What is ‘normal pressure’ hydrocephalus? Is it different from other types of hydrocephalus?

Normal pressure hydrocephalus (NPH) is caused by an excess of cerebro spinal fluid (CSF) in the brain. Symptoms of NPH include decreased mobility, some degree of dementia and, sometimes, urinary incontinence. The CSF builds up more slowly than other forms of hydrocephalus, and the ventricles (spaces where the fluid is made) expand slowly to accommodate it, but this can still damage the surrounding brain tissue. However, the slow onset means that people with NPH do not have the symptoms of raised intracranial pressure, such as headache, vomiting, nausea, sight disturbances or seizures, that children and young people with hydrocephalus have.

Why is it called ‘normal pressure’ hydrocephalus?

‘Normal pressure’ hydrocephalus (NPH) is so called because the three people first described with the condition (by Hakim and Adams in 1965) had normal pressure CSF, even though their brain ventricles had expanded.

Does hydrocephalus affect people of a certain age or gender?

The likelihood of developing normal pressure hydrocephalus increases slightly with age, so it usually affects people over the age of 50. It seems to affect males and females equally.

Now I have a shunt in place, will my condition deteriorate?

No - having a shunt will control your NPH. It will not, however, prevent those conditions that are part of normal ageing.

Will I need my shunt replaced after a certain period of time?

If your shunt malfunctions then it will need replacing. If everything goes as expected, your shunt should last you for life.

Is NPH hereditary?

No.

What are the short, medium and long term consequences of NPH?

The main problem seems to be lack of confidence in walking – particularly if you have had previous falls. Some people find it harder to remember previously learnt skills such as finding their way around or reading a map. However with practice these skills should return to some extent. Occasionally it is useful to have an assessment with a neuropsychologist who has a special interest in NPH. Your GP will be able to advise you on this. You may also need your shunt adjusting, though not everyone needs this.
What limitations may I experience?
You may find that you become much more tired than you would expect. Some people experience depression when coping with a long term condition, so talk to your GP if you are experiencing a low mood which doesn’t seem to shift.

How often should I have a check-up with the neurosurgeon?
Most neurosurgeons see their patients with NPH once or twice after the shunt is fitted. After that you will probably be discharged and advised to see your GP if you have any problems. The GP may then refer you back to the neurosurgeon, depending what the problem is.

Besides a shunt, are there any other medical and/or surgical remedies available to treat NPH?
A programmable shunt is the Gold Standard treatment for NPH. Trials with the drug acetazolamide have not been very successful and the medicines currently available are used for people who are not able to have surgery.

What should I do if my condition seems to be deteriorating?
Your shunt should stop your condition from getting worse, so if you have increasing difficulty walking, it can be a sign that the shunt needs adjusting. Go to your GP, who should refer you back to your neurosurgeon. However, your GP may be more familiar with the common signs of shunt problems seen in people with other forms of hydrocephalus (headache, vomiting, nausea, sight disturbance, seizures), so may not be aware that a gradual decline in walking is the key sign that a shunt needs adjusting with NPH.

The literature about NPH mentions dementia as a symptom – am I more likely to experience this even if I have been treated with a shunt?
Dementia is one of the symptoms that should lead your doctor to consider a diagnosis of NPH. Having a shunt fitted will usually halt the memory loss, and often the more extreme behaviours seen in some people with NPH.

Are there things I can do to improve my walking?
Just keep on walking! Remember the old saying ‘use it or lose it’. Some people find going to a gym useful – a programme of gentle walking and some leg exercises will help. If you’re having problems getting up and about, ask your GP to refer you to a physiotherapist.

Are there any things I can do to help improve my memory?
Think of your memory as another part of you that needs exercising, so doing things you enjoy such as crosswords, playing card games, reading books and newspapers can help. Joanna Iddon, a psychologist interested in hydrocephalus, has co-authored a book ‘Memory Booster Workout’ with lots of good ideas. Keep a notebook with you and write down everything that you need to remember. Use post-it notes as aides memoires.

I often walk with a ‘drunken’ gait – is this common?
Some people with NPH do experience a rolling gait, often when tired or stressed. If you experience this all the time, go to your GP, as your shunt may need adjusting. However, some people with NPH, especially if they already had poor mobility, do find that a rolling gait is the best they can achieve.
I have heard some people with NPH say they are affected by changes in atmospheric pressure, or that a full moon affects them. Is this common?
Anecdotally, people with all types of hydrocephalus report sensitivity to changes in atmospheric pressure, from headaches to being able to predict thunderstorms. There does not appear to be any science to support this or to suggest that the moon affects people. Some younger (pre-menopausal) women experience cyclical headaches or migraine, usually pre-menstrual.

Occasionally, usually at night, my ears have the sound of the sea, rather like putting a large seashell over your ear. Is this something to do with my shunt?
People with shunts often report hearing “swooshing” noises which they assume is a sign that the shunt is emptying properly. There has been a recorded case of someone with Meniere’s disease (an inner ear disease causing dizziness, nausea) and NPH who reported the noises (called hyperacusis) when the shunt was set with a high opening pressure and which decreased as the pressure was reduced (reported from Geneva 2009). However, there appears to be no scientific reason or research reported.

Can I go through the scanner at airports?
Modern programmable shunts are designed to withstand MRI scanners so should, in theory, be unaffected by airport scanners. But there have been many reports of shunts being accidentally reprogrammed so it is best to be cautious and to avoid airport scanners. You should show your Shunt Alert Card (available from Shine on 01733 555988) and follow the guidelines for people with heart pacemakers.

Can I dye my hair?
Yes you can dye or perm your hair as usual, once the shunt scar has healed. And you can sit under the old-fashioned driers with no ill effects.

At what stage after the onset of NPH should I notify the DVLA and, if so, will they automatically take my driving licence away?
You need to let the DVLA know as soon as you are diagnosed and have a date for surgery. You will not be allowed to drive until at least six months after your shunt is fitted. Contact the DVLA again six months after your surgery, as they will need to ask your neurosurgeon to confirm that you are fit to drive (i.e. have had no seizures or blackouts). If the top end of your shunt (the part in your brain) needs to be revised, you will need to inform the DVLA. You do not need to tell them if you only have your shunt pressure adjusted.

Which companies can I approach for travel insurance?
You will need to shop around, as companies tend to put up their premiums if they have to pay out for a shunt revision.

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