# Title of Case

Normal Pressure Hydrocephalus: under recognized and under diagnosed

# Authors of Case

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# Summary

Up to 150 words summarising the case presentation and outcome

Patient is a 71 year old white female who presented to the office with a several month history of gait instability and urinary difficulties. Normal Pressure Hydrocephalus (NPH) was suspected and patient was sent to an interventional radiologist for a spinal tap. Symptoms improved after spinal tap. Patient had MRI of brain and later a cysternography both of which indicated abnormal findings most consistent with NPH.

Patient was referred to a neurosurgeon for placement of a ventriculoperitoneal shunt.

# Background

Why you think this case is important – why you decided to write it up

Normal pressure hydrocephalus (NPH) is an important differential when assessing an older patient who has cognitive impairment, gait disturbance, and urinary incontinence. NPH is unique, in that with the appropriate diagnosis and treatment the dementia is often reversible, unfortunately, this syndrome is under recognized and under diagnosed. With more awareness of NPH, less people will be wrongfully diagnosed with other diseases that cause dementia like Alzheimer’s Disease and Parkinson’s Disease.

# Case Presentation

Presenting features, medical/social/family history

**HPI:**
- A.H. is a 71 y/o female who presented to the office with a several month history of gait instability and urinary difficulties.
- Over several months patient went from walking independently to using a walker to finally ending up in a wheel chair.
- A.H. has been falling more frequently over the past several months.
- Had lumbar puncture, MRI of brain, and cysternography, results of which were consistent with NPH.

**PMH/PSH:**
- Thrombophlebitis in rt leg (2007)
- Lower extremity edema
- Cataract rt eye
- Hypercholesterolemia
- Depression/ Anxiety
- Urinary difficulties (2008)
- Lumbar decompressive laminectomy and fusion L4/5
- Lt total knee arthroplasty (2006)
- Lumbar puncture (7-23-08)
- Nuclear medicine cysternography (9-22-08)

**Meds:**
- Altace 5mg qd  
- Lexapro 10mg qd  
- Fosamax 70mg  
- Vesicare 5mg qd  
- Coumadin 5mg (Mon-Sat, none Sun)  
- Xanax 0.5 mg prn  
- Lipitor 10mg qd  
- Ambien 10 mg qhs
Allergies:
- Penicillin and Latex

Family Hx:
- Mother ↓ 70’s, leukemia
- Father ↓ 80’s, AAA
- Sister 3↑, Type 2 DM
- Brother 4↑, Parkinson’s, DM1, CVD

Social Hx:
- Married, retired housewife
- G1P1
- Denies ETOH, tobacco, or illicit drug use.

ROS:
- Gen: + Weakness, fatigue
  - fever/chills/night sweats
- Cardiac: - chest pain, palpitations
- Resp: - dyspnea, cough
- GU: + frequency, nocturia, incontinence,
  - dysuria, hematuria, polydipsia
- Peripheral vascular: varicose veins, history of thrombophelbitits
- Neuro: + depression, weakness
- Otherwise, unremarkable.

PE:
- Vitals: P: 86 R: 14 BP: 110/60 Wt: 178
- Gen: Non-toxic, NAD, A&O X3. Patient is distressed about her gait instability
- HEENT: Neck supple, trachea midline, PERRLA, EOMI
- Heart: RRR, no MRG
- Lungs: CTA B/L
- Abd: Soft, distended. No guarding or rebound
- Peripheral Vascular: edema in lower extremities bilaterally
- Neuro: Motor strength 5/5.
- Gait: slow, cautious, wide-based

**INVESTIGATIONS** *If relevant*

Laboratory studies:
- Cytopathology report: (from cisternogram/LP). No acute inflammatory cells or malignant cells were observed.

Radiologic studies:
- MRI of brain with & without contrast: abnormal exam findings most consistent with NPH.
- LP: 17.5mL of clear fluid removed. Gait improvement, NPH likely.
- Nuclear Medicine Cysternography: early acumination of radiotracer within cerebral ventricles which persists at 24hrs and out to 48hrs. Findings compatible with NPH.

**DIFFERENTIAL DIAGNOSIS** *If relevant*
- Alzheimer’s Disease
- Parkinson’s Disease
- Demylenating Disease
- Chronic small vessel ischemia

**TREATMENT** *If relevant*

After the diagnosis of Normal Pressure Hydrocephalus was confirmed:
- The patient was referred to a neurosurgeon for the placement of a ventriculoperitoneal (VP) shunt.
OUTCOME AND FOLLOW-UP
Patient recovered well from VP shunt placement. Surgery was uncomplicated. A.H. was discharged home 3 days after surgery. Patient stated that she noticed an improvement in her gait instability. Both patient and husband were educated on possible complications from the VP shunt. Follow up appointment was scheduled.

DISCUSSION including very brief review of similar published cases (how many similar cases have been published?)

1. What are the current guidelines for the diagnosis and management of Idiopathic Normal Pressure Hydrocephalus?
2. How can one distinguish NPH from other diseases that can cause gait abnormalities and cognitive impairment?
3. What are positive and negative predictive factors for VP shunt placement and associated risks/complications of this surgery?
4. What are the differences between idiopathic normal pressure hydrocephalus (INPH) and secondary normal pressure hydrocephalus?

1. As of 2002, there was limited research that gave any in-depth discussion on guidelines for the diagnosis and management of NPH. Then, in 2002, an international NPH study group assembled to develop guidelines for the diagnosis of NPH. In 2005 the guidelines were published and covered four major topics: 1) the clinical diagnosis of INPH, 2) the value of supplementary diagnostic tests, 3) surgical management, and 4) outcome of CSF shunting in INPH².

Taking the above guidelines into account, patients can be divided into three different groups: A) Probable NPH, B) Possible NPH, and C) Improbable NPH. Walchenbach and colleagues go on to define the three subcategories in their study. Probable NPH consists of predominance of mental impairment, gait disturbance, rounded frontal horns on MRI, and urinary urgency or incontinence. For Possible NPH Walchenbach et al. describe findings that are not related to either Probable or Improbable. Clinical findings with Improbable NPH include marked dementia, gait disturbance, and the presence of another disease that could rule out NPH⁸. The ventricles on an MRI of a patient with Improbable NPH would look only slightly dilated.

2. NPH may present with symptoms that mimic other diseases like Alzheimer’s disease and Parkinson’s disease. It is important to perform a thorough history and physical exam on these patients, and if necessary refer them for a neurologic consult so that they do not get misdiagnosed. Neumiller et al. estimates that 375,000 Americans with NPH have been wrongfully diagnosed.

Gait abnormality can occur in both NPH and PD, but both have slightly different appearance. With PD, patients typically have narrow stance with a shuffling gait, whereas NPH patients have a wider stance with slow, wide-based gait⁴,⁷. Often times NPH patients will look as if their feet are stuck or glued to the floor⁵. NPH patients also lack the characteristic resting tremor and cogwheel rigidity seen with PD.

Neumiller et al. also looked at the cognitive impairment of NPH and compared it to that of AD and PD. The impairment that is seen with NPH typically includes a decrease in planning skills, verbalization and initiative. There are more behavioural issues seen in AD and PD including hallucinations, agitation and aggressiveness⁴. This same study also found that AD patients can also present with agnosia and aphasia, two characteristics that are not typically seen in NPH. With radiographic observation one can distinguish NPH from AD by noting the peri-hippocampal fissures (PHFs). The PHFs in a patient with NPH with normal or minimally dilated compared with marked dilation seen with AD⁶.
3. Published research indicates that there are certain predictive factors in whether VP shunt placement has a positive predicted outcome or a negative predicted outcome. In one study of 18 patients diagnosed with INPH, the onset of dementia predicted an unfavourable outcome with regards to shunt placement3. A more positive predicted outcome is seen with patients presenting with gait abnormalities, with or without urinary incontinence. The latter patient, with only gait abnormalities, usually presents earlier in the course of INPH, therefore improving the likelihood that shunt placement will be beneficial. Another study performed in 2005 concluded that there was still no reliable way to select patients whose NPH would improve5. In this study, Poca and colleagues performed shunt placement on patients who presented with traditionally poor prognosis factors and saw improvement, especially in gait and sphincter control.

There are several risks to VP shunt surgery. One study lists several risks associated with VP shunt placement and the percentage of time that they occur including: shunt malfunction (20%), subdural hematoma (2-17%), seizures (3-11%), infection (3-6%), and intracerebral hematoma (3%)4. The above complications can be divided into three groups including those related to the operation, those related to the shunt system, and those related to the flow of the shunt system2.

A risk benefit ratio should be taken into account before placement of a shunt. Gallia and colleagues used a risk benefit ratio that included life expectancy, comorbidities, and functional status. In their study Gallia et al. found that 75% of their patients had improvement in at least one INPH symptom following shunt surgery. At 18 months 46% had improvement in all INPH symptoms. Keeping in mind the risks and comorbidities of shunt placement, the benefits outweigh the risks in most individuals.

4. INPH does not have any known origin that leads to this disease. The major risk factor for INPH seems to be age because it is usually diagnosed in the sixth or seventh decade of life3. Another study listed diabetes and hypertension as other possible risk factors for INPH1. Marmarou and colleagues attribute several other mechanisms to INPH including white matter ischemic changes, reduction in metabolism and blood flow, and an increase in trans mantle pressure.

There was limited research on secondary NPH. In one study, Marmarou et al. defines secondary NPH being caused by events including head trauma, stroke, meningitis, and even subarachnoid hemorrhage. With any of the above symptoms the underlying cause needs to be diagnosed and treated. With secondary NPH there is less confusion on why the patient is experiencing certain symptoms and it is less likely to be confused with AD and PD.

LEARNING POINTS/TAKE HOME MESSAGES 3 to 5 bullet points

- Abnormal gait, urinary difficulty, and symptoms of dementia make up the triad that defines NPH, but it is common for a patient to present with only one or two of the above symptoms. Therefore, NPH should be in the differential when a patient presents with any one of aforementioned symptoms.

- NPH is a reversible disease that health care professionals need to become more familiar with so that it is not mistaken for non-reversible neurologic diseases like Alzheimer’s and Parkinson’s.

- Prompt referral to a neurologist or other multidisciplinary team, if NPH is considered, may help expedite treatment for this disease and a more favourable outcome is to be expected.

REFERENCES


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