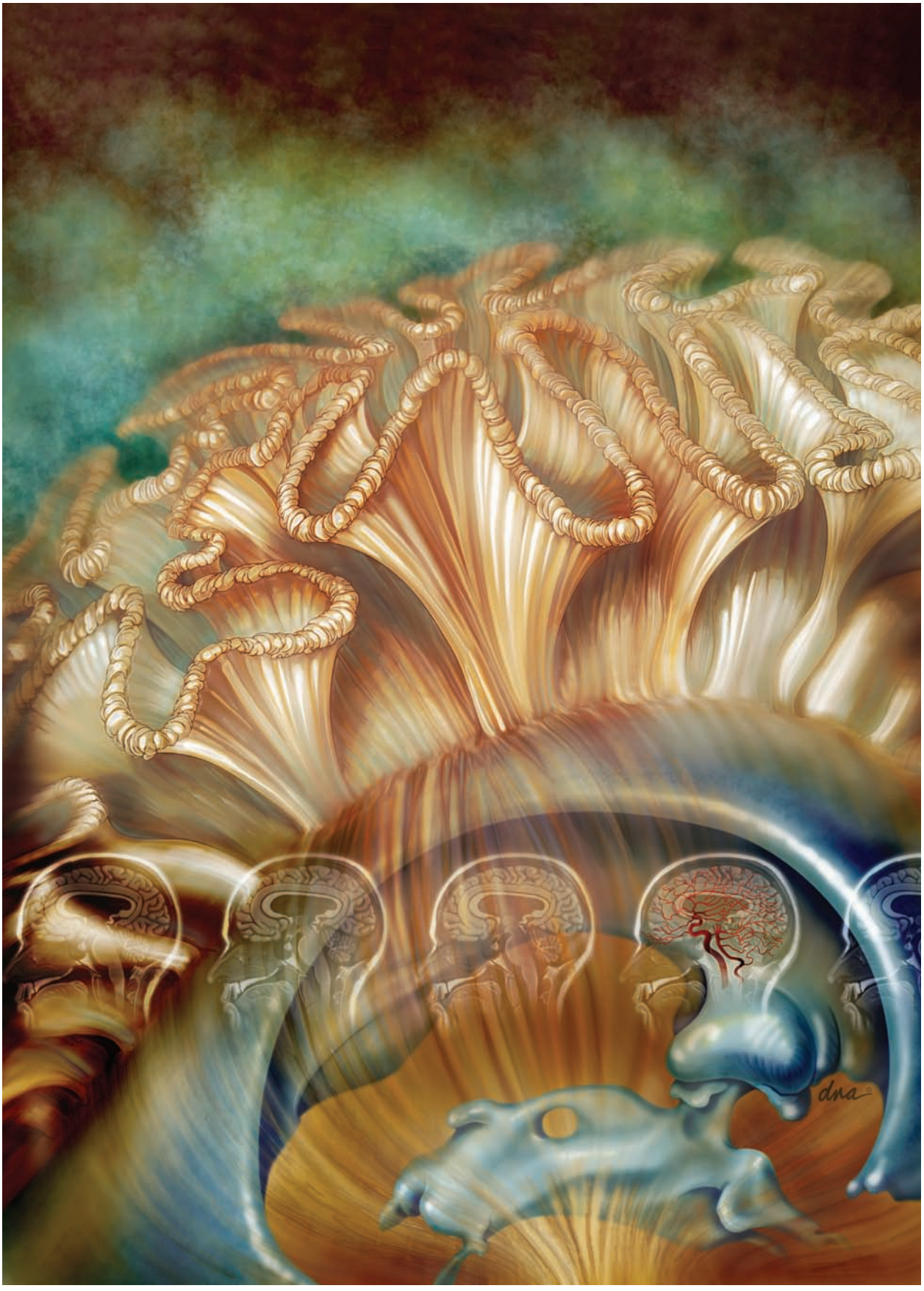

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Recognizing **normal pressure hydrocephalus** in older adults

By Carolyn E. Smith, MS, RN, CNRN

OFTEN MISDIAGNOSED as Parkinson disease or another neurologic disorder associated with aging, idiopathic normal pressure hydrocephalus (NPH) is most common in adults over age 60.[1] Because early detection and treatment with ventricular shunting can reverse dementia and other complications of NPH in many patients, nurses need to recognize a telltale triad of clues: gait disturbance, cognitive impairment, and urinary incontinence.

NPH can be either idiopathic or secondary to an underlying cause such as head trauma, subarachnoid hemorrhage, brain tumor, or an infection such as meningitis.[1] This article focuses on idiopathic NPH in older adults, using the case of Mr. S as an example.

A case in point

Mr. S, 78, is a widower who's struggled with frequent falls at home due to a shuffling gait and urinary incontinence. When talking with his family, he's been inattentive and slow to respond. Concerned about changes in Mr. S's mentation and declining health, his son has taken him to see his primary care physician (PCP) multiple times within the past 2 years. The PCP consistently attributed Mr. S's signs and symptoms to worsening of his Parkinson disease and onset of dementia.

After approximately 3 years, Mr. S's signs and symptoms remained unchanged. The son sought a second opinion from a neurosurgeon, who ordered magnetic resonance imaging (MRI) of his brain. The results showed larger than normal ventricles due to accumulated cerebrospinal fluid (CSF).

Suspecting NPH, the neurosurgeon also performed a lumbar tap test. After the removal of a small amount of CSF, Mr. S's signs and symptoms briefly improved: He became more attentive and had a steadier gait. He was diagnosed with

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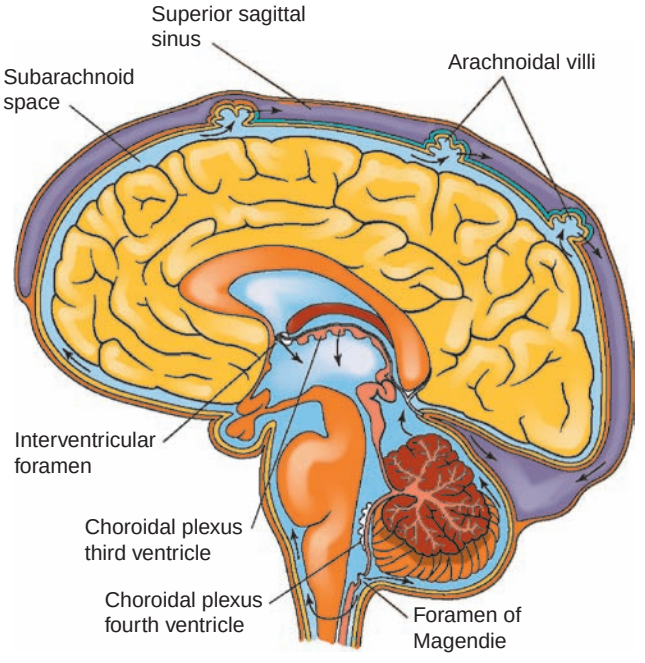
NPH and scheduled for surgery to place a ventriculoperitoneal (VP) shunt.

Characteristic triad

Mr. S experienced a triad of neurologic signs and symptoms characteristic of NPH: gait disturbance, cognitive impairment or dementia, and impaired bladder control. The disorder was first recognized in the 1950s before technology had been developed to continuously monitor intracranial pressure (ICP).[2] Because many patients presented with seemingly normal ICP, the disorder was termed "normal pressure" hydrocephalus. Although the name has stuck, it's somewhat misleading because patients' ICP can fluctuate widely.[3]

An estimated 750,000 Americans have NPH. From 9% to 14% of these patients reside in assisted-living facilities or other long-term-care facilities.[2] Approximately 20% or less of patients living with NPH receive the proper diagnosis and treatment.[4] Instead, they may be misdiagnosed with Parkinson disease, Alzheimer disease, or another form of dementia because signs and symptoms for these disorders are similar.

Normal flow of CSF



CSF circulation is a closed system. Most of the CSF produced daily by choroid plexuses in the two lateral ventricles is reabsorbed into the arachnoid villi, which are projections from the subarachnoid space into the brain's venous sinuses. CSF flows in one direction through the arachnoid villi and drains into the superior sagittal sinus.

The arachnoid villi have been compared to pressure-sensitive valves. When CSF pressure is greater than venous pressure, CSF leaves the subarachnoid space. As pressures equalize, the valves close. Dysfunction of CSF flow is seen in hydrocephalus.

Source: Hickey JV. *The Clinical Practice of Neurological and Neurosurgical Nursing.* 7th ed. Philadelphia, PA: Wolters Kluwer, Lippincott Williams & Wilkins; 2009:60.

By one estimate, 375,000 Americans diagnosed with dementia or Parkinson disease may in fact have NPH, but the true prevalence of NPH is difficult to determine because so many patients are misdiagnosed with other neurologic disorders.[5,6] The number of patients experiencing NPH is expected to rise due to an increase in adult longevity.[7]

Signs and symptoms of NPH are potentially reversible if the disorder is diagnosed and treated early. Unfortunately, 80% of NPH cases aren't recognized by PCPs.[4] Mr. S is an example of a patient whose signs and symptoms were incorrectly attributed to Parkinson disease and dementia.

How NPH develops

NPH is characterized by impairment of CSF absorption (see *Normal flow of CSF*). CSF accumulates in the ventricles of the brain largely because of decreased CSF reabsorption by the arachnoid villi. The enlarged ventricles can cause vascular and periventricular damage due to increasing edema. Neurologic signs and symptoms of NPH arise as the brain tissue is compressed from CSF accumulation and increasing edema.[5]

NPH causes physical and mental changes that lead to patient suffering and decreased quality of life. Untreated, it can also cause premature death. Signs and symptoms of NPH usually progress over months or years, but can occur suddenly if the patient experiences acute head injury or trauma.[8]

NPH is one of the few causes of dementia that may be reversible with treatment.[3] Early diagnosis provides the patient with the best chance to improve quality of life and return to an independent state of living.[8] To help patients achieve the best outcome, nurses and other healthcare professionals must be

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able to recognize its signs and symptoms, and PCPs should consider it in the differential diagnosis.[5]

The following discussion takes a closer look at the classic signs and symptoms of NPH and how they may be differentiated from other possible diagnoses. Keep in mind that a patient with NPH may not have all three manifestations.[7]

Gait disturbance. The most prominent clinical feature of NPH, and usually the first to appear, is gait disturbance or instability, which affects about 89% of patients with NPH.[7] Patients develop a shuffling gait and take wide-based steps, which puts them at an increased risk for falls.[9] Mr. S experienced frequent falls at home due to the changes in his ability to ambulate. Patients can also experience dizziness, causing a loss of balance when changing positions or turning; this increases the likelihood of a fall.[10] Because gait disturbances are progressive, patients who aren't treated will slowly lose the ability to walk.

When taking a health history, inquire about gait and problems ambulating. Listen for complaints of "heavy feet" or feeling as though the feet are "glued to the floor." These signs should alert clinicians to a possible NPH diagnosis.[4]

The gait impairments seen with NPH, such as a shuffling gait, are similar to those seen with Parkinson disease. However, patients with Parkinson disease have a narrow-based gait and less difficulty with feelings of imbalance. Additionally, symptoms of Parkinson disease improve with a daily regimen of levodopa, which doesn't affect gait disturbances in patients with NPH.[11]

Cognitive impairment. This may manifest as a subtle or severe change in mentation. Patients have trouble remaining attentive and recalling information. They're also slow to respond during conversation, as Mr. S's family noticed. Treatment with VP shunting may lead to full restoration of cognition.[12]

NPH is one of the few causes of dementia that may be reversible with treatment.

Mr. S's attentiveness improved briefly after the lumbar puncture when a small amount of CSF was removed, indicating he'd benefit from a surgical shunt. Patients who have Alzheimer disease wouldn't improve after a lumbar puncture and would exhibit other signs of memory difficulty, aphasia, and difficulty with simple tasks that aren't associated with NPH.[5] Other differential diagnoses that should be ruled out are vascular dementia, brain tumor or infection, hypothyroidism, multiple brain infarcts, and chronic alcoholism.[8]

Urinary incontinence. Patients experiencing incontinence due to NPH have difficulty with frequency and urgency. NPH-induced incontinence is easily misdiagnosed because many older adults suffer from this disorder due to other comorbidities, such as cystitis, bladder cancer, or benign prostatic hyperplasia (BPH).[3]

In the early stages of NPH, patients may experience urinary urgency rather than incontinence.[1,5] However, gait impairment may prevent them from getting to the bathroom in time, leading to incontinence.

Incontinence alone isn't diagnostic for NPH because it's often not seen in the early stages of NPH. In addition, older adults may not report incontinence as a potential problem if they attribute urinary difficulties with aging and other factors such as use of diuretics or BPH.[8] Patients who view incontinence as an embarrassment may not report it to the nurse or PCP.[10]

A complete health history and physical assessment should be conducted for patients exhibiting signs of NPH as well as a thorough neurologic evaluation.

Diagnostic testing

No gold standard test exists for the diagnosis of NPH.[1,13] Computed tomography and MRI are used to determine ventricle size and identify possible hydrocephalus, but these tests alone aren't always sufficient to confirm an NPH diagnosis. Other invasive diagnostic studies may be used, such as a lumbar tap test via a lumbar puncture, or continuous drainage of CSF in a hospital setting (also known as external lumbar drainage) for up to a week to monitor for improvement in signs and symptoms.[1,4,10] Over an extended period, ICP monitoring may reveal intermittent rhythmic pressure deviations.[1]

Treating NPH

After diagnosis, patients are evaluated for the best treatment option. Surgical placement of a VP shunt is the standard treatment for NPH.

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(See *How ventricular shunting takes the pressure off*.) One study found that patients have a 96% chance of symptom improvement after shunt placement.[14]

The risks and benefits of surgery are assessed for each patient. Patients who have multiple comorbidities, particularly those related to cardiovascular and cerebrovascular disorders, have a higher risk for surgical complications and poorer outcomes.[6,7]

An externalized ventriculostomy drain (EVD) may be placed before a patient undergoes shunt surgery to assess the benefits of placing a permanent VP shunt.[9] If a patient has a positive response to CSF drainage, either through the lumbar tap test or EVD, then the patient is considered an appropriate candidate for a VP shunt.

The goal of shunt surgery is to maintain adequate drainage of CSF in order to improve the patient's preoperative signs and symptoms and quality of life.[10] After VP shunt placement, 80% of patients experience improvement in signs and symptoms as early as 2 days postoperatively.[12] Surgical recovery and improvement in clinical features progress over a period of months, with gait disturbances improving in 30% to 95% of patients with properly functioning shunts.[5]

Possible complications of VP shunt placement include infection of the shunt catheter, intracerebral hematoma, seizures, and shunt malfunction, such as catheter misplacement or kinking. If the patient doesn't experience symptom improvement after shunting, follow-up imaging studies are performed to assess the ventricles and catheter.[5]

The neurosurgeon schedules Mr. S for placement of a VP shunt to help him regain his independence and improve his quality of life. The patient will require follow up with the neurosurgeon and post-op education from nursing staff.

Overdrainage of CSF is a common and potentially serious complication of VP shunting, particularly in the first year.

Post-op nursing responsibilities

After shunt placement, nurses should assess for clinical improvement as well as complications. Cognition improvement is usually the most apparent change after shunt placement, followed by reversal of incontinence, and lastly stability of gait.[9]

Nurses perform frequent neuro assessments in the inpatient setting to assess for complications after VP shunt placement. Postoperative headaches, fatigue, and insomnia may signal complications and should be reported promptly to the neurosurgeon.[5]

Nurses also monitor for signs of infection at the incision site and perform daily dressing changes.[15] Patients and families should be taught how to perform the dressing changes, if necessary, and to assess for signs and symptoms of infection such as erythema and edema at the incision site.

Nurses must ensure that patients and their families know when to notify the neurosurgeon; for example, if signs and symptoms experienced preoperatively begin to reappear. This could be a sign that the shunt is malfunctioning and may need to be revised.[5]

Overdrainage of CSF, which may lead to subdural effusion or hematoma, is a common and potentially serious complication of VP shunting, particularly in the first year.[1] Tell patients to report sustained or postural headaches. However, because this complication may be asymptomatic, also emphasize the importance of routine follow up with the surgeon as directed.

Nursing's key role

Untreated, patients with NPH will begin to lose vital functions, such

How ventricular shunting takes the pressure off

A ventricular shunt consists of a shunt catheter, a reservoir, and a one-way valve. A small incision is made behind the ear and the proximal portion of the shunt catheter is implanted into the lateral ventricle through a burr hole. The catheter is connected to an inline reservoir and one-way valve, which collect CSF and control the pressure and rate at which the system shunts fluid out of the ventricle. The distal portion of the shunt catheter is tunneled under the skin to a terminal point in the peritoneum, pleura, or vena cava. The shunt is left in place permanently unless it becomes dislodged, plugged, or infected, in which case it's surgically removed and replaced.

Source: Hickey JV. *The Clinical Practice of Neurological and Neurosurgical Nursing.* 7th ed. Philadelphia, PA: Wolters Kluwer, Lippincott Williams & Wilkins; 2009:323.

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as the ability to think clearly, walk independently, and maintain control of bodily functions.[11] But signs and symptoms of NPH are dramatically reversed for most patients who undergo ventricular shunting.[7] Early recognition of NPH signs and symptoms and prompt treatment support the best possible patient outcomes.

Nurses have a vital role in assessing patients for neurologic changes and identifying signs and symptoms suggesting NPH.[13] Whether neurologic changes are subtle or sudden, NPH should be considered as a possible cause to prevent misdiagnosis and a treatment delay.

The patient's family should be educated about signs and symptoms of NPH as well, because family members will note subtle changes sooner than healthcare providers. Nurses can promote education and awareness of NPH signs and symptoms for patients like Mr. S, whose families may be struggling to identify the cause of their loved one's neurologic decline. ■

REFERENCES

1. Graff-Radford NR. Normal pressure hydrocephalus. UpToDate. 2016. www.uptodate.com.

2. Wallenstein MB, McKhann GM. Salomon Hakim and the discovery of normal-pressure hydrocephalus. *Neurosurgery.* 2010;67(1):155-159.

3. Hydrocephalus Association. Normal pressure hydrocephalus. www.hydroassoc.org/normal-pressure-hydrocephalus.

4. Kiefer M, Unterberg A. The differential diagnosis and treatment of normal-pressure hydrocephalus. *Dtsch Arztebl Int.* 2012;109(1-2):15-25.

5. Fraser C, Stark SW. Gait disorder in older adults: is it NPH? *Nurse Practitioner.* 2011;36(3):14-20.

6. Paranathala MP, Sitsapesan H, Green AL, Cadoux-Hudson T, Pereira E. Idiopathic normal pressure hydrocephalus: an important differential diagnosis. *Brit J Hosp Med.* 2013;74(10):564-570.

7. Johnson M, Graham K. The diagnosis and surgical treatment of normal-pressure hydrocephalus. *J Am Acad Physicians Assistants.* 2010;23(5):51-56.

8. Siedlecki SL. Normal pressure hydrocephalus: are you missing the signs? *J Gerontol Nurs.* 2008;34(2):27-33.

9. Hickey JV. *The Clinical Practice of Neurological and Neurosurgical Nursing.* 7th ed. Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins; 2013.

10. Williams MA, Relkin NR. Diagnosis and management of idiopathic normal-pressure hydrocephalus. *Neurol Clin Pract.* 2013;3(5):375-385.

11. Bugalho P, Alves L, Miguel R. Gait dysfunction in Parkinson's disease and normal pressure hydrocephalus: a comparative study. *J Neural Transmission.* 2013;120(8):1201-1207.

12. Jaraj D, Rabiei K, Marlow T, Jensen C, Skoog I, Wikkelsø C. Prevalence of idiopathic normal-pressure hydrocephalus. *Neurology.* 2014;82(16):1449-1454.

13. Thynne K. Normal pressure hydrocephalus. *J Neuroscience Nurs.* 2007;39(1):27-32.

14. Halperin JJ, Kurlan R, Schwalb JM, Cusimano MD, Gronseth G, Gloss D. Practice guideline: idiopathic normal pressure hydrocephalus: response to shunting and predictors of response. *Am Acad Neurology.* 2015;85:2063-2071.

15. Hepburn-Smith M, Dynkevich I, Spektor M, Lord A, Czeisler B, Lewis A. Establishment of an external ventricular drain best practice guideline: the quest for a comprehensive, universal standard for external ventricular drain care. *J Neurosci Nurs.* 2016;48(1):54-65.

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