

## **Neurosurgery. Guidelines for the Diagnosis and Management of Idiopathic Normal-Pressure Hydrocephalus**

### **Development of Guidelines for Idiopathic Normal-pressure Hydrocephalus: Introduction.**

Neurosurgery. Guidelines for the Diagnosis and Management of Idiopathic Normal-Pressure Hydrocephalus. 57(3) SUPPLEMENT:S2-1-S2-3, September 2005.

Marmarou, Anthony Ph.D.; Bergsneider, Marvin M.D.; Relkin, Norman M.D., Ph.D.; Klinge, Petra M.D., Ph.D.; Black, Peter McL. M.D., Ph.D.

Abstract:

**OBJECTIVE:** There are no currently accepted evidence-based guidelines for the diagnosis and management of the normal-pressure hydrocephalus (NPH) patient. As a result, an independent study group was assembled to address this issue and determine the feasibility of developing standardized guidelines, which would be acceptable in the United States and abroad and would be based on the available scientific evidence. The guidelines were to encompass value of clinical presentation, value of supplementary diagnostic tests, surgical management, and outcome assessment.

**METHODS:** Initially, a series of 10 questions were formulated in the areas of pathophysiology, diagnosis, and treatment to obtain a consensus by panels of experts (see Acknowledgments) assembled in San Antonio, TX, in September 2000. This workshop provided significant insight into the difficulties in developing NPH guidelines, and a consensus was reached as to those questions involving expert opinion. Subsequently, evidentiary tables were developed on the basis of the available evidence. Only those studies with 20 or more idiopathic NPH (INPH) patients were included.

**RESULTS:** Sensitivity, specificity, and positive and negative predictive values for INPH diagnostic criteria were assessed, and guidelines were developed on the basis of the available evidence. Recommendations for classification of INPH and additional studies were documented.

**CONCLUSION:** The development of the guidelines was made difficult because systematic studies of INPH and patient numbers were few. It was decided to maintain the classification of NPH into two major categories, INPH and those of known cause (secondary NPH). Many studies "mixed" these classifications, and as a result, they could not be used in the evidentiary tables. Despite these problems, evidence-based

guidelines were developed, and it is hoped that they will be useful in guiding clinical management of the INPH patient.

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### **Diagnosing Idiopathic Normal-pressure Hydrocephalus.**

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Relkin, Norman M.D., Ph.D.; Marmarou, Anthony Ph.D.; Klinge, Petra M.D., Ph.D.; Bergsneider, Marvin M.D.; Black, Peter McL. M.D., Ph.D.

Abstract:

**OBJECTIVE:** The precise incidence and prevalence of idiopathic normal-pressure hydrocephalus (INPH) is not known, and evidence-based clinical diagnostic criteria have not been developed previously. This report contains evidence-based guidelines for clinical diagnosis of INPH that are intended to facilitate future epidemiological studies of INPH, promote earlier and more accurate diagnosis, and ultimately improve treatment outcome.

**METHODS:** The criteria for the diagnosis of INPH are based on evidence from the medical literature, supplemented as necessary by expert opinion. From 1966 to 2003, 653 publications on "normal-pressure hydrocephalus" were cited in MEDLINE, including 29 articles that met the more stringent criteria of including "idiopathic normal-pressure hydrocephalus" in their title. Additional studies were considered that explicitly identified INPH cases and/or specified the criteria for a diagnosis of INPH. Studies were graded according to the class of evidence and results summarized in evidentiary tables. For issues of clinical relevance that lacked substantive evidence from the medical literature, the opinions of consulting experts were considered and contributed to "Options."

**RESULTS:** Evidence-based guidelines for the clinical diagnosis of INPH have been developed. A detailed understanding of the range of clinical manifestations of this disorder and adherence to practice guidelines should improve the timely and accurate recognition of this disorder.

**CONCLUSION:** It is recommended that INPH be classified into probable, possible, and unlikely categories. We hope that these criteria will be widely applied in clinical

practice and will promote greater consistency in patient selection in future clinical investigations involving INPH.

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**The Value of Supplemental Prognostic Tests for the Preoperative Assessment of Idiopathic Normal-pressure Hydrocephalus.**

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Marmarou, Anthony Ph.D.; Bergsneider, Marvin M.D.; Klinge, Petra M.D., Ph.D.; Relkin, Norman M.D., Ph.D.; Black, Peter McL. M.D., Ph.D.

Abstract:

**OBJECTIVE:** The diagnosis and management of idiopathic normal-pressure hydrocephalus (INPH) remains unclear. Moreover, the value of supplementary tests to predict which patients would benefit from placement of a shunt has not been established. This report develops evidence-based guidelines for the use of supplementary tests as an aid in prognosis.

**METHODS:** MEDLINE searches from 1966 to the present were undertaken by use of the query NPH, normal-pressure hydrocephalus, lumbar drain, CSF [cerebrospinal fluid] tap test, and external CSF drainage in humans. This resulted in 242 articles. To provide a scientific, evidence-based review, we have chosen to restrict our analysis to clinically relevant studies usually consisting of large numbers of shunted NPH patients. Studies that did not specify INPH or secondary NPH were considered in a separate evidentiary table.

**RESULTS:** Evidence-based guidelines for use in supplementary tests have been developed. A positive response to a 40- to 50-ml tap test has a higher degree of certainty for a favorable response to shunt placement than can be obtained by clinical examination. However, the tap test cannot be used as an exclusionary test because of its low sensitivity (26-61%). Determination of the CSF outflow resistance via an infusion test carries a higher sensitivity (57-100%) compared with the tap test and a similar positive predictive value of 75 to 92%. Prolonged external lumbar drainage in excess of 300 ml is associated with high sensitivity (50-100%) and high positive predictive value (80-100%).

**CONCLUSION:** To date, a single standard for the prognostic evaluation of INPH patients is lacking. However, supplemental tests can increase predictive accuracy for prognosis to greater than 90%. Additional multicenter prospective randomized clinical trials are needed.

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### **Surgical Management of Idiopathic Normal-pressure Hydrocephalus.**

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Bergsneider, Marvin M.D.; Black, Peter McL. M.D., Ph.D.; Klinge, Petra M.D., Ph.D.; Marmarou, Anthony Ph.D.; Relkin, Norman M.D., Ph.D.

**Abstract:**

**OBJECTIVE:** To develop evidence-based guidelines for surgical management of idiopathic normal-pressure hydrocephalus (INPH). Compared with the diagnostic phase, the surgical management of INPH has received less scientific attention. The quality of much of the literature concerning the surgical management has been limited by many factors. These include retrospective analysis, small patient numbers, analysis of a mixed NPH population, and sometimes a lack of detail as to what type of shunt system was used. Many earlier studies predated our current understanding of the hydrodynamics of cerebrospinal fluid shunts, and therefore, the conclusions drawn may no longer be valid.

**METHODS:** A MEDLINE and PubMed search from 1966 to the present was conducted using the following key terms: normal-pressure hydrocephalus and idiopathic adult-onset hydrocephalus. Only English-language literature in peer-reviewed journals was reviewed. The search was further limited to articles that described the method of treatment and outcome selectively for INPH patients. Finally, only studies that included 20 or more INPH patients were considered with respect to formulating the recommendations in these Guidelines (27 articles).

**RESULTS:** For practical reasons, it is important to identify probable shunt responders diagnosed with INPH. If the patient is an acceptable candidate for anesthesia, then an INPH-specific risk-benefit analysis should be determined. In general, patients exhibiting negligible symptoms may not be suitable candidates for surgical management, given the known risks and complications associated with shunting INPH.

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 antisiphon device, with the understanding that underdrainage (despite a low opening pressure) may occur in a small percentage of patients because of the antisiphon device. On the basis of retrospective studies, the use of an adjustable valve seems to be beneficial in the management of INPH.

**CONCLUSION:** The treatment of INPH should not be considered lightly, given the seriousness of the potential complications. Within these limitations and the available evidence, guidelines for surgical management were developed.

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**Outcome of Shunting in Idiopathic Normal-pressure Hydrocephalus and the Value of Outcome Assessment in Shunted Patients.**

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Klinge, Petra M.D., Ph.D.; Marmarou, Anthony Ph.D.; Bergsneider, Marvin M.D.; Relkin, Norman M.D., Ph.D.; Black, Peter McL. M.D., Ph.D.

Abstract:

**OBJECTIVE:** To develop guidelines for assessing shunt outcome in patients with idiopathic normal-pressure hydrocephalus (INPH). To date, the literature available on this topic has been marked by disparate definitions of clinical improvement, varying postoperative follow-up protocols and periods, and substantial differences in the postoperative management. Because specific criteria for defining clinical improvement are seldom reported, conclusions drawn about shunt outcome may be subjective.

**METHODS:** A MEDLINE search back to 1966 was undertaken using the query NPH, normal-pressure hydrocephalus, shunting, shunt treatment, shunt response, outcome, and clinical outcome. The criteria for selection were studies that included INPH from 1966 to the present in which the outcome of INPH was reported in patient groups of 20 or more.

**RESULTS:** To date, there is no standard for outcome assessment of shunt treatment in INPH. The variable improvement rates reported are not only because of different criteria for selection of patients but also because of different postoperative assessment

procedures and follow-up intervals.

**CONCLUSION:** Studies that have established fixed protocols for follow-up have shown that short- and long-term periods after shunting are determined by many factors. Whereas short-term results were more likely to be influenced by shunt-associated risks, long-term results were independent of 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 1 year after surgery, these factors definitely influence the clinical effect of shunting, making the 1-year postshunt period a potential determinant of the shunt outcome. Guidelines for outcome assessment were developed on the basis of the available evidence and consensus of expert opinion.

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