If It’s Not NPH, What Is It?

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The NPH Syndrome

- Dementia or cognitive impairment
- Gait and balance difficulty
- Urinary incontinence

- These are the 3 most common symptoms of the elderly population

INPH is a comorbidity among comorbidities

- NPH clinically overlaps with many conditions of the elderly
  - Vascular dementia
  - Degenerative dementias or disease
  - Cervical stenosis/myelopathy
  - Lumbar stenosis
  - Peripheral neuropathy
- These conditions can co-exist with NPH and they can all mimic NPH symptoms

This is a common scenario

- Elderly patient with possible INPH, and
  - Coronary artery disease
  - Knee or hip prosthesis (or both)
  - Lumbar or cervical stenosis
  - Diabetes
  - Peripheral neuropathy (↓ pinprick in feet)
  - Periventricular white matter hyperintensities
  - Evans Index 0.33
  - Possible atrophy (?AD or FTD or Lewy body)
  - Prostate cancer (♂) or pelvic floor laxity (♀)
  - Hearing impairment
  - Mild cataracts
Disorders that may have all 3 symptoms
Disorders that may have 2 symptoms
Disorders that may have only 1 symptom
Disorders that aggravate symptoms

To answer the question...
- If it’s not NPH, it could be many things
- The job of the neurologist, neurosurgeon, or other evaluating physician is to answer this question
  - As part of the initial evaluation
  - As part of continuing care if the patient’s symptoms don’t resolve as expected
- Let’s break it down by symptom

What does “dementia” mean?
- When most people hear the term dementia, they automatically think of Alzheimer disease
- And when they think of Alzheimer disease, they think of “incurable”, “memory loss”, “loss of personhood”, “nursing home”

In reality
- Alzheimer dementia is not the only dementia
- Dementia has multiple potential causes, of which some are reversible and some are not
- Reversible:
  - Hypothyroidism, B12 deficiency, folate deficiency, depression, low blood pressure, normal pressure hydrocephalus, medication side effects
- Not reversible:
  - Alzheimer*, Fronto-temporal dementia, Lewy body dementia, vascular dementia, cortico-basal degeneration, progressive supranuclear palsy, etc
What is dementia?

- An insidiously progressive syndrome of significantly impaired intellectual functioning that interferes with normal activities and relationships
- Disturbance of multiple higher cortical functions, including memory, thinking, orientation, calculation, comprehension, learning capacity, language, and judgement

2011 NIH Criteria

- Dementia symptoms:
  - Interfere with the ability to function at work or at usual activities
  - Represent a decline from previous levels of functioning and performing
  - Are not explained by delirium or major psychiatric disorder
  - The cognitive or behavioral impairment involves a minimum of two “domains”

Dementia Symptom Domains

- Memory: ability to acquire and remember new information
- Cognitive Skills: Impaired reasoning and handling of complex tasks, poor judgment
- Perception: Impaired visuospatial abilities (e.g., recognizing faces, objects or getting lost in one’s own home)
- Language: Speaking, reading, writing
- Personality, behavior, or comportment

Examples

- Changes in social or occupational functioning, or in usual daily activities
  - Making mistakes with medications
  - Repeating questions or statements
  - Impaired driving skills (too slow, too fast, too erratic, getting lost)
  - Difficulty managing finances
  - Change in eating habits
  - Too friendly with strangers, making inappropriate statements, striking out
Dementia is not always recognized

- Family or primary physician may not recognize or diagnose dementia because:
  - Onset is insidious. An event, such as getting lost while driving, often triggers the recognition
  - Assumption that memory loss is a normal part of aging (it isn’t)
  - Assumption that all dementia is Alzheimer’s
  - Assumption that nothing can be done
  - Failure to refer to appropriate specialists
  - Failure of specialists to use comprehensive diagnostic approaches

Mild Cognitive Impairment

- Per the 2011 NIH Criteria
  - Concern about a change in cognition, compared to the person’s previous level
  - Evidence of lower performance in one or more cognitive domains that is greater than would be expected for the patient’s age and educational background
  - Preservation of independence in functional abilities
  - Not demented

Senior Moments

- Benign forgetfulness of aging is not dementia or MCI
  - Forgetting why you walked into a room
  - Tip-of-the-tongue phenomenon
  - Forgetting the name of someone you haven’t seen or thought of in a long time
  - Being slower than your children or grandchildren at learning new tasks...as long as you’re still capable of learning

Gait and Balance

- Gait is the term used to describe the pattern of a person’s walking
- Balance entails the postural reflexes necessary to keep the body upright and properly positioned for locomotion, and to protect against falls.
- Locomotion is the rhythmic, periodic movements of the limbs and trunk that move a person in a desired direction

We are bipeds

- Humans normally walk on two legs
- Nearly constant need to sense and correct axial posture and leg and foot position.
- This sensing and correcting involve neuronal circuits and pathways from the entire central and peripheral nervous system
  - Sensory input: visual, vestibular (inner ear), proprioception (position sense)
  - Motor coordination: Premotor planning, upper and lower motor neurons, cerebellar pathways, basal ganglia, and spinal cord

Gait Impairment

A Significant Health Problem

- 35% of adults >70 years have abnormal gait
- Abnormal gait has 2.2 times greater risk of death and institutionalization (J Am Geriatr Soc 54:255-61, 2006)
  - Mild 1.8x, Moderate to severe 3.2x
- Gait impairment increases the risk of falling 2.7x (JAMA 2010;303:258-66)
Risk of Institutionalization or Death Rises with Severity of Gait Impairment

Table 4. Risk of Death, Institutionalization, or Both over the 5-Year Study Period by Baseline Gait Status, Adjusted for Age and Sex

<table>
<thead>
<tr>
<th>Gait Status</th>
<th>N</th>
<th>Hazard Ratio</th>
<th>95% CI</th>
<th>P Value</th>
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</thead>
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<tr>
<td>Overall</td>
<td>469</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Normal</td>
<td>279</td>
<td>1.00 (Reference)</td>
<td></td>
<td></td>
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<tr>
<td>Neurologic</td>
<td>190</td>
<td>2.51 (1.53–4.08)</td>
<td>0.001</td>
<td></td>
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<tr>
<td>Nonneurologic</td>
<td>190</td>
<td>1.00 (Reference)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neuroclinical</td>
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<td>2.69 (1.64–4.40)</td>
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<tr>
<td>Nonclinical</td>
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<td>1.00 (Reference)</td>
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<tr>
<td>Neurological plus</td>
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<td>2.74 (1.62–4.65)</td>
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<td>Nonneurological plus</td>
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<td>1.00 (Reference)</td>
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<tr>
<td>Multisystemic</td>
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<td>Multisystemic plus</td>
<td>77</td>
<td>1.00 (Reference)</td>
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</table>


Michael A. Williams, MD

Neurologic vs Nonneurologic Gait Impairment

- **Neurologic**
  - Unsteady, ataxic, frontal, parkinsonian, neuropathic, hemiparetic, and spastic

- **Nonneurologic**
  - Arthritis, cardiac disease, chronic lung disease, and peripheral vascular disease
  - Low vision is a common contributor
  - Patients may have both
    - i.e., multifactorial gait impairment

Neurologic Gait Impairment

- Higher-level gait disorders involve difficulty with:
  - Integrating sensory information about the position of the body in its environment, including the effect of gravity, and
  - Properly selecting and executing motor plans for gait or postural reflexes

- Postural and locomotor reflexes are absent or inappropriate
  - Primary deficits of motor or sensory function are also usually absent.
  - Patients complain of weakness but they aren’t weak—they have poor motor control
  - The involved areas of the brain are basal ganglia and the frontal cortex and its connections to the basal ganglia and brainstem (e.g., white matter)

What does it look like?

- Difficulty initiating gait
- Feet stuck to the floor
- Altered posture—sometimes stooped, sometimes rigid, pushing backwards
- Slow, shuffling steps
- Shortened steps
- Difficulty with turns, or in tight spaces
- Difficulty with uneven surfaces

Gait and balance impairment is not always recognized

- Family or primary physician may not recognize or diagnose gait impairment because:
  - Onset is insidious. An event, such as falling, often triggers the recognition
  - Assumption that impaired gait is a normal part of aging (it isn’t)
  - Assumption that nothing can be done
  - Failure to refer to appropriate specialists
  - Failure of specialists to use comprehensive diagnostic approaches
Urinary Incontinence

- Urinary incontinence (UI) is loss of bladder control.
- Symptoms can range from mild leaking to uncontrollable wetting. It can happen to anyone, but it becomes more common with age. Women experience UI twice as often as men. (National Library of Medicine)

Types of Incontinence

- Stress Incontinence: If the muscles that keep the bladder closed are weak, patients may leak urine with sneezing, laughing or lifting heavy objects. (More common in women)
- Urge Incontinence or Overactive Bladder: If bladder muscles are too active, patients may feel a strong urge to go to the bathroom with difficulty inhibiting bladder emptying
- Other causes of incontinence exist, such as prostate problems and nerve damage

Urinary incontinence is not always recognized

- Family or primary physician may not recognize or diagnose incontinence because:
  - Onset is insidious. An event, such as an embarrassing accident, often triggers the recognition
  - Assumption that incontinence is a normal part of aging (It isn’t)
  - Assumption that nothing can be done except to wear a pad or adult diapers
  - Failure to refer to appropriate specialists
  - Failure of specialists to use comprehensive diagnostic approaches

Dementia

- Alzheimer’s disease
  - A type of neuro-degenerative disease
  - Disease in which certain groups of brain cells (neurons) deteriorate and die over a long period of time, gradually producing cerebral atrophy
  - Symptoms depend on the neurons affected

The brain scan may help

- Enlargement of the ventricles can occur in the absence of hydrocephalus
- In neuro-degenerative disorders, the lost brain volume (atrophy) is “replaced” by CSF, as nothing else can take up the space
- Patients with these disorders do not improve with shunt surgery
- And may experience complications

Other Types of Dementias

- Neuro-degenerative
  - Lewy Body Dementia
  - Fronto-temporal Dementia
  - Progressive Supranuclear Palsy (PSP)
  - Multi-System Atrophy (MSA)
  - Parkinsonism
- Other types
  - Vascular Dementia
  - Neuro-syphilis
Normal vs. Hydrocephalus

Vascular Dementia

Gait Impairment

• Neuro-degenerative Disorders
  • Lewy Body Dementia
  • Fronto-temporal Dementia
  • Progressive Supranuclear Palsy (PSP)
  • Multi-System Atrophy (MSA)
  • Parkinsonism
  • Spino-cerebellar degeneration

Cerebral Atrophy

Other Contributors

• Medication Side Effects!!!!!
• Sleep Apnea / Sleep Disorders
• Hypothyroidism
• Vitamin B₁₂ Deficiency
• Hearing Impairment
• Visual Impairment

Gait Impairment

• Other causes
  • Cervical Stenosis (chronic spinal cord compression)
  • Lumbar stenosis
  • Chronic lower back pain
  • Degenerative arthritis of the hips, knees, ankles
  • Peripheral neuropathy
  • Peripheral vascular disease
  • Inner ear (vestibular) disorders
Incontinence

- Other Causes
  - Women – Pelvic-floor abnormalities
  - Men – Prostatic hypertrophy/obstructive uropathy
  - Interstitial cystitis
  - Cervical stenosis
  - Lumbar stenosis
  - Peripheral neuropathy

Symptom patterns can be helpful

- A patient with dementia and no gait impairment is unlikely to have NPH
- A patient with dementia that developed early and gait impairment that developed late is unlikely to have NPH
- A patient with gait impairment and incontinence but no dementia should be evaluated for a spine disorder before tests for NPH are done

If someone you know has

- Cognitive impairment or dementia
- Gait and balance impairment
- Loss of urinary control with urgency

- We want to think of NPH
- But we need to consider many other causes so that we offer the appropriate treatments

Thank you!