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PRACTICE

EASILY MISSED?

Idiopathic normal pressure hydrocephalus

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A 74 year old man describes problems with his balance and walking for a year. His family say he is increasingly forgetful, and he has had urinary incontinence recently. He is referred to neurology, where a computed tomography (CT) scan of the brain reveals ventriculomegaly in the absence of substantial sulcal atrophy, consistent with normal pressure hydrocephalus.

What is idiopathic normal pressure hydrocephalus?

A triad of gait and balance impairment, cognitive impairment, and urinary incontinence characterise idiopathic normal pressure hydrocephalus. The symptoms progress insidiously over at least three months. There is ventriculomegaly without marked elevation in cerebrospinal fluid pressure. Normal pressure hydrocephalus may be secondary to alternative pathologies, for example, subarachnoid haemorrhage, meningitis, or traumatic brain injury.[1] The pathogenesis of idiopathic normal pressure hydrocephalus has been widely debated but no single theory has gained widespread acceptance.

Why is it missed?

Multiple case reports and a small, prospective study show that the condition is often overlooked as a cause of hypokinetic gait disorders in elderly people.[2-5] Presentation and severity of symptoms are variable. Often, only one or two symptoms of the triad manifest initially.[1 6] The symptoms may be confused for other conditions in the elderly, particularly Alzheimer's disease, vascular dementia, Lewy body dementia, frontotemporal dementia, Parkinson's disease, and "Parkinson's plus syndromes."[1 7] Additionally, patients may have coexisting pathology, including Alzheimer's disease or subcortical arteriosclerotic encephalopathy, making it difficult to pinpoint a diagnosis.[7 8]

Why does this matter?

Idiopathic normal pressure hydrocephalus is one of the few potentially reversible causes of dementia. Cerebrospinal fluid shunt surgery is the mainstay of treatment.[1 6] A systematic review of observational data after shunt surgery (3063 patients) showed improvement in one or more of the triad components in about 71% of patients.[9] However, there are no randomised controlled data, and most studies are surgical case series designed to evaluate a prognostic or imaging test. The studies vary in criteria for shunt insertion, shunt valve type, and outcome assessment, making it difficult to draw firm conclusions on treatment outcomes.

The natural course of the condition is unclear, but case studies and expert opinion indicate that symptoms deteriorate without surgery, and that longer duration of symptoms is associated with poorer surgical outomes.[1-10]

How is it diagnosed?

The diagnosis often relies on doctors being aware of the usual presentation and considering the condition as a differential diagnosis when a single symptom might be present. The overall clinical picture (see table 1⇓) coupled with suggestive neuroimaging is essential for diagnosis.[6 11]

Clinical features

Evidence summarised from hospital based studies indicates that the triad of gait and balance impairment, cognitive impairment, and urinary symptoms will be present at initial presentation in

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This is one of a series of occasional articles highlighting conditions that may be more common than many doctors realise or may be missed at first presentation. The series adviser is Anthony Harnden, professor of primary care, Department of Primary Care Health Sciences, University of Oxford. To suggest a topic for this series, please email us at practice@bmj.com.

What you need to know

- Consider the diagnosis in patients with progressive gait, balance, or cognitive problems, and/or urinary incontinence over three months
- Refer to neurology for exclusion of alternative diagnoses and neuroimaging. Ventriculomegaly in the absence of substantial sulcal atrophy is typical on neuroimaging
- Shunt surgery is the mainstay of treatment and improves symptoms, though long term data are lacking. Around 1 in 10 patients may experience complications of subdural haematoma, seizures, intracerebral haemorrhage, and infection in the postoperative period

How common is idiopathic normal pressure hydrocephalus?

- Evidence on the epidemiology of idiopathic normal pressure hydrocephalus is poor. Prevalence and incidence statistics are inaccurate and variable[1 6]
- It is classically seen in people >60 years old.[1 6] A retrospective Swedish study found that in those aged 70-79 years the prevalence is 0.2%, whereas in those aged 80-89 years the prevalence is 5.4%[2]
- It is one of the few reversible causes of dementia in the elderly[1]

about 60% of patients.[6] There is no published literature on the positive predictive value and sensitivities of symptoms.

Investigations

A CT scan or magnetic resonance imaging (MRI) of the brain is the next step for those with suggestive symptoms. In normal pressure hydrocephalus, ventricular enlargement is seen out of proportion to sulcal atrophy (fig 1⇓). Other features include widened temporal horns and periventricular lucency.[8 12] Secondary causes of normal pressure hydrocephalus can be identified. MRI has greater sensitivity in identifying changes in the brain in normal pressure hydrocephalus,[1-12] and is always performed before surgery.

A diagnosis of idiopathic normal pressure hydrocephalus is not based on neuroimaging alone, as mild internal hydrocephalus and narrow sulci may be present in healthy elderly individuals.

How is it managed?

A cerebrospinal shunt procedure, typically ventriculo-peritoneal shunt, is the mainstay of treatment.[1 6]

Case studies show that around one in 10 patients will experience subdural haematoma, seizures, intracerebral haemorrhage, and infection in the postoperative period.[9] Surgery is associated with a 1% mortality rate. Few studies assess long term outcomes after surgery. A retrospective study in 55 patients with idiopathic normal pressure hydrocephalus followed them for a mean of 5.9 years and found sustained improvement in all symptoms after shunt surgery, although nearly half of all patients required shunt revision.[13]

Patients are managed by a combination of neurology and neurosurgical input. Annual review by general practitioners should include an inquiry if mobility, memory, and continence are stable or improved. Ensure that patients are coping well in the community and have access to local support services.

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How patients were involved in the creation of this manuscript

We liaised with Shine UK and the National Hydrocephalus Foundation, and both provided us with patient reviewers: 10 patients with idiopathic normal pressure hydrocephalus read through our manuscript and endorsed its content.

Table

Table 1| Clinical symptoms in the diagnosis of idiopathic normal pressure hydrocephalus6

Symptom	Features	Diagnostic value
Gait and balance impairment	Typically the earliest and cardinal feature. Described as “magnetic,” “shuffling,” and “wide-based” Patients seldom have the hallmarks of Parkinson’s disease—that is, tremor, hypomimia, rigidity, and hand apraxia On clinical examination, inspect for a broad based gait, externally rotated foot posturing, and difficulty turning on the body’s long axis.	Evidence summarised from hospital based reports suggests that gait and balance impairment is present in 94-100% of patients at presentation
Cognitive impairment	Impaired attention and concentration, short term memory impairment, and psychomotor slowing Severe cortical deficit (such as aphasia, apraxia, agnosia, and amnesia) and absence of gait dysfunction suggest alternative cortical dementias, such as Alzheimer’s disease	Cognitive impairment seems to be present in 78-98% of patients
Urinary symptoms	Typically the last symptom to emerge in normal pressure hydrocephalus, this may manifest as urgency, frequency (nocturnal), or frank incontinence	Urinary dysfunction present in 76-83% of patients at presentation

Figure

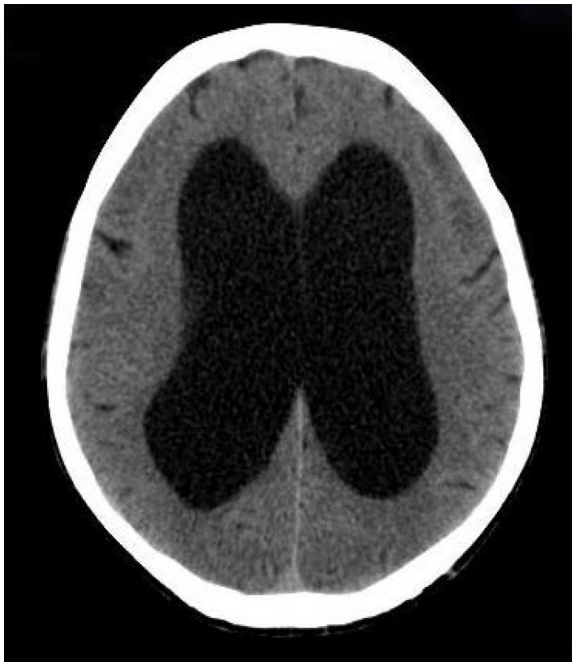

Fig 1 Computed tomography of the brain demonstrating ventriculomegaly that is out of proportion to sulcal atrophy