

## Normal pressure hydrocephalus

A topic in the Alzheimer's Association® series on understanding dementia.

### About dementia

Dementia is a condition in which a person has significant difficulty with daily functioning because of problems with thinking and memory. Dementia is not a single disease; it's an overall term — like heart disease — that covers a wide range of specific medical conditions, including Alzheimer's disease. Disorders grouped under the general term “dementia” are caused by abnormal brain changes. These changes trigger a decline in thinking skills also known as cognitive abilities, severe enough to impair daily life and independent function. They also affect behavior, feelings and relationships.

Brain changes that cause dementia may be temporary, but they are most often permanent and worsen over time, leading to increasing disability and a shortened life span. Survival can vary widely, depending on such factors as the cause of dementia, age at diagnosis and coexisting health conditions.

### Normal pressure hydrocephalus (NPH)

Normal pressure hydrocephalus (NPH) is a brain disorder in which excess cerebrospinal fluid (CSF) accumulates in the brain's ventricles, which are fluid-filled chambers. NPH is called “normal pressure” because despite the excess fluid, CSF pressure as measured during spinal tap is often normal. As brain ventricles enlarge with excess CSF, they can disrupt and damage nearby brain tissue, leading to difficulty walking, problems with thinking and reasoning, and loss of bladder control.

NPH can sometimes be treated with surgical insertion of a shunt, a long, thin tube that drains excess CSF from the brain to the abdomen. Surgery is most likely to help correct difficulties walking, but thinking changes and loss of bladder control are less likely to improve. Shunting doesn't help everyone with NPH, and there's uncertainty about how best to identify those most likely to benefit. There's also a lack of data showing how long the benefit of shunting may last for those whose symptoms improve.

### Prevalence

NPH primarily affects people in their 60s and 70s. The Hydrocephalus Association estimates that nearly 700,000 adults have NPH, but it is often misdiagnosed as Alzheimer's or Parkinson's disease. In fact, less than 20 percent of people with the disease are properly diagnosed.

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## Symptoms

The following symptoms are considered hallmarks of NPH:

- **Difficulty walking** that's sometimes compared to the way a person walks "on a boat," with the body bent forward, legs held wide apart and feet moving as if they're "glued to the deck."
- **Mild dementia** that involves loss of interest in daily activities, forgetfulness, difficulty completing routine tasks and short-term memory loss.
- **Decline in thinking skills** that includes overall slowing of thought processes, apathy, impaired planning and decision-making, reduced concentration and changes in personality and behavior.
- **Loss of bladder control**, which tends to appear somewhat later in the disease than difficulty walking and cognitive decline.

## Diagnosis

Because the symptoms of NPH are similar to Alzheimer's disease, Parkinson's disease and Creutzfeldt-Jakob disease, NPH is often overlooked or misdiagnosed. The three hallmark NPH symptoms are considered the "classic" clinical picture, but not everyone with NPH experiences all three symptoms. In a Mayo Clinic study, among 41 older adults with suspected NPH, all had difficulty walking, 30 experienced cognitive decline, and 14 reported loss of bladder control. Only 12 of the 41 had all three symptoms.

To confirm a diagnosis of NPH one or more of the following tests are done:

- **Brain imaging:** Imaging of the brain's structure to detect enlargement of the ventricles, often with magnetic resonance imaging (MRI) or CT scan, plays a key role in diagnosing NPH. Several brain disorders, including Alzheimer's disease, can cause overall brain tissue shrinkage that makes the ventricles look larger than normal. In NPH, brain tissue may not appear shrunken even though the ventricles are enlarged.
- **Clinical examination:** Because the clinical picture for NPH may vary and symptoms may overlap with those of Alzheimer's and other dementias, experts recommend that a person with suspected NPH undergo examination by a neurologist with extensive experience evaluating brain disorders that affect movement, thinking skills and physical functions.

- **Cerebrospinal fluid (CSF) tests:** CSF tests to predict shunt responsiveness and/or determine shunt pressure include lumbar puncture, external lumbar drainage, measurement of CSF outflow resistance, intracranial pressure (ICP) monitoring and isotopic cisternography.

### **Causes and risk factors**

In some cases, NPH is caused by other brain disorders such as a tumor, head injury, hemorrhage, infection or inflammation. But in most cases, the cause of the fluid buildup remains unknown.

### **Outcomes**

Few studies have explored the benefits of shunt insertion. Most of these studies were small and followed participants for a limited time. The SINPHONI-2 trial results, published in the June 2015 issue of *Lancet*, found that people with NPH might benefit from lumboperitoneal shunt surgery but further studies need to be done to confirm the findings. Available data suggests that difficulty walking is the symptom most likely to improve. Several studies found a significant rate of postsurgical complications and also showed that the short-term benefits of shunt insertion tend to decline over time.

### **Treatment**

NPH is one of the few causes of dementia that can be controlled or reversed with treatment. If symptoms and results from an evaluation and MRI point to NPH, a high-volume spinal tap may be used to identify if an individual has the potential to benefit from surgical insertion of a shunt. In this procedure, doctors remove a large amount of spinal fluid and observe the individual for 30 to 60 minutes, looking for any improvements in walking or thinking and reasoning. Most people originally suspected of having NPH do not improve following a CSF removal test.

Researchers have not found effective nonsurgical treatments for NPH. Drugs that remove excess fluid throughout the body, such as diuretics, don't appear to improve symptoms of NPH. More research is needed to understand the prevalence of NPH; show how the excess CSF involved in NPH causes symptoms affecting movement, thinking and bodily functions; and clarify the possible benefits and ideal targets of shunt insertion.

TS-0095 | Updated December 2018