NPH
NORMAL PRESSURE HYDROCEPHALUS

Early Diagnosis and Management

An overview for clinicians
Why read this brochure?

In your practice, you may be familiar with the signs and symptoms of Alzheimer’s and Parkinson’s disease, or with other conditions associated with the aging process. But do you know that these diseases and conditions share symptoms with a more treatable condition known as normal pressure hydrocephalus (NPH)?

The symptoms can be subtle and may not always occur together. NPH is challenging to diagnose and may be further complicated by co-existing illnesses. However, early diagnosis and treatment can relieve or even reverse its symptoms.

This brochure was created to help you recognize the signs and symptoms of NPH and to provide information on the next steps to take if you suspect your patient has this condition.

There are no definitive statistics, but it is estimated that about 5% of patients with dementia are likely to have NPH (Verrees 2004).
What is NPH?

Hydrocephalus is an abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles of the brain. NPH is a type of hydrocephalus that generally occurs in a person’s 60s and 70s.

When NPH is due to brain injury or disease, it is called secondary NPH. This form of the disorder is much easier to treat and has a better post-surgical outcome (Verrees 2004). Idiopathic NPH (INPH), by definition, occurs without a known cause, but there are a few theories about what does cause it. One accepted theory is that it is the result of an imbalance in CSF production and absorption. Others have suggested that it may be due to reduced blood flow in the small vessels of the brain (Vanneste 2000).

NPH is often characterized by the triad of gait disturbance, cognitive decline, and urinary incontinence. Sometimes the person may have one or two of the main symptoms but gait disturbance must be evident to make the diagnosis.
The NPH Triad

**Gait disturbance**
Often the first and most significant problem, gait disturbance may take the form of small shuffling steps, a tendency to fall, difficulty using stairs, or a feeling of heavy feet—many NPH sufferers describe the feeling of being “stuck to the floor.” Often patients and families will first seek medical attention when the person begins to have falls.

**Urinary incontinence**
Urinary incontinence may appear later in the progression of NPH, and in some cases this symptom may never develop. The symptoms may include frequency, urgency, or frank incontinence. Progression of the disease may lead to fecal incontinence.

“If any one of the symptoms is evident, normal pressure hydrocephalus should be considered and ruled out before moving on to a diagnosis of dementia or Alzheimer’s disease” (Smolinsky 2008).
**Mild dementia**

Mild dementia may take the form of forgetfulness, short-term memory loss, and loss of interest in things the person had previously enjoyed. In some instances there can be mood changes shown by withdrawal or having a flat effect which may seem like depression. Unlike other dementias, the dementia caused by NPH can sometimes be reversed or at least stabilized.

The onset of NPH is subtle and the symptoms may not appear simultaneously. It may appear to the caregiver, family, or the patients themselves simply to be part of the aging process. Another factor clouding the diagnosis of NPH is that it shares similarities with other conditions associated with an aging population, such as Parkinson’s or Alzheimer’s disease, peripheral neuropathy, osteoarthritis, among many others. Left untreated, NPH will progress and significantly impair a person’s quality of life.

Once a primary caregiver suspects NPH, additional tests are needed to confirm the diagnosis and to determine if shunting is an appropriate treatment option. At this point the care team should include a neurologist to facilitate the diagnostic procedures and interpretation of the results.
Diagnosing NPH

A routine diagnostic workup of a person suspected of having NPH usually consists of the following tests:

**Neuropsychological testing**

This is the most accurate and least invasive means of determining and documenting cognitive strengths and deficits. Common neuropsychological testing areas are:

- Attention
- Memory
- Language
- Reaction time
- Reasoning
- Emotional state

**CT or MR scan**

Neuroimaging to assess ventricular size is an essential part of diagnosing a patient suspected of having NPH.

**CSF lumbar drainage tests**

CSF lumbar drainage tests are used to predict the likelihood of a positive response by the patient to a shunting procedure. Though there is no way to accurately predict how an individual patient will respond, many doctors find the following tests helpful.

Lumbar Puncture or Spinal Tap is an invasive diagnostic test that allows CSF to be removed for examination and the pressure of the spinal column to be measured. Typically about 40–50 ml of CSF is removed to see if any of the symptoms are relieved. Generally, there is a response to the gait disturbance within about 6 hours, so the patient should expect to be re-examined the same day. Improvement of symptoms following a lumbar puncture is potentially indicative of a positive response to shunting.

“After a decade of depressing diagnoses I didn’t immediately accept that I had this relatively new, mysterious, reversible illness until my neurosurgeon had performed a lumbar puncture that induced an instantaneous, miraculous remission” (Conn 2007).
External Lumbar Drainage is an option if a patient does not respond to a simple lumbar puncture. There may be a need for more CSF to be removed or more time may be required to demonstrate a response. This procedure requires hospitalization for 2 to 3 days, so that CSF can be drained and regular assessments of the patient’s symptoms can be made, in an effort to determine if there will likely be a response to a surgical shunting procedure.

CSF Outflow Resistance is a more involved test that determines the capacity of the body to absorb excess CSF. The test involves a lumbar puncture, infusion of artificial spinal fluid, and specialized laboratory equipment that determines the body’s absorption capacity—sometimes called “outflow resistance.” If the outflow resistance is abnormally high, there is a good chance that the patient will improve with shunt surgery.
A ventriculoperitoneal (VP) shunt consists of a catheter placed into the ventricles. This is attached to a valve which is connected to a distal catheter. The distal catheter drains the CSF into the peritoneal cavity where it can be absorbed by the body.
Treating NPH

Implantable CSF-shunt therapy utilizes a relatively simple, efficient technology that has been used to treat hydrocephalus for over 50 years.

The only effective treatment for NPH is surgical diversion of CSF. This is accomplished with the surgical implantation of a shunt. A shunt is designed to drain CSF from either the brain’s ventricles or the lumbar subarachnoid space to a distal site, such as the peritoneal cavity where the CSF can be reabsorbed.

The most common devices used to treat NPH are ventriculoperitoneal (VP) shunts. The neurosurgeon will evaluate the factors for placing the shunt, including the risk/benefit ratio of the procedure, sites of the proximal and distal catheter, valve specifics, and shunt-related complications.

The two main types of shunt valves available are fixed pressure valves and adjustable pressure valves. Fixed pressure valves regulate the flow rate of the CSF based on a predetermined pressure setting. A major improvement to valve technology in recent years has been the development of adjustable pressure valves. These regulate the flow of CSF based on an externally adjustable pressure setting. By using custom magnetic tools, the doctor can fine tune the setting of a patient’s valve during an office visit according to the patient’s needs, thus avoiding unnecessary surgery.

Both of these types of valves can include overdrainage protection in the form of a siphon control device. The purpose of a siphon control device is to minimize excessive drainage due to gravity, which can cause more CSF to drain when the patient is in the upright position.

A study of 151 patients with NPH found that two-thirds improved with temporary drainage. After implantation of an adjustable shunt, 91% benefitted from the implant (Marmarou, et al. 2005).
Who Will Respond Well to Shunting?*

Probable NPH (from INPH Guidelines, Part II)

I. History

*Symptoms should be confirmed by a family member or someone familiar with the person’s previous and current condition.*

a. Insidious vs. acute onset
b. Symptoms begin after 40 years old
c. Person shows symptoms for at least 3 to 6 months
d. No preceding event such as head trauma, intracerebral hemorrhage, meningitis, or other known causes of secondary hydrocephalus.
e. Progression over time
f. No other neurological, psychiatric, or general medical conditions that can explain the presenting symptoms

II. Brain Imaging

a. Ventricular enlargement not completely caused by brain atrophy or congenital enlargement.
b. No obvious obstruction to CSF flow
c. At least one of the following:
   1. Enlargement of the lateral ventricles not entirely caused by brain atrophy.
   2. Callosal angle of 40 degrees or more
   3. Evidence of altered brain water content, including periventricular signal changes on CT and MRI not attributable to microvascular ischemic changes or demyelination
   4. An aqueductal or fourth ventricular flow void on MRI

III. Clinical

*The classical definition findings of gait/balance disturbance must be present, plus at least one other area of impairment in cognition, urinary symptoms, or both.*

Gait/Balance.

*At least two symptoms should be present and not due to other conditions.*

a. Decreased step height
b. Decreased step length
c. Decreased cadence (speed of walking)
d. Increased trunk sway during walking
e. Widened standing base
f. Toes turned outward on walking
g. Retropulsion (spontaneous or provoked)
h. En bloc turning (turning requiring three or more steps for 180 degrees)
i. Impaired walking balance, as shown by two or more corrections out of eight steps on tandem gait testing
III. Clinical (continued)

Cognition.
At least two symptoms should be present and not due to other conditions.
   a. Psychomotor slowing (increased response latency)
   b. Decreased fine motor speed
   c. Decreased fine motor accuracy
   d. Difficulty dividing or maintaining attention
   e. Impaired recall, especially for recent events
   f. Executive dysfunction
   g. Behavioral or personality changes

Urinary Incontinence.
Either one of the following should be present.
   a. Episodic or persistent urinary incontinence not attributable to primary urological disorders
   b. Persistent urinary incontinence
   c. Urinary and fecal incontinence
Or any two of the following should be present
   a. Urinary urgency as defined by frequent feeling of a need to void
   b. Urinary frequency as defined by more than six voiding episodes in an average 12-hour period despite normal fluid intake
   c. The need to urinate more than two times in an average night

IV. Physiological
CSF opening pressure in the range of 5-18 mm Hg (or 70-245 mm H2O) determined by lumbar puncture or comparable procedure.

Identifying Shunt-Responsive Patients (Marmarou 2005):
Supplemental Prognostic INPH Tests

- MRI shows no white matter lesions
- Tap test removal of 40 to 50 ml has a positive response
- Prolonged external lumbar drainage over 300 ml
- CSF outflow resistance (CSF Ro) greater than 18 mm Hg
- CSF-OP in the range of 105 to 190 mm H2O

Outcomes*

“The shunt virtually gave me my life back.”
- Ed Roe, valve recipient

- “Today, 70–80% of patients operated on with a CSF shunt improve postoperatively.”

- A 2005 study of 151 patients with NPH found that two-thirds improved with temporary drainage. After implantation of an adjustable shunt, 91% benefited from the implant.

- The INPH guidelines reported improvement rates of 30-96%

- Although all symptoms can resolve following shunt surgery, gait is the most likely to improve.

- A study by Gallia, Rigamonti and Williams found that 75% of patients had improvement in at least one symptom and 46% had improvement in all symptoms at 18 months.

- Time of intervention is critical: numerous studies have demonstrated that a longer duration of NPH symptoms is associated with lower likelihood of shunting response.

- Cognitive improvement was observed in more than 50% of patients following shunting.

*Gallia, et al., 2005
Potential Complications

As with any surgical procedure, there are potential complications associated with implanting a shunt. Unlike most surgeries, where the risks are highest during the operation itself, most of the common problems associated with shunting may occur at a later time. The major complications of shunting for NPH are subdural hematomas and infection. Occasionally, a shunt will become obstructed, but it is not life threatening. If an obstruction occurs, the patient will experience a gradual return of their original symptoms.

Patients, families, and caregivers should be alert to the signs and symptoms of shunt complications and recognize that shunt revisions may be necessary to replace a component that is not working.

According to a study published in the October 2005 issue of Neurosurgery, 132 patients received shunts for NPH and of these, 75% improved in at least one of their symptoms (McGirt 2005). The following complications were noted in the study: low-pressure headache, subdural hematoma, shunt obstruction, infection and overdrainage.
Bibliography


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