

NORMAL-PRESSURE HYDROCEPHALUS (NPH)

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Primary Disciplinary Field(s): Neurology, Neurosurgery, Geriatrics

1. Core Definition

Normal-Pressure Hydrocephalus (NPH) is a neurological syndrome characterized by the abnormal accumulation of cerebrospinal fluid (CSF) in the cerebral ventricles, resulting in ventricular enlargement, without the sustained presence of significantly elevated intracranial pressure (ICP). This paradoxical situation--ventriculomegaly coexisting with average ICP readings--is what lends the disorder its descriptive name. Though traditionally associated with elderly populations, NPH can affect adults of various ages, such as in the provided clinical example where a patient was diagnosed at 54. The condition is often referred to informally as **low-pressure hydrocephalus** due to the absence of the high-pressure spikes typical of acute obstructive hydrocephalus.

The core definition highlights a fundamental disturbance in CSF dynamics. While CSF is continuously produced within the choroid plexus of the ventricles, its normal absorption into the venous system via the arachnoid granulations is compromised in NPH. Although the pressure may register within the normal range when measured via standard lumbar puncture, the impairment in drainage leads to a chronic, subtle pressure gradient and mechanical stress on periventricular white matter tracts. This pressure, even if numerically "normal," is pathologically significant because it overcomes the brain's capacity for compliance, driving ventricular expansion over time.

NPH is broadly categorized into two types: idiopathic NPH (iNPH), where the cause is unknown and is most common in older adults, and secondary NPH (sNPH), which arises following a specific identifiable insult such as trauma, subarachnoid hemorrhage, or meningitis. The clinical importance of NPH lies in its potential reversibility; unlike many neurodegenerative disorders, NPH symptoms can often be significantly alleviated through surgical intervention, provided the diagnosis is accurate and timely.

2. Clinical Triad and Symptoms

The classical presentation of NPH involves the **Hakim Triad**, a cluster of three specific symptoms first described by Dr. Salomón Hakim: gait disturbance, cognitive impairment (dementia), and urinary incontinence. While the presence of all three symptoms is highly suggestive of NPH, patients frequently present with only one or two, complicating early diagnosis. The severity of symptoms can fluctuate, but they typically worsen progressively without treatment, significantly impacting the patient's quality of life and independence.

The most characteristic and often earliest symptom observed is **gait disturbance**. Patients typically exhibit a broad-based, shuffling gait (often termed magnetic gait), characterized by

difficulty lifting the feet and hesitation during initiation of movement (gait apraxia). This gait abnormality is disproportionate to any findings of muscular weakness or sensory loss and often resembles features seen in Parkinson's disease. Crucially, the gait disorder in NPH is generally the symptom most likely to show improvement following successful therapeutic shunting, making it a key prognostic indicator.

The **cognitive impairment** associated with NPH is predominantly subcortical dementia. This manifests as slowness of thought (bradyphrenia), executive dysfunction (difficulty planning, organizing, and sequencing tasks), and apathy. Memory loss is generally less pronounced in the early stages compared to typical Alzheimer's disease, although severe forms of NPH can lead to profound global cognitive decline. The third component, **urinary incontinence**, usually presents as urgency and frequency in the initial stages, progressing to frank incontinence as the disorder advances, reflecting the involvement of frontal lobe pathways controlling micturition.

3. Pathophysiology and Etiology

The underlying pathophysiology of NPH centers on chronic mechanical stress and alterations in cerebral blood flow caused by impaired CSF circulation. In iNPH, the exact mechanism is elusive but is thought to involve microvascular disease and increased resistance to CSF outflow at the level of the arachnoid villi, possibly due to age-related fibrosis or minor ischemic insults. This increased resistance prevents the CSF from being reabsorbed effectively, leading to back pressure and compensatory ventricular dilation.

The ventricular expansion exerts pressure on the surrounding brain tissue, particularly the **periventricular white matter** tracts that contain crucial motor and cognitive pathways. The stretching and potential demyelination of these fibers--including descending motor tracts and fronto-subcortical circuits--are directly responsible for the gait disturbance and executive dysfunction. Furthermore, the dilation may compromise the microcirculation in the periventricular zone, leading to chronic ischemia and secondary damage.

In cases of sNPH, the etiology is clearly linked to prior conditions that directly disrupt the subarachnoid space and the CSF drainage system. For example, a previous **subarachnoid hemorrhage** or bacterial meningitis can cause inflammation and scarring of the arachnoid villi, mechanically blocking the pathways responsible for reabsorption. In both idiopathic and secondary forms, the resulting hydrostatic pressure, even if appearing normal, is sufficient over time to deform the brain structures necessary for normal motor and cognitive function.

4. Diagnosis and Diagnostic Criteria

Diagnosing NPH is challenging because its clinical presentation overlaps significantly with more common neurodegenerative diseases like Parkinson's disease and Alzheimer's disease. A

comprehensive diagnosis relies on integrating clinical assessment, neuroimaging, and specific CSF dynamics testing. The primary goal of the diagnostic process is to confirm the presence of NPH and, crucially, to predict the likelihood of a positive response to therapeutic shunting.

Neuroimaging, typically via MRI or CT scan, is essential to document ventriculomegaly. A key imaging sign that supports the diagnosis is Disproportionately Enlarged Subarachnoid Spaces Hydrocephalus (DESH), which involves enlarged ventricles coupled with tight subarachnoid spaces over the high cerebral convexity and medial fissures, but with enlarged fissures around the basal cisterns. This pattern helps distinguish NPH from hydrocephalus ex vacuo (ventricular enlargement due to global brain atrophy). The patient's history, often revealing the onset in older adults, aids in refining the diagnostic probability curve.

The most definitive diagnostic step involves testing the patient's response to CSF removal. The **tap test**, or high-volume lumbar puncture (LP), involves removing 30-50 ml of CSF and observing for transient, measurable improvement in gait or cognition within 24 to 72 hours. A positive response strongly indicates that the patient will benefit from shunt surgery. In cases where the tap test is inconclusive, specialized monitoring techniques, such as continuous intracranial pressure monitoring or external lumbar drainage (ELD) over several days, may be employed to assess CSF dynamics and therapeutic potential more accurately. Reliable diagnostic criteria are paramount given the invasiveness of the primary treatment.

5. Treatment and Management (Shunting)

The definitive treatment for NPH is the surgical implantation of a shunt system, designed to divert excess CSF from the cerebral ventricles to another body cavity where it can be absorbed. The most common procedure is the implantation of a **ventriculoperitoneal (VP) shunt**, which routes CSF into the peritoneal cavity of the abdomen. Less common routes include ventriculoatrial (VA) shunts (diverting to the heart's right atrium) or lumboperitoneal (LP) shunts (diverting from the lumbar spine).

Shunt systems consist of a catheter placed in the ventricle, a valve mechanism that regulates flow, and a distal catheter. The valve is critical, as it controls the rate and pressure at which CSF drains. Modern systems often utilize **programmable valves**, allowing neurosurgeons to non-invasively adjust the pressure settings post-surgery to optimize drainage and minimize the risk of complications. This careful management is necessary because, although shunting often reaps advantages, problems might arise if the pressure is not meticulously regulated.

While shunting can yield dramatic improvements, particularly in gait disturbance, it carries inherent surgical risks and potential long-term complications. These complications include **shunt malfunction** (catheter blockage or breakage), which requires immediate surgical revision; infection (often by skin flora, requiring shunt removal and antibiotic treatment); and, critically, risks

associated with over-drainage. Over-drainage can cause the brain to shift rapidly, potentially leading to a subdural hematoma, requiring careful monitoring and adjustment of the valve settings to maintain optimal intracranial fluid balance.

6. Prognosis and Differential Diagnosis

The prognosis for NPH is relatively favorable compared to many other conditions causing dementia in older adults, earning NPH the moniker of "treatable dementia." Success rates for shunting, defined as significant and sustained improvement in at least one key symptom, typically range between 50% and 80%, with the best outcomes observed in patients whose primary symptom is gait disturbance and who show a robust positive response to the CSF tap test. However, the prognosis is poorer if cognitive symptoms are severe or if the patient has significant comorbid cerebrovascular disease.

Differential diagnosis is a crucial step in management due to the symptomatic overlap with other disorders. For instance, the gait disturbance in NPH can be mistaken for Parkinson's disease, while the cognitive decline may resemble Alzheimer's or vascular dementia. NPH is distinguished by the specific combination of the triad symptoms, the absence of early severe rigidity or resting tremor (characteristic of Parkinson's), and the positive response to CSF drainage testing. Given that the typical NPH patient is an older adult, careful differentiation is necessary to prevent unnecessary surgery in patients suffering from truly irreversible neurodegeneration.

Ongoing research focuses on identifying better biomarkers for iNPH to improve diagnostic accuracy and refine patient selection for shunting. These efforts aim to enhance the predictability of surgical success and reduce the frequency of complications, ensuring that the benefits of shunting are maximized for this frequently missed, yet treatable, condition.

Further Reading

[Normal-pressure Hydrocephalus \(Wikipedia\)](#)

[Normal-Pressure Hydrocephalus Diagnosis and Treatment \(Mayo Clinic\)](#)

[Normal-Pressure Hydrocephalus Fact Sheet \(National Institute of Neurological Disorders and Stroke\)](#)