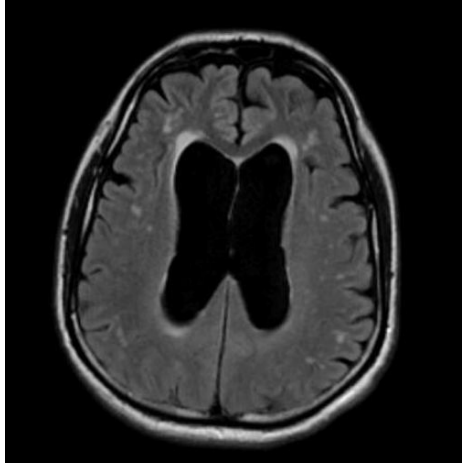
Ochsner
Neuroscience Institute



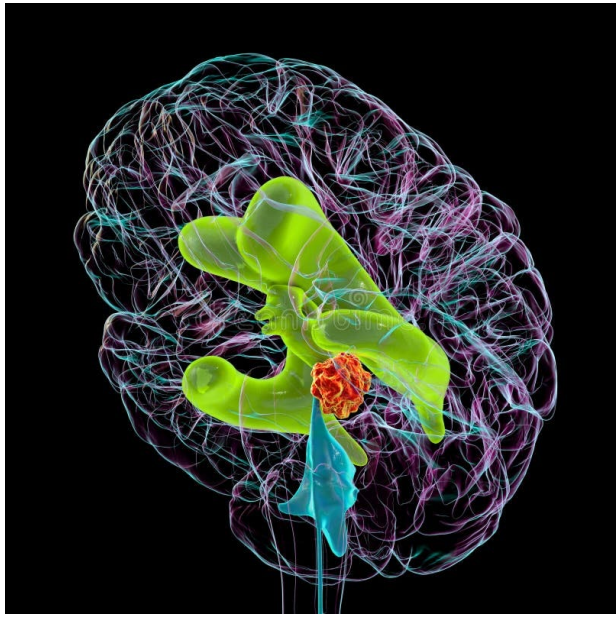
Understanding Normal Pressure Hydrocephalus (NPH)

Beyond "Wet, Wacky, Wobbly"

Erin Biro, MD, FAANS

Department of Neurosurgery

Ochsner Health



Neuroscience Symposium 2024

Disclosures: None

HIGH PERFORMING HOSPITALS
U.S.News & WORLD REPORT
2022-23
NEUROLOGY & NEUROSURGERY

Objectives:

- Define NPH
- Differential Diagnosis and Mimickers
- Discuss the pathophysiology, symptoms, and diagnosis
- Outline treatment options
- Outcomes
- Referral Guidelines

Source:

What is Normal Pressure Hydrocephalus ?

- Hydrocephalus: neurological disorder(s) that is characterized by increased accumulation of CSF with the ventricle system of the brain (cranial compartment). This accumulation leads to increased intracranial pressure (ICP) that can lead to a variety of neurological conditions including brain damage and even death if left untreated.

- Normal Pressure Hydrocephalus:

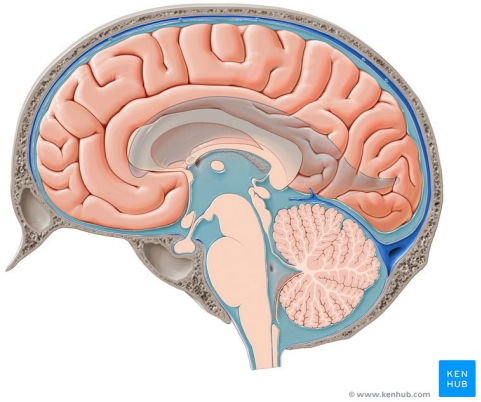
  - Specific “subtype” of hydrocephalus in which the ventricles are enlarged BUT the pressure is NOT elevated ICP

  - Impacts older adults (often underdiagnosed)

  - Triad of gait instability, cognitive impairment, and urinary incontinence

Source:

Pathophysiology:



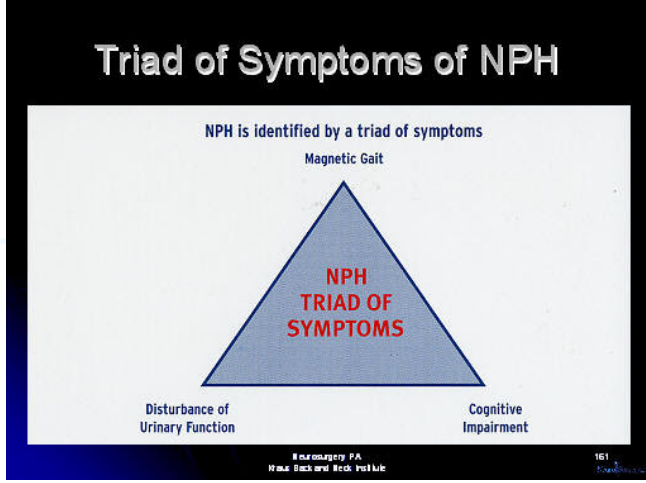
- Mechanism (?): Impaired CSF absorption paired with age-related cerebral atrophy resulting increased size of ventricle (+/-) trans-ependymal periventricular “edema” WITHOUT increase intracranial pressure
  - Ventricular Compliance or Hydraulic Conductance

- Etiology: Idiopathic or less commonly associated with distance history of trauma, subarachnoid hemorrhage, or meningitis. Currently no known gene or genetic link but there ongoing research

- NPH is complex interplay between CSF absorption issues, ventricular enlargement, white matter tract(s) compression, and disruption of neural networks -

Source:

Clinical Triad: “wet, wacky, wobbly”



- **Gait Disturbance: progressive gait apraxia
  - Hypokenetic and Broad-Based gait problem
  - Trouble taking first step, issues with turns, taking short steps, shuffling gait, difficult lifting feet
  - First symptoms to appear and first (and most likely) symptom to improve

- Cognitive Impairment: insidious but progressive cognitive decline
  - Short term memory issues
  - Slowed processing (math, multi-step tasks, etc…)
  - Resembles dementia

- Urinary Incontinence: typically urgency, frequency, progressing to incontinence
  - Not retention, not initiation difficulty
  - Usually last of 3 symptoms to appear and last to improve with treatment

Source:

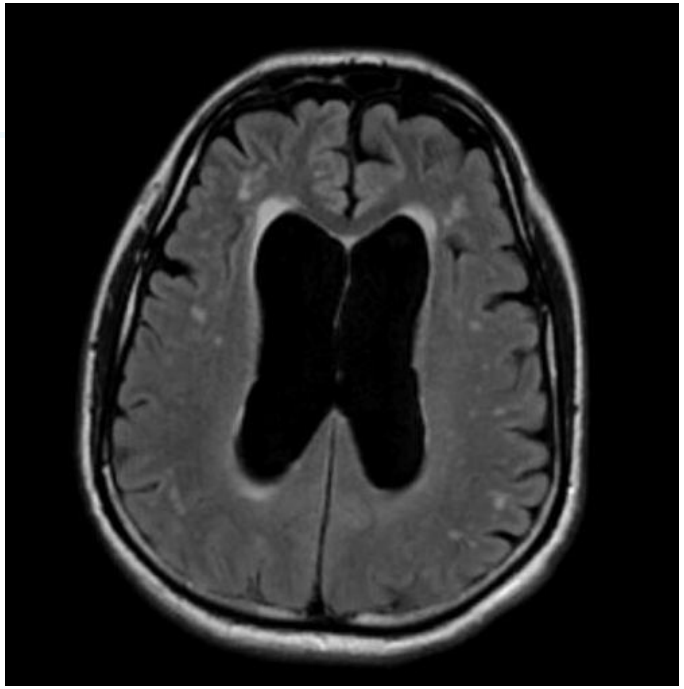
Ochsner Health Neuroscience Institute

Differential Diagnosis and Mimickers

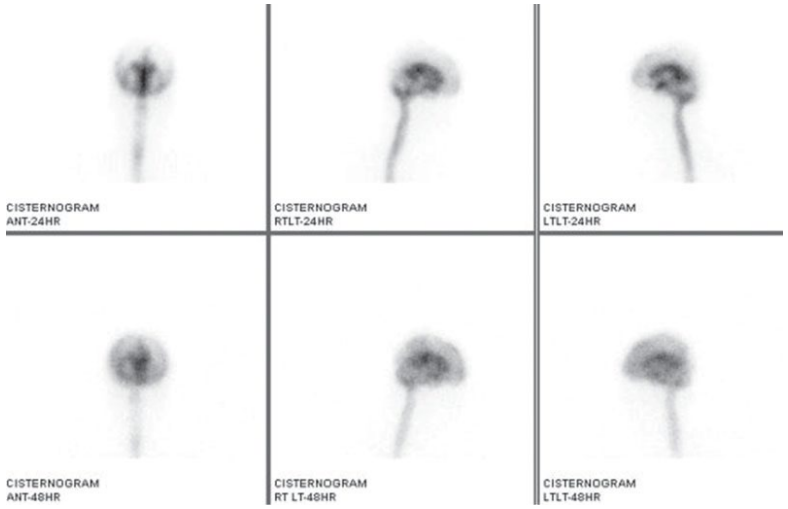
- * Data points to NPH being a significantly underdiagnosed neurologic pathology
- Alzheimer's Disease (memory more prominent than gait)
- Parkinson's Disease (tremors and response to dopaminergic meds)
- Vascular Dementia (h/o stroke, MRI show multiple infarcts or extensive white matter changes)
- Chronic Subdural Hematoma
- Binswanger's Disease (Subcortical Arteriosclerotic Encephalopathy) (h/o HTN, extensive small vessel disease on imaging)
- Cervical Myelopathy (hyperreflexia, increased tone)
- Frontotemporal Dementia (early changes in personality and language, frontal/temporal atrophy on imaging)

Source:

Diagnostic Work-Up

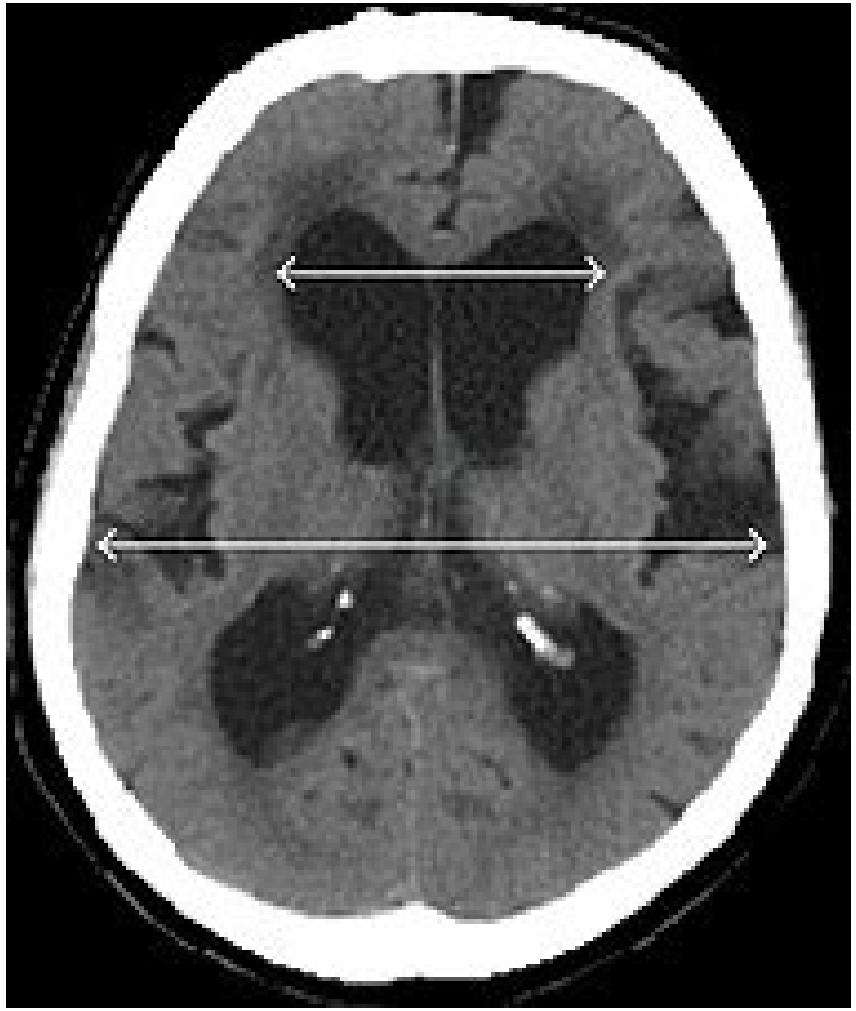


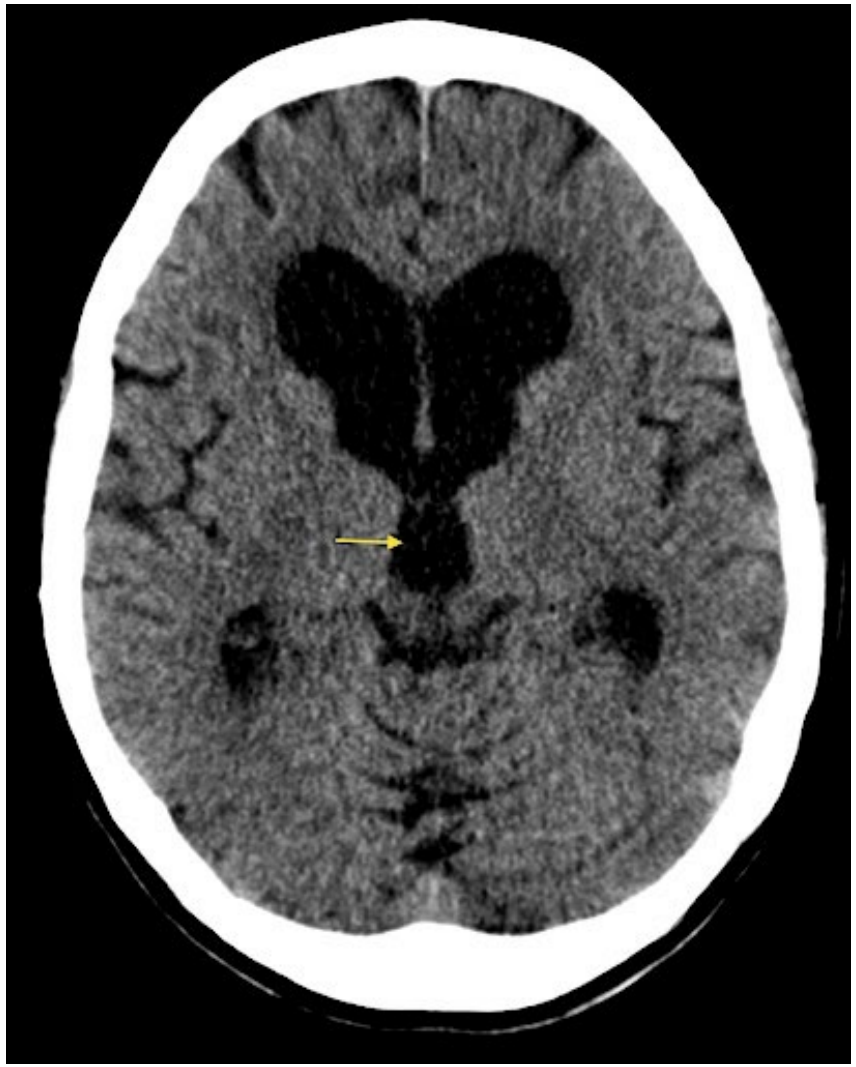
- Clinical Evaluation – Thorough history and neurological examination focusing on triad
- Neuroimaging – MRI of brain (gadolinium not needed) preferred but CT is fine if cannot get MRI
  - Non-Lesional Ventriculomegaly with periventricular signal changes
  - Evan's Index: ratio of widths of frontal horns to maximum width of inner table of skull (>0.3)
- Cisternogram – delayed clearing if isotope from lateral ventricle at 24 and 48 hours



- Response to CSF withdrawal – removal of 20-50 cc of CSF
- Neuropsychological Testing
- Formal Gait Analysis

Source:

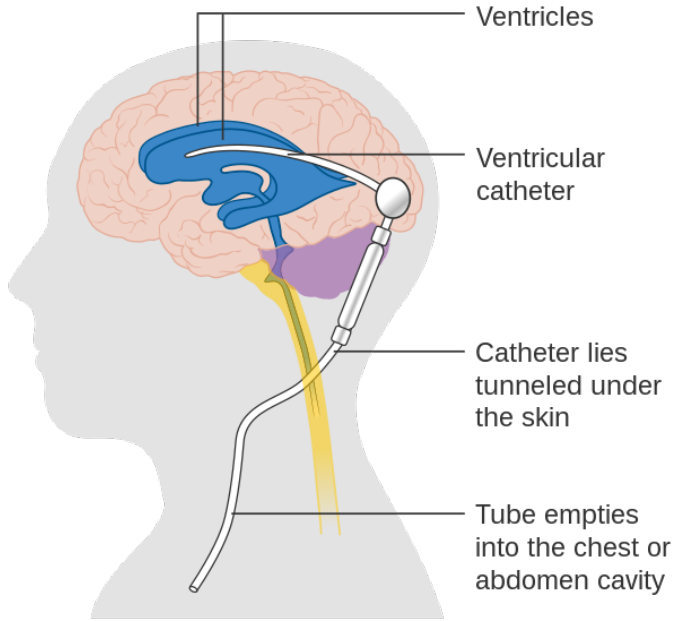




Source:

Treatment Options

- Monitor mild, early symptoms
- Medical Treatment NOT Effective – unlike pseudotumor cerebri, Diamox not effective)
- CSF Diversion
  - * Ventriculoperitoneal Shunt (VPS) placement – most common and highest success rate with lowest complication
    - 70-90% success rate, low shunt infection rate (compared to congenital shunted hydrocephalus)
  - Lumboperitoneal Shunt (LPS placement
  - Endoscopic Third Ventriculostomy
    - 30-50% success rate
    - No hardware, can still shunt
    - Patients with contra-indications to shunting



Source:

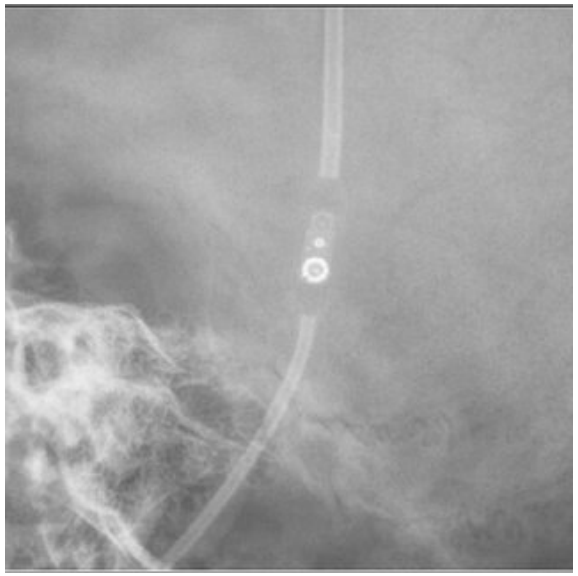
Lumbar Puncture or Lumbar Drain

- Pre-Procedural gait (+/- memory) analysis
- Sedated procedure with measurement of opening pressure and high volume CSF withdrawal (+/- CSF analysis)
- Post-Procedural gait (+/- memory) analysis
- Engage family in post procedural symptomatic improvement
- Regression of symptom improvement over time (1-2 weeks)

Source:

Advances in VP Shunting

- Minimally Invasive Techniques
- Programable Valves
- Antibiotic Impregnated Catheters and Tubing
- Neuronavigation & Neuroendoscopy
- Laparoscopy
- 20-30 minute procedure, less than 24 hour hospital stay



Source:

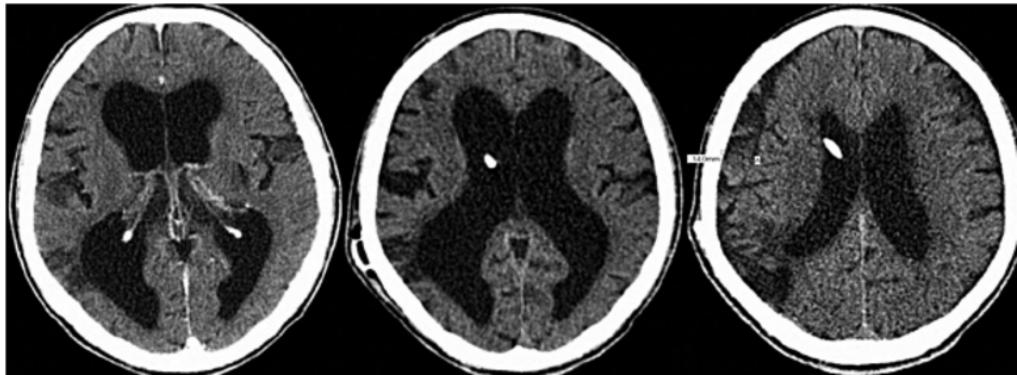
Outcomes & Complications

- **Over 85% success rate at Ochsner, >90% over last 6 years**
- Gait improvement is the first and longest lasting improvement
- Urinary symptoms last and lower improvement rate than gait and cognitive symptoms

- Requires valve programming optimization over time
- Outpatient PT

- Infection Rate - <4% compared to literature of 5-15% - lower than other shunt infection rates
- Subdural Hematoma – 6% compared to 5-15% in literature – due to over-drainage – most are treated non-surgically by dialing valve resistance up or valve to off
- Shunt re-operation rate for failure < 10% over lifetime

- Success rate > 10 years drops to 55-70%



Source:

Referral Guidelines

- Progressive symptoms do not have to be triad – gait and cognitive symptoms are main drivers
- Imaging (MRI or CT) of ventriculomegaly in setting of clinical symptoms
- NPH is underdiagnosed – referral to neurosurgery or neurology if clinical picture unclear – lots of mimickers
- Earlier intervention yield better results
  - Quality of life
  - Less falls and fall-associated hip fractures and other fractures
  - Mobility related health issues - DVT, PE, etc…

Source:

Ochsner Neurosurgery NPH Care Providers

- **Erin Biro – Ochsner Main Campus, Baptist**
- Lora Kahn – Ochsner Main Campus, Baptist
- Marcus Ware – Ochsner Main Campus, Westbank
- CJ Bui – Ochsner Main Campus
- Benjamin Brown – Ochsner Northshore – St. Tammany Parish Hospital
- Saul Morales – Ochsner Lafayette General

Source:

Questions or Case Discussion

Thank You!

- *Erin Biro*

- *C: (716)-491-7131*
- *O: (504) 842-4033*

- *ebiro@ochsner.org*

Source: