Idiopathic normal pressure hydrocephalus clinical features

Currently there is no pathological hallmark for iNPH 1)

The natural course of iNPH is symptom progression over time, with worsening in gait, balance and cognitive symptoms. This deterioration is only partially reversible.

Tipically a elderly presenting with gait abnormality, cognitive decline, and urinary incontinence, with enlarged ventricles of the brain but normal or slightly elevated cerebrospinal fluid (CSF) pressure 2) 3).

It can occur with varying combinations or degrees of each of the elements of the classic clinical triad first described by Hakim and Adams in 1965 4).

However, this complete triad is not always seen. Generally, gait disturbance plus one additional feature is required to consider the diagnosis

Postural stability in NPH is predominantly affected by deficient vestibular functions, which did not improve after spinal tap test. Conditions which improved best were mainly independent from visual control and are based on proprioceptive functions 5).

It is frequently present with cerebral vasculopathy; significantly increased prevalence of cardiovascular disease iNPH patients, which provide evidence that cardiovascular disease is involved as an exposure in the development of iNPH 6).

Idiopathic normal pressure hydrocephalus (iNPH) may present, besides the classic triad of symptoms, extrapyramidal parkinsonian like movement disorders.

Psychiatric manifestation of severe disabling anxiety 7).

Abnormal sleep breathing is frequently associated with iNPH. Validation in larger series is required but Román et al. from the Houston Methodist Hospital and Weill Cornell Medical College, Cornell University, New York, suggest including sleep evaluation in patients suspected of iNPH 8).

In SINPHONI—a Japanese multicenter cohort study looking at the validity of MRI findings in idiopathic NPH (iNPH) 9)—there were only 51% of patients with the complete triad of symptoms. Sexual dysfunction 10) neurological symptoms, psychiatric symptoms, or other infrequently reported signs have circumstantial relation to NPH but may hinder diagnostic processing 11).

Gait disturbance

Gait disturbance is assessed by raters of different professions or with different degrees of experience. Agreement studies are usually done by two raters or more, and comparisons among multiple groups of raters are rare.

Ishikawa et al. aimed to examine the agreement among multiple groups of raters on gait patterns and a grading scale through a video-assisted gait analysis in patients with iNPH. Fifteen participants with iNPH were enrolled. Gait was assessed according to seven patterns, including freezing and wide-
based gaits. The levels of severity (evident, mild, none) were rated by three groups of raters (two neurosurgeons [DR2], three experienced physiotherapists [PTe3], and two less experienced physiotherapists [PTl2]) through a simultaneous video viewing session. Severity of gait disturbance (GSg) was rated using the Japanese iNPH grading scale iNPHGS, and Krippendorff alpha was computed to assess agreement, with alpha ≥0.667 indicating good agreement and alpha ≥0.8 indicating excellent agreement. For group comparisons, 84%, not 95%, confidence intervals were applied. Among the seven gait patterns in the first assessment, excellent agreement was observed in wide-based and short-stepped gaits in only DR2. Good agreement was observed in four patterns, but the agreement by two groups was in shuffling and wide-based gait. There were no gait patterns showing good agreement among three groups. In the second assessment, excellent agreement was observed in three patterns but no gait patterns showed good agreement between two groups or more. Learning effect was observed only for standing difficulty in DR2. In contrast, good or nearly good agreement on GSg was observed among the three groups with excellent agreement in two groups. Agreement on gait patterns among the three groups of raters was not high, but agreement on the iNPHGS was high, indicating the importance of a precise description facilitating differentiation between neighboring grades.

Although no one feature is pathognomonic of the gait disturbance in NPH, the most common descriptors include “shuffling,” “magnetic,” and “wide-based”.

Disequilibrium and slowness of gait (due to short steps and gait apraxia) are common, and the latter feature is more likely to respond to shunting.

Slowness of both upper and lower extremities is common as well and can improve with shunting.

Appendicular tremor is present in 40% of NPH patients, is rarely of a parkinsonian (resting) quality, and does not respond to VPS.

**Urinary incontinence**

The bladder symptoms of iNPH are directly caused by detrusor overactivity, which can result in urinary frequency, urgency, or frank incontinence. Sakakibara et al. found that 95% of 41 patients with possible iNPH had urodynamic evidence of detrusor overactivity.

**Dementia**

*Idiopathic normal pressure hydrocephalus dementia*


3) Adams RD, Fischer CM, Hakim S, Ojemann RG, Sweet WH (1965) Symptomatic occult hydrocephalus


