

# About Hydrocephalus

*A Book for Families*



# FOREWORD

This booklet is written for parents of children with hydrocephalus and people with hydrocephalus in the hope that the information will give you a better understanding of the condition and how it can be managed. Although this book was originally written for parents, it contains basic information about hydrocephalus that is valuable to everyone—parents of children with hydrocephalus, families and individuals. In recent years, there have been remarkable advances in the treatment of hydrocephalus. With early detection and effective treatment, the outlook for children with hydrocephalus is promising. Many people with hydrocephalus lead normal lives with few limitations. Research and experience show that children with hydrocephalus have excellent opportunities to attain their full potential through programs that stimulate their development.

Hydrocephalus affects about one in every 500 to 1,000 children born. It is caused by a wide variety of medical problems, and the circumstances of each child's condition are unique. Each year, U.S. children with hydrocephalus alone generate 0.6% of hospital admissions, 1.8% of days, and 3.1% of charges. You will probably have many questions concerning your child's particular problems that are beyond the scope of this booklet, but you will find that your knowledge about the condition will increase steadily as time passes.

A number of experienced medical professionals and families of children with hydrocephalus participated in making this booklet. They have dealt with many of the issues facing you today. We hope that their experiences, knowledge and perceptions will help you discover your own path to understanding and coping with hydrocephalus.

*We wish to give special thanks to the people  
with hydrocephalus and their families  
who participated in the making of this booklet.*

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# INTRODUCTION

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*“When we were first given the shocking news about our child, it was hard for us to look beyond and realize that there was a lot of support out there, and that we would cope somehow.”*

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Hydrocephalus is an abnormal accumulation of fluid—cerebrospinal fluid, or CSF—within cavities called ventricles inside the brain. Hydrocephalus is commonly treated by a surgical procedure, performed by a neurosurgeon, in which a tube called a shunt is placed into the child’s body. The shunt channels the flow of fluid away from the brain or spinal cord into another part of the body, where the fluid can be absorbed and transported to the bloodstream. This is a relatively common operation—in fact, an average of 40,000 shunt operations are performed each year in this country. Using projections that incorporate inpatient and outpatient encounters we estimate the number of people in the U.S. who are treated for hydrocephalus each year ranges between 120,000 and 150,000. In most cases, the procedure successfully controls hydrocephalus, but, unlike many surgical procedures that can cure a condition, the placement of a shunt does not cure hydrocephalus. Except in rare cases, hydrocephalus is a lifelong condition. And as with any long-term medical condition, complications can occur to which parents must be alert. The changes that signal a possible complication require your understanding, because a complication left undiagnosed and untreated could cause severe brain damage, or threaten the life of your child.

In the following pages, we explain the nature and causes of hydrocephalus, its diagnosis, treatment protocols and follow-up care. We also provide important information about shunt malfunctions and infections, including a quick-reference table on page 29.

# ANATOMY AND PHYSIOLOGY

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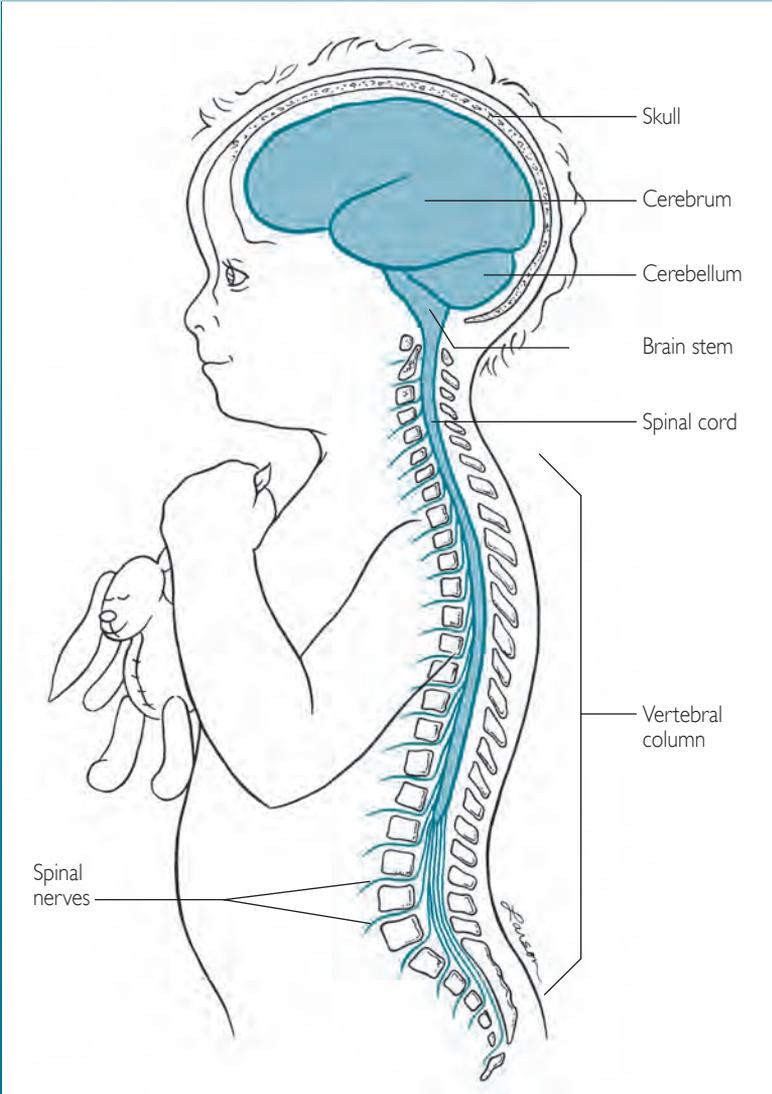
## Brain, Spinal Cord and Their Protective Coverings

The brain and spinal cord form the central nervous system. These vital structures are surrounded and protected by the bones of the skull and the vertebral column, as shown in Fig. 1. The bones of the skull are often referred to as the cranium. In infants, the skull is actually composed of separate bones, and an infant's soft spot (anterior fontanel) is an area where four skull bones nearly come together. The places where the bones meet and grow are called sutures. The vertebral column, which encases the entire spinal cord, is composed of bones called vertebrae. The spinal column begins at the base of the skull and extends all the way down to the tailbone.

The brain's major components are the cerebrum, the cerebellum and the brain stem. The cerebrum is the central processing area for the body's incoming and outgoing messages. It is also the area responsible for speech, thought and memory. The cerebellum primarily helps coordinate our body movements. The brain stem controls basic functions like heart rate, breathing and blood pressure. The spinal cord extends from the brain stem, through a very large opening (the foramen magnum) in the base of the skull, and down the spine. At the level of each vertebra in the spine, nerve fibers arise from the spinal cord and emerge through openings between the vertebrae. These are the spinal nerves, which carry messages to and from various regions of our bodies.

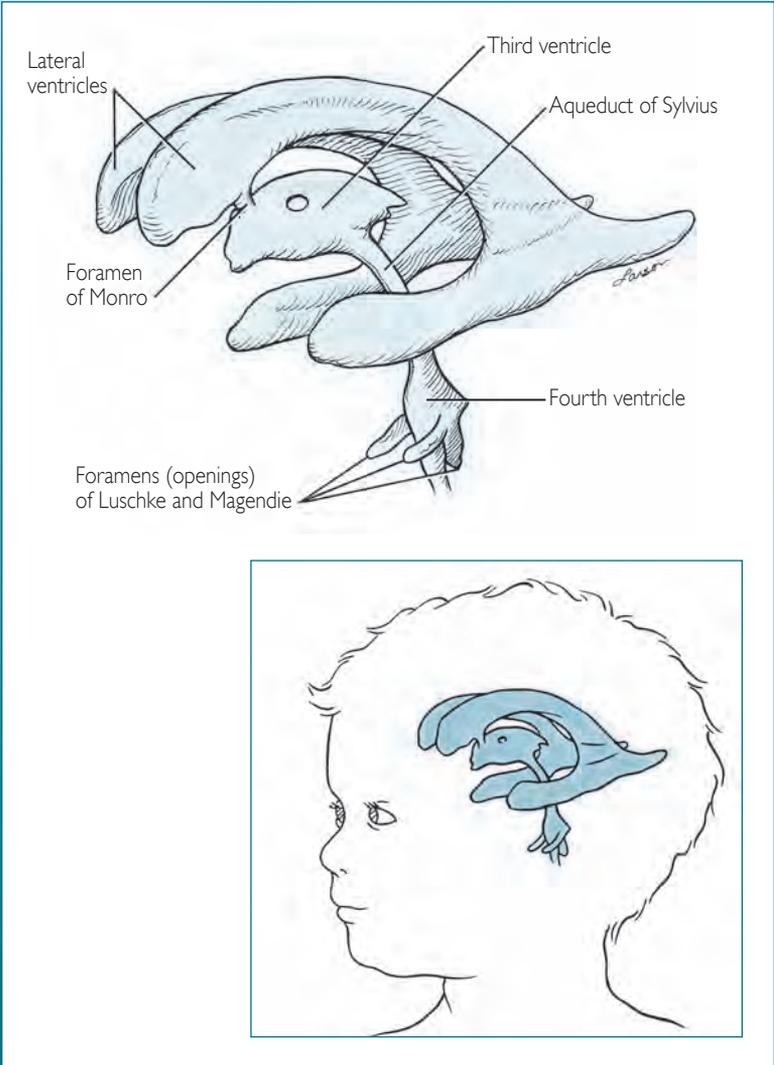
Lying between the brain and skull are three other protective coverings. These are the membranes (meninges), which completely surround the brain and spinal cord. An important fluid—the cerebrospinal fluid (CSF)—flows in a space between these membranes that is called the subarachnoid space. CSF is essentially salt water, and it is in con-

**Fig. I** The Brain and Spinal Cord and the Protective Bones Covering Them



**Fig. 2 Ventricular System**

The ventricles are interconnected by narrow passageways.



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*“..thank you all for a very, very special conference...for taking the time to make the conference a place where I can feel that he matters. Your efforts were felt and I believe that the association is moving in a direction that will really, serve our community, our tribe, our family.”*



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stant circulation and serves several important functions. The brain floats in CSF

## Ventricles

CSF is produced within the cavities of the brain that are called ventricles. Fig. 2 is a drawing of the ventricles. As you look at the drawing, imagine the ventricles as chambers filled with fluid. There are four in all: the two lateral ventricles, the third ventricle and the fourth ventricle. As you can see, the ventricles are interconnected by narrow passageways. Your neurosurgeon can learn valuable information about your child's condition by closely monitoring the size and shape of these ventricles.

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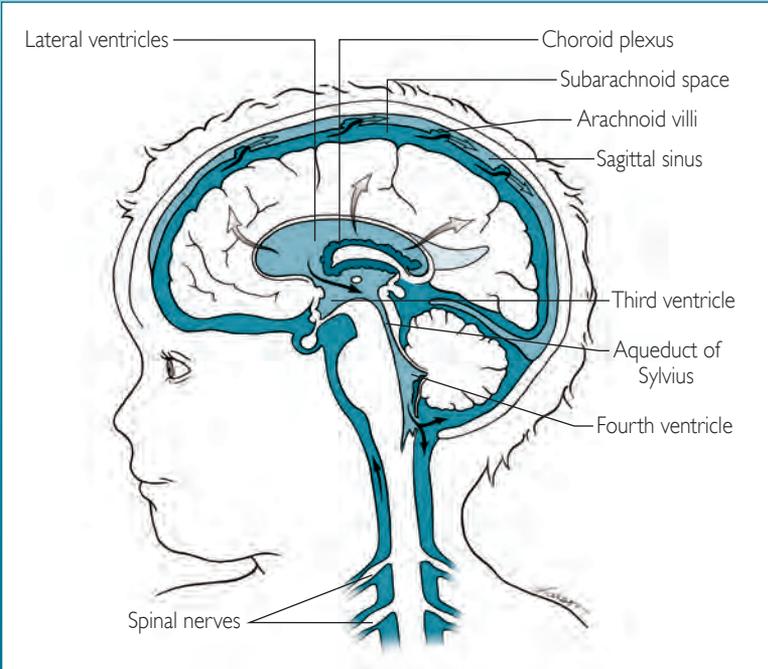
## Cerebrospinal Fluid Circulation and Absorption

CSF is formed within the ventricles by small, delicate tufts of specialized tissue called the choroid plexus. The solid arrows in the drawing on Fig.3 show the major pathway of CSF flow. Beginning in the lateral ventricles, CSF flows through two passageways into the third ventricle. From the third ventricle it flows down a long, narrow passageway (the aqueduct of Sylvius) into the fourth ventricle. From the fourth ventricle it passes through three small openings (foramina) into the subarachnoid space surrounding the brain and spinal cord. CSF is absorbed through blood vessels over the surface of the brain back into the bloodstream. Some absorption also occurs through the lymphatic system. Once in the bloodstream, it is carried away and filtered by our kidneys and liver in the same way as are our other body fluids.

The ventricular system is the major pathway for the flow of CSF. CSF also flows directly from the ventricles into the brain tissue surrounding them. This is shown by the broken arrows. Here the CSF passes through the spaces between the cells to where it eventually enters the subarachnoid space. It is believed that the brain tissue does not absorb any CSF, but simply provides another pathway for the fluid moving to the subarachnoid space. Some small amounts of CSF are also absorbed into lymphatic channels along the membranes covering the nerves (nerve sheaths) as they leave the brain stem and spinal cord.

**Fig. 3 Cerebrospinal fluid (CSF) Circulatory Pathway**

The drawing shows a view of the center of the brain. The solid arrows show the major pathway of CSF flow. The broken arrows show additional pathways.



## ABOUT HYDROCEPHALUS

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*“Since Tess was born with hydrocephalus, we have learned to live each day to its fullest and to put our faith in modern medicine and technology.”*

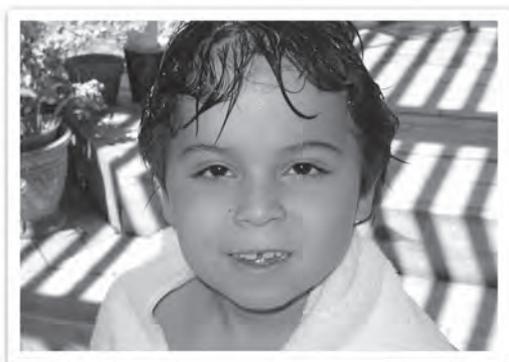
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Our bodies produce approximately a pint (500 ml) of CSF daily, continuously replacing CSF as it is absorbed. Under normal conditions there is a delicate balance between the amount of CSF that is produced and the rate at which it is absorbed. Hydrocephalus occurs when this balance is disrupted. Although there are many factors that can disrupt this balance, the most common is a blockage, or obstruction, somewhere along the circulatory pathway of CSF. The obstruction may develop from a variety of causes, such as brain tumors, cysts, scarring and infection. Specific causes will be discussed more fully in a later section.

Because CSF is produced continuously, when it is blocked it will begin to accumulate upstream from the site of the obstruction, much like a river swells behind a dam. Eventually, as the amount of fluid

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*Lucas is 8 years old. He was 1 year old when he was diagnosed with hydrocephalus, but the cause is not known. Lucas is very active, has many friends, loves sports and is teaching himself to play piano.*



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accumulates, it causes the ventricles to enlarge and pressure to increase inside the head. This condition is known as hydrocephalus.

Obstruction of the CSF pathway often occurs within the ventricles. Although it can occur anywhere in the ventricular system, the site of blockage usually lies either within the narrow passageways connecting the ventricles or where the CSF exits the fourth ventricle into the subarachnoid space. For example, because of its long, narrow structure, the aqueduct of Sylvius is especially vulnerable to becoming narrowed or obstructed, so that it blocks the flow of CSF. Likewise, when the small openings of the fourth ventricle fail to develop, or develop improperly, they also may obstruct the flow of CSF. Hydrocephalus of this kind is called ***noncommunicating hydrocephalus*** because the ventricles no longer provide free passage of CSF through them into the subarachnoid space.

In some cases of hydrocephalus, CSF flows unrestricted through the ventricles, but once it reaches the subarachnoid space its flow is impeded as it passes over the surfaces of the brain. In other cases, the absorptive sites (arachnoid villi) are blocked. Because the ventricles remain open and communicate with each other, this type of hydrocephalus is called ***communicating hydrocephalus***.

## Signs and Symptoms of Hydrocephalus

In an infant, the most obvious sign of hydrocephalus is an abnormal enlargement of the baby's head. The soft spot (fontanel) may be tense and bulging. The scalp may appear thin and glistening, and the scalp veins may appear to have unnatural fullness (prominence) as well. When you feel your baby's head along the suture lines, you may find that the bones are separated. Symptoms to watch for are vomiting, sleepiness, irritability and downward deviation of the baby's eyes (the sun-setting sign).

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*"I am in awe as to how one so young can endure so much yet radiate such warmth. Calina's energy and love for life is contagious. She is an angel." — Cindia*

*Calina was diagnosed with congenital hydrocephalus at 6 weeks old. A ventriculoperitoneal shunt was placed within a few hours after the diagnosis. Calina has had two revisions. Calina is an 8-year-old girl in second grade. She enjoys reading, writing, and art and loves entertaining others.*



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Toddlers whose sutures have not yet closed also show the signs of head enlargement. Older toddlers and children, once their sutures have closed, will show other symptoms of raised intracranial pressure (ICP) caused by their enlarged ventricles. Often these symptoms include headache, nausea, vomiting and sometimes blurred or double vision. The child might have problems with balance, delayed development in such areas as walking or talking, or poor coordination. As with infants, a child may be more irritable or tired than normal. The child may show a change in personality or be unable to concentrate or remember things, and their school performance may decline. Older children may have difficulty waking up and staying awake. While at times the symptoms are very noticeable, other times they can be very subtle and progress so slowly that only in retrospect are they appreciated.

# CAUSES

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A variety of medical problems can cause hydrocephalus. In many children the problem is there at birth—this kind of hydrocephalus is referred to as **congenital**. Most cases of congenital hydrocephalus are thought to be caused by a complex interaction of genetic and environmental factors. Hydrocephalus that develops later in life in some children, and even in adults, but is caused by a condition that existed at birth, is still considered a form of congenital hydrocephalus. When hydrocephalus develops after birth and is caused by a factor such as head injury, meningitis or a brain tumor, it is termed **acquired hydrocephalus**. Parents must not blame themselves for their child's hydrocephalus. In almost all cases the circumstances contributing to a child's condition are beyond the parent's control.

## Aqueductal Obstruction (Stenosis)

The most common cause of congenital hydrocephalus is obstruction of the cerebral aqueduct—the long, narrow passageway between the third and fourth ventricle. Aqueductal obstruction may result from

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*Solan was diagnosed with hydrocephalus and aqueductal stenosis at 11 months. Solan is now 5 years old. He loves music, particularly the guitar, the didgeridoo, and Native American music. He loves the outdoors, flying kites, school, and painting.*



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*“Having a child with a life-threatening health problem can be a very lonely experience. Finding an informed community has turned loneliness and fear into an opportunity for sharing and personal growth.”*

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narrowing or blockage of the aqueduct, or may be caused by infection, hemorrhage or a tumor. Fluid accumulates upstream from the obstruction, producing hydrocephalus.

### Neural Tube Defects or Myelomeningocele

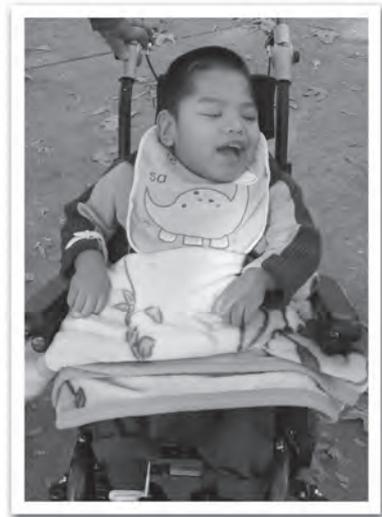
Spina bifida, meaning “open spine,” actually refers to the condition in which the structures (vertebrae, muscles, ligaments, etc.) supporting and protecting the spinal cord are impaired, not the spinal cord itself. Although commonly used, the term spina bifida is better replaced by the term **neural tube defect**, or NTD. A myelomeningocele is an open NTD wherein the spinal cord is exposed at birth and is often lacking CSF. This form of NTD is associated with widespread abnormalities of the central nervous system, including the Chiari II malformation and hydrocephalus that occur in 90 percent of NTDs. In the Chiari II malformation, part of the cerebellum and the fourth ventricle extend downward through the opening at the base of the skull, blocking the flow out of the fourth ventricle and therefore producing hydrocephalus.

### Intraventricular Hemorrhage

Intraventricular hemorrhage is an acquired form of hydrocephalus and most frequently affects premature newborns. It occurs when small blood vessels lying alongside the ventricular lining rupture. Blood may block or scar the ventricles or may plug the arachnoid villi, the sites of CSF absorption along the sagittal sinus. Less frequently, intraventricular hemorrhage may result from a malformation of blood vessels

*“Looking at William now, we feel great pride in how far William has come. His smile and joy for life are a testament to our faith in his abilities. William has been, and continues to be, an inspiration to his family, friends, and even strangers.” — Jennifer*

*William was born at 27 weeks' gestation. He suffered a bleed in his brain (grade 4 intraventricular hemorrhage) that caused hydrocephalus. At 4 months old, he was diagnosed with cerebral palsy (spastic quadriplegia type). William is now a 4-year-old in preschool. He loves playing on the computer and enjoys dancing and music. Most of all, he enjoys long walks outside in his wheelchair.*



within the brain, from a tumor lying near the ventricles or from injury to the head.

## Meningitis

Meningitis is an inflammation of the membranes (meninges) of the brain and spinal cord. It may be caused by bacterial infections or, less frequently, viral infections, which can scar the delicate membranes that line the CSF pathway. Hydrocephalus may develop following meningitis if this scarring restricts or obstructs the flow of CSF as it passes

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*“Networking is a vital thing. A lot of this is up to the parents. Parents have to start reaching out.”*

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through the narrow passageways of the ventricles or as it passes over the surfaces of the brain in the subarachnoid space.

### Head Trauma

A head injury can damage the brain’s tissues, nerves or blood vessels. Blood from these ruptured vessels may enter the CSF pathways. Because this blood causes inflammation, there may be scarring of the meninges, or blood cells may block the CSF absorptive sites. When this occurs, the CSF flow becomes restricted and hydrocephalus develops.

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*Jessica (right), enjoying herself with her sister Erin (left) at the 10<sup>th</sup> National Conference on Hydrocephalus in Park City, Utah. Jessica was diagnosed with congenital hydrocephalus at one week old. She is currently a junior at the University of California, Davis Campus; and is trying to live as normal a life as possible.*



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*“It was a real crisis for me. It was a grieving process. I was grieving for my perfect child, and had to let that vision go.”*

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## Tumors

In children, brain tumors most commonly occur in the back of the brain (posterior fossa). As a tumor grows it may fill or compress the fourth ventricle, blocking the flow of spinal fluid. In other areas of the brain a tumor may similarly block or compress the ventricular system, causing hydrocephalus.

## Arachnoid Cysts

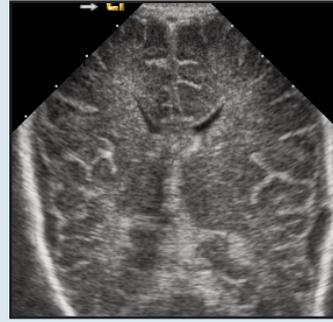
Arachnoid cysts are congenital in origin and may occur anywhere in the brain. In children, they are often located in the back of the brain and in the region of the third ventricle. They are CSF-filled cysts that are lined with the arachnoid membrane (one of the three meningeal coverings). Some arachnoid cysts are self-contained, while others may be connected by a passageway with the ventricles or subarachnoid space. The entrapped fluid may block the CSF pathways, producing hydrocephalus.

## Dandy-Walker Syndrome

In the Dandy-Walker syndrome, the fourth ventricle is enlarged because of partial or complete closure of its outlets. In addition, a portion of the cerebellum fails to develop. The Dandy-Walker syndrome can be associated with abnormal, or a lack of, development of other parts of the brain as well. Obstruction at the aqueduct may also occur. In some instances, two shunts are placed in the child's ventricles—one in the lateral ventricle and another in the fourth ventricle—to manage the hydrocephalus.

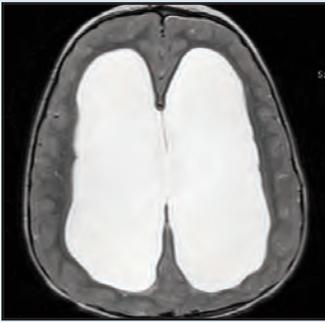


**Enlarged ventricles**

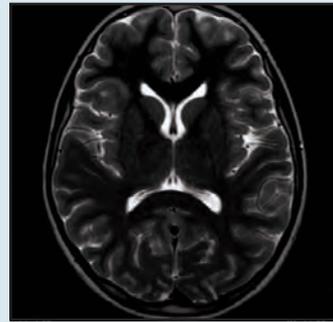


**Normal ventricles**

Ultrasounds showing the ventricles as viewed from the top of the head.



**Enlarged ventricles**

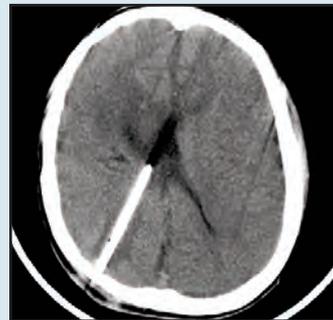


**Normal ventricles**

MRI scans showing the ventricles from the top view.



**Enlarged ventricles**



**Ventricles after shunt placement**

CT scans showing the ventricles as viewed from the top of the head.

# DIAGNOSTIC TESTS

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## Ultrasonography

Ultrasonography is a medical technique that uses high-frequency sound waves to outline structures within the head. It takes little time to perform and is a simple, painless procedure. By the passing of sound waves through the open fontanel of infants, good pictures (images) of the ventricles can be obtained to diagnose and follow the course of hydrocephalus. Because the skull blocks sound waves, ultrasonography cannot be used in an older child, once the fontanel is closed, unless there is a skull defect (a hole in the skull) through which the sound waves may pass.

## Computed Tomography (CT Scans)

CT scanning is a safe, reliable and painless procedure for diagnosing and assisting in the management of hydrocephalus. It is a sophisticated technique in which an x-ray beam is passed through a patient's body and pictures of the internal structures, in this case the brain, are made by the computer.

## Magnetic Resonance Imaging (MRI)

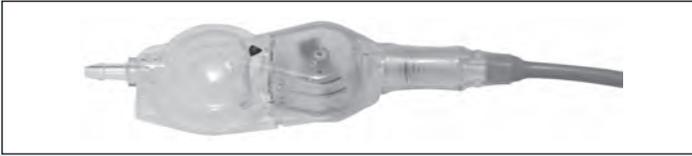
Like the CT scan, MRI is a diagnostic technique that produces images of the brain—but unlike CT scanning, MRI does not use x-rays. Instead, MRI uses radio signals and a very powerful magnet to scan the patient's body, and the signals are then formed into pictures by a computer. MRI is a painless procedure and has no known side effects. There are two types of MRI scans: The Single Shot Fast Spin Echo, which takes about three minutes and rarely requires sedation, is used to assess ventricular size. The full MRI, which takes 30 to 60 minutes and may require sedation, shows more minute details. Before the longer scans are performed, small children are given a sedative to minimize movement that would cause blurring of the images.

## EXAMPLES OF SHUNT VALVES



### **proGAV Adjustable Gravitational Valve**

Aesculap, Inc



### **CODMAN® CERTAS™ Programmable Valve**

Codman & Shurtleff, Inc., a Johnson and Johnson Company



### **Diamond Valve**

Phoenix Neuro



### **Polaris Adjustable Valve**

Sophysa



### **OSV II® Flow Regulating Valve**

Integra Neurosurgery



### **Strata Adjustable Valve**

Medtronic Neurosurgery

# TREATMENT

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Today, one of the best and most effective treatments for hydrocephalus is a surgical procedure in which a flexible tube called a shunt is placed into the child's CSF system. Recent medical and technological advances have led to a new and growing interest in another treatment, endoscopic third ventriculostomy (ETV), which is discussed on page 30.

## Shunting

### *Shunt Systems*

The shunt diverts the flow of CSF from the ventricles into another region of the body, most often the abdominal cavity or a chamber of the heart called the atrium. The shunt tube is about  $\frac{1}{8}$  inch in diameter and is made of a soft and pliable plastic (usually Silastic) that is well tolerated by our body tissues. Shunt systems come in a variety of models but have similar functional components. Catheters (tubing) and a flow-control mechanism (one-way valve) are components common to all shunts.

The parts of a shunt are named according to where they are placed in the body. The portion of the tube that is inserted into the ventricles is called the ventricular catheter. The peritoneal catheter is the portion of the tube that passes the CSF into the abdomen (peritoneal cavity). If the tube is placed into the right atrium of the heart, it is called the atrial catheter. A valve regulates the pressure of the CSF flow and prevents the backward flow of spinal fluid toward the ventricles. There are a number of different shunt systems currently available, examples of which are shown on the opposite page.

Most shunt systems have an access area—usually referred to as a reservoir—that allows easy entrance into the system with a fine-gauge needle in order to obtain CSF. This procedure, called tapping the shunt, allows the neurosurgeon to measure CSF pressure at that particular point in time. In some cases—but not all—this can give the neurosurgeon a sense of whether or not the shunt is functioning properly.

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The CSF that is removed from the reservoir can be sent for analysis, such as a study of white blood cell count or a culture to look for evidence of infection.

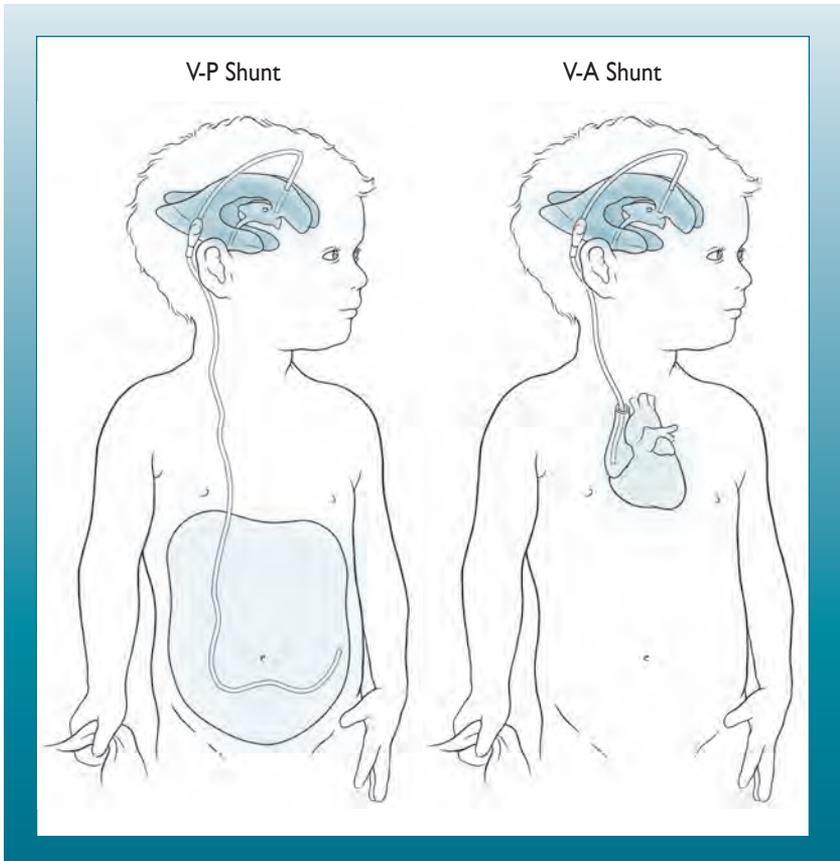
Reservoirs, which are recommended for all shunts, are most commonly associated with the ventricular catheter or incorporated into the valve system. Some reservoirs are compressible and can act as flushing devices. Depending upon the design of the system, the reservoir can push fluid either (most commonly) toward the head or away from the head. Some systems have a double reservoir, so that when the valve or reservoir is pumped, fluid can be pushed in either direction.

Unfortunately, whether or not the shunt pumps normally has little correlation with the functioning of the shunt. Many shunts may be working normally even though they don't pump well, while others that pump normally may be malfunctioning. It is not advisable to rely on the pumping characteristics as the sole measure of the shunt's functioning. Unless specifically advised by your neurosurgeon, pumping of the shunt is not recommended, as it can produce overdrainage or plugging of the system.

### ***Placement***

Like the parts of the shunt, the procedures used to place the shunt also are given their names according to where the shunt is placed in the body. The illustration at the right shows the placement of the shunt in the two most commonly performed procedures.

A ***ventriculoperitoneal (VP) shunt*** diverts CSF from the ventricles into the peritoneal cavity, the space in the abdomen where our digestive organs lie. The tip of the peritoneal catheter rests in this cavity near the loops of the intestine and bowel, but not inside them. The CSF shunted to this area is reabsorbed into the bloodstream.



A **ventriculoatrial (VA) shunt** diverts CSF from the ventricles into the right atrium of the heart. The atrial catheter is placed into a vein in the neck and then gently advanced through the vein into the atrium of the heart. Here the CSF passes directly into the bloodstream.

Placement of the VP shunt is generally the preferred procedure. On the whole, it has fewer risks and is easier to perform than the VA shunt procedure. Although other sites in the body may be used to divert the

flow of CSF from the ventricles (such as the chest cavity, in a ventriculopleural shunt), one would be chosen only if the usual sites for shunt placement cannot not be used or if the neurosurgeon determines it to be the most favorable for a particular child's circumstances.

### **Surgery**

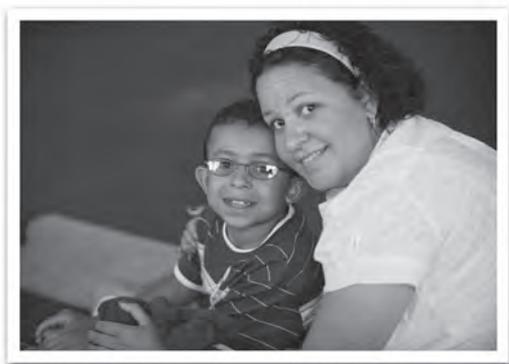
Your neurosurgeon should explain the type of shunt and placement procedure he or she plans to use. The insertion of a shunt is a relatively short and uncomplicated procedure. The child is brought to the operating room and is placed under general anesthesia. To ensure cleanliness, a small region of the scalp may be clipped or shaved, and, for a ventriculoperitoneal shunt, the entire area from the scalp to the abdomen is scrubbed with an antiseptic solution. Sterile drapes are placed over the child. Incisions are made in the head and abdominal areas. The shunt tube is passed beneath the skin, in the fatty tissue that lies just below the skin. A small hole is made in the skull, and the membranes between the skull and brain are opened. The ventricular end of the shunt is gently passed through the brain into the lateral ventricle. The abdominal (peritoneal) end is passed into the abdominal cavity through a small opening in the lining (peritoneum) of the

*At 22 weeks gestation Adora was diagnosed with congenital hydrocephalus due to aqueductal stenosis. The doctors gave Adora's parents a very grim prognosis not expecting her to live after her birth. However, she is now a very happy 4 year old, attending pre-school, loves reading books, singing songs, and playing music on her keyboard.*



*“Urian is a compassionate child who brings a positive outlook to life. He enjoys attending the Hydrocephalus Conferences and meeting other children with his same needs.” — Joanne*

*Urian was diagnosed with hydrocephalus at age 6 months. After a week of MRIs and various other tests, he was shunted. Urian also has Dandy-Walker variant. Urian is a bright 8-year-old boy currently attending second grade. He enjoys watching movies, spending time with mom and dad, and collecting cars and action figures.*



abdomen. This is where the CSF will ultimately be absorbed. The incisions are then closed. When the procedure is completed, sterile bandages may be applied to the incisions and the child is taken to the recovery room, where the anesthesia is allowed to wear off.

After the operation, the child will be watched closely as he or she recovers from surgery and the anesthesia. The neurosurgeon and nurses will check the child's vital signs and neurological status for signs of increased intracranial pressure (ICP) that would warn of a shunt malfunction. If the child is an infant, they will check the anterior fontanel for fullness and measure the head circumference at regular intervals. They will also watch the incisions for signs of infection. Some redness, swelling and tenderness are normal for the first week after surgery. The child may run a mild temperature for two or three days after the operation—this, too, is quite normal. If the child has a high fever or

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a fever that lasts for more than a few days, a surgeon or a pediatrician will do evaluations to determine what is causing it and how it should be treated. The neurosurgeon may specify that the child should stay in a certain body position for a period of time after the operation. For example, if greater drainage of CSF is necessary, the neurosurgeon may recommend that the child's head be elevated.

Shunt surgery usually involves minimal pain for the child. Some children experience neck and/or abdominal tenderness. Generally, mild analgesics such as acetaminophen are given. However, other medications are available to make the child comfortable, especially for the first few days after the operation. If all goes well and no complications arise, the child will be released from the hospital within one to three days.

After placement of a shunt, the size of the child's ventricles usually decreases. In infants, the fontanel becomes soft and may appear sunken, and the skull sutures will narrow or possibly even overlap. Except in infants, the shunt usually is not visible under the skin. A child whose hydrocephalus was diagnosed and treated early in infancy will have the same head size as his or her peers.

### Shunt Complications

Although hydrocephalus is almost always treated successfully with surgical placement of a shunt, shunt malfunction and, less frequently, infection occur in many cases. Shunt malfunction, which is caused by obstruction, simply means that the shunt is not able to divert enough CSF away from the ventricles in the brain. Shunt infection is caused by the child's own bacterial infection. These are serious problems and must be treated appropriately.

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*“It’s very important that every child have a good self-image. We feel our job as parents is to prepare all our children to lead a normal life when they leave the nest.”*

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### **Obstruction**

When shunt malfunction occurs, it is usually a problem with a partial or complete blockage of the shunt. The fluid backs up from the site of the obstruction and, if the blockage is not corrected, almost always results in recurrent symptoms of hydrocephalus. Shunt obstruction can occur in any of the components of the shunt. Most commonly, the ventricular catheter becomes obstructed by tissue from the choroid plexus or ventricles. The catheters or the valve may become blocked with blood cells or bacteria.

### **Infection**

Shunt infection usually is caused by contamination of the shunt at the time of surgery by the child’s own bacterial organisms; it is not acquired from exposure to other children or adults who are ill. The most common organism to produce infection is *Staphylococcus epidermidis*, which is normally found on the surface of the child’s skin and in the sweat glands and hair follicles deep within the skin. Infections of this type are most likely to occur one to three months after surgery but may occur up to six months after the placement of a shunt. Children with VP shunts are at risk of developing a shunt infection secondary to abdominal infection, whereas children with VA shunts may develop generalized infection, which can quickly become serious. In either case, the shunt infection must be treated immediately to avoid life-threatening illness or possible brain damage.

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*“Colin has the outward signs of surgeries (scars, shunt catheter) but other than those, you would never know how much this little boy has been through.” — Amy*

*Colin was diagnosed in utero with aqueductal stenosis as a result of an intraventricular hemorrhage. He also has Chiari malformation. At 7 days old, Colin had an endoscopic third ventriculostomy and placement of a ventriculoperitoneal shunt. He loves being in second grade and enjoys drawing, singing, playing piano, snowboarding, playing soccer, and being a cub scout.*



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### **Other Complications**

Shunts are very durable, but the components of the shunt can become disengaged or fractured as a result of wear or the child's growth, and occasionally they move within the body cavities where they originally were placed. More rarely, a valve will fail because of mechanical malfunction. However, it is possible that the valve pressure for a child's shunt system might drain fluid too rapidly or too slowly. To restore a balanced flow of CSF it might be necessary to replace the shunt with a new shunt containing a more appropriate pressure valve. Overdrainage of the ventricle could cause the ventricle to decrease in size to the point where the brain and its meninges pull away from the skull. If blood from broken vessels in the meninges becomes trapped between the

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*“When Julián was first diagnosed we wanted to do everything we could to allow Julián to have the best possible outcome, the Hydrocephallus association became an intricate part of our daily lives providing knowledge and resources helping us make decisions.” — Adriana*

*Julián was born prematurely and was diagnosed with hydrocephalus at 3 weeks. A VP shunt was placed at 5 weeks followed by two revisions by the time he was 10 months old. Julián just turned 5 years old and is very excited about going to kindergarten soon. After 5 years of various types of intervention such as speech and occupational therapy, Julián is age appropriate in every aspect. He loves sports – especially soccer.*



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brain and skull, resulting in a subdural hematoma, further surgery is required.

### **Signs of Shunt Malfunction and Infection**

Although symptoms of shunt malfunction vary considerably from child to child, a malfunction generally produces similar symptoms each time for a particular child. Shunt obstruction produces recurrent symptoms of hydrocephalus, increased intracranial pressure or fluid along the shunt tract.

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With infants, watch for such symptoms as a full and tense fontanel, bulging of the scalp veins and swelling or redness along the shunt tract. Also watch for unusual vomiting, irritability, sleepiness and decreased interest in feeding (infant appears to be less hungry, takes less volume, etc.). Once a child's head growth is complete and the sutures are closed, there are other symptoms to be alert for. Children and adults may experience headaches, vomiting, irritability and tiredness. Swelling along the shunt occurs less frequently. In the event of an abrupt malfunction, a child may develop symptoms rapidly, in a matter of hours or days. Older children and adults may become increasingly tired, may have difficulty waking up and staying awake and, unless treated promptly, may go into a coma.

Shunt infection frequently results in fever and may occur alone or in conjunction with shunt obstruction. Occasionally, shunt infection may produce reddening or swelling along the shunt tract.

Knowing what symptoms to watch for will help you become more at ease. Although the early symptoms of shunt malfunction or infection—fever, vomiting and irritability—are the same as for many childhood illnesses, you will learn to determine the symptoms associated with your child's shunt. Should you have any doubt about your child's symptoms, don't hesitate to call or visit your pediatrician for an evaluation. Remember, although shunt complications can be serious, they can almost always be treated successfully when they are discovered early. A review of symptoms to watch for is given on the next page.

### ***Shunt Revisions***

A shunt complication usually requires another operation to fix the shunt. Operations to fix a shunt are commonly called revisions. Depending on the cause of the complication, some or all of the components of the shunt will be replaced. In the event of infection, the child

## SYMPTOMS OF SHUNT MALFUNCTION OR INFECTION

This list of symptoms is for your reference only and is not a diagnostic aid. If you are in doubt about your child's medical condition, consult your physician immediately.

### Infants

- Enlargement of the baby's head
- Fontanel is full and tense when the infant is upright and quiet
- Prominent scalp veins
- Swelling along the shunt tract
- Vomiting
- Irritability
- Sleepiness
- Downward deviation of the eyes
- Less interest in feeding
- Fever\*
- Redness along the shunt tract\*

### Toddlers

- Head enlargement
- Vomiting
- Headache
- Irritability and/or sleepiness
- Swelling along the shunt tract
- Loss of previous abilities (sensory or motor function)
- Fever\*
- Redness along the shunt tract\*

### Children and Adults

- Vomiting
- Headache
- Vision problems
- Irritability and/or tiredness
- Personality change
- Loss of coordination or balance
- Swelling along the shunt tract
- Difficulty in waking up or staying awake
- Decline in academic performance
- Fever\*
- Redness along the shunt tract\*

*\*Fever and redness along the shunt tract both indicate infection.*

is given a course of antibiotic therapy and usually the entire shunt is replaced.

Although there are exceptional cases in which children receive a shunt and have no further need for revisions or replacements, these cases are rare. Experience shows us that some children undergo several revisions throughout their lives. Whether or not other complications will arise depends on your child's particular medical problems and bodily reactions to the surgical procedure and the shunt.

Hydrocephalus, left untreated, may cause brain damage with physical and severe cognitive challenges. We wish there were simple answers about when permanent damage can result—but much depends upon the timeliness and effectiveness of the treatment or the occurrence and severity of complications. The best way to prevent the possibility of your child's having brain damage is early detection of problems, should they occur. This is why it is so important that you learn the signs and symptoms of shunt malfunction and infection and have your child evaluated regularly by the pediatrician, neurosurgeon and

*Emma was just 1 year old when she was diagnosed with a brain tumor and hydrocephalus. At Children's Hospital of Wisconsin, Emma received surgery and a shunt, but by age 6 the tumor had returned. After having a second surgery, Emma now returns to Children's Hospital every 6 months to see Bruce Kaufman, MD, Medical Director of Neurosurgery, to make sure the tumor does not return.*



*Born prematurely, Waylon developed a brain bleed, which caused his hydrocephalus. Waylon is now 7 years old and is doing well. He is in first grade and he loves being a boy scout and playing baseball like his brother.*



neurologist. It is important to develop a strong relationship with the health-care team and to share information as well. Your pediatrician or family practice physician will provide your child's primary health care and will consult your neurosurgeon if a problem with the shunt is suspected. Your neurosurgeon will monitor your child's ventricles and will take care of problems associated with the shunt. A neurologist may follow your child's neurological status, as well as growth and development. You are an integral part of your child's health-care team, with your own knowledge of your child's health and history. Together with the medical professionals, you have the combined skills to provide excellent care for your child.

Some families find peace of mind—and a sense of control—in having their child carry or wear a medical identification device such as a surgical shunt I.D. card or a medical I.D. bracelet or necklace. Both methods of identification provide valuable medical information such as the

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*“To me, information always helped to soothe and make me feel like I could contribute something, rather than being out of control.”*

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names, addresses and telephone numbers of doctors to be contacted in an emergency; shunt type, manufacturer and pressure setting (if appropriate); and any additional information regarding medical conditions or allergies. Whether or not an identification device is important to you, it is crucial to keep your child’s medical reports up-to-date and easily accessible.

### **Endoscopic Third Ventriculostomy**

Endoscopic third ventriculostomy (ETV) is an alternate operation to treat hydrocephalus. The surgery entails making a tiny hole in the floor of the third ventricle to allow free flow of cerebrospinal fluid (CSF) into the spaces around the brain. This hole is made with the use of an endoscope, a long, thin instrument with a powerful magnifying lens, a light source, and narrow working channels that allow a surgeon to operate inside body cavities through very small openings. Improvements in endoscopic equipment, together with high-resolution MRI to visualize actual brain anatomy prior to the procedure have led to a new enthusiasm for ETV. As with all operations, the skill and experience of the neurosurgeon is important.

#### ***Who can be treated by endoscopic third ventriculostomy?***

ETV may be appropriate for hydrocephalus caused by obstructions of CSF flow in the aqueduct, the fourth ventricle, and its outlets as well as other cases. Some doctors report higher success rates in patients with aqueductal stenosis, the most common cause of congenital hy-

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*“If you deal with a life-threatening condition, it helps you appreciate each day, and what you have.”*

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drocephalus. Aqueductal stenosis is the obstruction of the long, narrow passageway between the third and fourth ventricles, which causes fluid to accumulate upstream from the obstruction. One technical requirement is that the third ventricle be wide enough for the endoscope. Shunted patients who have small ventricles, or so-called “slit” ventricles, may be candidates for ETV, but temporary disabling of the shunt to increase the size of ventricles may be necessary prior to surgery. This generally requires admission to an intensive care unit for several days, and it usually precipitates the symptoms that accompany acute hydrocephalus. It is generally reserved for patients who have been disabled by frequent episodes of shunt failure. Age is another factor in determining eligibility. Many neurosurgeons do not perform this procedure on infants or young toddlers.

***How is success defined?***

Five-year success rates for ETV are in the 50–80 percent range, depending upon the anatomy of the child’s brain and the cause of the hydrocephalus. Success is usually defined by patients and doctors alike as not requiring a shunt. Most doctors would categorize ETV as successful if a patient later shows clinical evidence of normal intracranial pressure (ICP) and structural evidence of stable or decreased ventricular size. If a patient has undergone ETV and still has a shunt in place, there may be some uncertainty about whether the ETV has been successful, whether the shunt is still draining, or both. Logically, this uncertainty can be resolved by removing the shunt, but additional surgery would not be in the patient’s best interest.

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*“It was hard not to be scared and worried all the time.  
We felt so much better after we got accurate  
information on hydrocephalus.”*

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Even when ETV is successful initially, it is very important for the child to have periodic neurosurgical evaluations. Success is usually determined within the first few months following the operation, but as with shunts, ETV can fail after years of successful control of symptoms. Many neurosurgeons recommend the same follow-up for children who have been treated by ETV as those with shunts.

***What are the potential complications of an ETV?***

Candid communication between the physician and parents/patient discussing possible complications as well as the definition of success is important when considering ETV. With new technologies the risks of ETV have been minimized.

The most common complications are fever and bleeding. Attempts to perforate the ventricular floor can lead to bleeding, as can damage to ventricular walls or perforation of the basilar artery. Large bleeds due to vessel injury under the third ventricle can be catastrophic, but they are rare. Short-term memory loss, which is typically temporary, is another complication, since the procedure may affect the hypothalamus and the areas of the mamillary body, which are responsible for memory. Because the area of the third ventricle, where the tiny opening is made, is responsible for some hormonal function, there is also a possibility of endocrinologic irregularities following ETV. This complication, too, is often short-lived.

ETV is not a cure for hydrocephalus, but rather an alternate treatment. There is a tendency for families and patients who have been treated successfully by ETV to forget that they ever had hydrocephalus. This is potentially dangerous. As noted above, ETV can fail after years of successful control of symptoms. Patients and their families must be watchful of recurrent symptoms of hydrocephalus, and as for patients with shunts, they should discuss with their care team what to do in case of emergency.



# CARING FOR YOUR CHILD

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*“The family is the central influence in the lives of children.”*

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All children have a need to be their own person as they mature and explore their world. A child with hydrocephalus is no different. It is essential that you treat your child as you would any other child, and that you afford him or her every opportunity to live as normal a life as possible. The shunt is a very durable device and should pose no special problems to normal handling or to childhood bumps and falls. Your child should be able to participate in most activities, with the possible exception of rough contact sports.

You will find that your knowledge and understanding of your child’s condition will increase together with your confidence and comfort in caring for him or her. When you have questions about hydrocephalus, write them down as they occur to you—and bring the list with you when you visit your doctor. You may find it helpful to talk to another family whose child has a similar problem. Also, realize that there are many resources available to families with children who have spe-

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*Anthony, who is now 4 years old, was diagnosed prenatally and born prematurely with aqueductal stenosis, causing his hydrocephalus. He has had one revision. He loves reading and collecting toy school buses.*



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*“Do not let your struggles with hydrocephalus be your burden in life...Let it be your success story.” — Mark*



*Mark was diagnosed prenatally and by the age of 22 he had endured many surgeries. He also has epilepsy. Despite these difficulties Mark has done well and is a proud recipient of the rank of Eagle Scout. Mark is currently attaining his Associates degree in English. He is engaged to Veronica and lives independently. For Mark the conditions of hydrocephalus and epilepsy have been challenges in his life, but he has not given up on his goals or dreams.*

*Natalie has had more than fifty shunt revisions; this is what she says about her hydrocephalus: “My family and I have never considered my hydrocephalus as a disability. It increased my motivation to do everything I ever wanted to do!”*

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cial needs. Begin by asking your nurse or doctor about some of these possibilities.

Friends and relatives can also offer valuable emotional support. And don't forget—all parents need to take time out for themselves. Allow a relative or a responsible sitter to care for your child from time to time. Leave important information and telephone numbers where you can be reached. When your family travels, get the names of medical resource personnel in the area to which you are going, and be sure to

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bring along important medical information as a safety measure. Although the likelihood of an emergency is remote, such preparedness will allow you greater peace of mind and will avoid unnecessary inconvenience should a problem arise. Some families choose to live in an area where access to a large medical center is convenient. If you live far from a major care center, you must plan to travel at times to ensure the best possible care for your child.

Depending on your child's medical problems, observation and consultation by other specialists may be needed. Your child may be referred to a neuro-ophthalmologist to have his or her vision examined. Sometimes parents who wish to have more children are referred to a specialist for a genetic consultation. The geneticist assesses parents and their child to find possible genetic causes for the child's medical problem and to determine the likelihood of another baby's being born with the same defect. Occupational therapists and educational psychologists can provide valuable assistance in your child's development. As advocates for your child, you and your physician must encourage strong communication and a team effort toward meeting your child's total health and developmental needs. Also, let your physician know your needs and concerns as a parent. And finally, make the effort to gather perspectives from the various health-care providers working with your child, in order to develop a framework of understanding about your child and this condition. This understanding, combined with your own experience as a parent, will enable you to offer the encouragement, support and resources your child needs to find acceptance and success in the world.

# AS YOUR CHILD GROWS UP

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As with all children, the age at which a child with hydrocephalus develops physical and intellectual skills varies. Many children with hydrocephalus have normal intelligence, physical development and coordination, but they may be slower in acquiring such skills as eye-hand coordination and in learning to walk. Each child is different, and each child's level of attainment in skills depends upon many factors. Your child's developmental progress will be influenced by the nature of the problem causing the hydrocephalus, by the degree of brain damage, if any that occurred before treatment and by infections or other complications. But a child's overall development and adaptation to the world also depends upon the individual child and the attitude and opportunities afforded him or her by parents and environment.

Seeking a specialized pediatric psychologist can help to maximize your child's physical, intellectual, emotional and social development. It is recommended your child be evaluated by a pediatric psychologist who has special skills in neuropsychological and emotional assessment. By giving your child a variety of diagnostic tests, the psychologist will be able to identify the strengths and weaknesses in his or her abilities. And because there are thought to be critical development stages at which optimum learning takes place, we urge that your child have regular evaluations.

As an infant, your child will be evaluated for such things as alertness, movement and tracking (responsiveness to sound and moving objects). As your child gets older, she or he will be evaluated for verbal, intellectual and reasoning skills, as well as for social and emotional growth. All are crucial to sound and full development. Early identification and intervention can help to compensate for known deficiencies and can stimulate your child's developing abilities, offering your child every opportunity to achieve his or her fullest potential. If your pediatrician and neurosurgeon are unfamiliar with a pediatric psychol-

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*“Max has helped me be a better person and confirms for me every day not to sweat the small stuff and be a joyful person.” — Michelle*

*Max was shunted at one year of age due to bacterial meningitis at 6 months. He has endured 10 shunt revisions in 12 years. At age nine, Max was also diagnosed with Marfan’s Syndrome. With his many challenges Max still loves to play basketball, golf, and baseball. He is an exceptional student in school and loves literature, math and sciences.*



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ogist who does such diagnostic testing, ask them to check with local resources or the nearest medical center and refer you to one.

As your child reaches school age, developmental testing can provide valuable information to help teachers meet your child’s educational needs. Federal law requires all public schools to address and provide for the educational needs of all children, including those with special needs. Very likely your healthcare team includes, or is supported by, a social worker who can be your child’s advocate as you seek the services that your child requires. If you notice a decline in your child’s academic performance, realize that many factors can contribute to these

changes, including shunt malfunction. Consult your health-care team for an evaluation. They may request an additional evaluation by a psychologist and, through their assessments, they will determine if your child has a problem with the shunt and may identify other contributing factors and offer helpful recommendations.

Challenge your child to seek out his or her potential. Your acceptance and love will have great impact on how he or she perceives him- or herself and, ultimately, succeeds in the world. Your positive attitude and encouragement will afford your child the greatest opportunity to live a full and happy life.



*Sarah and Stephanie are sisters who both have hydrocephalus due to aqueductal stenosis. Sarah (29) developed hydrocephalus and was shunted at age four. She has had over 30 revisions, along with a brain herniation. Sarah recently completed her B.S. in business administration/accounting. She enjoys knitting and going to movies with her brother and her friends. Stephanie (28) was diagnosed and shunted at six weeks of age. She has had three revisions. Stephanie completed her B.S. and M.S. in engineering and was recently married and is looking forward to beginning a new chapter in her life.*

# LOOKING TO THE FUTURE

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We must look to the future with vision and hope. Medical science is an advancing and dynamic field. Today we have solutions to medical problems that were not even dreamed of in the past. And through science and technology people will continue to expand the limits of what is possible. As we go forward, we must have faith in ourselves and our children. When faced with life's challenges, we discover not only personal strengths but also a greater capacity for compassion and love. It is from endeavors like these that we find true value and meaning in life.

The Hydrocephalus Association is positioned at the nexus of a movement to strategically drive hydrocephalus research. We are aligned with professional and peer advisors, with Congress and members of government, with other organizations and with key agencies. It is our intent to drive and support the research agenda by directly funding hydrocephalus research.

Research and experience show that children with hydrocephalus have excellent opportunities to attain their full potential through comprehensive integrated medical care and programs that stimulate their development. Individuals, families and professionals working together in an atmosphere of mutual trust and respect endure that an appropriate, comprehensive, ongoing care plan is in place.

# RESOURCES

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The Hydrocephalus Association is a national, 501 (c) (3) nonprofit organization founded in 1983. Our mission is to eliminate the challenges of hydrocephalus by stimulating innovative research and providing support, education and advocacy for individuals, families and professionals dealing with the complex issues of the condition.

The Association provides comprehensive services that empower individuals and families to seek out the best medical care, programs and resources that meet their needs now and in the future.

As the nation's largest and most widely respected organization dedicated solely to hydrocephalus, the Association has been instrumental in creating a community of individuals, families and health-care professionals addressing the complexities of hydrocephalus in all age groups—infants, children, young adults and adults. We continually update and expand our resources to keep pace with new technologies in the diagnosis and treatment of hydrocephalus and stay current with the needs of the individuals we serve.

Hydrocephalus is a chronic condition. With early detection, effective treatment and appropriate interventional services, the future for individuals with hydrocephalus is promising. We invite your inquiries.

# HEALTH RECORD

|  |                  |              |           |                   |
|--|------------------|--------------|-----------|-------------------|
| Name                                     | Phone            | Email        | Birthdate | Premature: Yes No |
| Parent/Guardian                          | Phone (home)     | Phone (work) | Email     |                   |
| Neurosurgeon                             | Phone            | Email        | Address   |                   |
| Pediatrician                             | Phone            | Email        | Address   |                   |
| Other Doctors                            |                  |              |           |                   |
| Name                                     | Phone            | Email        | Address   |                   |
| Name                                     | Phone            | Email        | Address   |                   |
| Type of Hydrocephalus/Other Conditions   |                  |              |           |                   |
| Shunt Name/Type                          | Pressure Setting |              | Allergies |                   |
| Brief Surgical/Medical History           |                  |              |           |                   |
| Medications                              |                  |              |           |                   |
| Developmental/Neuropsychological Testing | Date             | Follow-up    |           |                   |
| Notes/CT, MRI Scans or Head Ultrasounds  |                  |              |           |                   |

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## **Educational Booklets and Fact Sheets**

About Hydrocephalus: A Book for Families  
(available in English and Spanish)

Prenatal Hydrocephalus: A Book for Parents

Healthcare Transition Guide for Teens and Young Adults  
with Hydrocephalus

Hydrocephalus Diagnosed in Young and Middle-Aged Adults

About Normal Pressure Hydrocephalus: A Book for Adults  
and their Families

A Teacher's Guide to Hydrocephalus

More than 20 one- to four-page fact sheets that explain common  
questions about hydrocephalus and its complications

## **National Conference**

Biennial National Conference for Families and Professionals

## **Directories of Medical Professionals**

Pediatric Neurosurgeons

Neurosurgeons and Neurologists for Adults

Neuropsychologists

## **Support Network**

Local support groups all over the nation

## **Monthly E-Newsletter**

Available online or in print



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