The European Research Journal 2023;9(6):1464-1473 Original Article

DOI: 10.18621/eurj.1347626 *Neurosurgery*

Comparative retrospective analysis of patients with idiopathic normal pressure hydrocephalus and aqueductal web-related aqueductal stenosis

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ABSTRACT

Objectives: Aquaductal web (AW) is a special form of aqueductal stenosis with similar clinical presentation with idiopathic normal pressure hydrocephalus (iNPH). iNPH is indeed a communicating hydrocephalus syndrome whereas AW is a noncommunicating subtype. Here, we aimed to investigate the similarities and differences between these two different chronic hydrocephalus syndromes in terms of clinical signs and symptoms, response to shunt treatment and postoperative complications.

Methods: Forty-one patients who underwent shunt operation with the diagnosis of iNPH or AW at our clinic between January 2010-May 2019 were retrospectively analyzed. Patients were evaluated by age, gender, clinical sign and symptoms, comorbidities, intraoperative and postoperative complications, and early and late postoperative outpatient follow-up findings.

Results: Twenty-six patients were classified as iNPH group and 15 patients as AW group. Patients in the AW group were significantly younger (45.5 ± 15.6 years vs. 60.3 ± 15.4 years) than the iNPH group (p = 0.006). There was no statistical difference between the groups in terms of subdural effusion formation, need for shunt revision (p = 1.000). Chronic hydrocephalus symptoms regressed in 23 (88.5%) patients in the NPH group, and at least one of them improved. symptoms. This rate was 66.7% (n = 10) in the AW group. Both groups showed similar clinical improvement with VPS (p = 0.1169).

Conclusions: The placement of ventriculoperitoenal shunt is widely used in the treatment of iNPH. As iNPH and AW has clinical similarities despite the discrepancies between underlying pathophysiological mechanisms and both clinical entities respond similarly to shunt treatment we advocate VPS surgery in the management of AW as well.

Keywords: Normal pressure hydrocephalus, aqueductal stenosis, ventriculoperitoneal (VP) shunt

Idiopathic normal pressure hydrocephalus (iNPH) is a chronic hydrocephalus syndrome usually seen in the elderly population and typically presents with balance and gait disturbances, cognitive dysfunction, and

Received: ***August 22, 2023;*** *Accepted:* ***October 2, 2023;*** *Published Online:* ***October 9, 2023***

How to cite this article: *Türkkan A, Eser Ocak P, Altunyuva O, Sönmez B, Özpar R, Bekar A. Comparative retrospective analysis of patients with idiopathic normal pressure hydrocephalus and aqueductal web-related aqueductal stenosis. Eur Res J 2023;(6):1464-1473. DOI: 10.18621/eurj.1347626*

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urinary incontinence [1, 2]. It was first described by Hakim and Adam in 1965 [2]. It may present with varying combinations or degrees of each element of the classic clinical triad [3-5]. iNPH is not a rare clinical condition. The prevalence of iNPH in the elderly population is estimated to be approximately 0.2%-5% [6]. One of the main characteristics of iNPH is that cerebrospinal fluid (CSF) pressure is within normal ranges. Typical brain imaging shows ventriculomegaly, periventricular hyperintensities, wide opening of the sylvian fissure, narrowed subarachnoid space, and highly convex cortical sulci [7-9].

The pathophysiological processes of iNPH are poorly understood, leading to a lack of clear diagnostic criteria [10]. In addition, the clinical characteristics of iNPH can be confused with those of different neurologic extrapyramidal disorders such as Parkinson's disease, Alzheimer-type dementia, and leukoarthrosis [4, 7, 11]. This makes patient selection for surgical treatment and its differentiation from other neurodegenerative disorders difficult. Since it is the only variant of dementia disorders that can be treated surgically, it is recommended to diagnose and treat it as early as possible [1, 4, 12].

Aqueductal Web (AW) is a rare pathology that causes chronic hydrocephalus syndrome by causing stenosis in the distal part of the aqueductus cerebri. AW is a translucent membrane composed of clusters of ependymal cells and fibrillar neuroglia [13]. AW is a special form of aqueductal stenosis [13]. Very similar to iNPH, AW also clinically presents with gait disturbance, cognitive decline, and incontinence, except it often presents at younger ages. There is no globally accepted magnetic resonance imaging (MRI) protocol for evaluating patients with a prediagnosis of AW; therefore, it is often difficult to clearly define the etiology of hydrocephalus using routine MR images. Of note, 3D T2-weighted gradient-echo sequences of CSF flow MRI examination may help better assess the flow dynamics in CSF and may define a better characterization of the aqueductal opening and associated abnormalities [14-16].

Although iNPH and AW harbor clinical and radiological similarities, iNPH is indeed a communicating hydrocephalus syndrome whereas AW is a noncommunicating subtype [17]. Since both iNPH and AW can benefit from CSF diversion, it is important to differentiate them from other causes of dementia with ventriculomegaly.

Therefore, we aimed to investigate the similarities and differences between these two different chronic hydrocephalus syndromes in the present study in terms of clinical signs and symptoms, diagnosis, response to shunt treatment, and complications. In addition, we examined the results of ventriculoperitoneal shunt surgery in the treatment of both patient groups.

METHODS

This retrospective examination was approved and authorized by the hospital clinical research ethics committee with the number 2023/03 and was subsequently performed by the regulations of the Declaration of Helsinki. Forty-one adult patients who underwent shunt operation with the diagnosis of iNPH or AW at our clinic between January 2010 and May 2019 were retrospectively analyzed. The patients were evaluated by age, gender, clinical signs and symptoms, comorbidities, neurologic examination, intraoperative and postoperative complications, and early and late postoperative outpatient follow-up.

The patients who had ventriculomegaly (Evan's index > 3) on brain computed tomography (CT) or magnetic resonance imaging (MRI) either accompanied or not accompanied by the classical triad symptoms (difficulty in walking, urinary incontinence, and dementia) were further examined for a chronic hydrocephalus syndrome. Gait disturbance was assessed using the 10-m walk test, and dementia was assessed using the Mini-Mental State Examination (MMSE). In addition, urinary continence was assessed by interviewing the patients and/or their caregivers. In all patients, the surgical decisions were supported by gait assessment after a lumbar tap test including drainage of 40 ml CSF by lumbar puncture. Recovery after lumbar puncture was defined as subjective improvement reported by patients and/or their family members. This test was considered supportive, not mandatory, for shunting decisions in the patients.

For radiologic evaluation, brain MRI, CSF flow MRI (3D T2-weighted gradient-echo sequences), diffusion tensor MRI, and brain CT imaging were performed in all patients. The Evans index was calculated

in each patient by dividing the maximum width between the frontal horns of the lateral ventricles by the distance between the two inner tabulae. Patients with CSF flow MRI and brain MRI revealing a membranous structure at the aqueductal level were diagnosed with AW. Those with Evans index < 0.30 were excluded from the study. Patients with a potential underlying cause for hydrocephalus such as a history of head trauma, intracranial hemorrhage, stroke, meningitis, primary malignancy, etc. were also excluded.

Ventriculoperitoneal Shunting Protocol

All patients included in the study underwent ventriculoperitoneal shunt (VPS) placement surgery. The surgical procedure included the placement of a ventricular catheter through a burr hole in the right frontal Kocher's point and a peritoneal catheter through a midline or paraumbilical mini-laparotomy. Codman programmable valve (Johnson and Johnson, MA, USA) was used in all patients.

After VPS surgery, all patients underwent regular follow-up examinations at 3, 6, and 12 months and then annually. Lengths of hospital stay, complications, or readmissions related to the ventriculoperitoneal shunting procedure were documented.

Shunt Response Assessment

Response to VPS was assessed approximately 6 months after surgery during outpatient follow-up visits. A 20% improvement in the 10-m walk test was considered a significant improvement. In addition, an increase of 2 or more in the MMSE score was considered a significant improvement. However, due to the study's retrospective design, postoperative objective measures of gait and cognition were only available in approximately 80% of the patients. For the remaining patients, scores were obtained by interviewing the patients and/or their caregivers.

Statistical Analysis

Continuous variables were expressed in terms of mean ± standard deviation. Independent samples t-test was used for comparisons between the two groups according to normality test results. Categorical variables were presented as frequency and percentage values [n (%)] and compared using the Pearson Chi-square test. Statistical analysis was performed using GraphPad Prism 7 (GraphPad Software, San Diego, CA, USA). A p value of <0.05 indicated statistical significance.

RESULTS

Our study included 41 patients with chronic hydrocephalus. Twenty-six patients (14 males and 12 females) which included patients with clinical, laboratory, and routine cranial MR findings suggestive of iNPH were classified as iNPH group. The mean age of the patients in this group was 60.3 ± 15.4 years. Those with a membranous structure at the aqueductal level in their CSF flow MRI and brain MRI were classified as AW group and this group included 15 (7 males and 8 females) patients. The mean age of the patients in this group was 45.5 ± 15.6 years. The AW group patients were significantly younger than the iNPH group (p = 0.006).

In the iNPH group, the main presenting complaint was dementia (n = 22, 84.6%) followed by ataxia and gait disturbance (n = 20, 76.9%) and urinary incontinence (n = 16, 61.5%). In the AW group, 11 (73.3%) patients presented with dementia, 8 (53.3%) with ataxia and gait disturbance, and 8 (53.3%) with urinary incontinence. The initial symptom developed in both groups was dementia (n = 18 vs. n=7; 69.2% vs. 46.7%, respectively), followed by ataxia and gait disturbance (n = 6, 23.1% vs. n=5, 33.3%), and urinary incontinence (n = 2, 77% vs. n = 3, 20%). Naturally, dementia was the symptom with longest duration in both the iNPH and AW groups (28.8 ± 45.5 months vs. 22.3 ± 33.1 months).

In the AW group, headache was observed in 7 (46.7%) patients and nausea/vomiting in 4 (26.7%) patients, which were the symptoms that might have potentially been of significance for increased intracranial pressure. However, in the iNPH group, 9 (34.6%) patients had headaches, and only 1 (3.8%) had nausea/vomiting. The distribution of presenting symptoms was similar between groups (p > 0.05).

Type-2 diabetes mellitus was the most common comorbidity which was seen in 14 (38.5%) patients in the iNPH group and 4 (26.7%) patients in the AW group (Table 1).

Radiologic examinations revealed ventriculomegaly without enlargement of the sulci in the iNPH

Table 1. General demographic characteristics and clinical conditions of patients with iNPH and AW

Variables	iNPH (n = 26)	AW (n = 15)	*p* value
Sex, n (%)			
Female	12 (46.1)	8 (53.3)	0.7513
Male	14 (53.8)	7 (46.7)	
Mean age (years)	60.3 ± 15.4	45.5 ± 15.7	**0.006**
Symptoms at presentation, n (%)			
Dementia	22 (84.6)	11 (73.3)	0.433
Gait disturbance	20 (76.9)	8 (53.3)	0.167
Urinary incontinence	16 (61.5)	8 (53.3)	0.744
Paraparesis	16 (61.5)	6 (40)	0.211
Headache	9 (34.6)	7 (46.7)	0.517
Dizziness	1 (3.8)	4 (26.7)	0.051
Nausea/vomiting	1 (3.8)	4 (26.7)	0.051
First symptom, n (%)			
Dementia	18 (69.2)	7 (46.7)	0.194
Gait disturbance	6 (23.1)	5 (33.3)	0.490
Urinary incontinence	2 (7.7)	3 (20)	0.336
Mean duration of symptoms (months)			
Dementia	28.8 ± 45.5	22.3 ± 33.1	0.602
Gait disturbance	17.2 ± 24.9	21.3 ± 35.5	0.697
Urinary incontinence	15.7 ± 21.6	13.7 ± 30.6	0.826
Comorbidities, n (%)			
Diabetes	10 (38.5)	4 (26.7)	0.511
Hypertension	9 (34.6)	3 (20)	0.479
Coronary artery disease	5 (19.2)	-	0.139
Thyroid goiter	3 (11.5)	1 (6.7)	1.000
Parkinson	3 (11.5)	0	0.286
Alzheimer's sisease	2 (7.7)	0	0.524
Benign prostate hypertrophy	1 (3.8)	1 (6.7)	1.000
Cerebrovascular disease	1 (3.8)	1 (6.7)	1.000
Atrial fibrillation	1 (3.8)	1 (6.7)	1.000
Schizophrenia	-	1 (6.7)	0.365

Data are shown as mean ± standard deviation or n (%). AW = Aqueductal web, iNPH = Idiopathic normal pressure hydrocephalus

group. In CSF flow measurements of all patients, a sinusoidal flow pattern symbolizing forward-backward movement was found in the aqueductus cerebri, while no-signal void (hyperdynamic CSF flow) reflecting rapid CSF flow was detected at the aqueductal level.

All patients underwent shunt surgery using a programmable VPS. In the early postoperative period, one (3.8%) patient in the iNPH group developed an intraventricular hematoma, and one patient (6.7%) in the AW group suffered from internal carotid artery (ICA) infarction. The patient who developed intraventricular hematoma was followed up in the intensive care unit for 13 days with an external ventricular drainage. The patient with ICA infarction underwent thrombectomy by interventional radiology. Both patients were discharged home without additional neurological deficits.

In 14 (53.8%) patients in the iNPH group, the shunt settings required adjustment due to subdural effusion or inadequate clinical improvement during outpatient follow-ups. In five of these cases (19.2%), shunt pressure changes were insufficient in treating subdural effusion and subdural drainage was required. Shunt revision was performed in one (4%) case. In seven (46.7%) patients in the AW group, the shunt settings were adjusted due to subdural effusion or inadequate clinical improvement. Subdural drainage was

Table 2. Overall early (postoperative first 24 hours) and late postoperative (postoperative 6 months) complications following ventriculoperitoneal shunt surgery for idiopathic normal pressure hydrocephalus and aqueductal web

	iNPH (n = 26)	AW (n = 15)	All patients (n = 41)
Early Complications, n (%)			
ICA infarct	1 (3.8)	0	1
Intraventricular hematoma	0	1 (6.7)	1
Late Complications, n (%)			
Subdural effusion/hematoma requiring drainage	5 (19.2)	3 (20)	8 (19.5)
Shunt revision	2 (7.7)	1 (6.7)	3 (7.3)
Wound discharge	0	1 (6.7)	1 (2.4)

AW = Aqueductal web, ICA = Internal carotid artery, iNPH = Idiopathic normal pressure hydrocephalus

needed in two (13.3%) of these cases, one (6.7%) of which also underwent shunt revision (Table 2).

During a mean follow-up period of 37.8 ± 27.1 months (42.4 ± 30.2 vs. 29 ± 20.4 months), 23 (88.5%) of the patients in the iNPH group showed regression of chronic hydrocephalus symptoms and improvement in at least one of the symptoms. In the AW group, this rate was 66.7% (n = 10). There was no statistical difference between the groups in terms of the occurrence of subdural effusion (p = 1.000), and the need for shunt revision (p = 1.000). Both groups showed similar clinical improvement with VPS (p = 0.1169) (Table 3).

DISCUSSION

The third edition of the iNPH guidelines published in Japan in 2021 proposes a new classification for NPH [4]. According to this newly revised classification, the iNPH category is clearly distinguished from NPH by its congenital/developmental and acquired etiologies [4]. Diagnosing acquired NPH is not difficult, which occurs after a specific etiologic event. On the other side, the differential diagnosis of iNPH is difficult due to the similarity of nonspecific symptoms with many other diseases. However, AW which causes hydrocephalus in adulthood, is defined as late-onset congenital hydrocephalus [4]. AW is actually a special form of aqueductal stenosis [13]. The web consists of a thin ependymal membranous septum, and the cause of its formation is unknown.

Although there are large series in the literature about iNPH, the literature data regarding AW mainly consists of case reports. For example, Chen *et al*. [18] reported that 21 of 2009 patients treated for obstructive hydrocephalus had a membranous structure at the aqueductal level. Matsuda *et al*. [19] presented a case

Table 3. Clinical outcome of patients who underwent ventriculoperitoneal shunt surgery for idiopathic normal pressure hydrocephalus and aqueductal web

	iNPH (n = 26)	AW (n = 15)	*p* value
Mean follow-up time (months)	42.4 ± 30.2	29 ± 20.4	0.099
Adjustment in valve pressure	14 (53.8)	7 (46.7)	0.751
Clinical improvement	23 (88.5)	10 (66.7)	0.116

Data are shown as mean ± standard deviation or n (%). AW = Aqueductal web, iNPH = Idiopathic normal pressure hydrocephalus

of late-onset aqueductal membranous occlusion hydrocephalus. Terada *et al*. [20] presented an adult case of hydrocephalus caused by a membranous structure at the aqueductal level. However, most of these studies reported cases that were diagnosed intraoperatively. Therefore, patients with adult-type AW may not have been adequately identified due to flaws in the preoperative diagnosis. In this context, our study is one of the most extensive series of patients treated for adult-type AW-related hydrocephalus who were diagnosed with the presence of an AW preoperatively.

On the other side, iNPH is highly prevalent, particularly in the elderly population. In a population-based study in West Sweden, the prevalence of iNPH was estimated to be between 0.2%-5.9% in the seventh and eighth decades [6]. The average onset for iNPH is around 70 years of age, and men and women are affected equally [6, 21]. However, AW is usually detected in childhood due to the obstructive type of hydrocephalus. As AW is extremely rare in adults, it is believed to present in these patients since childhood, and become symptomatic after years of compensated ventriculomegaly [13, 19, 20]. AW often needs clarification with iNPH. Nevertheless, the age of onset is much earlier than that of in iNPH patients. In consistence with the literature, in the current study, we found that patients diagnosed with adult-type AW were significantly younger compared to the group of patients diagnosed with iNPH.

Since iNPH and AW both cause chronic hydrocephalus, they display clinical similarities although they differ pathophysiologically [22]. iNPH is a nonobstructive hydrocephalus, and the exact underlying pathophysiological mechanisms remain unclear although several different theories have been proposed so far. According to the most accepted theory, there is an increased resistance to CSF absorption from the arachnoid villi [23].As a result, lack of adequate CSF absorption leads to ventricular enlargement. CSF leaks through the ependymal spaces into the periventricular white matter and causes a decrease in tissue metabolism, resulting in ischemia and hypoxia in white matter axons. Eventually, demyelination and neural apoptosis secondary to chronic ischemia and hypoxia occur [23]. Kuriyama *et al*. [10] reported that retrograde flow in the internal jugular vein during the Valsalva maneuver was significantly higher in patients with iNPH compared to the control group. They emphasized that this condition may cause the resultant picture with increased central venous pressure and decreased CSF absorption [10].

Contrarily, AW causes obstructive hydrocephalus due to the presence of a translucent membrane formed by clusters of ependymal cells and fibrillary neuroglial cells in the distal part of the aquaeductus cerebri [13, 24]. For its development, various previous inflammatory processes as well as congenital glial cell occlusion have been blamed [20]. At the diagnostic stage, many AW patients are mistaken for iNPH patients as both groups show findings known as the classic clinical triad of Hakim and Adams [2] which consists of a symmetrical gait disorder with slow, short, and wide-based steps, urinary incontinence, and dementia. In the literature, gait disturbance has been reported to be the most common abnormal walking pattern [4, 25, 26]. However, this gait pattern is also common in Parkinson's disease as well as other degenerative and vascular diseases affecting the CNS such as peripheral neuropathies, spinal stenosis, myelopathy, vitamin B12 deficiency, and sometimes even combination of these diseases [4, 7, 11]. Of note, less than two-thirds of patients with iNPH have been reported to display all three components of the triad [4, 5, 11].

In the present study, gait disorders were found in 76.9%, urinary incontinence in 61.5%, and cognitive impairment in 84.6% of the iNPH patients. Clinical symptoms caused by increased intracranial pressure, such as headache and nausea/vomiting were more common in patients with AW (46.7%) than in the patients with iNPH (36.6%). A similar occurrence of Hakim's triad was present in slightly smaller percentages of patients in the AW group as well with a ratio of 53.3%, 53.3%, and 73.3% for gait impairment, urinary incontinence, and cognitive impairment respectively. The most common presenting symptom and first symptom in both iNPH and AW was forgetfulness. The ratios for the presence of other symptoms and mean symptom duration were similar between groups (Table 1). In this respect, there is a high degree of clinical similarity between iNPH, a nonobstructive chronic hydrocephalus syndrome, and AW, an obstructive hydrocephalus syndrome.

Obviously, we need accurate imaging techniques to show CSF pathways in detail in patients for whom treatment is planned with the diagnosis of hydrocephalus. In fact, iNPH and AW cannot be differenti-

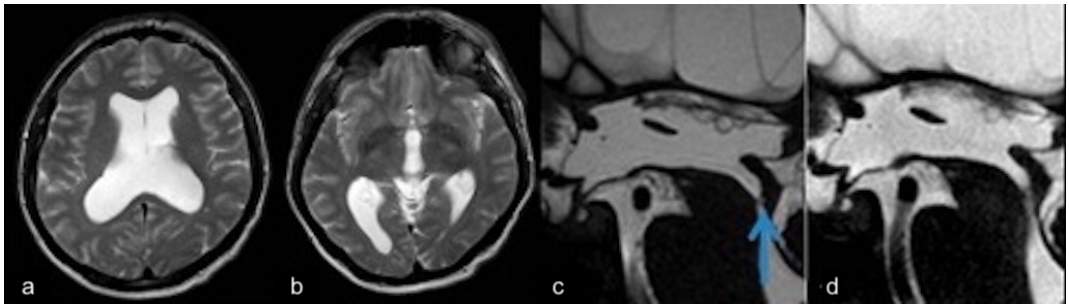

Fig. 1. CSF Flow MRI image of a patient with an aqueductal web. There is chronic hydrocephalus in the lateral ventricles (a) and third ventricle (b) on T2-weighted images. (c) In the 3D Balanced Heavy T2 image there is membrane formation distal to the aqueduct (blue arrow). (d) No expected flow void area of aqueductal CSF flow in flow-sensitive T2 DRIVE sequence.

ated from each other based on conventional radiologic examinations such as cranial CT and MRI [7, 17, 27]. Therefore, patients with AW may be overlooked. Invasive methods such as radionuclide cisternography, CT cisternography with iodinated contrast or air, and MR cisternography with gadolinium-based contrast have been described and used so far. However, these methods have not gained widespread acceptance due to their invasive nature. In examinations such as 3D T2-weighted gradient-echo sequences of CSF flow MRI, the membrane formation of the aqueductal level can be detected in patients with AW, and the expected signal void area due to CSF flow may be absent (Fig. 1). On the other hand, iNPH can be diagnosed by detecting the signal void area representing rapid CSF flow at the aqueductal level (Figs. 2 and 3). According to the current literature, CSF flow MRI supports the diagnosis of iNPH and helps diagnose AW [3, 7, 14, 27]. In the present study, all patients underwent 3D T2-weighted gradient-echo sequences of CSF flow MRI preoperatively to confirm our diagnosis. We found that 36.6% of the patients for whom we planned surgery with a prediagnosis of iNPH had indeed an AW in the radiologic examinations. This was in line with the study by Giordan *et al*. [16] who reported the presence of aqueductal stenosis in a low percentage of iNPH patients (10%) and suggested investigation with high-resolution MRI in patients with iNPH. We observed AW in 36.6% of our chronic hydrocephalus patients. We believe that further investigations, such as 3D T2-weighted gradient-echo sequences of CSF flow MRI are more sensitive to display web formation at the aqueductal level, leading to higher rates of AW diagnosis. Although there are studies on various CSF biomarkers for the diagnosis of iNPH, the relationship between AW and these biomarkers has yet to be stud-

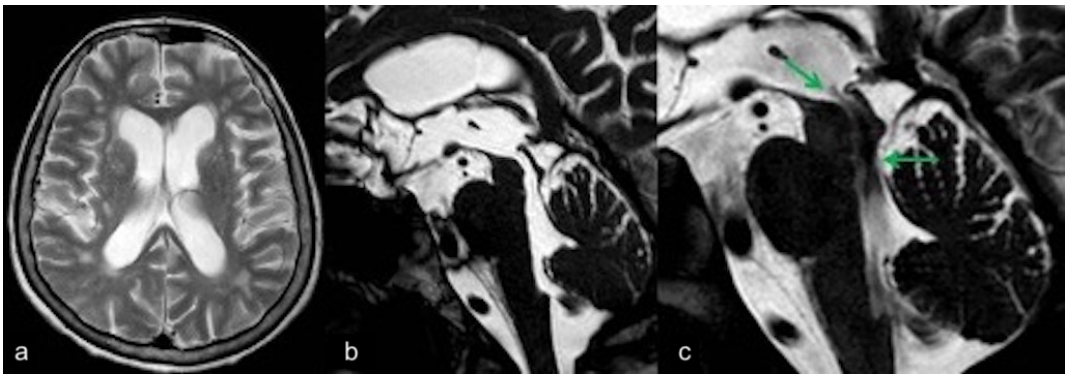

Fig. 2. CSF Flow MRI image of a patient with iNPH. (a) T2-weighted image shows chronic ventriculomegaly in both lateral ventricles. (b) No obstruction is seen at the aqueductal level in the 3D Balanced Heavy T2 image obtained for morphological evaluation. (c) Flow void reflecting rapid CSF flow at the aqueductal level (arrows) in flow-sensitive T2 DRIVE image.

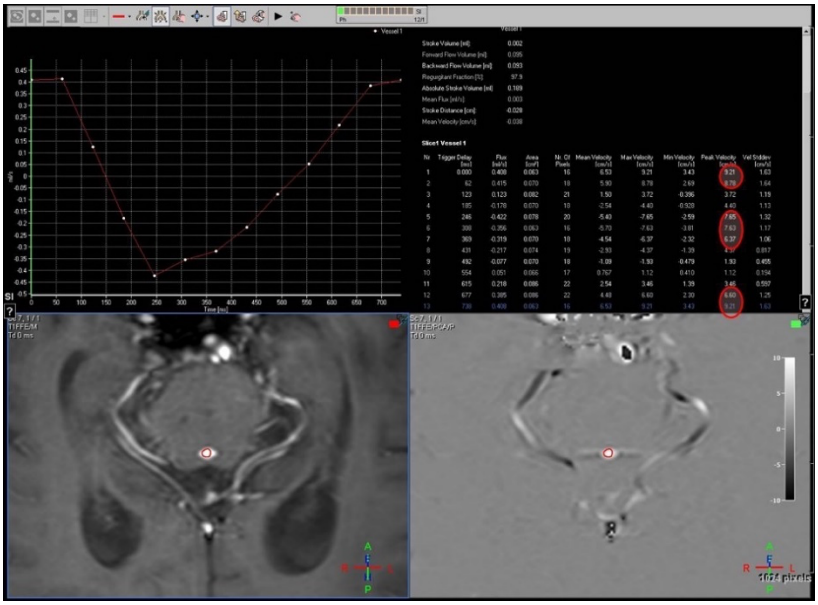

Fig. 3. Increase in expected peak systolic velocities (normally 4-6 cm/sec) in quantitative transverse phase contrast imaging obtained from the aqueductal level of a patient with iNPH (round inside).

ied [14]. Therefore, comparing patients with radiologically differentiated AW or iNPH will contribute to the literature in the future.

Systemic diseases such as hypertension, diabetes mellitus, hyperlipidemia, ischemic heart disease, atherosclerotic cerebrovascular disease, and peripheral arterial disease may cause additional morbidity in patients with iNPH. At the same time, neurodegenerative diseases including Alzheimer's and Parkinson's disease as well as cerebrovascular white matter diseases may be observed together with iNPH [4, 11, 16, 28]. These comorbid diseases may increase the severity of iNPH symptoms. The presence of concomitant neurologic disorders may also affect recovery and long-term prognosis after treatment with shunt placement as well [4].

In our study, comorbid diseases were more common in patients with iNPH, a disease of advanced age [1, 4, 8, 9] compared to the group of AW patients, albeit with a nonstatistical significance (Table 1).

Ventriculoperitoneal shunt surgery remains current in the surgical treatment of iNPH [1, 3, 4, 16, 26, 29]. In cases with AW, the membranous structure at the aqueductal level can be removed by endoscopic methods [30]. Surgical complications and recurrences may be seen in fiberoptic ventriculoscopy, endoscopic third ventriculostomy, and posterior transcranial approaches used in the treatment of AW [15, 16, 30]. In addition, endoscopic applications require good anatomical knowledge and experience. The learning process is long and technically requires neurological surgical equipment that is not available in all centers. For this reason, shunt surgery for AW can be applied as an alternative option to other surgical methods. Ventriculoperitoneal shunt surgery is technically easier to perform and more noninvasive. In addition, patients with AW are younger and usually have fewer comorbid diseases. From this point of view, a higher surgical performance can be expected in shunt surgery in patients with AW patients.

With the advancements in technology, the use of shunt systems with programmable pressure valves has proven to be successful in the treatment of hydrocephalus [3, 29]. Skalicky *et al*. [3] has reported that, compared to fixed pressure shunts, the use of programmable valves results with significantly lower rates of revision surgeries. Early and late complications following shunt surgery in the treatment of iNPH have been reported in the literature [4, 16]. In a recent meta-analysis, Giordian *et al*. [16] has reported that the development of subdural effusion requiring drainage was the most common complication followed by infections after VPS surgery for iNPH.

In our study, we used VPSs with programmable

valves in all patients with iNPH and AW. Using a ventriculoperitoneal shunt with a programmable valve gave us advantages in preventing the development of subdural effusion while closely monitoring clinical improvement. During the follow-up period for patients with iNPH and AW who underwent VPS surgery of 42.4 ± 30.2 months vs. 29 ± 20.4 months, respectively chronic hydrocephalus symptoms regressed in 88.5% of iNPH patients and 66.7% of AW patients (Table 3). Statistically, we did not observe a difference between the two groups in terms of response rate to the shunt surgery. During outpatient follow-ups, shunt settings were adjusted in almost half of the patients in both the iNPH and AW group (53.8% vs. 46.7%, respectively) due to the occurrence of subdural effusion. Yet, some patients required 19.2% vs. 13.3%) burr hole drainage as shunt pressure adjustment failed to manage the effusion. One patient per group underwent shunt revision due to malfunction in the present series. (Table 2). According to our results, these two groups of patients did not show a difference in the rate of development of subdural effusion after shunting as well as the rate of shunt revisions. In addition, the low need for revision surgery due to the use of shunts with programmable valves was consistent with the literature. The rates of infection in the present series (2.4%) were similar to the literature [1, 16].

Limitations

There are some limitations of this study. First, the study was not double-blinded. Due to the retrospective design of the study, only approximately 80% of patients had objective postoperative measures of gait and cognition available. For the remaining patients, scores were obtained by interviewing the patients and/or their caregivers. Second, the group size was small, reflecting both the relatively low incidence of NPH and the smaller subgroup of complex NPH within the overall disease spectrum. Third, this was a single-center study. Studies with better design and more participants are needed to confirm our findings.

CONCLUSION

iNPH and AW display similar clinical presentation although the underlying pathophysiological mechanisms are largely different, and they occur at different age groups. The placement of VPS is a widely accepted treatment of iNPH. Based on the similar clinical response and postoperative success rates, we advocate VPS surgery in the management of AW, which seems a more advantageous technique considering the technical difficulties of other surgical methods.

Authors' Contribution

Study Conception: AT, PEO; Study Design: AT, PEO; Supervision: AB; Funding: N/A; Materials: AT, AB; Data Collection and/or Processing: AA, OA, BS; Statistical Analysis and/or Data Interpretation: AT, PEO, RÖ; Literature Review: OA, BS, RÖ; Manuscript Preparation: AT, PEO, OA, BS, RÖ and Critical Review: PEO, AB.

Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

Financing

The authors disclosed that they did not receive any grant during conduction or writing of this study.

Acknowledgment

The abstract of this paper was presented as oral presentation at 7th Congress of the European Academy of Neurology- Virtual 2021, June 19-22.

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