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

	Normal	**NPH**
CSF formation	0.4 ml/min	0.25
CSF volume	150 ml	300
Turnover rate (ml/day)	4	1.2

Pathophysiology

- **Increased venous resistance**

Bateman GA. Vascular compliance in normal pressure hydrocephalus. AJNR Am J Neuroradiol 2000;21:1574–1585.

- Altered expression of molecules regulating CSF production and absorption

  TNF α, TGF β (increased)

Tarkowski E, Tullberg M, Fredman P, et al. Normal pressure hydrocephalus triggers intrathecal production of TNF-alpha. Neurobiol Aging 2003;24:707–714

Epidemiology

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

  Brean A et al (2008)

- 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

Bugalho P, Guimaraes J. Gait disturbance in normal pressure hydrocephalus: a clinical study. Parkinsonism Relat Disord 2007;13:434–437.

- Appendicular tremor : 40% of NPH patients
    - Do not respond to Ventriculo peritoneal Shunt

Krauss JK, Regel JP, Droste DW, et al. Movement disorders in adult hydrocephalus. Mov Disord 1997;12:53–60.

Pathophysiology of gait disturbances

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

Nakayama T, Ouchi Y, Yoshikawa E, et al. Striatal D2 receptor availability after shunting in idiopathic normal pressure hydrocephalus. J Nucl Med 2007;48:1981–1986.

- Compression of brainstem structures, such as the pedunculopontine nucleus

Mocco J, Tomey MI, Komotar RJ, et al. Ventriculoperitoneal shunting of idiopathic normal pressure hydrocephalus increases midbrain size: a potential mechanism for gait improvement. Neurosurgery 2006;59:847–850.

Urinary incontinence

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

Sakakibara R, Kanda T, Sekido T, et al. Mechanism of bladder dysfunction in idiopathic normal pressure hydrocephalus. Neurourol Urodyn

- 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

Hellstrom P, Edsbagge M, Archer T, et al. The neuropsychology of patients with clinically diagnosed idiopathic normal pressure hydrocephalus. Neurosurgery 2007;61:1219–1226.

- 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

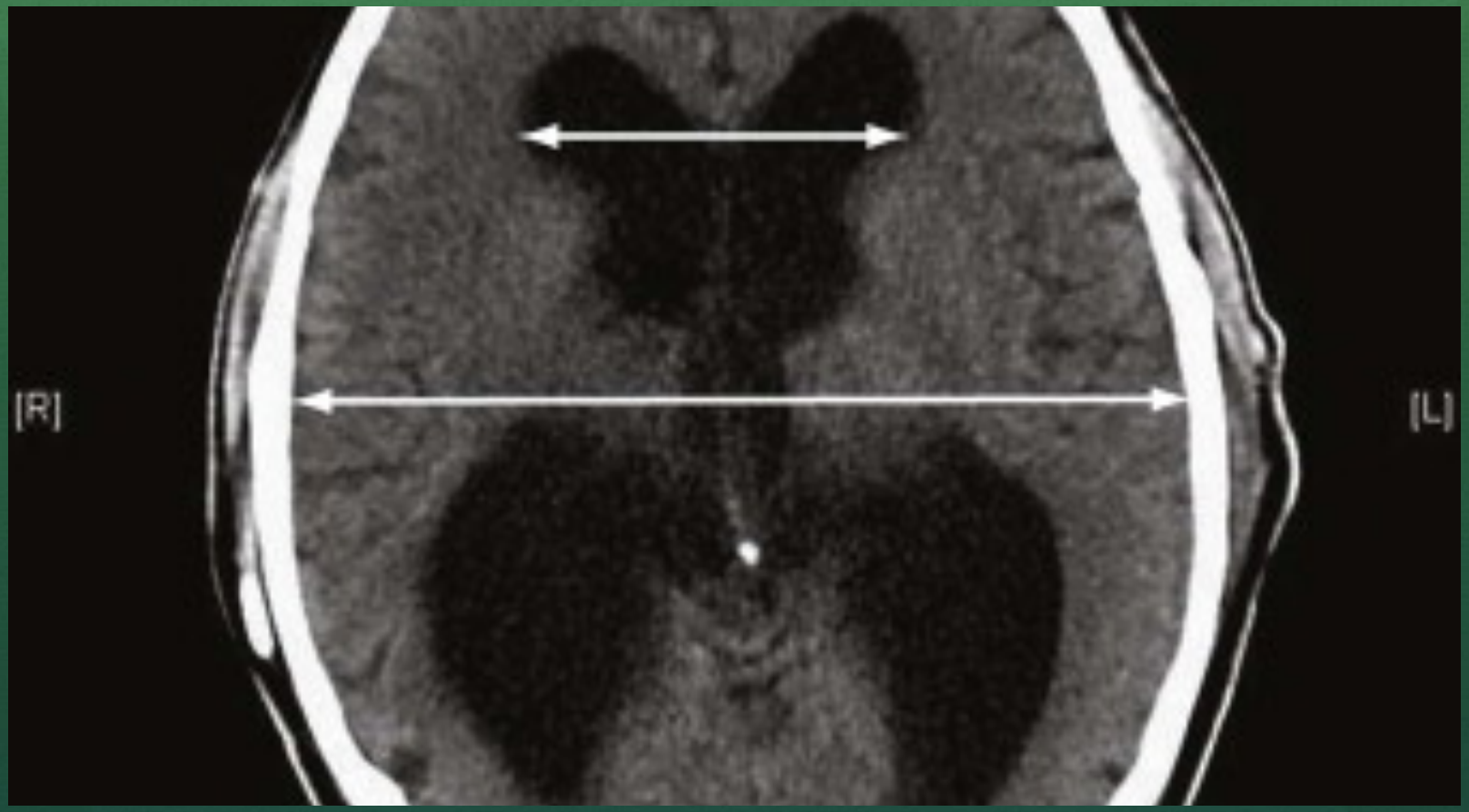
- Evan's index >0.3
- Bicaudate ratio > 0.25
- Temporal horn enlargement
- Periventricular signal changes
- Aqueduct or fourth ventricle flow void
- Supportive / not required

  Radionucleotide cisternogram: delayed clearance

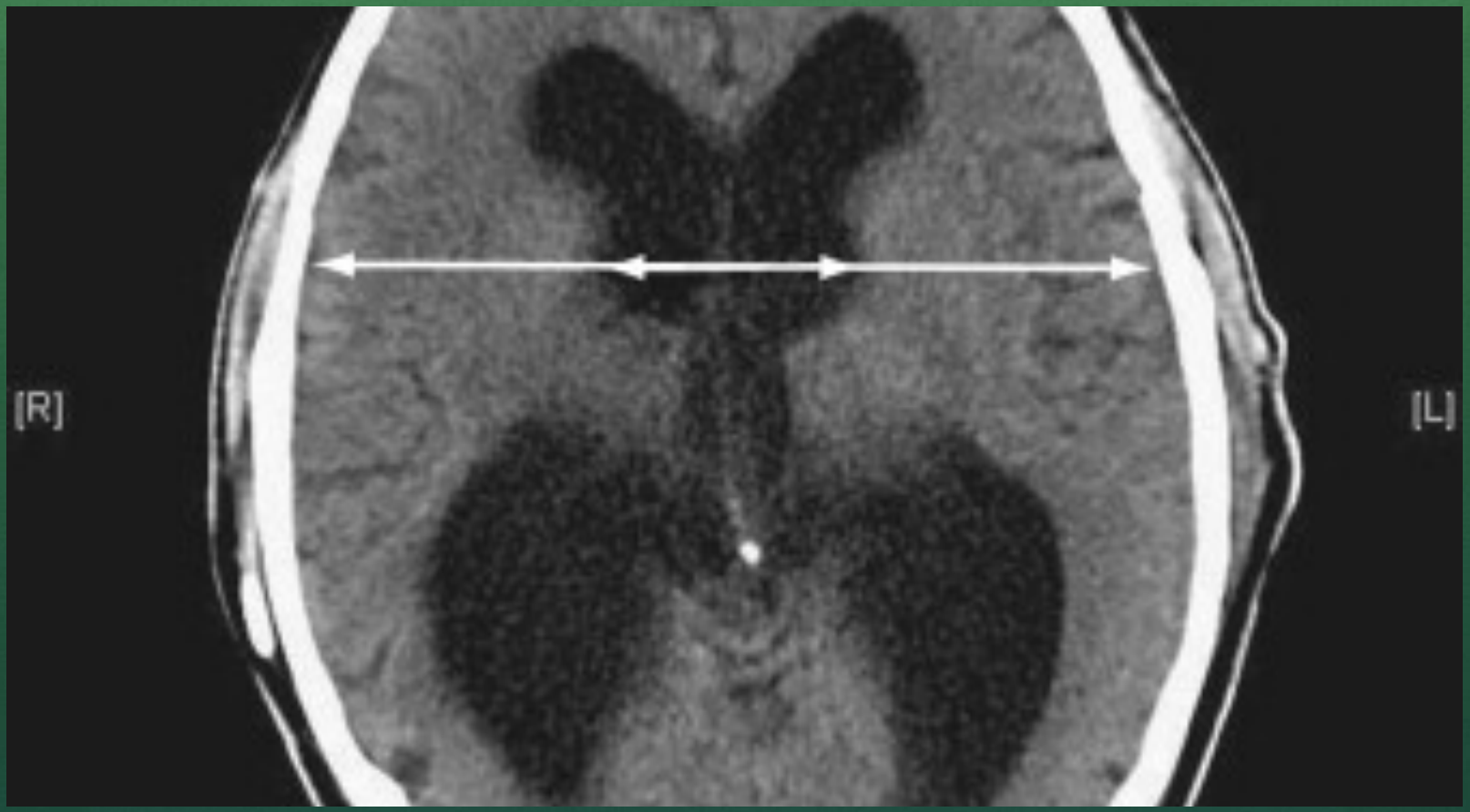
  Cine MRI: increased ventricular flow rate

  SPECT- acetazolamide: Decreased periventricular perfusion not reversed with acetazolamide.

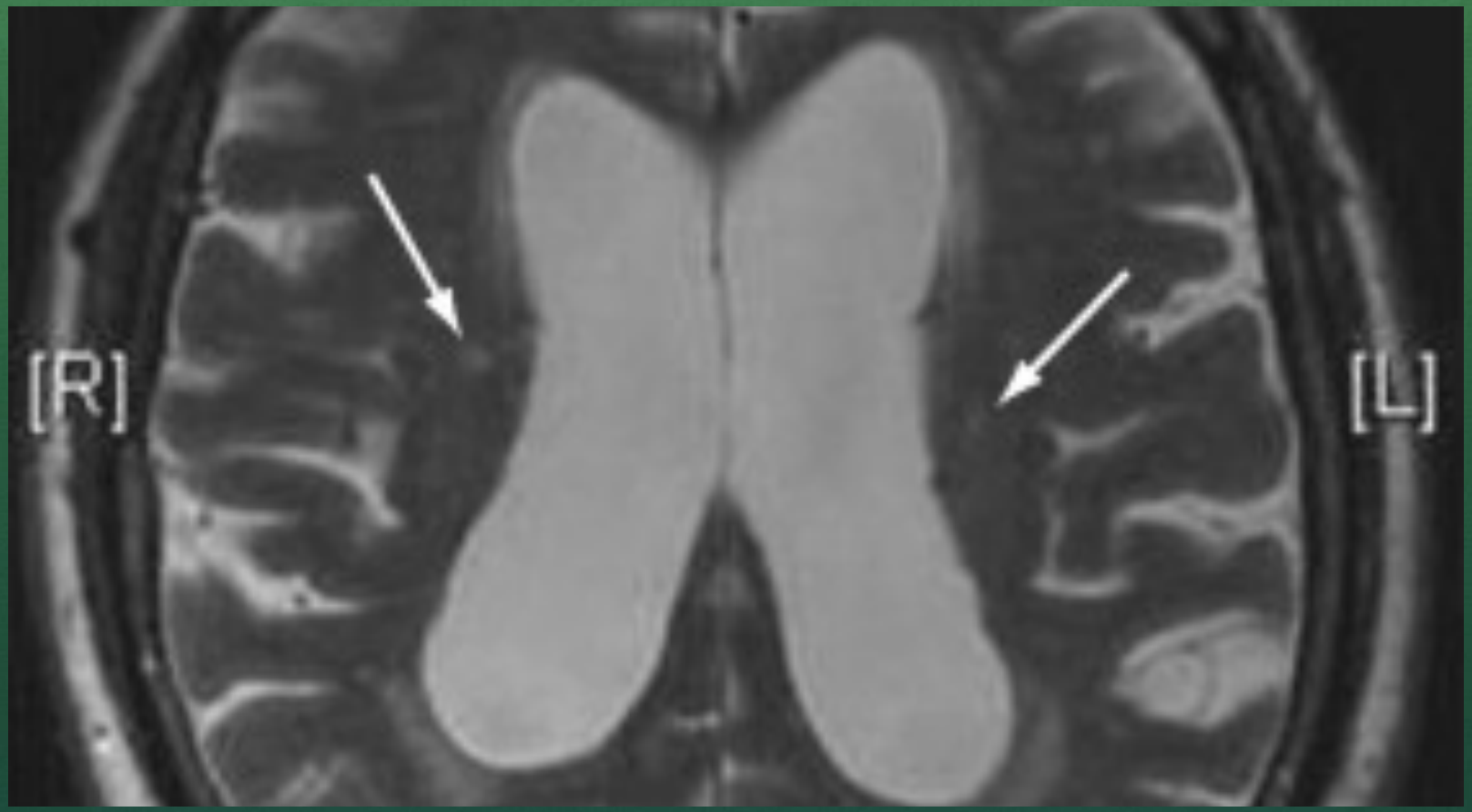
Evan's Index



Bi-caudate Ratio



MRI in NPH



Diagnosis

Probable	Possible	Unlikely
Age > 40	< 40	Papilledema
Symptoms >3 mo	< 3mo	Other reasonable causes
Gait disorder most	Non progressive symptoms	No triad
Urinary incontinence or Dementia	+/-	No ventriculomegaly
OP 70 -245 mm H20	Abnormal / NA	
Evan's index >0.3	Cerebral atrophy	
Temporal horn enlargement		
PVO		
Aqueductal / Fourth ventricle flow void		
Callosal angle > 40		

Relkin N, Marmarou A, Klinge P, et al. Diagnosing idiopathic normal-pressure hydrocephalus. Neurosurgery 2005;57:S4–S16.

Prognosis tests

- Age should not be considered an exclusionary criterion in those without other surgical risk factors.
- **Lumbar puncture:**
  - Sensitivity : 26%, Specifity : 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.

Boon AJ,Trans JT,Delwel EJ,et al, Dutch- normal pressure hydrocephalus study:Randomized comparison of low and medium size shunts, j neurosurg, 1998;88:490

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

Marmarou A, Bergsneider M, Klinge P, et al. The value of supplemental prognostic tests for the preoperative assessment of idiopathic normal-pressure hydrocephalus. Neurosurgery 2005;57:S17– S28.

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

Boon AJ,Trans JT, Delwel EJ et al, Dutch- normal pressure hydrocephalus study:Randomized comparison of low and medium size shunts, j neurosurg,1998;88:490

ETV

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

Gangemi M,Maiuri F,Buona Massa S,etal: Endoscopic third ventriculostomy in idiopathic normal pressure hydrocephalus , Neurosurg 2004;55:129

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%

McGirt MJ, Woodworth G, Coon AL, et al. Diagnosis, treatment, and analysis of long-term outcomes in idiopathic normal-pressure hydrocephalus. Neurosurgery 2005;57:699–705. [PubMed: 16239882] discussion 699–705

Outcome

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

Hebb AO, Cusimano MD. Idiopathic normal pressure hydrocephalus: a systematic review of diagnosis and outcome. Neurosurgery 2001;49:1166–1184.

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

Kahlon B, Sjunnesson J, Rehncrona S. Long-term outcome in patients with suspected normal pressure hydrocephalus. Neurosurgery 2007;60:327–332.

THANK YOU