Chapter 6
Hydrocephalus

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Introduction

Hydrocephalus is the abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles (i.e., the fluid filled cavities within the brain) and subarachnoid spaces (i.e., the fluid filled space around the brain). It is often associated with dilatation of the ventricular system and increased intracranial pressure (ICP; i.e., increased pressure within the brain). The incidence of pediatric hydrocephalus as an isolated congenital disorder is approximately 1/1,000 live births. Pediatric hydrocephalus is often associated with numerous other conditions, such as myelomeningocele, tumors and infections. Hydrocephalus is almost always a result of an interruption of CSF flow and is rarely because of increased CSF production. In this chapter, we will discuss the clinical features, diagnosis, and treatment of pediatric hydrocephalus.

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Clinical Features

Signs and symptoms of progressive hydrocephalus depend on age. The following outlines the signs and symptoms of hydrocephalus in premature infants, full-term infants, and older children.

Premature Infants

Hydrocephalus in premature infants is predominantly caused by post-hemorrhagic hydrocephalus (PHH), which occurs due to malabsorption of the CSF within the brain. Infants with PHH may have no symptoms or may exhibit increasing spells of apnea and bradycardia. If ventriculomegaly progresses and ICP increases, the anterior fontanelle becomes convex, tense, and nonpulsatile; and the cranial sutures splay and the scalp veins distend (Table 6.1).

Full-Term Infants

The common causes of hydrocephalus in full-term infants include aqueductal stenosis, Dandy-Walker syndrome, arachnoid cysts, tumors, and cerebral malformations. Symptoms include irritability, vomiting, and drowsiness. Signs include macrocephaly, a convex and full anterior fontanelle, distended scalp veins, cranial suture splaying, frontal bossing, cracked pot sound on percussing over dilated ventricles (positive Macewen's sign), poor head control, and the "setting-sun" sign, in which the eyes are inferiorly deviated (Table 6.1).

Older Children

Hydrocephalus after infancy is usually secondary to trauma or tumors. The predominant symptom is usually a dull and steady headache, which typically occurs upon awakening. It may be associated with lethargy, and often improves after vomiting. The headaches slowly increase in frequency and

Table 6.1 Signs and symptoms of hydrocephalus in children

Premature infants	Full-term infants	Older children
Apnea	Irritability	Headache
Bradycardia (i.e., low heart rate)	Vomiting	Vomiting
Tense fontanelle	Drowsiness	Lethargy
Distended scalp veins	Macrocephaly	Diplopia
Globoid head shape	Distended scalp veins	Papilledema
Rapid head growth	Frontal bossing	Lateral rectus palsy
	Macewen's sign	Hyperreflexia/clonus
	Poor head control	
	Lateral rectus palsy	
	"Setting-sun" sign	

severity over days or weeks. Other common complaints include blurred or double vision, decreased school performance and behavioral disturbances (Table 6.1).

Diagnosis

Hydrocephalus can be diagnosed by cranial ultrasonography in infants with open scalp fontanelles, and by CT and MR imaging, which will demonstrate increased ventricular size, as well as the site of pathological obstruction if present (e.g., tumors that obstruct the ventricles and produce ventriculomegaly).

Treatment

The treatment of hydrocephalus can be divided into non-surgical approaches and surgical approaches, which in turn can be divided into non-shunting or ventricular shunting

procedures. The goals of any successful management of hydrocephalus are: (1) optimal neurological outcome and (2) preservation of cosmesis. The radiographic finding of normal-sized ventricles should not be considered the goal of any therapeutic modality.

Non-surgical Options

There is no non-surgical medical treatment that definitively treats hydrocephalus effectively. Historically, acetazolamide and furosemide have been used to treat hydrocephalus. Although both agents can decrease CSF production for a few days, they do not significantly reduce ventriculomegaly, and can lead to potential side effects such as lethargy, poor feeding, tachypnea, diarrhea, and electrolyte imbalances. While acetazolamide has been used historically to treat premature infants with PHH, recent studies have shown it to be ineffective in avoidance of ventricular shunt placement and to be associated with increased neurological morbidity.

Surgical: Non-shunting Options

Whenever possible, the obstructing lesion that causes the hydrocephalus should be surgically removed. For example, the resection of a tumor that obstruct the ventricles often treats the secondary hydrocephalus. Unfortunately, in most cases of congenital hydrocephalus, the obstructive lesion is not amenable to surgical resection.

For CSF obstruction at or distal to the aqueduct (e.g., tectal plate tumors, acquired aqueductal stenosis, or posterior fossa tumors), a potential surgical treatment is the endoscopic third ventriculostomy. By surgically creating an opening at the floor of the third ventricle, CSF can be diverted without placing a ventricular shunt. Recent studies report a high success rate for endoscopic third ventriculostomies among pediatric patients with hydrocephalus secondary to aqueductal stenosis.

Table 6.2 Common indications for ventricular shunt placement

Congenital hydrocephalus
Persistent post-hemorrhagic hydrocephalus
Hydrocephalus associated with myelomeningocele
Hydrocephalus associated with Dandy-Walker cyst
Hydrocephalus associated with arachnoid cyst
Hydrocephalus associated with posterior fossa tumor

Surgical: Ventricular Shunts

The following outlines the components of a ventricular shunt, and the potential shunt complications. In addition, Table 6.2 lists some of the common indications for placement of a ventricular shunt.

Components

CSF shunts are silicone rubber tubes that divert CSF from the ventricles to other body cavities where normal physiologic processes can absorb the CSF. Shunts typically have three components: a proximal (ventricular) catheter, a one-way valve that permits CSF flow out of the ventricular system, and a distal catheter that diverts the CSF to its eventual destination (i.e., peritoneal, atrium or pleural space). The most common type of ventricular shunt in use today is the ventricular to peritoneal shunt (i.e., shunt tubing from the ventricles to the peritoneal cavity which is the potential space around the organs in the abdomen).

Valves come in a variety of different pressure and flow settings depending on the manufacturer. However, a recent advance in shunt valve technology has been the introduction

of programmable valves, which allows one to adjust the opening pressure settings of the implanted shunt valve without the need to subject the child to an additional surgical procedure to change valves.

Shunt Complications

Shunt complications and failure remain a significant problem in treating hydrocephalus. The goal in treatment of hydrocephalus with a ventricular shunt is to decrease intracranial pressure and associated brain damage and simultaneously prevent complications associated with the ventricular shunting procedure. Shunt complications fall into three major categories: (1) mechanical failure of the device, (2) functional failure because of too much or too little flow of CSF, and (3) infection of the CSF or the shunt device. However, the two most common complications are infection and obstruction, which are further explained below.

Shunt Infection

Despite the numerous measures used to decrease the risk of infection, in general, approximately 1–15 % of all shunting procedures are complicated by infection. Approximately three-quarters of all shunt infections become evident within 1 month of placement. Nearly 90 % of all shunt infections are recognized within 1 year of the last shunt manipulation, as it is believed that most bacteria are introduced at the time of surgery.

The most effective and widely used treatment of a shunt infection is to remove the infected shunt hardware and place an external ventriculostomy drain (i.e., placement of a tube within the ventricles and connecting it to an external sterile collection bag outside one's body). The patient is then treated with the appropriate intravenous antibiotics based on culture and sensitivity results. When the infection is cleared, a new ventricular shunt system is implanted, and the external ventriculostomy is removed.

Shunt Obstruction

Shunt obstruction is another common complication. Shunt devices are to be viewed as mechanical devices that can become obstructed or malfunction anywhere in their course and anytime during their lifetime. The most common scenarios occur weeks, months, or years after insertion, when choroid plexus or debris has occluded the proximal ventricular catheter tip. Another common shunt malfunction scenario is the child who has obstructed his distal catheter or has outgrown his peritoneal catheter, and presents with an obstruction after the distal catheter tip has slipped out of the peritoneal cavity. In addition, shunt valves can malfunction, and shunt tubing can break, disconnect or dislodge from its previous location.

Common symptoms of shunt obstruction depend on the age of the child. A child with a shunt malfunction often presents with signs and symptoms of increased brain pressure. Infants with a shunt malfunction usually present with irritability, poor feeding, increased head circumference, and/or inappropriate sleepiness. Children with a shunt malfunction usually present with headache, irritability, lethargy, nausea, and/or vomiting. However, it is important to inquire if the signs and symptoms that the child is presenting with are the same as those during a shunt malfunction in the past. The child can present with waxing and waning symptoms, or can alternatively present with a progressively worsening picture that does not improve until the shunt is revised. A child complaining of pain with a clinical picture consistent with shunt obstruction should not be given narcotics because of possible respiratory depression or arrest.

When a shunt malfunction is suspected, head imaging studies should be obtained after a careful history and physical examination. A head CT/MRI and anteroposterior and lateral skull, chest, and abdominal x-rays, are obtained to evaluate for increased ventricular size and shunt hardware continuity, respectively. Children who are diagnosed with a shunt malfunction are taken promptly to the operating room for shunt revision.

Summary

Signs and symptoms of progressive hydrocephalus depend on age. Symptomatic ventricular shunt malfunction should be evaluated, recognized and treated promptly to avoid undue morbidity. Ventricular shunt infection currently occurs in 1–15 % of children who have shunts placed or revised, and the majority of infections are detected within the first 1–6 months after a shunt procedure. The prognosis of pediatric hydrocephalus is dependent primarily on the underlying brain morphology (i.e., a child with relatively normal brain organization has a better outcome than a child with abnormal morphology).

Suggested References

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