Review Article

Tetraventricular noncommunicating hydrocephalus: Case report and literature review

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ABSTRACT

Background: Tetraventricular hydrocephalus is a common presentation of communicating hydrocephalus. Conversely, cases with noncommunicating etiology impose a diagnostic challenge and are often neglected and underdiagnosed. Herein, we present a review of literature for clinical, diagnostic, and surgical aspects regarding noncommunicating tetrahydrocephalus caused by primary fourth ventricle outlet obstruction (FVOO), illustrating with a case from our service.

Methods: We performed a research on PubMed database crossing the terms "FVOO," "tetraventriculomegaly," and "hydrocephalus" in English. Fifteen articles (a total of 34 cases of primary FVOO) matched our criteria and were, therefore, included in this study besides our own case.

Results: Most cases presented in adulthood (47%), equally divided between male and female. Clinical presentation was unspecific, commonly including headache, nausea, and dizziness as symptoms (35.29%, 21.57%, and 9.80%, respectively), with ataxic gait (65%) and papilledema (40%) being the most frequent signs. MRI and CT were the imaging modalities of choice (11 patients each), often associated with CSF flow studies, such as cine MRI and CT ventriculogram. Endoscopic third ventriculostomy (ETV) was both the most popular and effective surgical approach (50.85% of cases, with 18.91% of recurrence) followed by ventricle-peritoneal shunt (16.95% of patients, 23.0% of recurrence).

Conclusion: FVOO stands for a poorly understood etiology of noncommunicating tetrahydrocephalus. With the use of ETV, these cases, once hopeless, had its morbimortality and recurrence reduced greatly. Therefore, its suspicion and differentiation from other forms of tetrahydrocephalus can improve its natural course, reinforcing the importance of its acknowledgment.

Keywords: Fourth ventricle outlet obstruction, Hydrocephalus, Tetraventriculomegaly

INTRODUCTION

Tetraventricular hydrocephalus is a common presentation of communicating hydrocephalus. Detecting, among tetraventricular hydrocephalus, cases with noncommunicating etiology may open the possibility of different surgical treatment strategies other than performing ventricular shunting. Herein, we report a challenging case of tetraventricular noncommunicating hydrocephalus caused by primary fourth ventricle outlets obstruction (FVOO) that had undergone multiple shunt revisions. Furthermore, we offer a review of

literature searching for clinical and imaging clues for early diagnosis.
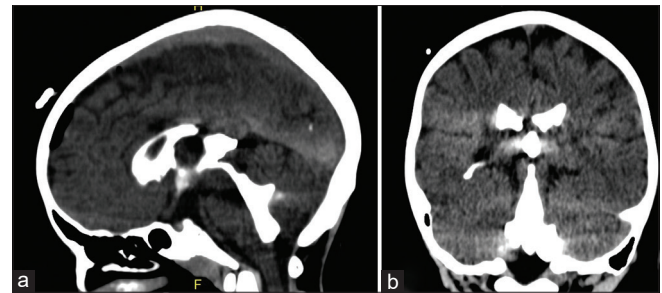
CASE REPORT

A 7 year-old male patient with tetraventricular hydrocephalus presenting learning difficulties had been submitted to an implantation of a programmable ventricle-peritoneal shunt (VPS), Sophysa®, in October 2014. In January 2015, he was readmitted to the hospital presenting abdominal pain, vomits, and VI and VII cranial nerve palsy. MRI showed tetraventricular enlargement. The VPS was revised but no signs of infection or obstruction were detected. The VPS was then replaced for a medium pressure valve. Two weeks later, under a new onset of intracranial hypertension (ICH), the VPS was removed and an external ventricular drainage (EVD) was implanted. The patient persisted somnolent, with apathy and intense nausea during the 1st days postsurgery and after stabilization of the neurological status, a low-pressure ventriculoatrial shunt (VAS) was installed in March 2015. Three months later, a VAS revision was necessary due to obstruction of the proximal catheter that was attached to the choroid plexus. An EVD was implanted. After VAS revision, the EVD was closed for 3 days, then removed, and the patient was discharged from the hospital. On the next day, the patient returned to the hospital with symptoms and signs of ICH. The former neurosurgeon contacted our neurosurgical team, and the patient was transferred to our neurosurgical service. He was admitted in July 2015 presenting a decreased level of consciousness (GCS = 12), vomiting, and bradycardia. After a new EVD procedure, intracranial pressure was monitored for 3 days and recorded within normal values. The neurological status improved and the EVD was withdrawn. Two days later, he presented an episode of apparent tonic–clonic seizure, accompanied by bradycardia, arterial hypotension, vomiting, and became aphasic. The previous implanted VAS was withdrawn and an EVD was reimplanted. As we noticed that the MRI showed third ventricular dilation with downward bulging of its floor [Figure 1], a typical characteristic of noncommunicating hydrocephalus, we decided to perform a CT ventriculography that showed a complete FVOO [Figure 2]. An endoscopic third ventriculostomy (ETV) was then successfully performed. Intracranial pressure monitoring after surgery showed 48 h of ICH and then normalized. The clinical and neurological status became progressively better, and no more vomits or seizures were observed. The patient was discharged from the hospital after 10 days. He is still nowadays shunt free.

DISCUSSION

There are only a few cases reported, each one presenting different and variable signs and symptoms, and equally variable treatment strategies to obtain successful
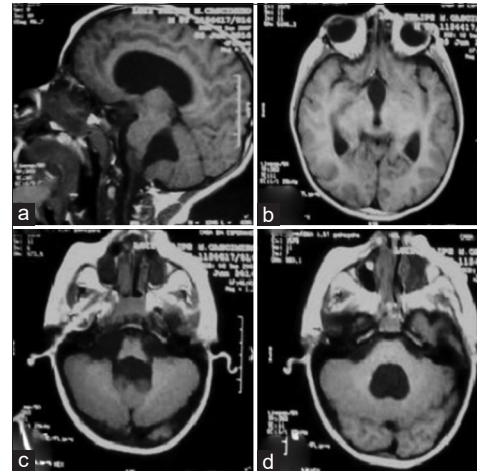
Figure 1: Sagittal (a) and axial (b-d) MRI scans showing tetrahydrocephalus and the third ventricular floor downward bulging, which can be suggestive of noncommunicating hydrocephalus.

Figure 2: Sagittal (a) and coronal (b) CT ventriculography showing the flow obstruction of contrast circulation ate the level of the fourth ventricle apertures.

outcomes.[20,25] We report a case of primary FVOO misdiagnosed as communicating hydrocephalus and provide a literature review to clarify clinical clues for the diagnosis and best treatment strategies for this condition.

FVOO is an uncommon situation usually associated with posterior fossa congenital malformations (i.e. Dandy–Walker syndrome and Chiari malformation); tuberous sclerosis; infection (i.e. meningoencephalitis and arachnoiditis); trauma; postsurgical adhesions; or tumors.[7,11,20,25,26,28,38]

Background

Dandy was probably the first to describe in paper, in early 1920s, FVOO as a cause of tetrahydrocephalus. In a time, while a great proportion of the scientific community was still debating whether fourth ventricle apertures (Luschka's and Magendie's foramina) were naturally present or simply artefact of dissection and whether the ventricle system had a communication or worked isolated from the subarachnoid space,[12] Dandy described a series of patients which

presented with tetrahydrocephalus and did not show, in his own recently developed pneumoencephalogram, airflow from the fourth ventricle to cisterna magna, suggesting an obstructive etiology, posteriorly confirmed during surgery or necropsy.

Dandy was also perceptive in observing that even though FVOO was an uncommon cause of noncommunicating hydrocephalus and usually secondary, there was also another group of patients that shared a primary, or idiopathic, etiology, when it could not be linked to another associated pathology.

Later, in 1958, Holland and Graham[19] described what seemed to be a distinct case of congenital atresia of the foramina of Luschka and Magendie in a case report that did not reveal any other malformation that could contribute to the diagnosis of Dandy–Walker syndrome, besides the foraminal atresia and the hydrocephalus itself. During the investigation, a cystic structure on the cerebellopontine angle, continuous with the lateral recesses and the fourth ventricle, was discovered and thought to be a result of chronic intraventricular hypertension. The natural assumption of a secondary fibrosis of the foramina as the cause of this condition was dismissed by the patient's negative clinical history and by the absence of inflammatory signs during the surgery. Thus, it advocated Dandy's theory that FVOO could also, in fact, be a primary condition.

After many years, Amacher and Page,[1] in 1971, published a series of four cases that shared similarities with the case described by Holland and Graham.[19] Amacher and Page revealed that, after craniotomy, a thin membrane was found to be the cause of the CSF flow disturbance. That membrane, it is important to say, had already been observed by Holland and Graham and would be described by Rifkinson-Mann *et al.* in 1987.[32] The summary was the postulation that the late onset of the symptoms observed in a group of patients who presented this condition would possibly take place due to the "porosity" or semipermeability of the membrane wall covering the foramina. It would allow a minimum flow through the aperture, enough to be compatible with life, but causing the chronic hypertension responsible for the dilation of the ventricular system and the hydrocephalus.[10]

Physiopathology

The most solid theory in vogue in the matter so far postulates that the development of semipermeable membranes obstructing the foramina could be responsible for the condition. Theoretically, the intermittent CSF flow would allow good clinical tolerance for some time, maybe even decades, until some event, such as hemorrhage, meningitis, and arachnoiditis, would lead to a decompensated permeability, and, following it, CSF accumulation upstream.[32] Yet, supposing none of those events came to reality, which

would be the case of the subject of this study, fibrosis itself could progressively decrease the permeability of the foramina, resulting in a more chronic process of hydrocephalus, or even the complete and acute CSF obstruction after the membrane ultimately adheres to the dura by progressive hydrocephalus. The contact between the foramina of Magendie and Luschka, respectively, with the suboccipital and the petrosal dura, gives rise to a definite arachnoidal membranous organization, with disappearance of CSF due to progressive tetraventricular and a downward displacement of the posterior fossa structures (fourth ventricle, tonsil herniation, and blockage of the foramen magnum). This is a mechanism of functional blockage that may be similar to the outlet occlusion that is observed in the Chiari malformation: displacement of the cerebellar tonsils leads the outlets to face the dura, reducing the physiological CSF flow and dilating the fourth ventricle.[28,33] That could, therefore, explain how a group of patients present with acute onset of hydrocephalus and ICH, while others develop a much more chronic course, with a slow instead of rapid progressing ICH.

CASE REVIEW OF LITERATURE

Inclusion criteria

We performed a research on PubMed database crossing the terms "FVOO," "tetraventriculomegaly," and "hydrocephalus" in English. Inclusion criteria included cases of tetrahydrocephalus without a concomitant malformation or other secondary cause that could correspond to a confounding factor to primary or idiopathic FVOO. Excluding criteria were secondary or etiologic nonspecified cases, communicating hydrocephalus or case reports that did not provide enough information to allow differentiation of the clinical course, radiological findings, treatment of choice, and outcomes between primary and secondary etiologies. Fifteen articles (a total of 34 cases of primary FVOO) matched our criteria and were, therefore, included in this study besides our own case.

Epidemiological and clinical findings

In our series of 35 cases [Table 1], 34 had their age described in paper (mínium 1 month old, maximum 73 years old, mean age 31.87 years), with most cases presenting in adulthood (a total of 16/34 cases or 47%) followed by childhood (with 13/34 cases – 38.3%) and elderhood (with 5/34 cases – 14.7%). Gender was balanced between this patients, with a 1:1 male: female ratio among the 34 cases, showing no sex predominance.

When it comes to this entity, clinical signs and symptoms are variable and little can offer to diagnostic confirmation. The most common symptoms observed were headache,

Table 1: Part 1: Case reports found in English literature that matched our criteria.

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Holland and Graham, 1957[19]	1	31 yo	Male	OFV	Pneumoencephalography	Symmetrical dilation of the entire ventricular system and absence of air flow over the cerebral hemispheres	Failure: recurrence (new FV was done) and death.	16 months	Autopsy: thin membrane over the roof of the fourth. Bilateral cysts emerging from CPA with approximately 2 cm in diameter continuous with Luschka foramina and FV. Foramina of Magendie not identified.
Amacher and Page, 1971[1]	1	13 yo	Female	OFV	Pneumoencephalography	Gross, symmetrical hydrocephalus above a very large FV which was normal in position, no air passing beyond it	Failure: death (cause not described).	18 months	Cisterna magna was small and the cerebellum appeared normal. After separating the tonsils, a membrane was found to be the cause of the oclusion of foramina of Magendie
	2	5 mo	Female	OFV	Pneumoencephalography	Severe dilation of the entire ventricular system with no air passing out of the FV	Failure: recurrence (reexploration followed by ventriculo-cervical and lumbar-ureteral shunt was done) and death (meningitis).	3.5 years	Moderately large cisterna magna with clear CSF, no evidence of previous infection or hemorrhage, a thin membrane above the foramen of Magendie
	3	8 yo	Male	OFV	Pneumoencephalography	Large and high FV with no air passing beyond it	Failure: recurrence (VPS was done).	2 years	A transparent membrane across the lower FV and another near the aqueduct

(*Contd...*)

Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
	4	21 yo	Female	OFV	Pneumoencephalography	Appeared to show a lobulated mass obstructing the lower FV	Successful.	1 year	A translucent membrane above the foramen of Magendie totally obstructing the lower portion of the FV.
Rifkinson-Mann et al., 1987[32]	1	42 yo	Male	OFV	CT ventriculography	Enlarged FV with associated hidrocephalus	Successful.	1 year	A transparent membrane in the area of the foramen of Magendie without signs of infection
	2	52 yo	Male	OFV	CT ventriculography	Nonenhancing hypodense cyst assumed to be an enlarged FV. Moderate hydrocephalus of lateral and third ventricles. Little flow to the subarachnoid space	Successful.	1 year at least	The midline area was covered by a sheet, extending from the inferior medullary velum
Mohanty et al., 1999[29]	1	32 yo	Male	ETV	CT ventriculography	Dilation of all ventricles with disproportionate dilation and ballooning of the third and FVs. No contrast in basal cisterns. Significant periventricular lucency	Successful.	2 years	-
	2	20 yo	Male	ETV	CT	Dilation of all ventricles with periventricular lucency	Successful.	15 months	-
	3	45 yo	Female	ETV	CT/MRI	Dilation of all ventricles	Successful.	12 months.	-

(Contd...)

Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Huang *et al.*, 2001[20]	1	15 yo	Female	OFV	CT/MRI	CT: enlargement of the entire ventricular system. MRI: which revealed a dilated FV with a membranous-like obstruction in the region of the foramen of Magendie	Successful.	14 months	A membrane was seen covering the foramen of Magendie
Carpentier *et al.*, 2001[8]	1	58 yo	Female	ETV	MRI	Increased ventricular dilation, downward displacement of the tonsils without true herniation, enlarged Sylvian aqueduct with increased CSF flow. No flow shown through Luschka and Magendie foramina.	Successful.	3 years	-
Suehiro *et al.*, 2002[37]	1	9 mo	Male	ETV	CT/CT ventriculography/ MRI/cine MRI	CT: dilated ventricles and a bilateral low-density space in CP angle. MRI: dilated ventricles and bilateral cysts in CP angle, periventricular T2 hyperintensity. Cine MRI: absence of flow through Magendie foramina. CT ventriculography showed no communication between the ventricles and the subarachnoid space.	Failure: procedure could not be performed (VPS was done).	3 months	-

(Contd...)

Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Karachi *et al.,* 2003[25]	1	21 yo	Female	ETV	MRI	Tetrahydrocephalus with important FV enlargement with its lateral apertures dilated into CPA, parenquimal structures were normal. Good patency of aqueduct and obstruction of FV outflow. MRI: asynchronous CSF velocities measured in the FV and cisterna magna.	Successful.	26 months	-
	2	53 yo	Female	ETV	MR/CT ventriculography	Tetrahydrocephalus with important FV enlargement with its lateral apertures dilated into CPA, parenquimal structures were normal. Good patency of aqueduct and obstruction of FV outflow. Ventriculography: stenosis and dilation of the foramina of Luschka and Magendie with no passage of contrast agent from the cisterna magna	Successful.	24 months	-

(Contd...)

Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
	3	68 yo	Male	ETV	MRI/CT ventriculography	Tetrahydrocephalus with important FV enlargement with its lateral apertures dilated into CPA, parenquimal structures were normal. Good patency of aqueduct and obstruction of FV outflow. MRI: asynchronous CSF velocities measured in the FV and cisterna magna. Ventriculography: stenosis and dilation of the foramina of Luschka and Magendie with no passage of contrast agent from the cisterna magna	Successful.	58 months	-

FV: Fourth ventricle, ETV: Endoscopic third ventriculostomy, VPS: Ventricle-peritoneal shunt, OFV: Open fourth ventriculostomy

(Contd...)

Table 1: Part 2: Case reports found in english literature that matched our criteria.

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Longatti et al., 2009[28]	1	53 yo	Male	ETV	CTI+MRI: 7 patients, Cine MRI: 8 patients	CT+MRI: obstructive tetraventricular hydrocephalus, with downward displacement of the third ventricle floor (four patients), obliteration of the retrocerebellar CSF space (six patients), slight displacement of the cerebellar tonsils without herniation (two patients). In four patients, the interpeduncular and prepontine cisterns were significantly compressed and reduced in volume by the abnormally dilated fourth ventricle. Cine MRI: in complete absence of CSF flux at the obex level (three patients); CSF flow uncertain (two patients); normal flow at the obex (three patients). Closure of the fourth ventricle outlets was suggested by the cine MRI study in 62.5% of cases.	Failure: recurrence (new ETV was done).	12 years	Reduced size of Magendie foramina wich was covered by a membrane. Only the left Luschka foramina were visible.
	2	49 yo	Female	ETV			Successful.	7 years	Reduced size of Magendie foramina wich was covered by a membrane. Both Luschka foramina were visible.
	3	70 yo	Male	ETV			Failure: recurrence (VPS was done).	6 years	Reduced size of Magendie foramina wich was covered by a membrane. None of Luschka foramina were visible.
	4	73 yo	Male	ETV+Aqueductoplasty			Successful.	6 years	Reduced size of Magendie foramina wich was covered by a membrane. Only the left Luschka foramina was visible.
	5	36 yo	Male	ETV			Successful.	2 months	-
	6	65 yo	Female	EFV			Successful.	34 months	Reduced size of Magendie foramina wich was covered by a membrane. Both Luschka foramina were visible.
	7	58 yo	Male	ETV			Successful.	2 months	Reduced size of Magendie foramina wich was covered by a membrane. Both Luschka foramina were visible.

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Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Giannetti *et al.,* 2011[16]	1	8 yo	Female	Endoscopic occipital fourth ventriculostomy (previous VPS)	MRI	Marked enlargement of the fourth ventricle, significant dilation of the lateral recesses of the fourth ventricle with bulging into the cerebellopontine angle and patent cerebral aqueduct.	Successful.	4 years	Fine translucent membranes were observed, which occluded the foramina.
Roth *et al.,* 2012[33]	1	Not described	Not described	ETV	MRI/CT ventriculography	Tetraventricular dilation, out-pouching of the Luschka foramina, ballooning of the fourth ventricle, and lack of flow artifact at the outlet of the fourth ventricle	Successful.	26 months	-
Torres-Corzo *et al.,* 2014[40]	1	1 mo	Female	ETV, EFV	Not described	Not described	Successful.	24 months	-
	2	4 mo	Male	ETV, EFV	Not described	Not described	Successful.	30 months	-
	3	1 yo	Female	ETV, EFV	Not described	Not described	Failure: recurrence (Aqueductoplasty was done).	26 months	-
	4	12 yo	Female	ETV, EFV	Not described	Not described	Successful.	24 months	-
	5	46 yo	Female	ETV, EFV	Not described	Not described	Successful.	24 months	-
	6	48 yo	Female	ETV, EFV	Not described	Not described	Failure: recurrence (Aqueductoplasty was done).	28 months	-
Hashimoto *et al.,* 2014[18]	1	1 yo	Male	ETV	CT/MRI/ Cine MRI/CT ventriculography	CT: tetrahydrocephalus. MRI: tetrahydrocephalus, fourth ventricular outlet obstruction, a visible membranous obstacle at the foramen of Magendie.	Failure: recurrence (new ETV was done).	Not described.	-

(Contd...)

Table 1: *(Continued)*

Author	Case	Age	Gender	Surgery technique	Imaging modality	Imaging findings	Outcomes	Follow-up	Surgical findings
Ishi *et al.*, 2015[23]	1	3 yo	Male	ETV	MRI/CT ventriculography	MRI: enlargement of all ventricular systems associated with the dilatation of the foramina of Magendie and Luschka/CT ventriculography: contrast was accumulated in the foramina of Magendie	Failure: recurrence (new ETV was done).	20 months	-
Shimoda *et al.*, 2017[34]	1	66 yo	Female	VPS	MRI/CT	Tetraventriculomegaly and CSF flow obstruction at the aqueduct, foramen of Magendie, and bilateral foramina of Luschka	Failure: recurrence (ETV was done).	Not described	-
Rosa *et al.*	1	7 yo	Male	ETV (2 previous VPS and 1 previous VAS)	MRI/CT ventriculography	Tetrahydrocephalus with fourth ventricle outlet obstruction	Successful.	6 years	-

ETV: Endoscopic third ventriculostomy, EFV: Endoscopic fourth ventriculostomy, VPS: Ventricle-peritoneal shunt

nausea and vomiting, dizziness, vision disturbance, and incontinence [Table 2]. Ataxic gait and papilledema were the most observed clinical signs followed by ideomotor slowdown and increased head circumference [Table 3]. It becomes clear, therefore, that the clinical presentation does not differ from other forms of hydrocephalus, being somewhat typical of increased CSF pressure and cerebellar compression, and incapable of pointing to a definite etiology.

Imaging diagnosis

It is easy to identify dilation or augmented CSF collection in the fourth ventricle as a characteristic radiological finding in cases of FVOO.[20,25,33,28] However, the unquestionable presence of a membranous obstruction, even with high-quality MRI sequences, in the absence of other obstructive images, is much harder to prove.[25,28,29] In this scenario, opinions diverge in the most recent publications. Karachi *et al.* argued that the confirmation of membranous obstacles would not be necessarily required for the diagnosis of this condition, as obstruction of Magendie's and Luschka's foramen can be confirmed using options such as ventriculography and/or MR flow images. Others stand by a direct exploration of the fourth ventricle, whenever possible, using a flexible endoscope through an open and wide aqueduct.[28,31] Longatti *et al.* postulated that this would be a method of absolute sensitivity, opposing to 62.5% sensitivity in the cine MRI. Oertel *et al.*,[31] on the other hand, still defended that CT ventriculography with injection of contrast medium through a ventricular catheter would be the most sensitive method, based on the fact that serial CT images after injection show collected contrast medium in the outlets of the fourth ventricle and subsequent interruption of its diffusion to the prepontine cistern and cervical subarachnoid space. Here, some precaution should be taken, especially in pediatric patients, because of the radiation exposure. It would be sensible to selectively choose children presenting certain backgrounds, such as patients with inconsistent MRI findings suggestive of an obstructive etiology, for example, the ballooning of the third ventricle with downward bowing of the third ventricular floor and tetraventriculomegaly with a flow signal of the aqueduct and low or no flow at the fourth ventricular outlet. If a patient presents with a large and open fontanel, ventriculography would be rather easily performed; otherwise, a cisternography through lumbar puncture (LP) could be considered.[33] Although LP is contraindicated in obstructive hydrocephalus, the use of a smaller needle to aspirate a small volume of CSF and the maintenance of the patient in recumbent position following the procedure could prevent pressure gradient an morbidity justifying the validation of the technique. Finally, some authors have demonstrated the validity of MRI ventriculography following ventricular injection of gadolinium as a safe and effective diagnostic method, in alternative to the CT ventriculography,

Table 2: Clinical symptoms by the time of admission.

Symptoms in admission	Number of patients	Percentage
Headache	18	35.29
Nausea	11	21.57
Dizziness	5	9.80
Vision alteration	4	7.84
Incontinence	4	7.84
Dysesthesias	2	3.92
Hemiparesis	2	3.92
Memory impairment	2	3.92
Cervical pain	1	1.96
Depression	1	1.96
Amenorrhea	1	1.96
Total	51	100.00

Table 3: Clinical signs by the time of admission.

Signs in admission	Number of patients	Percentage
Ataxic gait	13	65.00
Papilledema	8	40.00
Increasing head circumference	4	20.00
Ideomotor slowdown	4	20.00
Lateralized cerebellar signs	3	15.00
Altered state of consciousness	3	15.00
Postural instability	2	10.00
Behavioral issues	2	10.00
Hyperreflexia	2	10.00
Seizures	2	10.00
Worsening in school performance	2	10.00
Nystagmus	1	5.00
Dysdiadokokinesis	1	5.00
Hyporeflexia	1	5.00
Dehydration	1	5.00
Aphasia	1	5.00
Bradycardia	1	5.00
Total	20	100.00

suggesting that it should be more widely implemented in the routine of investigation, preventing unnecessary radiation exposure.[24,31]

In our series, 47 different radiological examinations were used in the 35 patients, with MRI and CT being the most commonly observed (11 patients each), followed by CT ventriculography and cine MRI (10 patients each), and pneumoencephalography, due its historical role in diagnosing such patients (five cases). Perhaps, because of its widely availability and fast technique, CT is still frequently used as a first radiological examination in these cases, even though it does not seal the FVOO as the etiology for hydrocephalus, once the patients usually arrive without any previous examination or diagnosis and in poor general condition. When available, MRI and cine MRI are commonly used as

Surgical techniques

The first surgical approaches of primary FVOO were focused on the logical idea that if obstruction of the foramina was the cause of the condition, then restoring the physiological pathway would be the natural choice.[19] Holland and Graham in 1958 performed a posterior craniotomy and a wide opening of the membrane, reestablishing the flow between the fourth ventricle and the subarachnoid space. This technique was repeated in Amacher and Page's series[1] in 1971. Even though patients initially improved with reduction of ICH and reacquiring normal ventricles sizes, this technique was followed by frequent recurrence of the hydrocephalus.[9,39]

Contemporarily, shunting techniques took place and became a preferable method in hydrocephalus cases, especially the communicating forms. The first types of shunts, very popular in the past, such as the ventriculoureteral shunting in the 1950s,[22] were replaced by either peritoneal or atrial shunting. These are standard techniques used nowadays to allow the drainage of the CSF directly to the peritoneum or atrium, reducing the intracranial pressure without the need of reabsorption, which is usually impaired in communicating hydrocephalus.

The use of shunts, however, is also associated with numerous complications, once it involves the maintenance of a functioning catheter connecting the subarachnoid space and the peritoneum or atrium. The obstruction and malfunction of the catheter can lead to recurrence of the hydrocephalus and the need to reassess the procedure; overdrainage can lead to headache and intracranial bleeding;[5,6] its presence itself could lead to infection (meningitis, endocarditis, and peritonitis),[15,30,35] migration (ventriculoperitoneal shunting was associated with catheter migration to and/or perforation of both hollow and solid viscera and genitourinary tract),[3,4,14,17] and local complications (ascites, pseudocysts, intestinal and urinary tract obstruction, and inguinal hernia, if peritoneal; cardiac arrhythmias, thrombosis, and further embolization, if atrial).[2,5,6,20,21,34,41] In addition, the FVOO, such as other forms of noncommunicating hydrocephalus, has a CSF reabsorption maintained, while the flow through the ventricles is compromised. Consequently, shunting may not be the best indicated approach taking into consideration the possible complications inherent to these procedures and the pathophysiology of the condition that diverts completely from the communicating forms of hydrocephalus.

Some decades after, in the 1990s, Chai[9] suggested a new type of fourth ventricular-cisternostomy, aiming to prevent recurrence of the obstruction: they used a polyethylene tube to maintain the communication between the fourth ventricle and the subarachnoid space. Unfortunately, in the 26-year follow-up, two out of the 12 patients in the study died because of malfunction of the tubes. This outcome, along with the possible complications of the use of an intracranial catheter, leads to restriction and later absolut interruption in research studies regarding this technique.

It was Mohanty *et al.* in 1999[29] that first used the ETV, in a series of three cases, for the treatment of FVOO. Its use, until then, had been restricted to obstructive cases of hydrocephalus due to aqueductal stenosis, with FVOO being managed by shunts, as in communicating forms of hydrocephalus. This technique, a more secure and less invasive procedure, is defended by several references in literature[8,13,18,23,28,31,37,40] and received support by personalities as Kulkarni *et al.*[27] and Spennato *et al.*[36] who postulate that, when compared to shunts, ETV lacks problems in the areas of disconnection, occlusion, high infection rate, overdrainage and valve dysfunction, thrombosis, and migration [Table 4].

As a final consideration, it is important to bring up the newest approach in vogue: the transaqueductal endoscopic fourth ventriculostomy (EFV).[16] Here, we stand before a more bold and edgy technique that allows opening of all three ventricular exits, regardless of the anatomy and dilation of the supratentorial ventricles; permits a more satisfying control of bleedings, especially if irrigation is necessary, among other advantages, when compared to ETV. With the use of a flexible endoscope, the fourth ventricle is approached safely and a direct exploration of ventricle anatomy and its apertures is possible. However, it is important to reinforce that ETV is more secure and, for this reason, EFV could be considered a better option only in cases when ETV is not feasible, since the additional manipulation of the third ventricle, mesencephalic aqueduct, and fourth ventricle

Table 4: Procedures used for treating the 35 patients.

Procedure	Number of patients	Percentage
Endoscopic third ventriculostomy	30	50.85
Shunt	10	16.95
Open fourth ventriculostomy	8	13.56
Endoscopic fourth ventriculostomy	7	11.86
Aqueductplasty	3	5.08
Endoscopic occipital fourth ventriculostomy	1	1.69
Total	59	100.00

cannot undergo without also increased risk of local damage, and, consequently, sequelae.

CONCLUSION

FVOO stands for a neglected etiology of noncommunicating tetrahydrocephalus although the first reports of this entity can be traced to the early 20th century.

The clinical features are not specific and do not allow its differentiation from other forms of hydrocephalus. However, with modern techniques of diagnosis, specially MRI, we can rely on noninvasive and highly elucidative examinations for its recognition, once there's a suspicion.

Several surgical techniques were once used to treat the condition. Initially, open exploration of posterior fossa was performed, with poor, unsatisfactory results. VPS was also very frustrating as an approach of choice in these patients. Only after the addition of endoscopic techniques to the surgical repertoire, these cases, once apparently hopeless, presented a reproducible improvement in morbidity and mortality, with ETV being considered the surgery of choice, followed, when not feasible, by EFV.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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