Idiopathic normal pressure hydrocephalus: the benefits and problems of shunting

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The syndrome of idiopathic normal pressure hydrocephalus (INPH) is characterized by ventriculomegaly in the absence of raised intracranial pressure and presents as the clinical triad of gait disturbance, urinary incontinence and cognitive impairment. Diversion of the cerebrospinal fluid (CSF) through implantation of a shunt is the mainstay of therapy for INPH. However, a number of difficulties face the medical practitioner in the selection of patients for shunt surgery and provision of optimum follow-up. We outline these issues and describe the methods by which we believe clinicians can ensure the best possible outcomes from shunting for INPH.

One of the first issues that confronts practitioners is the fact that diagnosing INPH is itself problematic—symptoms are often attributed to aging or to other conditions that mimic hydrocephalus, causing the diagnosis of INPH to be missed. To ensure the selection of those patients who are most likely to benefit from shunt placement, it is critical, therefore, that a sensitive and accurate method of selection is used.[1] If shunt placement was offered solely on the basis of patient history, examination, and presence of ventriculomegaly on neuroimaging, only 46–61% of patients with possible or probable INPH would benefit from surgery. To improve these odds, practitioners have supplemented these investigations with additional selection criteria based on physiological testing (such as the determination of CSF outflow resistance) or functional testing (such as the CSF tap test and extended lumbar drainage [ELD]). ELD, whereby small amounts of CSF are drained daily over a period of 3 days via a spinal catheter, has been shown to have the highest sensitivity and specificity (60–100% and 50–100%, respectively).[2] In fact, the additional quantifiable improvement in patient function during ELD denotes a much greater chance of surgical success than the mere presence of the clinical and radiological appearance of INPH. In a prospective study of 151 patients, preoperative use of ELD as a supplementary test had a 90% predictive value for good outcomes after shunt surgery for INPH.[3] The test was most accurate in patients with predominant gait symptoms and less accurate with increasing number and severity of symptoms in the INPH triad. In a similar study, ELD predicted improvement when gait impairment was the primary symptom, and when the duration of symptoms was short.[4] These data suggest that ELD can accurately select patients who will benefit from shunting, and this is currently the best supplementary test available.

Careful selection of patients for surgery and accurate monitoring after shunt placement is possible only if tests are available that can detect and measure changes in function after treatment of INPH. Current tools—for instance, the commonly used Mini Mental State Examination and the Tinetti balance scale—are unable to quantify the subtle changes in cognition and gait that occur in patients with INPH. We therefore strongly recommend the use of more-sensitive cognitive tests that rely on the measurement of delayed auditory recall and visual-constructive abilities,[5,6] and use of computerized gait analysis that provides a more-objective assessment of gait before and after shunt placement.[3,7]

Improvements in anesthesia and post-surgical care have greatly decreased the morbidity of surgery in general; however, it is the advent of newer technologies that has radically improved outcomes after shunt surgery. Although there is no consensus on the type of valve that should be employed in INPH, the majority of surgeons agree that the use of adjustable valves can prevent overdrainage of CSF, which is a cause of subdural hematoma, or underdrainage of CSF, which is a cause of inadequate relief of INPH symptoms.[8,9] Furthermore, the use of antibiotics-impregnated catheters seems to reduce the risk of infectious complications. Close monitoring of patients, leading to prompt recognition of shunt malfunction and timely correction of shunt obstruction, will maximize recovery after shunt surgery and minimize complications.[10] Recently published data suggest that endoscopic third ventriculostomy (ETV) is effective as a treatment for INPH and should be

offered as an alternative to shunting.[11] However, while ETV is the procedure of choice for obstructive hydrocephalus, its reported efficacy in patients with INPH might be explained by the presence of a subarachnoid or ventricular obstruction that is not visualized without advanced neuroimaging. We believe that firmer criteria are needed that can identify the subset of patients with INPH who are likely to benefit from ETV before this procedure is widely used in this group.

Occasionally, outcome after shunting is unsatisfactory despite a clinical improvement during ELD and excellent surgical technique—because of, for example, progression of associated comorbidities or late recognition of INPH.[2,3] In general, however, adherence to strict criteria, leading to careful selection of patients for surgery, will result in favorable outcomes after shunt placement. Several authors have reported sustained overall improvement of symptoms following shunting of 24–80% and 50% at follow-up of 3 years and 5 years, respectively,[10] and many patients are able to return to independence following successful reversal of INPH through shunting.[12] In a series of 55 patients with INPH selected for shunting with ELD as a supplementary test, with a mean follow-up of 6.9 years, 80% of patients had a persistent improvement in gait after surgery. Sustained improvements in cognition and urinary incontinence occurred in a respective 76% and 60% of the patients.[10] Successful reversal of the hydrocephalus syndrome through shunting benefits individuals and also produces remarkable cost savings for society.[12]

The success of this management strategy for INPH could, however, be counterproductive if the resultant enthusiasm were to cause a relaxation of the selection criteria. In addition, the unrelenting advertising that often comes with the discovery of a new promising avenue of treatment can prime physicians and the public to expect benefits beyond those originally indicated. It is natural for patients who are experiencing progressive cognitive decline to hope that shunting might stop or reverse their deterioration, setting the stage for the creation of a strong placebo effect on the patient and on the family. This combination of factors, if not controlled by the application of firm diagnostic criteria, could persuade the untrained surgeon to offer surgery in situations in which it is unsuitable, potentially leading to a surge in complications and a decline in the perceived usefulness of the procedure even in the most appropriate cases.

To conclude, INPH can be successfully managed with use of strict selection criteria based on clinical and radiological characteristics corroborated by properly documented functional improvement during ELD. CSF shunting in INPH produces long-lasting, excellent outcomes in many patients. The potential for abuse of the shunting procedure, however, looms large and demands firm adherence to high-quality standards in the selection process and honest appraisal of the limitations of the procedure.

Competing interests
The authors declared no competing interests.

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