How can you best diagnose idiopathic normal pressure hydrocephalus?

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Evidence-based answer

Diagnose idiopathic normal pressure hydrocephalus (INPH) by clinical history, brain imaging, physical findings, and physiological criteria.

The clinical examination must show the characteristic gait disturbance and either impaired cognition or impaired urinary continence (strength of recommendation [SOR]: **B**, based on systematic review of small randomized controlled trial [RCT] and prospective trials).

The cerebrospinal fluid (CSF) opening pressure should be between 70 and 245 mm H_2O (SOR: **B**, based on systematic review of small RCT and prospective trials). No single test has sufficient sensitivity to rule out the diagnosis of INPH (SOR: **B**, based on systematic review of small RCT and prospective trials).

Clinical commentary

Subtle clues help make the diagnosis

Normal pressure hydrocephalus is primarily diagnosed clinically. The classic triad of gait instability, cognitive dysfunction, and urinary incontinence, however, seldom present together. The only promising diagnostic and therapeutic intervention is the response observed with a ventriculoperitoneal shunt. However, this intervention is invasive and not without risks. Neuroimaging plays a role, but only when the clinical suspicion is high.

Therefore, understanding the subtleties in the character of the gait, the time of onset, the progression of dementia, and the onset of urinary incontinence in relationship to one another helps in making the final diagnosis.

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FAST TRACK

To diagnose idiopathic normal pressure hydrocephalus, an exam must reveal the characteristic gait and impairment in either cognition or urinary continence

Evidence summary

Current uncertainty in diagnostic criteria makes estimates of the incidence of INPH unclear, but it is thought to cause fewer than 5% of cases of dementia.[1]

Two systematic reviews have looked at the question of diagnosing INPH.[2,3] Unfortunately, there is no definitive test or physical finding for INPH. For patients over 40 years of age, INPH has an insidious onset, a progressive course, and lacks an identifiable antecedent cause. A brain imaging study reveals ventricular enlargement not attributable to other causes. Some suggest that the diagnosis be assessed as "probable," "possible," and "unlikely" based on the degree of fulfillment of a set of historical, imaging, clinical, and physiological criteria (**TABLE**).[3]

CONTINUED

FAST TRACK

In idiopathic normal pressure hydrocephalus, brain imaging studies will reveal ventricular enlargement not attributable to other causes

TABLE

Categorizing the likelihood of idiopathic normal pressure hydrocephalus[3]

Probable INPH

HISTORY (MUST FULFILL ALL)

- Insidious onset over age 40
- Progression over at least 3 to 6 months
- No evidence of an antecedent event known to cause secondary hydrocephalus
- No other neurological, psychiatric, or medical condition sufficient to explain the presenting symptoms

BRAIN IMAGING (MUST FULFILL ALL)

- A CT or MRI study showing evidence of ventricular enlargement not entirely attributable to cerebral atrophy
- No macroscopic obstruction to cerebrospinal fluid flow
- At least 1 of the following: enlargement of the temporal horns of the lateral ventricles, colossal angle >40 degrees, evidence of altered brain water content, flow void on MRI

CLINICAL

- Evidence of a gait/balance disturbance must be found, plus at least 1 other area of impairment in cognition, urinary continence, or both

Gait/balance should reveal *at least* 2 of the following 9 items:

1. Decreased step height
2. Decreased step length
3. Decreased speed of walking
4. Increased trunk sway during walking
5. Widened standing base
6. Toes out when walking
7. Retropulsion
8. Turning *en bloc*
9. Impaired walking balance

Tests of cognition should show evidence of *at least* 2 of the following 7 characteristics that are not fully attributable to other conditions:

1. Psychomotor slowing
2. Difficulty dividing or maintaining attention
3. Decreased fine motor speed
4. Decreased fine motor accuracy
5. Impaired recent memory formation or executive function, such as insight, performance of multistep procedures, or formation of abstractions/similarities
6. Behavioral changes
7. Personality changes

Symptoms of urinary incontinence not attributable to other primary urological disorders should be present:

- Episodic or persistent urinary incontinence
- Fecal incontinence
- Any 2 of the following: urinary urgency, urinary frequency (>6 voids per 12 hours), nocturia (>2 voids per night)

PHYSIOLOGICAL

- Cerebrospinal fluid opening pressure on lumbar puncture should be in the range of 70–245 mm H_2O (or 5–18 mm Hg)

Which patients will benefit from shunting?

Supplemental prognostic tests have been developed to help decide which patients are most likely to benefit from a ventriculoperitoneal shunt. Complicating comparisons between the various tests is the lack of a standard set of measures of function in gait, cognition, and urination; nor is there agreement on how long after shunting the clinician should make these measurements.

A systematic review[4] of the most commonly used prognostic tests identified a response to a large-volume (40–50 mL) CSF tap test as having a positive predictive value (PPV) between 73% and 100% but a negative predictive value (NPV) of only 23% to 42%. Thus, observing an improvement of function after such a test is a good predictor of improvement after shunting, but many patients who do not respond to the test respond to shunting.

TABLE

Categorizing the likelihood of idiopathic normal pressure hydrocephalus[3] (continued)

Possible INPH

HISTORY (MUST FULFILL ALL)

- Reported symptoms begin earlier than 40 years of age or show lack of progression
- There are remote antecedent events such as head trauma, intracerebral hemorrhage or meningitis
- The coexistence of other neurological, psychiatric, or medical conditions that make it difficult to attribute symptoms to just idiopathic normal pressure hydrocephalus

BRAIN IMAGING (MUST FULFILL ALL)

- Cerebral atrophy is sufficient to potentially explain observed hydrocephalus
- Structural lesions are present that may influence ventricular size

CLINICAL

- There are symptoms of incontinence or cognitive impairment without observable gait/balance disturbance
- Isolated gait/balance disturbance or cognitive impairment is observed

PHYSIOLOGICAL

- Opening cerebrospinal fluid pressure has not been measured or it falls outside the required range for probable idiopathic normal pressure hydrocephalus

A variation of the CSF tap test is the extended lumbar drainage test, which involves placing a lumbar intrathecal catheter and allowing the drainage of 10 mL of CSF/hour for 72 hours. The PPV for this test ranges from 80% to 100%, and NPV from 66% to 100%.

A third possible test is the measurement of resistance to an infusion of saline into the lumbar subarachnoid space (CSF Ro test). This test has multiple variations of technique. Reported values for PPV are 75% to 92%, and for NPV of 27% to 92%.

Other tests, such as radionuclide cisternography or magnetic resonance imaging CSF flow void, have predictive values too low or have too few studies to be recommended.[4]

Recommendations from others

A recently published expert consensus statement proposes that the diagnosis of INPH be made using the history, clinical examination, and neuroimaging.[4] Cases of probable INPH can proceed directly to ventriculoperitoneal shunt, or supplemental testing can be used to improve the certainty of a positive shunt response. A positive CSF tap test should lead to shunting.

Follow up negative tap tests with the extended lumbar drainage test or the CSF Ro test (or both). A positive response to any of the tests should lead to shunting; negative responses to all the tests indicates a low chance (<10%) of responding to a ventriculoperitoneal shunt.[4] ■

FAST TRACK

Improved function after a large-volume CSF tap indicates a good chance the patient will respond to shunting

References

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