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Idiopathic normal pressure hydrocephalus. A brief review

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ABSTRACT
Idiopathic normal pressure hydrocephalus (INPH) is a neurodegenerative disease which affects the elderly, with a significant prevalence in the general population (0,2% - 5,9%), thus a common pathology encountered by neurologists and neurosurgeons, alike. Although the widespread availability of modern imaging techniques has facilitated the diagnosis of this disorder, the clinical manifestations can often be misleading. Also, an overlap with other degenerative or psychiatric diseases can make the differential diagnosis even more challenging. Cerebrospinal fluid (CSF) diversion procedures are the first line of treatment for INPH. Nowadays, there are several shunting options available, including: ventriculoperitoneal (the most commonly used), ventriculoatrial, ventriculopleural, ventriculosternal, lumboperitoneal, endoscopic third ventriculostomy. Choosing a procedure tailored to the individual patient is essential for therapeutic success. Although they are generally straightforward surgical interventions, they associate a high rate of failure, regardless of procedure used, which emphasizes the need for regular clinical and imagistic follow-up. Thus, INPH remains a disease where there is significant room for improvement, both in diagnosis and treatment.

Keywords
idiopathic normal pressure, hydrocephalus, CSF shunting

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DEFINITIONS
Hydrocephalus is produced by an excessive accumulation of cerebrospinal fluid (CSF) at the level of the ventricular system that determines a specific clinical picture (depending on the patient's age) and enlargement of ventricles on imaging studies.

Normal pressure hydrocephalus, also known as chronic adult hydrocephalus, is clinically characterized by the classic triad: gait, urinary and cognitive dysfunction, associated with ventriculomegaly.

Normal pressure hydrocephalus can be idiopathic or secondary.

Idiopathic normal pressure hydrocephalus (INPH) is a neurodegenerative disease which affects the elderly, representing a treatable cause of dementia with a prevalence of 0,2% up to 5,9%1,2.

CLINICAL PICTURE
In 1965, Hakim and Adams described the clinical picture of this disease, characterized by walking, urinary and cognitive dysfunction (3).

The patients present an ataxic gait, bradykinetic, with small steps and low cadence, as well as balance disorders. They have difficulty in ascending or descending stairs, in getting up from a sitting position or turning around. In advanced stages, patients become hypertonic, bradykinetic.

Urinary disfunctions generally manifest as incontinence. In the early stages of the disease, the urinary symptoms consist of urgency in emptying the bladder, but subsequently it is replaced by urinary incontinence, more frequently than fecal incontinence. In advanced stages, patients become indifferent to their urinary disorders.

Cognitive disorders consist of memory impairment, slow ideation, attention deficits, apathy, depression and aggressiveness.

It is important to obtain a correct diagnosis because other diseases present a similar clinical picture. INPH symptoms are also encountered in other disorders that must be included in the differential diagnosis, such as: Parkinson's disease, Alzheimer's disease and vascular dementia. Similar cognitive impairment can be encountered in schizophrenic patients and the two conditions can also coexist. In Vanhala's opinion, INPH is three times more frequent in this category of patients4.A list of other disorders that must be differentiated from INPH is presented by Bech- Azeddine5.

According to Israelsson et al, the prevalence of depression is four times higher in patients with INPH compared to the elder population without INPH2.

Imagistic diagnosis

Modern imagistic methods reveal ventriculomegaly. The size and span of ventricles can be appreciated by calculating certain indices:

- Evans index (bifrontal index) is the distance between the lateral walls of the frontal horns of the ventricle divided by the internal diameter of the skull at the same level. In INPH, the value of this index is ≥ 3
- Bicaudate index is equal to the ratio between a line drawn between the two ends of the caudate nuclei and the internal diameter of the skull at the same level
- Fronto-occipital horn index represents the ratio between the sum of the distances between the lateral walls of the frontal horns and occipital horns of the ventricles respectively and twice the largest internal diameter of the calvaria in the horizontal plane

CSF physiology and INPH physiopathology

Briefly, it is known that CSF is produced at the level of the choroid plexus located in the ventricular system with a rate of 0,3 - 0,5 ml/min, representing approximately 500 ml every 24 hours.

CSF travels from the lateral ventricles to the fourth ventricle through the aqueduct of Sylvius and subsequently, through the Magendie foramen reaches the subarachnoid spinal space and cerebral convexities.

CSF is reabsorbed at the level of Pacchioni's granulations located parasagitally on the cerebral convexities and afterwards reaches the superior sagittal sinus, entering the blood stream.

In the case of INPH, one hypothesis suggests that enlargement of ventricles compresses or deforms the axons of the central motor neurons that pass through the medial portion of the corona radiata.

Parkinson's disease type symptoms of INPH patients are caused by the dysfunction of the nigrostriatal dopaminergic pathways caused by abnormal CSF pulsations which affect the substantia nigra and determine the disorder of motor planning.6

The cognitive disorders are produced by involvement of the frontostriatal system, of the projection fibers which pass in the proximity of the lateral ventricles.

Treatment

The majority of INPH patients benefit from surgical treatment of CSF diversion.

Preoperatively, there must be a careful selection of patients that respond to this treatment. In order to do so, there are multiple methods to evaluate potential responsiveness and tests which evaluate the degree of clinical improvement:

1) CSF pressure: is measured during a lumbar puncture, with the patient positioned in left lateral decubitus, with the help of a manometer. In healthy individuals, the opening pressure is 122 mmH2O, while in INPH patients, the pressure varies between 60 – 240 mmH2O, with a mean of 150 mmH2O. This pressure is not constant, a value higher than 245 mm mmH2O indicating secondary noncommunication hydrocephalus, not INPH1,7.

2) Alleviation of symptoms after repeated

lumbar punctures with removal of 40 ml CSF is a common method for estimating responsiveness, but nonetheless a subjective one.

3) External CSF drainage for 72 hours, which determines clinical improvement. It is carried out by inserting an intrathecal catheter in the lumbar area and drainage of 10 ml CSF every hour for 72 hours8.

4) Postoperative clinical improvement can be evaluated by comparing results of several tests:

- Timed Up and Go (TUG)
- Timed Up and Go Cognition (TUG-C)
- Berg Balance Scale (BBS)
- Performance Oriented Mobility Assessment (Tinetti)
- Mini Mental Status Examination (MMSE)
- NPH Japanese Scale
- Minimal Clinically Important difference (MCID)
- Geriatric Depression Scale 15

Surgical treatment is not risk free. Therefore, a risk-benefit evaluation must be carried out for each patient. Patients under anticoagulant treatment associate a high risk for intracranial haemorrhage. Those with cerebrovascular disease have a low rate of response to CSF drainage.

Types of CSF drainage

- Ventriculoperitoneal (VP) shunt is the most frequently used.
  There are however other shunt options, each with its advantages and disadvantages:
- Ventriculoatrial (VA)
- Ventriculopleural
- Ventriculosternal
- Lumboperitoneal (LP)
- Endoscopic third ventriculostomy (ETV)

Ventriculoperitoneal shunting is not recommended for patients with a history of multiple abdominal surgery or peritonitis, due to the presence of adhesions and low CSF absorption. Also, VP shunting is difficult to perform in obese patients.

Ventriculoatrial shunting can be associated with chronic infections, which if undiagnosed, can lead to shunt nephritis.

Lumboperitoneal shunt has a similar efficiency compared to ventriculoperitoneal shunt in INPH treatment. It is indicated for INPH patients who are not ideal candidates for intracranial procedures. LP shunts are technically more difficult to perform in obese patients, in ones with spinal deformity (kyphosis, scoliosis), degenerative lumbar stenosis. In these patients, repeated lumbar dural sac punctures may be necessary, which can lead to venous plexus lesions with blood-stained CSF and also radicular lesions.9

Endoscopic third ventriculostomy can be performed "d'emblée" or in the case of VP shunt malfunction. A rigid neuroendoscope is introduced through a burr hole placed in the right Kocher point (10-13 cm posterior from the nasion and 2-3 cm lateral from the median line) up to the third ventricle, through Monro's foramen. The floor of the third ventricle is entered between the mammillary bodies and the infundibular recess and the fenestration is enlarged by inflating a Fogarty catheter's balloon, thus creating a communication between the third ventricle and the prepontine cistern, representing a 4-6 mm diameter ventriculostomy. The patients who, intraoperatively, immediately after ETV present no pulsations on the floor of the third ventricle ("flag signal"), will not improve after ETV and need immediate VP shunting10.

Ventriculosternal shunting

In critical situations, when vascular access cannot be performed, intraosseous fluid infusion is a procedure that has been performed for many years11.

From this technique, Tubbs conceived ventriculosternal shunting. The distal catheter is implanted in the sternal manubrium, after a 4 cm tunnelization. From this point, CSF is drained through the internal mammary veins and the azygos vein12.

The ventriculosternal shunt represents an alternative to other types of shunting, when they are contraindicated. Risks of fat embolism, sternal iatrogenic fracture, brachiocephalic vein injury or vagus nerve injury are disadvantages of this method13.

Types of valves

CSF pressure must be measured intraoperatively and a valve corresponding to that pressure must be chosen or in the case of a programmable valve, the opening pressure will be adjusted to the measured pressure.

There are low, median and high pressure valves with a range of opening pressure of 20 - 40 mmH2O, 50 - 90 mmH2O and 100 – 140 mmH2O, respectively.

CSF overdrainage can occur in the case of VP shunts because of the hydrostatic pressure of the liquid column, when the patient is standing. Anti-siphon systems, placed distally from the valve, prevent overdrainage and intracranial pressure decrease.

In the case of programmable valves, pressure values can be adjusted noninvasively, thus avoiding revision surgery to replace the existing valve with a different pressure one.

Postoperative course

Preoperatively, patients that will benefit from the shunting procedure must be selected and the surgery must be tailored to the patient's particularities (type of shunting/valve). In the postoperative period, all patients need clinical and imagistic follow-up in order to evaluate their response to surgical treatment and to obtain an early diagnosis of shunt system malfunction or possible complications.

Out of the cardinal symptoms of INPH, gait disturbances are the first to improve. Evaluation of the cognitive component requires psychometric testing. Stein and Laughfitt scale, Black scale or Rankin scale can be used14,15.

Approximatively ¼ of operated patients experience shunt malfunction in the first year after surgery and up to 60% require shunt revision during the next years. The rate of shunt failure in the first year is 20 - 50%, with a mean of 40%16. Two causes contribute to shunt malfunction: inadequate catheter placement and infection. In order to prevent proximal catheter malposition, when the catheter is located paraventricularly or partially intraventricular, adequate placement of ventricular catheter can be achieved with the help of image guidance, stereotaxis, neuronavigation or endoscopy. A correctly placed catheter must float at equal distance from the ventricular walls, far from the choroid plexus and must have a straight trajectory from the burr hole. All catheter perforations must be located intraventricularly in order to prevent its obstruction with paraventricular tissue16.

Recently, the concept of prevention of readmission and reoperation has been introduced in neurosurgery, from which Preventable Shunt Revision Rate was derived and introduced in the US in 2016. The fact that a part of shunt failures are caused by intraventricular catheter malposition was noted. Therefore, at the end of each operation a control of catheter position with imagistic guidance is recommended.

The rate of shunt infection reported in the literature varies between 1% and 15%. This rate is lower when: antibiotic impregnated catheters are used, 2 layers of gloves are worn by the operators, the number of medical personnel in the operating room is limited, prophylactic antibiotherapy is administered, antibiotics are applied in the surgical wound (Bacitracine) etc. All these aforementioned measures can prevent bacterial inoculation during surgery.

In the case of febrile patients (temperature > 38oC) with meningeal irritation signs, CSF must be obtained through lumbar puncture in order to obtain cultures and determine the microorganisms that are present as well as their antibiotic sensitivity.

Overdrainage, which occurs more frequently in standing position, in the case of an inadequate valve can determine the formation of a subdural fluid collection, followed by the development of subdural hematoma. In other patients, the drainage is insufficient and the valve must be replaced with a low pressure one.

Conclusions

INPH is a frequently encountered condition that is still challenging to diagnose and treat despite significant progress in imaging diagnosis and development of different surgical techniques. Although surgical treatment consists of generally straightforward procedures, it associates a high rate of failure, regardless of technique used, which emphasizes the need for regular clinical and imagistic follow-up. Thus, INPH remains a disease where there is significant room for improvement, both in diagnosis and treatment.

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