NORMAL PRESSURE HYDROCEPHALUS
Introduction

- Classic clinical triad first described by Hakim and Adams in 1965:
  - Gait disturbance
  - Urinary incontinence
  - Dementia.

- Generally, gait disturbance plus one additional feature is required to consider the diagnosis.

- Secondary forms of NPH
  - Trauma, Hemorrhage, Infection, Mass lesions, or Delayed aqueductal stenosis
## CSF dynamics

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>NPH</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF formation</td>
<td>0.4 ml/min</td>
<td>0.25</td>
</tr>
<tr>
<td>CSF volume</td>
<td>150 ml</td>
<td>300</td>
</tr>
<tr>
<td>Turnover rate (ml/day)</td>
<td>4</td>
<td>1.2</td>
</tr>
</tbody>
</table>
Pathophysiology

- **Increased venous resistance**


- Altered expression of molecules regulating CSF production and absorption

  TNF \( \alpha \), TGF \( \beta \) (increased)

Epidemiology

- Prevalence: 21.9 / 100,000
- Incidence: 5.5 / 100,000


- Race and sex not associated
Gait disturbances

- Most common initial symptom
- Present in 90% patients
- Initially unsteadiness/frequent falls/slow/difficulty initiating/difficulty on turning
- Magnetic gait: broad base, slow, short steps
- Maintained arm swing
- Increased tone, Exaggerated Reflexes, weakness unusual
**Gait**

- **Slowness of gait is responsive to shunt**
  

- **Appendicular tremor: 40% of NPH patients**
  
Pathophysiology of gait disturbances

- Compression of internal capsule fibers by distended third ventricle (Yakovlev 1947)
- Disturbances in basal ganglia pathways


- Compression of brainstem structures, such as the pedunculopontine nucleus

Urinary incontinence

- Frequency, urgency, urge incontinence

- Sakakibara et al. found that 95% of 41 patients with possible iNPH had urodynamic evidence of detrusor overactivity.


- Due to involvement of sacral fibres of corticospinal tracts
D/D of Urinary Incontinence

- BPH
- Autonomic dysregulation
- Anticholinergics
- Diuretics
Dementia

- <5% of all cases of dementia.
- Subcortical frontal dysexecutive syndrome
- Memory loss, decreased attention, impaired planning, slowness of thought & apathy.
- Apraxia, aphasia, agnosia: AD / multi-infarct dementia / fronto temporal dementia
- Asymmetric tremor / lead pipe rigidity: lewy body disease
Dementia

- Even MMSE >25 can have deficits.
- Correlates with vascular risk factors


- Progressive Dementia with normal gait: consider other diagnosis
D/D of gait disturbances and dementia

- **Vascular**: stroke/ Binswanger’s disease
- **Degenerative**: Parkinson’s/ AD / CADASIL
- **Misc**: CSM, LCS, Diabetic neuropathy
Imaging

- Evan’s index > 0.3
- Bicaudate ratio > 0.25
- Temporal horn enlargement
- Periventricular signal changes
- Aqueduct or fourth ventricle flow void
- Supportive / not required

Radionucleotide cisternogram: delayed clearance
Cine MRI: increased ventricular flow rate

SPECT- acetazolamide: Decreased periventricular perfusion not reversed with acetazolamide.
Evan’s Index
Bi-caudate Ratio
MRI in NPH
# Diagnosis

<table>
<thead>
<tr>
<th>Probable</th>
<th>Possible</th>
<th>Unlikely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age &gt; 40</td>
<td>&lt; 40</td>
<td>Papilledema</td>
</tr>
<tr>
<td>Symptoms &gt;3 mo</td>
<td>&lt; 3mo</td>
<td>Other reasonable causes</td>
</tr>
<tr>
<td>Gait disorder most</td>
<td>Non progressive symptoms</td>
<td>No triad</td>
</tr>
<tr>
<td>Urinary incontinence or Dementia</td>
<td>+/-</td>
<td>No ventriculomegaly</td>
</tr>
<tr>
<td>OP 70 -245 mm H20</td>
<td>Abnormal / NA</td>
<td></td>
</tr>
<tr>
<td>Evan’s index &gt;0.3</td>
<td>Cerebral atrophy</td>
<td></td>
</tr>
<tr>
<td>Temporal horn enlargement</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PVO</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aqueductal / Fourth ventricle flow void</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Callosal angle &gt; 40</td>
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Age should not be considered an exclusionary criterion in those without other surgical risk factors.

Lumbar puncture:
- Sensitivity: 26%, Specificity: 100%

Extended lumbar drainage
- 50-80% sensitive, 80% specific, PPV: 80-100%

CSF outflow resistance measurement
- > 18 mm Hg/ml/min 46% sensitive, 87% specific

Cine phase-contrast MRI:
- Insufficient evidence to correlate ventricular Stroke volume with outcome
Tap test

- 40-50 cc tapped.
- Gait was formally assessed pre- and post-tap with the Gait Scale
- Gait scale = walking score + step score + time score
- The Step Score is based on the number of steps required to walk 10 m.
- The Time Score is based on the amount of time required to walk 10 m.
- Cognitive function was also assessed pre- and post-tap with mental status screening (Folstein Mini Mental State Exam).
- Post-tap assessments were conducted within 2-4 h after the TT.

External lumbar drainage

- CSF is drained at a rate of 10 to 15 cc per hour for 72 hours.
- **Risk includes** headache, radiculopathy, and bacterial meningitis.
- **Positive predictive value 90% , Negative predictive value 78%**.
- **Positive ELD: shunt**
- **Negative ELD: Risk – benefit ratio**
Practice guidelines

- High CSF pressure should prompt investigation for a secondary cause of hydrocephalus.
- Response to a 40-mL to 50-mL (high-volume) lumbar tap suggests a potential benefit to shunting.
- An ELD may be used to evaluate those who do not respond to a high-volume tap.
- There is no substantial predictive value to MRI CSF flow studies.

Treatment

- **Medical**: (Temporizing measures only)
  - Acetazolamide
  - High volume tap
- **Surgery for patients with favorable risk benefit ratio.**
  - Surgery
    - Shunt
    - ETV
Shunt in NPH

- Ventriculo peritoneal shunt
- Low pressure
- Programmable shunt preferred
- No study has shown significant benefit with a particular type of shunt or valve

ETV

- Patients with relative aqueduct stenosis
- Triventricular hydrocephalus with NPH
- Gangemi et al reported 72% improvement and low rate of complications (4%).

Complications

- 3-4% risk of ICH
- 1-2% mortality
- 2-17% subdural hematoma
- Shunt blockage
- Shunt infection
- Hardware disconnection
- Shunt tube material allergy
Outcome

- Over a period of 10 years and 99 procedures,
  - Rates of death 1%
  - Subdural hematoma 3%
  - Infection 12%
  - Shunt infection 6.7%
  - Need for shunt revision were 33%

**Outcome**

- The pooled mean response rate to shunting for iNPH was 59% in a meta-analysis.


- In those with good long-term survival, sustained improvement is possible, with a rate of 39% documented after 5 years.

THANK YOU