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 lifespan. 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 ventricle, which are hollow, 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. Difficulty walking is the symptom most likely to improve after surgery. Thinking changes and bladder control are less likely to get better. Shunting doesn't help everyone with NPH, and there's uncertainty about how best to identify those most likely to benefit. The effectiveness of shunting in NPH has never been demonstrated in a randomized clinical trial. 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. Scientists aren't certain how many older adults have this disorder, in part because common symptoms of NPH area also common in other brain disorders.

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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."
- **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

There are no standard diagnositic criteria for NPH. The three hallmark NPH symptoms are considered the "classic" clinical picture, but not everyone with NPH has all of these symptoms. In a recent Mayo Clinic study, among 41 older adults with suspected NPH, all had difficulty walking, 30 had cognitive decline and 14 had loss of bladder control. Only 12 of the 41 had all three symptoms.

Imaging of the brain's structure to detect enlargement of the ventricles, often with magnetic resonance imaging (MRI), plays a key role in diagnosing NPH. Several brain disorders, inlcuding Alzheimer's disease, can cause overall brain tissue shrinkage that makes the ventricles look larger than normal. In NPH, altough the ventricles are enlarged, brain tissue may not appear shrunken.

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 neurologist with extensive experience evaluating brain disorders that affect movement, thinking skills and physical functions

If symptoms evaluation and an MRI strongly suggest NPH, a large-volume spinal tap may be used to identify those with a reasonable likelihood of benefiting from surgical insertion of a shunt. In this procedure, doctors remove a larger-than-usual amount of spinal fluid, and then observe the person for 30 to 60 minutes to note any improvements in walking or thinking and reasoning. Most people originally suspected of having NPH do not improve following a CSF removal test.

Causes and risk factors

In some cases, NPH is caused by other brain disorders such as hemorrhages, infections or inflammation. But in most cases, the fluid buildup happens for unknown reasons.

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Outcomes

There are few studies of the benefits of shunt insertion, and none of them are randomized clinical trials. Most of these studies were small and followed people for a limited time. Available data suggest that difficulty walking is the symptom most likely to improve. Several studies found a significant rate of postsurgical complications. Finding also showed that short-term benefits of shunt insertion tended to decline over time.

Treatment

Researchers have not found effective nonsurgical treatments for NPH. Drugs that remove excess fluid throughout the body, such as diuretics, haven't been shown to help NPH. More research is needed to understand the prevalence of NPH; to show how the excess CSF involved in NPH causes symptoms affecting movement, thinking and bodily functions; and to clarify the possible benefits of shunt insertion and who is most likely to benefit.

TS-0095 | Updated May 2015

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