

Normal pressure hydrocephalus

Normal pressure hydrocephalus (NPH) is a rare and somewhat controversial neurological condition, which is said to typically affects older adults and is a potentially reversible cause of dementia.

The brain and spinal cord are surrounded by a clear fluid, called CSF, which is produced and stored in cavities within the brain (ventricles). Hydrocephalus is defined as an abnormal accumulation of CSF in the ventricles of the brain, usually when the natural system for draining and absorbing excess CSF is impaired.

NPH is sometimes suspected when there is a triad of symptoms - walking disturbance, dementia, and urinary incontinence. In NPH the CSF is thought to accumulate gradually and the ventricles enlarge slowly, therefore, there is no increase in pressure within the skull, accounting for the term "normal pressure".

Symptoms

NPH is traditionally characterised by the gradual onset of three symptoms – walking disturbance, dementia, and urinary incontinence. The cardinal symptom is disturbance of balance and walking, including a "shuffling" gait, unsteadiness and falls, which may develop insidiously over months or years. The degree of cognitive impairment is variable, ranging from subtle behavioural and mood changes, to difficulty with reasoning and memory loss. Urinary dysfunction is a common late symptom, including frequency urination, urgency to urinate and incontinence.

Causes

In the majority of cases, the cause of NPH is unknown. However, in some cases NPH may be related to previous a head injury, brain infection, or stroke. These conditions may cause scarring, which interferes with the way CSF drains from the brain.

Controversies about NPH

Some doctors are not sure that NPH does represent a separate condition. Everyone agrees that there are undoubtedly patients with hydrocephalus who present later in life with the symptoms usually associated with NPH. However, it is not clear that these patients do have 'normal pressure', especially if they have a diagnostic test called 'intracranial pressure monitoring' when the pressure is measured over a 24 hour period.

In addition there are many patients who develop the same symptoms as NPH, not because of hydrocephalus but because their brains are getting smaller through old age or vascular disease. As the brain gets smaller the fluid filled spaces inside the brain enlarge and can give an appearance that looks like hydrocephalus.

In this situation it can be difficult to be completely sure that the patient doesn't have hydrocephalus. Because age related brain shrinkage is much more common than hydrocephalus this tends to be the final diagnosis in most patients suspected of having NPH.

Tests

The diagnosis of NPH (or hydrocephalus) may be difficult, as the symptoms are very similar to those of other disorders, such as Alzheimer's dementia or Parkinson's disease. There are several tests available to establish the diagnosis. The most common test is a CT scan or MRI scan of the brain, which will show enlargement of the ventricles, usually without evidence of much 'space' around the outside of the brain.

A procedure called a lumbar puncture (spinal tap), which involves removing CSF from the area around the spinal cord in the lower back, has traditionally been undertaken to see if this affects mobility and thinking. However this is probably not a reliable test. If patients are suspected of having hydrocephalus they will normally need to see a neurosurgeon. Intracranial pressure monitoring can be carried out but even this may not predict who will respond well to surgery.

Treatment

In the first instance, patients should discuss their symptoms with the GP, who should consider referral to a neurologist for further investigations. NPH is a long term condition, which generally cannot be cured. However, symptoms may be improved with medicines and practical support to relieve mood and behavioural problems, and cope with physical problems, such as incontinence and immobility.

In certain cases, a shunt operative may be undertaken – this involves placing a fine tube, known as a shunt to drain out excess CSF and reroute it under the skin to another part of the body, usually to abdomen. The shunt remains in place indefinitely.

This may occasionally lead to substantial relief of symptoms in some patients; however, unfortunately, shunt operations are not beneficial for all patients and are associated with quite high rates of complications (30-50%) such as infections, bleeding and shunt blockages. It is difficult to predict who will benefit from a shunt operation, but patients with mainly walking and urinary symptoms tend to have better outcomes than those with cognitive symptoms.