



About Normal Pressure Hydrocephalus (NPH)

A Book for Adults and Their Families



About Normal Pressure Hydrocephalus—A Book for Adults and Their Families was written for adults with NPH, their families, friends and caregivers with the intention of providing information about the diagnosis and treatment of adult-onset normal pressure hydrocephalus (NPH). It is a companion piece to our booklet *About Hydrocephalus—A Book for Families*, the most widely distributed resource on infant and childhood hydrocephalus in the United States.

It is our belief that individuals and families dealing with the complex issue of adult-onset NPH must educate themselves 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. For making clinical decisions, patients must rely on the guidance and recommendations of the clinicians providing their care.

Foreword

Hydrocephalus is a condition characterized by the expansion of the cavities or ventricles in the brain, caused by an abnormal accumulation of cerebrospinal fluid (CSF). Hydrocephalus can develop with high or normal CSF pressure. Normal pressure hydrocephalus (NPH), originally described by Salomon Hakim, MD, PhD, in 1964, is a clinical condition which principally affects the elderly. It is characterized by a triad of symptoms: motor disturbances (mostly gait impairment), incontinence and dementia, associated with ventricular enlargement in the absence of elevated intracranial pressure. The diagnosis of NPH does not require all three symptoms to be present.

Almost 50 years after the original description of NPH, there still remain many unknowns, including its diagnostic criteria. It is not uncommon to find many of the symptoms of NPH in the elderly. These may occur as an isolated finding or associated with other diseases. Changes in some of the intellectual functions are expected to occur during the process of aging in the same way that other physiologic functions of the body become altered with advancing age. In spite of all this, NPH is a known and unique clinical entity justifying its own differential diagnosis with other brain atrophies. There is great importance in identifying patients who have NPH and had previously been diagnosed as hopeless cases of degenerative brain disease, Alzheimer's or Parkinson's disease, since there is the opportunity to provide them with a treatment for NPH that will allow them to have a better quality of life.

During the last years, much has been learned to help provide a more accurate diagnosis and better treatment for NPH. The families of patients who might have NPH should be well informed of the symptoms which are characteristic of this clinical entity, since it is with them that the process of making a diagnosis starts. The families should be encouraged to take a first step, since many patients with NPH have experienced "miraculous" recoveries after treatment and are now living much fuller lives.

My family has been very involved in the field of hydrocephalus for many years and we have seen many persons recover dramatically after being treated for NPH. The most rewarding experience has always been to help someone with NPH. Even though sometimes a patient might not recover after treatment as was expected, the results are many times very rewarding and definitely worth the effort. As has been previously said, “There is life after NPH.”

—Carlos Hakim, PhD

Because some of the symptoms of NPH can appear similar to Alzheimer’s or Parkinson’s or are often associated with the aging process, NPH can be misdiagnosed or go undiagnosed for years. Milt Newman went 13 years before being diagnosed with NPH and successfully treated with a shunt. “The difference in how I feel is like night and day. Now I’m living; I wasn’t living before. I waited for my wife Phyllis to do everything for me. Now I can do things I couldn’t do before.”

—DR. MILT NEWMAN



What Is Hydrocephalus?

Hydrocephalus is a condition characterized by an abnormal accumulation of cerebrospinal fluid (CSF) or “spinal fluid” within cavities called ventricles that are inside the brain. CSF surrounds the brain and spinal cord. The functions of CSF include physical support or cushioning of the brain, excretion of some waste products and distribution of important substances within the central nervous system. The average adult produces about one pint (500 cc) of clear spinal fluid daily. When the circulatory path of the CSF is blocked, fluid begins to accumulate, causing the ventricles to enlarge and the pressure inside the head to increase, resulting in hydrocephalus.

What Is Normal Pressure Hydrocephalus?

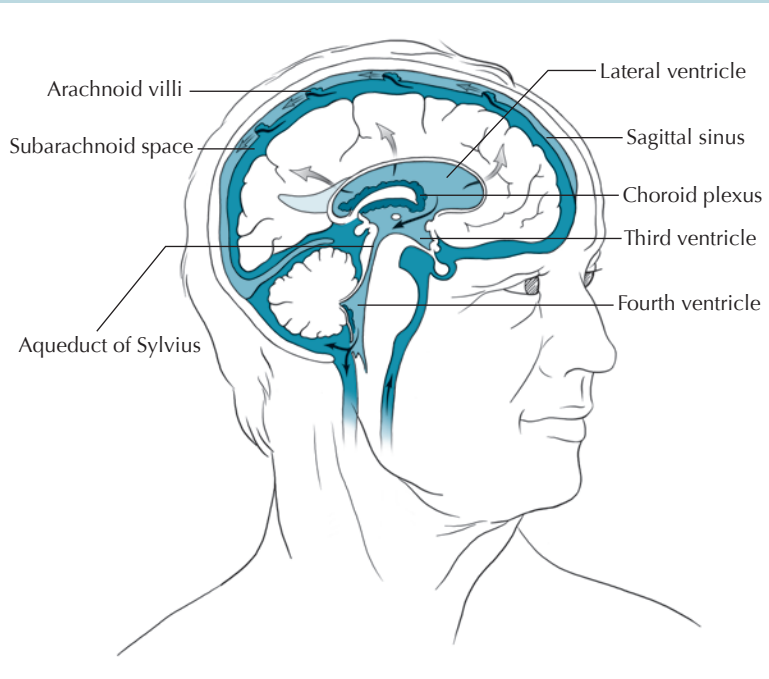
Normal pressure hydrocephalus (NPH) is an accumulation of cerebrospinal fluid that causes the ventricles in the brain to become enlarged, sometimes with little or no increase in intracranial pressure (ICP). It is most commonly seen in older adults, and is accompanied by some or all of the following triad of symptoms:

- Gait disturbances
- Mild dementia
- Impaired bladder control

In most cases of NPH, it is not clear what causes the CSF absorptive pathways to become blocked.

The name “normal pressure hydrocephalus” came out of Dr. Salomon Hakim’s 1964 paper describing certain cases of hydrocephalus where a triad of neurologic symptoms occurred in the presence of “normal” CSF pressure. This was before continuous pressure-recording techniques were available. We now know that the phrase “normal pressure” is misleading, because many patients have fluctuations in CSF pressure ranging from high to nor-

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.



mal to low. However, normal pressure hydrocephalus, or NPH, continues to be the common name for the condition.

What Causes Normal Pressure Hydrocephalus?

The majority of cases of NPH are **idiopathic**, meaning of unknown cause (also known as **primary NPH**).

NPH can also develop as the result of a known cause, in which case it is referred to as **secondary NPH**. Some of these causes are head injury, cranial surgery, subarachnoid hemorrhage, tumor or cysts, as well as subdural hematomas, bleeding during surgery,

meningitis and other brain infections. All of these predisposing conditions can cause inflammation that affects the CSF pathways, impeding CSF flow.

What Are the Symptoms?

Normal pressure hydrocephalus is usually characterized by a triad of symptoms: gait disturbance (difficulty walking), mild dementia and impaired bladder control. These symptoms may not occur all at the same time, and sometimes only one or two symptoms are present.

- **Gait disturbances** range in severity from mild imbalance to the inability to stand or walk at all. Gait is described as a “magnetic gait,” often wide-based, short-stepped, slow and shuffling. People with NPH may have trouble picking up their feet, making stairs and curbs difficult and frequently resulting in falls. They may also have difficulty turning around, and turn very slowly with multiple little steps. Gait disturbance is often the most pronounced symptom and the first to become apparent.
- **Mild dementia** can be described as a loss of interest in daily activities, forgetfulness, difficulty dealing with routine tasks and



“The placement of my shunt provides me with new hope, physical balance and freedom from falling, allowing me to live a normal lifestyle including visiting family, travel and serving as an arbitrator.”

—DR. JAMES O’GRADY

short-term memory loss. The cognitive symptoms associated with NPH are usually less severe than full-blown dementia, and are often overlooked for years or accepted as an inevitable consequence of aging. People with NPH do not usually lose language skills, but they may be less aware of their deficits than those around them, and may even deny that there are any problems. Not all individuals have an obvious cognitive impairment. In mildly affected cases, conversational skills may be preserved and thinking abilities may be relatively unchanged. In some cases, cognitive changes may only be detectable with formal neuropsychological testing.

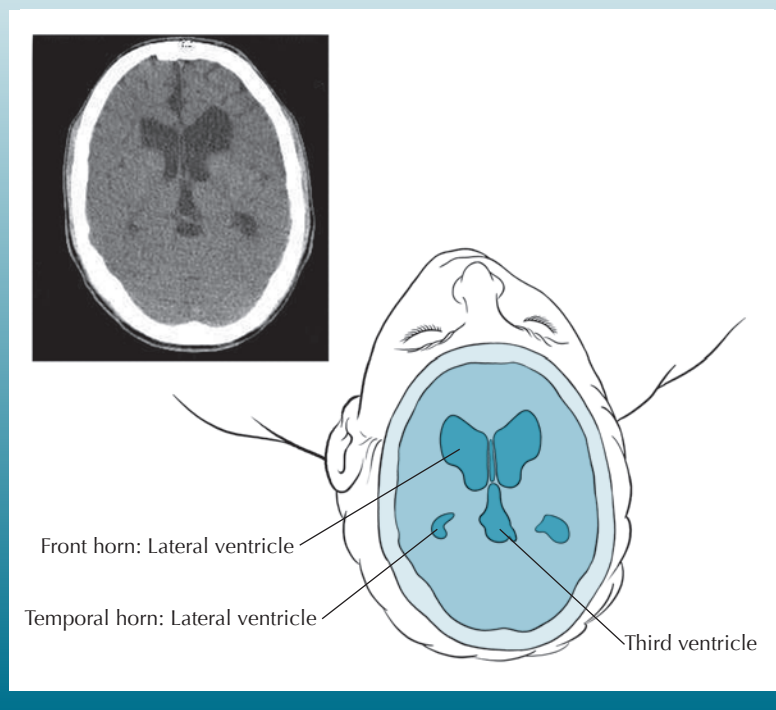
- **Impairment in bladder control** is usually characterized by urinary frequency and urgency in mild cases, whereas a complete loss of bladder control (urinary incontinence) can occur in more severe cases. Urinary frequency is the need to urinate more often than usual, sometimes as often as every one to two hours. Urinary urgency is a strong, immediate sensation of the need to urinate. This urge is sometimes so strong that it cannot be held back, resulting in incontinence. In very rare cases, fecal incontinence may occur. Some people never display signs of bladder problems.

Because the triad of symptoms are often associated with the aging process in general, and a majority of the NPH population is older than 60 years, people often assume that they must live with the problems and adapt to the changes occurring within their bodies as they age.

Symptoms of NPH can also resemble those of other conditions affecting the elderly. For example, the cognitive deficits of NPH can resemble those associated with early Alzheimer's, and the gait disturbances of NPH can look similar to those of Parkinson's. (In some cases, NPH can occur in combination with these diseases.)

There are challenges to distinguishing disease processes that mimic NPH symptoms. Any type of senile or pre-senile dementia,

View of enlarged ventricle: CT scan (top); orientation of CT scan in the head (bottom)



including Alzheimer's, may be associated with atrophy or shrinking of the brain, resulting in large CSF spaces that are visible on MRI and CT scans. Because the appearance is quite similar to NPH, differentiating these diseases can be very difficult. People with Parkinson's may have the typical gait disturbance, dementia and incontinence associated with NPH, but they rarely have the enlarged CSF spaces. People with spinal stenosis, a condition in which the nerves supplying the bladder and lower limbs are compressed from arthritic changes in the lumbar spine, may have incontinence and gait disturbance. However, they do not necessarily have enlarged CSF spaces.

Symptoms can be present for months or even years before a

person sees a physician. The symptoms of hydrocephalus seem to progress with time. The rate of progress is variable, and it is often a critical loss of function, or disability, that brings a person to seek evaluation and treatment. It seems that the longer and more severe the symptoms, the less likely it is that treatment will be successful. As a general rule, the earlier the diagnosis, the better the chance for successful treatment; however, some patients who have had symptoms for years can improve with treatment.

How Is Normal Pressure Hydrocephalus Diagnosed?

It is often the affected person or close family member who first brings the symptoms of NPH to the attention of a doctor. Occasionally, enlarged ventricles are discovered on a brain image performed for another purpose. Once NPH is suspected by a primary physician, a variety of tests may be recommended to confirm the diagnosis and assess the person's candidacy for shunt treatment.

At this point, it is important that a neurosurgeon and/or a neurologist (or neuropsychologist) become part of the medical team. The involvement of these medical professionals 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 and risks of surgery.

The decision to order a given test may depend on the specific clinical situation, as well as the preference and experience of the medical team. Not all of the tests described here need to be performed in order to make a diagnosis.

Clinical exams to evaluate symptoms consist of an interview and/or a physical/neurologic examination. In a clinical examination, a doctor might:

- discuss and observe walking and turning to determine the extent and type of gait disturbance;

-
- assess cognition by asking a few questions or administering a full neuropsychological evaluation using pencil-and-paper tests to probe such qualities as attention, reaction time, memory, reasoning, language and emotional state; and
 - verbally assess urinary urgency and frequency or incontinence.

The presence of all three symptoms is not necessary to the diagnosis of NPH. The medical team will consider the pattern and severity of impairments, along with results of other tests mentioned below, in differentiating NPH from other conditions.

Brain images to detect enlarged ventricles commonly include magnetic resonance imaging (MRI) and computerized tomography (CT). Each of these imaging techniques is described in more detail below.

- **MRI** uses radio signals and a very powerful magnet to create a picture of the brain. It is safe for most people, reliable and painless, but takes longer than a CT scan and some may find the physically confining circumstances of being in an MRI machine challenging. MRI can detect enlarged ventricles as well as evaluate the CSF flow and provide information about the surrounding brain tissues. MRI scans can also assess how fast CSF moves through a particular part of CSF pathways called the cerebral (or Sylvian) aqueduct (the CSF flow void sign). Some physicians believe that high CSF flow through the aqueduct predicts improvement with treatment of NPH. The MRI provides more information than the CT, and is therefore the test of choice in most cases, but people with cardiac pacemakers or certain other metallic implants may not be able to have MRI scans because of potential interference with these devices.
- **CT** (or CAT) scan is a picture of the brain created by using X-rays and a special scanner. An X-ray beam passes through

the head, allowing a computer to take a picture of the brain. It is safe, reliable, painless and relatively quick (about 15 minutes). A CT scan will show if the ventricles are enlarged.

CSF tests to predict shunt responsiveness and/or determine shunt pressure include lumbar puncture, external lumbar drainage, measurement of CSF outflow resistance, intracranial pressure (ICP) monitoring and isotopic cisternography. Though there is no way to accurately predict a particular patient's responsiveness to a particular shunt, many doctors find these tests helpful in determining the likelihood of a positive response to shunting. Each test is described in more detail below. People who have abnormal bleeding tendencies or take medications that affect bleeding should talk with their medical team about any special precautions before invasive procedures are performed.

- **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 lower back. Up to 50 cc of CSF is removed to see if symptoms are temporarily relieved by this CSF volume reduction. If removal of some CSF dramatically improves symptoms, even temporarily, then surgical treatment is likely to be successful. A limitation of lumbar puncture and removal of a small volume of CSF as a screening test for NPH is that some people may have little or no improvement after the test, and yet may still improve with a shunt. When the response to a lumbar puncture is “negative” or uncertain, further evaluation may be helpful.
- **External lumbar drainage, also called lumbar catheter insertion or continuous lumbar drainage**, is a variation of the lumbar puncture where a thin, flexible tube, called a catheter, is left in place to drain CSF. The procedure, which is performed in the hospital, allows for either intermittent or continuous removal of spinal fluid over several days to imitate the effect that

a shunt would have. It also allows for more accurate recording of CSF pressure. With an intermittent drainage protocol the person is free to move around when the fluid is not being drained.

Spinal fluid drainage over time can be thought of as a “test drive” of a shunt without actually undergoing shunt surgery. However, because it requires hospitalization and has associated risks it may not be recommended for all patients. People who respond dramatically to such spinal fluid drainage are likely to respond to shunt surgery, which will be necessary for long-term treatment of NPH. Some physicians advocate using the pressure results for selecting the type of shunt or initial shunt setting for programmable and adjustable shunts.

- The **measurement of CSF outflow resistance** is a more involved test that requires a specialized clinical setting. This test begins with a lumbar tap and assesses the degree of blockage of 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, since the shunt mimics the function of the body’s normal CSF drainage pathways.
- For **intracranial pressure (ICP) monitoring** or **spinal pressure monitoring**, a small pressure monitor is inserted through the skull into the brain or ventricles or in the lumbar region to measure the ICP. Pressure monitoring, either by the lumbar catheter or the intracranial method, requires admission to a hospital. It can detect an abnormal pattern of pressure waves as well as low or high pressure. It is possible for NPH to occur even when CSF pressure is not measurably high. The results of this test can also be used to select initial shunt pressure if a shunt is being implanted.

- **Isotopic cisternography** is no longer in frequent use because a “positive” cisternogram result does not reliably predict whether a patient will respond to implantation of a shunt, and results are often ambiguous. It involves having a radioactive isotope injected into the lower back through a spinal tap, in order to monitor the absorption of CSF over a period of several days. This test is done in the hospital.

What Treatment Is Available?

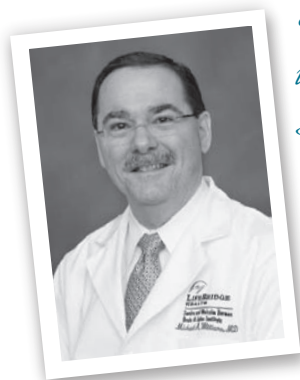
The most common and usually the only available treatment for NPH is the surgical implantation of a **shunt**, a device that channels CSF away from the brain to another part of the body where it can be absorbed. Most shunt systems consist of three components: (1) a collection catheter situated within the cerebral ventricles or the lumbar spinal canal; (2) a valve mechanism to control how much CSF flows; and (3) an exit catheter to drain the CSF to another part of the body where it can be absorbed (see drawing on page 18). The most common part of the body for drainage is the peritoneal (abdominal) cavity, and the most common system is a **ventriculoperitoneal (VP)** shunt (from the cerebral ventricle to the peritoneum). The drainage catheter can also be placed in a vein that leads to the heart, a configuration called a **ventriculoatrial (VA) shunt**. After surgery is completed, all components of the shunt system are entirely under the skin, and nothing is exposed to the outside.

The shunt valve is a critical component of the shunt system. The design that has been in use the longest is a fixed **differential-pressure (DP) valve**. This valve opens when the fluid pressure at the inlet of the valve exceeds the pressure at the outlet by a certain amount. Traditionally, DP valves have come in low, medium and high pressure varieties. For the adult with hydrocephalus, sometimes the valve is overwhelmed by the effects of gravity, leading to the drainage of too much CSF. This is commonly known as siphoning. Siphoning is sometimes asymptomatic, sometimes associated with headaches or nausea in the upright position, and in

some instances, can cause so much spinal fluid to drain from the head that small blood vessels between the brain and skull are disrupted, causing a type of bleeding known as subdural hematoma, a serious complication of shunting.

In order to counteract this potential problem, valve mechanisms have been designed that incorporate **anti-siphon** and **gravity-compensating mechanisms** or **flow-regulated mechanisms**. Anti-siphon devices are triggered by pressure change; gravity-compensating devices are triggered by postural change. Anti-siphon devices are not effective with lumbar shunts, but gravity-compensating devices are. Both devices are designed to counteract overdrainage of CSF when sitting or standing.

The **flow-regulated valve** is designed to minimize overdrainage by limiting flow to the approximate rate of CSF production when conditions of overdrainage occur (when sitting, standing or straining). This valve acts like a differential-pressure valve when the risk of over-



“Caring for patients with NPH and their families is a rich and rewarding experience. One of the greatest joys for me is when an elderly couple comes back after the shunt operation and tells us, ‘You’ve given us our lives back’ because they can now travel, and socialize, and enjoy their families in ways they couldn’t before the NPH was treated. And for those patients we evaluate who turn out not to have NPH, while we’re disappointed that we can’t help them get better, they and their families are almost always grateful that we undertook the time and effort to investigate the possibility.” —MICHAEL WILLIAMS, MD

Examples of Shunt Valves



ProGAV
Aesculap, Inc.



Hakim Programmable Valve
Codman, a Johnson and Johnson Company



Diamond Valve
Vygon Neuro



Polaris Adjustable Valve
Sophysa



OSV® SmartValve™
Integra



Strata Adjustable Valve
Medtronic Neurosurgery

(in the same spinal region where a spinal tap is done). The tubing, which is narrower than that used in ventricular shunts, is tunneled under the skin to the abdomen, where it is inserted into the peritoneal cavity, much like a VP shunt. Some people who are particularly apprehensive about the insertion of a ventricular shunt through the brain may be more comfortable with an LP shunt, yet it is worth noting that insertion of a ventricular shunt rarely harms the brain. Although LP shunts may have some potential advantages, in general they are more prone to obstruction over the long term and if there is a problem with the shunt, it is more difficult to assess what the problem is.

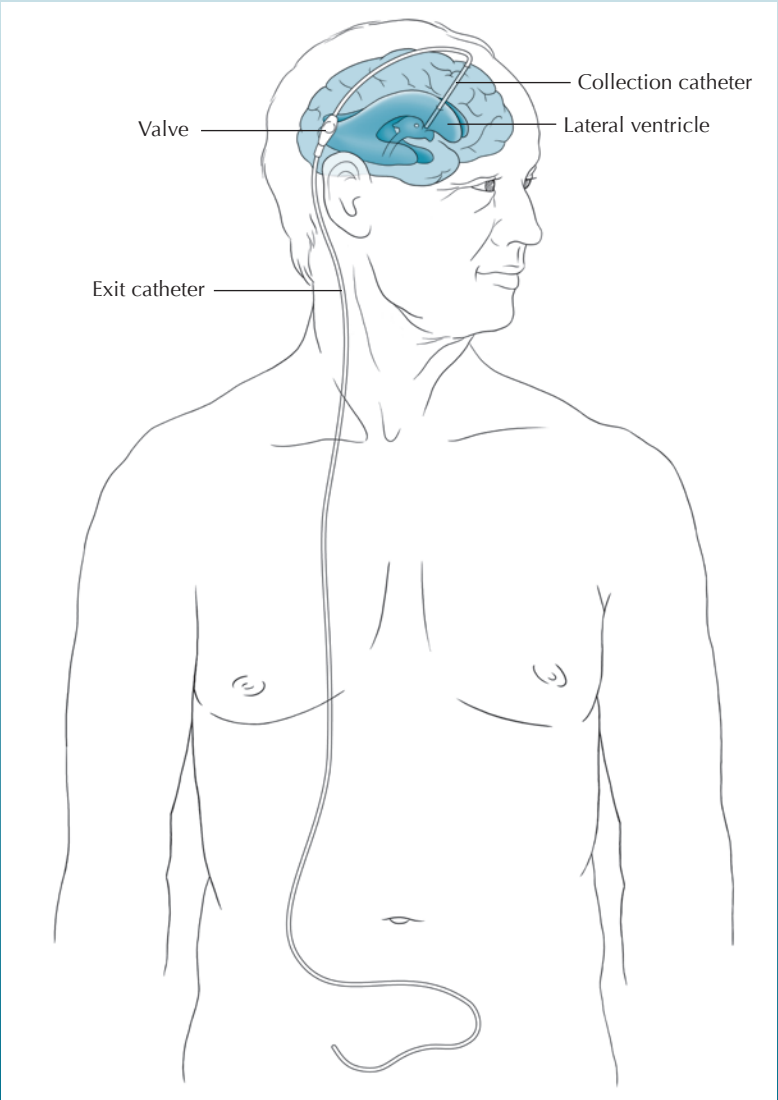
For people who have aqueductal stenosis, a surgical procedure called **endoscopic third ventriculostomy (ETV)** may be considered as an alternative to a shunt. In this procedure, the neurosurgeon uses a special endoscope to create an alternative CSF passageway that bypasses the obstruction at the cerebral aqueduct. The determination of aqueductal stenosis can be made by MRI. The success of ETV in adults is variable, and some people who have a third ventriculostomy may later require shunt surgery to treat their symptoms. Clinical trials are currently exploring ETV as a treatment for NPH.

Will Shunting Help?

This question cannot be answered definitively. Many tests and evaluation criteria have been proposed, but, unfortunately, no one single factor is reliable in predicting success from implantation of a shunt for NPH. The following findings are generally associated with a better outcome following shunt placement:

- The onset of gait disturbance as the first and most prominent symptom
- A known cause for NPH, such as trauma or hemorrhage
- The scan (MRI or CT) shows the ventricular size to be disproportionately larger than the CSF in the subarachnoid space

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 abdominal (peritoneal) cavity.



- Removal of spinal fluid via lumbar puncture or lumbar catheter gives dramatic, temporary relief of symptoms
- ICP or spinal fluid pressure monitoring shows an abnormal range or pattern of spinal fluid pressure or an elevated CSF outflow resistance
- Minimal evidence of disease of the small blood vessels nourishing the brain

Because some people with NPH have additional medical or neurological problems, it is important for them, their families and their neurosurgeons to discuss their expectations of shunt surgery. Does “success” mean that they will regain the levels of motor skill or mental ability they had before the symptoms presented themselves? Does it mean that the condition will not worsen? Or does it mean something else? The definition of success must be individualized, and it is important to know that it is possible for any or all of the hydrocephalus symptoms to improve with shunting. One way to evaluate the success of shunt surgery is to consider whether it has reduced the disabilities that were present before surgery and increased the individual’s functional abilities.

Although everyone hopes for a complete recovery, it is not often seen, and many individuals and their families are satisfied when shunt surgery results in reduced disability or dependence than they had before surgery, or prevention of further neurological deterioration. People with NPH, their families and their physicians need to be supportive and hopeful, but they should also know the possible complications, risks and realities of shunt implantation.

Not all people with enlarged ventricles need treatment. Some people with enlarged ventricles have no symptoms and no neurologic deficits at all, even when evaluated by neurosurgeons and neurologists who specialize in hydrocephalus. This is a condition often called “compensated hydrocephalus.” People who have compensated hydrocephalus cannot be “made better” by treating the hydrocephalus, and in such circumstances, there are no ben-

efits that would offset the potential risks of treatment (see below). People with compensated hydrocephalus may, however, develop symptoms later in their lives, and may benefit from treatment with shunting at that time. Therefore, it is important for individuals with compensated hydrocephalus to see a neurosurgeon or neurologist periodically to assess whether subtle symptoms are developing.

What Does a Shunt Operation Entail?

VP: The surgical procedure to implant a VP shunt usually requires less than an hour in the operating room. After the patient is placed under general anesthesia, his or her scalp overlying the shunt insertion site is shaved and the patient is scrubbed with an antiseptic from the scalp to the abdominal area. These steps are taken in order to reduce the chances of an infection. Small incisions are made in the head and in the abdomen to allow the neurosurgeon to pass the shunt tubing through the fatty tissue just under the skin. A small hole is made in the skull, opening the membranes between the skull and brain to allow the ventricular end of the shunt to be passed through the brain and into the lateral ventricle. The abdominal (peritoneal) end is passed through a small opening into the abdominal cavity where the excess CSF will eventually be absorbed. The incisions are then closed and sterile bandages are applied.

LP: Under general anesthesia, the LP shunt catheter is inserted using a hollow needle between two vertebrae in the lumbar CSF region of the spine. The valve is implanted below the skin and the drainage catheter is directed into the peritoneal cavity. Once in place the LP shunt drains the excess cerebrospinal fluid removed from the lumbar region and transports it to the peritoneal cavity, where it is eventually absorbed by the organs and passed out of the body during urination. Patients with lumboperitoneal shunts are left with two scars: a vertical scar down the lower (lumbar) part of the spine and a horizontal scar across the upper abdomen.

The patient stays under careful neurological observation for the



“Following a stroke at the age of 59, I was eventually correctly diagnosed with NPH after seeing my family doctor, a psychologist “and taking several meaningless tests and wasting about a month. Once a neurologist made the correct diagnosis, a shunt was installed and my symptoms immediately disappeared.”

—MARTIN MITTLEMAN

first 24 hours following a procedure. The incisions are monitored for signs of infection. The patient will stay in the hospital from one to seven days. Follow-up visits will be necessary to check post-operative status and resolution of symptoms. After surgery, physical therapy, occupational therapy and other rehabilitation strategies may be advised to help patients attain as much resolution of symptoms as possible. People should talk with their neurosurgeon about his or her particular protocols following surgery.

How Successful Is Shunting?

The symptoms of gait disturbance, mild dementia and bladder control problems may improve within days of shunt surgery, or may take weeks to months to abate. There is currently no way to predict how fast, or to what extent, this improvement will occur.

For those who do improve, changes are often seen in the first weeks, although there are late responders and some people take longer to recover from surgery. In addition, this improvement may range from mild to dramatic. It is not possible to predict how long the improvement will last, as the course of clinical improvement varies for each person. Some people seem to reach a plateau, while others improve for months but then seem to decline again. Unfortunately, there are no guarantees. A recurrence of symptoms in a

person who had improved should prompt consideration of shunt malfunction or one of the complications described below.

The rate of success for shunting normal pressure hydrocephalus is quite variable. Neurosurgeons do not agree on the factors that lead to a successful procedure, nor do they have similar rates of success. Although the success rate for shunting is higher when proper diagnostic and treatment procedures are followed, it is not possible to predict how much a patient will improve after surgery. It is important to note that if initial success is followed by a recurrence of symptoms, it may be due to a valve or shunt failure, or the need for a lower pressure valve—rather than failure of the procedure.

The presence of other neurological or medical conditions may also affect the outcome after shunt surgery. One of the most common is the long-term effect of high blood pressure on the brain, which can cause multiple tiny strokes to the same areas of the brain that are affected by hydrocephalus, causing virtually the same symptoms. Other factors to consider in recovery are arthritis involving the back, hips or knees; impaired sensation in the legs and feet (neuropathy); other causes of urinary dysfunction; or the presence of Alzheimer's disease. The issue is often not whether the symptoms are caused by one diagnosis or another, but how much is caused by one diagnosis and how much by another. The more the symptoms are due to NPH, rather than the associated conditions, the greater the likelihood of successful recovery after shunting.

What Are the Complications and Risks Involved with Shunting?

Although the insertion of a shunt is a relatively simple neurosurgical procedure, the decision to undergo surgery should not be taken lightly. The treatment of normal pressure hydrocephalus carries greater risks compared to the treatment of hydrocephalus in children; therefore, this operation should be undertaken only if the degree of disability or the progression of the disorder warrants

Hazel's Story

"My mom is a classic case of misdiagnosis for NPH. Her symptoms began to appear in 2007. The doctors thought she had a dropped bladder and suspected a stroke, but there was no evidence of either. Later on, they diagnosed her with Parkinson's then with Alzheimer's disease. I had done some volunteer work in nursing homes with Alzheimer's and Parkinson's patients and I knew my mom didn't act like a patient with either one.



"During my dad's illness and after his death, my mom came to live with me. I set her up with a general practitioner and she agreed to send my mother to a neurologist to run further tests. Finally the neurologist diagnosed my mother with normal pressure hydrocephalus and they referred her for surgery. It took us a while to find the right surgeon, but when we finally went to the Cleveland Clinic and met Dr. Mark Luciano, my mom fell in love. With her permission, the surgery was performed by Dr. Luciano over the live video feed as part of the 11th National Conference on Hydrocephalus.

"Mom still gets frustrated because the change is taking time. But for me it's remarkable. It's as if she went back in time five years and each day she improves a little more. She hasn't cooked in about eight years, but last night she said, 'I think it is about time I start cooking again—I think I'll start with something easy, shrimp dip.' I know it doesn't sound like much to most people, but to me, it's amazing." —RICHARD CHARD

such intervention. An additional factor to consider is whether the expected benefits of surgery outweigh the risks. Ways to assess the expected benefits include diagnostic tests that reveal information about the abnormal CSF flow that the shunt is intended to correct, such as the response to CSF removal, abnormal CSF pressure patterns or insufficient CSF absorption. Most people feel more comfortable proceeding with shunt surgery if they have good reason to expect a favorable outcome from the surgery.

The potential complications of shunt surgery include those related to the actual operation, as well as those that may occur days to years later. Unlike many other operations, in which the surgical risks are highest during the operation, most of the problems associated with shunting can occur weeks or even years after the surgery.

One of the most common problems with shunt systems is obstruction (the shunt becomes clogged). It is not possible to predict which patients will have a shunt obstruction, nor when the obstruction will occur. A shunt obstruction is usually suspected when the original symptoms of NPH reappear over a period of days, weeks or months. Fortunately, shunt **obstruction** in NPH is usually easily fixed and rarely results in serious problems, although further surgery may be necessary.

Another common complication of shunt surgery is **shunt malfunction**, which can occur when either end of the shunt is incorrectly positioned, the valve fails to function properly or the CSF is not efficiently reabsorbed. The typical remedy involves a further surgical procedure.

It is important for people with NPH and their families to know the symptoms they experienced prior to shunt implantation and to alert their physician if the symptoms return after the shunt is implanted, as this may be a sign of an obstructed or malfunctioning shunt.

Additional complications include an **infection** involving the surgical wound, the shunt or the CSF (meningitis); **bleeding** into the brain or ventricles; or a **seizure**. A shunt infection may be in-

icated by fever, redness or swelling along the shunt track. In very rare instances, a person can have a reaction to the implanted shunt materials. Fortunately, these complications are uncommon and can be managed successfully in almost all cases, although treatment may require additional surgery.

One of the more serious complications that can occur following insertion of a shunt is a **subdural hematoma** (blood clot). The risk of a subdural hematoma in people with NPH and a shunt is approximately 5 to 10 percent. Because most shunts drain CSF from the center of the brain (the ventricles), this may cause the surface of the brain to pull away from the skull, thus stretching and tearing blood vessels that go from the scalp to the surface of the brain. This can sometimes be seen on a CT scan as a fluid space between the brain and the skull called a hygroma. Although a hygroma may not have any clinical symptoms, it may increase the risk of hematoma. The symptoms of a subdural hematoma may vary from headache to paralysis or even coma or death. Shunt-related subdural hematomas most commonly occur following a fall, even a minor one with no apparent injuries at the time of the fall.

The interval between the fall and the onset of symptoms can be as long as days or weeks. Subdural hematoma can also be caused by overdrainage, when a shunt drains too much CSF. Therefore, people with NPH should not hesitate to seek medical attention if they develop worrisome symptoms.

In patients who have had multiple abdominal operations, there is a rare complication of injury to the bowel.

Given these potential complications, individuals and their families need to assess their own situations to determine if the possible benefits of surgery outweigh the possible risks.

NPH Left Untreated

People with NPH usually present with progressive symptoms, and there is no reason to believe that the progression will stop on its own. No one is able to predict how fast the symptoms

“My wife and I did quite a bit of research once I made the decision to have the surgery, which precipitated many questions for my surgeon. Confidence in my surgeon by both my wife and I was crucial, and I must say that my surgeon is the person who finally put me at ease with the surgery along with talking to a person who already had the procedure and could give me some insight as to what I could expect afterward. The surgery was a success and my recovery from it was relatively brief (a few weeks).” —PETER MORRIS



will progress, and the seriousness of symptoms may vary day to day. Evidence suggests that the longer the symptoms have been present and the more severe the symptoms are, the less likely it is that the treatment will be successful. As a general rule, the earlier the diagnosis and treatment, the better the chance of recovery. However, some patients who have had symptoms for years can improve with treatment. For patients whose symptoms are very mild, their doctor may advocate closely monitoring the clinical condition without proceeding immediately to a shunt operation.

The Importance of Family Support

The process of diagnosing possible normal pressure hydrocephalus can often be frustrating. The symptoms of gait disturbance, mild dementia and bladder control problems can also occur with a number of other conditions that affect people over 60. Sometimes these conditions coexist with hydrocephalus, making the diagnosis and treatment even more difficult. Families and individuals who stay informed and ask questions of their physicians will likely feel more involved in the management of their care. It is important to be op-

timistic about surgical treatment. We live in a society where people are living longer, and more and more adult children are being asked to care for their aging parents. The frustration and complexity of being ill, or caring for an ill parent or spouse, can be stressful for all involved. Adults with hydrocephalus may resent being dependent on their spouse, children and family. Families often feel better and cope better by acknowledging and discussing their emotional responses to a chronic, and possibly disabling, condition such as hydrocephalus.

What Can We Expect for the Future?

There is currently a great deal of interest in the diagnosis and management of NPH, with the expectation that the reported incidence of this condition will increase dramatically as the baby boomers reach retirement age and beyond. Physicians, medical-device manufacturers and patient advocates are cooperating on a variety of levels to improve methods of diagnosis, techniques and devices for treatment, and public awareness of NPH. It is anticipated that these efforts will result in years of better quality of life for people affected by this condition.

Conclusions

The Hydrocephalus Association is positioned at the nexus of a movement to strategically drive hydrocephalus research. We are aligned with professionals 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.

There are many unknowns surrounding the diagnosis and treatment of adult-onset normal pressure hydrocephalus. Although the success rate for shunting is higher when proper diagnostic and treatment procedures are followed, it is not possible to predict the degree of improvement that will follow shunt surgery. However, NPH is not a hopeless condition. Advances in shunt technology and surgical techniques are continually being

introduced and developed. Adults diagnosed with normal pressure hydrocephalus, and their families, should be encouraged to ask questions, gather information and network with others.

The Hydrocephalus Association was formed to support and foster these goals by bringing together individuals and families such as yours. Because normal pressure hydrocephalus can be a complex medical condition, it is essential that everyone involved learn as much as possible about the particular case so that informed decisions can be made.

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 called 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.

communicating hydrocephalus: Hydrocephalus in which the openings between the ventricular spaces and between the fourth ventricle up to the subarachnoid space are functioning.

choroid plexus: The structures in the lateral, third and fourth ventricles that produce cerebrospinal fluid.

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 brain stem 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.

hygroma: A sac, cyst or bursa distended with fluid. A subdural hygroma is a collection of fluid between the brain and the skull.

intraventricular hemorrhage: Bleeding into the ventricles.

lateral ventricle: One of two normal cavities within the cerebral hemispheres that contain 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; this 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; 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.

Index

- abdominal cavity, 13, 17, 19
- adjustable valve. *See* valves
- aging, 2, 3, 7, 26
- Alzheimer's disease, 2, 3, 7, 8, 21, 22
- anesthesia, 19
- anti-siphon devices. *See* valves
- aqueduct of Sylvius, 5, 10, 28, 30
- aqueductal stenosis, 16, 28
- arachnoid villi, 5, 28
- artificial spinal fluid, 12

- bladder control, 4, 6, 7, 20, 25, 29
 - impairment, 4, 6, 7
 - loss of, 7
- blockage, 12, 30
- blood clot, 24

- catheter, 11, 12, 13, 17, 18, 19, 28
- CAT scan. *See* CT scan
- cerebral aqueduct, 16, 28, 29, 30
- cerebrospinal fluid. *See* CSF
- choroid plexus, 5, 28
- clinical exam, 9
- coma, 24
- compensated hydrocephalus, 18, 19
- complications, 18, 21, 23, 24
- computerized tomography. *See* CT scan
- continuous lumbar drainage. *See* lumbar drainage
- cranial surgery, 5
- CSF, 2, 4 - 6, 8, 10 - 14, 16 - 19, 23, 24, 28
 - flow void sign, 10
 - outflow resistance, 11, 12, 18
 - removal, 11, 18, 23
- CT scan, 8, 10, 11, 24
- cysts, 5

- death, 22, 24
- dementia, 2, 4, 6, 7, 8, 19, 25, 29
- diagnosis, 2, 9 - 13, 20, 21, 25, 26
- diagnostic tests, 9 - 13, 23
- differential-pressure valve. *See* valves

- endoscopic third ventriculostomy (ETV), 16, 28
- enlarged ventricles, 4, 8, 9, 10, 18, 29
- estimation of CSF pressure, 11

- external lumbar drainage. *See* lumbar drainage

- family support, 25
- fever, 24
- flow-regulated valve. *See* valves

- gait disturbance, 2, 4, 6, 7, 8, 9, 16, 20, 25, 29
- gravity-compensating device. *See* valves

- headache, 13, 24
- head injury, 5
- hematoma, 5, 14, 24, 29
- hemorrhage, 5, 16, 29
- hydrocephalus
 - compensated, 18, 19
 - defined, 4
 - normal pressure. *See* NPH
- hygroma, 24, 29

- ICP, 4, 11, 12, 18
- ICP pressure monitoring, 12
- idiopathic, 5
- incisions, 19, 20
- incontinence, 2, 7, 8, 10
- infections, 6, 19, 20, 23, 24, 29
- meningitis, 6, 23, 29
- shunt infection, 23
- intracranial pressure. *See* ICP
- isotopic cisternography, 11, 13

- LP shunt, 16, 19
- lumbar catheter, 11, 12, 18
- lumbar drainage, 11
- lumbar puncture, 11, 18
- lumbar puncture. *See* LP shunt

- magnetic resonance imaging. *See* MRI
- malfunction. *See* shunt malfunction
- meningitis, 6, 23, 29
- metallic implants, 10
- mild dementia, 4, 6, 20, 25
- MRI, 8, 10, 16

- nausea, 13
- neurologist, 9, 18 - 20, 22
- neurosurgeon, 9, 15, 18 - 21, 34
- normal pressure hydrocephalus. *See* NPH

NPH

- causes, 5
- defined, 4, 5
- diagnosis, 9 – 13
- symptoms, 6 – 9
- treatment, 13 – 16
- untreated, 24, 25

obstruction. *See* shunt obstruction

overdrainage, 14, 24

pacemaker, 10

paralysis, 24

Parkinson's disease, 2, 3, 7, 8, 22

peritoneal cavity, 13, 16, 17, 19

pressure monitoring, 12, 18

programmable valves. *See* valves

redness, 24

removal of spinal fluid. *See* CSF removal

shunt

- infection, 19, 20, 23
- malfunction, 21, 23
- obstruction, 16, 23, 29
- responsiveness, 11
- surgery, 12, 13, 16, 18, 20 – 26
- types of, 13 – 17
- valve. *See* valves

siphoning, 13

spinal cord, 4, 28, 29

spinal fluid pressure, 13, 18

spinal needle, 11

spinal tap, 11, 13, 16

subdural hematoma, 5, 14, 24

subarachnoid hemorrhage, 5

subarachnoid space, 5, 18, 30, 31

surgery, 7. *See also* shunt surgery

surgical risks, 9, 12, 18, 21 – 24

swelling, 24

symptoms of NPH, 2, 3, 7, 9, 23, 6 – 9, 10, 11

third ventriculostomy. *See* endoscopic third ventriculostomy

trauma, 16

triad of symptoms, 2, 4, 6, 7, 29

tube, 11, 28

tumor, 5

urinary frequency, 7, 10

urinary urgency, 7, 10

valves

adjustable, 12, 15

anti-siphon device, 14

differential-pressure, 13, 14

flow-regulated, 14

gravity-compensating device, 14

mechanism, 13, 14

programmable, 12, 15

VA shunt, 13

ventricle

fourth ventricle, 5, 28

lateral ventricle, 5, 8, 17, 19

third ventricle, 5, 8

ventriculoatrial shunt. *See* VA shunt

ventriculoperitoneal shunt. *See* VP shunt

ventriculostomy. *See* endoscopic third ventriculostomy

VP shunt, 13, 16, 17, 19

walking, 6, 9. *See also* gait disturbance

X-rays, 10

Resources

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 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 healthcare 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.

About Normal Pressure Hydrocephalus—A Book for Adults and Their Families was originally published by the Hydrocephalus Association in 1998, with financial support from The George H. Sandy Foundation.

Revision of this booklet was made possible through funds contributed by Codman, a Johnson & Johnson Company, The Medtronic Foundation and Medtronic Neurological Technologies.

Editorial review was provided by:

Marvin Bergsneider, MD	Harold L. Rekate, MD
Peter M. Black, MD, PhD	Norman Relkin, MD, PhD
Michael D. Heafner, MD	Marvin L. Sussman, PhD
Sharon Lamb, RN	Charles Teo, MD
Edward R. Laws, Jr., MD	Marion L. Walker, MD
Anthony Marmarou, PhD	Michael Williams, MD

2010 Edition

Graphic Design: Briar Levit, BriarMade

Photography: Our special thanks to the many families who graciously allowed the use of their photos for this booklet.

Editor: Rachel Fudge

Illustrations: Lynne Larson © 2002

© Copyright 2010 Hydrocephalus Association

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*
- *A Teacher's Guide to Hydrocephalus*
- More than 20 one- to four-page fact sheets that answer common questions about hydrocephalus and its complications

Directories of Medical Professionals

- Pediatric Neurosurgeons
- Congenital Neurosurgeons
- Neurosurgeons and Neurologists for Older Adults
- Neuropsychologists

Hydrocephalus Association

870 Market St, Suite 705
San Francisco, CA 94102

Toll-free: (888) 598-3789
E-mail: Info@HydroAssoc.org
Website: www.hydroassoc.org