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The Primary Care Guide To Assessment And Management Of Normal Pressure Hydrocephalus (NPH)

1. Overview of NPH

Normal pressure hydrocephalus is a chronic condition that is uncommon in most clinical practices. NPH achieves health consumer recognition because shunting may improve neurological function or cognition in selected patients. The actual frequency of NPH is undetermined although this clinical condition is relatively uncommon in postmortem series. The diagnosis of NPH requires a multifaceted evaluation (1). The clinical criteria that distinguish individuals who will benefit from shunting procedures, from those with static disease remain controversial. Multiple studies have examined the diagnosis, natural history, and responsiveness to surgical intervention of NPH. The peer-reviewed data on NPH is limited by a small number of studies that include a small number of patients using a variety of diagnostic methodologies and surgical interventions. Few studies provide tissue diagnosis to confirm a diagnosis of NPH. Younger patients with chronic obstructive hydrocephalus manifest symptoms similar to those of older individuals with NPH (2).

2. Diagnostic Features of NPH

The clinical triad of NPH includes instability of gait, urinary incontinence, and cognitive impairment. Brain imaging usually identifies ventriculomegaly, consistent with hydrocephalus in the absence of lesions that would have produced obstruction of CSF flow.

The diagnosis of NPH is actually a misnomer because the pressure is not normal. This name occurred because opening pressures during lumbar puncture often appear within the normal range. The pressure effect is mediated by pressure waves that occur over time. The second component to diagnosing NPH includes either imaging, diagnostic trial of CSF drainage to determine clinical improvement or other studies, such as CSF outflow conductance.

3. Dynamics of CSF

Cerebral spinal fluid is produced in the choroid plexus throughout life and flows through the Foramen of Monroe through the third ventricle and into the cerebral aqueducts. The CSF passes through the fourth ventricle in the brain stem and out through the Foramen of Luscha and Magendie where it circulates around the spinal cord and brain and up to the arachnoidal granulations which are embedded in the sagital sinus. Obstructive hydrocephalus can occur from obstruction in the flow of CSF or disruption in the reabsorbption within the arachnoidal granulations. Inflammatory processes produced by infection or hemorrhage in the arachnoidal space can produce thickening of the lepto-meninges that reduce or obstructs effective re-absorption of CSF (3).

4. Treatment of NPH

The therapeutic intervention for NPH is ventriculoperitoneal (VP) shunting. VP shunting has also been proposed as potentially beneficial to persons with Alzheimer's disease. The drainage of CSF from the ventricular systems was theorized as reducing the overall amyloid burden in the brain.

VP shunting is relatively safe and shunt failure is relatively uncommon. A small percentage of elderly individuals will develop subdural hematomas (2-3%) or spontaneous hemorrhages as a consequence of VP shunting. CNS infections are a rare complication from this procedure (4).

Functional brain imaging, such as SPECT scanning of NPH, demonstrates diffuse hypometabolism that does not improve with reduction of CSF pressure (9). MRI assessments of patients with NPH fail to demonstrate comorbid white matter lesions that would explain the gait abnormality.

The overall success rate of VP shunting in persons with NPH depends upon the criteria used to identify "success" and the selection of patients who undergo procedure **(See Table 1).** The number of studies and study populations are small. Insufficient data is available for an adequate meta analysis.

Table 1. Outcomes from Shunting for Elders with Normal Pressure Hydrocephalus (NPH)

n	Duration	Outcome	Ref.
151	1 yr.	Improved gait and ADL function	1
29	3 mos.	72% improved gait and 29% improved cognitive testing	8
11	6 mos.	Variable results with some cognitive improvement	7
95	1 yr.	Variable results: CSF outflow resistance is best prognosticator for improvement	6
12	1 yr	Improvement: 100% gait, 90% sphincter control, 33% dementia	5
132	3 mos.	33% improvement global function	10
	6 mos.	60% improvement global function	10
	24 mos.	75% improvement global, 93% gait	10

Duration: time from shunt to follow-up assessment

5. Pathological Diagnosis of NPH

The pathological diagnosis of NPH includes evidence for ventriculomegaly as well as absence of diseases that could explain enlarged ventricles (3). Biopsy assessment during shunt placement show that almost half (42%) have the histopathology of AD (4). Ventriculomegaly is a common pathological finding in most dementias because the ventricles enlarge as brain tissue is lost. Individuals with NPH should have no other disease process that could explain enlargement of ventricles through loss of brain parenchyma. The actual obstruction to outflow is rarely identified in postmortem specimens of clinically confirmed NPH.

6. Conclusions About NPH

NPH is a difficult clinical diagnosis to confirm because many persons with Alzheimer's disease have ventriculomegaly and difficulties with gait. Additional diagnostic procedures are encouraged to confirm a diagnosis of NPH rather than performing a shunt based on clinical history and ventricular enlargement on brain imaging. Therapeutic expectations are modest for most individuals and clinicians can advise families that gait and incontinence may improve with shunting.

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