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Guidelines for management of idiopathic normal pressure hydrocephalus: progress to date

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Summary

The aim of this project was to develop evidenced based guidelines for the diagnosis and management of idiopathic normal pressure hydrocephalus (iNPH). An advisory panel consisting of the authors assisted by international experts met on several occasions and formulated preliminary guidelines for iNPH managemen. The authors developed evidentiary tables based on available literature from 1966 to the present. Additional meetings to refine the evidentiary tables and incorporate expert opinion when necessary resulted in the development the iNPH guidelines. Evidence based guidelines identifying the value of clinical examination, brain imaging, Tap Test, CSF drainage, ICP monitoring and Surgical Management in diagnosing and treating the iNPH patient were developed. These are the first international evidence based guidelines focused on iNPH. Class I data were scant and guidelines relied mostly on class II and III evidence. It became clear that more prospective randomized studies are needed to resolve some of the controversial issues such as iNPH classification and sensitivity of diagnostic tests for identifying shunt responsive iNPH.

Keywords: Guidelines for idiopathic normal pressure hydrocephalus; NPH; shunt responders.

Introduction

The symptoms of gait disturbance, incontinence and dementia associated with Normal Pressure Hydrocephalus (NPH), a syndrome introduced by Hakim and Adams [1, 2], seriously impacts upon the quality of life of senior citizens. However the diagnosis of NPH remains controversial, as there are no specific guidelines for diagnosis as well as management. In part, this controversy has evolved as a result of mixing idiopathic NPH from those cases of known cause resulting from trauma, stroke, subarachnoid hemorrhage. Another reason for controversy is the general acceptance by many clinicians that an accurate diagnosis of NPH depends upon the response to shunting. Taking these issues in concert, it is clear that an evidenced based set of guidelines for diagnosis and management of NPH is needed. This report provides an overview of the progress made in development of these guidelines and provides recommendations for future research.

Methods

The guidelines were compiled from a review of the MEDLINE literature in combination with references provided by an expert panel made up of clinical scientists from the U.S., Europe and Japan. Articles from 1965 to the present were considered and the evidence was classified as class I, II or III. Class I evidence was derived from prospective, randomized well-controlled clinical trials. Class II evidence was derived from a prospective data collection with retrospective analysis of clearly reliable data and class III data referred to retrospective data, chart reviews, databases or registries and expert opinion. When expert opinion was required, it was obtained from advisory panels made up of international experts.

Results

The absence of prospective randomized clinical trials of any aspect of NPH required that the guidelines be constructed primarily on class II and III data. A decision was made to classify NPH into two major categories, idiopathic (INPH) and secondary (SNPH) following the initial separation by Black [3, 4].

Clinical diagnosis of INPH

It was also decided that the diagnosis of INPH required convergent evidence from the clinical history, physical examination and brain imaging. The symptoms of INPH typically manifest during adult life as an insidiously progressive chronic disorder that lacks an antecedent cause. Gait and/or balance impairments are usual symptoms and findings may also include disturbances in cognition or control of urination. Documention of ventricular enlargement (Evans Index ≥ 0.30) by brain imaging is necessary but not sufficient in itself to establish an INPH diagnosis. Results of neuroimaging must be interpreted in conjunction with the clinical history and physical findings in order to accurately diagnose INPH and differentiate it from other disorders. It was recommended that it may be useful to classify INPH into Probable, Possible and Unlikely categories, operationally defined by the extent to which the expected elements of INPH are present and diagnostic cofounders can be excluded. Note that shunt responsiveness did not enter into this diagnostic formulation and was considered separately.

Supplementary tests for identifying shunt responders

Drainage of cerebrospinal fluid (CSF) via a lumbar tap can be of prognostic value if the response is significant. Lumbar puncture tap tests should withdraw 40–50 cc since lesser volumes (25 cc or less) have low sensitivity [5, 6]. However, the tap test cannot be used as an exclusionary test due to the inherent low sensitivity (25–61%). The prognostic value of this procedure for identifying patients who will benefit from shunt diversion of fluid increases as greater amounts of fluid are removed by external lumbar drainage. The highest sensitivity and specificity is associated with prolonged controlled lumbar drainage (500 cc/3 days). (50–100%) [7]. The utilization of methods to compute outflow resistance of cerebrospinal fluid (Ro) is also helpful in identifying shunt responders. The informationavailable for assessing the usefulness of Ro in INPH is limited and the two reports cited for INPH are the largest series of data currently available. Although data are scant, the reported accuracy of resistance measures may be higher than that of the CSF tap test [8]. Therefore, determination of CSF outflow resistance may be helpful in increasing prognostic accuracy for identifying SRINPH when tap test results are negative.

Identifying shunt responsive patients: summary

Step 1 – clinical evaluation

Based on the history, neurological exam, and basic neuroimaging (CT and/or MRI), the patient is categorized as Probable, Possible, or Unlikely INPH. In an otherwise healthy patient in whom the clinical diagnosis appears highly probable, it may not be unreasonable to proceed directly to treatment (the placement of a shunt) without supplemental tests keeping in mind the 50 to 61% degree of certainty.

Step 2 – supplemental testing

To avoid complication and improve the certainty of a positive shunt response beyond 50 to 61%, all Probable and Possible INPH patients should be considered for supplemental testing (CSF tap test, Infusion study, ELD).

Step 3 – tap test

Given its ease, it is reasonable to proceed initially with a CSF tap test. A positive response to a 40–50 cc tap test has a higher degree of certainty for a favorable response to shunt placement than that which can be obtained by clinical examination. However the tap test cannot be used as an exclusionary test due to its low sensitivity.

Step 4 – resistance testing

Determination of the CSF outflow resistance via an infusion test carries a higher sensitivity (57–100%) compared to the tap test but a similar positive predictive value of 75 to 92%.

Step 5 – external lumbar drainage

Prolonged external lumbar drainage in excess of 300 cc is associated with high sensitivity (50–100%) and high positive predictive value (80–100%). It is a most effective test for identifying SRINPH but requires hospital admission and carries a higher complication rate than CSF tap or Resistance studies. Of the three recommended supplemental tests, the prognostic value of the ELD is likely retained even with Possible and Unlikely INPH clinical designations.

Surgical considerations

There are no Class I studies that have addressed the question of comparing operative versus conservative management of INPH. The risk-benefit ratio must be individualized for each patient with the following issues in mind: 1) shunt-responsive INPH exists with reasonable certainty, 2) there are low surgical risks related to co-morbidities, and 3) the degree of INPH-

related morbidity warrants the shunt-related risks. The two most commonly used configurations are the VP and VA shunts. The choice of valve type and setting should be based on empirical reasoning and a basic understanding of shunt hydrodynamics. The most conservative choice is a valve incorporating an anti-siphon device (ASD) with the understanding that under-drainage (despite a low opening pressure) may occur in a small percentage of patients due to the ASD. Based on the results of retrospective studies, the use of an adjustable valve may be beneficial in the management of INPH due to the ability to non-operatively manage both under- and over-drainage problems.

Studies of outcome following shunt surgery for INPH

To date, there is no standard for outcome assessment of shunt treatment in idiopathic NPH. The variable improvement rates reported are not only due to different criteria for selection of patients but also due to different postoperative assessment procedures and follow-up intervals. Studies that have established fixed protocols for follow-up have shown that short- and long-term periods after shunting are determined by many factors. While short-term results were more likely to be influenced by shunt-associated risks, long-term results were independent upon factors inherent to the shunt procedure and shunt complications, i.e. death and morbidity related to concomitant cerebrovascular and vascular diseases. Studies have shown that beyond one year post surgery these factors definitely influence the clinical effect of shunting, making the one year post shunt period a potential determinate of the shunt-outcome.

A firm description of shunt outcome can be based on the documentation of either the clinical impairment, improvement following treatment or both. Grading of either the functional status of the idiopathic NPH patient or grading the clinical criteria of gait, incontinence and dementia should be performed. Examples of reported scales are: Black, Stein and Langfitt, Boon, Mori and Krauss [9–13]. Besides gait, improvement in cognition is also correlated with the patient's daily function. Neuropsychological testing may be of value in evaluating subtle cognitive deficits or changes with treatment. The latter have the advantage of having established norms for age and education level, however, the contribution of the various neuropsychological tests in the assessment of clinical outcome of shunt treatment remains to be elucidated. Efforts should be made to investigate how and when clinical outcome from shunt treatment is best assessed with respect to short- (3, 6 months) and long-term (1 year or greater) prognosis. The long-term prognosis may be affected by life expectancy and co-morbid factors not related to the shunt procedure.

In addition, there is a need of standardized reporting of shunt related complications and their effect on both the clinical outcome and the benefit of shunt treatment in INPH.

Conclusion

Although much has been learned regarding the diagnosis and treatment of idiopathic NPH, prospective, randomized multi-center trials are needed to resolve many of the issues regarding selection of patients for shunt surgery, type of valve configuration, value of resistance testing, effectiveness of probable, possible and unlikely diagnostic categories. These studies are necessary to elevate the guidelines from class II and III data to class I.

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