Alexandria Journal of Medicine (2014) **50**, 341–344

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Alexandria Journal of Medicine

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Endoscopic third ventriculostomy in idiopathic normal pressure hydrocephalus

Mohammed Ahmed Eshra *

Department of Neurosurgery, Faculty of Medicine, Alexandria University, Egypt

Received 20 June 2013; accepted 20 November 2013
Available online 22 December 2013

KEYWORDS

CSF;
Idiopathic normal pressure hydrocephalus;
Endoscopic third ventriculostomy

Abstract *Objective:* To determine the efficacy of endoscopic fenestration of the third ventricle in the treatment of idiopathic normal pressure hydrocephalus.
Methods: 16 patients with idiopathic normal pressure hydrocephalus were treated by endoscopic third ventriculostomy. This study lasted three years. All patients were over 50 years old complaining of 2 or more of the famous triad of gait disturbance, urinary incontinence and dementia. All patients had ventricular dilatation with negative history of infection, brain injury or hemorrhage.
Results: 8 patients showed marked improvement in their clinical conditions, 3 patients showed mild improvement, while 5 patients had no improvement without any deterioration in their preoperative state. Thus the overall improvement after ventriculostomy occurred in 11 patients (68%).
Conclusion: Endoscopic third ventriculostomy is as effective as shunt in treating idiopathic normal pressure hydrocephalus. Patients must be treated as early as possible before permanent damages occur. Further studies are needed to clarify more about the pathophysiology, CSF dynamics and new management.
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1. Introduction

Normal pressure hydrocephalus (NPH) is a clinical syndrome characterized by gait disturbance, bladder incontinence, and deterioration of the mental power (dementia). Its clinical importance lies in the possibility of being one of the reversible causes of dementia. First described by Hakim in 1965, NPH describes hydrocephalus in the absence of papilledema and with normal cerebrospinal fluid (CSF) opening pressure on lumbar puncture.[1]

This syndrome is characterized by mild intracranial hypertension due to increased CSF accumulation in the ventricular system of the brain causing ventricular enlargement.

This is followed by gradual falling of the intracranial pressure till reaching the high normal level of 150–200 mm H_2O. So when measuring the ICP, the results are usually not elevated.

The real pathophysiological way of occurrence of this type of hydrocephalus is not fully understood but most researchers consider it as a type of communicating hydrocephalus with impaired CSF reabsorption at the arachnoid granulations.

* Tel.: +20 1001529424.
E-mail address: eshrawyalatool@yahoo.com
Peer review under responsibility of Alexandria University Faculty of Medicine.

http://dx.doi.org/10.1016/j.ajme.2013.11.004
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There are 2 types of normal pressure hydrocephalus: idiopathic and secondary. The term idiopathic means that there is no detectable cause. The secondary type of NPH can be due to a subarachnoid hemorrhage, head trauma, tumor, CNS infection, or as a sequel of cranial surgery.[2] NPH can occur at any age but commonly in the elderly people with an incidence of about 5.5 patients per 100,000 of people per year.[3–5]

Diagnosis of NPH is done by clinical examination, imaging studies including CT and MRI to confirm or rule out any organic lesions in the brain which may be responsible for such clinical syndrome and the procedure in the clinical test which helps in the diagnosis is to do a lumbar puncture to estimate the opening pressure and to estimate the clinical improvement after CSF removal which has a high predictive value for subsequent success with shunting. This is called the ''lumbar tap test'' or Miller Fisher test.[19] On the contrary, 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. This test may be completed by continuous CSF drainage by insertion of intrathecal drain with tidal drainage for 2 or 3 days.[19]

CT scan may show ventriculomegaly without atrophy of the brain parenchymal mass. MRI may show some degree of periventricular haze due to transependymal migration of CSF surrounding the ventricles on T2/FLAIR sequence. Imaging however cannot differentiate between pathologies with similar clinical picture like Alzheimer's dementia, vascular dementia or Parkinson's disease.[6]

Infusion test is a test that may have higher sensitivity and specificity than a lumbar puncture, but is not performed in most centers. The outflow conductance (Cout) of the cerebrospinal fluid (CSF) system is a parameter considered by some centers to be predictive in selection for hydrocephalus surgery. Cout can be determined through an infusion test. This is not a test that is normally performed prior to shunting, but may become more accepted.

In some centers, External lumbar drainage has been shown to have the highest sensitivity and specificity with regard to predicting a successful outcome following surgery.[7]

2. Patients and methods

Sixteen patients with idiopathic normal pressure hydrocephalus (INPH) were treated by endoscopic third venticulostomy (ETV) over a period of three years. 13 patients were males and 3 were females. All patients were over 50 years old complaining of two or more symptoms of the classic triad of dementia, gait disturbance and urinary incontinence for less than one month. All patients had ventricular dilatation with negative history of infection, brain injury or hemorrhage. All patients underwent clinical examination, brain imaging and ICP measurement through lumber puncture. In all patients, ICP were less than 15 cm H_2O (8–14 cm H_2O).

MRI revealed moderate dilatation of the four ventricles i.e., ventricles dilated out of proportion to any sulcal enlargement (which distinguishes it from atrophy), slightly enlarged sylvian fissure and slight cortical atrophy.[20]

All patients were evaluated pre and postoperative by the grading system of the Japanese Committee for Scientific Research on Intractable Hydrocephalus (JCSRIH),[21] as shown in Table 1.

The total score of the triad of symptoms was used to evaluate the severity of the clinical syndrome pre and postoperative. Tap test was done in all patients and a noticeable improvement occurred in ten patients (negative test in 6 patients). We operated on all 16 patients as 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.[20] Postoperative phase contrast MRI was done after the endoscopic surgery. No statistical analysis of the results was applied due to small number of patients.

Table 1 Grading system of the Japanese Committee for Scientific Research on Intractable Hydrocephalus (JCSRIH).

Grade	Definition
Gait disturbance	
0	Normal
1	Unstable but independent
2	Walking with one cane
3	Walking with two canes or a walker frame
4	Walking not possible
Dementia	
0	Within normal range
1	No apparent dementia but apathetic
2	Socially dependent but independent at home
3	Partially dependent at home
4	Totally dependent
Urinary incontinence	
0	Absent
1	Absent but with urgency
2	Sometimes only at night
3	Sometimes during the day
4	Frequent

2.1. Surgery

All patients were operated upon under general anesthesia. After ventricular access, we measured the ventricular pressure which did not differ from that measured through the lumbar puncture. The standard Carl Storz 6 mm rigid endoscopy was used to perform a wide ventriculostomy in the third ventricle floor (tuber cinereum) anterior to the two mammillary bodies. Any secondary membrane was violated. After the ventriculostomy the transmitted pulsations of the basilar artery were observed through the floor.

3. Results

According to the grading system, 3 patients were found to be of grade 2, one patient grade 5, 6 patients grade 6; one patient grade 7, 3 patients grade 8, and 2 patients grade 9.

No intra operative complications were encountered. The patients were followed up both clinically and radiologically based on 1–1.5 month intervals with total follow up ranging from 7 to 26 months.

8 patients showed marked improvement in their clinical conditions that started immediately postoperatively and continued to progress up to one week later and this improvement was manifested by reduction in their grade by two up to seven degrees compared to the preoperative grade. If no improvement after one week, we considered the result as failure.

The gait disturbance improved in all ten patients, urinary incontinence improved in 6 patients (of the 10) whereas dementia improved in 2 patients (of the 10).

3 patients showed mild improvement in their clinical conditions manifested by reduction in their grade by one to two degrees compared to the preoperative grade. Only one of them needed further management in the form of ventriculoperitoneal shunt.

5 patients had no improvement without any deterioration in their preoperative state. They underwent a ventriculoperitoneal shunt insertion resulting in improvement of only 2 patients.

Thus the overall improvement after ventriculostomy occurred in 11 patients (68%).

4. Discussion

The pathophysiologies of occurrence of NPH were studied by many authors and the majority of them have explained the syndrome on the basis of both mechanical and ischemic factors.

Some authors suggested that the ventriculomegaly leads to vascular affection by stretching, and the decreased compliance and high pulse pressure lead to local "barotrauma" or "tangential shear stress". It has been postulated that the purpose of the shunt is to add additional capacitance to the system, increasing perfusion, not to decrease the pressure (which is already normal).[8–15]

The acetazolamide challenge test, which normally increases CBF, fails to do so in NPH patients, particularly in the periventricular white matter. After CSF diversion, CBF in white matter generally improves, as does the response to acetazolamide.[16]

This lack of the usual vasomotor response to carbonic anhydrase inhibitors (or to inhaled CO_2) probably indicates that the arterioles are already maximally dilated as a result of local ischemia.[17]

The VP shunt modulates the pulse pressure; there will be decreased interstitial edema, decreased interstitial pressure, improved perfusion, and decreased ischemia.[18]

Today, most surgeons agree with the shunting of the secondary forms of normal pressure hydrocephalus but shunting of the idiopathic form is controversial because of the difficulty in distinguishing the idiopathic form from other neurodegenerative conditions.[20]

Endoscopic third ventriculostomy is used widely and successfully nowadays in treating obstructive hydrocephalus and to less extent in treating communicating hydrocephalus.

Mitchell et al. have reported the use of ETV in treating INPH with a good and favorable outcome with overall success rate of about 75%.[18] However Michelangelo Gangemi et al. reported a total improvement in 72% of cases and these results are similar to our results to a large extent.[20]

Performing ETV leads to minimal reduction in the intra ventricular CSF pressure and similar to the shunt operation; this leads to an increase of the cerebral blood flow and cerebral perfusion pressure leading to improvement of the patient symptoms.

Michelangelo Gangemi et al. in 2004 have reported that the rapid transmission of the pressure wave through the ventriculostomy toward the basal cistern could restore the CSF dynamics. They observed that before ventriculostomy, the CSF contained in the ventricles was still but after ventriculostomy, there were pressure waves and pulsations of the floor of the third ventricle. Also they reported an increase of the CSF flow at the tentorial incisura in patients with INPH after third ventriculostomy.[20]

CSF tap test was performed before ventriculostomy and had a good predictive value. However, Michelangelo et al. think that the CSF tap test is not useful before ETV because this procedure is not associated with CSF withdrawal and diversion. But to my opinion, the ETV done in INPH is associated with minimal CSF diversion.

The infusion test was not done as it is invasive and carries the risk of infection especially in elderly patients.[19]

Postoperative clinical improvement after ETV, indirectly confirms that in these patients with INPH, the CSF absorption is not severely compromised and is sufficient to ensure a significant improvement of the CSF dynamics. ETV may improve this dynamics only in the early stage, before significant damage to the periventricular brain parenchyma occurs. So ETV must be performed in patients with clinical evolution of not more than one year before diagnosis.[20]

ETV is performed as the first successful procedure before shunt in patients with short preoperative symptoms with prevalence of gait disturbances with little or no dementia. These criteria suggest the presence of a fair grade of brain compliance and little damage of the brain parenchyma of the periventricular structures. On the contrary patients with a long history of symptoms and significant dementia may be candidates for shunt procedure from the start.[20]

Alan R. Cohen in his comment on Gangemi study reported that: NPH is a form of communicating hydrocephalus. How can ETV lead to clinical improvement in these patients? We will need to modify our thoughts about hydrocephalus and its treatment. Perhaps a fenestration in the third ventricular floor, creating a communication with the basal subarachnoid space, modifies the CSF pulse pressure wave and thereby leads to clinical improvement. Whatever the mechanism, this stimulating and controversial study offer a new possible treatment for selected patients with NPH and will certainly serve as an impetus for further investigations.[20]

5. Conclusion

Endoscopic third ventriculostomy is as effective as shunt in treating idiopathic normal pressure hydrocephalus. Patients must be treated as early as possible before permanent damages occur. The parameters that determine the improvement include short preoperative symptoms with prevalence of gait disturbances with little or no dementia and positive CSF tape test. Further studies are needed to clarify more about the pathophysiology, CSF dynamics and new management.

Conflict of interest

None.

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