



*About* ***Normal Pressure  
Hydrocephalus***

***A Book for Adults and  
Their Families***

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***About Normal Pressure Hydrocephalus—A Book for Adults and Their Families*** was written for adults, 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 Parents*, 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 normal pressure hydrocephalus 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.

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## Foreword

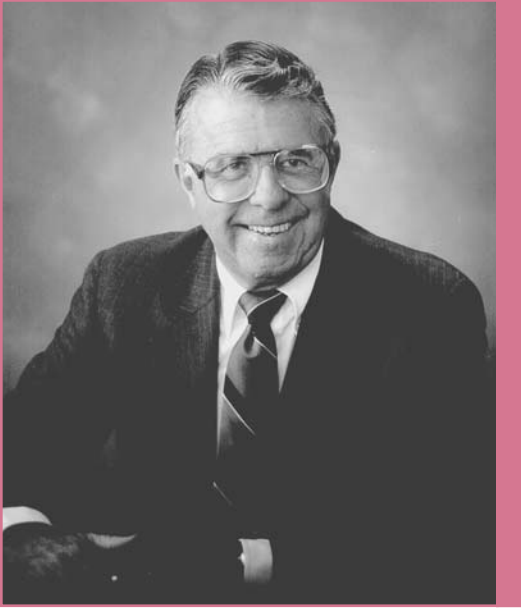
**T**he characteristic symptoms were clearly present—the stuttering walk, urinary incontinence, ataxia and the first traces of dementia and memory loss. Yet it would be several years before my husband, Richard, would be correctly diagnosed with adult-onset normal pressure hydrocephalus. Those years would prove to be extremely frustrating and painful for all of us.

Often I would catch sight of Dick—a man ordinarily stoic by nature—with his teeth tightly clenched and his hands twisted into fists. Bafflement and defeat seemed to be his constant companions. Neither of us could understand what was happening to him. For my part, I suspected Alzheimer’s disease and I would draw the children aside to voice that judgment, but it was one I, as well as they, was loath to accept.

At one point we had a physical therapist come to the house to assist Dick with his walking, little knowing that all the training and exercise in the world would never again allow those feet to walk normally. Even more demoralizing to his spirit was the loss of bladder control, the terrible urgency that resulted in many accidents and changes of clothing and bed linens.

So over the years, with the help of a hired driver with strong arms, we floundered, consulting a corps of physicians—but to no avail. It was not until we met with a neurologist, young and bright and thorough, that we saw a ray of hope for diagnosis. At once he ordered a CT scan and a radionuclide cisternogram. The resultant findings were consistent with normal pressure hydrocephalus. At last we had a diagnosis. Yet the name of Dick’s disease held words of meaning as foreign to us as Jabberwocky. But help was on the way!

The children were of vast assistance. Immediately they were on the Internet, talking to other doctors and patients and ultimately discovering the Hydrocephalus Association, based in San



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*Dick Kranz was diagnosed with normal pressure hydrocephalus in 1998, at age 72, and underwent shunt surgery shortly thereafter.*

Francisco. With support and encouragement from the Association we were gently led on to the next step in treating NPH—a shunt inserted into Dick’s cranium to drain the cerebrospinal fluid into the abdominal area, a procedure often associated with malfunction and other complications. The concept was scary, but it seemed the only recourse and the neurosurgeon was optimistic. Coming home that sunny blue day from our appointment with the neurosurgeon, following along Lake Erie’s shore, Dick said, “Oh, just to get back in the boat on that lake again!” There were tears in his eyes. He was so hopeful.

The shunt insertion was successful—for four days. For four days the witty, dynamic, clear-thinking Richard was with us again. But then we lost him to a stroke, a sad terminus to a renewed life so eagerly awaited.

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Later, after Dick's death, we discovered a CT scan taken six years prior to the final diagnosis of NPH. It distinctly pictured the telltale abnormality of enlarged ventricles. To this day that CT scan remains a mystery. Who read—or didn't read—it? Why was this condition not recognized? If we allowed ourselves, we could play the "what-if" game. What if adult-onset normal pressure hydrocephalus had been diagnosed back then, giving Dick a six-year head start? The what-ifs could go on and on. . . . Suffice it to say, the earlier this condition is detected, the brighter the treatment results.

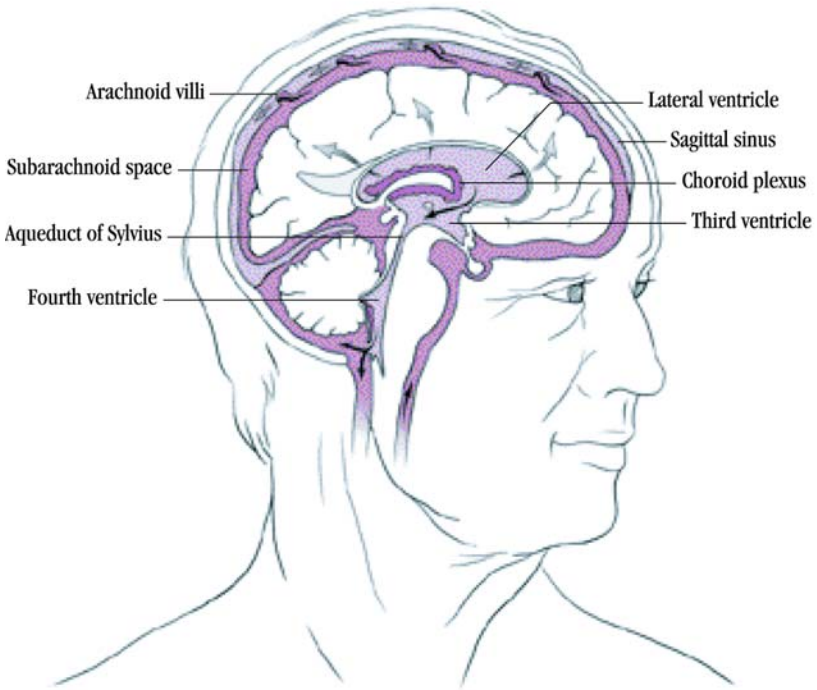
Granted, adult-onset NPH is not always readily diagnosed, its symptoms so similar to other diseases and conditions of aging. Yet to my mind, consideration and testing for NPH should be mandatory in the assessment of any apparently "psychiatric" illness of old age.

Thanks to the brilliant work of several neurosurgeons and neurologists, NPH—while not yet a household term—is more commonly recognized and diagnosed. Shunt technology, too, continues to advance, including a shunt that is now externally programmed and regulated, a giant step forward in treating the disease.

Through the efforts and research of dedicated doctors (many of whom contributed to this booklet) and hard-working patient advocates like the Hydrocephalus Association, much has been learned about NPH and its treatment. Thanks to them, there is hope for a more promising consequence for other new patients.

Dick's struggle is being vindicated.

*-Jo Kranz  
Rocky River, Ohio, October 2000*



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**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.*

## About Normal Pressure Hydrocephalus

### ***What Is Hydrocephalus?***

Hydrocephalus is a condition characterized by an abnormal accumulation of cerebrospinal fluid (CSF) within cavities called ventricles 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 disturbance, mild dementia and 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 normal to low. However, normal pressure hydrocephalus, or NPH, continues to be the common name for the condition.



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## ***What Causes Normal Pressure Hydrocephalus?***

The majority of cases of NPH are idiopathic (meaning of unknown cause). NPH can also develop as the result of a 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?***

The syndrome of normal pressure hydrocephalus is usually characterized by a triad of symptoms: complaints of 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 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 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

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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 is 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.

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 presenile dementia, including Alzheimer's, may be associated with atrophy or shrinkage of the brain, resulting in large CSF spaces that are visible on MRI and CT scans. Because the appearance is quite similar to

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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 their family 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, one or more of the following tests is usually 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. 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 and risks of surgery.

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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 the tests described here need to be performed in order to make a diagnosis.

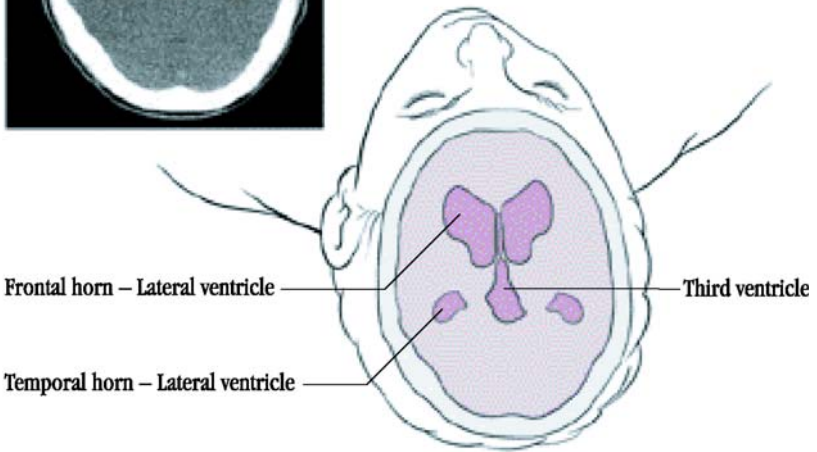
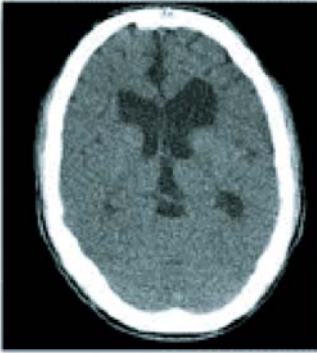
**Clinical exams to evaluate symptoms** consist of an interview and/or a physical/neurologic examination. Some common tests a doctor might perform in a clinical exam are:

- 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 15 minutes. 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



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*Top: CT scan showing enlarged ventricles*

*Bottom: Orientation of CT scan in the head*

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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 make 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 any particular patient’s responsiveness to any particular shunt, many doctors find the following 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.

- **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. Up to 50 cc of CSF is removed to see if symptoms are temporarily relieved. 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

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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 (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 (e.g., the removal of 10cc at the beginning of every hour), 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 therefore greater 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 clinic 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 drainage pathways.

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- For **intracranial pressure (ICP) monitoring**, a small pressure monitor is inserted through the skull into the brain or ventricles 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. Remember that 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.
  - **Isotopic cisternography** 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. It is not used much anymore because a “positive” cisternogram result does not reliably predict whether a patient will respond to implantation of a shunt, and results are often ambiguous.

### ***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 (*see drawing on page 20*). 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.



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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 valve**. This valve opens if the fluid pressure at the inlet of the valve exceeds the pressure at the outlet by a certain amount. Traditionally, differential-pressure valves have come in low, medium and high pressure varieties. For the adult with hydrocephalus, sometimes the effects of gravity on CSF flow through the shunt overwhelm the valve, 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.

In order to counteract this potential problem, valve mechanisms have been designed that incorporate **anti-siphon** and **gravity-compensating 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 prevent 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 overdrainage is low (when the body is prone). It is designed specifically to minimize excess CSF drainage during strain—such as bowel movements, coughing, sneezing or sexual exertion.

**Adjustable** and **programmable valves** are differential-pressure valves that can be adjusted externally, using magnetic program-

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mers or adjustment tools. These valves allow the opening pressure of the shunt to be fine-tuned without additional surgery and may help to optimize treatment and avoid repeated surgery in some cases. The valves can be adjusted within a range of differential pressures, from low to medium to high, in multiple steps. In some cases, adjustable valves include an anti-siphon or gravity-compensating device. Because adjustable or programmable valves are generally reset with magnetic devices, they should be checked after an MRI or exposure to other strong magnets.

It is important to realize that each of these valve designs has potential advantages and disadvantages, and that there is no single design or setting that works in all patients. The valve a neurosurgeon selects for an individual depends on a number of factors, including age, size of ventricles, intracranial pressure, the availability of the valve and the experience of the neurosurgeon. People with NPH may want to ask their neurosurgeon to demonstrate a sample shunt and discuss the pros and cons of the system being recommended.

As an alternative to a VP or VA shunt, some neurosurgeons may recommend a lumboperitoneal (LP) shunt. An LP shunt is inserted into the spinal space in the lower back (in the same place a spinal tap is done). The tubing, which is smaller 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

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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.

### ***Who Is a Likely Candidate for Shunting?***

The answer to this question is still uncertain. Many tests and evaluation criteria have been proposed, but, unfortunately, no one single factor is reliable in predicting success from implantation of a shunt. The following findings are generally associated with a better outcome following shunt placement:

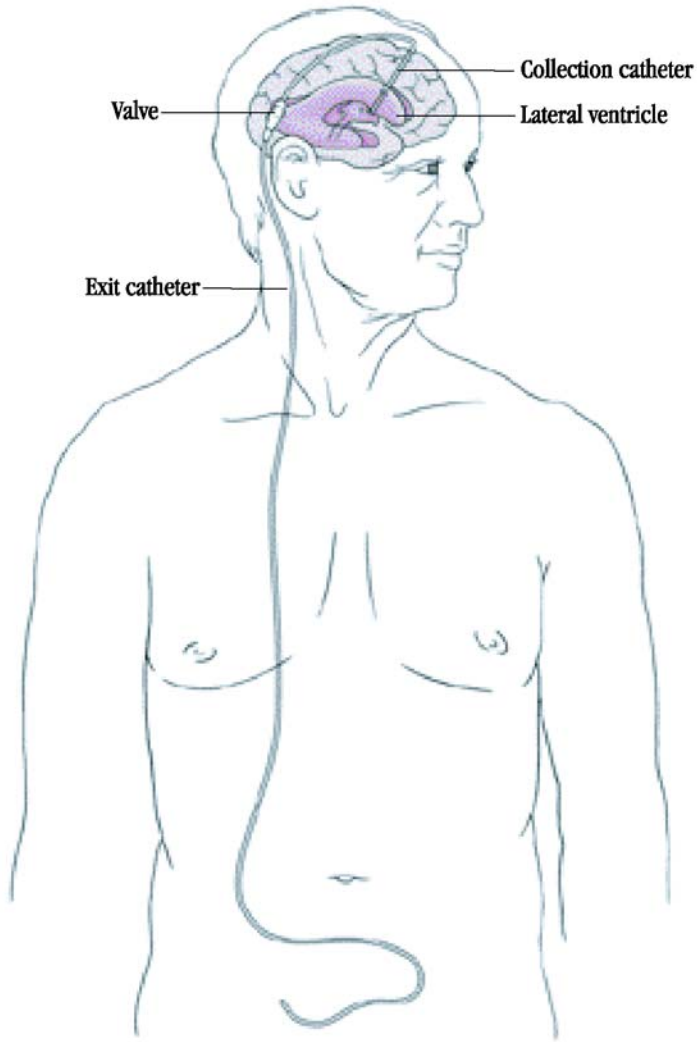
- The onset of gait disturbance as the first and most prominent symptom
- A known cause for NPH, such as trauma or hemorrhage
- The scan shows the ventricular size to be disproportionately larger than the CSF in the subarachnoid space
- Removal of spinal fluid via lumbar puncture or lumbar catheter gives 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

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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 he or she 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 he or she 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 benefits 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.



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*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.*

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## ***What Does a Shunt Operation Entail?***

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, their 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 then made on 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.

The patient generally stays under careful neurological observation for the first 24 hours following the procedure. The incisions are monitored for signs of infection. Patients generally 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.

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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.

Other factors that affect the outcome after shunt surgery are the presence of other neurological or medical conditions. 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.

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## ***What Are the Complications and Risks Involved with Shunting?***

Although shunt surgery is a relatively simple neurosurgical procedure, the decision to undergo the insertion of a shunt should not be taken lightly. The treatment of normal pressure hydrocephalus carries greater risks compared to the treatment of children with hydrocephalus; therefore, this operation should be undertaken only if the degree of disability or the progression of the disorder warrants 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. A complication can be thought of as any unwanted event related to the surgical procedure itself or the presence of the shunt. 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 that they can become obstructed (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 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.



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Another common complication of shunt surgery is shunt malfunction, which can occur when either end of the shunt is malpositioned, 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 indicated 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

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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, a person with NPH should not hesitate to seek medical attention if he or she develops worrisome symptoms.

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 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. If your symptoms are very mild, your 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 even more difficult. Families and individuals who stay informed and ask questions of their physicians will

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likely feel more involved in the management of their care. It is important to be optimistic about surgical treatment without expecting miracles. We live in a society where people are living longer, and more and more adult children are being asked to care for their aged 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. Many families 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.

## Conclusion

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 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.

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- 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 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.

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## 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)

*Hydrocephalus Diagnosed in Young and Middle-Aged Adults—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

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*serving individuals, families and professionals since 1983*

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