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Normal pressure hydrocephalus and the predictive value of presurgical tests

Hidrocefalia de pressão normal e o valor preditivo dos testes pré-cirúrgicos

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Normal pressure hydrocephalus (NPH) is characterized by gait disturbance, progressive mental deterioration and urinary incontinence associated with enlargement of the ventricular system and normal cerebrospinal fluid (CSF) pressure, although episodes of increased CSF pressure do occur. In NPH, the excessive accumulation of CSF in the ventricular system is due to an impairment of CSF flow distally to the fourth ventricle ("communicating"). About 50% of NPH cases are "secondary" (due to meningitis, subarachnoid hemorrhage, trauma) while the other 50% are "idiopathic" (iNPH), usually occurring in the 7th decade of life. Normal pressure hydrocephalus is a rare cause of dementia (less than 5% of all demented patients), with iNPH showing an annual incidence between 1.8/100,000 and 5.5/100,000 inhabitants, and prevalence ranging from 0.2% to 2.9% among individuals aged 65 years or older, based on surveys in northern Europe and Japan.

Typical cases present with the triad of (1) gait disturbance as the first and most salient sign; followed by (2) an astheno-emotional syndrome characterized by difficulties in concentrating, increased irritability and fatigability, emotional instability, forgetfulness (more rarely, mild dementia), with psychomotor retardation, apathy, and sometimes a parkinsonian or depressive appearance; and, later on, (3) urinary urgency and/or incontinence. Gait disturbance is the cardinal sign in iNPH, typically a broad-based, short-step, slow magnetic gait with start hesitation and instability that is worst on turning, often with falls[1]. It is not a genuine gait apraxia, since the patient can execute the walking movements without difficulty when lying down, despite freezing their gait when standing on their feet and trying to initiate walking[2]. This gait impairment has been considered a gait ignition failure due to a frontal dysfunction or, more specifically, due to a frontal-basal ganglia disconnection, with uninhibited antigravity reflexes and co-contraction of agonists and antagonists during walking[3]. Most of the iNPH syndrome is explained by a reduction of blood flow and metabolism in the frontal lobes, basal ganglia, medial thalamus, hippocampus, and anterior parts of periventricular white matter[4].

In the typical patients, the diagnosis is almost straightforward and, after shunting, they are the most likely to improve, mainly in their gait disturbance. However, particularly in cases with atypical or incomplete clinical manifestations, there may be differential diagnostic difficulties, as the triad can be mimicked by other conditions such as Parkinson's disease, progressive supranuclear palsy, and subcortical arteriosclerotic encephalopathy (Binswanger's disease), which are much more common than NPH. In the elderly, other more common conditions may also explain the gait difficulties (arthrosis or arthritis, peripheral neuropathy, vestibular disease), urinary incontinence (prostate disease, chronic urinary tract infection), and mental deterioration (Alzheimer's disease), particularly when the cognitive impairment predominates and precedes the motor and urinary disturbance. Thus, these diagnostic difficulties require more accurate methods and criteria for selection of patients for shunt surgery, which can benefit up to 80% of these patients but has up to 50% complication rates that dissuade us from shunting every case of suspected NPH. Among the diagnostic and prognostic supplementary tests, the most used are neuroimaging, intracranial pressure monitoring, the lumbar infusion test, and the CSF tap test.

Neuroimaging with computerized tomography or magnetic resonance imaging in iNPH show the following signs that are decisive for an NPH diagnosis and selection of shunt-responsive patients: ventricular enlargement out of proportion to the cerebral atrophy (with Evans index > 0.3), and associated ballooning of the frontal horns; periventricular hyperintensities; corpus callosum thinning and elevation, with a callosal angle between 40º and 90º (if greater than 90º, it suggests brain atrophy, as in Alzheimer's and Lewy body dementias); widening of the temporal horns not entirely explained by hippocampal atrophy; aqueductal or fourth ventricular flow void; enlarged Sylvian fissures and basal cistern, and narrowing of the sulci and subarachnoid spaces over the high convexity and midline surface of the brain[5,6]. On the other hand, other imaging methods such as radionuclide cisternography, single-photon emission computed tomography, positron emission tomography, and even diffusion tensor imaging or resting-state functional magnetic resonance imaging, though compatible with an NPH diagnosis, as yet do not improve the accuracy of identifying shunt-responsive patients. A "positive" radionuclide cisternography may be seen in other dementias and even in healthy elderly subjects, and it has questionable predictive value.

Intracranial pressure monitoring usually shows peak elevations of CSF pressure (B waves), considered to predict a good postsurgical outcome, especially when they occur in more than 50% of the intracranial pressure recording time[7].

In the lumbar infusion test, saline or artificial CSF infused into the ventricle or lumbar subarachnoid space raises the resistance to CSF outflow with subsequent increase of CSF pressure, which in NPH reaches higher levels than the plateau seen in normal individuals. A resistance to CSF outflow of 18 mm Hg/ml per minute or higher[8], and/or CSF pressure pulsatility[9] are considered predictors of a good surgical outcome.

The CSF tap test consists of quantitative testing of gait and cognitive functions before and after the drainage of 30-50 ml lumbar CSF. It is the only test that can temporarily simulate the effect of an actual shunt, and is able to predict not only the outcome of surgery but also the degree of improvement. The one-tap CSF tap test has a high positive predictive value (up to 100%) but low sensitivity (26-61%), which can be improved by performing a repeated or continuous three-day external lumbar drainage (minimum of 150 ml CSF drained daily), with high sensitivity (50-100%) and high positive predictive value (80-100%)[10]. In the European iNPH Multicentre Study the predictive values of the CSF tap test and lumbar infusion test in 115 iNPH patients did not correlate with the outcome after 12 months (except for an increase in the gait task of the CSF tap test)[11].

In this issue of *Arquivos de Neuro-Psiquiatria*, Souza et al.[12] present the results of a well-designed study of the effect of the CSF tap test on 15 gait variables of 25 NPH patients, showing gait speed as the most responsive parameter, followed by cadence, step length, *en bloc* turning, and step height. Gait speed as the parameter with best response to CSF removal has already been established in the neurological literature. Further studies are needed to disclose postural and gait parameters, other than speed, that, isolated or combined, could best predict surgical results, by analyzing CSF tap test data of larger samples of patients who subsequently had good postsurgical outcomes, as the "gold standard" for iNPH diagnosis remains as the clinical improvement after CSF shunting.

Thus, the most relevant issue is whether or not the patient will benefit from shunt surgery. In this regard, the best predictors are a short disease duration, high cognitive scores (mild or no dementia), a gait disturbance preceding mental deterioration, typical neuroimaging findings, and a positive CSF tap test. Even though the most reliable prediction is achieved with a positive repeated or continuous CSF removal, a negative CSF tap test cannot be used to exclude patients from surgery, if the other predictors indicate a good outcome.

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