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Review of normal-pressure hydrocephalus

Dr. Jamelah Ahmed Alghalbi[1], Dr. Norah Aljalal[2], Dr. Hussain Alkuly Alghamdi[3], Dr. Nedaa Bantan[4], Dr. Peter Bernad[5]

[1] Consultant Neurology/Stroke, King Abdullah Medical Complex, Jeddah, Saudi Arabia
[2] Consultant Neurology, Dammam Hospital Complex, Dammam, Saudi Arabia
[3] Specialist, King Abdullah Medical Complex, Jeddah, Saudi Arabia
[4] Pediatric, King Fahad Forced Armed Hospital, Jeddah, Saudi Arabia
[5] Neurology consultant, George Washington university hospital, United States

Abstract
The clinical triad of normal pressure hydrocephalus (NPH), a condition that affects the elderly and is identified by ventriculomegaly and deep white matter ischemia (DWMI) on magnetic resonance imaging, includes gait disturbance, dementia, and urinary incontinence (MRI). The differential diagnosis of neurological diseases must include it as a key factor. The frequency of dementia is increasing in line with the rise in life expectancy worldwide. The pathophysiology of NPH is still a topic of debate. Although guidelines and recommendations for best practices are well established, there is an ongoing debate on the use of various imaging modalities to pinpoint specific symptoms of the illness and predict how effective CSF shunting will be. Despite the development of diagnostic methods, treatment approaches, and more than 50 years of study, NPH remains a problem for experts. Although we should differentiate between sNPH and iNPH based on the result as well as the clinical, pathophysiological, and epidemiological aspects, they shouldn't be seen as separate entities. The patient's medical history, neurological examination, and brain imaging are still used to make the diagnosis in both kinds of NPH. Ventriculoperitoneal and ventriculoatrial shunts are two of the most common CSF shunt procedures.

Keywords: hydrocephalus (NPH), normal-pressure, (MRI)

Introduction

Hakim and Adams first identified the syndrome of gait apraxia, dementia, and incontinence as normal pressure hydrocephalus (NPH) in 1965 along with dilated ventricles and normal CSF pressures [1]. The majority of those affected by this condition are elderly, and some of them may exhibit symptoms of both NPH and Alzheimer's or small-vessel disease. The diagnostic difficulty in managing each case is weighing the risks of surgery in this age group against the potential advantages of treating the component that could be resolved by CSF diversion [8].

NPH is a syndrome that may be reversible, it is clinically characterized by enlarged cerebral ventricles (ventriculomegaly), cognitive decline, gait apraxia, and urinary incontinence [1]. Patients who may benefit from CSF diversion can be identified through neuropsychological profiling and CSF infusion studies; however, shunt responsive NPH patients typically, but not always, have higher resistance to CSF absorption [1].

It is a crucial component of the differential diagnosis of neurological illnesses. In keeping with the global increase in life expectancy, dementia prevalence is rising [4]. There is still controversy surrounding the pathophysiology of NPH. Although standards and suggestions for best practices are well established, there is still disagreement over the use of distinct imaging modalities in identifying particular aspects of the illness and forecasting the effectiveness of CSF shunting [8]. NPH remains a challenge for the professionals despite the development of diagnostic techniques and therapeutic strategies and more than 50 years of research [4].

Definition

Gait disturbance, dementia, and urine incontinence are the clinical triad of normal pressure hydrocephalus (NPH), a disease that affects the elderly and is defined by ventriculomegaly and deep white matter ischemia (DWMI) on magnetic resonance imaging (MRI). According to estimates, NPH can cause up to 10% of dementia cases. It is important because ventriculoperitoneal shunting can be used to cure NPH. Those with meningitis or hemorrhage, which are established causes of chronic communicating hydrocephalus, typically respond faster than patients with the so-called "idiopathic" variety, most likely as a result of poor selection criteria in the past. Hyperdynamic cerebrospinal fluid (CSF) flow through the aqueduct has been linked to positive shunt response [4].

MRI and a coronal head scan (left) (right) In the left image, the medial cisterns (red circle) and tight convexity (red circle) over the convexity towards the vertex are narrowed at the level of the posterior commissure; these are typical findings of NPH. On the right image, however, the medial cisterns (green arrow) and the CSF gaps above the convexity towards the vertex (red arrow) are expanded, a finding consistent with brain atrophy. The callosal angle is indicated by the blue lines in both photos; a callosal angle less than 90o is indicative of NPH (left image), whereas a callosal angle larger than 90o is indicative of brain atrophy (right image). Periventricular signal changes are indicated by the blue arrows. These changes are unilateral in incidence (right image), which indicates they are most likely the result of vascular encephalopathy. The anomalies visible in the left image might be transependymal CSF diapedesis as a result of NPH [6].

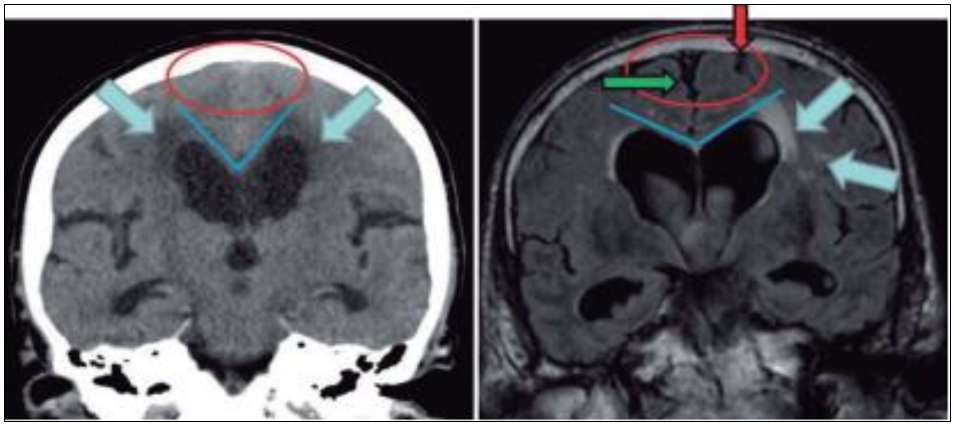

Fig 1

Epidemiology

NPH is a rare condition, according to a number of recent epidemiologic research, yet having large ventricles is a common sign of aging. NPH may have multiple causes, such as vascular disease, congenital conditions, and poor CSF absorption. The expanded ventricular diameter and CSF fluid collection outside of the ventricles, which are not the result of atrophy, are MRI characteristics of NPH. A "tight high convexity" and enlargement of the CSF gaps in the sylvian fissure are predictive MRI characteristics in NPH that have been referred to as disproportionately increased subarachnoid space hydrocephalus (DESH) [2].

Idiopathic Normal Pressure Hydrocephalus (iNPH)

The most typical type of hydrocephalus in adults is idiopathic normal pressure hydrocephalus (iNPH). Patients experience a condition that includes dilated cerebral ventricles along with deteriorated walking, cognition, and bladder control (urgency and incontinence). A CSF shunt, which is often placed between the lateral ventricle and the abdomen's ventriculoperitoneal [VP] shunt, is the only successful treatment for iNPH [7].

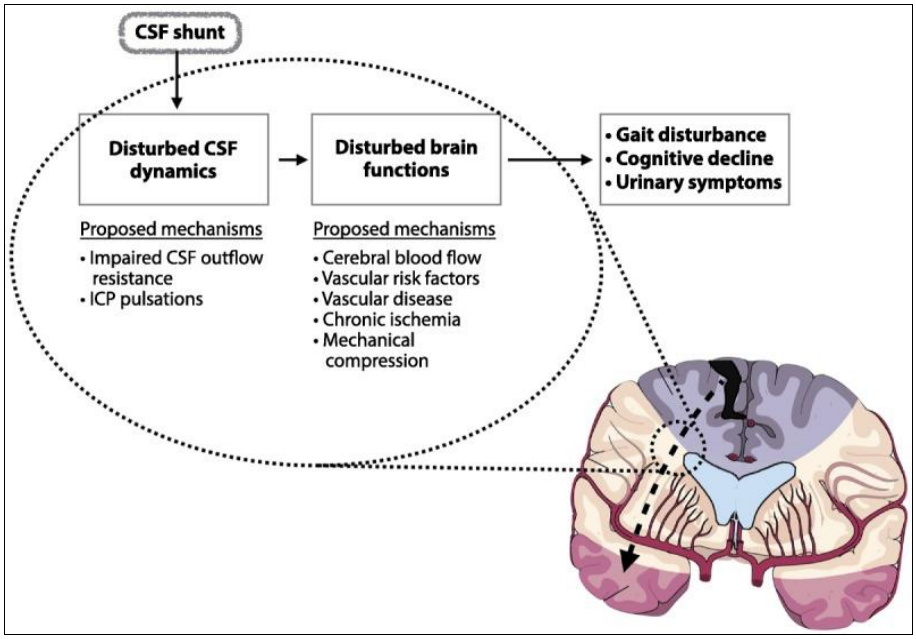

Fig 2

The idea of iNPH has frequently come up in conversation. One woman (age 63) had suspected meningeal carcinomatosis, one guy (age 62) had a IIIrd ventricle cyst, and only two of the six initial patients (males, ages 16 and 43) had idiopathic NPH (male, age 52 and woman, age 63). They all presented with frontal dementia along with a gait impairment resembling Bruns apraxia or frontal ataxia, as well as frontal-type urine and fecal incontinence, along with symptoms of brain edema and brain damage, including akinetic mutism [3]. Neurosurgical treatment for all of these symptoms, such as ventriculoatrial shunting or the Torkildsen technique, had a strikingly positive effect (ventriculo-cisternostomy). Upon spinal tap, all patients exhibited normal opening CSF pressure, and the majority of them became better when CSF was drained. Pneumo-encephalograms showed a symmetrical and huge expansion of the entire ventricular system, including the aqueduct and IVth ventricle, without air in the subarachnoid space in all of the original cases. This finding revealed that the hydrocephalus was communicating [1].

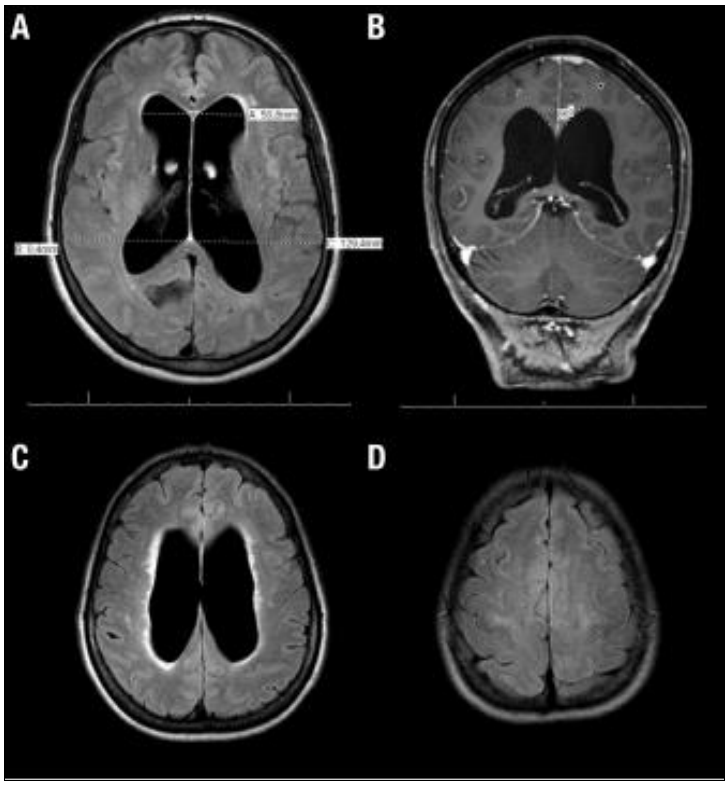

Fig 3

Neuroimaging in NPH

a. The ratio of the greatest width of the frontal horns of the lateral ventricles and the maximal internal diameter of the skull at the same level on axial CT or MRI images demonstrates a considerable ventriculomegaly with increased Evans Index. Evans index is 0.39 in this instance (abnormality > 0.3).
b. Reduced callosal angle on a T1-weighted coronal gadolinium-enhanced MRI scan.
c. An axial FLAIR MRI scan showing trans-ependymal edema is likely the cause of the dilated lateral ventricles and strong signal in the neighboring white matter.
d. Axial FLAIR MRI of the frontoparietal areas demonstrating shrinking of the sulci and subarachnoid spaces along the high convexity and midline surface.

Differential diagnosis of NPH

The following criteria are used to determine whether someone has NPH: a history of gait disturbance, progressive mental decline, and urinary urgency or incontinence; hydrocephalus, which is indicated by an Evans' ratio on computed tomography (CT) or magnetic resonance imaging (MR) imaging that is greater than 0.30; and a CSF opening pressure (properly measured) of less than 24 cm of water [6].

International Guidelines recommend using the essential imaging findings listed below to diagnose NPH [1]:

1. Ventricular hypertrophy with an Evans's index greater than 0.
2. There are no macroscopic CSF flow obstructions.
3. At least one of these auxiliary characteristics:

a. Callosal angle of 40° or more.
b. Enlarged temporal horns of the lateral ventricles that are not totally caused by hippocampal atrophy.
c. Modified brain water content causes periventricular signal abnormalities on CT and MRI that aren't wholly attributable to demyelination or microvascular chemical changes.
d. An MRI showing a flow void in the fourth ventricle or Sylvian aqueduct.

e. When the clinical signs of the trio are abnormal or lacking, or when they are imitated by other disorders, differential diagnostic challenges may occur. In fact, a variety of other conditions, including vascular dementia, corticobasal degeneration, parkinsonism, Lewy body disease, progressive supranuclear palsy, neurosyphilis, multiple system atrophy, and drug side effects, may result in the full triad. These conditions may also occur in conjunction with other illnesses, especially cerebrovascular and Alzheimer's disease, which can be present in up to 75% of INPH patients [6]

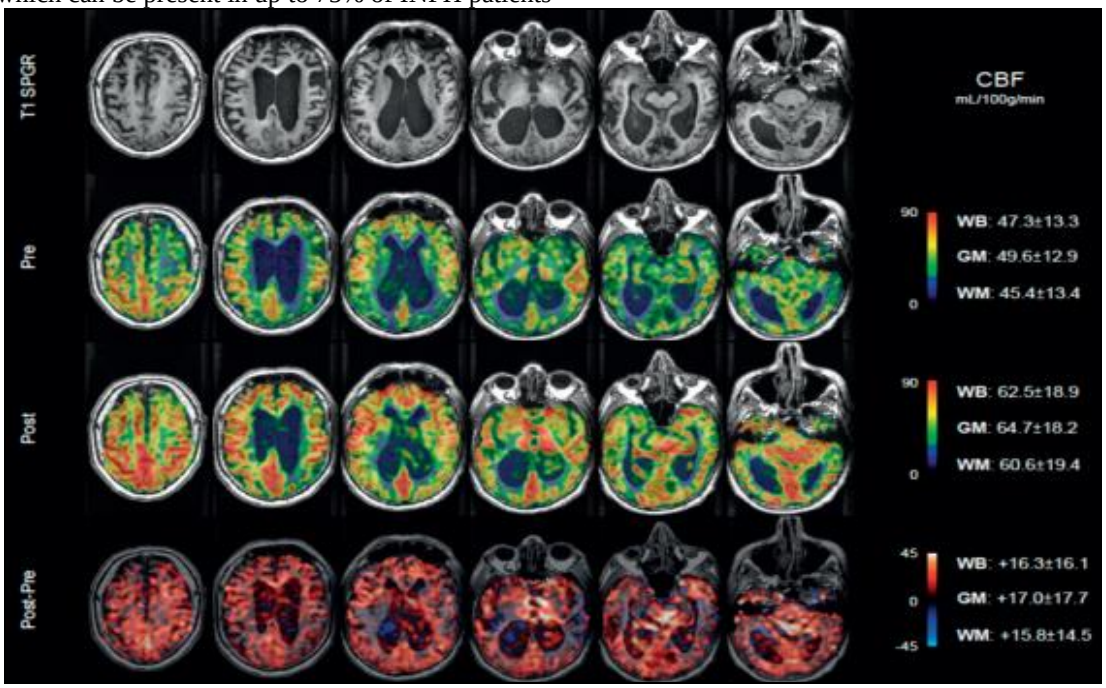

Fig 4

An illustration of an ASL-MRI showing a relationship between improved CBF and clinical recovery following a significant spinal tap (unpublished data).

Alzheimer's disease

Increased ventricles, a compromised blood-brain barrier, and decreased CSF clearance are likely to blame for the neuronal degeneration, which results in the buildup of neurotoxins including beta-amyloid and tau protein. The Alzheimer-like alterations in the brain of iNPH patients and rats with persistent hydrocephalus can be explained by this compromised turnover [1]. AD and the NPH syndrome can have quite different clinical presentations, despite the fact that both are significant disorders in the differential diagnosis [7].

The existence of "cortical" symptoms such hippocampal forgetfulness, agnosia, apraxia, and aphasia distinguishes AD from other conditions including inattention, apathy, memory loss, and psychomotor slowness. Additionally, motor and urinary complaints frequently come before and take a back seat to AD dementia [6].

However, 4 months after VPS, 32% of patients who had moderate-to-severe AD pathology exhibited less postoperative improvement in NPH symptoms and lower baseline cognitive test scores. 35 The NPH triad symptoms may be seen in advanced stages of AD due to the co-occurrence of NPH and AD. Conversely, overlapping AD features are present in roughly 75% of seriously demented NPH patients [2].

Secondary normal pressure hydrocephalus (sNPH)

Subarachnoid hemorrhage (SAH), which affected 46.5% of patients, head trauma, 29%, intracranial malignancies, 6.2%, meningoencephalitis, 5%, and cerebrovascular illness, which affected 4.5% of patients, were the leading causes of sNPH. The insertion of a ventriculoperitoneal shunt was used to treat sNPH in 71.9% of patients, while a ventriculoatrial shunt was used in 24.4% of patients [3]. Elevated Evans' ratio was the most frequent radiological sign prior to the onset of symptoms, and sNPH includes all patients in whom an etiology is determined [1].

It has become challenging to provide useful guidelines for maximizing the treatment and diagnosis of the condition because sNPH might have a variety of causes [5]. On the other hand, CSF viscosity is increased by proteins and other substances in brain tumors and inflammatory responses, especially neuro-cysticercosis in tropical regions. As a result, CSF reabsorption through arachnoid granulation is compromised, which results in NPH [1]. Although, only 0.51% of those with secondary normal pressure hydrocephalus under the age of 61 exhibited symptoms, after 4 to 8 years 25% of the asymptomatic participants developed symptoms that were consistent with secondary normal pressure hydrocephalus [2].

Conclusions

Based on the outcome as well as the clinical, pathophysiological, and epidemiological traits, we should distinguish between sNPH and iNPH, however they shouldn't be treated as independent entities. In both types of NPH, the diagnosis is still made on the basis of the patient's medical history, neurological examination, and brain imaging. CSF shunt surgeries including ventriculoperitoneal and ventriculoatrial shunts are the major kinds of treatment.

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