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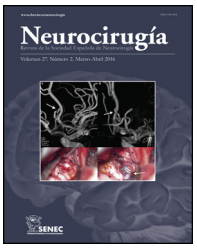

Special article

Long-term prognosis of idiopathic chronic adult hydrocephalus: I. The University Hospital Marqués de Valdecilla diagnostic and therapeutic protocol☆

Rubén Martín-Láez[a,b,c,*], *Alfonso Vázquez-Barquero*[a,b,c]

[a] *Servicio de Neurocirugía-Unidad de Raquis Quirúrgico, Servicio de Neurocirugía, Hospital Universitario Marqués de Valdecilla, Universidad de Cantabria, Santander, Cantabria, Spain*

[b] *Instituto de Investigación Sanitaria Marqués de Valdecilla (IDIVAL), Santander, Cantabria, Spain*

[c] *Observatorio de Salud Pública de Cantabria, Fundación Marqués de Valdecilla, Santander, Cantabria, Spain*

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ABSTRACT

Introduction: Despite the existence of published guidelines for more than a decade, there is still a substantial variation in the management of idiopathic normal pressure hydrocephalus due to its diagnostic and therapeutic complexity.

Development: The diagnostic and therapeutic protocol for the management of idiopathic normal pressure hydrocephalus in use at the Department of Neurosurgery of the University Hospital Marqués de Valdecilla is presented. The diagnostic process includes neuropsychological testing, phase contrast cine MRI, urodynamic evaluation, continuous intracranial pressure monitoring, cerebrospinal fluid hydrodynamics by means of lumbar infusion testing, and intra-abdominal pressure measurement. A patient is considered a surgical candidate if any of the following criteria is met: mean intracranial pressure >15 mmHg, or B-waves present in >10% of overnight recording; pressure–volume index <15 ml, or resistance to cerebrospinal fluid outflow (R_{OUT}) >4.5 mmHg/ml/min in bolus infusion test; R_{OUT} >12 mmHg/ml/min, intracranial pressure >22 mmHg, or high amplitude B-waves in the steady-state of the continuous rate infusion test; or a clinical response to high-volume cerebrospinal fluid withdrawal.

Conclusions: The implementation of a diagnostic and therapeutic protocol for idiopathic normal pressure hydrocephalus management could improve various aspects of patient care. It could reduce variability in clinical practice, optimise the use of health resources, and help in identifying scientific uncertainty areas, in order to direct research efforts in a more appropriate way.

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* *Corresponding author.*
E-mail address: rmlaez@yahoo.es (R. Martín-Láez).

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Pronóstico a largo plazo de la hidrocefalia crónica del adulto idiopática: I. Protocolo diagnóstico-terapéutico del Hospital Universitario Marqués de Valdecilla

Palabras clave:
Derivación ventrículo-peritoneal
Hidrocefalia crónica del adulto
Presión intracraneal
Punción lumbar evacuadora
Resonancia magnética
Test de infusión
Test neuropsicológicos
Urodinámica

RESUMEN

Introducción: A pesar de la existencia de guías clínicas desde hace más de una década, la complejidad diagnóstica y terapéutica de la hidrocefalia crónica del adulto idiopática hace que la variabilidad en su manejo sea elevada.

Desarrollo: Se presenta el protocolo diagnóstico-terapéutico empleado en el Servicio de Neurocirugía del Hospital Universitario Marqués de Valdecilla para evaluar a los pacientes remitidos por sospecha diagnóstica de hidrocefalia crónica del adulto idiopática. El proceso diagnóstico incluye valoración neuropsicológica, RM craneal con secuencias de Cine-RM por contraste de fase, estudio urodinámico, registro continuo de presión intracraneal, hidrodinámica licuoral mediante test de infusión lumbar y medición de la presión intraabdominal. Se consideran candidatos quirúrgicos a los pacientes que cumplen cualquiera de los siguientes criterios: presión intracraneal media >15 mmHg u ondas B en >10% del registro nocturno; índice presión-volumen <15 ml o resistencia al drenaje del líquido cefalorraquídeo (R_{OUT}) >4,5 mmHg/ml/min en el test de bolos; R_{OUT} >12 mmHg/ml/min, presión intracraneal >22 mmHg o presencia de ondas B de alta amplitud en la meseta del test de Katzman; o respuesta a la evacuación licuoral de alto volumen.

Conclusiones: La implementación de protocolos diagnóstico-terapéuticos podría mejorar varios aspectos del proceso asistencial de la hidrocefalia crónica del adulto idiopática, no solo al disminuir la variabilidad en la práctica clínica sino también al optimizar el uso de recursos sanitarios y ayudar a la identificación de áreas de incertidumbre científica, permitiendo dirigir los esfuerzos en investigación de una forma más adecuada.

Introduction

Since its description as a differentiated nosological entity five decades ago by Salomon Hakim,[1] idiopathic normal pressure hydrocephalus (iNPH) has been shown to be one of the main treatable causes of motor disorders, cognitive impairment and urinary dysfunction in the elderly. However, many key aspects of this disease including its epidemiology, pathophysiology, or diagnosis remain full of unknowns difficult to clear up.

Over the age of 65, the prevalence appears to be about 1.3% of the general population and the incidence rate reaches 120 cases/100,000 residents-year.[2] The recent discovery of an exponential increase with age means that iNPH emerges as a disease related to ageing, as occurs with other neurodegenerative diseases such as Parkinson's Disease or Alzheimer's Disease.[3] Additionally, up to 52.4% of iNPH cases do not have typical clinical-radiological characteristics,[3–5] leading to it not being suspected in a considerable percentage of patients, especially in those over the age of 80. The socio-economic consequences of this under-diagnosis should not go unnoticed, as it is estimated that the annual health expenditure due to non-detection and treatment of patients with iNPH exceeds €300 million in Europe and $170 million in the United States.[2,6]

In this context, the need to formalise the diagnostic and therapeutic process of iNPH appears evident. Already in 1972, the article published in the *North Carolina Medical Journal* by Avant and Toole[7] entitled "Diagnostic guidelines in hydrocephalic dementia" showed a diagnostic sequence to determine whether a patient could be a candidate for derivative treatment. However, until the past decade no scientific society at a national or supranational level had developed clinical guidelines for the management of this disease. The Japanese Society of Normal Pressure Hydrocephalus was the first to publish a consent document about the diagnosis and treatment process of iNPH in 2004,[8,9] and was also the first to make an update in 2012.[10] However, the greatest impact was obtained by the International iNPH Guidelines Committee when it published the "International Guidelines for the Management of Idiopathic Normal Pressure Hydrocephalus" in 2005.[11,12] These guidelines have been assumed by much of the international neurosurgical community and have permeated other scientific societies such as the European Federation of Neurological Societies and the European Neurological Society.[13]

In spite of the existence of clinical guidelines for more than a decade, the study by Yoshiyama et al.[14] observed that in Japanese cognitive impairment units, adherence to the recommended diagnostic and therapeutic process was lower than 50%. This situation appears to be reproduced in the literature, since in the bibliographic review on epidemiology of iNPH performed by Martín-Láez et al., only 40% of the articles published since 2005 followed the international or Japanese clinical guidelines.[2] However, the advantages of normalising the management of these patients are evident: they reduce the variability of clinical practice, optimise the use of resources