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Mortality and neurological worsening in idiopathic normal pressure hydrocephalus after CSF diversion: prospective study with long-term follow-up

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Resumen

Introduction: Idiopathic normal pressure hydrocephalus (iNPH) is a rare disease and many aspects of its pathophysiology, diagnosis and treatment remain unclear. Studies dealing with long-term outcome after shunt treatment are scarce, as also is the literature available about mortality, the rate of neurological worsening and their causes.

Methods: In 1999, a protocol evaluation and treatment of patients referred with suspicion of iNPH to the Department of Neurosurgery of the University Hospital "Marqués de Valdecilla" was introduced. This protocol included neuropsychological and neuroradiological evaluation, overnight continuous ICP monitoring and hydrodynamic lumbar infusion tests.

Results: Between January 1999 and December 2012, 175 patients were operated on with the diagnosis of iNPH. The estimated incidence of iNP in Cantabria was 2.49+-1.58 cases/100,000 inhabitants-years. Postoperative complications appeared in 14.8% of patients, with a perioperative mortality rate of 2.7%. The rate of revision surgery was 8.5 cases/100 patients-years. During the first year, 16.15% of patients required at least one shunt revision, with a subsequent annual rate of revision of 3.20%. The revision rate of ventricular-jugular shunts against the venous flow doubled that of other shunt configurations. The neurological worsening free survival was slightly less than three years, with an incidence of 14.9 cases of neurological worsening per 100 patients-years. The severity of clinical manifestations and the predominance of cognitive symptoms were identified as independent predictive variables for its development. The mortality of untreated iNPH is almost eight times higher than that of general population. Surgical treatment modifies the natural history of this disease, decreasing the likelihood of death by 53.3% and placing it at rates similar to that of stroke or ex-vacuo ventricular dilation. Multivariable analysis identified gender and extent of white matter hyperintensities as independent predictors of mortality.