

Clinical Guidelines for Idiopathic Normal Pressure Hydrocephalus

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Abstract

Idiopathic normal pressure hydrocephalus (iNPH) is a syndrome characterized by gait disturbance, dementia, and/or urinary incontinence without causative disorders, and ventricular enlargement due to disturbance of the cerebrospinal fluid (CSF) circulation. The number of patients with iNPH will increase with the aging of the population in Japan. However, iNPH is often difficult to differentiate from other senile disorders such as lumbar canal stenosis, parkinsonism, and so on. Clinical guidelines for iNPH are required to improve understanding and provide for patients' quality of life and social care. These guidelines propose three levels of iNPH: possible, probable, and definite. Possible iNPH includes one or more of the classical triad and ventricular dilation in middle aged and elderly patients with closing of the CSF space at high convexity on magnetic resonance imaging. Probable iNPH shows improvement of the symptoms after CSF removal in patients with possible iNPH. Definite iNPH shows clinical improvement after CSF shunt operation. The CSF tap test is a major diagnostic measure because of the simplicity and less invasiveness. Use of the programmable valve is recommended to decrease CSF overdrainage. These guidelines are helpful for the diagnosis and treatment of iNPH.

Introduction

Idiopathic normal pressure hydrocephalus (iNPH) is a syndrome characterized by gait disturbance, dementia, and/or urinary incontinence without causative disorders, and ventricular dilation due to disturbance of the cerebrospinal fluid (CSF) circulation. The Japanese Society of Normal Pressure Hydrocephalus began to develop clinical guidelines for iNPH based on methods of evidence-based medicine in February 2002.

Method

Research questions were made and 517 papers on diagnosis and 587 papers on therapy were collected using MEDLINE. The major effort was focused on

papers dealing with cases of iNPH. Although definite diagnosis of iNPH is made after the CSF shunt operation, preoperative diagnosis is possible and is proposed by these guidelines. A flowchart of diagnosis is provided for general physicians, neurologists, and neurosurgeons (Fig. 1).

Results

Gait disturbance is a major symptom and is characterized by a short stepped, magnetic gait with instability. Gait disturbance is seen in most patients and can be the only symptom of iNPH. Dementia is noted in about two thirds of patients and is usually mild. Urinary disturbance is seen in almost half of patients.

Critical reviews were made of the diagnostic tests

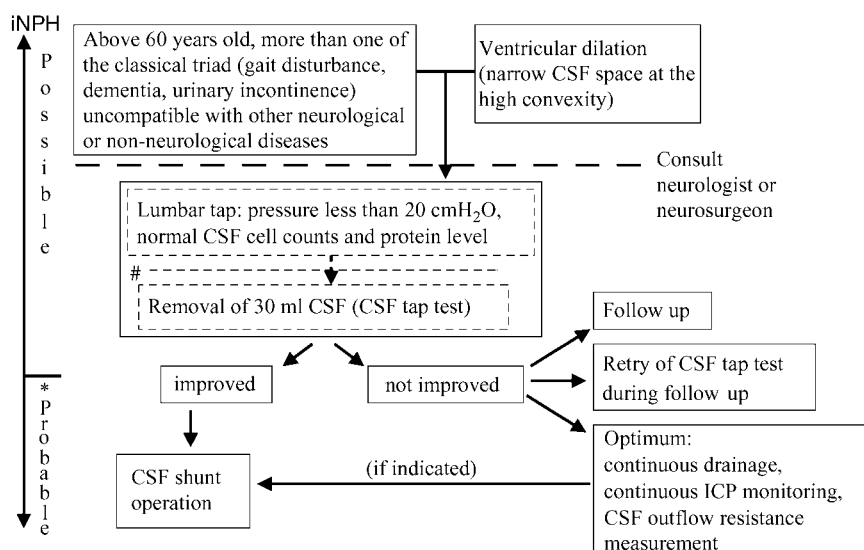


Fig. 1 Flowchart of diagnosis of idiopathic normal pressure hydrocephalus (iNPH). #Tap test can be performed if cerebrospinal fluid (CSF) is watery clear. *Probable iNPH is diagnosed if tap test or optional tests are positive.

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including computed tomography (CT), magnetic resonance (MR) imaging, CSF circulation study, CSF pressure dynamics, and so on. Mild to moderate ventricular dilation is essential but non-specific. Periventricular change on CT and MR imaging is non-contributory. Closing CSF space at the high convexity in contrast to the basal cistern and/or sylvian fissure on coronal sections of MR imaging is an indicative finding for iNPH.²⁾ CT/radioisotope cisternography and cerebral blood flow study may not be confirmatory diagnostic tests. CSF removal via lumbar puncture with assessment of gait disturbance (CSF tap test) is highly sensitive and less invasive, but has low specificity.^{1,3)} Continuous CSF drainage shows almost the same sensitivity and higher specificity,³⁾ but is more invasive. Measurement of CSF outflow resistance has high sensitivity and high specificity,¹⁾ but this is not popular in Japan. Detection of monoamines and peptides in the CSF is non-diagnostic.

Discussion

These guidelines propose three levels of diagnosis: possible, probable, and definite iNPH. The diagnosis of "possible" iNPH is based on the presence of one or more classical symptoms, ventricular dilation with closing sulci at the high convexity, and clear CSF with normal CSF pressure in middle aged and elderly patients. The diagnosis of "probable" iNPH is based on improved gait after the CSF tap test or continuous CSF drainage. The diagnosis of "definite" iNPH is based on improvement of symptoms after the CSF shunt operation.

Surgical indications should be determined by the clinical symptoms, findings on MR imaging, and CSF tap test. Attention should be paid to the

patients' general condition and social background. CSF shunt operation is a single therapeutic measure. The recent development of the programmable valve provides higher effectiveness with external pressure adjustment. Gait disturbance is the most common symptom (more than 80% to 90% of patients), followed by dementia and urinary disturbance. The latter symptoms usually show delayed improvement in contrast to gait disturbance. Long-term follow up ranges from 3 to 5 years, and reveals that clinical improvement is persistent in about two thirds of patients.

The population of elderly people is increasing in Japan, and nursing care for elderly patients becomes more and more important. The present guidelines are helpful for the diagnosis and treatment of iNPH.

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