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NORMAL PRESSURE HYDROCEPHALUS: HOW CAN IT BE TOLD APART FROM NEURODEGENERATIVE DISEASES OF THE ELDERLY?

Raphael C. Zohn

ABSTRACT

Normal pressure hydrocephalus (NPH) affects more of the older population than people recognize. The underestimation of this neurological condition is due in most part to the overlap of its symptoms to other forms of dementia as well as many other geriatric conditions. The objective of this paper was to research and contrast various methods of differentiation in the diagnosis of normal pressure hydrocephalus as well as find pretreatment indicators of successful surgery. Methods included reviewing of articles and studies done to evaluate which symptoms are most commonly presented in normal pressure hydrocephalus and their subtle differences from the symptoms of other neurodegenerative diseases. There are also comparisons of different theories as to the prevalence of normal pressure hydrocephalus and which, if any, symptoms are indicative of a correct diagnosis. Conclusions were as follows: there are guidelines, although controversial, that can be followed in trying to distinguish normal pressure hydrocephalus; there are some symptoms that are better prognosticators of successful surgery than others, and while surgery is often followed by the subsequent relapse of symptoms, this is possibly due to the comorbidity of other disorders with normal pressure hydrocephalus. Surgery should therefore be approached cautiously while weighing the risks versus the benefits. Normal pressure hydrocephalus seems to be fairly prevalent and when appropriate, some older people might be able to reverse their symptoms.

INTRODUCTION

If an elderly man were to walk into his doctor's office complaining of walking difficulties, urinary incontinence, as well as having trouble with memory and hard thinking, the doctor might initially presume the man has Alzheimer's disease. Upon further review, he would be greatly mistaken. Extensive testing can lead to a drastic change in the diagnosis. Intermittently elevated levels of cerebrospinal fluid (CSF) pressure in the lateral ventricles of the brain along with ventriculomegaly (enlargement of the ventricles without a noticeable amount of cortical atrophy) would tell the doctor that the man is suffering from hydrocephalus. Unfortunately, this example is very common. Despite the fact that a patient might be presenting the abovementioned classic clinical triad of symptoms for hydrocephalus, errors all too likely. Normal pressure hydrocephalus (NPH) is a subcategory of communicating hydrocephalus, one of the four main categories of the disease. NPH has primary (idiopathic) and secondary (known) causes. Secondary causes include complications following head trauma, subarachnoid hemorrhage, meningitis, brain tumor, or previous neurosurgical procedures. While people of all ages are affected by secondary NPH (sNPH), idiopathic NPH (iNPH) distinguishes itself as a disease of the elderly. About 50% of NPH cases are idiopathic and half come from a known event. When most think of hydrocephalus, the swelled head of a child comes to mind. Few realize how widespread iNPH is and that many older patients suffer from it. Many doctors don't even notice it and diagnose it as something else. However, if caught early on, iNPH can be treated with a much greater chance of success. The problem arises when it comes to pinpointing all the characteristics of iNPH that can aid in its diagnosis. Its symptoms are not specific to the diagnosis and occur commonly in many other neurodegenerative conditions, such as Parkinson's and Alzheimer's disease (Klassen and Ahlskog 2011). Furthermore, it is often comorbid with other neurological disorders that are actually present in geriatric patients. This doubt makes it

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difficult to elect for surgery and is the primary reason why 80% of cases of hydrocephalus go unrecognized and untreated (Kiefer and Unterberg 2012). Studies done on its prevalence, along with case studies and accounts show that it is far more common than would appear at first glance due mainly to mistakes and the fact there is little awareness of its commonality. The very fact that iNPH is believed to be uncommon, leads to false diagnoses, which in turn causes skewed statistics. While this disease is perceived to be rare, attempts to identify all patients with iNPH overlook a large number of cases (Conn 2011). This paper will explore just how prevalent it is, as well as discuss literature on ways it can be better recognized and diagnosed. Studies indicate that certain symptoms are more indicative of iNPH than others. There are many characteristics of NPH that are specific to it, and can aid in its diagnosis. How can we differentiate a potentially reversible disorder from the more common forms of dementia?

BACKGROUND OF NPH HISTORY

The first mention of hydrocephalus dates back all the way to 2500 BC where references can be found in ancient Egyptian medical literature. It was not until 1000 AD, however, that an operative procedure was performed. An Arab surgeon at that point in time clearly describes the removal of spare cerebrospinal fluid from the skulls of hydrocephalic children. He writes that “the volume of the skull then increases daily, so that the bones of the skull fail to close” (Aschoff et al. 1999). A real treatment for the condition did not enter the scene until the 20th century when shunts and other neurosurgical procedures became popular. The first description of NPH came in 1965 when a small group of patients presented with various neurological symptoms, ventricular enlargement, and what looked like normal cerebrospinal fluid pressure during lumbar puncture. After shunting, their symptoms improved (Vacca 2007). The shunt treatment does not fully heal the patient though, and, to this day, there is no complete cure.

ANATOMICAL DEFINITION

Hydrocephalus can be broadly defined as an abnormal expansion of the lateral ventricles in the brain caused by an accumulation of cerebrospinal fluid. Cerebrospinal fluid cushions the brain and spinal cord in the subarachnoid space between the arachnoid and pia maters that cover them. Cerebrospinal fluid is produced by specialized capillaries known as the choroid plexus. Cerebrospinal fluid flows from the two lateral ventricles through their respective foramina of Monro into the third ventricle. From there it passes through the aqueduct of Sylvius into the fourth ventricle, which is located in the posterior fossa, and then the central canal of the spinal cord. It enters the space in between the meninges through small openings in the ventricular system and covers the brain and spine, acting as a cushion and protecting the brain from shock. From the subarachnoid space, cerebrospinal fluid is absorbed by clusters of arachnoid villi, sometimes called arachnoid granulations, close to the top of the brain, and eventually drains into the venous system from the superior sagittal sinus (Tortora and Derrickson 2006). The cerebrospinal fluid in a normally functioning person flows around the superior sagittal sinus and gets reabsorbed by the arachnoid villi due to pressure gradient differential. When cerebrospinal fluid is being blocked at any point during this flow cycle, it causes the ventricles in the brain to get stretched and become enlarged, affecting portions of subcortical brain tissue as well as white matter. There is approximately a pint of cerebrospinal fluid produced in the brain each day and the turnover rate of the cerebrospinal fluid is more than three times a day. However, because the production of the fluid is independent of the absorption, a lack of absorption results in an accumulation of cerebrospinal fluid in the ventricles (See Figure 1).

ETIOLOGY

There are several different types of hydrocephalus. Some are born with it, while others develop it much later in life. It can be genetic or acquired through physical or mental trauma. It can make one's head swell up like a balloon or shrink the brain in some cases. There are different causes for hydrocephalus and thus several different categories. Reasons could be either disrupted flow of the fluid, a problem with reabsorbing the fluid, or too much production. The most common is flow obstruction. There are four main categories of hydrocephalus: congenital, acquired, non-communicating, and communicating. NPH is a form of communicating hydrocephalus meaning there is no obstruction in the actual pathway and is therefore extremely difficult to detect. The etiology of NPH is unclear. While some cases can be attributed to known neurological injuries like subarachnoid hemorrhage and meningitis, most are idiopathic. Its symptoms most likely are due to ventricular dilation. The leading theory behind this dilation is the lack of reabsorption of cerebrospinal fluid into the venous system.

Others suggest that the cerebral vasculature may have a role in the pathogenesis of NPH (Siedlecki 2008). Although the intracranial pressure gradient increases between the fluid in the subarachnoid space and the ventricles, it remains normal. It is this increasing pressure that leads to cerebral ischemia and a stretching of the periventricular white matter. Scarring and fibrosis on the arachnoid granulations can disrupt the absorption of cerebrospinal fluid.

DIAGNOSIS

NPH is characterized by the chronic elevation of intracranial pressure due to increased cerebrospinal fluid becoming stable and therefore not exhibiting some of the usual hydrocephalic symptoms, such as headaches, vomiting, nausea, or altered consciousness. The big three symptoms, referred to as the Hakim triad after the doctor who first described them, include gait impairment, cognitive dysfunction, and urinary incontinence. NPH can be considered when the triad of symptoms is present. All three symptoms, however, are not required to suspect NPH. In the past, NPH would be diagnosed only if presenting the complete triad. Now, however, it can be diagnosed and treated in the presence of only two cardinal symptoms or even just one. This change in attitude resulted from the recognition that the longer NPH remains untreated, the worse the prognosis gets, with the complete triad always signifying an advanced stage of the disease (Kiefer and Unterberg 2012). Typically, gait impairment occurs first, with cognitive impairment and urinary incontinence occurring later. The difficulty with gait is usually described as a shuffling or magnetic walk and an exaggerated wide stance. It is also usually first to improve after shunt surgery. If left untreated, gait may deteriorate until the patient can no longer walk and is limited to a wheel chair. Dementia, when present, is subcortical. Cortical dementia, such as Alzheimer's has characteristics such as severe memory loss, an inability to recall words and understand common language. In contrast, subcortical dementia presents as inattention, latency in recall, and lack of spontaneity (slowness in response and reaction). These are known as abulic traits (Byrd 2006). The urinary disorder associated with NPH starts with urinary frequency or

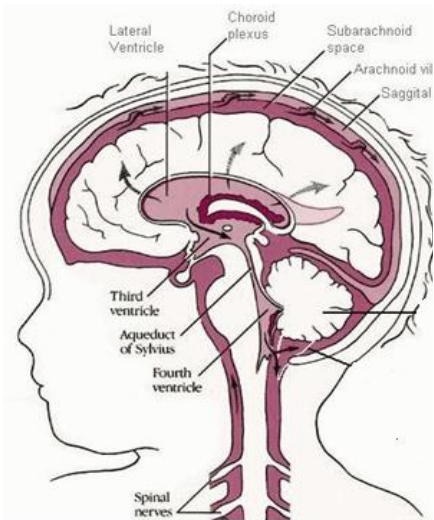


Figure 1: Anatomy of the ventricle system. Source: National Fibromyalgia Research Association

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urgency and then develops into complete loss of bladder control in the later stages of the disease. Incontinence often will not appear in early stages, and may go unreported as patients will attribute it to other prostatic issues and normal aging. It is also possible that what is chalked up to incontinence is merely an inability to reach the bathroom because of the slowed gait. These three clinical manifestations are likely caused by pressure on the structures adjacent to the ventricles. For example, pressure on the frontal lobes and their interconnections, including structures such as the limbic system, may cause symptoms of dementia. Pressure on the cortical center for bladder control may be the reason for incontinence, and pressure on the corticospinal tract, whose fibers lateral to the ventricles supply motor function to the legs, may cause the gait disturbance. The fibers of the corticospinal tract passing closest to the lateral ventricles in the corona radiata may be the reason why gait is first to improve.

The earlier on NPH is diagnosed, the more reversible it seems to be. Physical examination, assessment of cortical and subcortical function, and careful analysis of gait, along with a complete record of previous health, are the most important tools used in diagnosing NPH. Diagnoses aren't simple because these symptoms overlap with other disorders. Today, radiographic imaging studies are used as the main determining factor. In patients with clinical features of NPH, CT scans or MRIs should be used to measure ventricular size, rule out ventricular obstruction, and look for other possible causes like tumors, infections, and structural abnormalities. Diagnostic guidelines for monitoring symptoms have been published to pick out probable NPH patients from the possible (Siedlecki 2008). Screening of prospective surgery candidates is done to decide if shunt surgery would successfully reverse their symptoms. There have been conflicting reports as to the effectiveness of shunting procedures. However, cerebrospinal fluid monitoring and a trial of controlled, continuous cerebrospinal fluid drainage of 150-200mL per day has proven to make diagnoses more accurate (McGirt et al. 2005). Invasive diagnosis includes a spinal tap test: lumbar puncture with the removal of 30 to 70 mL of cerebrospinal fluid. This can be repeated for two or three days in a row. Modern forms of treatment lead to improvement in 70-90% of patients according to some studies (Kiefer and Unterberg 2012).

TREATMENT

The foremost method of treatment is the surgical installment of a ventriculoperitoneal shunt, a tube that connects the ventricles of the brain to an alternate drainage site, usually the abdominal cavity. A small hole is made in the skull and a catheter is placed in one of the lateral ventricles. It is then tunneled subcutaneously behind the ear and down to the abdominal cavity. The shunt contains a cap and valve ensuring there is no back-flow. In a way, some non-communicating hydrocephalic patients have an advantage because the only issue is the blockage of the subarachnoid space. In such cases, the shunt need only bypass that blockade and can empty into the subarachnoid space. Patients that have shunts implanted need life-long monitoring to ensure that it is doing its job and there is no recurring hydrocephalus. Shunt surgery is a very invasive procedure and is not without risk. Included in these risks are hemorrhage and stroke. There are some drugs that help delay the need for surgery: acetazolamide, furosemide, and digoxin, to name a few. Symptoms tend to improve upon removing the excess fluid at least for the short-term. It has been found to be the most effective available treatment for NPH. It is believed that shunting improves symptoms by removing the pressure from the parenchymal absorption pathway at the areas of the ventricles where cerebrospinal fluid is produced, reducing interstitial swelling and pressure, which then reduces ischemia.

iNPH

Hydrocephalus is special in that it is one of the few reversible causes of dementia. iNPH mainly affects people over 60 years of age. The big problem with iNPH is that it goes unnoticed and thereby untreated in many cases. Folks who think there is no cure for their dementia would be shocked to find out that it may actually be quite reversible. Diagnosis is made very difficult by the fact that there is overlap with many other diseases of the elderly and the surgery is quite invasive and can create complications. Often, after surgical implantation of a shunt, the symptoms will return, leading to questions whether NPH was the problem to begin with. It further complicates things when NPH is actually comorbid with some of the diseases that it is confused with, such as Alzheimer's and Parkinson's. It is estimated that more than 750,000 Americans have NPH, but less than 20% receive an appropriate diagnosis and treatment. This is largely due to its misdiagnosis in the first place and it is believed that an astounding 5 to 10% of people living with dementia, in fact have NPH (Siedlecki 2008).

PREVALENCE

The true prevalence of NPH is debatable. The available epidemiological data are inconsistent partly because there is no one set of diagnostic criteria. Moreover, 75% of those with NPH also have vascular dementia or Alzheimer's disease. A study done recently in Germany found one person in 80 to be demented. About 250,000 people receive a new diagnosis of dementia each year and NPH is thought to account for about 6% of all cases of dementia. A study of demented patients in nursing homes found that 9% to 14% of them had the symptoms typical of NPH. The incidence of NPH is approximated at 0.2 to 5.5 new cases per 100,000 people per year. Its prevalence is said to be 0.003% in patients under 65, and 0.2% to 2.9% for 65 and up. The prevalence of NPH rises significantly with age (Kiefer and Unterberg 2012). In one community-based study, data suggest that real clinical and imaging features that strongly suggest NPH are relatively uncommon and that most patients originally suspected of having NPH do not respond to a cerebrospinal fluid removal trial. The authors come to the conclusion that despite "relatively high prevalence figures (0.4%–14%)...studies that simply tabulate gait disturbance, dementia, incontinence, and ventriculomegaly without broader reference to the overall clinical context probably overestimate NPH prevalence" (Klassen and Ahlskog 2011). This is interesting because they seem to partially dismiss the triad of symptoms and neuroimaging showing enlarged ventricles as being indicative of NPH. Most other studies find that differential diagnosis based the clinical triad of symptoms and reading of imaging tests is more subtle. It is possible that their sample sizes are too small to be able to draw these conclusions from them.

DIFFERENTIAL DIAGNOSIS

The diagnosis of NPH and how its symptoms manifest themselves is controversial to say the least. Many other diseases have similar clinical characteristics to NPH, either by themselves or together with others. The most closely related are Alzheimer's disease, Parkinson's disease, diffuse Lewy body disease, vascular dementia, frontotemporal dementia, chronic alcoholism, carcinomatous meningitis, subdural hematoma, lumbar canal stenosis, and endocrine disorders such as hypothyroidism and Addison's disease. It is possible that many diseases can be alternate causes of symptoms of NPH as well as coexist with it. For example, spinal stenosis, a gait-associated condition, a cognition-associated condition such as subdural hematoma, and urinary diseases such as benign prostatic hypertrophy together at the same time in a patient and will appear to be NPH. The following is a contrast between the symptoms of NPH and other neurological disorders in numerous steps to be followed during diagnosis:

IMAGING AND CEREBROSPINAL FLUID STUDIES

Features on CT scans that suggest NPH are the presence of bigger temporal horns, overly enlarged ventricles (ventricles tend to enlarge with age anyway), and brain tissue shrinkage. Ventricles enlarge with age, but if the ratio of the maximum width of the anterior ventricular horns to the maximum width of the calvarium is greater than 0.3 (called the Evans ratio), the ventricles are considered abnormally enlarged (Factora and Luciano 2008). NPH can be characterized by findings on MRIs such as transependymal resorption, T2-weighted increased intensities in brain parenchyma next to the ventricles, and preservation of hippocampal tissue. Typical findings of NPH also include a coronal section at the level of the posterior commissure revealing a narrowed subarachnoid level surrounding the brain, called a tight convexity, and narrow medial cisterns. In addition to the lateral ventricles, the third ventricle is usually enlarged as well, while the fourth may or may not be enlarged. In addition, a higher level of cerebrospinal fluid flow through the aqueduct of Sylvius and a forceful cerebrospinal fluid flow through the foramina in the cerebrum. Spinal tap using a radionuclide isotope introduced into the subarachnoid space through a lumbar puncture has been one of the most used tests for diagnosing NPH. The test detects increased resistance to cerebrospinal fluid absorption via the arachnoid granulations, as seen in NPH. The standard spinal tap test collects between 20 and 50 mL of cerebrospinal fluid, measures the opening pressure, and conducts cerebrospinal fluid analysis. Normal cerebrospinal fluid protein and glucose levels, white blood cell count, and an opening pressure in the range of 60 to 240 mm H₂O, is highly suggestive of NPH. In all, there are six cerebrospinal fluid-related tests that can be used to figure out if NPH is present and also help determine if the patient is likely to respond to shunting. These include the standard spinal tap, large-volume spinal tap, temporary external lumbar drainage, extended intracranial monitoring, cerebrospinal fluid outflow assessment, and measurement of aqueductal cerebrospinal fluid flow (Osei-Boamah 2011). A particularly effective prognosticator of surgery outcome is the continuous spinal drainage of 150 to 200 mL of cerebrospinal fluid per day for 2 to 7 days. This test is considered to be positive if the number of steps taken in a 10-meter gait test, and the time needed to walk 10 meters, are reduced by 20% or more and/or there is an improvement of at least 10% in psychometric tests.

GAIT DYSFUNCTION

Diseases that share similar gait symptoms with NPH include: Parkinson-plus syndromes, Parkinson's disease, vascular Parkinsonism, visual impairment (cataracts, glaucoma, and macular degeneration), lumbar canal stenosis, large joint osteoarthritis, and peripheral neuropathy deconditioning (Factora and Luciano 2008). How can NPH's specific gait defects be told apart from the walking difficulties older people with these illnesses encounter? NPH patients initially complain of dizziness, trouble walking on stairs, and difficulty getting up from or sitting down on a chair. As the disease progresses, the patient's gait deteriorates significantly, becoming slow, broad-based, short-stepped, and glue-footed. Freezing during walking and difficulty with turning or starting to walk also suggest disease progression. Often, as a result of these gait abnormalities, frequent falls will call for the need of a cane or walker. In the later stages of NPH, motor deficit is usually worse because of the cognitive deficits. Sometimes, it is so severe that the patient can't walk at all. Of major importance for the differential diagnosis, are the following: Externally rotated posture of the feet, particular difficulty turning on the body's long axis, and absence of apraxia (Kiefer and Unterberg 2012). This is confusing because in the past, some actually referred to the symptom as gait apraxia because it looks like the person has forgotten how to walk and has trouble with basic components of walking (DiCecco 2009; Osei-Boamah 2011;

Siedlecki 2008). Apraxia is a disorder of motor planning and should not be confused with ataxia, a lack of coordination of movement, or abulia, the lack of desire to carry out an action. One doctor, however, writes in his findings that the short stride, slow gait, and difficulty with turning present challenging similarities between the gait of NPH and that of Parkinson's disease and cerebellar ataxia. Ways to help identify NPH include the absence of extrapyramidal signs such as cogwheel rigidity and no response to levodopa. In addition, NPH patients do not exhibit a resting tremor. Unlike patients with Parkinson's disease, NPH patients do not respond to visual and acoustic cues. Cerebellar ataxia is different in that it has other features such as dysarthria, gaze-evoked nystagmus, and appendicular dysmetria. NPH has none of those symptoms (Osei-Boamah 2011). NPH patients also have poor posture with a forward rounding of the shoulders and arms that hang loosely at their sides. They appear to always be watching their feet as they attempt to move. This limpness of the extremities is a telling sign of NPH, whereas Parkinson's is usually coupled with rigidity and stiffness of the limbs and trunk (Siedlecki 2008). In addition, the gait with Parkinson's disease tends to be more narrow-based, and the balance problems and disequilibrium are not as apparent (Ropper and Brown 2005). Many believe that a detailed gait history is extremely important for an accurate diagnosis. Specifically, the onset of the gait problems should be examined. Was the onset a progressive decline or more sudden? Typically, gait impairment in NPH is more gradual. Did the patient exhibit symptoms of tremor or bradykinesia (a general slowness of movement) suggesting Parkinson's? If the patient has back or cervical neck pain, it suggests the presence of lumbar canal stenosis or cervical myelopathy respectively (Factora and Luciano 2008). Parkinson-plus syndromes have features such as gaze palsy or autonomic dysfunction in addition to the shuffling gait that reduce the likeliness of diagnosis of NPH. In the elderly it is more likely to have multiple etiologies for these gait symptoms. For example, patients who have symptoms of NPH also can have lumbar canal stenosis. Therefore, one should always consider which of the disorders is more likely to be the cause for the impeded gait. In this case, it should also be considered that lumbar canal stenosis can interfere with a trial of cerebrospinal fluid removal used in predicting shunt outcome in NPH and limit potential for improvement after shunt placement (Factora and Luciano 2006).

COGNITIVE IMPAIRMENT

Disorders with similar cognitive manifestations as NPH include: Alzheimer's disease, diffuse Lewy body disease, vascular dementia frontal lobe dementias, depression, and prion disease. Cognitive impairment associated with NPH is often mild, and might initially be attributed to normal aging. The problems are mainly subcortical in nature. Short-term memory loss, short attention and concentration spans, apathy, and slow processing are some results of subcortical dementia. In other words, difficulty forming words, being unable to carry out simple tasks sequentially, or difficulty interpreting sensory stimuli are all cortical features and do not appear in NPH dementia. Any of these symptoms would distinguish NPH from cortical dementias, such as Alzheimer's disease (Byrd 2006). If other signs of cortical dementia, such as aphasia, agnosia, or apraxia are present, Alzheimer's disease should be considered. Dementia is also the dominant clinical feature of Alzheimer's disease. Alzheimer's and NPH can occur together, however, and the incidence of Alzheimer's disease in patients with suspected NPH is between 33% and 50% according to one study's estimate (Savolainen et al. 1999). Vascular dementia (stroke, multi-infarct dementia, vertebrobasilar insufficiency) and dementia with Lewy bodies are other causes of dementia to consider. Vascular dementia usually presents as a stepwise decline over an extended period, with greater loss of higher-order cognitive functions such as visuospatial perception and executive function. Lewy body dementia is especially hard to

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tell from NPH, as it has both gait and cognitive deficits. However, its hallmarks are hallucinations and cognitive fluctuations which are rare in NPH patients (Johnson and Graham 2010). When the comorbidities are associated with subcortical deficits like in vascular dementia, depression, and frontotemporal dementia, it is much more difficult to separate those characteristics that imply NPH. Table 1 shows the differences between cognitive deficits one comes across in NPH and the symptoms Alzheimer's disease and vascular dementia classically present (Factora and Luciano 2006). The timing of the cognitive impairment's development compared to that of gait is useful diagnosing NPH and determining if shunting will be successful. Normally dementia does not precede gait impairment in NPH, and when it does it signifies a weaker response to surgery. As previously discussed in regards to Alzheimer's disease, in cases where dementia is the predominant feature of all symptoms, consider evaluation for another neurodegenerative disease (Siedlecki 2008). History of cognition in a patient plays a role as well. A history of slowly progressing fading of memory and other areas of cognition bad enough to affect daily activities supports a diagnosis of Alzheimer's disease. These symptoms may also appear years after the onset of Parkinson's. A known history of strokes and a stepwise decline with each might mean vascular dementia. Mental status testing is used to confirm cognitive impairment is present and see how severe it is. The Mini-Cog and the Short Portable Mental Status Questionnaire are used to screen for the presence of cognitive impairment. The Folstein MiniMental State Examination can also be used to quantify how severe the impairment is. A clock-drawing test is sometimes used to assess visuospatial skills and executive function.

Table 1: Comparison of dementia characteristics.

	Alzheimer's disease	Vascular dementia	Normal pressure hydrocephalus
Memory impairment	X	X	Impaired retrieval
Executive dysfunction	X	X	X
Impaired visuospatial process	X	X ^a	
Impaired language	X	X ^a	Bradyphrenia
Impaired complex motor skills	X	X ^a	
Psychomotor slowing			X
Impaired attentiveness			X

a- Can occur based on location of infarction (tissue death)

These are not problems present in NPH patients according to most. Use of a screening test such as the Geriatric Depression Scale can weed out patients with depression that can affect cognitive function. Neuropsychological tests can be very useful in distinguishing between the

cortical and subcortical dementias (Factora and Luciano 2008; Siedlecki 2008). Early cerebrospinal fluid shunting can improve the cognitive deficits in as many as 80% of patients with NPH, but improvement is far less likely if the patient also has vascular dementia or Alzheimer's disease (Kiefer and Unterberg 2012).

URINARY INCONTINENCE

In the elderly population, incontinence is very prevalent. Causes include benign prostatic hypertrophy resulting in bladder outlet obstruction, retention coming from neurogenic bladder, either from long-standing diabetes or related to Parkinson's, pelvic relaxation that helps lead to stress incontinence, and cystitis. Identifying the specific type of incontinence (urge, stress, overflow, or functional) can help find the true cause. The use of cystoscopy and urodynamic testing can be used to eliminate other diagnoses (Factora and Luciano 2006). Incontinence in NPH comes from detrusor hyperactivity which results from absence of central inhibitory control but there is no evidence of defective bladder sphincter control. It starts as increased urinary frequency. It later develops into urge incontinence and then permanent urinary incontinence. Fecal incontinence is rare in NPH and points to another type of disorder. Urinary symptoms usually come later than the other two symptoms and are lacking entirely in some patients. In patients with NPH, the slow gait can compound the problem, making it difficult to reach the bathroom in time and can make it difficult in determining the real cause (Byrd 2006). Diuretics can cause urinary frequency, alpha-agonists obstruction, and some medications with anticholinergic side effects may produce urinary retention. Evaluation of urinary incontinence can target certain symptoms. A urinalysis can easily be done to exclude the presence of a urinary tract infection. Getting a postvoid residual can identify those with urinary retention who are experiencing stress symptoms or any sensation of incomplete void. Urodynamic testing to evaluate urge incontinence has not been helpful in distinguishing NPH from other causes of detrusor instability. Cerebrospinal fluid shunting can improve bladder dysfunction in as many as 80% of NPH patients if surgery is done early on, but if performed in an advanced stage less than 50% to 60% respond positively (Kiefer and Unterberg 2012).

PROGNOSIS

The prognosis for NPH depends on a variety of factors, including age of onset and timing of surgery. Overall, most reports say people with suspected NPH have a 50% chance of benefiting (for the long term) from the installation of a shunt (Longe 2006). That figure includes those with known causes. iNPH in particular, as previously discussed, has several indicators of good shunting outcome that include early diagnosis, gait as the predominant symptom, and a positive response to large-volume cerebrospinal fluid draining. The presence of B waves during the majority of continuous intracranial monitoring, cerebrospinal fluid outflow resistance of more than 18 mm Hg/mL/min., and increased aqueductal cerebrospinal fluid flow are also indicative of potential success with shunting (Osei-Boamah 2011). A study by Malm et al. (1995) found that the number of patients with iNPH who improved from surgical implantation of the ventriculoperitoneal shunt went from 64% at 3 months to 26% at 3 years. This showed that a shunt may be effective only temporarily, lasting from 1 to 3 years. However, shunting can still make an enormous difference in quality of life for many of these patients. In the short term, the success rate is usually reported between 50% and 90% with all patients suffering a decrease over the following years. The high variability in reports is likely due to differences in selection, method of shunting, care in following up, and how success is defined. Much failure over time is caused by the development or progression of other neurologic or systemic disease (Factora and Luciano 2008). Some reports of symptom improvement after ventriculoperitoneal shunt surgery

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are as low as 30% in patients with iNPH and 50% to 70% for patients with sNPH (Johnson and Graham 2010). Studies have shown that up to 93% of patients have major improvements in gait shortly after surgical treatment of NPH, as opposed to only 50% improving in cognitive symptoms (Yusim et al. 2008). Indeed, many seem to think the gait symptom is the primary indicator of NPH as well as ventriculomegaly (See Figure 2). In one study, among 41 patients with suspected NPH, all had gait dysfunction, 30 had cognitive impairment, and 14 had urinary incontinence. Twenty out of forty one presented with two of the symptoms, while twelve had all three parts of the triad. Nineteen were found to have at least moderate postural instability (defined by a positive pull test). All 40 patients with available results had documented ventriculomegaly. Only 13 of these underwent shunt placement, as it was offered only to those with gait improvement after a trial of cerebrospinal fluid removal, and only 4 of these had all elements of the classic NPH triad (Klassen and Ahlskog 2011). However, despite these positive symptoms, more than half did not respond well to the shunt in the long run. Of note is the fact that no patient with postural instability showed definite improvement after shunting, and 4 out of 5 (not including one who died during surgery) were later given an additional diagnosis. It may be that this symptom is a trademark of underlying neurodegenerative disease. Shunting may not be the best course of action for these patients (Klassen and Ahlskog 2011). Many feel that the best predictor of ventriculoperitoneal shunt outcome is external lumbar drainage; it is said to be about 85% accurate in identifying patients that would benefit from surgery, and is becoming more and more popular with neurosurgeons (Byrd 2006). Any surgery in the elderly population is going to have risks. Studies have risks as high as 30% to 40% in shunting, and severe morbidity or death at 6% to 8% (Factora and Luciano 2006).

CONCLUSIONS

NPH is a lesser-known medical condition for which a relatively small amount of research is conducted to improve treatment for. It was once thought that its prevalence hovered around 0.5% (Brean and Eide 2008). Recent studies show that it is closer to being between 1 and 2%. The reason for the discrepancy seems to be an oversimplification of the terminology of the symptoms (Conn 2011), and there do seem to be more subtle tells when it comes to diagnosing NPH. Identifying symptoms in the triad is simple, but determining which patients truly have NPH and will benefit from shunting is not straightforward. Shunting outcomes are not quite related to which patients had NPH to begin with due to comorbidity with other diseases. Although patients need not have all three symptoms for NPH to be considered, the gait deficit seems the most crucial for diagnosis. The mere presence of enlarged ventricles does not seem to be a valid indicator as they tend to enlarge with age. There are multiple conflicting theories and therefore many disagreements as to the true symptoms of NPH. Thus, the differential diagnosis of NPH can be quite difficult. The criteria for a diagnosis of NPH seem clear to everyone only with the following findings that make the diagnosis of NPH less likely: intracranial pressure above 25 cm H₂O (this rules out iNPH by definition), age under 40, cortical deficits, progressive dementia without gait disturbance, and lack of progression of symptoms. The progression of symptoms is a controversial point, as

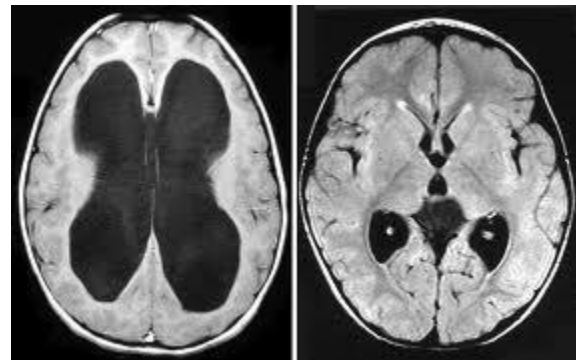


Figure 2: CT scan of ventriculomegaly vs. normal in adult

authors differ on what is considered a normal time of progression. A patient is considered a potential ventriculoperitoneal shunt candidate if he or she has symptoms consistent with NPH (not explained by other diagnoses) and has enlarged ventricles on imaging. The doctor should try to rule out as many other possible disorders as possible, and consider surgery by carefully weighing the risks and potential benefits as well as factoring in the chance for success.

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