).[7–10]

History suggestive of chronic elevations in ICP, which is the most common presentation of pediatric hydrocephalus, may include insidious development of symptomatology, chronic progressively worsening headaches, and delayed

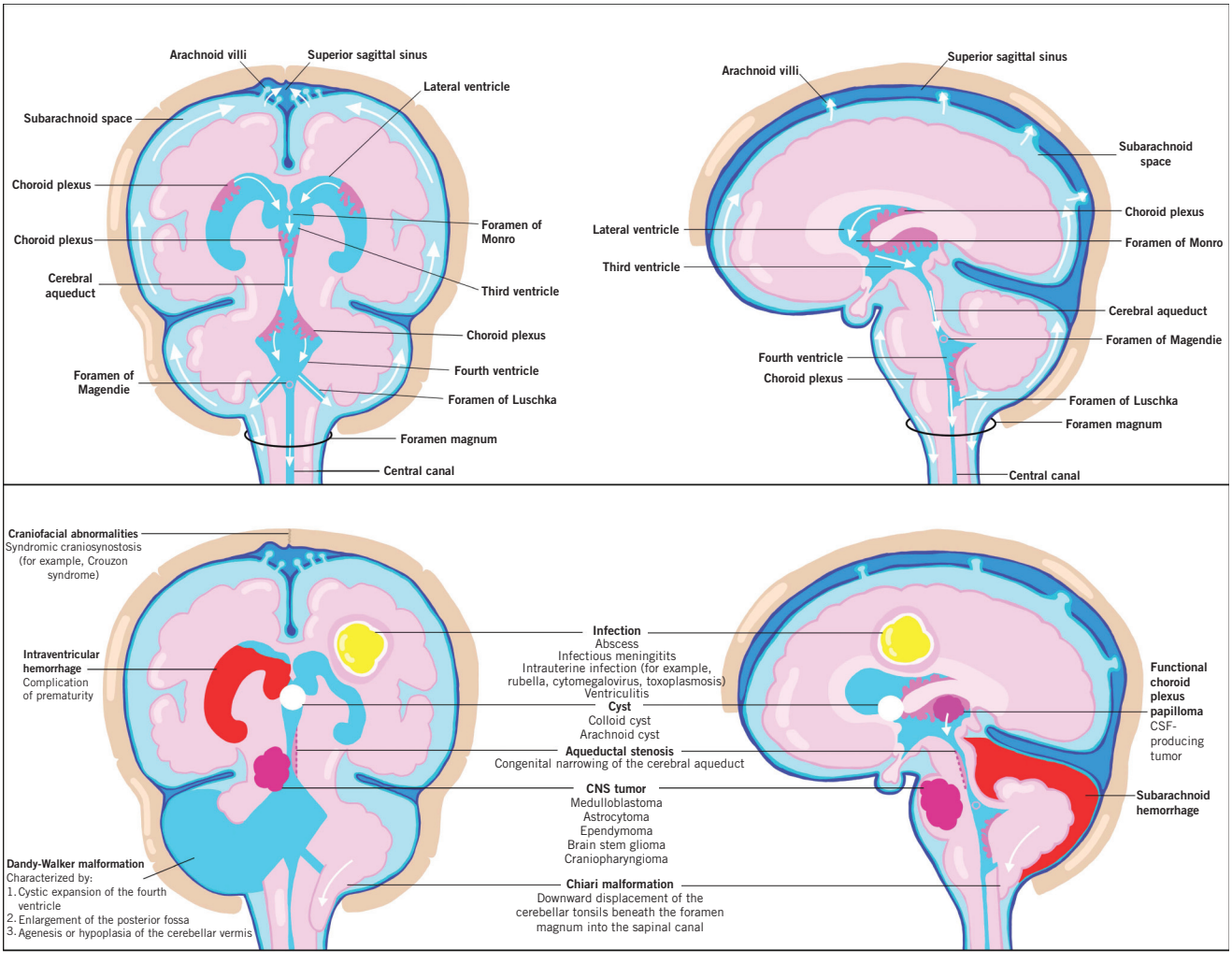


FIGURE 1. Normal circulation of cerebrospinal fluid (top) and pathologic states leading to development of hydrocephalus (bottom)

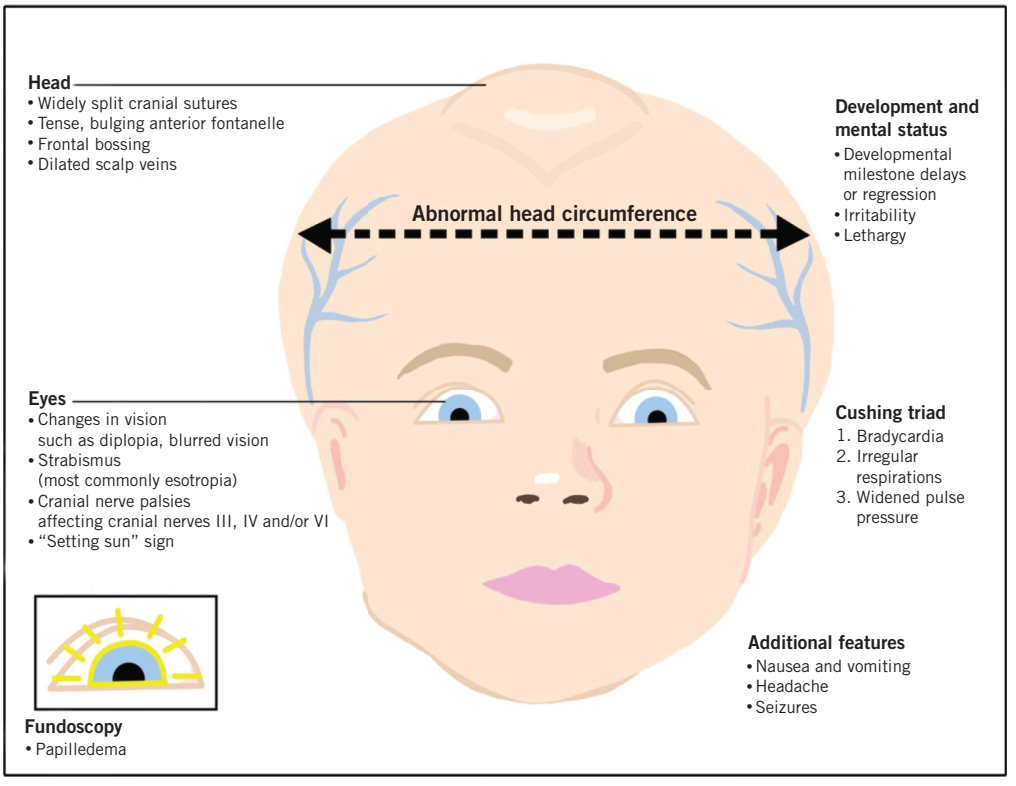


FIGURE 2. Clinical features of hydrocephalus

developmental milestones or regression in developmental milestones (**Table 1**). Infants and toddlers also may present with a history of macrocephaly. Acute onset of symptoms, such as sudden changes in mentation, may be suggestive of acute increases in ICP.

If hydrocephalus is suspected, assess the patient's vital signs, looking for bradycardia and hypertension. Cranial percussion may reveal Macewen sign, a characteristic "cracked-pot" sound, indicating high intracranial fluid volume.[8] Findings of elevated ICP unique to infants and toddlers include tense or bulging fontanelle, prominent scalp veins, frontal bossing, and palpable splitting of the cranial sutures.[8,9] These findings depend on the presence of an open fontanelle, which will typically close between ages 4 and 24 months in a healthy child.

Papilledema may be present on a fundoscopic examination, but its absence is not exclusionary.[11] In a patient with chronic ICP elevations, papilledema may be accompanied by blurred vision and/or visual field defects.[12] Ophthalmology consultation often is indicated in the workup of a patient with potentially elevated ICP. Cushing triad (bradycardia, irregular respirations, and widened pulse pressure) describes late findings of acutely increased ICP and is generally associated with poor prognostic outcomes.[13]

SCREENING AND DIAGNOSIS

The mainstay of screening for hydrocephalus includes fetal ultrasound and head circumference monitoring. Most patients are diagnosed *in utero* during a routine anatomy scan between 15 and 35 weeks gestation. During this study, the lateral ventricles are measured for ventriculomegaly, dilation of one or both lateral ventricles to 10 mm or greater.[14] This is suggestive of congenital obstruction, most commonly from aqueductal stenosis. If dilation is evident on ultrasound, antenatal MRI can be used for confirmation. Further workup also should be conducted in the postnatal period.

Occipitofrontal circumference measurements are included in the standard of care for all children early in life. The CDC recommends serial occipitofrontal circumference measurements from birth to age 36 months.[15] The American Academy of Pediatrics recommends that a child's occipitofrontal circumference be measured at least eight times during the first 24 months of life.[16] Full-term infants will typically have an occipitofrontal circumference between 33 and 37 cm (10.8 to 12.1 in).

In the primary care setting, an occipitofrontal circumference greater than 3 standard deviations from average or greater than the 97th percentile for a given age and sex is clinically significant and warrants referral to pediatric neurosurgery for further evaluation.[17] In premature infants, use age-adjusted reference ranges until the child is at least age 24 months.[18] A curved pattern in occipitofrontal circumference measurements is expected throughout the course of development because of a deceleration in the rate of cranial growth.[15] Neurosurgical referral is warranted in patients with significant increases in head circumference that do not follow the patient's expected growth curve.

To measure head circumference, place the starting end of the measuring tape at the occiput, the most prominent point on the back of the head. Wrap the measuring tape around to the supraorbital ridges, located superiorly to the eyebrows, and back around until the two ends meet.

In children with head circumference measurements that are concerning for hydrocephalus, the neurosurgery team may choose to proceed with neuroimaging or to act conservatively by closely monitoring developmental milestones and performing serial occipitofrontal circumference measurements.

Neuroimaging can involve MRI, head ultrasound, or ocular ultrasound, depending on patient age, severity of symptoms, and institutional availability. MRI generally is the imaging modality of choice for the initial diagnosis of hydrocephalus.[19,20] MRI is used to assess CSF flow dynamics and visualize CSF-containing cavities.[9] Transependymal edema, or a smooth, hyperintense halo, is suggestive of outflow of CSF into surrounding brain tissue.[21] This finding typically is best seen on a fluid-attenuated inversion recovery sequence. Depending on institutional availability, ultrafast MRI, which is only 1 minute in length per sequence, can be used as an alternative to standard MRI to reduce motion artifacts without the need for sedation.[20]

Head ultrasound can be performed in place of MRI for children under age 1 year, depending on the presence of adequate opening of the anterior fontanelle.[22] Head ultrasound often is preferred over MRI in this age group and can be used to quantify, monitor, and confirm ventricular dilation as well as to aid in the determination of its cause. Head ultrasound also may be performed as a screening measure in newborns with myelomeningocele.

Optic nerve sheath diameter is a noninvasive indirect measure of ICP that can be obtained via ocular ultrasound.[23,24] Increases in optic nerve sheath diameter occur quickly after the onset of ICP elevation, making this test appropriate for patients presenting to the pediatric emergency and critical care settings in critical condition with history suggestive of increased ICP.[24]

Definitive evaluation of ICP can be achieved by ICP monitoring with an intraparenchymal monitor or an external ventricular drain (EVD), which can be both diagnostic and therapeutic. Due to its invasive nature, ICP monitoring should be limited to select patients. ICP monitoring may be appropriate in patients with inconclusive imaging despite high clinical suspicion. ICP monitoring also can be used when there is a need to observe changes in ICP over time. This often is indicated following surgical correction of the underlying cause, to monitor for resolution of associated hydrocephalus. Because of its therapeutic nature, ICP monitoring with EVD may be warranted in patients with severe, sudden onset of hydrocephalus.

ICP may vary based on patient age, weight, and physical position. Elevated ICP generally is defined as pressure exceeding 20 mm Hg for 5 minutes or longer.[25,26] Also track cerebral perfusion pressure (CPP), which can be calculated by subtracting ICP from mean arterial pressure.[7] A low CPP (below 45 mm Hg) is suggestive of poor cerebral perfusion and a high risk for ischemic brain injury.[27]

DIFFERENTIAL DIAGNOSIS

Before initiating management for suspected hydrocephalus, which often is invasive in nature, consider these alternative potential diagnoses.

Benign extra-axial fluid of infancy (also called benign enlargement of subarachnoid spaces, benign macrocephaly, and benign external hydrocephalus) is a common cause of macrocephaly.[28] Studies have demonstrated a strong familial link, suggesting a genetic component. When taking patient history, ask the patient's parents about a family history of macrocephaly in order to help differentiate this process from hydrocephalus (**Table 1**). Although the pathophysiologic processes are not well understood, macrocephaly is thought to occur because of underdevelopment of arachnoid villi. As the arachnoid villi mature, head size typically stabilizes, with complete resolution by age 2 years.[28]

The typical presentation of a patient with benign extra-axial fluid of infancy is a developmentally normal infant, age 6 months or younger, with macrocephaly. However, a growing number of researchers have suggested a correlation between increased extra-axial fluid and autism spectrum disorder.[29,30] These studies have shown prolonged retention of fluid volumes lasting through ages 3 to 4 years.[30] Imaging findings of benign extra-axial fluid of infancy typically include increased volume of the subarachnoid space, with the largest expansion along the frontal convexities.[17] Lateral ventricles generally are normal or slightly enlarged.

Migraines are a common neurologic condition in children.[31] In contrast to headaches associated with hydrocephalus, migraines in children present with a prodromal period beginning hours to days before the migraine's onset, consisting of combinations of fatigue, poor concentration, irritability, neck stiffness, blurred vision, nausea, phonophobia, and photophobia.[32] Similarly to migraines, headaches associated with pediatric hydrocephalus may present with photophobia. Unlike migraines in adults, which typically present with unilateral pain, pediatric migraines often follow a bifrontal or bitemporal pain distribution.[33] Pediatric migraines also may be associated with an aura. Typical auras present with positive manifestations followed by negative manifestations.[31] The most commonly reported features are visual in nature; for example, shimmering, flashing lights and/or visual hallucinations (such as uniform patterns or shapes) followed by scotomata.[31] Symptoms can include sensory and speech/language manifestations. Patients

TABLE 1. Eliciting an accurate patient history

If hydrocephalus is suspected in a child, ask parents:

- Is your child having episodes of fatigue so severe that you cannot wake the child up for hours at a time?
- Is your child having episodes of irritability and inconsolable crying so severe that you cannot calm or distract the child?
- Is your child having bouts of projectile vomiting unrelated to any attributable causes, such as feedings or gastrointestinal illnesses?
- Have you noticed any changes in your child's eye movements? For example, the eyes moving in different directions or getting stuck in the downward position?

often experience auras that include multiple categories of features. The International Classification of Headache Disorders, 3rd edition provides diagnostic criteria for migraine diagnosis.[34] Features of headaches that are concerning for secondary pathologies such as hydrocephalus and may warrant further workup include progressively worsening headaches, headaches that are more prominent in the morning on awakening, headaches that may be relieved with sitting up, and headaches that awaken the patient from sleep.[34]

Abusive head trauma is associated with a considerable risk for morbidity and mortality. Keep a high index of suspicion in infants and young children presenting without a known cause of common, potentially related findings.[35] Mechanisms of abusive head trauma include isolated shaking, direct impact, or a combination.

The classic triad of violent shaking injuries include encephalopathy, subdural hematoma, and retinal hemorrhage; however, the validity of this presentation has been debated in recent literature.[36] Rib fractures also are commonly associated with shaking injury attributed to squeezing of the child's chest during the action.[37] Clinical features can include general findings such as vomiting and irritability. Apnea is a common finding in patients with severe abusive traumatic brain injuries.[38] This may be explained by brainstem dysfunction, which is theorized to be secondary to the cervical spine injury that occurs in 15% to 46% of patients with abusive head trauma.[38] Between 28% and 50% of patients have seizures.[38] As with other causes of head trauma, abusive head trauma may lead to development of hydrocephalus; therefore, the confirmation of hydrocephalus itself is not sufficient to rule out abusive head trauma. The Pediatric Brain Injury Research Network's four-variable abusive head trauma clinical prediction rule can be used to predict risk for abusive head trauma and should be considered regardless of the presence of diagnostic findings consistent with hydrocephalus.[39] This screening assessment should raise suspicion of abusive head trauma in any child under age 3 years admitted to the pediatric ICU (excluding those with known mechanisms of unintentional injury) who presents with one or more of the following:

- Respiratory compromise before admission
- Bruising of the ears, neck, and torso
- Bilateral or interhemispheric subdural hemorrhages and/or fluid collections
- Skull fractures (excluding those with typical accidental characteristics: isolated, unilateral, nondiastatic, linear, parietal location).[39]

Note that this screening tool has been found to have a high rate of false positives.

In patients with suspected abusive head trauma, obtain a CT of the head and cervical spine to assess for fractures and hemorrhage. MRI may be used for further investigation. Skeletal survey should always be used to assess for additional bony injury. Subdural hemorrhages found on imaging in abusive head trauma often are mixed density and located in various locations.[37] Bony findings on CT head suspicious for direct cranial impact include unexplained fractures, bilateral skull fractures, occipital skull fractures, and multiple fractures in different stages of healing.[36]

MANAGEMENT AND FOLLOW-UP

The mainstay of treatment for children with hydrocephalus involves surgical diversion of CSF.

Short-term approaches Temporary interventions, such as ventriculosubgaleal (VSG) shunting, EVD, and CSF tapping, are indicated to provide immediate decompression in patients with acute or decompensated hydrocephalus.[40] Although this is successful in preventing the need for internal shunting in some patients, permanent measures often are needed.

VSG shunting involves the insertion of a catheter to create a connection between the lateral ventricle and the subgaleal space.[41] CSF then accumulates in the subgaleal pouch and eventually is absorbed by the bloodstream. VSG shunts often are used in neonates with hydrocephalus secondary to germinal matrix hemorrhage or infection to allow time for the body to become more suitable for ventriculoperitoneal shunt (VPS) insertion, a long-term approach described below.[42]

EVDs are a temporary option that can be diagnostic and therapeutic for patients with elevated ICP. EVDs consist of a catheter with its proximal end inserted, most commonly, into the lateral ventricle through Kocher point (which is at the intersection of the tragus line and the midpupillary line). The distal end of the catheter connects to an external collecting system.[43] EVD placement can be performed in the OR at bedside and with or without ultrasound guidance. Depending on institutional policy, PAs practicing in neurosurgery or neurocritical care can independently place EVDs under general supervision. Studies have demonstrated equivalent accuracy and complication rates in EVDs placed by PAs and NPs versus senior neurosurgeons.[44]

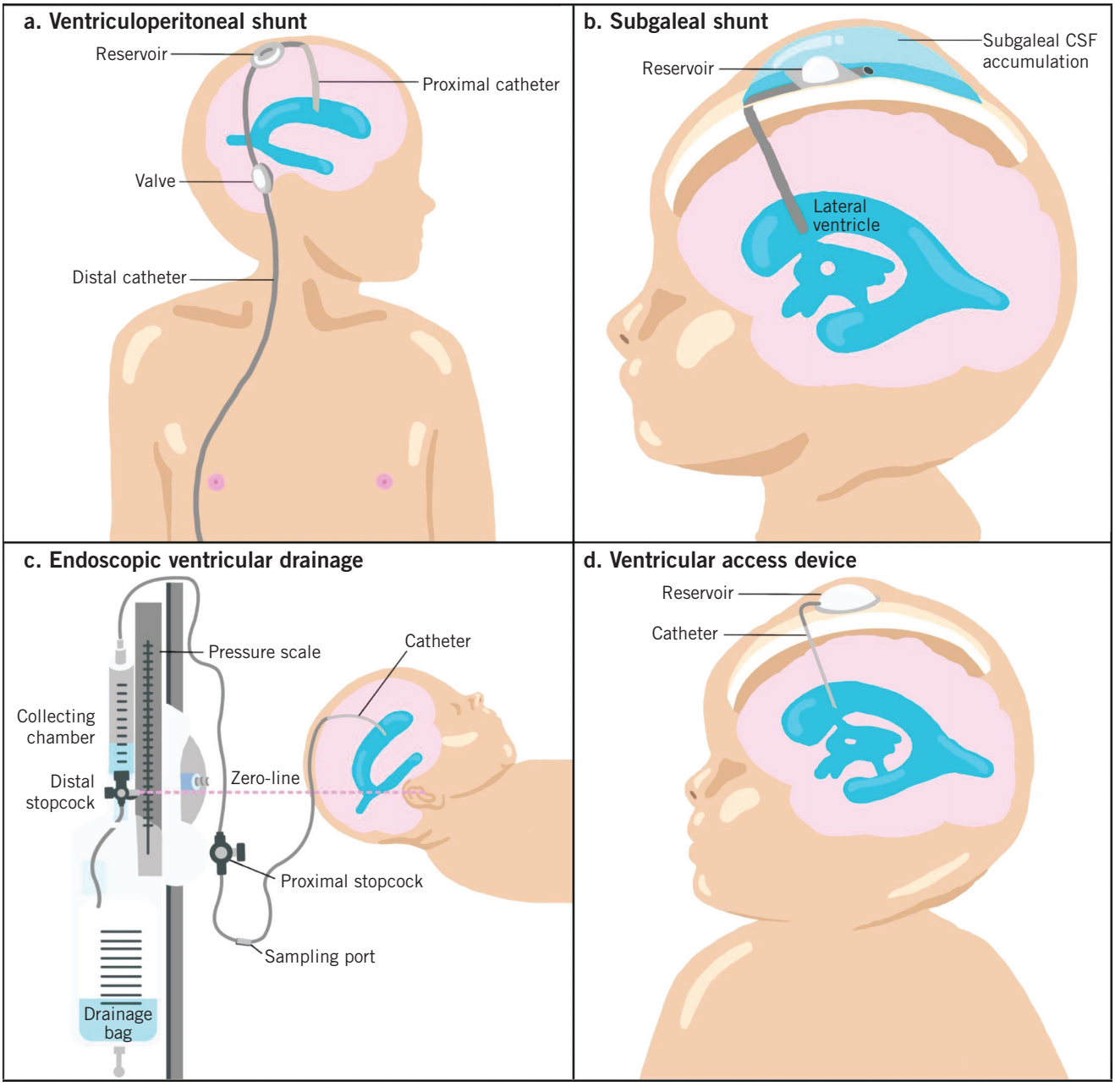
Following EVD placement, the pressure transducer is zeroed at the level of the tragus horizontally, often using a laser built into the device for precision. The device should remain at this level relative to the patient at all times so it can drain at the intended settings. Any changes in the height of the pressure transducer of the EVD relative to the height of the tragus will alter the pressure needed for CSF to drain into the chamber. The EVD should be clamped during events such as transportation or therapy sessions when the patient is moving.

Serial CSF tapping can be performed to help reduce CSF volume. In young children with open fontanelles, CSF can be accessed directly through the lateral angle

of the anterior fontanelle. In children with closed fontanelles, a ventricular access device, such as an Ommaya reservoir, can be used. A ventricular access device can be tapped using a standard 23- to 25-gauge butterfly needle.

Long-term approaches VPS insertion is the gold standard long-term intervention for the management of hydrocephalus (**Figure 3**). The proximal catheter of the shunt is inserted in a lateral ventricle. The reservoir serves as an access point to tap the shunt using a standard 23- to 25-gauge butterfly needle. Accounting for the valvular component of the shunt, traditionally, a fixed pressure valve has been used. With advancements in VPS technology, programmable shunt valves are increasingly used, and allow for noninvasive adjustments in valvular pressure settings using magnets. Studies have shown similar data in the incidence of shunt revisions in patients with both valve types.[45] A lateral plain radiograph can be used to visualize a valve in order to identify whether a valve is fixed or programmable. The distal catheter in a VPS is tracked along the side of the neck and down into the peritoneal cavity where the CSF is emptied.

FIGURE 3. CSF diversion procedures for hydrocephalus



Ventriculoatrial shunts (VAS) and ventriculopleural (VLP) shunts can be used in patients with contraindications to distal catheter placement in the peritoneal cavity. The differentiating feature of these shunts is simply the endpoint of the distal catheter—the VAS drains into the atria and VLP shunts drain into the pleural space. These alternative procedures are second line because of the additional associated risks.

The most common early complication of CSF shunting is infection, which typically occurs within weeks to months of shunt insertion, though later-onset infections have occured.[46] Shunt failure and malfunction are common long-term complications that can arise at any point in life following VPS placement. Failure and malfunction can be caused by obstruction, kinks, or knots in the catheter tubing, migration of proximal or distal shunt components, or valve malfunction.[47] Studies show an increased likelihood of shunt failure occurring in patients with neonatal and infantile shunt placement. To determine this, during routine physical examinations of patients with shunts, palpate along the cranial end of the distal catheter along the scalp and neck to feel for kinks in the catheters; however, this may be limited in some patients due to body habitus.

In a patient with suspected shunt malfunction, obtain a shunt series radiography and ultrafast MRI or CT of the head. Ultrafast MRI is becoming more commonly used in the workup of these patients because it does not require the ionizing radiation that is associated with CT scans. Shunt reservoirs can be tapped to assess proximal shunt function and provide an estimated ICP.[48] Thermal flow evaluation also has been shown to be a useful adjunct to imaging by detecting and assessing CSF flow through the distal catheter. Thermal flow evaluation is used at large facilities including Boston Children's Hospital and Children's Hospital of Philadelphia.[49] Studies have found this technique to have a high sensitivity for recognizing cases of catheter obstruction.[49] CSF shunt scintigraphy studies also can be performed to assess shunt patency.[48]

Constipation can result in transient VPS malfunction, and should be prevented or treated promptly in patients after VPS insertion. This can be particularly complicated in children with neurogenic bowel dysfunction.[50] Closely monitor all children with VPSes for constipation and establish a bowel regimen promptly if present.

Endoscopic third ventriculostomy (ETV) provides an alternative to CNS shunt insertion that is free of exogenous material by internally bypassing the site of obstruction in patients with obstructive hydrocephalus. During the ETV procedure, an endoscope is used to create a hole in the floor of the third ventricle, letting CSF drain into the subarachnoid space and ultimately pass into venous circulation via the arachnoid villi.

ETVs generally have a higher incidence of short-term failure but a lower incidence of long-term failure compared with VPS.[51] The ETV success score (ETVss), developed in 2009 by Kulkarni and colleagues, is widely used to predict the likelihood of success of ETV (**Table 2**).[52,53] To derive an estimated 6-month probability of ETV success, ETVss uses age, cause, and history of previous shunt placement. Scores are calculated in intervals of 10: a score of 0 to 40 indicates low likelihood of ETV success; 50 to 70 indicates moderate likelihood; and 80 or greater indicates high likelihood of success.

ETV/choroid plexus cauterization (ETV/CPC) involves ETV with the addition of cauterizing the choroid plexus in the lateral ventricles to decrease CSF production. ETV/CPC has been shown to have higher success compared with ETV alone in infants and young children ages 3 months to 24 months.[54] ETV/CPC success in this age range has been found to vary based on the cause of hydrocephalus, with higher rates of success among patients with aqueductal stenosis and myelomeningocele, and lower rates in patients with preterm intraventricular hemorrhage.[55]

TABLE 2. ETVss[53,56]

A score of 0 to 40 indicates low likelihood of ETV success; 50 to 70 indicates moderate likelihood; and 80 or greater indicates high likelihood of success.

Variable		Points
Age	<1 month	0
	1 month to <6 months	10
	6 months to <1 year	30
	1 year to <10 years	40
	≥10 years	50
Cause	Infection	0
	Myelomeningocele, intraventricular hemorrhage, or nontectal brain tumor	20
	Aqueductal stenosis, tectal tumor, or other pathology	30
History of previous shunt placement	Yes	0
	No	10

FOLLOW-UP

Patients with hydrocephalus should follow up closely with neurosurgery for long-term surveillance. Head circumference and developmental progress monitoring are essential. Routine follow-up with ophthalmology for annual fundoscopic examinations often is indicated as a measure of screening for increased ICP.

CONCLUSION

Pediatric-onset hydrocephalus is a common neurosurgical problem with lifelong implications. Although prompt neurosurgical intervention can allow patients with hydrocephalus to lead relatively normal lives, patients face a

lifelong risk of surgical complications and treatment failure. Clinicians must be familiar with the presentation of new-onset hydrocephalus as well as the signs of shunt failure or malfunction and use up-to-date diagnostic modalities to provide an appropriate workup. Familiarity with modern management techniques provides the foundation to recognize treatment failure, malfunction, or complications. **JAAPA**

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