Primary care providers (PCPs) often see elderly patients in their practice with symptoms of dementia, usually because of irreversible conditions. Normal pressure hydrocephalus (NPH) is an often unrecognized cause of dementia symptoms, and can be reversible with appropriate diagnosis and treatment, specifically ventriculoperitoneal (VP) shunting. It has been estimated that 375,000 Americans misdiagnosed with dementia or Parkinson's disease have symptoms that are actually caused by normal pressure hydrocephalus. Some experts suggest that as many as 6% of nursing home residents may have NPH. These figures, however, are controversial since no precise data on the incidence and prevalence of NPH have been published. The diagnosis of NPH can be difficult, and is often based on a combination of clinical signs and symptoms, radiographic findings, and confirmatory testing.

The term “normal pressure hydrocephalus” was originally described by Hakim and Adams in 1965 and refers to a clinical triad of gait disturbance, dementia symptoms, and urinary incontinence accompanied by radiographic hydrocephalus. The diagnostic challenge is to differentiate NPH from other neurologic problems, such as Alzheimer's disease and Parkinson's disease, that can present with similar symptoms. Normal pressure hydrocephalus can be idiopathic or secondary. Secondary NPH is associated with an identifiable cause, such as subarachnoid hemorrhage, meningitis, or brain injury. Idiopathic NPH occurs without a clearly defined etiology and primarily affects the elderly.

Pathophysiology
The exact pathophysiology of NPH is unclear. Cerebrospinal fluid dynamics are affected by both production and absorption. Cerebrospinal fluid production seems to be normal in NPH, but there appears to be a problem with CSF absorption. The majority of the CSF is formed by specialized tufts in the ventricles, called the choroid plexus, which produces CSF at a rate of approximately 20 to 25 mL per hour. The CSF circulates from two lateral ventricles to a single midline third ventricle into the fourth ventricle, which is located within the posterior fossa of the brain. The CSF exits the ventricular system and passes into the subarachnoid space surrounding the brain and around the spinal cord (see Figure: “Normal CSF Flow”). The CSF in the subarachnoid space provides a cushioning effect to the brain. Most of the CSF is absorbed into the arachnoid villi, which are permeable and allow the CSF to exit from the subarachnoid space into the venous sinuses.

For reasons not clearly understood, in NPH the arachnoid villi fail to maintain an adequate removal of CSF. One theory proposes that fibrosis and scarring impair reabsorption. Others suggest that the cerebral vasculature may have a role in the pathogenesis of NPH. Clinical deterioration may be due to slow, progressive impairment of the periventricular blood flow. This may explain why the prevalence of arterial hypertension and cerebral arteriosclerosis is increased in patients with NPH. Studies have also shown that NPH may be associated with injury to neurotransmitter and neuropeptide systems, which affect cognitive performance.

Some experts suggest that the term “normal pressure hydrocephalus” is a misnomer. Studies have shown that while CSF pressure is normal at one spinal tap, episodes of slightly increased CSF pressure occur in NPH. Over the years, other terms have been suggested, such as “adult hydrocephalus syndrome,” “adult symptomatic hydrocephalus,” or “chronic hydrocephalus,” but the term “normal pressure hydrocephalus” has prevailed. Whatever the etiology, the end result is dilatation of the ventricles (or ventriculomegaly), resulting in a communicating hydrocephalus. A communicating hydrocephalus is a condition in which the arachnoid villi are un-
It has been estimated that 375,000 Americans misdiagnosed with dementia or Parkinson's disease have symptoms that can be reversed with appropriate diagnosis and treatment.
able to reabsorb CSF sufficiently without an obstruction to CSF flow in the ventricular system. Non-communicating, or obstructive, hydrocephalus occurs when there is impedance to flow, usually caused by a mass lesion, such as a tumor, within or adjacent to the ventricular system. In NPH, the progressive ventriculomegaly may cause compression of structures adjacent to the ventricles, resulting in the clinical manifestations of the disease. For example, pressure on the frontal lobes may cause the dementia symptoms; pressure on the cortical center for bladder control may cause incontinence; and pressure on the corticospinal fibers lateral to the ventricles may cause the gait disturbance.

- Clinical Features and Differential Diagnosis

The syndrome of NPH is characterized by the classic clinical triad of gait disturbance, dementia symptoms, and urinary incontinence (see Table: "Clinical Triad of NPH"), although not all patients will present classically. Gait impairment is often the most pronounced symptom of NPH and may be the initial manifestation. It is characterized by slow, wide-based, shuffling steps, as if the feet were "glued to the floor." This causes problems in ascending and descending stairs and curbs. Patients also describe a sense of dizziness and disequilibrium, resulting in balance problems, difficulty turning, and a tendency to fall backwards. Frequent falls can result from this gait disturbance, leading to other injuries.

Patients with Parkinson's disease demonstrate a gait disturbance similar to that associated with NPH. Both can present with a hypokinetic gait and decreased stride length, manifested by shuffling. The gait with Parkinson's disease, however, tends to be more narrow-based, and the balance problems and disequilibrium are not as apparent. Lack of arm swing and stooped posture are more prominent in Parkinson's disease than in NPH. Parkinson's patients will improve with a trial of a levodopa drug (such as Sinemet), which will have no effect on patients with NPH. Patients with Parkinson's disease may also exhibit other symptoms, such as resting tremor and rigidity, that can help differentiate the two conditions.

Dementia symptoms in the early stages of NPH may be subtle and primarily frontal in nature, including apathy, dullness in thinking and actions, and inattention. Lack of spontaneity and slowness in verbal or motor response are frequently encountered. Memory deficits are usually a component of NPH, which can lead to a false diagnosis of Alzheimer's disease. Alzheimer's disease is a type of cortical dementia characterized by aphasia, apraxia, and agnosia, which NPH patients do not typically exhibit. In other words, NPH dementia does not include word formation difficulties, inability to sequentially carry out simple tasks, or trouble interpreting sensory stimuli. These clinical features can assist in distinguishing NPH from other dementias, such as Alzheimer's disease.

Urinary symptoms associated with NPH often appear later than the other symptoms and are absent in some patients. Initially, they usually consist of urinary urgency and frequency and can progress to complete loss of bladder control, and in rare cases, fecal incontinence. Bladder dysfunction is due to stretching of the periventricular nerve fibers and subsequent partial loss of inhibition of bladder contractions. Urodynamic studies have shown hyperreflexia and instability of the bladder detrusor muscle but no evidence of defective bladder sphincter control. In patients with NPH, the slow gait can compound the problem, making it difficult to reach the bathroom in time. The differential diagnosis in elderly patients with urinary incontinence should include benign prostate hypertrophy, pelvic relaxation, and cystitis (see Table: "NPH Differential Diagnosis").

Normal pressure hydrocephalus is usually considered a syndrome associated with older adults, but up to one-quarter of patients are under 50 years of age. There are some reports of NPH in children and adolescents although these findings have been disputed. It seems to be more prevalent in males than in females.
Clinical Triad of NPH

<table>
<thead>
<tr>
<th>Clinical Manifestations</th>
<th>Characterized By:</th>
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<tbody>
<tr>
<td>Gait disturbance</td>
<td>Wide-based, shuffling gait</td>
</tr>
<tr>
<td></td>
<td>Difficulty picking up feet</td>
</tr>
<tr>
<td></td>
<td>Problems with stairs, curbs</td>
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<tr>
<td></td>
<td>Instability with turning</td>
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<tr>
<td></td>
<td>Frequent falls</td>
</tr>
<tr>
<td>Dementia symptoms</td>
<td>Short-term memory deficits</td>
</tr>
<tr>
<td></td>
<td>Loss of interest in daily activities</td>
</tr>
<tr>
<td></td>
<td>Difficulty with routine tasks</td>
</tr>
<tr>
<td></td>
<td>Lack of spontaneity in actions and verbal response</td>
</tr>
<tr>
<td></td>
<td>Slow processing of information</td>
</tr>
<tr>
<td></td>
<td>Problems with retention of language skills</td>
</tr>
<tr>
<td>Urinary incontinence</td>
<td>Urinary frequency and urgency (early sign)</td>
</tr>
<tr>
<td></td>
<td>Urinary incontinence (late sign)</td>
</tr>
</tbody>
</table>

CT Scan in NPH

Patient with normal sized ventricles (left). Patient with the ventriculomegaly of normal pressure hydrocephalus (right).

Diagnosing NPH

Accurately diagnosing NPH has presented challenges and controversies over the years, and currently there is no clear consensus regarding diagnostic confirmation of NPH. Computed tomography (CT) or magnetic resonance imaging (MRI) is often the first diagnostic study performed if the clinical picture suggests NPH. In NPH, the CT or MRI will reveal ventricular enlargement out of proportion to sulcal atrophy as well as rounding of the frontal horns (see Figure: "CT Scan in NPH"). It is often difficult to distinguish the etiology of the ventricular enlargement caused by NPH from brain atrophy as a result of other degenerative cortical conditions. However, patients with Alzheimer's disease tend to have a more significant overall loss of brain tissue as well as focal atrophy in the hippocampus. Magnetic resonance imaging is more sensitive than CT scan in differentiating the etiology of ventriculomegaly. MRI is also essential to confirm the absence of other pathologic conditions, such as subdural hematoma, intracranial neoplasms, or infections.

Because of the lack of definite diagnostic criteria for NPH, a variety of confirmatory tests have been used to assist in determining which patients would benefit from VP shunting. Since many patients with NPH are elderly, often with comorbid conditions, the goal is to shunt those patients who would clearly improve with surgery and to spare those who would not benefit from unnecessary surgical risk. Confirmatory tests include: nuclear cisternography, the lumbar tap test, the lumbar infusion test, intracranial pressure monitoring, and external continuous lumbar drainage.

Nuclear cisternography, in which a radioisotope is injected via a spinal tap, has been used in an attempt to select patients appropriate for shunting. While patients with NPH may show slow cortical uptake of the isotope, it has fallen out of favor since it seems to be more useful in diagnosing known causes of communicating hydrocephalus and less useful in diagnosing idiopathic NPH. Cisternography is currently considered by most experts to be an unreliable predictive test for NPH and has been replaced by other testing procedures. The lumbar tap test (or CSF tap test) consists of the removal of approximately 40 to 50 mL of CSF via a lumbar puncture. The test is considered positive if the symptoms temporarily improve after the withdrawal of CSF. The lumbar infusion test measures CSF pressures after mock CSF or saline is infused into the lumbar subarachnoid space. Other facilities use long-term recording of intracranial pressure to identify suitable VP shunt candidates. Unfortunately, the accuracy of these methods is difficult to evaluate because the patient samples were small and the techniques varied among the studies.

Several recent studies have reviewed the predictive value of external continuous lumbar drainage. Patients are hospitalized for a 3-day period, a small lumbar catheter is inserted, and about 5 to 10 mL of CSF is drained hourly. The patient’s symptoms, particularly the gait, are observed closely during the lumbar drainage trial. An objective improvement...
### NPH Differential Diagnosis

**Common conditions that may present similarly to NPH:**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Clinical Manifestation</th>
<th>Comparison to NPH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alzheimer’s disease</td>
<td>• Cognitive impairments characterized by aphasia, apraxia, and agnosia</td>
<td>• Cognitive impairment characterized by slowness of thought, inattentiveness, and apathy</td>
</tr>
<tr>
<td></td>
<td>• Gait impairment usually not a predominant feature</td>
<td>• Gait impairment is usually the first and most pronounced feature</td>
</tr>
<tr>
<td></td>
<td>• Urinary incontinence may appear in later stages</td>
<td>• Urinary urgency/incontinence may occur later</td>
</tr>
<tr>
<td>Parkinson’s disease</td>
<td>• Gait impairment is usually narrow-based, shuffling, with bradykinesia, decreased arm swing, and stooped posture</td>
<td>• Gait impairment usually more wide-based; bradykinesia, decreased arm swing, and stooped posture not as apparent</td>
</tr>
<tr>
<td></td>
<td>• Dementia symptoms may appear later in the disease</td>
<td>• Symptoms usually will not improve with levodopa medications</td>
</tr>
<tr>
<td>Vascular dementia (stroke, multi-infarct dementia, vertebrobasilar insufficiency)</td>
<td>• Usually presents with focal neurologic deficits attributable to a specific area of cerebral damage</td>
<td>• CT/MRI will show evidence of infarct(s)</td>
</tr>
<tr>
<td></td>
<td>• Gait disturbance may be due to motor weakness or limb ataxia</td>
<td>• Will not usually present with the classic clinical triad of NPH</td>
</tr>
</tbody>
</table>

in the symptoms is considered a positive test. Usually, the patients are discharged home after the trial, and if the symptoms return to baseline within 1 to 2 weeks after the catheter is removed, this is considered further confirmatory evidence. External lumbar drainage has been found to have a predictive value of about 85% for identifying patients that would benefit from shunt surgery, and is becoming more popular with neurologists and neurosurgeons. Some specialists prefer to first try a lumbar tap test on an outpatient basis, and if the patient improves, offer VP shunting. If the patient does not approve, they may proceed with the external lumbar drainage trial, which requires a hospital stay.

#### Treatment

The definitive treatment for NPH is placement of a VP shunt. Ventriculoatrial and ventriculopleural shunts can also be used in certain cases but are less common. The VP shunt involves placement of a catheter into one of the lateral ventricles, draining CSF from the ventricle into the catheter and through a valve placed under the scalp. From the valve, the catheter is tunneled under the skin to the peritoneal cavity, where the CSF is reabsorbed (see Figure “Ventriculoperitoneal Shunt”). More recently, programmable valves have been designed that help prevent the potential problems of overshunting and undershunting. Postoperative overshunting may result in ventricular collapse, causing a subdural hematoma, which can be a serious complication. Patients who are undershunted may not have optimal improvement of their symptoms. Programmable VP shunts can be easily adjusted during the postoperative period based on clinical and radiographic findings.

Other factors that determine suitable candidates for VP shunting involve a risk versus benefit analysis of each individual patient. Advanced age and comorbid conditions such as cardiovascular disease, pulmonary disease, and stroke can increase the postoperative complication rate. Shunt infections can occur, although the incidence of shunt infections is relatively low. Patients with coexisting Alzheimer’s disease may not receive enough improvement in their cognitive and functional status to justify the surgery. In general, coexisting cerebrovascular disease or Alzheimer’s disease are predictors of poor outcome following shunting.

The success rate of VP shunting varies widely in the literature, anywhere from 31% to 96%. Many of these studies, however, were reported using less stringent patient selection criteria than is currently recommended. A careful selection process can greatly increase the chances of a meaningful functional improvement. In general, patients with a shorter history of mental deterioration, patients in which the gait disturbance preceded the mental impairment, and patients with clinical improvement after either a lumbar tap test or external lumbar drainage tend to have a better surgi-
Ventriculoperitoneal Shunt

In the abdominal area, requiring routine incisional care. The staples or sutures will be removed at about 10 days postoperatively. Most patients report very little postoperative pain, which can usually be managed with short-term analgesics.

A follow-up CT scan without contrast is recommended postoperatively to check shunt placement and to assess for any hemorrhage. If the patient's clinical symptoms are improving, a CT scan at about 3 and 12 months postoperatively will also be performed, primarily to evaluate for any subdural fluid collection that may occur in a delayed fashion. Patients without clinical improvement after shunting should be followed more carefully. In patients with programmable valves, the valve can be adjusted if the patient fails to improve as expected, necessitating a repeat CT scan after each adjustment.

Discussion

Normal pressure hydrocephalus is a potentially reversible cause of gait disturbance, dementia symptoms, and urinary incontinence that primarily affects the elderly population. In the recent literature as well as the media, there has been increased interest in this syndrome, which is felt to be underrecognized and underdiagnosed. The process of diagnosing NPH begins with a thorough history and physical examination, usually in the primary care setting. When the clinical evaluation suggests NPH, a noncontrast CT or MRI should be ordered. If ventriculomegaly is present on imaging studies, prompt referral to a neurologist or neurosurgeon for confirmatory testing is recommended.

Many patients with NPH report years of insidious symptoms before finally being diagnosed and treated for the condition. The ability to walk, think coherently, and control bodily functions is vital to independence and self-worth. Patients who undergo a successful VP shunt procedure may have a varying degree of improvement. Realistic expectations of the probable outcomes of the procedure should be discussed thoroughly with the patient and the family. While many patients have remarkable improvement after VP shunt insertion, others have minimal or no improvement. Each patient and family is unique, and their expectations are unique as well.

REFERENCES


AUTHOR DISCLOSURE
The author has disclosed that she has no significant relationship or financial interest in any commercial companies that pertain to this education activity.

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