

Normal pressure hydrocephalus: A Primary Health Care management proposal

Hidrocefalia de pressão normal: proposta de manejo na Atenção Primária à Saúde

Hidrocefalia de presión normal: propuesta de manejo en Atención Primaria de Salud

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Abstract

Normal pressure hydrocephalus – NPH is one of the potentially reversible causes of dementia. In this work, we provide the clinical tools for the management of NPH to the Primary Health Care – PHC physicians. The PubMed and Cochrane databases were reviewed, and 40 articles were chosen, which include randomized clinical trials, metanalysis and systematic reviews. The symptoms of the classic triad are nonspecific and found in the elderly with other diseases. Gait apraxia is the earliest symptom, followed by cognitive impairment and urinary incontinence. The concomitance of the three symptoms occurs in about 60% of the cases in the advanced stages. Apraxia of gait is presented with short steps, without sensorial, vestibulocerebellar or muscle strength changes, which allows it to be distinguished from other pathological marches. Cognitive impairment is subcortical – psychomotor slowness, inattention, loss of working memory and executive function and preservation of episodic memory – unlike cortical dementias, such as Alzheimer's disease. Urinary incontinence must be distinguished from overflow by prostatism and stress incontinence in women. The PHC physician, when faced with the triad, should request a CT scan or brain MRI and, in the presence of one or two symptoms, rule out other diagnoses. Patients with NPH seek help from the physician in the PHC who must know how to identify and differentiate their symptoms. In the case of suspicion, the study of neuroimaging is mandatory and referral to neurosurgery or neurology is recommended.

Keywords: Hydrocephalus, Normal Pressure; Disease Management; Diagnosis; Primary Health Care.

Resumo

A hidrocefalia de pressão normal – HPN é uma das causas de demência potencialmente reversíveis. Nosso objetivo é fornecer as ferramentas clínicas para manejo da HPN pelo médico da Atenção Primária a Saúde – APS. Fez-se uma revisão nas bases de dados PubMed e Cochrane e foram escolhidos 40 artigos para a apresentação desta proposta, que incluem ensaios clínicos randomizados, metanálises e revisões sistemáticas. Os sintomas da tríade clássica são inespecíficos e encontrados em idosos com outras enfermidades. A apraxia da marcha é o sintoma mais precoce, seguido pelo comprometimento cognitivo e pela incontinência urinária. A concomitância dos três sintomas ocorre em cerca de 60% dos casos nas fases avançadas. A apraxia da marcha se apresenta com passos curtos, sem alterações da força muscular, sensitivas ou vestibulocerebelar, o que permite diferenciá-la de outras marchas patológicas. O comprometimento cognitivo é subcortical – lentidão psicomotora, desatenção, perda da memória de trabalho e da função executiva e preservação da memória episódica –, diferente das demências corticais, como a doença de Alzheimer. A incontinência urinária deve ser diferenciada do transbordamento pelo prostatismo e da incontinência de esforço na mulher. O médico da APS, diante a tríade deve solicitar exame de tomografia ou ressonância cerebral e, na presença de um ou dois sintomas, afastar outros diagnósticos. Os pacientes com HPN buscam na APS auxílio do médico que deve saber identificar e diferenciar sua sintomatologia. Diante da suspeita, o estudo de neuroimagem é mandatário e o encaminhamento à neurocirurgia ou neurologia é recomendado.

Descritores: Hidrocefalia de Pressão Normal; Gerenciamento Clínico; Diagnóstico; Atenção Primária à Saúde.

Resumen

La hidrocefalia de presión normal – HPN es una de las causas potencialmente reversible de demencia. Nuestro objetivo es brindar las herramientas clínicas para el manejo de la HPN por parte del médico de Atención Primaria de Salud - APS. Se realizó una revisión en las bases de datos PubMed y Cochrane y se eligieron 40 artículos para la presentación de esta propuesta, que incluían ensayos clínicos aleatorizados, metaanálisis y revisiones sistemáticas. Los síntomas de la tríada clásica son inespecíficos y se encuentran en ancianos con otras enfermedades. La apraxia de la marcha es el síntoma más precoz, seguido del deterioro cognitivo y la incontinencia urinaria. La concomitancia de los tres síntomas ocurre en aproximadamente 60% de los casos en las etapas avanzadas. La apraxia de la marcha se presenta con pasos cortos, sin alteraciones de la fuerza muscular, sensorial o vestibulocerebelar, lo que permite diferenciarla de otras marchas patológicas. El deterioro cognitivo es subcortical: lentitud psicomotora, falta de atención, pérdida de la memoria de trabajo y función ejecutiva, con preservación de la memoria episódica, a diferencia de las demencias corticales, como la Demencia de Alzheimer. La incontinencia urinaria debe distinguirse del rebosamiento por prostatismo y la incontinencia de esfuerzo en las mujeres. El médico de APS, delante de la tríada, debe solicitar una tomografía computarizada o una resonancia cerebral y ante la presencia de un o dos síntomas, descartar otros diagnósticos. Los pacientes con HPN suelen procurar primero la APS, por lo que los médicos de la APS deben saber identificar y diferenciar sus síntomas. Delante la sospecha, el estudio de neuroimagen es necesario y se recomienda el encaminar para neurocirugía o neurología.

Palabras-claves: Hidrocefalia, Presión Normal; Manejo de la Enfermedad; Diagnóstico; Atención Primaria de Salud.

Introduction

Normal pressure hydrocephalus – NPH initially described by Hakim and Adams, in 1965¹ is a syndrome characterized by the classic triad of dementia, urinary incontinence and gait apraxia, symptoms that are bound to improve after ventricular bypass – VB. Brain imaging (BI), a diagnostic requirement, identifies a dilation of cerebral ventricles.²⁻¹⁰ NPH is responsible for 5 to 6% of all cases of dementia and is considered as a potentially reversible

cause of dementia. Its early diagnosis and treatment are decisive for a favorable prognosis.^{9,11-13}

It is estimated that approximately 750,000 Americans have NPH, with higher frequency as life expectancy increases and among them, 80% were not identified in Primary Health Care – PHC. Still, it is reported that 375,000 Americans diagnosed with dementia or Parkinson's disease – PD have NPH.⁹

NPH can be classified due to its etiopathogenesis as idiopathic and secondary (congenital or acquired). Idiopathic NPH – NPHi is a type of communicating (non-obstructive) hydrocephalus that develops with advancing age, in general, from the sixth or seventh decade of life, with a slow and progressive clinical course. Its incidence ranges from 0.5 to 2.9% in the elderly population, corresponding to 50% of the cases of NPH.^{2,3,7,14-16} Its prevalence is 1,300 cases per 100,000 individuals over 60 years of age.¹⁵ Secondary NPH (NPHs) has heterogeneous demographic characteristics and its causes include: subarachnoid haemorrhage (SAH); traumatic brain injury (TBI); brain tumor and central nervous system (CNS) infections. Its evolution is faster and is observed in different age groups according to the etiology.^{6,7,9,17}

The urinary, cognitive and motor symptoms of NPH result in difficult differentiation with other diseases of the elderly population,^{2,18} which could explain the diagnostic errors, especially in the early stages of the disease. In PHC, it has been observed the confusion of initial symptoms of NPH with “memory loss due to age”, urinary incontinence with prostatism or cystocele and gait ataxia with other changes in the elderly's ambulation. Therefore, a high index of suspicion and clinical knowledge of NPH is warranted.

Almost all the reviewed articles are intended for neurologists/neurosurgeons and none for the PHC physician. On the other hand, most patients with NPH, in the initial stage, go to the PHC doctor. Their initial symptoms generate errors and delay in diagnosis/treatment, which leads to worse prognosis and irreversibility.

The arguments addressed motivated us to carry out this research with the objective of providing the clinical approach of NPH to the PHC physicians, facilitating its identification and early referral. It is of interest to the general practitioner since he/she is the gateway to the health system for most patients with initial symptoms of Normal Pressure Hydrocephalus. It addresses a syndrome considered to be a Public Health problem, due to its prevalence in the elderly population and because it is one of the few potentially reversible causes of dementia. In addition, the early identification of the disease is essential to avoid diagnostic errors and contribute to its favourable prognosis. Our article articulates the work of the non-generalist specialist with family medicine, which is so necessary in these times of the gigantic teaching-assistance gap between medical specialties.

Methodology

In order to present the management of Normal Pressure Hydrocephalus to the Primary Health Care physician, a reflective theoretical research based on argumentation and personal interpretation was carried out. The partial bibliographic survey was obtained from PubMed/MEDLINE and Cochrane databases from January 2012 to December 2020. The following keywords were used: "Hydrocephalus, normal pressure"; "Management"; "Diagnosis"; "Treatment"; "Primary health care". The selection criteria for the articles were: randomized clinical trials; meta-analysis; systematic and literature reviews; articles with age of both sexes and greater than or equal to 60 years.

From the research, 90 results were obtained. The descriptor "Hydrocephalus, normal pressure" generated 64 results, from which 37 articles were selected. The descriptor "Hydrocephaly, normal pressure" associated with "Management" generated 5 results, of which 2 were used. The descriptor "Hydrocephaly, normal pressure" associated with "Diagnosis" generated 9 results, of which 5 were used. The descriptor "Hydrocephaly, normal pressure" associated with "Treatment" generated 12 results, of which 6 were used. None of the articles found were directed to PHC. Thirty-seven articles inherent to the NPH (Table 1)¹⁻⁴⁰ – 35 were written in English, 1 in Spanish and 1 in Portuguese. To describe the anatomy of the ventricular system, an article and a textbook were used. To describe the semiology of differential diagnoses, a textbook was used. This article is presented in the main: anatomy and physiology of the ventricular system (VS); pathophysiology; clinical manifestations and differential diagnosis; complementary exams; diagnostic criteria; treatment; final considerations.

Table 1. Summary table of the bibliographical references found in the databases

Authors	Year of publication	Title	Journal
Pereira <i>et al.</i>	2012	Hydrocefalia de pressão normal: visão atual sobre a fisiopatologia, diagnóstico e tratamento	Rev Med
Mori <i>et al.</i>	2012	Guidelines for Management of Idiopathic Normal Pressure Hydrocephalus: Second Edition.	Neurol Med Chir
Ghosh & Lippa	2014	Diagnosis and Prognosis in Idiopathic Normal Pressure Hydrocephalus	Am J Alzheimers Dis Other Demen
Picascia <i>et al.</i>	2015	A review of cognitive impairment and differential diagnosis in idiopathic normal pressure hydrocephalus	Funct Neurol

Authors	Year of publication	Title	Journal
Eide & Sorteberg	2016	Outcome of Surgery for Idiopathic Normal Pressure Hydrocephalus: Role of Preoperative Static and Pulsatile Intracranial Pressure	World Neurosurg
Chankaew <i>et al.</i>	2016	Bulbar dysfunction in normal pressure hydrocephalus: a prospective study	Neurosurg Focus
Daou <i>et al.</i>	2016	Revisiting secondary normal pressure hydrocephalus: does it exist? A review	Neurosurg Focus
Williams & Malm	2016	Diagnosis and Treatment of Idiopathic Normal Pressure Hydrocephalus	Continuum (Minneapolis)
Smith	2017	Recognizing normal pressure hydrocephalus in older adults	Nursing
Saper	2017	Is there even such a thing as "Idiopathic normal pressure hydrocephalus"?	Ann Neurol
Paranathala <i>et al.</i>	2013	Idiopathic normal pressure hydrocephalus: an important differential diagnosis	Br J Hosp Med
Suchorska <i>et al.</i>	2015	Optimized surgical treatment for normal pressure hydrocephalus: comparison between gravitational and differential pressure valves	Acta Neurochir
Aragonès <i>et al.</i>	2018	Hidrocefalia normotensiva idiopática: alta incidencia en mayores de 80 años	Rev Esp Geriatr Gerontol
Graff-Radford	2016	Is normal pressure hydrocephalus becoming less idiopathic?	Neurology
Martín-Láez <i>et al.</i>	2015	Epidemiology of Idiopathic Normal Pressure Hydrocephalus: A Systematic Review of the Literature	World Neurosurg
Anderson <i>et al.</i>	2017	Challenges in diagnosing normal pressure hydrocephalus: Evaluation of the diagnostic guidelines	eNeurologicalSci
Shaw <i>et al.</i>	2016	Clinical outcomes in the surgical treatment of idiopathic normal pressure hydrocephalus	J Clin Neurosci
Tudor <i>et al.</i>	2015	Endoscopic third ventriculostomy (ETV) for idiopathic normal pressure hydrocephalus (iNPH)	Cochrane Database Syst Rev
Silva <i>et al.</i>	2005	Sistema ventricular: anatomia e patologia aplicadas ao diagnóstico por imagem	J Bras Neurocirurg
Machado & Haertel	2014	Neuroanatomia funcional. 3 ed.	Atheneu
Wang <i>et al.</i>	2020	Pathogenesis and pathophysiology of idiopathic normal pressure hydrocephalus	CNS Neurosci Ther

Authors	Year of publication	Title	Journal
Engel <i>et al.</i>	2018	Paradigm-shift: radiological changes in the asymptomatic iNPH-patient to be: an observational study	Fluids Barriers CNS
Hellstrom <i>et al.</i>	2012	A new scale for assessment of severity and outcome in iNPH	Acta Neurol Scand
Oliveira <i>et al.</i>	2019	Normal-pressure hydrocephalus: A critical review	Dement Neuropsychol
Schirinzi <i>et al.</i>	2018	Cerebrospinal fluid biomarkers profile of idiopathic normal pressure hydrocephalus	J Neural Transm
Kiefer & Unterberg	2012	The Differential Diagnosis and Treatment of Normal-Pressure Hydrocephalus	Dtsch Arztebl Int
Pyykko <i>et al.</i>	2018	Incidence, Comorbidities, and Mortality in Idiopathic Normal Pressure Hydrocephalus	World Neurosurg
Nikaido <i>et al.</i>	2017	Intermittent gait disturbance in idiopathic normal pressure hydrocephalus	Acta Neurol Scand
Peterson <i>et al.</i>	2016	The effect of shunt surgery on neuropsychological performance in normal pressure hydrocephalus: a systematic review and meta-analysis	J Neurol
Porto CC.	2013	Semiologia médica. 7a ed	Guanabara Koogan
Shaji <i>et al.</i>	2018	Clinical Practice Guidelines for Management of Dementia	Indian J Psychiatry
Kockum <i>et al.</i>	2018	The idiopathic normal-pressure hydrocephalus Radscale: a radiological scale for structured evaluation	Eur J Neurol
Mihalj <i>et al.</i>	2016	CSF tap test — Obsolete or appropriate test for predicting shunt responsiveness? A systemic review	J Neurol Sci
Oliveira <i>et al.</i>	2013	Programmable valve represents an efficient and safe tool in the treatment of idiopathic normal-pressure hydrocephalus patients	Arq Neuropsiquiatr
Liu <i>et al.</i>	2016	Clinical outcomes after ventriculoatrial shunting for idiopathic normal pressure hydrocephalus	Clin Neurol Neurosurg
Keong <i>et al.</i>	2016	Imaging normal pressure hydrocephalus: theories, techniques, and challenges	Neurosurg Focus
Toma <i>et al.</i>	2013	Systematic review of the outcome of shunt surgery in idiopathic normal-pressure hydrocephalus	Acta Neurochir
Halperin <i>et al.</i>	2015	Practice guideline: Idiopathic normal pressure hydrocephalus: Response to shunting and predictors of response	Neurology

Authors	Year of publication	Title	Journal
Vakili <i>et al.</i>	2016	Timing of surgical treatment for idiopathic normal pressure hydrocephalus: association between treatment delay and reduced short-term benefit	Neurosurg Focus
Pfanner <i>et al.</i>	2017	Cerebrospinal Fluid Biomarkers as Predictors of Shunt Response in Idiopathic Normal Pressure Hydrocephalus: A Systematic Review	Can J Neurol Sci

Source: Prepared by the authors (2021).

Development

Anatomy and physiology of VS

The VS, through which cerebrospinal fluid (CSF) circulates, is composed by lateral ventricles – LV, third, fourth ventricles and connections between them. The communication between the LV and the third ventricle takes place through the foramina of Monro and from the third to the fourth ventricle through the Sylvius aqueduct. The fourth ventricle continues through the central channel of the spinal cord and with the subarachnoid space through the foramen of Magendie and Luschka¹⁹ (Figure 1).

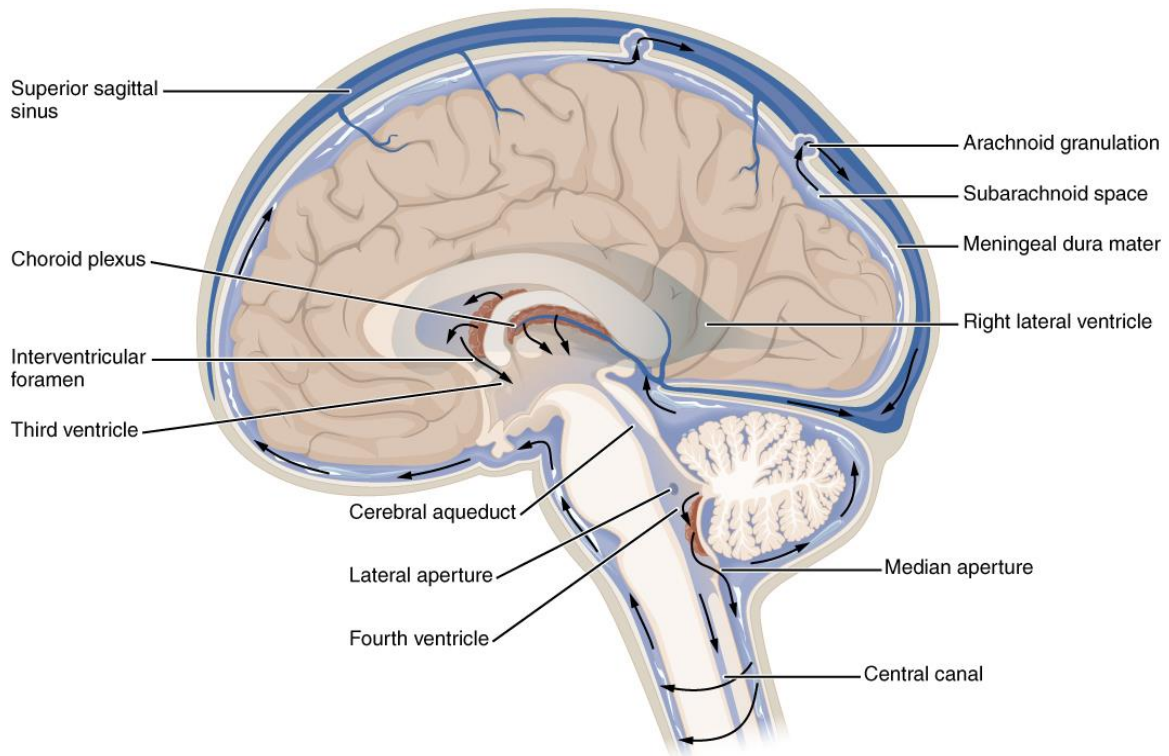


Figure 1. Anatomophysiology of CSF dynamics

Source: http://common.wikimedia.org/wiki/File:1317_CSF_Circulation.jpg.

CSF is produced in the choroid plexus and in the ependyma, circulating throughout the VS, the central channel of the spinal cord and subarachnoid space. Its resorption occurs in the subarachnoid and dura mater granulations. Its volume ranges from 100-150 mL and its main function is mechanical protection.²⁰

Pathophysiology

There is a disorder of CSF dynamics, with dilation of the VS that leads to a reduction in blood flow and brain metabolism, in addition to changes in neuronal conduction by compression of the periventricular white matter. Some of the causes of changes in CSF flow are: fibrosis of the leptomeninges; destruction of arachnoid granulations; sclerotic changes of the vessels and reflux of the contents of the jugular vein.^{2,9,21,22} Despite the identification of such dysfunctions, the pathophysiology of NPH is still poorly understood.^{6,23}

The gray substance appears to be preserved and there is a slow destruction of the white periventricular fibers that generate subcortical dementia. The greater dilation of the frontal horns would explain the predominance of frontal symptoms, such as lower processing speed, sluggish motor activity, urinary incontinence and gait changes, described as Bruns' frontal apraxia.^{21,24}

Clinical manifestations and differential diagnosis

Many diseases are confused with NPH, such as vascular dementia – VD, Alzheimer's disease – AD, Parkinson's disease – PD, chronic alcoholism, brain tumors and CNS infections.^{17,25} The challenges on distinguishing NPH from other CNS diseases justify that about 80% of cases remain undiagnosed and untreated, being one of the most underdiagnosed diseases. The time from onset of symptoms to diagnosis is around 15 months. In addition, about 75% of patients with severe NPH suffer from other brain disorders.^{2-4,9,16,17,26,27}

NPH can be expressed by the classic triad: apraxia of gait, loss of cognitive function and urinary incontinence. The incidence of the triad is unclear. Its symptoms are not specific and are found in the elderly with other diseases. The manifestations are characterized by a slow and progressive course, with gait disorder being the earliest symptom (94 to 100%), followed by cognitive impairment (78 to 98%) and urinary dysfunction (76 to 83%). The concomitance of these three symptoms occurs in about 60% of cases, particularly in the late stages of the disease.²⁵

In addition to the triad, there are other psychiatric and neurological manifestations. Psychiatric disorders include apathy, anxiety, emotional instability and a depressive state. The neurological ones include bradykinesia, hypokinesia, stiffness,

changes in postural and osteotendinous reflexes.^{2,6,9} The more advanced stages include paraparesis, increased deep reflexes and the presence of primitive reflexes, such as the glabellar reflex, handgrip reflex, palmomental and hyperreflexia of the masseter reflex. These primitive reflexes are related to the process of de-frontalisation with the progress of the disease.⁷

The gait in the NPH is characterized by slowness, short steps and an enlarged base with the feet dragged as if they were glued to the ground (magnetic gait). The instability is more pronounced during the change of direction and there is little influence from external factors, such as verbal commands or visual marks, with no learning effect being observed before neurosurgical treatment. The progression of the disease can lead to total inability to walk. The anatomical and functional substrate of the gait disorder are changes in the corticospinal tract and the striatum.^{2,9,16,23,28} There is a lack of loss of muscle strength, sensitivity or signs of cerebellar and vestibular origin, which allows differentiate apraxia from walking with other pathological marches, such as pareto-spastic, polyneuritic, cerebellar or vestibular. Special mention for possible errors in the elderly with osteoarthritis of the hip and knees, which cause changes in ambulation due to functional impotence in the absence of neurological signs. In the early stages of NPH, the balance of the upper limbs is preserved, which allows to rule out parkinsonism. Also, in the advanced stages, secondary parkinsonism may appear with bilateral and symmetrical symptoms such as: plastic stiffness, rest tremors and hypokinesia. These manifestations are confused with PD.⁹

Cognitive impairment presents itself as psychomotor slowness, decreased attention, loss of working memory and executive function. The language is usually slow and monotonous, with reduced verbal fluency. Recognition and episodic memory are relatively preserved in the early stages. The corpus callosum, the striatum, the upper frontal gyrus and the medial portion of the frontal lobe are possible anatomical regions involved in NPH dementia.^{2,24,29} It is understood that this dementia is of the "subcortical" type, which differentiates it from DAD, typically "cortical", expressed by the loss of episodic memory, aphasia, agnosia and apraxia. Furthermore, motor and urinary symptoms of AD, in general, present in advanced stages.

In addition, the neuropsychiatric symptoms of NPH are confused with major depression – MD and subcortical vascular type dementia – VD. In MD there is sadness, anhedonia, pessimism, feeling of guilt and suicidal ideation. A satisfactory response to antidepressant treatment would be expected in MD. VD should be considered in patients with vascular risk factors, history of previous stroke and signs of neurological focus.

Urinary dysfunction consists on overactive bladder with nocturia, urinary urgency, reduced urinary flow, increased residual volume and reduced bladder capacity. In the early

stages, urgent symptoms predominate over incontinence, however, they may not be present in mild cases.² It is recommended to investigate the existence of involuntary urinary loss and to look for urine smell or urine marks on clothes.

Urinary incontinence is present in 30–70% of the elderly and it is necessary to differentiate it from paradoxical or overflow urinary incontinence, typical of prostatism – incomplete emptying of the bladder, terminal drip, thin and weak urinary stream – as well as stress incontinence in women.³⁰ In this sense, in men and women, a good practice would be to perform the examination of external and internal genitalia in the presence of urinary incontinence, to rule out urethral, prostatic or uterine prolapse, according to gender.

Complementary assessment

Laboratory tests are indicated to rule out causes of cognitive impairment: blood count, erythrocyte sedimentation, thyroid, liver and kidney function tests, urinary sediment analysis exam, dosage of vitamin B1, B12 and folic acid, sodium, potassium, total and ionic calcium and phosphorus.³¹ When the clinic suggests encephalopathy from the HIV/AIDS or Neurosyphilis, serological test should be ordered.

The BI is essential for diagnosis and treatment. Cerebral magnetic resonance imaging (MRI) is the most used to visualize ventriculomegaly and define the state of the brain. It is more useful than computed axial tomography – CT, as it allows a better assessment of the posterior fossa, cerebral and hippocampal volumetry. CT is used when there is a contraindication – pacemaker and presence of metals in the body – or unavailability of MRI.^{2,3} It is prudent to explain the Evans index, which is the ratio between the maximum width of the frontal prominences of the lateral ventricles and the width maximum of the internal board of the skull (Figure 2). The index value >0.3 is a hydrocephalus marker.^{2,16,22,32}

Finally, the dynamic investigation of CSF is performed as a test in the diagnosis of NPH.¹⁶ The tap test is a widely used procedure for draining 30 to 50 mL of CSF through lumbar puncture – LP, with documentation of cognitive function and 2 to 3 hours walk before and after the LP.³ The tap test has a sensitivity of 28 to 62%; a low negative predictive value and a specificity of 33 to 100%. The largest volume test performed over days – continuous drainage tests – with extraction of 300 to 500 mL of CSF, has greater reported sensitivity, but specificity comparable to the tap test. The CSF cytochemistry and pressure (180 to 200 mmHg) should be normal. In specialized centers, biomarkers are dosed to ward off AD, such as β 42 amyloid, total tau and phosphorylated tau.^{2,3,9,24,25,33}

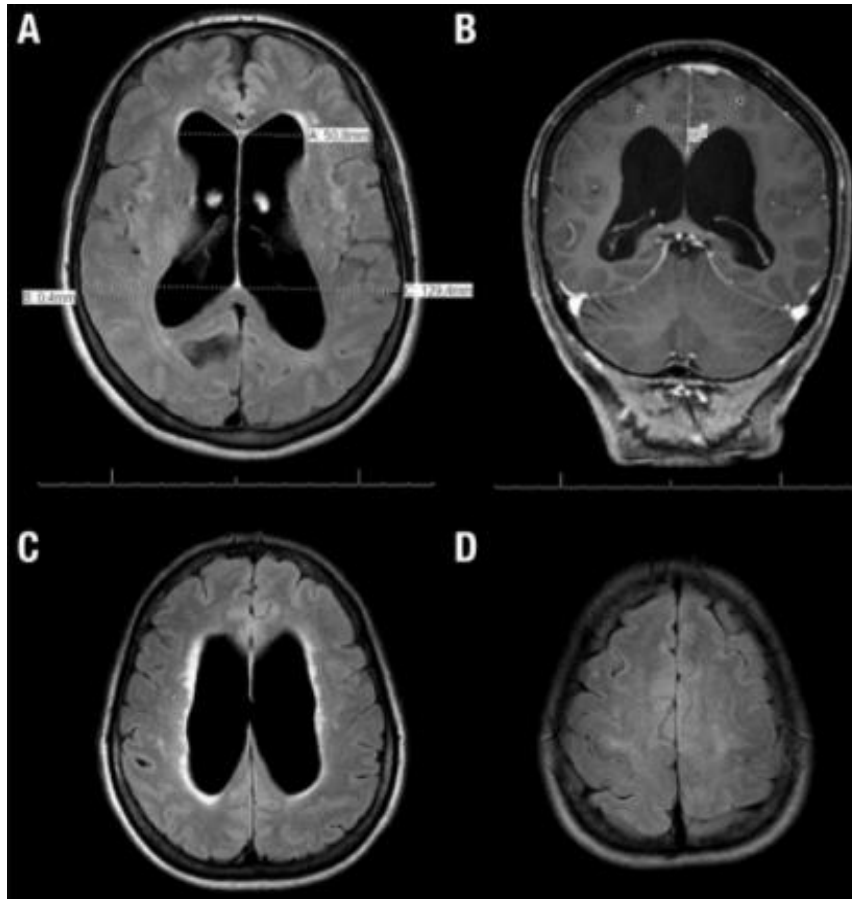


Figure 2. Neuroimaging in HPN. **A.** MRI Flair in axial section showing ventriculomegaly with increased Evans index. **B.** T1-weighted MRI image with gadolinium contrast showing reduction of the callous angle. **C.** Axial MRI showing a bright sign around the white matter, suggesting transependymal edema. **D.** Axial MRI showing narrowing of the grooves and subarachnoid space at high convexity and midline in the region of the frontoparietal surface

Source: Oliveira LM, Nitrini R, Román GC. Normal-pressure hydrocephalus: a critical review. *Dement Neuropsychol.* 2019;13(2):133-43. 10.1590/1980-57642018dn13-020001.

Diagnostic criteria

For the diagnosis, at least two symptoms of the triad and the presence of ventriculomegaly in the BI are required in the absence of CSF flow obstruction. In addition, at least one of these support criteria for brain imaging must be present: enlarged temporal horns in the absence of hippocampal atrophy; callous angle $\geq 40^\circ$; changes in periventricular signal secondary to ependymal transudation, away from demyelinating lesions and microangiopathy.^{7,13,16,32,34}

NPHi is an exclusion diagnosis. The identification of characteristic findings on imaging is called asymptomatic ventriculomegaly (non-obstructive and without secondary causes), characteristic of NPHi.^{2,15,16,25,35,36}

Therapeutic conduct

In case of improved cognition and gait after tap test or continuous CSF drainage, the patient is considered a candidate for ventricular bypass – VB, with a 50 to 96% chance of improvement, considering that no drug treatment is proven effective for a NPH.^{2,7,9,25,37} Ventriculoperitoneal – VPB, ventriculoatrial – VAB and lumboperitoneal – LPB derivation are used. VPB is currently the preferred route, and more studies are being conducted regarding the benefits of LPB, as it presents a lower risk of infection.

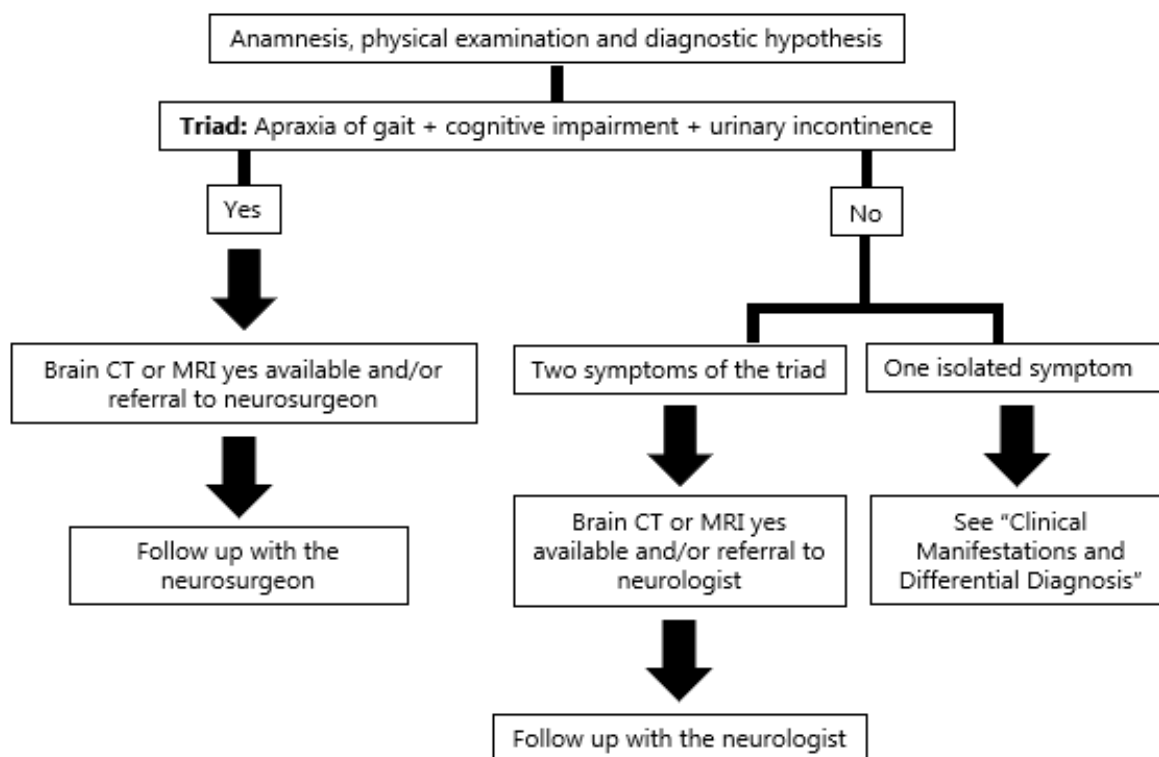
The duration of pre-bypass symptoms has been reported as a favorable prognostic factor, as well as the normality of the biomarkers in the CSF. Age may not be a prognostic factor. VB is the only treatment that has a high evidence.^{7,38-40} After VB, ventricular pressure is reduced, leading to improved cerebral perfusion.³⁴ The effectiveness of the treatment maintained for 3 to 6 months was identified in 64 to 99% of the patients, while in the treatment maintained for 3 to 5 years, it was observed in 28 to 91% of the patients. Gait disorder is the most responsive to VB, followed by the urinary incontinence and cognitive symptoms. Memory, verbal learning and psychomotor speed are benefited, unlike executive function, which undergoes minor changes. Memory improvement after VB occurs mainly in visual and spatial memory.^{2,3,5,9,17,27,29}

As drug treatment, the use of acetazolamide and osmotic diuretics has been recommended as an attempt to control hydrocephalus and memantine for symptomatic treatment of neuropsychiatric symptoms. However, in the performed review, we did not find guidelines that recommend these medications.^{3,24}

Final remarks

NPH is one of the potentially reversible causes of dementia. Its initial symptoms can be presented at numerous pathologies, generating diagnostic errors. The classic triad of dementia, urinary incontinence and apraxia of gait is not always present, which makes its diagnosis difficult, with consequent delay in treatment and worsening of the prognosis. The possibility of reversing the clinical condition with VB justifies the need for an early diagnosis.

Most patients at the beginning of their complaints search for PHC, therefore, the PHC physician must be aware of how to identify the symptoms of NPH, interpret the BI report and its main differential diagnoses. In view of clinical suspicion, imaging is mandatory and referral to neurosurgery or neurology is recommended (Figure 3).



Note: CT = Computed Tomography. MRI = Magnetic Resonance.

Figure 3. Management of normal pressure hydrocephalus by the Primary Health Care physician
 Source: Prepared by the authors (2021).

Field and prospective studies are recommended to define the degree of knowledge of PHC doctors in relation to NPH and to define the time between the onset of symptoms, diagnosis and VB.

Authors' contributions

In an equivalent way, the authors contributed in all stages of the work, as well as all approved the version submitted for publication.

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