

# B.R.A.I.N. Tool

## Idiopathic Normal Pressure Hydrocephalus (iNPH)

### Understanding Hydrocephalus in Adults

Your brain contains billions of cells and is the control centre of your mind and body. It is protected by your skull, cushiony membranes, and an essential liquid called cerebrospinal fluid, or CSF. We all need CSF. It delivers important nutrients and chemicals from the blood to the brain, removes waste products from the brain, and protects this complex, essential organ.

CSF is continuously produced inside four ventricles, or chambers, in the brain. Normally, CSF flows freely from one ventricle to the next before it exits the brain via the spine and is reabsorbed. However, when the flow of CSF to the outside of the brain is interrupted or blocked, too much CSF accumulates. This causes the ventricles to swell which puts pressure on the brain and can cause serious damage. The excess accumulation of CSF is called hydrocephalus.

### Adult Onset Hydrocephalus

When hydrocephalus is diagnosed in an adult, it may be associated with acquired causes, may be associated with previously unrecognized congenital causes or it may be idiopathic Normal Pressure Hydrocephalus (iNPH).

The acquired causes of hydrocephalus in adults may be similar to its causes at other ages. They include:

- brain cysts or tumours
- meningitis or encephalitis
- traumatic brain injury
- certain strokes and brain hemorrhages

The previously unrecognized congenital causes of hydrocephalus in adults may include:

- aqueductal stenosis
- brain cysts and certain tumours

Hydrocephalus does not always appear immediately after these causes occur, and it may be years or decades before the symptoms become evident.

### Symptoms of Adult-Onset Hydrocephalus

- headaches
- nausea
- blurry eyes, focusing
- irritability
- drowsiness
- seizures
- drowsiness
- unsteady walk or gait
- leg weakness
- sudden falls

### What is Idiopathic Normal Pressure Hydrocephalus?

Never heard of idiopathic Normal Pressure Hydrocephalus (iNPH)? You're not alone. iNPH is not widely known and is frequently misdiagnosed as "old age problems" or Alzheimer's or Parkinson's disease. A neurological disorder, INPH affects more than 1 in every 200 adults over the age of 65. This means



thousands of people, who have much to live for, may be unnecessarily losing their independence, time with loved ones and admitted to long term care facilities or die prematurely. If you suspect someone you know may have INPH, be sure to ask about it specifically and insist the proper diagnostic testing be done.

A significant challenge in getting appropriate care for possible iNPH is lack of awareness. Getting an early diagnosis and proper treatment can be difficult because the public and most family physicians are unfamiliar with the condition.

Unfortunately, too many people with INPH are incorrectly diagnosed or are misdiagnosed with diseases

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associated with aging. A delayed diagnosis can be devastating and cause irreversible brain damage.

### How Do You Recognize INPH?

INPH is characterized by three classic symptoms. Once they're identified, a CT or MRI scan is the first critical step towards an accurate diagnosis.

### Symptoms of INPH?

#### **Difficulty Walking**

Usually the most pronounced and first to appear, gait disturbances range from mild imbalance or instability to an inability to stand or walk. Walking difficulties occur in almost all patients with INPH. Dementia without walking difficulties usually does not occur with INPH. A person's gait may be wide-based, short-stepped, slow and shuffling. Individuals have trouble picking up their feet and often trip and fall. They describe the feeling as "feet stuck to the floor." Turning around often requires many small steps.



#### **Problems with Memory**

This forgetfulness, difficulty performing routine tasks and short-term memory loss may be moderate or severe. Cognitive symptoms may be overlooked for years or accepted as part of aging. People with INPH may have obvious cognitive impairment or conversational skills and thinking abilities may be unchanged. Cognitive changes may be detectable only with formal neuropsychological testing.

#### **Loss of Bladder Control**

This can range from urinary frequency and urgency in mild cases to complete loss of bladder control in severe cases, when urinary urgency is strong and cannot be controlled. Not all individuals with INPH display obvious signs of bladder problems.

### How is INPH Diagnosed?



Once a physician suspects INPH, testing is usually carried out to confirm the diagnosis and assess the individual's suitability for treatment. It is important that a neurosurgeon or neurologist be part of the medical team at this point, to interpret test results and discuss surgery and its risks. The medical team will consider the pattern and sever-

ity of impairments, along with the results of other tests, in differentiating INPH from other conditions such as Alzheimer's or Parkinson's disease.

Physical and neurological examinations will be used to evaluate the symptoms. There will be discussion and observation of walking and turning to determine the extent and type of gait disturbance. Asking questions or administering neuropsychological evaluation will test cognition. Areas of examination include attention span, reaction time, memory, reasoning, language and emotional state. There will be verbal assessment of bladder urgency and frequency or incontinence.

An MRI or CT scan will examine the brain to determine if enlarged ventricles exist. Other testing could include CSF tests such as lumbar puncture (spinal tap), external lumbar drainage (ELD), or measurement of CSF outflow resistance and intracranial pressure.

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### Common Tests

Gait Analysis involves walking 10 meters (30 feet) while a neurologist or neurosurgeon observes. They are looking for the telltale shuffling gait. Short steps with feet “stuck to the floor.”

### Neuropsychological Testing (cognitive testing)

A series of assessments to determine if there is a loss of brain function including memory, concentration and problem-solving. These may take from 10 minutes to 2 hours to complete

### INPH is Treatable!

Early diagnosis can increase the chances of successful treatment. Once a physician suspects INPH, testing is usually conducted to confirm the diagnosis and determine suitability for treatment.

If you have any of the symptoms we described, ask your family physician for a referral to a neurologist or neurosurgeon. The evaluation may involve:

- Discussion and observation of walking and turning for gait disturbance;
- Asking questions/administering neuropsychological evaluations for cognition;
- Verbal assessment of urinary urgency and frequency or incontinence;
- MRI or CT scan to detect enlarged ventricles;
- CSF tests such as lumbar puncture (spinal tap) or external lumbar drain (ELD).

If left untreated, INPH usually leads to a progressive decline with worsening symptoms. Falls are common with risk of head injury or broken bones. Often death occurs due to medical causes such as pneumonia, heart disease or stroke.



### Treatments Options for Idiopathic Normal Pressure Hydrocephalus

The seriousness of INPH symptoms may vary from day to day, but the progression is not likely to stop on its own. The only treatment for INPH involves a surgical procedure to implant a shunt. A shunt is a system that is inserted into brain or spinal CSF space to drain CSF into another part of the body, usually the abdomen, where it can be reabsorbed. Shunts typically consist of two tubes with a valve between them. As the fluid pressure inside the brain or spine increases, the valve opens and redirects the excess fluid from the brain to another part of body. Shunt surgery can usually be performed in less than an hour.

There are 3 main types of shunts:

**Ventriculoperitoneal shunt** that drains from the brain into the abdomen.

**Ventriculoatrial shunt** that drains from the brain into a chamber of the heart.

**Lumboperitoneal shunt** that drains from the lower spine (back) to the abdomen.

And there are 2 types of shunt valves:

**Fixed pressure valves** that regulate the flow of CSF on a predetermined pressure setting.

**Adjustable (programmable) valves** can be adjusted non-in-

vasively with magnetic tools to change the pressure setting after surgery.

There are different shunts on the market and your neurosurgeon will choose the option they feel will work best for you. Most people with INPH now have a programmable shunt implanted.

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### Which Treatment is Best?

Many tests and evaluation criteria are considered but unfortunately, no single factor is completely reliable in predicting success from shunt implantation or ETV. The choice of which treatment will best suit a patient depends on the following factors:

- onset of gait disturbances is the first and most prominent symptom;
- monitoring of intracranial pressure or spinal fluid pressure shows an abnormal range or pattern of spinal fluid pressure or an elevated CSF outflow resistance;
- removal of spinal fluid gives significant temporary relief of symptoms;
- there is minimal evidence of other significant brain diseases.

Many people with INPH have additional medical and/or neurological problems. It is important to discuss with the medical team what outcomes can be expected with regard to regaining balance, walking, mental ability or bladder function. While everyone hopes for a full recovery, it may not be seen and establishing reasonable expectations is essential. Many individuals with INPH, appropriately selected to undergo surgery will be satisfied with reduced levels of disability or dependence, or the prevention of further neurological deterioration.

### Success of Treatment

While the CSF removal testing usually predicts that a shunt will improve symptoms, there is currently no way to accurately predict how quickly or to what extent gait disturbance, mild dementia and bladder control problems may improve. Improvements may start within days of surgery, or it may take a few months. Some individuals may quickly reach a plateau, while others may improve over 3-12 months.

Success may also be affected by the presence of other neurological or medical conditions such as blood vessel diseases, high blood pressure, Alzheimers or Parkinson's Disease.

### Complications and Risks Of Surgery

Complications of surgery are rarely life-threatening. It is important to review the specific risks associated with surgery while deciding whether to proceed. These complications may include:

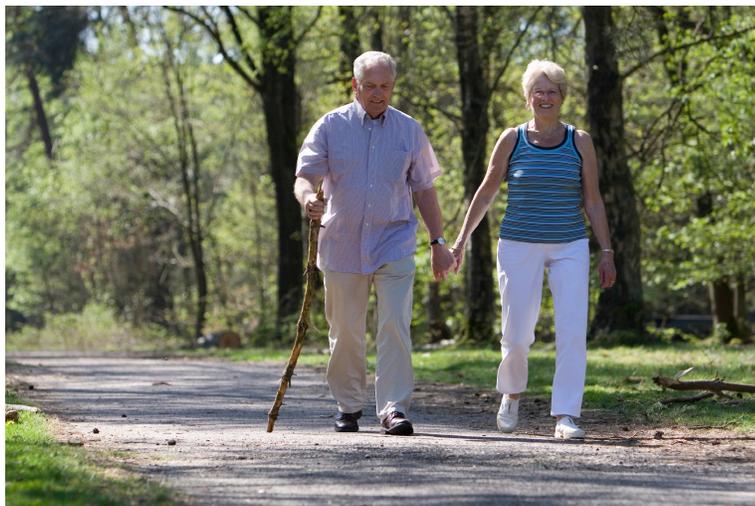
- obstruction and malfunction of the shunt requiring a surgical repair
- shunt/csf infection
- surgical bleeding
- seizures
- reaction to anesthesia
- mild abdominal pain

### Recovery from Hydrocephalus Surgery

After surgery, hospital stay could be 2 to 7 days, depending on how badly mobility was affected before surgery and the rated of clinical progress.

Your doctor will evaluate your condition post-surgery. You may need some rehabilitation therapy to get the most improvement. Recovery may be limited by the extent of damage and

your brain's ability to heal. Hydrocephalus is a life-long condition that requires long-term follow-up by a neurosurgeon. Follow-up could include MRI, CT



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Scan or X-rays to determine if proper drainage of CSF is being maintained.

Potential worrisome postoperative symptoms which should be reported and investigated as soon as possible include:

- redness, tenderness, pain or swelling of the skin along the shunt track and incision
- fever
- irritability or drowsiness
- nausea, vomiting, headache or double vision
- abdominal pain

In addition, if there is a return of previous symptoms, the individual should consult their neurosurgeon or neurologist to ensure that there is no malfunction of the shunt, or other complications such as valve failure or the need for a change of a programmable valve to a higher or lower pressure setting.

### Is INPH Preventable?

Presently, there is no way to prevent INPH. Getting treatment as soon as symptoms appear is key to increase the chance for a full or partial recovery.

### Possible Questions For Patients To Ask

- How much experience do you have in diagnosing and treating INPH?
- Do you see many patients with INPH in your practice? Could you connect me with them?
- Is there another health care provider I should consult with?
- Is this surgery necessary?
- If I don't have the surgery, what can I expect health wise?
- I've never had surgery before. Is there anything you can tell me about your personal experiences with these procedures that will lower my fear?
- What are the risks/complications for shunting in my case?

- What steps will be taken to reduce complications?
- Will you be my neurosurgeon? If not, can I meet with the neurosurgeon prior to my surgery?
- Will you need to cut or shave my head?
- How do you thread the tube under my skin and into the abdomen/heart?
- Will you be using sutures, staples or stitches?
- How long will my surgery be? What should I expect after surgery?
- How long will I be in the hospital?
- When can I bathe/shower after surgery? Do I need to protect the incision area?
- Will I need to do rehabilitation therapy after surgery? Why?
- Will home care be arranged to look after my incision?
- What do I do if I suspect infection?
- Will the sutures/staples/stitches need to be removed? When?
- How long will I need to recover, before I can go back to my daily routine? Work? School? Physical activity?
- What is your surgery follow-up procedure?
- Who do I contact if I have any questions or issues once I leave the hospital? You/your nurse?
- I don't live close to this hospital. If I have an emergency which hospital should I go to?
- Thank you for taking the time to answer my questions. If I have more, can I contact you directly?

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### An Important Reminder....

The good news is that when INPH is diagnosed early and managed properly the life altering symptoms in many cases, can be reversed.

Speak with your doctor as soon as possible if you, or someone you love, is experiencing challenges with:

- memory
- mobility and
- bladder control

Ask your doctor if INPH could be the cause and if testing for INPH is appropriate. Your doctor can start the process by requesting a CT scan or MRI of the brain.



### About Hydrocephalus Canada

Our goal is to empower individuals impacted by hydrocephalus to experience the best life possible. We believe everyone affected by hydrocephalus:

- has the right to our attention, compassion and commitment
- has value and deserves to be treated with dignity
- requires and deserves access to safe, effective care
- benefits from, and offers benefit to, collaborative communities
- has the responsibility to help everyone understand hydrocephalus

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