What is Normal Pressure Hydrocephalus (NPH)?

The brain and spinal cord are surrounded by a clear fluid called cerebrospinal fluid (CSF). This fluid is produced and stored in cavities in the brain called ventricles. It circulates around the brain, moving from ventricle to ventricle. The purposes of the fluid are to cushion and protect the brain and spinal cord, to supply them with nutrients, and to remove some of their waste products. Any excess fluid drains away from the brain and is absorbed by other tissues.

Hydrocephalus is a condition in which there is too much CSF in the ventricles. This occurs when the natural system for draining and absorbing extra CSF does not work right. The ventricles enlarge to accommodate the extra fluid and then press on different parts of the brain, causing a number of different symptoms. Hydrocephalus has many different causes. Some people are born with the condition, while others develop it during their lives.

Normal pressure Hydrocephalus (NPH) is a type of Hydrocephalus that occurs in adults, usually older adults. The average age of people with NPH is older than 60 years. NPH is different than other types of Hydrocephalus in that it develops slowly over time. The drainage of CSF is blocked gradually, and the excess fluid builds up slowly. The slow enlargement of the ventricles means that the fluid pressure in the brain may not be as high as in other types of Hydrocephalus. However, the enlarged ventricles still press on the brain and can cause symptoms. (The term “normal pressure” is somewhat misleading.)

The parts of the brain most often affected in NPH are those that control the legs, the bladder, and the “cognitive” mental processes such as memory, reasoning, and problem solving, and speaking. This decline in mental processes, if it is severe enough to interfere with everyday activities, is known as dementia. Other symptoms include abnormal gait (difficulty walking), inability to hold urine (urinary incontinence), and, occasionally, inability to control the bowels.
Note: The symptoms of NPH can be similar to those of diseases such as Alzheimer’s and Parkinson’s. Experts believe that many cases of NPH are misdiagnosed as one of these diseases.

**What causes NPH?**

Normal Pressure Hydrocephalus can occur after a head injury, bleeding around the brain (due to a blow to the head), stroke, meningitis (infection of a protective layer of tissue around the brain), or brain tumour. It can also happen after surgery on the brain. How these conditions lead to NPH is not clear. In most cases, the cause of NPH is never known.

**What are the symptoms of NPH?**

At first, the symptoms of normal pressure Hydrocephalus are usually very subtle. They worsen very gradually.

Symptoms of dementia include:
- Memory loss
- Speech problems
- Apathy (indifference) and withdrawal
- Changes in behaviour or mood
- Difficulties with reasoning, paying attention, or judgment

Walking problems
- Unsteadiness
- Leg weakness
- Sudden falls
- Shuffling steps
- Difficulty taking the first step, as if feet were stuck to the floor
- “Getting stuck” or “freezing” while walking

Urinary symptoms
- Inability to hold urine
- Inability to hold stool, or faeces (less common)
- Frequent urination
- Urgency to urinate
Normal Pressure Hydrocephalus

The following symptoms can be related to increased pressure in the brain:
- Headache
- Nausea
- Difficulty focusing eyes

When to Seek Medical Care for NPH

Some people think that memory loss, difficulty finding words, walking problems, or urination problems are normal parts of aging. In many cases, however, these are symptoms of treatable conditions. Any of these problems, or changes in mood or behaviour, warrants a visit to your health care provider.

How is NPH Diagnosed?

The symptoms of Normal Pressure Hydrocephalus can occur in Alzheimer’s disease and Parkinson’s disease. However, the combination of dementia-like symptoms, walking problems, and urinary problems should alert your health care provider to the possibility of NPH. Making the distinction is very important because the treatments for these conditions are quite different. Tests are available that can diagnose NPH. At any point in this process, your health care provider may refer you to a specialist in brain disorders (neurologist or neurosurgeon) to complete the evaluation and begin treatment.

What is the Treatment for NPH?

Normal Pressure Hydrocephalus generally cannot be cured. It is a long-term condition. However, many people with the condition obtain substantial relief through surgical treatment. For those who are not candidates for surgery, treatment consists of measures to relieve mood and behavioural problems, cope with physical problems such as incontinence and walking difficulties, and maximize physical, mental, and social functioning.

Surgery for NPH

Occasionally the cause of the Hydrocephalus can be treated directly through surgery. For example, a brain tumour blocking drainage of the CSF can be removed. In most cases, however, the underlying problem is not known or cannot be treated. The treatment in these cases is a shunt operation.
Normal Pressure Hydrocephalus

A shunt is a thin tube that is implanted in the brain by a neurosurgeon. It is inserted into the ventricles to drain excess CSF away from the brain. The tube is routed under the skin from the head to another part of the body, usually the peritoneum (the lower belly). The shunt is equipped with a valve that opens to release fluid when the pressure builds up. The fluid drains harmlessly and is later absorbed by the bloodstream. The pressure setting on the valve sometimes must be readjusted. The newer shunts allow adjustment without another operation.

A shunt operation is not a cure. It does not treat the underlying cause of NPH. It can, however, relieve the symptoms. The shunt remains in place indefinitely. If properly implanted, the shunt often is not obvious to other people.

Shunt operations do not work for everyone with NPH. Many people who undergo a shunt operation have substantial symptom relief. In some, the symptoms improve and then start to worsen again. Others benefit little, if at all. Even the experts are not able to predict perfectly who will benefit and who will not. Many surgeons perform a spinal tap before surgery to test whether the symptoms get better with removal of fluid. In some cases, the person is hospitalized for a few days while fluid is drained slowly through a small tube called a catheter. This is another way of checking whether removing extra fluid will help symptoms.

The earlier the NPH is diagnosed, the better the chances that the surgery will help. In general, people with milder symptoms have better outcomes with this surgery. Like any surgery, the shunt operation can cause complications. Such complications include infection of the shunt and blood clots around the brain. Your neurologist or neurosurgeon will discuss the pros and cons of this operation and whether it might work for you.

Another operation is sometimes used instead of shunt placement. In Endoscopic Third Ventriculostomy, an endoscope (thin tube with a lighted camera on the end) is used to create a small hole in the floor of the ventricles. The hole provides another way for CSF to drain from the brain.
Normal Pressure Hydrocephalus

Prevention of NPH

There is no known way to prevent NPH. A healthy lifestyle, including not smoking, maintaining a healthy weight, and regular exercise, may help avoid conditions such as high blood pressure, heart disease, diabetes, and stroke that might contribute to NPH. Wearing a seatbelt and safety helmet when indicated can help avoid head injury, another cause of NPH.

For further information on Shunts and Endoscopic Third Ventriculostomy please see “What is a Shunt” and “What is Endoscopic Third Ventriculostomy”