



*Hydrocephalus Diagnosed in
Young and Middle-Aged Adults*

A Book for Adults and Their Families

Hydrocephalus Diagnosed in Young and Middle-Aged Adults—A Book for Adults and Their Families was written for adults, their families and friends with the intention of providing information about the diagnosis and treatment of hydrocephalus diagnosed in adulthood. It is a companion piece to our booklet *About Hydrocephalus—A Book for Parents*, the most widely distributed resource on infant and childhood hydrocephalus in the United States.

It is our belief that families and individuals dealing with the complex issue of hydrocephalus diagnosed in young and middle-aged adults must become educated about the condition in order to make informed decisions regarding treatment and care. While each case differs, the information presented in this booklet is intended to give a general overview of the condition without making judgments or recommendations for individual care.

HYDROCEPHALUS ASSOCIATION

SAN FRANCISCO, CALIFORNIA

SECOND EDITION, JANUARY 2003

Editor: Rachel Fudge

Illustrations: Lynne Larson © 2003

Medical Consultants: Harold L. ReKate, M.D.
Barrow Neurological Institute, Phoenix, AZ

Michael A. Williams, M.D.
Johns Hopkins Hospital, Baltimore, MD

Special thanks to our medical reviewers:

Rick Abbott, M.D.; Alexa Canady, M.D.; and Joseph Piatt, M.D.

The following references were consulted in the writing of this booklet:

- Drs. J. A. Cowan, D. Rigamonti and M. A. Williams: "Clinically Distinct Syndrome of Hydrocephalus in Young and Middle-Aged Adults," presented at the 3rd International Hydrocephalus Workshop, Kos, Greece, May 2001.
- Drs. G. P. Prigatano and H. L. ReKate: "Long-Standing Overt Ventriculomegaly of the Adult: Neuropsychological Considerations," presented at the 6th National Conference on Hydrocephalus, Phoenix, AZ, May 2000.
- Drs. T. Fukuhara and M. Luciano: "Clinical Features of Late-Onset Idiopathic Aqueductal Stenosis," *Surgical Neurology* 2001; 55: 132-7.
- Drs. N. Buxton, D. Macarthur, M. Vloeberghs, J. Punt, I. Robertson: "Neuroendoscopic Third Ventriculostomy for Hydrocephalus in Adults: Report of a Single Unit's Experience with 63 Cases," *Surgical Neurology* 2001; 55: 74-8.

Publication of this booklet was made possible by a generous grant from the George H. Sandy Foundation.

© 2003 Hydrocephalus Association, San Francisco, California

All rights reserved. No part of this booklet may be used or reproduced in any form or by any means, or stored in a database or retrieval system, without prior written permission of the Hydrocephalus Association. Making copies of this booklet is against the law. Additional copies of this booklet may be obtained for a small fee from the Hydrocephalus Association.

*Hydrocephalus Diagnosed in
Young and Middle-Aged Adults*

Introduction

FOR many years, people with hydrocephalus, their families and their doctors have worked unceasingly to increase public awareness of hydrocephalus, its causes and its treatment. In the past half-century, great advances have been made in the diagnosis and treatment of hydrocephalus, especially as it occurs in infants. Normal pressure hydrocephalus (NPH), which occurs primarily in the elderly, is now receiving a great deal of media and medical attention, leading to more efficient and timely diagnosis and treatment. Hydrocephalus is now regularly detected in utero, before the baby is born. New treatment protocols, including advances in shunt and endoscopic technology, make it more likely than ever that people with hydrocephalus—whether diagnosed in infancy or adulthood—will live full, rich lives.

Despite these advances over the past 50 years, there is still much to be learned about hydrocephalus and the subtle forms it can take. Even now, hydrocephalus is most commonly understood to occur in infants or the elderly—at the extremes of life. But in actuality, hydrocephalus can occur at any time in life, and as a result of a variety of causes. And now doctors are beginning to identify and describe a distinct form of hydrocephalus that arises in young and middle-aged adults.

Vastly different from hydrocephalus diagnosed in infancy and early childhood, or adult-onset normal pressure hydrocephalus found in older adults (typically age 60 and older), hydrocephalus in young and middle-aged adults is a unique and often confusing condition. Though it has only recently begun to receive formal attention from the medical community, hydrocephalus in this age group presents a host of challenges and opportunities for patients and medical professionals alike. The challenge goes far beyond routine or specialized medical care, encompassing psychosocial, emotional and occupational issues, all of which will be discussed in this booklet.

A Note on Terminology

Medical professionals are only just beginning to identify and describe the distinct syndrome of hydrocephalus in adults. As yet, there is no universally agreed-upon term, such as normal pressure hydrocephalus, to describe this population. We have chosen to use the term coined by Michael A. Williams, M.D.: the syndrome of hydrocephalus in young and middle-aged adults (SHYMA). Other terms used to describe this and similar populations are late-onset idiopathic aqueductal stenosis, longstanding overt ventriculomegaly of the adult, and late-onset aqueductal stenosis.

About This Booklet

Some of the information in this booklet is adapted or reprinted from *About Hydrocephalus—A Book for Families* and from other materials published by the Hydrocephalus Association. We begin with a detailed description of hydrocephalus in general, including its causes. Chapter 2 discusses symptoms and diagnosis, and Chapter 3 provides information about treatment of hydrocephalus. Social and emotional issues resulting from SHYMA are outlined in Chapter 4. At the very end is a detailed list of further resources to consult.

This is our first attempt to provide a detailed overview of SHYMA. Because this condition is only just beginning to be understood and identified, we realize that we all have much to learn. A revised edition will almost certainly be published within the next few years, as SHYMA becomes better understood. We welcome your feedback.

1 ~ About Hydrocephalus

Anatomy and Physiology

The 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. The bones of the skull are often referred to as the cranium. 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 to the tail bone.

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 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 called the subarachnoid space. CSF is in constant circulation and serves several important functions. Because it surrounds the brain and spinal cord, CSF acts as a protective cushion against blows to the head and spine. Though it is clear and colorless, CSF contains many nutrients and proteins that are needed for the nourishment and normal function of the brain. It also carries waste products away from surrounding tissues.

Ventricles

CSF is produced within the cavities of the brain that are called ventricles. Imagine the ventricles as chambers filled with fluid. There are four in all: the two lateral ventricles, the third ventricle and the fourth ventricle. The ventricles are interconnected by narrow passageways. Your neurosurgeon or neurologist can learn valuable information about your condition by closely monitoring the size and shape of these ventricles as shown by CT or MRI scans.

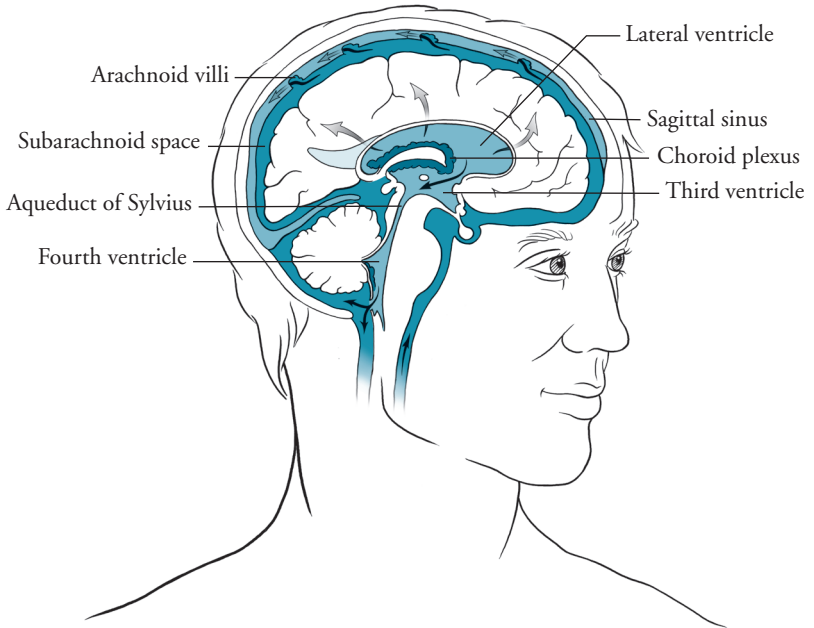
Cerebrospinal Fluid Circulation and Absorption

CSF is formed within the ventricles by small, delicate tufts of specialized tissue called the choroid plexus. Beginning in the lateral ventricles, CSF flows through 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. Most of the CSF is absorbed through tiny, specialized cell clusters (arachnoid villi) near the top and midline of the brain. CSF passes through the arachnoid villi into a large vein (the superior sagittal sinus) and is absorbed into the bloodstream. The ventricular system is the major pathway for the flow of CSF.

Hydrocephalus

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 rate at which CSF is produced and the rate at which it is absorbed. Hydrocephalus occurs when this balance is disrupted and the rate of absorption is less than the rate of production. 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 below.

Because CSF is produced continuously, when its flow is blocked it will begin to accumulate upstream from the site of the obstruction,



Cerebrospinal fluid (CSF) circulatory pathway: *The drawing shows a view of the brain. The black arrows show the major pathway of CSF flow. The gray arrows show additional pathways.*

much like a river swells behind a dam. Eventually, as the amount of fluid 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** or **obstructive hydrocephalus** because the ventricles no longer provide free passage of CSF through them into the subarachnoid space.

Another type of hydrocephalus is **communicating** or **extraventricular obstructive hydrocephalus**. It usually results from a thickening of the arachnoid around the base of the brain, which blocks the flow of CSF from the spinal to the cortical subarachnoid spaces. CSF flows unrestricted through the ventricles, but a blockage between the spine and the fluid around the outside of the brain prevents the free flow of CSF through the subarachnoid space.

Etiology (Causes)

Hydrocephalus that is **congenital** (present at birth) is thought to be caused by a complex interaction of environmental and perhaps genetic factors. (Aqueductal stenosis and spina bifida are two examples.) Some people with SHYMA are classified as having **decompensated congenital hydrocephalus**. That is, the hydrocephalus may have been present at birth, and perhaps even treated in early childhood, but remained largely compensated and asymptomatic for many years. Congenital hydrocephalus can be diagnosed by assessing head circumference. If the head circumference is significantly larger than normal, according to standard charts and references, it is reasonable to suspect that hydrocephalus has been present since infancy, even though it may not have been symptomatic in infancy.

Acquired hydrocephalus arises after birth, and results from intraventricular hemorrhage, meningitis, head trauma, encephalitis, tumors or cysts.

Sometimes doctors are unable to pinpoint the cause of the hydrocephalus. In this case, the hydrocephalus is deemed to be *idiopathic*, meaning with no known cause.

It is not known whether genetic factors play any role in SHYMA, although inherited forms of hydrocephalus are virtually unknown.

The causes of SHYMA are similar to the causes of hydrocephalus at all ages, including processes that obstruct the ventricles, such as cysts or tumors, and processes that impair the flow of spinal fluid through the subarachnoid space, such as meningitis, encephalitis, concussion, head injury, or certain strokes and brain hemorrhages.

Hydrocephalus does not always occur immediately after one of these predisposing conditions has occurred. In many instances, years or decades may pass before the symptoms of SHYMA become evident.

2 ~ Symptoms & Diagnosis

THE symptoms of SHYMA are similar in some ways to those of normal pressure hydrocephalus (NPH) in the elderly, but are often much more subtle. And yet they can have a most profound effect on patients' lives, rendering them unable to work or to have difficulty functioning in day-to-day life.

Although the medical literature describing SHYMA and related conditions is limited, it appears that diagnosis is often difficult, causing physical and mental distress and possibly serious long-term health problems due to an extended period of pressure on the brain. Sometimes, symptoms are disregarded as manifestations of mid-life crisis or other psychological/emotional issues. Additionally, as symptoms worsen and the condition goes undiagnosed, one's ability to function at work can be affected, and employment can be compromised.

Symptoms

The most common symptoms of SHYMA are disturbances in gait, cognition and bladder control. Chronic headaches are also frequently reported.

Gait disturbances include a sense of clumsiness, and difficulty walking on uneven surfaces or stairs. Subtle gait abnormalities may be seen during a careful examination, but obvious gait impairment or apraxia is rare.

Cognitive complaints are often described as "dullness," decline in organizational skills and dependence on lists, and may lead to difficulty with job duties. And in fact, a great number of people with SHYMA report that their symptoms resulted in decreased job performance. Depending on the length of time hydrocephalus has gone untreated, cognitive decline can vary greatly.

Chronic headaches that are hard to relieve with pain medications (such as aspirin or acetaminophen) are also frequently reported. Other symptoms include visual complaints and syncope (fainting).

Patients' complaints are often quite prominent, in contrast to the doctors' clinical findings, which can be subtle—a discrepancy that may impede accurate diagnosis of SHYMA. The average time to diagnose—that is, the time between the onset of symptoms and a diagnosis of hydrocephalus—appears to be significant, and is often years.

Some people with SHYMA may have been diagnosed with hydrocephalus as children or infants, and perhaps even shunted, yet displayed no further symptoms and hence were not treated for years. Perhaps their shunts malfunctioned or stopped working somewhere along the way, setting the stage for a relapse of symptoms. The slow, insidious return of hydrocephalus symptoms in SHYMA suggests that the hydrocephalus was probably compensated at the time of shunt obstruction, and it is only with the passage of time (sometimes years) that decompensation occurs and symptoms re-emerge. If the hydrocephalus were not compensated, then shunt obstruction would produce fairly rapid onset of hydrocephalus symptoms. The first generation of children shunted for hydrocephalus in the 1950s and '60s may be at particular risk for SHYMA, as some of them may not have received any neurosurgical care since childhood.

The degree of symptoms and their resultant effect varies widely among patients. If symptoms have been present for years, the patient may be more seriously disabled. Early diagnosis can be a factor in successful resolution of symptoms.

Diagnosis

SHYMA is diagnosed using a combination of brain scans, intracranial pressure monitoring and clinical evaluation of symptoms.

Once symptoms of gait disturbance, mild dementia or bladder control have been identified, a physician who suspects hydrocephalus may

recommend one or more additional tests. At this point in the diagnostic process, it is important that a neurologist and a neurosurgeon become part of your medical team, along with your primary care physician. Their involvement from the diagnostic stage onward is helpful not only in interpreting test results and selecting likely candidates for shunting but also in discussing the actual surgery and follow-up care, as well as expectations of surgery. The decision to order a given test may depend on the specific clinical situation, as well as the preference and experience of your medical team.

These tests may include computerized tomography (CT), magnetic resonance imaging (MRI), lumbar puncture, continuous lumbar CSF drainage, intracranial pressure (ICP) monitoring, measurement of cerebrospinal fluid outflow resistance or isotopic cisternography. Neuropsychological evaluation may also be recommended.

CT Scan

CT is a picture of the brain created by using x-rays and a special scanner. It is safe, reliable, painless and relatively quick (about 15 minutes). An x-ray beam passes through the head, allowing a computer to make a picture of the brain. A CT will show if the ventricles are enlarged or if there is obvious blockage.

Unexpected enlargement of the ventricles is often the first clue that SHYMA may be present, but this finding alone is usually not a sufficient reason to proceed to surgical therapy. It is important to remember that hydrocephalus can be compensated or uncompensated, and further testing is often needed. It is potentially harmful to put a shunt into a person with compensated hydrocephalus, just as it is potentially harmful *not* to put a shunt into a person with decompensated hydrocephalus.

MRI

MRI is also safe and painless, but it will take longer than 15 minutes. MRI uses radio signals and a very powerful magnet to create a picture of the brain. Again, it will be possible to detect if the ventricles are enlarged as well as evaluate the CSF flow and provide information about the surrounding brain tissues. (The MRI provides more information than the CT, and is therefore the test of choice in most cases.) MRI scans

can also assess how CSF moves through a particular part of the brain called the cerebral aqueduct (“the CSF flow void sign”).

In idiopathic forms of hydrocephalus, it is essential to perform an MRI with and without contrast enhancement. This allows the doctor to look for subtle signs of tumor or chronic infection, which can cause adult-onset hydrocephalus and which may require additional treatment besides shunt surgery.

Lumbar Puncture

Lumbar puncture, or spinal tap, allows an estimation of CSF pressure and analysis of the fluid. Under local anesthetic, a thin needle is passed into the spinal fluid space of the low back. Removal of up to 50 cc of CSF is done to see if symptoms are temporarily relieved. If removal of some CSF dramatically improves symptoms, even temporarily, then surgical treatment may be successful. The use of a lumbar puncture as a screening test for SHYMA is not advocated by all physicians, since many patients who experience little or no improvement after the test may still improve with a shunt.

This test is extremely important if the cause of the hydrocephalus is not obvious. Occult infections or tumors with CSF spread may be found from culturing and analyzing the CSF for tuberculosis or fungi.

Lumbar catheter insertion is a variation of the lumbar puncture. A spinal needle is inserted in the spinal fluid space of the low back, then a thin, flexible tube (catheter) is passed into the spinal fluid and the needle is removed. The lumbar catheter allows for continuous and more accurate recording of spinal fluid pressure, or for continuous removal of spinal fluid over several days to imitate the effect that a shunt would have. Patients who respond dramatically to such spinal fluid drainage are likely to respond to shunt surgery. Lumbar catheter insertion requires hospitalization.

Intracranial Pressure Monitoring

Intracranial pressure monitoring requires admission to the hospital. A small pressure monitor is inserted through the skull into the brain or ventricles to measure the ICP. The pressure is not always high, and con-

tinuous pressure monitoring over 24 or 48 hours (either by the lumbar catheter or the intracranial method) may be necessary to detect an abnormal pattern of pressure waves.

The measurement of CSF outflow resistance is a more involved test that requires a specialized hospital setting. In essence, this test assesses the degree of blockage to CSF absorption back into the bloodstream. It requires the simultaneous infusion of artificial spinal fluid and measurement of CSF pressure. If the calculated resistance value is abnormally high, then there is a very good chance that the patient will improve with shunt surgery.

Isotopic Cisternography

Isotopic cisternography involves having a radioactive isotope injected into the lumbar subarachnoid space (lower back) through a spinal tap. This allows the absorption of CSF to be evaluated over a period of time (up to 96 hours) by periodic scanning. This will determine whether the isotope is being absorbed over the surface of the brain or remains trapped inside the ventricles. Isotopic cisternography involves lumbar puncture and is considerably more involved than either the CT or MRI. This test has become less popular because a “positive” cisternogram result does not reliably predict whether a patient will respond to shunt surgery. Its utility in SHYMA is not known.

Neuropsychological Testing

Neuropsychology is the study of brain-behavior relationships. When a patient’s primary problems include thinking, emotion and behavior, neuropsychological evaluation can be an important complement to clinical work in making a diagnosis. An evaluation can also help family members and physicians understand the impact of hydrocephalus on a patient’s everyday function.

A neuropsychological assessment typically includes a thorough interview with the patient and one or more family members, as well as a close review of medical records and studies. A series of tests is administered to assess various aspects of cognitive function, including attention, memory, language, visual-spatial ability and executive function (the ability to reason, plan and modulate behavior). The ultimate goal is to understand

how changes in brain structure and function are affecting the patient's behavior.

In the case of SHYMA, neuropsychological testing can help determine whether or not a patient would benefit from immediate surgical intervention or should simply be monitored for a longer period.

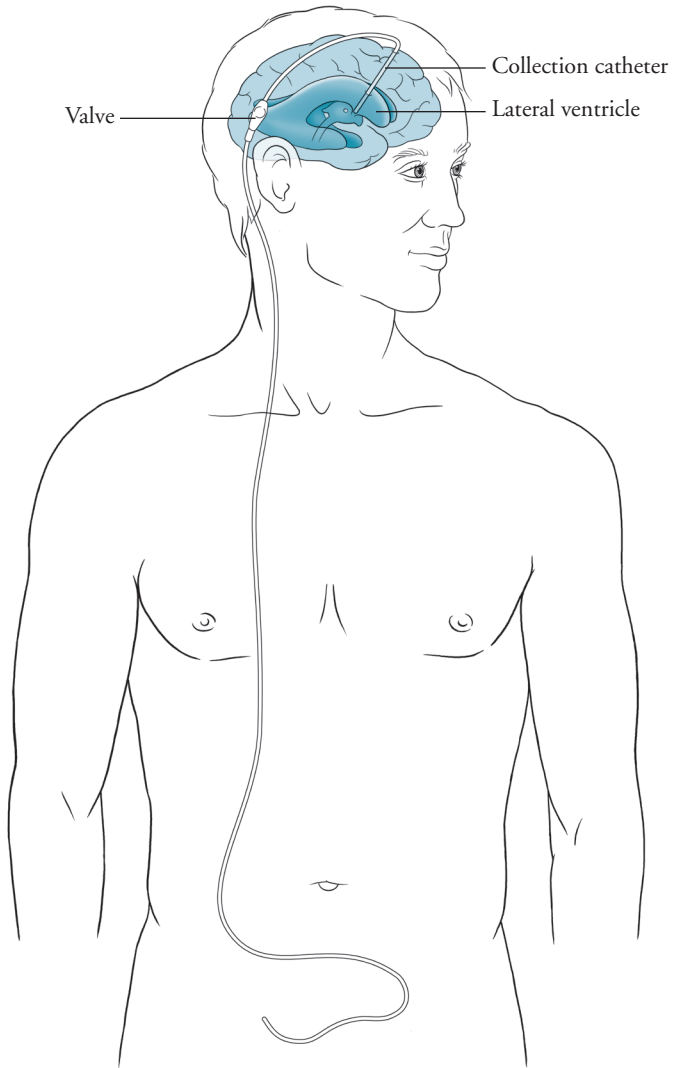
3 ~ Treatment

IN many cases, prompt treatment can reverse many of the symptoms of hydrocephalus, restoring much cognitive and physical functioning. If left untreated, however, symptoms can become quite disabling, leading to severe cognitive and physical decline. It appears that the length of time between onset of symptoms and diagnosis is a factor in the success of treatment. Another, as yet unmeasurable, factor that affects the outcome of treatment is the extent of reversible versus irreversible brain injury caused by hydrocephalus. Treatment is most successful when little irreversible injury has occurred. Transient improvement of symptoms after lumbar puncture or spinal fluid drainage by lumbar catheter is one way to demonstrate that some of the brain injury is still reversible.

Shunting

The most common treatment for SHYMA, as with all forms of hydrocephalus, is shunting. A shunt is a flexible tube placed into the ventricular system that diverts the flow of CSF into another region of the body where it can be absorbed, such as the peritoneal (abdominal) cavity or the right atrium of the heart. The shunt tube is about one-eighth inch in diameter and is made of a soft and pliable plastic 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 valve in the shunt maintains the CSF at normal pressure within the ventricles.

The surgical placement of a shunt, which is performed by a neurosurgeon, is a relatively short procedure. The patient is brought to the operating room and is placed under general anesthesia. 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 patient. 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.



With the VP shunt in place, cerebrospinal fluid (CSF) flows into the collection catheter and down the exit catheter, which shunts the fluid into the peritoneal cavity.

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 ventricle. The abdominal (peritoneal) end is passed into the abdominal cavity through a small opening in the lining (peritoneum) of the 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 patient is taken to the recovery room where the anesthesia is allowed to wear off.

Success of Shunting

In some studies of patients with SHYMA, shunting has had extremely high rates of success, reversing marked decline and returning patients' lives to "normal." However, due to the lack of data collected on this specific population, statistics are hard to come by.

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. These are serious problems and must be treated appropriately.

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, or the distal (emptying) catheter becomes obstructed. The reason for the distal catheter obstruction depends on the site of the distal catheter. If it is in the abdominal (peritoneal) cavity or the cavity between the chest wall and the lining of the lung (pleural cavity), then obstruction can occur from debris or scarring around the tip of the

catheter. In the jugular vein, debris or an organized blood clot can form on the catheter.

Infections

Shunt infection usually is not acquired from exposure to others who are ill. Instead, it is caused by bacteria that normally reside on the skin of *all* persons. The most common organism to produce infection is *Staphylococcus epidermidis*, which is normally found on the surface of the 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. People with VP shunts are at risk of developing a shunt infection secondary to abdominal infection, whereas people 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.

Other Complications

Shunts are very durable, but their components can become disconnected or fractured as a result of wear, 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 person'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 brain and skull, resulting in a subdural hematoma, further surgery is required. This is a potentially life-threatening complication, and it is more common in adults than in children.

Signs of Shunt Malfunction and Infection

Although symptoms of shunt malfunction vary considerably from person to person, a malfunction generally produces similar symptoms each time for a particular person. Shunt obstruction produces recurrent symptoms of hydrocephalus, increased intracranial pressure or fluid along the shunt tract. The nature of the symptoms will depend on: 1) whether the hydrocephalus is obstructive or communicating, and 2) the spectrum of symptoms the person had initially. Generally, shunt obstruction causes a person's original symptoms to return.

Adults with obstructive (noncommunicating) hydrocephalus may experience headaches, vomiting, irritability and fatigue. Swelling along the shunt occurs less frequently. In the event of an abrupt malfunction, a person may develop symptoms rapidly, in a matter of hours or days. He or she may become increasingly tired, may have difficulty waking up and staying awake and, unless treated promptly, may go into a coma.

Adults with communicating hydrocephalus will experience the gradual, almost unnoticeable, return of their symptoms. In many instances, they or their family suddenly recognize the symptoms have returned, and then when they think carefully about it, they realize the symptoms have been returning over days or weeks.

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.

The fear of possible shunt obstruction can be a source of persistent anxiety and apprehension for patients and families. Knowing what symptoms to watch for can help you be more at ease. Although the early symptoms of shunt malfunction or infection—fever, vomiting and irritability—are the same as for many illnesses, a person with shunted hydrocephalus will learn to determine the symptoms associated with his or her shunt. A doctor should always be consulted by phone or in person if there is any doubt about symptoms. 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 below.

Signs and Symptoms of Shunt Infection or Malfunction

Vomiting
Headache
Vision problems
Irritability and/or fatigue
Personality change
Loss of coordination or balance
Swelling along the shunt tract
Difficulty in waking up or staying awake
Decline in cognitive function
Fever*
Redness along the shunt tract*

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

Endoscopic Third Ventriculostomy

Endoscopic third ventriculostomy (ETV) is a relatively new surgical procedure for the treatment of hydrocephalus. The surgery involves making a hole in the floor of the third ventricle to allow free flow of spinal fluid into the basal cisterns for absorption. This concept is an old one, and other procedures utilizing this type of approach have been tried for many years. The improvement in endoscopic equipment combined with the ability of MRI to visualize actual brain anatomy prior to the procedure have led to a new enthusiasm for ETV.

ETV is generally considered to be appropriate for treating obstructive (noncommunicating) hydrocephalus. It is controversial as to whether it is effective in treating non-obstructive (communicating) hydrocephalus, although some neurosurgeons have used it successfully in these cases. In order to perform the procedure, the ventricles must be large enough to see the appropriate brain structures.

Success of ETV

Because the identification of SHYMA is so recent, and the population treated with ETV so small, it remains to be seen how successful ETV is for treating SHYMA. However, some small-scale studies have indicated

that ETV can be effective in treating late-onset idiopathic aqueductal stenosis.

It is important to realize that ventricular volume (size) does not always decrease after ETV or shunt surgery, even among patients who seem to have relief of symptoms, so the success of the procedure cannot be assessed simply by imaging (CT or MRI). SHYMA patients in particular may have nonspecific symptoms or symptoms out of proportion to imaging findings. If ETV does not relieve all symptoms completely, determining whether the treatment has been adequate or whether the patient should go on to a shunt can be very difficult.

Studies assessing the relative merits of shunting versus ETV in children (not adults) suggest that ETV does not “cure” hydrocephalus any better than shunts do. While ETV may eliminate the future need for a shunt, and the associated risks of shunt malfunction, available data indicates that children with hydrocephalus who have undergone ETV return for additional surgical procedures as often as shunted patients do.

Even when ETV is initially successful, it is crucial for the patient to have periodic neurosurgical evaluations.

Finding a Neurosurgeon/Neurologist

Establishing a relationship with a health-care team is important to the success of hydrocephalus treatment. While each individual’s needs vary, two important components for a successful relationship are communication and trust. Just as you need a doctor who is experienced in hydrocephalus and open and willing to discuss your particular situation, doctors need you to be candid and knowledgeable about the specifics of your situation and needs.

There are many competent and skilled neurologists and neurosurgeons to choose from. Unfortunately, outside of the pediatric neurosurgery community, it can be difficult to find a doctor with experience and expertise in treating hydrocephalus.

In cases of young and middle-age onset of hydrocephalus, the neurologist is oftentimes the “gatekeeper”—the first doctor, after the primary care provider, a patient gets referred to when symptoms appear, or persist. A neurologist can be both a principal care provider and a consultant to other physicians. For disorders that require frequent care, such as Parkinson’s disease, Alzheimer’s disease or multiple sclerosis, a neurologist is often the principal care provider. For conditions such as hydrocephalus, stroke or concussion, the neurologist acts more in a consulting role, providing diagnostic testing and referral to other specialists when necessary.

Because most neurologists and primary care providers do not see many patients with hydrocephalus, they may not be aware of diagnostic protocols and treatment options, and may not properly recognize the condition or recommend additional consultation with a neurosurgeon. That is why it is very important for you and your family to become educated about hydrocephalus in general and be willing and able to speak knowledgeably with the neurologist, or the primary care provider. If you discover that the doctor is not experienced or interested in your condition, it’s time to get a second or even a third opinion.

Neurosurgeons are usually principal care providers, offering not only operative management but also diagnosis, evaluation and critical care. In the case of hydrocephalus, the neurosurgeon will order and review CT and MRI scans and often will work in consultation with the neurologist on other diagnostic tests. It is the neurosurgeon who will place the shunt, or perform the endoscopic third ventriculostomy, and follow the patient once surgery is done.

Finding a neurosurgeon who is knowledgeable and experienced with hydrocephalus is critical, and oftentimes difficult for adults. Because hydrocephalus is more often identified in infants and young children, neurosurgeons without a specialty in pediatrics are not as likely to have experience in this area.

Whether you are newly diagnosed, still undiagnosed but suspecting hydrocephalus, or in need of a second opinion, start by asking friends, family and colleagues for help. They can do research at the library (a university or medical center library is best) or on the Internet; look up

all words related to your diagnosis. Gather the names of doctors who have written articles related to hydrocephalus. Research professional medical associations such as the American Association of Neurological Surgeons, Congress of Neurological Surgeons, American Academy of Neurology—often the websites of these types of organizations let you search for physicians by name or location. Contact support groups such as the Hydrocephalus Association for listings and directories of doctors who treat adults with hydrocephalus.

Do your homework before meeting with the doctor. Have your medical records with you, including copies of any scans or tests. Write down any and all questions that you have and don't be shy about asking the doctor about his or her experience with hydrocephalus in your age group and his or her protocols for diagnosis and treatment. Above all, take a friend or family member with you to all medical appointments—you will be collecting a lot of information and the person who goes with you can not only act as your advocate but be a second set of ears.

It's important to understand that hydrocephalus is a complex condition and the doctors may not be able to answer all your questions with certainty. What's necessary, however, is that you feel validated and respected—comfortable that the doctor is interested in having you as a patient and cares about your long-term health issues. Most people with hydrocephalus find that the neurosurgeon becomes the team leader, providing not only diagnostic expertise and surgical management but ongoing follow-up care as well. Whether it's the neurosurgeon, neurologist or a primary care physician, remember that the most important member of the health-care team is you.

4 ~ Social & Emotional Issues

WHILE our knowledge of infant/childhood-onset hydrocephalus has greatly expanded in the past decade, when it comes to adult forms of hydrocephalus, there is much less information. Social services, support systems, interventional and educational programs are in place for families of infants and children; for adults, there are few, if any, services available to help them with their complex needs.

A diagnosis of hydrocephalus in young adulthood to middle age can present serious psychosocial and emotional problems. If you are a parent, your parenting skills and confidence in these skills may be affected. Concerns about dying and leaving your children and their spouse without financial security and social support are real. On the flip side, other people may worry about being a burden on their families, relying upon aging parents, or busy siblings, for long-term care and support.

Not a Midlife Crisis

Anecdotal evidence and a few studies indicate that it is not uncommon for people with undiagnosed SHYMA to be told that their symptoms are “all in their head.” These symptoms, admittedly unusual in this age group, are frequently dismissed as manifestations of job dissatisfaction, midlife crisis, depression or other psychiatric problems. In fact, some people who have been diagnosed with SHYMA report feeling great relief that their complaints are validated—that their symptoms are real.

Workplace Concerns

The decline in cognitive functioning that occurs in many people with SHYMA often manifests itself in decreased job performance. Forgetfulness makes it harder to do a job right, as does the often-reported “mental dullness,” an unspecific but undeniably real symptom.

As if a growing difficulty performing one's job-related tasks were not enough, people with untreated SHYMA may also worry about the security of their job and their future abilities. Employers are not familiar with the syndrome, and may not readily believe that medical factors are behind the decrease in work performance, further adding to the tension in the workplace.

Once a diagnosis of SHYMA is made, you may still worry about your long-term health and ability to provide for yourself and your family.

Conclusion

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 families. 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 a national, 501(c)(3) nonprofit organization founded in 1983 to provide support, education and advocacy to individuals, families and professionals. Our goal is to provide comprehensive services that empower individuals and families to seek out the best medical care, programs and resources that will meet their needs now and in the future.

As the nation's most widely respected hydrocephalus support group, the Hydrocephalus 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. However, with early detection, effective treatment and appropriate interventional services, the future for individuals with hydrocephalus is promising. We invite your inquiries.

Glossary

- aqueductal stenosis:** a narrowing of the aqueduct of Sylvius. This is one cause of obstructive hydrocephalus; it may be treated using a CSF shunt or by a surgical procedure known as an endoscopic third ventriculostomy (ETV).
- arachnoid:** the middle layer of the meninges. It covers the brain and spinal cord smoothly without conforming to the irregularities of their surfaces; CSF flows within the arachnoid space.
- arachnoid villi:** small projections in the dura mater that project into the dural venous (blood) sinuses. CSF is reabsorbed from the arachnoid space by passing through the arachnoid villi and entering the venous system. Also known as arachnoid granulations.
- catheter:** flexible, hollow tube used to shunt fluid. For CSF shunting, the proximal catheter of a shunt is the inflow catheter and the distal catheter is the outflow catheter.
- cerebral aqueduct (aqueduct of Sylvius):** a narrow channel for CSF flow in the midbrain that connects the third and fourth ventricles.
- cerebrospinal fluid (CSF):** clear, colorless liquid secreted primarily by the choroid plexus and contained within the ventricles and the subarachnoid space. CSF functions primarily to float and cushion the brain and spinal cord.
- choroid plexus:** the structures in the lateral, third and fourth ventricles that produce cerebrospinal fluid.
- communicating hydrocephalus:** hydrocephalus in which the openings between the ventricular spaces and between the fourth ventricle up to the subarachnoid space are functioning.
- dura mater (or dura):** the outermost and heaviest layer of the meninges covering the brain and spinal cord; this layer is closest to the skull.
- foramen of Monro (interventricular foramen):** an opening between the lateral ventricle and third ventricle through which CSF flows from the lateral ventricle into the third ventricle.
- fourth ventricle:** a cavity within the brain that is situated between the brainstem and the cerebellum. The fourth ventricle receives CSF from the cerebral aqueduct, and CSF exits the fourth ventricle via the foramina of Luschka and Magendie into the subarachnoid space.

hematoma: a localized collection of blood, usually clotted.

hemorrhage: the escape of blood from blood vessels.

hydrocephalus: an abnormal condition that occurs when there is an imbalance between the rate of CSF production and the rate of absorption, leading to gradual accumulation of CSF.

intraventricular hemorrhage: bleeding into the ventricles.

lateral ventricle: one of two normal cavities within the cerebral hemispheres that contains cerebrospinal fluid. CSF flows from the lateral ventricles into the third ventricle via the foramen of Monro.

meninges: membranous coverings of the brain and spinal cord consisting of the dura mater, arachnoid and pia mater.

meningitis: inflammation of the meninges. Meningitis can result from bacterial or viral infection. Scarring of the arachnoid that results from meningitis can restrict or block CSF flow and absorption.

noncommunicating hydrocephalus: hydrocephalus in which there is obstruction of the flow of CSF through the cerebral aqueduct or from the fourth ventricle to the subarachnoid space.

normal pressure hydrocephalus (NPH): a syndrome characterized by enlarged ventricles and a triad of symptoms including gait disturbance, dementia and impaired bladder control; a form of hydrocephalus that occurs most often in middle-age and older persons.

obstructive hydrocephalus: hydrocephalus caused by a blockage along the CSF flow pathway.

third ventricle: a midline cavity within the brain that is situated between the right and left thalamus. It receives CSF from each lateral ventricle via the foramen of Monro, and CSF exits the third ventricle via the cerebral aqueduct (aqueduct of Sylvius).

third ventriculostomy: a surgical operation to create an opening through the membranous floor of the third ventricle, permitting CSF to exit the third ventricle directly into the subarachnoid space at the base of the brain.

ventricle: a cavity within the brain that contains cerebrospinal fluid.

Resources

The Hydrocephalus Association is a national, 501(c)(3) nonprofit organization founded in 1983 to provide support, education and advocacy to families, individuals and professionals. Our goal is to provide 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.

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.

Resources

About Hydrocephalus—A Book for Families (English or Spanish)

About Normal Pressure Hydrocephalus—A Book for Adults and Their Families

Prenatal Hydrocephalus—A Book for Parents

A Teacher's Guide to Hydrocephalus

Directory of Neurosurgeons

LINK Directory

Quarterly Newsletter

The Resource Guide

Fact and Information Sheets

Annual Educational Scholarships

Annual Neurosurgical Resident's Prize

Biennial National Conference for Families and Professionals

Hydrocephalus Association

serving individuals, families and professionals since 1983

870 Market Street • Suite 705 • San Francisco, CA 94102

Tel. 415-732-7040 • Toll-free 888-598-3789 • Fax 415-732-7044

Email: info@HydroAssoc.org • Web: www.hydroassoc.org

■ **HYDROCEPHALUS** ■
ASSOCIATION

870 Market Street ■ Suite 705 ■ San Francisco, CA 94102
Tel. 415-732-7040 ■ Toll-free 888-598-3789 ■ Fax 415-732-7044
Email: info@HydroAssoc.org ■ Website: www.hydroassoc.org