Normal pressure hydrocephalus
In 1964, Colombian neurosurgeon Salomón Hakim and colleagues described a syndrome of:

- Progressive cognitive decline
- Gait difficulties
- Urinary incontinence
- Ventricular dilatation
- Normal cerebrospinal fluid (CSF) pressure during lumbar puncture
Normal pressure hydrocephalus

- One of the few causes of reversible dementia
  - Gait impairment
  - Urinary incontinence and
  - Dementia

- Only a small percentage $\rightarrow$ NPH

Most common disorders in geriatric population
The prevalence of NPH is uncertain

- **In Sweden**: 1 per 60,000 persons
  - (1.67 per 100,000) per year underwent shunting for presumed NPH between 1996 and 1998
- **In German**: 1.8 per 100,000 citizens per year
1% to 1.6% of dementia was attributed to NPH

A study from Denmark suggests that 4% of patients referred for dementia were ultimately diagnosed with NPH

Experts have suggested that NPH constitutes as much as 5% of the dementia

Etiology

- Idiopathic NPH → 2/3
- Secondary NPH → 1/3
  - Meningitis
  - Encephalitis
  - Head injury (including concussion)
  - Subarachnoid hemorrhage
  - Other processes causing an inflammatory response in the subarachnoid space
- Normal CSF production by the choroid plexus
  - 0.3 to 0.6 mL/min, and
  - Normal aging - minimal decrease in production
  - An equilibrium between CSF production and resorption maintains CSF pressure at 6 to 20 cm H₂O
Disruption in the CSF circulation leading to gradual enlargement of the ventricles and emergence of symptoms.
- Disruption of CSF flow within the subarachnoid space
- Diminished conductance of CSF to the arachnoid granulations
CSF accumulation

Stretching of periventricular white matter

Changes in subcortical white matter

Decreased cerebral blood flow

Clinical symptoms
Hakim invoked Pascal’s law
(force = pressure \times area)

\[ \text{Force} = \text{ICP} \times \text{ventricular surface area} \]

constant

increases (response to increased ventricular volume)

lower (sustain the force)
Gait impairment

- typically appear before urinary incontinence and cognitive decline

- Gait apraxia
  - loss of ability to produce normal walking movements despite intact strength and sensation

- Gait ataxia
Gait impairment

- **History**
  - Difficulty in walking at the expected pace, climbing stairs, and halting ambulation down a sloping surface
  - Difficulties getting into and out of a chair or bed
  - Walking speed, difficulty navigating obstacles, falls, and reliance on walking aids or need for assistance
Gait impairment

Examination

- Reduced gait velocity
- Shorter stride length
- Broad base
- Difficulty turning - en bloc, multistep
- Decreased foot clearance
- External rotation of the feet
- Poor dynamic equilibrium
“Magnetic gait”

- A wide base
- Tiny steps
- Shuffling with minimal clearance

As though the floor maintained a magnetic pull
### Gait impairment

<table>
<thead>
<tr>
<th>Normal pressure hydrocephalus</th>
<th>Parkinson’s disease</th>
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<tr>
<td>bradykinesia, short stride length, and freezing</td>
<td>same</td>
</tr>
<tr>
<td>Widened base</td>
<td>Narrowed base</td>
</tr>
<tr>
<td>Normal arm swing</td>
<td>Decreased arm swing</td>
</tr>
<tr>
<td>gait rarely improves with verbal or environmental cueing</td>
<td>responded favourably to external cues by improving gait</td>
</tr>
<tr>
<td>Tremor - rare</td>
<td>Tremor - common</td>
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Urgency, frequency - present initially
Incontinence - typically late symptom of INPH

- Enlarged ventricles stretching the periventricular nerve fibers, particularly the frontal horns of the lateral ventricles
- Partial loss of inhibition of bladder contractions (detrusor hyperreflexia)
Urinary urgency and incontinence

- Referral for evaluation by a urologist and urodynamic studies
  - Recurrent urinary tract infections in women
  - Gynecologic anomalies
  - Prostatism in men
  - Bladder dystonia/dysautonomia in patients of either gender
A primarily subcortical dementia
Decline in immediate and delayed recall with preserved memory storage
Impaired complex information processing and psychomotor slowing
Visuospatial perception and visuoconstructive skills are also affected
- Neuropsychologic testing can be helpful in identifying the severity and potential causes of the dementia
- Mini-Mental State Examination was designed to reveal cortical dementia, patients who have subcortical dementia can have a normal score
- Clock Drawing Test or Trail-Making Test -- > visuospatial $ executive
The Montreal Cognitive Assessment (MoCA) may also be helpful in quickly detecting subtle cognitive dysfunction associated with early NPH.
**Diagnosis**

- **Adam’s triad:**
  - Gait deficit - most crucial for diagnosis
  - Cognitive deficits without gait involvement, the diagnosis of NPH is highly unlikely
  - Urinary dysfunction urinary frequency with urgency is noted first, before the development of incontinence

Patients need not have all three symptoms for NPH
CT or MRI is needed to confirm the presence of enlarged ventricles

- MRI may be more useful than CT in identifying other CNS disorders and providing greater detail

- MRI cine CSF flow - monitored to identify blockages
  - Hyperdynamic flow demonstrated in the aqueduct supports a diagnosis of NPH
Communicating hydrocephalus

- Rounding of the lateral and third ventricles
- Upward bowing and thinning of the corpus callosum
- Pulsation artifact (flow void) in the Sylvian aqueduct

"ventricular enlargement out of proportion to cortical atrophy"
The ratio of the maximum width of the anterior horns of the lateral ventricles to the maximum width of the calvarium at the same level of the foramen of Monro.

- A ratio greater than or equal to 0.3 defines ventriculomegaly.
TABLE 3A Criteria for the diagnosis of INPH

**Signs and symptoms:**
- Gait and/or balance disturbance (mandatory)
- Cognitive dysfunction (optional, most often present)
- Urinary urgency or frequency (optional)

**Neuroimaging (preferably MRI):**
- Communicating hydrocephalus without obstruction of the CSF pathways
- Enlarged ventricles (Evan’s index ≥ 0.3)
- No severe white matter destruction nor extensive cortical atrophy.

**Cerebrospinal fluid:**
- Lumbar CSF pressure between 6–24 cm H₂O
- Raised CSF outflow resistance
- Normal protein and cell count

**Exclusion of:**
- Major traumatic brain injury
- Subarachnoid haemorrhage
- Bacterial meningitis
# Differential diagnosis

## Table 1 Alternative causes to symptoms of NPH

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<td>Alzheimer's disease</td>
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<tr>
<td>Diffuse Lewy body disease</td>
</tr>
<tr>
<td>Vascular dementia</td>
</tr>
<tr>
<td>Parkinson's disease-related dementia</td>
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<tr>
<td>Parkinson-plus syndromes (multiple system atrophy, progressive supranuclear palsy, corticobasal degeneration)</td>
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<tr>
<td>Frontotemporal dementia</td>
</tr>
<tr>
<td>Depression</td>
</tr>
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<td>Medication adverse effect</td>
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<tr>
<td>Prion disease</td>
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<td>Traumatic brain injury</td>
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## Gait impairment
- Parkinson-plus syndromes
- Parkinson’s disease
- Vascular parkinsonism
- Visual impairment (cataracts, glaucoma, macular degeneration)
- Lumbar canal stenosis
- Large joint osteoarthritis
- Peripheral neuropathy Deconditioning
- Fear of falling

## Urinary incontinence
- Benign prostatic hypertrophy
- Pelvic floor insufficiency
- Detrusor instability
- Urinary tract infections
- Medication adverse effect
Confirmation test and determine outcome

- Tracer cisternogram
- High-volume lumbar puncture
- External lumbar drainage
- The CSF infusion test

Other
- cine phase flow MRI
- extended intracranial pressure monitoring and B-waves
- analysis of CSF content
High volume lumbar puncture

- Tap Test
  - Most widely used technique for predicting shunt response
  - removal of 30 to 50 mL of CSF, can be done in an outpatient setting

A positive Tap Test
  - Clinical improvement post–Tap Test (hours to days) after the procedure
  - Correlates with an increased likelihood of a positive response to shunt placement

- No contraindications to lumbar puncture
  - Uninterruptible anticoagulation
  - Signs of pathologically elevated intracranial pressure

**Assessment of response**

- Gait test
- Cognitive testing

Before and between 2 to 4 hours after the lumbar puncture
Table 3
Characteristics of ancillary studies used in diagnosis of normal pressure hydrocephalus and determination of benefit for surgical intervention

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<tr>
<td>Sensitivity</td>
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External lumbar drainage

- Continuous or prolonged external lumbar CSF drainage (ELD)
  - Hospital admission
  - A catheter placed temporarily in the lumbar region
  - Rate of 10 mL/h for 36 to 72 hours (400 to 800 mL of CSF)
  - The greater risks and inconvenience
  - Use in patients that have a negative or an equivocal CSF Tap Test response but in whom there is a high clinical suspicion of NPH
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Determining the pressure volume index (PVI) and resistance to CSF outflow (Rout)
- increase the diagnostic and prognostic accuracy when results of a Tap Test are equivocal
Continuous intracranial pressure monitoring

- The presence of increased pressure spikes (B waves) during sleep that are considered pathognomonic for NPH
- Invasive nature and controversies in interpretation
- Limited application in clinical practice
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Factors favoring clinical improvement in normal pressure hydrocephalus after shunting

Secondary NPH
Gait disturbance preceding cognitive impairment
Mild impairment in cognition
Short duration of cognitive impairment
Clinical improvement (usually in gait) after LP or continuous lumbar CSF drainage
Rcsf outflow of 18 mm Hg/mL/min or greater during continuous lumbar CSF infusion test
Presence of B waves for 50% of the time or greater during continuous lumbar CSF monitoring
Factors weighing against clinical improvement after shunting
Moderate or severe cognitive impairment
Dementia present for 2 or more years
Cognitive impairment preceding gait disturbance
Presence of aphasia
History of ethanol abuse
MRI with significant white matter involvement or diffuse cerebral atrophy
Factors of unproved significance
Long duration of gait abnormality
Absence of aqueductal flow void despite patent aqueduct (on MRI)
No clinical improvement after LP
Cisternography
CBF measurements
Pharmacotherapy

- Acetazolamide (carbonic anhydrase inhibitor) and diuretic
  - Reduces the production of CSF by 30% to 50%
  - Palliate cases of mild hydrocephalus
  - Cannot be used for definitive treatment
CSF shunting

- Ventriculoatrial shunting
- **Ventriculoperitoneal shunting**
- Lumboperitoneal shunting
- Ventriculopleural shunting
- Ventriculovenous shunting
Lateral ventricle
Third ventricle
Fourth ventricle

Shunt system to drain spinal fluid from ventricles

Normal

Compression of brain due to build-up of spinal fluid in ventricles

Hydrocephalus

Spinal fluid drains into abdomen
CSF shunting

- Single valve setting (fixed resistance)
  - The simplest first-generation valves offered a to CSF flow to avoid overdrainage

- Programmable shunts (variable resistance)
  - Magnetic mechanism
  - CSF flow → less CSF flow (higher pressure setting) for patients who have evidence of overdrainage (postural headaches or presence of subdural hygroma or hematoma)
Programmable Valves
Fig. 2. (A) Brain of a patient who had NHP before shunt placement; note the large collections of CSF in the sulcal spaces. (B) Brain with NPH in same patient after shunt placement (right lateral ventricle)—sulcal collections of fluid are reduced significantly after shunting.
Figure 2 Development and noninvasive treatment of bilateral subdural hygromas following placement of a programmable shunt for idiopathic normal pressure hydrocephalus. (A) Postoperative head CT shows no evidence of subdural fluid collections. (B) Head CT demonstrating bilateral subdural hygromas (arrows). The patient noted worsening gait and balance similar to her preoperative symptoms. To treat the hygromas, the pressure setting of the shunt was increased. (C) Head CT scan 4 weeks later demonstrating resolution of the subdural fluid collections. The patient's symptoms concomitantly improved.
Endoscopic third ventriculostomy (ETV)

- Mid or late adulthood with NPH Patients
- Long-standing aqueductal stenosis
- Blockage in CSF flow between the third and fourth ventricle
  - A neuroendoscope $\rightarrow$ lateral ventricle via a right frontal burr hole $\rightarrow$ floor of the third ventricle $\rightarrow$ subarchnoid space
Complications related to surgery and anesthesia, such as myocardial infarction and deep vein thrombosis

Acute intracerebral hemorrhage 1% to 2%

Infection of the shunt  8% to 10%, *Staphylococcus epidermidis, Propionibacterium acnes*

Subdural hematoma  <5%

Subdural hygroma (sometimes these may have a small hemorrhagic component)

Seizures

Shunt malfunction

Headache

Hearing loss

Tinnitus

Oculomotor palsies

Damage to intra-abdominal organs

*Neurol Clin 25 (2007) 809–832*
Outcome

- **Improvement in gait** - most frequent observable benefit
- Improvements in cognition, memory, and alertness
  - *less impressive* than the gait improvements
- **Urologic symptoms can improve dramatically**, restoring urinary continence and improving patient quality of life
Prognosis and follow-up

- The success rate: 50% and 90%

- The high variability in reports
  - Differences in selection, shunting method and materials, vigilance in follow-up, and definition of success.

A 2001 meta-analysis

- 59% → improvement immediately after shunting (range, 24%–100%)
- 29% → prolonged improvement (range, 10%–100%),
- 38% → shunt complications (range, 5%–100%),
- 22% → surgical revision (range, 0%–47%),
- 6% → permanent neurologic deficit or death (range, 0%–35%)

In 2005, Marmarou and colleagues

- 90% of patients selected based on response to controlled CSF drainage experienced improvement after shunt surgery

132 patients diagnosed using a protocol of ICP monitoring and controlled CSF drainage

- 33% - improvement in at least one symptom 3 months after shunt surgery
- 60% at 6 months
- 75% at 18 +/- 13 months

All NPH symptoms improved in 46% of patients at 18 +/- 13 months.

Plain radiograph

- Disconnected: Proximal catheter, the valve mechanism, and the distal catheter, including its tip
- which occurs rarely in adults
Fig. 2. 74 year old man presented with return of original NPH symptoms. Plain skull x-rays demonstrate a gap between the proximal end of the shunt valve and the ventricular catheter, as shown by the arrows.
Evaluation of shunt malfunction

- A nuclear medicine shunt patency study
  - injection of a tiny amount of radioisotope into the shunt reservoir followed by observation of flow through the shunt system

- If the shunt is obstructed, flow is absent or significantly delayed
- Improvement
  - over weeks and months
  - can persist as long as the shunt is working and other diseases do not cause functional limitation
Thank you

Hydrocephalus
(from Hess, 1922)