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Evaluation and Management of Normal Pressure Hydrocephalus in the Primary Care Setting

Normal pressure hydrocephalus (NPH) is an abnormal accumulation of cerebrospinal fluid (CSF) in the brain which causes the ventricles to minimally enlarge. This article will briefly discuss clinical symptoms, diagnostic features, clinical outcomes and limitations of prognostic studies that every primary care provider should know in order to diagnose this condition in a timely manner and potentially prevent the development of dementia and significant morbidity in their patients.

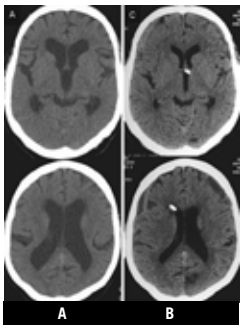
Clinical Symptoms

NPH is characterized by a triad of symptoms including abnormal gait, early stage dementia, and urinary incontinence. Patients are typically diagnosed during their 6th or 7th decade of life. However, the symptoms of NPH can be variable and subtle, and the classic triad of symptoms may not be present in all patients.

Typically, gait apraxia is the first presenting symptom and can be a significant problem. It is manifested by a slow gait, shorter shuffling steps and imbalance offset by a wide stance – sometimes referred to as "magnetic gait". Additional movement disorder symptoms including apraxias (inability to perform learned tasks or movements) of limb and trunk movements, bradykinesia and other parkinsonism symptoms may be present.

Symptoms of incontinence generally occur later in the course of the disease. In the early stages, urinary urgency and frequency is present in most patients but sphincter muscles are unaffected. These initial urinary symptoms are difficult to differentiate because they could be present with several other common disorders in this patient population, like prostatism, recurrent urinary infections, bladder dystonia, or pelvic floor disorders.

Mental status changes may often resemble depression and cognitive symptoms may be subtle. Slowing of thought, inattentiveness, apathy and recall problems are characteristic to the subcortical dementing process. The presence of dementia is extremely variable and cognitive symptoms are usually least responsive to intervention. Surgical treatment is usually discouraged in patients with advanced degree of dementia, even in the presence of incontinence and gait dysfunction.



Brain CT before (A) and after shunting (B)

Diagnosis

Although the related literature is not entirely conclusive about NPH, it is suggested that early diagnosis is optimal for a variety of reasons. Various conditions can be ruled out, some of which – subdural hematoma, brain tumors, or intracranial infection – may be life threatening. Other diagnoses are frequently confused with NPH including Parkinson's disease (PD), Alzheimer's disease (AD), and atherosclerosis. Early diagnosis allows for timely treatment in those patients that are most likely to benefit.

Alzheimer's disease and NPH can often be distinguished by imaging studies, clinical assessment and neuropsychological testing. Mental impairment symptoms are "cortical" (e.g. aphasia, apraxia, agnosia) and diagnosed early in the course of AD. Conversely, NPH causes "subcortical" abnormalities (e.g. slow information processing, difficulty with complex tasks, memory impairment with intact recognition) that manifest later in the course of the disease.

Parkinson's disease and NPH can cause similar gait disturbances – hypokinesia, freezing, imbalance, extrapyramidal symptoms – which could be attempted to differentiate with a trial of levodopa. A number of other conditions have similar clinical presentation and should be included in a patient work-up such as depression, subcortical arteriosclerotic or other vascular encephalopathies, peripheral neuropathy and B12 or folate deficiency.

While MRI or CT scans reveal ventricle enlargement, imaging tests alone are not sufficient to establish the diagnosis of NPH and all clinical symptoms should be considered. Ventriculomegaly is a common finding, but is itself not diagnostic of NPH. Other radiographic findings may include temporal horn enlargement, periventricular edema and signal changes, or an aqueductal/fourth ventricle flow void. Imaging studies also help to rule out ventricular obstruction, structural lesions and other often coexisting conditions such as cerebrovascular disease.

A cerebrospinal fluid opening pressure measured by a lumbar puncture is another useful diagnostic tool, which typically falls within the normal or slightly elevated range, but should not exceed 24.5 cm H2O.

A positive response to shunt placement is suggested as a reliable tool to diagnose NPH, but this method does not exclude the NPH diagnosis in patients with concomitant conditions. High volume lumbar puncture or the 72-hour external lumbar drainage test, "temporary" shunting, may be used to evaluate a potential benefit of the treatment.

The presence of certain symptoms could also help to rule out the diagnosis of NPH, which includes papilledema indicating increased intracranial pressure, or seizures and headaches presenting as manifesting symptoms.

According to evidence-based guidelines and classification developed by an NPH independent study group in 2005 (Relkin et al, Neurosurgery 2005), there are three categories of idiopathic NPH: probable, possible and unlikely. Unfortunately, these categories are exclusively symptom-based and more research is needed to evaluate their prognostic value.

Treatment

Normal pressure hydrocephalus is most commonly treated with a surgical placement of a shunt system, which drains CSF into another area of the body (ventriculoperitoneal, lumboperitoneal, ventriculoatrial shunting) where the body can absorb it.

Surgical interventions performed in the early stages of the disease seem to have the greatest benefit. As with any surgical procedure, a risk-to-benefit ratio should be carefully considered before offering patients a CSF shunting procedure. Such factors as advanced age and function, comorbidity, immune incompetence and functional and coagulation status should be taken into account. It is advised to stop systemic anticoagulation perioperatively, thus the expected improvement from shunting should justify the added risk in anticoagulated patients. A CSF shunting procedure may not always prevent further progression of the disease, but even a temporary improvement may make a significant difference in a patient's life. Even patients with a progressive decline in gait lacking the other aforementioned triad symptoms who have enlarged ventricles on imaging studies that can not be explained by any other conditions should be considered candidates for CSF shunting.

Clinical Outcomes and Prognosis

Overall, improvement rates regarding CSF shunting range from 30% to 96% in the literature (Klinge et al., Neurosurgery, 2005). This wide variation in outcomes most likely suggests that divergent definitions of clinical improvement were used and emphasizes the need for careful patient selection. The development of safer and more effective programmable shunts may reduce the rates of associated complications, especially those related to both overshunting and undershunting.

Response to shunting varies significantly between patients and further supports the importance of correctly identifying patients who most likely would benefit from shunting. Patient selection should be based on clinical and diagnostic findings, although sometimes additional adjunctive testing is necessary to confirm the diagnosis. Some of these tests may be beneficial in predicting a positive response to the CSF shunting procedure, but cannot indicate its sustainability over time.

Based on the current literature, the best and more consistent results are obtained in patients with the typical clinical triad in addition to the imaging criteria with improvement rates ranging from 61% to 77% (Black, J Neurosurg 1980; Benzel et al., Neurosurgery, 1990). In a long-term study that compared clinical outcomes, the shunt-treated patients had a lower need for care than the non-shunted patients (Savolainen, et al, Acta Neurochir (Wien), 2002). Early identification and selection of good candidates for the shunting procedure can potentially reverse or relieve symptoms and provide these patients with an improved quality of life.

This brief summary is not intended to be comprehensive and should not take the place of specialized evaluation and management of cranial and spinal conditions. For more information and to discuss a potential patient in further detail please call us at 303.938.5700, or email us at info@bnasurg.com. Information is also available on our website at www.bnasurg.com.

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