Diagnosis And Management Of Normal Pressure Hydrocephalus

An Essay
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Contents:

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Abbreviations

ADC: *Apparent diffusion coefficient*
ASD: *Antisiphon device*
CBF: *Cerebral blood flow*
Cout: *Conductance outflow*
CSF: *Cerebrospinal fluid*
CT-Scan: *Computerized tomography scan*
DWMI: *Deep white matter ischemia*
ELD: *External lumbar drainage*
FR: *Flow rate*
INPH: *Idiopathic normal pressure hydrocephalus*
IP: *Intracranial pressure*
LP: *Lumbar puncture*
MRI: *Magnetic resonance imaging*
OEF: *Oxygen extracted fraction*
OP: *opening pressure*
pDfr: *Peak diastolic flow rate*
PET: *Position emission tomography*
PSfr: *Peak systolic flow rate*
SAH: *Subarachnoid hemorrhage*
VA: *Ventriculo-atrial*
VP: *Ventriculo-peritoneal*
VPS: *Ventriculo-peritoneal shunt*

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Introduction

Normal pressure hydrocephalus (NPH) is a chronic type of communicating hydrocephalus whereby the increase in intracranial pressure (ICP) due to accumulation of cerebrospinal fluid (CSF) becomes stable and that the formation of CSF equilibrates with absorption. The ICP gradually falls but still maintains a slightly elevated level and the CSF pressure reaches a high normal level of 150 to 200 mmH_2O. **(Levine, 1999)**

Measurements of CSF, therefore, are not usually elevated. Because of this equilibration, patients do not exhibit the classic signs of increased intracranial pressure such as headache, nausea, vomiting, or altered consciousness. However, patients do exhibit the classic triad of gait difficulties, urinary incontinence, and mental decline as first described by Hakim and Adams in 1965. **(Finney, 2009)**

It is often misdiagnosed as Parkinson's disease, Alzheimer's disease, and senility due to its chronic nature and its' presenting symptoms. Although the exact mechanism is unknown, normal-pressure hydrocephalus is thought to be a form of communicating hydrocephalus with impaired CSF reabsorption at the arachnoid villi. **(Silverberg et al, 2006)**

The clinical manifestations of NPH may exhibit the classic triad (also known as Adam's triad) of urinary incontinence, gait disturbance, and dementia. **(Factora, 2006)**

Gait disturbance and apraxia is the first symptom of the triad and may be progressive, due to expansion of the ventricular system, particularly at the level of the lateral ventricles, leading to traction on the lumbosacral motor fibers that run in this region. Often, this takes on the form of unsteadiness and impaired balance, especially on stairs and curbs. **(Bugalho and Guimaraes, 2007)**

Weakness and tiredness may also be part of the complaint, although this is very vague. NPH gait disturbance is often characterized as a "magnetic gait," in which feet appear to be stuck to the walking surface until wrested upward and forward at each step. The gait may mimic a Parkinsonian gait, with short shuffling steps and stooped, forward-leaning posture, but there is no rigidity or tremor. A broad-based gait may be employed by the patient in order to compensate for the ataxia. **(Factora and Luciano, 2008)**

Dementia is predominantly frontal lobe in nature, with apathy, dullness in thinking, and slight inattention. Memory problems are usually the main problem, which can lead to the misdiagnosis of Alzheimer's disease. **(Byrd, 2006)**

Urinary incontinence appears late in the illness, consisting of increased frequency and urgency.

Diagnosis of NPH is usually first led by a Lumbar puncture, followed by the evaluation of clinical response to removal of CSF. This can be followed by a CT, MRI, CSF flowmetry and continuous external lumbar CSF drainage during 3 or 4 days. **(Marmarou et al, 2005)**

Lumbar puncture is usually the first step in diagnosis and the opening pressure measured carefully. In most cases, CSF pressure is usually above 155 mmH_2O. Clinical improvement after removal of CSF (30 ml or more) has a high predictive value for subsequent success with shunting. This is called the "lumbar tap test" or "Fisher test". A "negative" test has a very low predictive accuracy, as many patients may improve after a shunt in spite of lack of improvement after CSF removal. **(Lim et al, 2007)**

Infusion test is the test with higher sensitivity and specificity than lumbar puncture. The outflow conductance (Cout) of the cerebrospinal fluid (CSF) system is a parameter considered to be predictive in selection for hydrocephalus surgery. Cout can be determined through an infusion test. **(Brean and Eide, 2008)**

CT scan may show enlarged ventricles without convolutional atrophy.

MRI may show some degree of transependymal migration of CSF surrounding the ventricles on T2/FLAIR sequence. **(Chatzidakis et al, 2008)**

CSF flowmetry, the measurement of CSF flow in the aqueduct has been a focus of interest since the development of MR imaging (MRI) techniques for this purpose in diagnosing idiopathic normal-pressure hydrocephalus (iNPH). **(Al Zain et al, 2007)**

Patients with a known cause of NPH are more likely to have improvement from ventriculoperitoneal shunting than those who lake a defined etiology. Signs or symptoms of short duration are more likely to be ameliorated with CSF diversion than longstanding impairment. Patients whose symptoms improve following a large volume lumber puncture have a greater chance of responding to shunting than patients whose deficits remain unchanged following removal of CSF. **(Chang et al, 2006)**

A patient presenting primarily with gait instability, followed by incontinence and mild dementia of short duration, with imaging showing ventricular dilatation but a preserved cortical mantle, has a reasonable likelihood of benefiting from ventriculoperitoneal shunting. A floridly demented patient whose ineffective ambulation and incontinence likely are secondary to cognitive decline accompanied by radiography demonstrating significant, generalized cortical atrophy will not likely respond to CSF diversion. **(Hashimoto MA, 2008)**

The decision to shunt should not be taken lightly. Accurate patient selection is the key to a successful outcome. If symptoms involving gait, incontinence, and/or dementia are present for a short period of time, patients have a higher chance of amelioration of these symptoms with shunting than patients who have had symptoms for a long time. However, improvement has been recognized in patients even after years of cognitive decline. Therefore, treatment should not be withheld, if otherwise indicated, solely on the basis of symptom duration. Yet, with NPH, no strict algorithm or definitive test provides an ironclad answer. In the absence of absolute proof remain practical indications and clinical likeliness to formulate decisions. **(Marmarou A et al, 2005)**

Aim of the work

The aim of this work is to study how to diagnose and manage the cases of normal pressure hydrocephalus. In addition to review the factors that affect the prognosis of such cases to avoid the misdiagnosis and mismanagement.

<u>Anatomy of the Ventricular System</u>

The cerebral ventricular system consists of a series of interconnecting spaces and channels within the brain which are derived from the central lumen of the embryonic neural tube and the cerebral vesicles to which it gives rise. Within each cerebral hemisphere lies a large C-shaped lateral ventricle. Near its rostral end the lateral ventricle communicates through the interventricular foramen (foramen of Monro) with the third ventricle, which is a midline, slit like cavity lying between the right and left halves of the thalamus and hypothalamus. Caudally, the third ventricle is continuous with the cerebral aqueduct, a narrow tube that passes the length of the midbrain, and which is continuous in turn with the forth ventricle, a wide tent-shape cavity lying between the brain stem and the cerebellum. Caudally, the fourth ventricle is continuous with the vestigial central canal of the spinal cord. **(Gray's Anatomy, 2005)**

<u>Neural Relationships:</u>

Each lateral ventricle is a C-shaped cavity that warps around the thalamus and is situated deep within the cerebrum. Each lateral ventricle has five parts: the frontal, temporal, and the occipital horns, the body, and the atrium. Each of these five parts has medial and lateral walls, a roof, and a floor. In addition, the frontal and temporal horns and the atrium have anterior walls. These walls are formed predominantly by the thalamus, septum pellucidum, deep cerebral white matter, corpus callosum, and two C-shaped structures, the caudate nucleus and the fornix, that warp around the thalamus. **(Rhoton, 2002)**

The thalamus is located in the center of the lateral ventricle. The body of the lateral ventricle is above the thalamus, the atrium and occipital horn are posterior of the thalamus (Fig A and B). The superior surface of the thalamus forms the floor of the body, the posterior surface of the pulvinar of the thalamus forms the anterior wall of the atrium, and the inferior surface of the thalamus is situated at the medial edge of the roof of the temporal horn**. (Rhoton, 2002)**

The caudate nucleus is an arched C-shaped, cellular mass that wraps around the thalamus and constitutes an important part of the wall of the lateral ventricle. It has a head, body, and tail. The head bulges into the lateral wall of the frontal horn and body of the lateral ventricle. The body forms part of the lateral wall of the atrium, and the tail extends from the atrium into the roof of the temporal horn and is continuous with the amygdaloid nucleus near the anterior tip of the temporal horn. In the body of the lateral ventricle, the caudate nucleus is superolateral to the thalamus; in the atrium, it is posterolateral to the thalamus; and in the temporal horn, it is inferolateral to the thalamus (Fig A and B). The stria terminalis, a fiber tract that runs parallel and deep to the thalamostriate vein, arises in the amygdaliod nucleus and courses along the border between the caudate nucleus and the thalamus in the wall of the ventricle from the temporal horn to the body. **(Rhoton, 2002)**

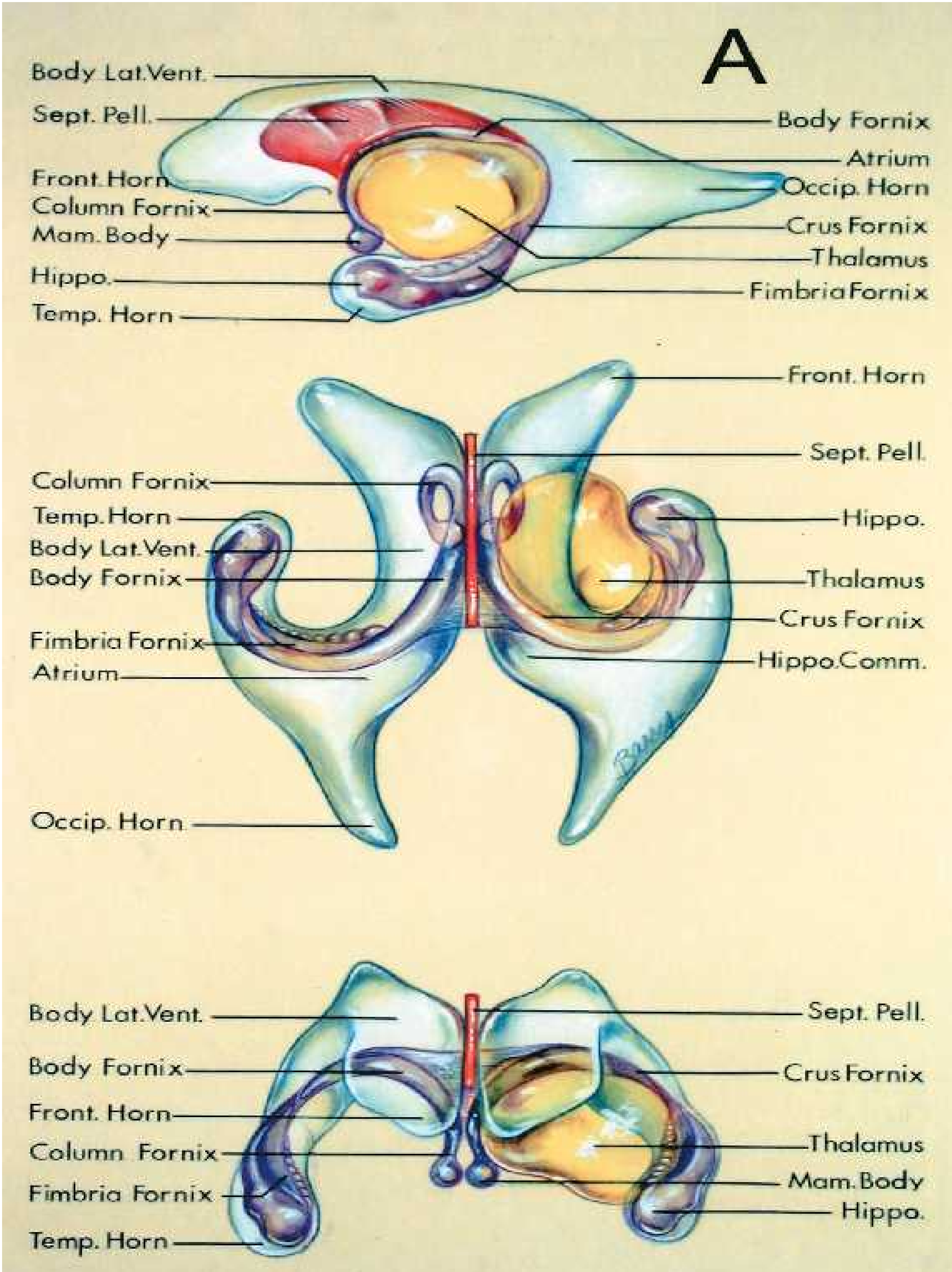
The fornix is another C-shaped structure that wraps around the thalamus in the wall of the ventricle. The fornix consists mainly of hippocampomammillary tract fibers that originate from the hippocampus, subiculum, and dental gyrus of the temporal lobe. The fimbria arises in the floor of the temporal horn on the ventricular surface of the hippocampal formation and passes posteriorly to become the crus of the fornix. The crus wrap around the posterior surface of the pulvinar of the thalamus and arches superomedially towards the lower surface of the splenium of the corpus callosum. At the junction of the atrium and the body of the lateral ventricle, the paired crura meet to form the body of the fornix, which runs forward along the superomedial border of the thalami in the medial wall of the body of the lateral ventricle. The body of the fornix separates the roof of the third ventricle from the floor of the bodies of the lateral ventricles. At the anterior margin of the thalamus, the body of the fornix separates into two columns that arch along the superior and anterior margins of the foramen of Monro in their course towards the mammillary bodies. In the area below the splenium, a thin sheet of fibers interconnects the medial margins of the crura to form the hippocampal commissure. In the body of the lateral ventricle, the body of the fornix is in the lower part of the medial wall; in the atrium, the crus of the fornix is in the medial part of the anterior wall; and in the temporal horn, the fimbria of the fornix is in the medial part of the floor (Fig B). **(Rhoton, 2002)**

The corpus callosum, which forms the largest part of the ventricular walls, contributes to the wall of each of the five parts of the lateral ventricle. The corpus callosum has two anterior parts, the rostrum and genu, a central part, the body, and a posterior part, the splenium. The rostrum is situated below and forms the floor of the frontal horn. The genu has a large bundle of fibers, the forceps minor, that forms the anterior wall of the frontal horn as it sweeps obliquely forwards and lateral to connect the frontal lobes. The genu and the body of the corpus callosum form the roof of both the frontal horn and the body of the lateral ventricle. The splenium contains a large fiber tract, the forceps major that forms a prominence, called the bulb, in the upper part of the medial wall of the atrium and occipital horn as it sweeps posteriorly to connect the occipital lobes. Another fiber tract, the tapetum, which arises in the posterior part of the body and splenium of the corpus callosum, sweeps laterally and inferiorly to form the roof and lateral wall of the atrium and the temporal and occipital horns. The tapetum separates the fibers of the optic radiations from the temporal horn. **(Rhoton, 2002)**

The septum pellucidum which is composed of paired laminae, separates the frontal horns and bodies of the lateral ventricles in the midline. In the frontal horn, the septum pellucidum is attached to the rostrum of the corpus callosum below, the genu anteriorly, and the body above. In the body of the lateral ventricle, the septum is attached to the body of the corpus callosum above and the body of the fornix below (Fig A). The septum pellucidum is tallest anteriorly and shortest posteriorly, disappearing near the junction of the body and crura of the fornix where the crura and hippocampal commissure fuse with the lower surface of the corpus callosum. The anterior-posterior length of the septum pellucidum varies from 28 to 50 mm. there may be a cavity, the cavum septum pellucidum, in the midline between the laminae of the septum pellucidum. **(Rhoton, 2002)**

The lateral Ventricles:

The lateral ventricle is customarily divided into a body and anterior, posterior and inferior horns (Fig. A and B). the anterior (frontal) horn lies within the frontal lobe. It is bounded anteriorly by the posterior aspect of the genu and rostrum of the corpus callosum, and its roof is formed by the anterior part of the body of the corpus callosum. The anterior horns of the two ventricles are separated by the septum pellucium. The coronal profile of the anterior horn is roughly that of a flattened triangle in which the rounded head of the caudate nucleus forms the lateral wall and floor. The anterior horn extends back as far as the interventricular foramen. **(Gray's Anatomy, 2005)**



Relations of the lateral ventricles (lateral, anterior and inferior view)

(Rhoton, 2002)