

What is **Normal Pressure Hydrocephalus?**



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Normal Pressure Hydrocephalus (NPH) is a condition which mainly affects people over 60.

A clear fluid, cerebrospinal fluid (CSF) is constantly produced and absorbed, and flows through chambers within the brain, (ventricles) and around the outside of the spinal cord. CSF protects the brain and spine, provides them with nutrients, and removes toxins.

When more CSF is produced than absorbed, hydrocephalus occurs. In NPH the ventricles become enlarged by too much CSF building up, although the pressures inside the head are often found to be normal. The condition is associated with several symptoms, including difficulty walking, cognitive impairment/dementia, and urinary incontinence.

What causes NPH?

In many people, the cause of NPH is not known but it is usually associated with older age.

In some people NPH can be caused by a previous head injury, a bleed in the brain, or because of infection or a tumour blocking the flow of the CSF. In some cases, the cause can be reduced CSF flow owing to stenosis (narrowing) of the aqueduct which is



a part of the ventricular system. In NPH the excess fluid and enlarged ventricles press on the blood vessels and brain tissue. This can alter the way the brain works, and prevent the brain from removing toxins. These toxins can then build up in the brain and begin to harm brain cells, leading to dementia and loss of brain volume.

Important to know:

There are 2 main forms of NPH:

Idiopathic

In most people with NPH, no cause can be found. This is termed Idiopathic.

Secondary NPH

Secondary NPH occurs as a result of previous blockage or head injury. It may occur many years after injury, or develop when untreated hydrocephalus that has been stable for many years begins to cause symptoms.

Symptoms

Increasing difficulty walking is often the first symptom noticed. The pattern of walking (gait) can become slow and shuffling, with difficulty picking the feet up (sometimes described as 'magnetic gait' or 'sticky feet') or a wide based stance.

Difficulty walking occurs because the enlarging ventricles press on the part of the brain that controls movement of the legs. Balance problems and falls (especially backwards) are also common.

Cognitive impairment in NPH may appear to be like Alzheimer's but it generally progresses at a slower rate, and usually follows difficulties with walking rather than being the first symptom to appear. In addition to memory difficulties, there may be problems with attention and slowness of thought. Some people's behaviour may change, especially if the frontal lobe is under pressure. This part of the brain usually helps us monitor our behaviour so it is appropriate to the situation we are in,

people with frontal lobe impairment may behave with fewer inhibitions or regard for social norms than previously.

Many people also experience issues with their **mental health**, such as depression or anxiety, or with regulating their mood, becoming agitated, angry or frustrated.

Urinary leakage is more common than bowel leakage, but some people may have either or both. Leakage of urine occurs because the area of the brain controlling bladder function is situated just above the ventricles so is easily affected if the ventricles expand. People with bladder involvement describe urgency and frequency of needing to pass urine, with sudden leakage. If combined with walking difficulties, this can make getting to the toilet in time very difficult.

Headaches and fatigue may occur and can vary in intensity between people. There may also be a wide range of neurological symptoms such as numbness, tingling or weakness in any of the arms, legs, or face.



How is it diagnosed and what other tests may I have?

Imaging

A CT scan of the brain would show the ventricles are larger than expected. Enlarged ventricles are a common finding in older people, as the brain tissue tends to shrink. You may need to have an MRI scan to look in more detail for the cause of the larger ventricles (people with certain metal implants may not be able to have an MRI). The MRI may include an additional scan to look at the blood flow and CSF flow in the brain.

Based on the results of the CT and MRI scans you may then have a consultation with a neurosurgeon. Surgery may be offered at this stage, especially if a disproportionately enlarged subarachnoid space hydrocephalus (DESH) is seen on the scan. This means that there is a larger than expected space below the brain in the skull with a narrowing of the space above the brain, as the brain is pushed upwards by the large amount of CSF. This sign can help identify that a shunt may be beneficial. However, other tests may also be performed first.

Lumbar Puncture

One of these further tests may be a lumbar puncture to measure the pressure in the spine, which closely relates to the pressure in the brain. CSF is removed by inserting a needle into the small of the back. Often tests

of walking speed and/or cognition are conducted before and after the removal of CSF by lumbar puncture. With NPH, there may be a rapid improvement in symptoms following CSF removal, and this can indicate that symptoms are likely to improve with insertion of a shunt.

Sometimes CSF is drawn slowly from the lumbar spine over 24 or 48 hours, to mimic how a shunt would work, with walking and cognitive tests before and after. This is called Extended Lumbar Drainage (ELD).

An infusion test can also be performed via lumbar puncture, adding a little fluid and measuring how quickly the CSF is absorbed. This can demonstrate an impaired CSF absorption, supporting a diagnosis of NPH.

Cognitive tests

You may be referred to a neuropsychologist (a psychologist specialising in the effects of brain function on our thinking and behaviour). Their tests assess memory, language use, speed of thinking and spatial skills. The assessment will help with diagnosis, since NPH has a distinct pattern of changes, and give a baseline of your cognitive function before treatment.



Recommended reading...

What other conditions could this be?

NPH can present in a similar way to other conditions experienced by older people, for example Alzheimer's and Parkinson's diseases. NPH is a potentially reversible cause of dementia and could account for around 6% of people with dementia so it is important to consider NPH as a possible cause of dementia symptoms. Other conditions may also be present as well as NPH. It is possible to have both NPH and Parkinson's disease, or NPH and vascular dementia at the same time. This can make diagnosis and predicting someone's improvement after surgery difficult.

What treatment options are there for NPH?

Surgery to divert excess CSF can be offered to people whose tests show it is likely to improve their NPH symptoms.

There are 2 main surgical options:

Shunt

A thin tube from the ventricles to the abdomen to drain CSF. Most people with NPH are fitted with a shunt that can be adjusted to control the pressure (programmable shunt).

ETV (endoscopic third ventriculostomy)

An option for some people with secondary NPH caused by certain blockages such as aqueduct stenosis.



For more information about shunts, visit: www.shinecharity.org.uk/shunts



Why is surgery not an option for me?

Before any procedure is offered, there needs to be a careful consideration of the potential benefits and risks.

Both procedures involve risks and surgery would not be proposed if it is unlikely that it would improve your symptoms.

For some people with additional health conditions, such as heart or breathing difficulties, uncontrolled diabetes, or stroke, surgery may not be offered because of the increased risks associated with having an operation and anaesthesia.

As we all get older, our brains get firmer and less able to expand or squash (compliance). Sudden reductions in pressure in the skull can create a vacuum at the surface of the brain and result in bleeding in or on the surface of the brain, for example subdural haemorrhage. This is more common in older

people, because of the increased firmness of the brain. Age itself is not a barrier to surgery, provided you are in good enough health for the operation.

Any operation in the brain can result in seizures or a stroke. Any procedure could potentially make you worse, although the aim of any procedure would be to improve your symptoms or delay their progression.

Important to know:

If it is felt by your specialist team that surgery is too high risk, or would be unlikely to help, you should be offered other options to help ease and manage your symptoms.

This may include physiotherapy, occupational therapy, or medication. You should not be left without support, and should remain under specialist care, often from the Neurology team, and you should have regular reviews.

Will having surgery completely reverse my symptoms?

A shunt will not necessarily reverse all your symptoms but may slow down their progression. Symptoms may continue to worsen, or some may improve while others stay the same or worsen. It is different for everyone, but generally the earlier treatment is started the better.

Walking is the symptom most likely to improve, and bladder issues the least likely to improve after shunting, so if you experience bladder issues, it is best to seek medical support in addition to your treatment for NPH. If you begin to see a return of your original symptoms after having a shunt, let your neurosurgery team know, as your shunt may need adjusting or checking. Some members have told us that seeing a neurophysiotherapist helped improve their walking and balance.

Why are my symptoms still getting worse after having a shunt/ETV?

It can be very difficult to predict whose condition will improve after surgery. Some people whose tests show a good chance of improvement find they don't improve, or even deteriorate. Some people find their improvement takes several weeks to begin. It is possible, as mentioned above, for other conditions to occur at the same time. These conditions will not be improved by surgery and may continue to worsen. An underlying dementia or Parkinson's disease may become more apparent once NPH is managed effectively.

If your symptoms continue to worsen, it may be due to a failure of the first procedure. If you have had ETV, it may be possible to place a shunt if the ETV doesn't work, so discuss this with your specialist

team. Your shunt may need to be changed due to blockage or infection.

Important to know:

Studies have shown that shunt failures, blockages and infections are much less common in older adults compared to babies and children.

Shunt malfunction will usually lead to a gradual return of your original symptoms, rather than sudden, acute deterioration needing urgent care, that you may have read about in the context of other forms of hydrocephalus.

What happens if I do not have surgery?

If surgery is not an option, there may be the option to take medication to try to improve symptoms and to continue to have physiotherapy and occupational therapy to improve your balance and memory. Your local bladder and bowel service will be able to advise and offer support for incontinence. Over time you may find your symptoms worsen but it is not possible to predict when or how long this would be.

It may be helpful to think about your lifestyle and home environment, and identify changes you might make to ensure you can continue doing things you enjoy, and are important to you, if your mobility declines further. Some people may decide to downsize their home, or move to a home without steps or stairs. Other people may opt to stay put, with adaptations such as a stairlift or downstairs WC. An OT would be able to advise on changes that will keep you safe and independent for as long as possible. This service can be contacted through your local council's Social Services department. If you think you need more care, contact your Social Services and request an assessment of your needs.

It may also be time to consider what would happen if you become unable to make decisions on your treatment or finances. Talking to your solicitor about Lasting Power of Attorney

can give you and your family peace of mind. This would need to be completed while you are still able to make your own decisions, as would creating a Will, so consider talking this over with your family.

What else would be useful to know?

All shunts have different properties, and at certain times, such as preparing for an MRI, this information will be needed. There are also some precautions you need to take with some, but not all shunts, around strong magnetic fields, so it is definitely worth finding out what shunt you have.

Shine has a Shunt Alert Card, on which you or your hospital can record your shunt type and carry with you. This would let emergency services know about your condition and your shunt if you were taken ill.

To order a shunt alert card, email: firstcontact@shinecharity.org.uk call **01733 555988** or visit: shinecharity.org.uk/shuntalertcard



Recommended reading...

If you have a shunt inserted, read Shine's information on shunts. 'Know Your Shunt' explains the importance of knowing the make and model of your shunt www.shinecharity.org.uk/knowyourshunt

Driving and NPH

You must notify DVLA if you have NPH with any symptoms, and complete Form B1. If you have surgery, you will need to surrender your licence for 6 months, or until you are fit to drive again, whichever is longer. You will need to reapply for your licence, and DVLA will request confirmation from your Neurosurgeon that you are fit to drive again. You are always responsible for ensuring you are fit to drive, even if your medical team gives permission.

You do not need to notify DVLA after lumbar puncture, ICP monitoring or having the setting changed on your programmable shunt.



What other support is available?

Occupational Therapy (OT)

This service can assess and advise you on any adaptations or equipment which may make your daily life easier, for example showering, dressing or accessing your bathroom. Disabled Facilities Grants can help pay for adaptations to your home. Contact your local Social Services department for more information.

Physiotherapy

A Physiotherapist can advise on exercises and activities to keep you mobile for as long as possible, as well as mobility aids.

Mental Health

Contact your GP if you need support with your mental health or mood.

Financial support

There are several benefits available for people who have a long-term physical or mental health condition, or disability, such as Attendance Allowance and Personal Independence Payment. Eligibility depends on many factors. See Shine's Benefits section on the website, or contact Shine on 01733 555988 to discuss what is available with one of Shine's Support and Development Workers.

Shine's specialist services

If you have Normal Pressure Hydrocephalus, live with or care for someone who does, or are a professional providing support - we're here to help.

Support and Development Workers (SDWs)

Support and Development workers are on hand in England, Wales and Northern Ireland to help you make informed choices and to provide confidential information, advice and support on any of life's ups and downs.

Common requests include:

- Understanding health conditions and treatment options
- Continence issues
- Support for unpaid carers to access respite and assistance with getting care assessments/care packages
- Housing adaptations
- Benefits for the person with NPH and their unpaid carer

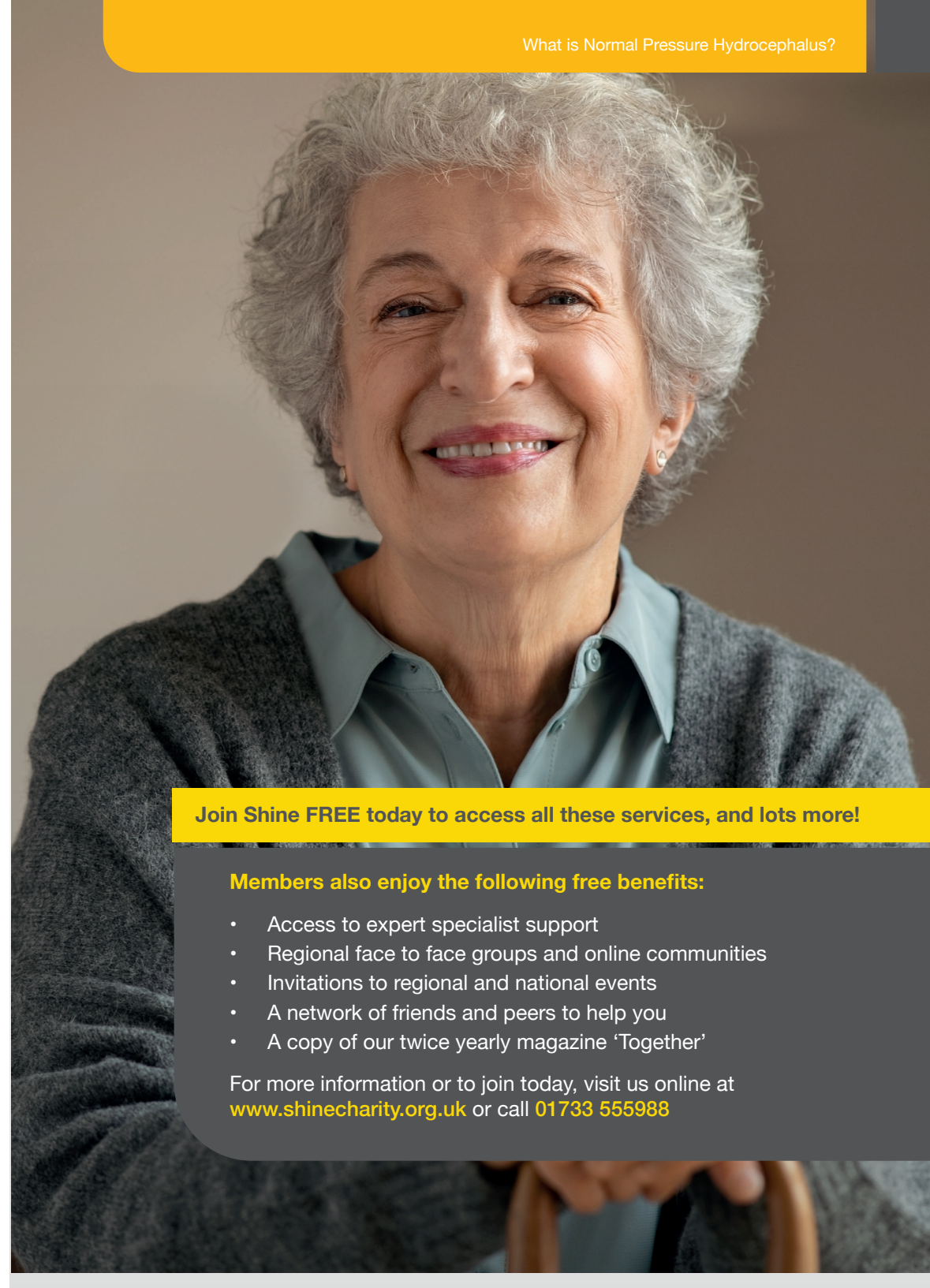
Through our Support and Development Workers you also have access to Shine's additional services, including our Health Team.

Shine's Health Team have significant experience in the conditions we represent and understand the concerns, frustrations and possible choices you might face in gaining an accurate diagnosis and treatment plan for NPH.

As well as supporting NHS specialist clinics, arranging training and information for Shine members, and fellow professionals, and arranging one-stop health checks through Shine's Virtual Health Hubs, they can advise individuals and their loved ones/carers on a one to one basis, on matters such as:

- Getting a diagnosis for NPH
- Shunts and ETVs (endoscopic third ventriculostomy)
- Continence
- Depression or anxiety
- Orthopaedics
- Physiotherapy
- Equipment and adaptations

In addition - for the health-care professionals in your life – our CPD accredited e-learning course on Normal Pressure Hydrocephalus can be accessed from the Professionals area of the Shine website to give professionals a more detailed insight into the condition.



Join Shine FREE today to access all these services, and lots more!

Members also enjoy the following free benefits:

- Access to expert specialist support
- Regional face to face groups and online communities
- Invitations to regional and national events
- A network of friends and peers to help you
- A copy of our twice yearly magazine 'Together'

For more information or to join today, visit us online at www.shinecharity.org.uk or call **01733 555988**

Who are Shine?

With around 12,000 members across England, Wales and Northern Ireland, Shine are Europe's leading charity for people affected by spina bifida and hydrocephalus.

For over 50 years, we've been at the centre of developments which have improved the lives of thousands of people, enabling and empowering our members to lead the lives they want to live.

**Get in touch and
join today for FREE!**



**Many of Shine's
services are entirely
funded by the generosity
of the public**

Without your support, we could not offer vital services that help improve the lives of adults affected by Normal Pressure Hydrocephalus, and their families. Please continue to support us in giving our members the best quality of life.

Donate today at:
shinecharity.org.uk

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