



## Idiopathic Normal pressure Hydrocephalus. How Much Are We Missing

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### SUMMARY

Normal pressure hydrocephalus (NPH) IS a type of brain malfunction caused by expansion of the lateral ventricles with distortion of the corona radiate fibers. The pathophysiology of idiopathic Normal pressure hydrocephalus remains unclear but is considered to be a communicating hydrocephalus due to reduced CSF absorption. It is theorized that shearing stress forces are exerted on periventricular vessels leading to ischemic changes or it is possibly secondary to interstitial edema with impaired blood flow. It was first described by Hakim and Adams in 1965 [1]. There are 2 types of NPH: Idiopathic (iNPH) and secondary to trauma, tumor, subarachnoid hemorrhage, meningitis...etc. [2]. Symptoms are summarized by Hakim's triad of gait disorder, dementia and urinary incontinence or "Weird Walking Water".

Gait is usually slow, wide based, with foot dragging (magnetic gait) and outwardly rotated feet. Motion in joints is reduced and arm swing can be exaggerated. Falls are common, usually backwards, and is sometimes compensated

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by a forward leaning posture of the trunk [3]. Idiopathic NPH can be described as lower body Parkinsonism, with no rigidity or tremors.

Dementia is frontal lobe, subcortical in type with inattention, apathy, forgetfulness, dullness in thinking and actions [4].

Urinary incontinence occurs late in the course of the disease, is usually urgency in type and secondary to removal of supraspinal inhibition with detrusor over activity [4].

Prevalence of NPH among residents of assisted living facilities was estimated to range between 9-14% [5].

CT scan head is usually used as a screening tool and is one of the reason CT scan is ordered with cognitive impairment or recurrent falls, however its sensitivity and specificity to detect iNPH is unknown. Ventriculomegaly out of proportion to sulcal enlargement has been historically quantified by Evans index (transverse diameter of frontal horns of the lateral ventricle divided by transverse diameter of the skull, usually above 0.3) but is far from being an ideal measurement as it depends on the level at which frontal horns and inner skull diameters are measured (Figure 1). MRI brain is the best modality to diagnose iNPH. Beside ventriculomegaly, it may show periventricular high signal on T2 weighted sequences reflective of trans ependymal migration of CSF [6], thinning of corpus callosum and aqueductal flow void due to increased CSF velocity across the aqueduct of Sylvius, where the CSF appears black on T2 weighted images (Figure 2). Nuclear medicine Cisternography and CT Cisternography are less important in diagnosing iNPH.

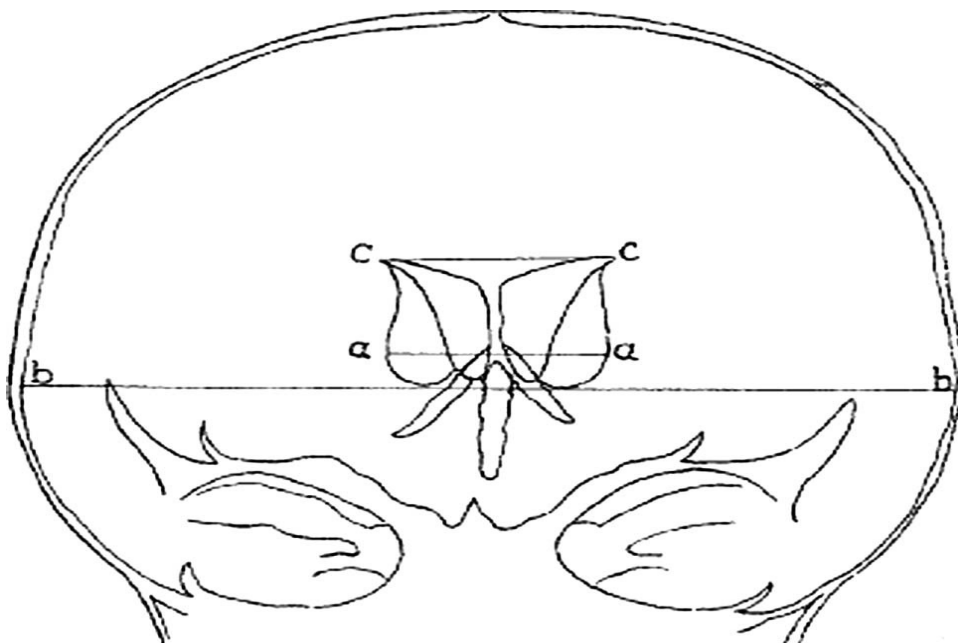


Figure 1: Evans' index revisited

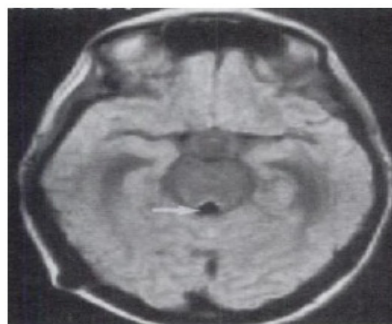
Note: Evans Index=  $cc/bb$  and is normally less than 0.3 [7].

## MRI flow void

- Loss of MRI signal due to flow of CSF



Normal aqueduct



Abnormal aqueduct

Figure 2: Normal-pressure-hydrocephalus

High volume lumbar puncture (Miller Fisher test) where at least 30 ml of CSF is removed should be done in all patients with suspicion of iNPH. It usually showed an opening pressure in the high range of normal limits, i.e. 150-200 mmH<sub>2</sub>O. Gait before and after lumbar puncture should be tested as it has prognostic implications and can predict success of shunting procedure. However, it has low negative predictive value, i.e. lack of improvement in gait speed doesn't rule out the diagnosis of iNPH.

External Lumbar drainage (ELD), when an intrathecal catheter is inserted allowing drainage of CSF for 3-4 days, has the highest sensitivity and specificity to diagnose the condition but carries a risk of infection and catheter dislodgement. Infusion test when Ringer lactate solution is infused via an intrathecal catheter while continuously monitoring pressure is another option permitting the measurement of outflow conductance (C<sub>out</sub>). Continuous Intracranial pressure monitoring may identify patients who would benefit the most from shunt placement and shows intermittent intracranial hypertension in some patients..

Prognostic tests such as Miller Fisher test, ELD and infusion test sometimes yield false results if CSF is altered by severe spinal stenosis [8]. This is particularly important since both cervical and lumbar spinal stenosis can both be in the differential diagnosis of iNPH and may need to be ruled out.

Once the diagnosis of iNPH is established, a shunting procedure, usually a ventriculoperitoneal (VP) shunt is considered. Ventriculomegaly will diminish 3-4 days after placement of the shunt, however only 21% will show

marked improvement in symptoms, usually those with gait disturbances, mild or no urinary incontinence and mild dementia. More recent studies showed that if patients with iNPH, shunt placement yielded beneficial outcomes in 86% of patients (81% in gait disturbances, 70% in urinary incontinence and 40% in cognitive impairment) [9,10].

Both CSF biomarkers [11] and Neuroimaging in general could not predict the ELD outcome, although there is possibly a role for soluble Amyloid precursor protein (APP) alpha in CSF [12]. CSF biomarkers are mainly used to rule out associated disease, e.g. Alzheimer's disease.

In summary, the prevalence of the disease, lack of biomarkers and/or specific neuroimaging findings, and favorable outcome to shunting procedure in a well selected population should guide our assessment in patient with gait disturbances, as follows:

1. Our target should be the population above 65 Y old where the incidence of iNPH is about 0.5%, with peak onset in 6th and 7th decades [13]. A large Norwegian study showed a significant increase in incidence with increasing age (3.3 per 100K for people 50-59 Y of age, up to 181.7 per 100K for people 70-79 Y of age [14].
2. Selection of ideal patients for shunting procedure is crucial. Favorable prognostic factors include [14]:
  - short duration of symptoms (less than 6 months)
  - onset of gait abnormality preceding dementia
  - temporary symptom relief after high volume Lumbar puncture, especially gait
  - absence of significant cerebrovascular disease
  - aqueductal flow void on T2 weighted images.

Given the facts outlined above, one has to wonder how many patients with iNPH, potentially reversible disease, are missed, especially in the elderly population with gait disturbances/recurrent falls who end up in assisted living or Skilled nursing facilities.

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