Normal Pressure Hydrocephalus and Parkinsonism: Preliminary Data on Neurosurgical and Neurological Treatment

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OBJECTIVE: **Idiopathic normal pressure hydrocephalus (iNPH) may present, besides the classic triad of symptoms, with extrapyramidal parkinsonianlike movement disorders. We present a randomized prospective study comparing adjustable ventriculoperitoneal (VP) shunt insertion plus dopamine oral therapy (group A) versus VP shunt alone (group B) in patients affected by iNPH associated with parkinsonism.**

METHODS: **A detailed screening process included neurologic, neurosurgical, and neuropsychological evaluations, followed by a cerebrospinal fluid tap test and resistance outflow measurement. Outcome was evaluated through the Japanese NPH Grading Scale-Revised (JNPHGSR) and the motor (third) section of the Unified Parkinson's Disease Rating Scale, Motor Section (UPDRS-m). Friedman analysis of variance with a Wilcoxon post hoc test was used to evaluate the difference in JNPHGSR and UPDRS-m scores between pretreatment and follow-up (12 months) in the 2 groups, and a Kruskal-Wallis statistic and post hoc Mann-Whitney test were used to compare the change in JNPHGSR and UPDRS-m scores between the 2 groups.**

RESULTS: **Thirty-two of 54 (59%) patients (mean age, 73.2 years) screened in 36 months met the inclusion criteria, but only 30 were enrolled (2 refused surgery) (15 in each group). Preoperative ^{123}I-ioflupane-cerebral single-photon emission computed tomography (DaTSCAN) revealed striatal dopaminergic deficit in 14/30 patients (46.5%). At the final 12 months follow-up, both groups improved JNPHGSR and UPRDS-m scores. The UPDRS-m score improvement was significant in both groups, but greater in group A ($P = 0.003$); JNPHGSR score improvement was similar in the 2 groups.**

CONCLUSIONS: **iNPH associated with parkinsonism may be a frequent finding. In these cases, patients may benefit from VP shunt plus dopamine oral therapy.**

INTRODUCTION

Life expectancy has increased in the last 50 years in developed countries, signifying a great achievement for medicine and health care systems but a critical issue for twenty-first century physicians. The percentage of elderly patients and, consequently, the incidence of neurodegenerative diseases, are constantly increasing.[1] Hence, neurologists and neurosurgeons are facing increasingly more patients affected by age-related diseases.

Idiopathic normal pressure hydrocephalus (iNPH) is a potentially treatable neurologic disorder of the elderly. It comprises disturbances of gait and balance, urinary control, and cognition (the so-called classic triad of symptoms of the disease) in

Key words
- Cerebrospinal fluid
- DaTSCAN
- Normal pressure hydrocephalus
- Parkinsonism
- UPDRS
- Ventriculoperitoneal shunt

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
DaT: Dopamine transporter
DaTSCAN: ^{123}I-Ioflupane-cerebral SPECT
FAB: Frontal Assessment Battery
iNPH: Idiopathic normal pressure hydrocephalus
JNPHGSR: Japanese NPH Grading Scale-Revised
MMSE: Mini Mental State Examination
MODA: Milan Overall Dementia Assessment
SPECT: Single-photon emission computed tomography
TUG test: Timed Up And Go test
UPDRS-m: Unified Parkinson's Disease Rating Scale, Motor Section
VP shunt: Ventriculoperitoneal shunt

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Citation: World Neurosurg. (2016) 90:348-356.
http://dx.doi.org/10.1016/j.wneu.2016.03.004

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

combination with enlargement of the cerebral ventricles.[2] Gait and balance disturbances are the most common clinical findings and may occur alone or together with cognitive and urinary symptoms.[2-4] Diagnosis of iNPH is often challenging because of its varying presentation and overlapping with other disorders common in the elderly such as cerebrovascular and neurodegenerative diseases, urologic dysfunctions, lumbar stenosis.[5,6] The most effective treatment offered to patients is surgical shunting with ventriculoperitoneal (VP) shunt,[7,8] usually with programmable valves.[9]

Furthermore, not all patients do show the classic triad of symptoms; many patients often present signs of movement disorders, such as parkinsonism.[10-12] Diagnosis and treatment become even more challenging in this group of patients and adequate management requires both the neurologist and the neurosurgeon.[13] These patients may have been treated surgically with a VP shunt or with L-dopa medications; however, there is a conspicuous lack of information concerning the outcome and, more important, the proper therapy.[5,8,14]

The incidence of this association is not clear, varying from 10% to 70% of cases in previous reports.[12,14] This condition is probably underestimated because the diagnosis, as mentioned earlier, is challenging and requires 2 skilled specialists.

The pathophysiology of parkinsonian symptoms in iNPH has not been conclusively understood. The abnormal pulsation of cerebrospinal fluid (CSF) flow occurring in hydrocephalus may

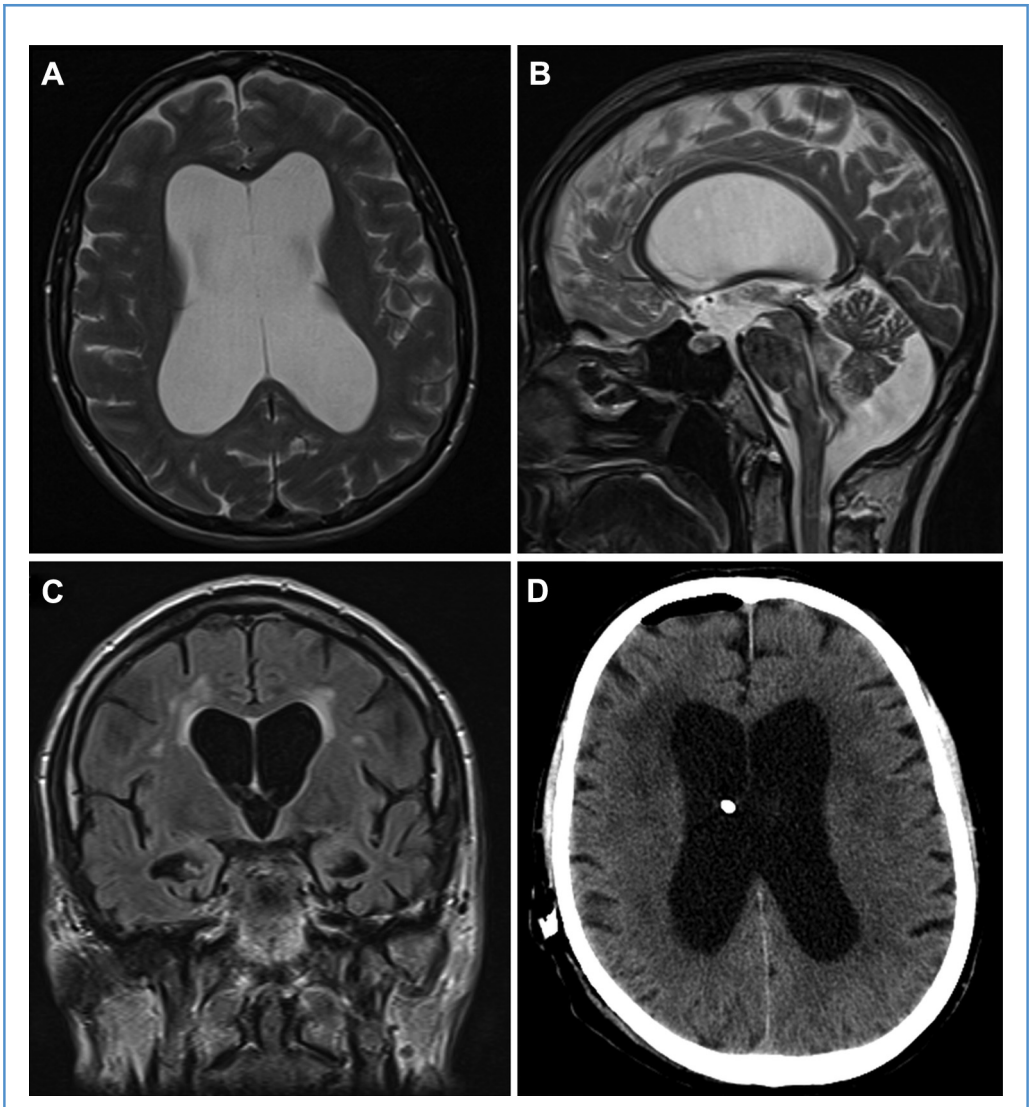

Figure 1. (**A**) Preoperative axial T2-weighted magnetic resonance image (MRI) showing enlargment of lateral ventricles. (**B**) Preoperative sagittal T2-weighted MRI; note how the corpus callosum is stretched by the hydrocephalus. (**C**) Preoperative coronal fluid-attenuated inversion recovery MRI; note the mild transependimal cerebrospinal fluid reabsorption. (**D**) Postoperative axial computed tomography scan after ventriculoperitoneal shunt with the ventricular catheter inserted into the righ lateral ventricle through a right precoronal burr hole access.