Infant Hydrocephalus

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PRACTICE GAPS

Infant hydrocephalus is a highly heterogenous diagnosis that will most likely be encountered by pediatric physicians in their practice. Infant hydrocephalus encompasses a complex pathophysiology and can manifest in many different acquired and congenital forms. Furthermore, there are many parallel signs and symptoms as well as accompanying diagnoses that need to be distinguished to best triage infant hydrocephalus in general. Understanding the similarities and differences of these types and the available surgical treatments is key to achieving optimal management and outcomes.

OBJECTIVES *After completing this article, readers should be able to:*

1. Describe basic hydrocephalus pathophysiology.
2. List the clinical features of the most common acquired and congenital types of infant hydrocephalus.
3. Highlight the outcomes of surgical treatment of infant hydrocephalus.

ABSTRACT

Hydrocephalus is a neurosurgical condition that is highly prevalent in pediatric medicine. In the infant population, there is a distinct set of features that all primary pediatricians would benefit from understanding. Infant hydrocephalus can present prenatally on imaging and postnatally with symptomatic enlargement of the head and associated skull features and raised intracranial pressures. The 2 major pathophysiology models of infant hydrocephalus are the bulk flow and the intracranial pulsatility models. The most common acquired forms of hydrocephalus include posthemorrhagic hydrocephalus, postinfectious hydrocephalus, and brain tumor. The most common congenital forms of hydrocephalus include those due to myelomeningocele, aqueductal stenosis, and posterior fossa malformations. There are various evaluation and treatment algorithms for these different types of hydrocephalus, including cerebrospinal fluid shunting and endoscopic third ventriculostomy. The aim of this review was to elaborate on those features of hydrocephalus to best equip primary pediatricians to diagnose and manage hydrocephalus in infants.

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ABBREVIATIONS

CIM	Chiari type I malformation
CPC	choroid plexus cauterization
CSF	cerebrospinal fluid
DWM	Dandy-Walker malformation
ETV	endoscopic third ventriculostomy
ICP	intracranial pressure
IVH	intraventricular hemorrhage
MMC	myelomeningocele
PHH	posthemorrhagic hydrocephalus
PIH	postinfectious hydrocephalus

INTRODUCTION

Hydrocephalus (Greek *hydro* = water, *kefala* = head) remains one of the most common pediatric and infant conditions that requires specialized pediatric attention. Hydrocephalus remains a global issue, with annual specialized health expenditures costing approximately $2 billion in the United States, and beyond $1 billion in sub-Saharan Africa. (1)(2) Hydrocephalus occurs when there is abnormal buildup of cerebrospinal fluid (CSF) in the ventricular spaces of the brain, leading to increased intracranial pressures (ICPs), splaying of cranial suture lines, and various neurologic symptoms and deficits. (3) The morbidity burden of this condition can be very high, especially in infants (<12 months of age). Serious long-term and permanent neurologic sequelae are possible, including cerebral palsy, epilepsy, and cognitive and behavioral delays. (4) Timely detection, evaluation, and surgical treatment can minimize the risk of these issues.

Historically, Hippocrates was one of the first to describe accumulation of water in the brain as hydrocephalus in 400 BC. (5) By the 1800s, it had become accepted that this water was indeed CSF, and the symptoms of hydrocephalus were a result of increased ICP, (6) as demonstrated by symptom relief with direct drainage of CSF (either from the brain as a ventriculostomy or from the thecal sac as a lumbar puncture). (7) But it was only by the early 1900s that surgical treatments were proved to be more permanently effective and safe, first with third ventriculostomies followed by ventricular shunting to the peritoneum and other destinations. (8)

When discussing hydrocephalus, the terms *ventriculomegaly* and *macrocephaly* are frequently mentioned; the distinction between these terms is critical. By definition, hydrocephalus refers to the clinical manifestations of increased ICPs, which include increasing head circumference and irritability in infants. Ventriculomegaly refers to the radiographic increase in ventricle system size. Ventriculomegaly contrasts to macrocephaly, which refers to a physical head circumference that is above the 98th percentile age-standardized measurement. (9) To be clear then, hydrocephalus can present with ventriculomegaly and macrocephaly; however, ventriculomegaly and macrocephaly can present due to other causes as well and are not exclusive to the hydrocephalus diagnosis only.

The first objective of this review was to summarize our current understanding of how hydrocephalus develops and can present in infants. The second objective was to highlight the most common forms of hydrocephalus that can present to a practicing primary pediatrician. Finally, the most common treatment options for these patients are described.

EPIDEMIOLOGY

It is estimated that there are almost 400,000 new cases of infant hydrocephalus diagnosed each year worldwide, which approximates to a rate of 20 to 110 per 100,000 live births. (10)(11)(12) There are geographic and socioeconomic biases to consider, however, as the burden of disease is not distributed evenly. Up to 75% of the global burden falls in the countries of Africa, Latin America, and Southeast Asia. With respect to the pooled incidence of congenital hydrocephalus, correspondingly the highest values are seen in those same regions, up to 5 times more than in North America, where the burden is the lowest. (10) The incidence is higher in low- and middle-income countries (123 per 100,000 births) than in high-income countries (79 per 100,000 births). (10) It has been posited that drivers of these disparities derive from quality and access to prenatal care and education, with endemic infections, poorer diets, and limited access to diagnostic imaging in particular regions all leading to specific types of infant hydrocephalus being more prevalent in some regions over others. (2)(13)(14) Furthermore, how hydrocephalus can be treated varies worldwide too. In low- and middle-income countries, materials such as imaging equipment, shunt valves, and endoscopy equipment are unavailable, limiting the efficacy of hydrocephalus treatment. (15) There are likely many more patients in these countries who do not receive timely treatment as in high-income countries. (16)

CLINICAL PRESENTATION AND EVALUATION

There are very few clinical signs or symptoms of infant hydrocephalus before delivery of the infant. While in utero, fetal ventriculomegaly can be detected on surveillance ultrasonography. Fetal ventriculomegaly is defined as a greater-than-normal diameter of the lateral ventricle atria, typically greater than 10 mm. (17) Specialist interpretation of prenatal imaging is required, however, because ventriculomegaly can also result from abnormal brain development and not necessarily reflect hydrocephalus caused by increased ICPs.

In infants, signs of hydrocephalus are more subtle than the classic symptoms seen in older children and adults. This is because neonates are born with open fontanelles and skull sutures, which can accommodate increases in ICP more so than a more mature skull that is fixed in intracranial volume. As such, early signs of hydrocephalus in infants are mostly physical rather than symptomatic in

nature and include a growing head circumference crossing multiple percentiles over time, bulging fontanelles, and separated (splayed) cranial sutures. As hydrocephalus progresses untreated, these patients can present with vomiting, irritability, lethargy initially and then seizures, downward deviated gaze, and episodes of apnea and bradycardia.

An infant with evolving hydrocephalus will commonly present to the primary pediatrician with a history of irritability and vomiting. These complaints can be often confused with infant colic or viral infection, and as such, a physical examination is crucial. Physically, parents may note increasing head size, and a "full head," which refers to the fontanelles being pushed outward due to increasing ICPs. At this point, head imaging is the most useful procedure to diagnose or rule out hydrocephalus. Head imaging is useful because a large head circumference or macrocephaly alone can arise due to other conditions as well, such as benign anatomic macrocephaly, which is a benign variant of skull development and does not require any intervention. (9) Other considerations include benign subdural collections, such as hygromas. (18) This distinction is important to recognize during the evaluation of macrocephaly because it can often be misconstrued as traumatic in nature, which is not the case. (19)

On imaging of hydrocephalus in infants, enlarged ventricular systems can often be appreciated. In neonates, ventriculomegaly can often be seen satisfactorily by head ultrasonography. (20) Head ultrasonography is typically the preferred initial imaging modality given the lack of radiation exposure and the ease by which this can be completed at the bedside. Further information can be gained from performing magnetic resonance imaging without contrast, particularly by using the cine flow sequence that can identify more subtle flow-related abnormalities that help diagnose certain hydrocephalus types. (21)

PATHOPHYSIOLOGY

CSF is a clear, acellular liquid derived from plasma and circulates in the subarachnoid space of the central nervous system, covering the brain and spine. CSF plays a crucial role in protecting, nourishing, and removing waste from the central nervous system, as well as providing a degree of mechanical shock absorbance from accelerative vectors. (22)

Total CSF volume in neonates and infants are within the ranges of 20 to 40 mL and 60 to 90 mL, respectively, with mean production rates approximately 1 mL/h and 10 mL/h, respectively. (23) CSF itself is generated mostly by the choroid plexus, which is made up of cuboidal epithelium enveloping highly vascular connective tissue. (24) These plexi are located in the lateral ventricles of the brain, which are structured collections of CSF. There exist 2 lateral ventricles that drain CSF through the foramina of Monroe into the third ventricle and then into the fourth ventricle. CSF then circulates from the fourth ventricle into the subarachnoid cisternal spaces and eventually throughout the subarachnoid space of the thecal sac. Ultimately, most CSF returns to the subarachnoid spaces cranially to be absorbed by arachnoid granulations that channel the CSF into major draining sinuses. (25)

Known as the bulk flow model, the most traditional view on the cause of hydrocephalus in infants involves some element of obstruction anywhere from the origins of CSF production to the final point of absorption into the systemic system. (26) This obstruction results in abnormal accumulation of CSF in the brain, leading to hydrocephalus. If the obstruction exists in the ventricular spaces, most commonly in the form of a lesion or a narrowing in the path of CSF circulation, this type of hydrocephalus is referred to as *obstructive* or *noncommunicating* hydrocephalus. If there is obstruction in the subarachnoid spaces involving the CSF circulation and absorption sites, this type of hydrocephalus is referred to as *nonobstructive* or *communicating* hydrocephalus.

ACQUIRED FORMS OF HYDROCEPHALUS

Posthemorrhagic Hydrocephalus

One of the most common causes of acquired infant hydrocephalus is intraventricular hemorrhage (IVH) of prematurity. (27) Estimates suggest that among all infants born prematurely, up to 40% will present with IVH at birth, which can increase to as high as 50% in neonates weighing 1,000 g and less. (27)(28) The pathophysiology behind hemorrhage development is believed to involve immaturity or dysfunctional development of the germinal matrix in the ventricles; this matrix is a highly vascular component beneath the ventricle lining from which cells migrate during development. (29) The immature matrix is more prone to rupture at the time of premature birth, leading to blood products accumulating in the ventricular spaces (Fig 1).

With these blood products layering in the ventricle, 1 significant sequela of this IVH associated with prematurity is the subsequent occurrence of posthemorrhagic hydrocephalus (PHH). PHH is believed to occur in at least 30% of all premature IVH cases, with incidences even higher with poor prenatal care. (30) The mechanism for PHH is suspected to be that intraventricular blood products aggravate the ventricular lining by multiple inflammatory processes, leading to dysfunction of the ventricular ependymal lining, thereby disrupting CSF flow

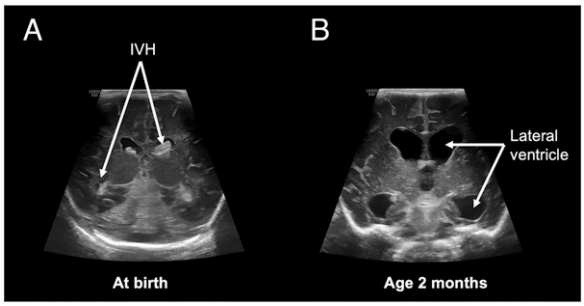

Figure 1. Posthemorrhagic hydrocephalus will often evolve from intraventricular hemorrhage (IVH) often seen secondary to premature birth. Coronal head ultrasonographic image of the lateral ventricles and third ventricle in a 32-week-gestation infant at birth (A) and 2 months later (B). There is IVH seen as hyperechoic lesions in the ventricle at birth. By 1 month, these have largely resolved; however, there is a clear increase in the caliber of all ventricles, resulting in ventriculomegaly and hydrocephalus.

and resulting in hydrocephalus. (4) PHH is typically seen as a consequence of prematurity, and its prophylaxis parallels good management of prematurity in general. Good prenatal care includes administering maternal glucocorticoids and magnesium sulfate in patients at risk for premature delivery, which has been shown to reduce the incidence of IVH. (31)

Postinfectious Hydrocephalus

In the setting of acute or chronic meningitis, it is estimated that 1% to 2% of infants will develop postinfectious hydrocephalus (PIH). This is a worldwide estimate (32); however, there is a strong geographic bias. In North America, for example, the incidence of PIH is effectively negligible today due to high accessibility to prenatal and postnatal infectious disease care. In other countries with lesser quality of care, the incidence of PIH after meningitis may be as high as 30% to 60%. (33)(34) It is believed that the inflammatory response to infection results in the development of fibrous adhesions and scarring along the ventricle wall and arachnoid villi. (35) This development leads to CSF dysfunction and hydrocephalus.

The prevention of PIH is driven by providing adequate infectious disease coverage at the time of diagnosis of meningitis. In the prenatal setting, adequate infectious disease coverage can also include treatment of the mother before birth. Detailed prenatal and maternal history taking is highly recommended if a pediatric specialist suspects PIH.

Obstruction by Tumor

In rare situations, infants born with brain tumors that occur as an intraventricular mass or a mass pressing against the ventricular system can develop obstructive hydrocephalus (Fig 2). Infant brain tumors are indeed rare; of all pediatric brain tumors, they make up less than 10% of cases and are most commonly detected within the first months after birth secondary to altered mental status and physical examination findings related to hydrocephalus. (36)(37) These physical findings commonly occur with a posterior fossa tumor, with pilocytic astrocytoma or ependymoma being the common pathologies in older children. (38) However, the most common pathology in infants is a teratoma, which composes up to one-half of all infant cases. The next most common tumors are astrocytic gliomas and choroid plexus tumors. (39)(40) Although more common in older toddlers and children, medulloblastomas and atypical teratoid/rhabdoid tumors are not typically seen in the infant age group.

Specific to this acquired type of hydrocephalus, an early prenatal sign of mass effect causing obstruction will be polyhydramnios on prenatal ultrasonography. In this case, polyhydramnios is due to poor swallowing ability in utero because of a compressive mass effect on the hypothalamus and resulting neurologic dysfunction. (41) Definitive

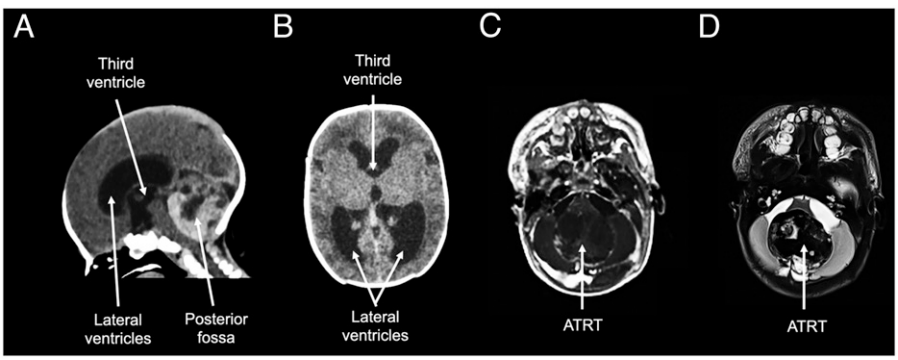

Figure 2. Imaging of a 1-day-old infant presenting with irritability and macrocephaly. Sagittal computed tomographic scan (A) shows a posterior fossa lesion with accompanying enlarged ventricles, which are confirmed on the axial view (B). Then, a T1-weighted magnetic resonance image, axial view with contrast (C), shows a non–contrast-enhancing heterogenous lesion in the posterior fossa causing obstruction by mass effect, which is then reconfirmed on axial T2-weighted imaging (D). Such findings are compatible with obstructive hydrocephalus, which in this case required surgical resection for definitive treatment. Final pathology was atypical teratoid/rhabdoid tumor (ATRT).

management of this hydrocephalus type is surgical resection to relieve the obstruction.

CONGENITAL FORMS OF HYDROCEPHALUS

Myelomeningocele

Myelomeningocele (MMC) is a severe form of spina bifida, which represents a neural tube defect in which the neural tube that forms the spinal column does not completely close. Specifically in MMC, there is a defect in the vertebral arches, resulting in protrusion (*-cele*) of both spinal cord (*myelo-*) and meninges (*meningo-*) to the skin. This typically occurs at the lumbar level in infants, (42) with incidence estimates ranging from 20 to 500 per 100,000 live births worldwide. (43)(44)(45) This is highly geographically dependent, with sub-Saharan Africa and North America known to demonstrate the highest and lowest age-standardized rates of incidence, respectively. (46) These differences are partly due to the disparities in global efforts to improve folate diet supplementation, which has proved to drastically minimize the incidence of MMC. (47)

Most patients with MMC will present with ventriculomegaly and hydrocephalus, either prenatally or postnatally. (48) The defect itself and its associated ventriculomegaly can often be seen on prenatal imaging (Fig 3A). It is thought that disruption to the normal developing CSF circulation due to the open MMC defect in utero disrupts the normal hydrodynamics and pressures of the CSF drainage pathways, resulting in abnormal CSF accumulation by the time of birth. (49)

Currently, both prenatal (in utero) and postnatal repair are options to treat MMC depending on time of detection and resources available. There are advantages to both the prenatal and postnatal approaches. With respect to prenatal repair, the rates of both hindbrain herniation and hydrocephalus requiring shunting decrease. (50)(51) Conversely, with this approach the rates of preterm delivery, closure dehiscence, and abscess formation are higher. In addition, prenatal repair requires advanced maternal and cardiac monitoring and, as such, is not readily available or performable in all countries. Most centers around the world that can perform these repairs are located in high-income countries. (52) With respect to postnatal repair, the standard of care is open surgical closure of the placode, followed by closure of the dura, muscle, fascia, and skin (Fig 3B). (47) Many efforts continue on a global stage to improve the quality of prenatal care and education surrounding maternal folate supplementation to prevent MMC and, indirectly, its associated hydrocephalus too. (53)

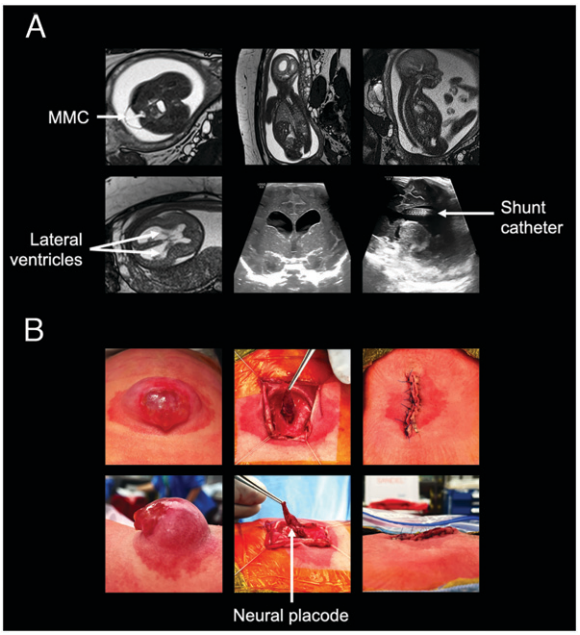

Figure 3. Myelomeningocele (MMC) defects are known to present with hydrocephalus early on. A. Radiologic T2-weighted magnetic resonance images (MRIs) of prenatal MMC at 25 weeks' gestation showing axial (top left), coronal (top middle), and sagittal (top right) views. Then, the ventricles are viewed: the prenatal axial view on T2-weighted MRI (bottom left), the coronal head postnatal ultrasonographic image (bottom middle), and the sagittal head ultrasonographic image after a shunt has been placed for hydrocephalus (bottom right). B. Operative images for postnatal repair at 1 day of age showing anteroposterior (top row) and lateral (bottom row) views of the MMC before surgery (left), during surgery (middle), and after surgery (right). The neural placode is isolated during surgery and internalized. It represents the primitive spinal cord that has not closed properly.

The incidence of hydrocephalus has been shown to greatly range from 50% to 90% of patients with MMC (42)(48)(50)(54) and has been shown to also depend on the timing of MMC repair. A seminal cohort study published in 2013, the Management of Myelomeningocele Study, randomized patients with MMC to compare the safety and efficacy of prenatal repair of MMC with that of standard postnatal repair. (50) The investigators found that at age 12 months, 40% of patients in the prenatal repair group required surgical treatment for hydrocephalus, whereas 82% of patients in the postnatal repair group required surgical treatment for hydrocephalus.

Aqueductal Stenosis

The midline cerebral aqueduct connects the third ventricle to the fourth ventricle, through which the CSF circulates from the third to the fourth ventricle. When there is congenital narrowing or stenosis of this aqueduct, then CSF outflow is limited, causing abnormal accumulation in the proximal ventricles that can lead to hydrocephalus (Fig 4).

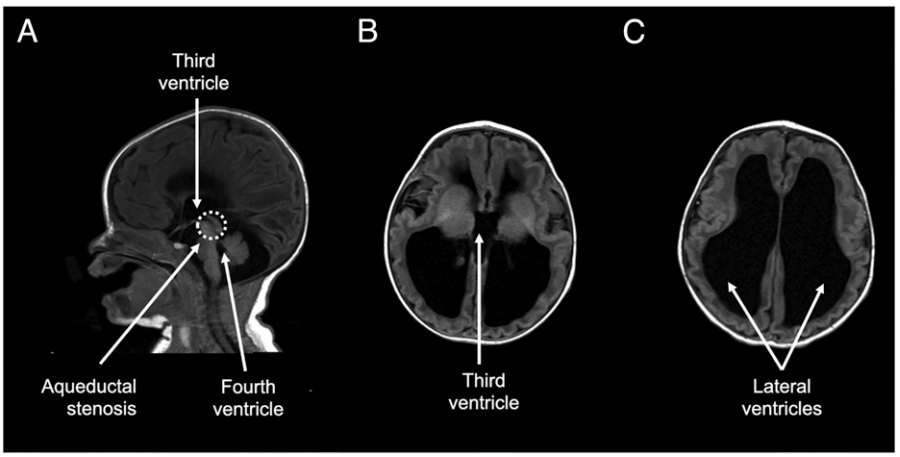

Figure 4. Aqueductal stenosis is a common cause of obstructive congenital hydrocephalus. T1-weighted fluid-attenuated inversion recovery magnetic resonance imaging of a 1-day-old infant. A. Sagittal view shows the stenosis of the central aqueduct (dotted circle) between the third and fourth ventricles. Axial views show increased ventricle size of the third ventricle (B) and lateral ventricles (C). This patient presented with signs of bulging fontanelles.

Aqueductal stenosis has been attributed to more than half the cases of infant hydrocephalus. (55) Most of these cases are thought to be idiopathic, resulting from spontaneous developmental errors in which abnormal folding of the neural plate causes narrowing of the neural tube. (56) However, it is estimated that 10% to 25% of these cases are attributable to an X-linked genetic point mutation in the neural L1 cell adhesion molecule responsible for neuronal migration and axon guidance. (12)(57) More common in boys due to the X-linked nature, mutated L1 cell adhesion molecule results in several structural malformations that obstruct CSF flow, most commonly at the level of the aqueduct. (58) Aqueductal stenosis may be detectable on prenatal imaging; however, its genetic nature is typically a postnatal determination.

Given the anatomic nature of this stenosis, this type of hydrocephalus is more amenable to an endoscopic third ventriculostomy approach than other hydrocephalus types because this condition provides a clear alternative conduit for CSF flow between the third ventricles and the basal cisterns, which lie beyond the fourth ventricle. (59)

Posterior Fossa Malformations

Developmental malformations that occur in the posterior fossa can obstruct the central aqueduct and fourth ventricle and CSF outflow, leading to hydrocephalus. The 2 most common examples of posterior fossa malformations are Dandy-Walker malformation (DWM) and Chiari type I malformation (CIM).

DWM is the most commonly encountered posterior fossa malformation, occurring in approximately 3 in 100,000 live births. (60) DWM is classically defined as a triad of 1) a hypoplastic upward rotated cerebellar vermis with 2) cystic dilatation of the fourth ventricle and 3) an enlarged posterior fossa with upward displacement of nearby structures, including the sinus and tentorium (Fig 5). (61)(62) It is believed that this congenital condition results from abnormal development of the rhombencephalon and foramina of the fourth ventricle, leading to the associated vermian hypoplasia and dilatation of the fourth ventricle, respectively. (63) Collectively, the features of DWM can lead to a mass effect on the nearby central aqueduct and obstruction of CSF flow. It has been estimated that up to 90% of infants with DWM will present with symptoms of hydrocephalus, most presenting in the first 3 months after birth, and most commonly presenting with macrocephaly. (64)(65)

CIM exists within a spectrum of hindbrain herniation malformations described by Hans Chiari in 1891. (66) CIM specifically describes what is now typically seen on imaging as the caudal descent of the cerebellar tonsils of 5 mm or more below the foramen magnum (Fig 6). It is most commonly considered to result from the underdevelopment of the posterior cranial fossa. (67) To date, hydrocephalus is estimated to occur in up to 20% of all patients with CIM, which is likely to manifest due to obstruction of the fourth ventricle with or without concurrent aqueductal stenosis. (68)(69) Estimates suggest that 1% of all pediatric patients will possess a CIM (70); however, only a very small proportion of these patients will be detected to have CIM as infants. (71) It should be noted that the typical surgical treatment for CIM is decompression, which can also relieve the obstruction of the CSF circulation. As such, studies have shown that neurosurgery for pediatric patients with CIM can result in resolution of any concurrent hydrocephalus in approximately

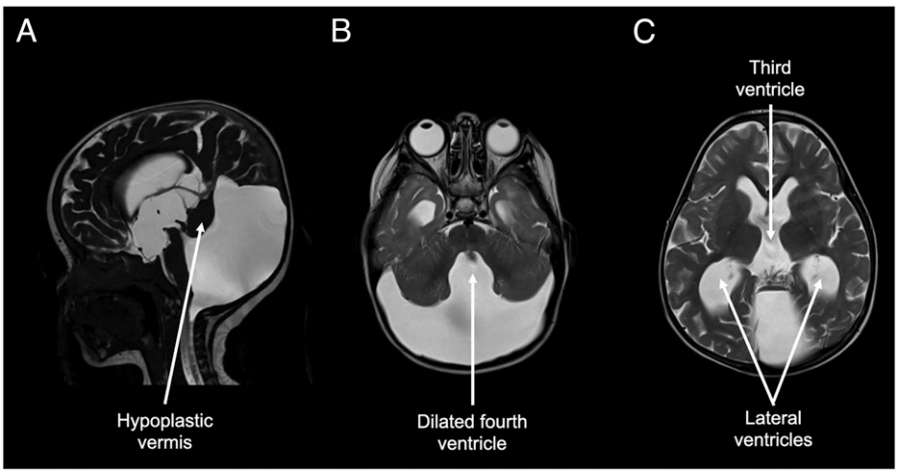

Figure 5. Congenital malformations often take time to manifest clinical symptoms of increased intracranial pressures. T2-weighted magnetic resonance image sequences of an 11-month-old with increasing head circumference demonstrating Dandy Walker malformation with the sagittal view showing hypoplastic vermis (A), the axial view showing a dilated fourth ventricle (B), and the axial view showing an enlarged ventricular system (C).

85% of patients without the need for further dedicated hydrocephalus surgical treatment. (72)

SURGICAL TREATMENT

There are 2 major definitive surgical treatments for infant hydrocephalus: CSF shunting and endoscopic third ventriculostomy (ETV). These surgical treatments are in addition to the primary treatment of any associated conditions already mentioned, such as MMC repair, tumor resection, and CIM decompression. These definitive treatments can be preceded by more temporary measures, such as an external ventricular draining and Ommaya reservoir taps.

Temporary Measures

If a patient presents with severe hydrocephalus and is classically obtunded or severely compromised, an external ventricular drain is a rapid bedside procedure that can be done to relieve CSF from the lateral ventricles. This draining procedure is performed by placing a proximal catheter into the ventricle and connecting the catheter to an external drainage system from which CSF can be collected and drainage controlled. In the neonatal setting, other options to temporize an infant until the infant is an appropriate candidate for definitive surgery are as follows. Because the fontanelle of infants is accessible through the open suture

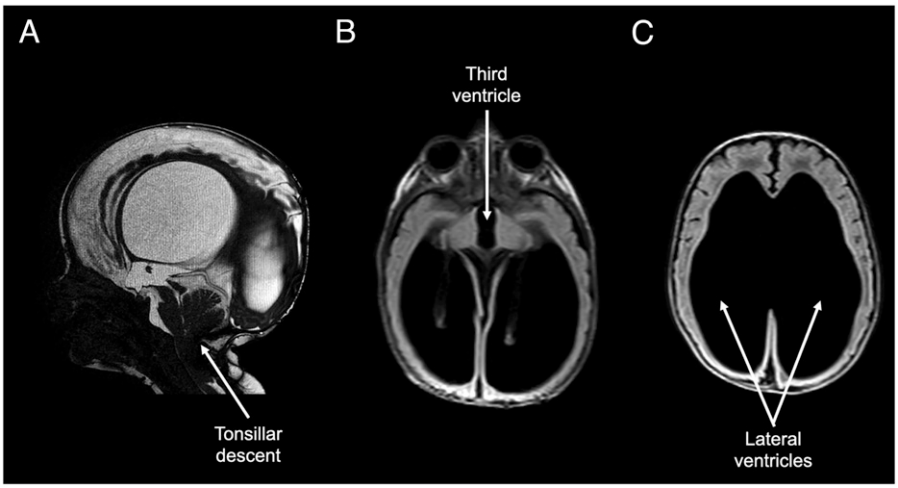

Figure 6. The inferior part of the cerebellum is called the tonsils, and the tonsils can descend downward, creating a Chiari malformation with accompanying hydrocephalus. Magnetic resonance image of a 6-week-old with increasing head circumference demonstrating Chiari type I malformation with a T2-weighted sequence sagittal view showing tonsillar descent below the foramen magnum, a T1-weighted fluid-attenuated inversion recovery (FLAIR) sequence axial view showing a dilated third ventricle (B), and a T1-weighted FLAIR sequence axial view showing enlarged lateral ventricles (C).

lines, a transfontanelle tap can be performed at the bedside. A transfontanelle tap involves piercing a fine needle through the fontanelle to access the lateral ventricles and remove CSF. If continuous CSF removal is required, another option that is more sustainable than repeated transfontanelle taps is placement of an Ommaya reservoir. This surgical procedure involves placing a proximal catheter into the lateral ventricle and connecting the catheter to a subcutaneous reservoir below the skin. This reservoir can be accessed by needle at the bedside repeatedly.

CSF Shunting

Shunting CSF from the lateral ventricles to a distal destination outside the CSF space is the most well-known surgical treatment for hydrocephalus. (3) The shunt setup consists of a proximal catheter placed in the ventricles, a valve to control forward flow and prevent back flow, and a distal catheter placed into a body cavity capable of absorbing the shunted CSF. The most common and most preferred distal destination is the peritoneal cavity (ventriculoperitoneal shunt) due to the peritoneal cavity's accessibility and the volume of CSF it is able to absorb. Contraindications to the peritoneal cavity, such as active peritonitis, scarring, or previous failure, require an alternative destination. In infants, alternative destinations include the right atrium (ventriculoatrial shunt) via the subclavian vein and the pleural cavity (ventriculopleural shunt).

Infants are often eligible for shunting once they reach appropriate weight and are cleared of any surgical contraindications, such as an active infection. This procedure, however, is not without morbidity. In infants alone, irrespective of hydrocephalus type, estimates suggest that in the first year, 30% to 40% of shunts will fail, by 2 years 40% to 50% of shunts will fail, and by 10 years, the average failure rate is 80% to 90%. (73) Unfortunately, the average number of times an infant is expected to undergo shunt revision due to failure is 2.5 times during childhood. Failure can occur due to multiple reasons, including obstruction by brain debris, bacterial infection, and abdominal pseudocysts.

Shunt failure can present with many symptoms similar to that of initial hydrocephalus, but also can present with infectious symptoms, such as vomiting and fever in the case of meningitis. A common concern of neurosurgeons is that unnecessary shunt tapping or sampling increases this real risk of infection, which can lead to early shunt failure. Hence, when there is a concern for failure, a more wholistic initial approach should be taken before tapping or sampling a shunt, including basic laboratory examination, imaging of the shunt system (typically radiographs), and searching for a systemic infection. This initial approach can be most efficiently performed in an emergency department setting.

Shunt independence is a concept that, in the future, patients can have their shunt system removed or ligated (surgically closed). (74) Currently, it remains unclear as to how many infant patients will progress to true independence because cases of recurrent hydrocephalus have been reported decades after apparent successful shunt removal. (75) Nevertheless, in a large series of 300 patients with congenital hydrocephalus, it was deemed that 17% did not require shunting. (75) Currently, there is no strong evidence that can predict shunt independence years after initial surgery, and any attempt to do so is not advised. Rather, continuous and faithful follow-up will prove to be the most useful in managing shunts long-term.

Endoscopic Third Ventriculostomy

More recently, ETV has emerged as a minimally invasive alternative to CSF shunting in treating specific infant hydrocephalus types. This surgery involves making a frontal scalp incision through which an endoscope is passed into the third ventricle and making a hole in the ventricular floor (ventriculostomy) to communicate the third ventricle with the basal cisterns, which then allows CSF to circulate throughout the thecal sac. This approach is theoretically ideal to bypass CSF flow obstruction that is at the level of the central aqueduct and fourth ventricle, such as that seen with aqueductal stenosis and posterior fossa malformations. However, neonates and infants have the highest rate of ETV failure observed. In patients younger than 6 months, the chances of success are at best 50% depending on the type of hydrocephalus. (76) In the case of ETV failure, CSF shunting most commonly becomes the default surgical treatment. Similar to CSF shunting, it is critical for infants treated by ETV to be seen by both their pediatric specialist and surgeon regularly for surveillance purposes.

Because access to the ventricles containing choroid plexus is achieved with the ETV approach, there is also an adjunct procedure that can be performed known as *choroid plexus cauterization* (CPC). The rationale for adding this component to the ETV approach is that the choroid plexus generates most of the CSF and, therefore, its cauterization will stall CSF production and relieve the hydrocephalus burden. (77) In a large infant series involving hydrocephalus, ETV with CPC resulted in favorable outcomes, with failure rates of approximately 50% across acquired and congenital types of hydrocephalus. (78) The indications for the addition of CPC to an ETV approach remain surgeon-dependent for most hydrocephalus types.

Summary

- Infant hydrocephalus is common worldwide, with greater incidences seen in African, Latin American, and Southeast Asian regions. (Based on some research evidence as well as consensus) (10)
- Infant hydrocephalus can prenatally present on imaging and postnatally present with symptomatic enlargement of the head, associated skull features, and raised intracranial pressures. (Based on strong research evidence) (17)
- Two acquired forms of hydrocephalus are post-hemorrhagic and postinfectious hydrocephalus, which are associated with prematurity and meningitis, respectively. (Based on strong research evidence) (27)(30)
- Obstructive forms of hydrocephalus can arise secondary to aqueductal stenosis, brain tumors, and posterior fossa malformations. (Based on strong research evidence) (3)
- Myelomeningoceles are common congenital causes of hydrocephalus, and their timely repair can also address associated hydrocephalus. (Based on strong research evidence) (50)
- The gold standard of hydrocephalus surgical treatment is cerebrospinal fluid shunting, with endoscopic third ventriculostomy as a viable alternative for specific hydrocephalus types. (Based on strong research evidence) (3)

Take the quiz! Scan this QR code to take the quiz, access the references, and view and save images and tables (available on August 1, 2024).

PIR QUIZ

1. A 4-month-old girl is brought to the clinic by her parents for a routine follow-up visit. The patient is the product of a full-term pregnancy and spontaneous vaginal delivery with no perinatal complications. She was seen in follow-up in the clinic at 2 weeks and at 8 weeks of age before today's visit. The parents report that the baby has been feeding well but is occasionally fussy, colicky, and spits up after feedings. On physical examination her height and weight are at the 25th percentile for age and her head circumference is at the 90th percentile. The clinician noted that at her previous 2 visits at 2 and 8 weeks of age, the head circumference was at the 15th and 75th percentiles, respectively. An imaging study shows fluid collections in the subdural space bilaterally. Which one of the following is the most likely diagnosis in this patient?

   A. Benign congenital hygromas.
   B. Communicating hydrocephalus.
   C. Nonaccidental head trauma.
   D. Noncommunicating hydrocephalus.
   E. Normal variant.

2. You are seeing a 4-month-old term baby for a health supervision visit in your office and you notice that his head circumference is greater than the 98th percentile for age. The parents report that, on occasion, the patient exhibits some fussiness and intermittent vomiting, which they attribute to spit-ups. The infant is otherwise feeding well and is meeting all expected milestones. Which one of the following is the most appropriate immediate next step in management?

   A. Computed tomographic scan of the head.
   B. Head circumference at the next health supervision visit.
   C. Magnetic resonance imaging of the head.
   D. No intervention is needed.
   E. Ultrasonography of the head.

3. A 6-month-old infant is seen for the first time in the clinic for follow-up. The baby is a former 24-weeks premature infant with a history of hydrocephalus and ventriculoperitoneal shunting. Which one of the following is the most likely cause of hydrocephalus in this patient?

   A. Aqueductal stenosis.
   B. Brain tumor.
   C. Intraventricular hemorrhage.
   D. Meningitis.
   E. Myelomeningocele.

4. Various types of hydrocephalus can be the result of various causes. In which of the following causes of hydrocephalus is an endoscopic third ventriculostomy approach indicated?

   A. Aqueductal stenosis.
   B. Chiari type 1 malformation.
   C. Communicating hydrocephalus.
   D. Dandy-Walker syndrome.
   E. Noncommunicating hydrocephalus.

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5. A 3-year-old boy with a history of hydrocephalus and a ventriculoperitoneal shunt is brought to the emergency department by his parents with a 1-day history of vomiting. On physical examination he is well-appearing and shows no clinical signs of dehydration. There is no history of fever and no known sick contacts. Which one of the following is the next step in management?

   A. Cerebrospinal fluid analysis from a shunt tap.
   B. Cerebrospinal fluid analysis from a spinal tap.
   C. Order no studies and discharge on oral ondansetron.
   D. Schedule an appointment with neurosurgery for outpatient follow-up in 1 week.
   E. Radiography of the shunt system.