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The entity of the trapped fourth ventricle: A review of its history, pathophysiology, and treatment options

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Abstract:

An isolated or trapped fourth ventricle is a relatively rare, although serious, adverse effect of hemorrhagic, infectious, or inflammatory processes that involve the central nervous system. This entity usually occurs after successful shunting of the lateral ventricles and may become clinically evident with the development of delayed clinical deterioration. This decline of the neurological status of the patient is evident after an initial period of improvement of the relevant symptoms. Surgical treatment options include cerebrospinal fluid shunting procedures, along with open surgical and endoscopic approaches. Complications related to its management are common and are related with obstruction of the fourth ventricular catheter, along with cranial nerve or brainstem dysfunction. We used the keywords: "isolated fourth ventricle," and "trapped fourth ventricle," in PubMed® and Web of Science®. Treatment of the trapped fourth ventricle remains a surgical challenge, although the neurosurgical treatment armamentarium has broadened. However, prompt recognition of the clinical and neurological findings that accompany any individual patient, in conjunction with the relevant imaging findings, is mandatory to organize our treatment plan on an individual basis. The current experience suggests that any individual intervention plan should be mainly based on the underlying pathological substrate of hydrocephalus. This could help us to preserve the patient's life, on an emergent basis, as well as to ensure an uneventful neurological outcome, maintaining at least the preexisting level of neurological function.

Keywords:

Endoscopic procedures, fourth ventricular shunting, trapped fourth ventricle

Introduction

The entity of isolated or trapped fourth ventricle represents a relatively rare, yet important, neurosurgical condition which is usually manifested in individuals who had previously been treated successfully with shunting of the lateral ventricles. It deserves special mention because of the fact that its swift diagnosis along with the adoption of an appropriate management protocol is of paramount importance for the resuscitation of the patient's life and its neurological integrity. When the clinical history of the majority of the affected individuals is reviewed, we ascertain that it is related with a severe inflammatory reaction within the ventricular system, due to hemorrhage or microbial infection.[1] After initial successful management of hydrocephalus, all the compartments of the ventricular system may become decompressed, as well as the inner walls of the aqueduct of Sylvius, which may become apposed. The combination of these anatomical alterations along with the existence of an evolving ependymal

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inflammation could be the offending mechanism for the development of intraventricular adhesions.

Attempting a brief description of the anatomical boundaries of the fourth ventricle along with a review of the cerebrospinal fluid (CSF) circulation through this anatomical structure is of paramount importance. Our basic tenet is to elucidate the pathophysiology of this entity and delve into the issue of the proposed treatment modalities.

The fourth ventricle exhibits three outlets, namely the paired foramina of Luschka laterally, and the foramen of Magendie in the midline. Anatomically, the dorsal wall of the foramen of Magendie is constituted by the caudal extension of the cerebellar hemispheres and vermis, whereas the ventral and lateral borders are formed by the obex of the medulla. The foramen of Magendie is located in the midline and normally communicates with the cisterna magna, although in some individuals, this outflow is not patent.[2] The anatomic substrate which constitutes the prerequisite for the development of the syndrome of trapped fourth ventricle (TFV) refers to obstruction of all of the fourth ventricular outlets. Namely, this refers to the level of the cerebral aqueduct proximally and distally to the foramina of Magendie and Luschka. The net result of the disrupted CSF circulation consists of progressive dilatation of the fourth ventricle. This is attributed to the ongoing CSF production by the choroid plexus of the fourth ventricle and to a ball valve, one-way mechanism (related to CSF flow) at the level of the cerebral aqueduct.

Attempting to review the wide spectrum of pathological processes which share in common the development of obstructive hydrocephalus due to occlusion of the fourth ventricular outlets, we could classify them into several subcategories [Table 1].

A wide spectrum of different inflammatory processes is implicated, most of them being related with infection and intraventricular hemorrhage (IVH).[3-5] In case of preservation of the patency of the cerebral aqueduct, retrograde flow of CSF results in enlargement of all of the supratentorial compartments of the ventricular system, without entrapment of the fourth ventricle.[6] Initial insertion of ventriculoperitoneal (VP) or ventriculoatrial (VA) shunt could allow for decompression of all the ventricular cavities. Nevertheless, as dilation of the cerebral aqueduct is relieved, apposition of the inflamed ependymal surfaces may result in adhesions which predispose to obstruction, resulting to acquired aqueductal stenosis.[7] The combination of discontinuation of retrograde flow through the aqueduct along with the already established restriction of the outflow of the fourth ventricle establishes a pathophysiological mechanism which could potentially result in the entrapment of the fourth ventricle.[2,7,8] Apart from that, the ependyma and choroid plexus of the fourth ventricle continue the unhindered production of CSF, resulting in an ongoing accumulation of CSF. The net result of this process is the progressive dilatation of the ventricular compartments.

Table 1: Conditions related to or predisposing to trapped fourth ventricle

Offending pathological mechanism	Examples
Congenital	Dandy-walker malformation
Posthemorrhagic	Subarachnoid hemorrhage
	Intraventricular hemorrhage
Infectious/postinfectious	Bacterial meningitis/ventriculitis
	Mycobacterium tuberculosis
	Fungal (e.g., candidiasis)
	Parasitical (e.g., cysticercosis)
	Ventriculoperitoneal shunt infection
Postinflammatory	Sarcoidosis
Neoplastic	Carcinomatous meningitis (leptomeningeal spread)

Enlargement of the fourth ventricle may be responsible for the development of unilateral or bilateral dysfunction of the third, fourth, seventh, and bulbar cranial nerves. In addition, it could be the offending mechanism for the evolving compression of the reticular activation system, which could be responsible for a clinical entity constituting of an altered level of consciousness and eventually coma. A recrudescence of these clinical and neurological signs may indicate the necessity of surgical treatment, which should manage the obstruction-isolation of the fourth ventricle.

The most common clinical scenario relevant to these patients is that of an infant with a history of prematurity, who developed IVH and was treated with shunt placement.[9-12] An early bibliographic report exists, which described and correlated the occurrence of acquired aqueductal stenosis in cases with known, and shunted, communicating hydrocephalus, on the basis of pneumoencephalographic findings.[7] To the best of our knowledge, Hawkins *et al*. was the first author who reported a small series of patients with cerebellar signs after an initially satisfactory response to shunt placement, a clinical scenario which strongly resembles that of postshunting TFV.[13]

The entrapment of the fourth ventricle related to posthemorrhagic hydrocephalus is a well-known and analyzed clinical entity. The frequency of TFV among premature patients suffering from IVH is relatively high (15.4%). Most patients with TFV are asymptomatic at presentation and can be managed

Table 2: Pivot table, presenting studies centered on the entity of entrapped fourth ventricle, including their relevant number of patients included, their clinical presentation, treatment modality performed, and relevant success rate of each individual treatment option

Title	Authors	Year	Number of patients	Clinical presentation	Treatment method	Results
Isolated ventricles following intraventricular hemorrhage	Theodore W, *et al*.	1982	2	Respiratory distress	Multiple shunts	All points showed clinical improvement
An isolated fourth ventricle in neurosarcoidosis: MRI findings	Volker Hesselmann, *et al*.	2002	1	Deterioration of neurological status	Ventricular drainage	Immediate clinical improvement
Trapped fourth ventricle in an adult: Radiographic findings and surgical treatment	Jerry L. Hubbard, *et al*.	1987	1	Deterioration of mental and neurological status	A bilateral suboccipital craniectomy and C-1 and C-2 laminectomies+shunt	Clinical improvement
Hydrocephalus due to idiopathic stenosis of the foramina of Magendie and Luschka. Report of three cases	C. Karachi, *et al*.	2003	3	Headache, vertigo, or gait disorders, sphincter disorders, disorders of higher functions	ETV	All 3 patients became asymptomatic during the weeks following the surgical procedure and remained stable at a mean follow-up interval of 36 months
Suboccipital endoscopic management of the entrapped fourth ventricle: Technical note	Alaa Raouf, *et al*.	2013	13		Suboccipital endoscopic trans-fourth ventricular aqueductoplasty	Short stent was used in 8 patients. Stent migration occurred in five of them. Three of these 5 patients developed posterior fossa compression manifestations due to aqueduct restenosis
Endoscopic transaqueductal or interventricular stent placement for the treatment of isolated fourth ventricle and preisolated fourth ventricle	Hideki Ogiwara, *et al*.	2013	9		Endoscopic stent placement	Three patients (33%) underwent reoperation due to obstruction of the abdominal catheter, partial occlusion of the ventricular catheter, and retraction of the fourth ventricular catheter
Endoscope-assisted placement of a multiperforated shunt catheter into the fourth ventricle through a frontal transventricular approach	Kristen Upchurch, *et al*.	2007	4		Endoscope-assisted placement of a multiperforated shunt catheter	Neurological and radiographically verified improvement in all patients
Endoscopic aqueductoplasty and interventriculostomy for the treatment of isolated fourth ventricle in children	Michael J. Fritsch, *et al*.	2004	18	Impairment of consciousness, tetraparesis, and ataxia	Endoscopic aqueductoplasty, endoscopic aqueductoplasty with a stent, endoscopic interventriculostomy (lateral ventricle or third ventricle to fourth ventricle), and endoscopic interventriculostomy with a stent	Seven patients required reoperation because of restenosis
Endoscopic cerebral aqueductoplasty: A trans-fourth ventricle approach	Jason M. Sansone, *et al*.	2005	9		Cerebral aqueductoplasty procedures through the foramen magnum trans-fourth ventricle	9/9 resolution of their preoperative symptoms, recurrent aqueductal stenosis developed in one patient twice, requiring the placement of an aqueductal stent through the same approach
Endoscopic treatment of the trapped fourth ventricle	C. Teo, *et al*.	1999	16		Shunt procedure; the next eight had endoscopic procedures	The patients with shunts appeared to have a higher than expected rate of revision (50%)

Contd...

Table 2: Contd...

Title	Authors	Year	Number of patients	Clinical presentation	Treatment method	Results
Avoiding complicated shunt systems by open fenestration of symptomatic fourth ventricular cysts associated with hydrocephalus	Alan T. Villavicencio, *et al*.	1998	6		Suboccipital craniectomy and open fenestration	5 of the 6 patients (83%) have remained asymptomatic
Endoscopic treatment of isolated fourth ventricle: Clinical and radiological outcome	Matthias Schulz, *et al*.	2012	19		Aqueductoplasty or, in cases with a supratentorially extended fourth ventricular component	27.3% of patients experienced complete resolution of presenting symptoms, whereas 68.3% demonstrated partial resolution 2; insufficient catheter placement and subdural hygroma) and a need for long-term stent revisions (*n*=3; stent retraction and shunt revision for other causes
Bilateral abducens and facial nerve palsies following fourth ventricle shunting: Two case reports	Spennato. P, *et al*.	2005	2	Bilateral abducens and facial nerve palsies	A "Y" connector, endoscopic aqueductal stenting	Clinical improvement
Cranial nerve palsies after shunting of an isolated fourth ventricle	Alexandre Simonin, *et al*.	2015	2	Abducens and facial nerve palsies	A "Y" connector	Clinical improvement
Progressive cranial nerve palsy following shunt placement in an isolated fourth ventricle	Dachling Pang, *et al*.	2005	1	Delayed, progressive palsies of the 6th, 7th, 10th, and 12th cranial nerves	Suboccipital craniectomy and C-1 laminectomy	Within 2 weeks, the sixth and seventh nerve palsies began to improve
Isolated fourth ventricle as a complication of ventricular shunting	John C. Hawkins Iii, *et al*.	1978	3	Cerebellar dysfunction combined with signs suggestive of shunt malfunction	Shunting of the fourth ventricle	All patients returned to normal function
Spectrum of the syndrome of the isolated fourth ventricle in posthemorrhagic hydrocephalus of the premature infant	H.E. James, *et al*.	1990	10	Increasing head size, fontanelle fullness, irritability difficulty with swallowing, vomiting, hypoactivity, headaches and lethargy	Fourth ventricle shunt catheter and connection to the existing shunt	Resolution of symptoms
Reversible progressive multiple cranial nerve paresis in the isolated fourth ventricle following placement of fourth ventricle shunt: Case report and review of the literature	Ravi Thakker, *et al*.	2019	1	Bilateral facial and multiple lower cranial nerve paresis with bilateral internuclear ophthalmoplegia	Nonoperatively	Completely recovered at 9 months
Feasibility of a fourth ventriculopleural shunt for diversion of an isolated fourth ventricle: A technical note	Courtney Suzanne Lewis, *et al*.	2018	1	Headaches, nausea, vomiting, and 6th nerve palsy	Fourth VPL shunt	Immediate relief of headaches and mild improvement of 6th nerve palsy. Resolution of her 6th nerve palsy, at 1-year follow-up
Stereotactically guided fourth ventriculoperitoneal shunting for the isolated fourth ventricle	Colpan M.E, *et al*.	2003	1	Vomiting, nausea, diplopia, truncal ataxia, vertical nystagmus	Stereotactically guided fourth ventriculoperitoneal shunt	All symptoms gradually resolved

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Table 2: Contd...

Title	Authors	Year	Number of patients	Clinical presentation	Treatment method	Results
The endoscopic trans-fourth ventricle aqueductoplasty and stent placement for the treatment of trapped fourth ventricle: Long-term results in a series of 18 consecutive patients	Pasquale Gallo, *et al*.	2012	18		Endoscopic aqueductoplasty and stent placement	At a mean follow-up of 90.8 months, all patients experienced a stable clinical improvement. Only two complications were observed: A transient diplopia due to disconjugate eye movements in one patient and a transient trochlear palsy in another one
Posterior fossa craniotomy for trapped fourth ventricle in shunt-treated hydrocephalic children: Long-term outcome	Suhas Udayakumaran, *et al*.	2011	12		Posterior fossa craniotomy/craniectomy and opening of the TFV into the spinal subarachnoid space	One patient underwent a second FTFV 21 months after the initial procedure. All 12 patients (100%) showed clinical improvement
Endoscopic aqueductoplasty and placement of a stent in the cerebral aqueduct in the management of isolated fourth ventricle in children	Giuseppe Cinalli, *et al*.	2006	7	Lethargy, vomiting, shunt infection, diplopia, developmental delay	Endoscopic aqueductoplasty alone or combined with placement of a stent in the cerebral aqueduct	Restenosis of the aqueduct occurred in two patients in whom stents had not been placed
Endoscopic aqueductal stent placement for the treatment of a trapped fourth ventricle	Leszek M. Sagan, *et al*.	2006	5	Ataxia and nystagmus, shunt malfunction, lethargy, headache, vomiting	Endoscopic aqueductal stent	Parinaud syndrome, rotatory nystagmus, and abducens nerve palsy developed postoperatively, in 1 patient; these deficits resolved after a preexisting supratentorial shunt was upgraded
Isolated fourth ventricle in neurocysticercosis: Pathophysiology, diagnosis and treatment	Colli B, *et al*.	1993	4	Signs of intracranial hypertension	4th ventricle drainage	All clinical signs improved
Trapped fourth ventricle in coccidioidal meningitis	H. Robert Harrison, *et al*.	1982	1	Vomiting, A left and ataxia, papilledema	4th ventricle shunt	Clinical improvement
The isolated fourth ventricle in children: CT and clinical review of 16 cases	Giuseppe Scotti, *et al*.	1980	16	Posterior fossa signs	4th ventricle shunt	Improved of the clinical condition in six of 14 children
Isolated fourth ventricle: To shunt or stent	Aaron Mohanty, *et al*.	2017	25	13 were asymptomatic	6 FVPS and 19 AST	Stent migration was observed in 2 patients, FVPS group, 1 had 2 shunt revisions while another developed reversible cranial nerve paresis, the extent of reduction was more with FVPS
Double compartment hydrocephalus-a new clinical entity	E. L. Foltz, *et al*.	1980	6	Anorexia, dysarthria, diplopia, lethargy, nystagmus	Removal of a veil occlusion of the upper 4th ventricle aqueduct	Immediate recovery in 5 of 6 patients

FVPS: Fourth ventriculoperitoneal shunt, AST: Aqueductal stents, CT: Computerized tomography, MRI: Magnetic resonance imaging, ETV: Endoscopic third ventriculostomy, TFV: Trapped fourth ventricle, VPL: Ventral posterolateral nucleus, FTFV: Fenestration of trapped fourth ventricle

without surgery. Symptomatic patients should be treated surgically for decompression of the fourth ventricle.[14] Predisposing factors which make patients susceptible to the development of TFV include, in the vast majority of patients, intraventricular inflammatory reactions, including bacterial infection or hemorrhage. When the diagnosis of TFV is delayed, it could have deleterious effect for the patient, leading to severe neurological dysfunction and/or death. Another common clinical scenario associated with TFV is its existence in the context of Dandy–Walker syndrome. This refers to a congenital malformation involving mainly the cerebellar vermis.[15,16] These patients typically exhibit a large cystic dilatation of the fourth ventricle, in concordance

with lateral ventriculomegaly. This entity is related embryologically to defects in the roof plate of the rhombencephalon, which prevents the normal inward turning of the inferior vermis and choroid plexus into the fourth ventricle.[17]

In addition to the causal relationship of TFV with these pathological processes, other entities which share in common their association with severe inflammatory reaction, i.e., neurosarcoidosis,[4] cysticercosis,[18] and coccidioidal meningitis[19] should be stated. The net result of all of these pathophysiologic pathways is the expansion of the fourth ventricle, which causes compression of the adjacent neural structures, including the cerebellum, brainstem, and lower cranial nerves. The clinical equivalent of this consequence ranges from the appearance, at the initial stages, of irritability, headache, and emesis, along with bulbar and cerebellar dysfunction. As the clinical situation is evolving, additional signs are recorded, such as opisthotonos, lethargy, respiratory instability, bradycardia, and a variety of ocular findings. When the spectrum of all of these potentially devastating neurological complications is established, urgent surgical treatment of this entity is necessitated. However, considerable debate still exists as to the most appropriate surgical strategy.

Materials and Methods

We used the keywords: "isolated fourth ventricle," and "trapped fourth ventricle," in PubMed® and Web of Science®. We reviewed the results, mainly being interested in the collection of data regarding clinical features, pathophysiology, along with available treatment options for the TFV. In addition, specific attention was given to literature reports dedicated to complication avoidance and advances in ventriculoscopy and frameless stereotaxy. To perform a narrative review addressing the issue of TPV, we executed a title-specific search using Thomson Reuters Web of Science database to identify the articles centered on that issue. We used all the relevant terms ("isolated fourth ventricle" and "trapped fourth ventricle") as our search criterion without setting any restrictions regarding publication dates. Retrospective, institutional studies, prospective studies, case reports, and cross-sectional studies were enrolled. We included all relevant publications, regardless of the language in which the manuscript was written.

Discussion

The entity of the trapped fourth ventricle has been reported in the past with several synonyms, such as double compartment hydrocephalus,[20,21] isolated fourth ventricle,[13,22] and encysted fourth ventricle[16,21] and has been recognized to be secondary to a wide spectrum of pathological conditions. More precisely, shunting of the lateral ventricles for communicating hydrocephalus, aqueduct stenosis,[22] and obstruction of the outlets of the fourth ventricle[13] have long been identified as inherently associated with this. Regarding its pathophysiologic substrate, it is common concept that pathological changes, of mechanical or inflammatory origin, at the aqueduct or at the fourth ventricular outlets are implicated. These could potentially render the fourth ventricle completely isolated from the other compartments of the ventricular system and subarachnoid space. Early reports exist[7] which state that shunting of the lateral ventricles due to communicating hydrocephalus can produce aqueductal obstruction due to kinking or chronic inflammatory changes secondary to the development of infectious disease. Raimondi *et al*.[16] presented a small series of patients who manifested this complication, pointing out that a significant percentage of them harbored Dandy–Walker syndrome as their underlying pathology.

The most common scenario, regarding the age at presentation of these cases, refers to premature infant who suffers from posthemorrhagic (communicating) hydrocephalus related to intraventricular hemorrhage due to prematurity. They are initially managed with ventriculoperitoneal shunt placement and occur in only a minority of infants.

Another commonly involved pathophysiologic mechanism is that related to the existence of the Dandy–Walker syndrome, a congenital malformation that affects the vermis of the cerebellum.[23] The most common imaging presentation of these patients includes a large cystic dilatation of the fourth ventricle, combined with an associated enlargement of the supratentorial ventricular system. Embryologically, the pathophysiologic substrate is implicated by defects in the roof plate of the rhombencephalon, which prevents the normal inward turning of the inferior vermis and choroid plexus into the fourth ventricle. Apart from that pathophysiological settings, other relevant mechanisms responsible for the development of TFV have been implicated. Among them, the most common pathway is the contribution of severe inflammatory reaction, irrespective of the underlying cause (i.e., neurosarcoidosis, cysticercosis, and meningitis).

Irrespective of the offending pathophysiologic mechanism, the most common clinical sequela is related to the expansion of the volume of the fourth ventricular cavity, with the associated compression that is administered on the nearby neural structures, that is cerebellum, brainstem, and lower cranial nerves. The net result of that phenomenon is a clinical presentation that is equivalent to that of an enlarging posterior fossa

mass, called the posterior fossa syndrome. Initially, patients suffer from irritability, headache, and emesis, along with bulbar and cerebellar dysfunction. Later, on the course of the disease, other neurological signs are recorded, including opisthotonic posture, lethargy, respiratory instability, bradycardia, and a wide spectrum of abnormalities related to oculomotor function. A more comprehensive catalog of associated disturbances follows:

1. Mutism (and speech changes)
   a. Complete loss of speech
   b. Decreased speech production
   c. Dysarthria
   d. Changes in speech quality, prosody, initiation.
2. Ataxia
   a. Unsteady gait/balance
   b. Hypotonia.
3. Behavioral symptoms
   a. Regressed personality
   b. Physical agitation (e.g. restlessness)
   c. Poverty of spontaneous movement
   d. Withdrawal.
4. Emotional lability
   a. Dysphoria
   b. Apathy
   c. Distress
   d. Inconsolability
   e. Tearfulness
   f. Giggling
   g. Distractibility
   h. Irritability.

It is widely accepted that an estimated amount of approximately 140 ml of CSF circulates, surrounding the brain and the spinal cord. We currently hypothesize that about 480 ml of CSF are produced every day, and CSF is recycled every 8 h. It is mainly produced by the choroid plexus (about 80%), and the remainder is thought to be secreted by the brain parenchyma. The lateral ventricles produce the vast majority of CSF, which then passes through the foramina of Monro to the third ventricle, and from there to the fourth ventricle through the aqueduct of Sylvius. The fourth ventricle is bounded by the pons and the upper part of the medulla anteriorly and the cerebellar vermis posteriorly. Inferiorly, it exhibits a continuation with the central canal of the spinal cord. The fourth ventricle has two lateral apertures (named, the foramina of Luschka) and one median aperture (the foramen of Magendie) through which the CSF passes to the subarachnoid space. Absorption of CSF is performed through the arachnoid villi, which protrude into the superior sagittal venous sinus, and then, it is redirected to the venous circulation. When both the aqueduct of Sylvius along with the foramina of Luschka and Magendie are occluded, the fourth ventricle becomes isolated from the remaining ventricular system, as well as from the circulation of the CSF through the pathway of the subarachnoid space. Irrespective of that, CSF production from the isolated choroid plexus of the fourth ventricle continues, resulting in a progressive and relentless dilation of the fourth ventricle. This, occasionally, could manifest with clinical signs of an expanding lesion within the boundaries of the posterior fossa. Figures 1 and 2 are used to explain the anatomical features of endoscopic third ventriculostomy (ETV).

We attempted to perform a comprehensive overview and we mention that the first relevant case[21] reported in the literature refers to a patient with cysticercosis, subsequent development of meningitis, and consequently, communicating hydrocephalus. Shunting of the lateral ventricles was performed and a few months later, signs of a posterior fossa mass developed. Imaging and surgical findings revealed cystic dilation of the fourth ventricle, related with occlusion of its outlets, combined with occlusion of the aqueduct. After the first description of the isolated fourth ventricle in 1975, several other similar documented cases were published.[22,24]

Another issue that deserves special caution is related to the consequences of the isolation of the fourth ventricle and the determination of the indications that render its management of paramount importance. The first manifestation of progressive fourth ventricle dilatation relates to compression and posterior displacement of the cerebellum. Following that, brain stem distortion begins, which is accompanied by anterior displacement of that entity. A common finding is the detection of superior herniation of the ballooned fourth ventricle through the tentorial hiatus into the supratentorial compartment. The clinical consequences of these alterations regarding the anatomical relationships of the fourth ventricle can vary widely. Isolated fourth ventricle (IFV) can either be associated with overt neurological signs or be asymptomatic and detected at routine follow-up. The common clinical features include truncal instability, poor feeding, disconjugate eye movements, and somnolence. An acute or subacute isolation of the fourth ventricle is more rarely detected and is associated with infection or hemorrhage at the time of diagnosis.[11,23,25]

Regarding the indications for decompression of an IFV, it seems that they are not so clearly determined. It seems to be common practice that surgery is reserved only for patients with signs and symptoms that could be attributed to the underlying pathology.[26-29] The overall assessment and the final judgment appear to be more straightforward in older children and adults, as it is easier to clarify if the dilated fourth ventricle is symptomatic. On the other hand, in infants or younger children with developmental delay, this association is more difficult to be established, and the condition can be recognized only after the clinical and neurological

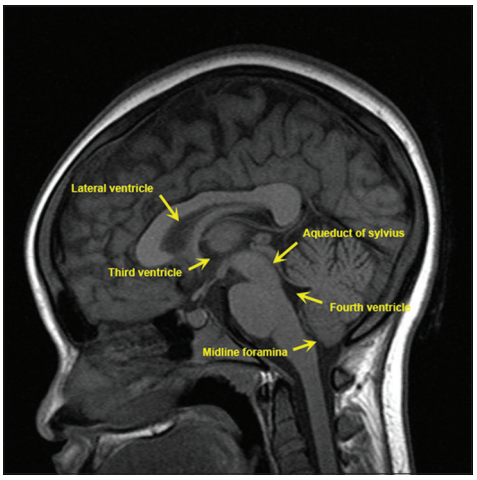

Figure 1: Anatomic structures in the midline of the brain, related to the fourth ventricle

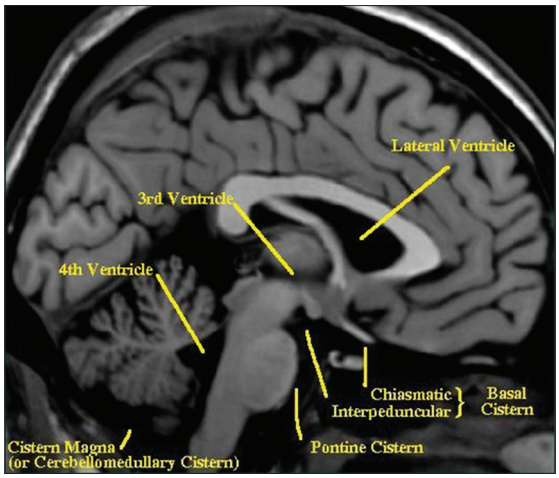

Figure 2: Anatomic relationships of the ventricular system and of the main brain cisterns where cerebrospinal fluid circulates

deterioration is considerable. Following that concept, it is common practice to operate on these patients when the fourth ventricle remains consistently dilated, associated with significant brain stem compression, as well as when a progressive dilatation was demonstrated during follow-up.

The diagnostic modality of choice for the evaluation of these patients and the determination of the underlying pathology seems to be magnetic resonance imaging (MRI). According to a recent review,[23] all patients that were considered for surgical intervention were evaluated preoperatively through the aid of focused thin overlapping sagittal section T2-weighted MRI imaging at the level of the aqueduct. This imaging was targeted on the assessment of the underlying offending pathology and degree of the aqueductal obstruction. The aqueductal obstruction was classified as (a) short-segment aqueductal obstruction (<5 mm in length), (b) long-segment obstruction (>5 mm in length), and (c) aqueductal web. The combination of a patent aqueduct with an enlarged fourth ventricle, the recognition of imaging characteristics compatible with Dandy–Walker malformation as well as posterior fossa cysts associated with another pathology constituted exclusion criteria from the diagnosis of isolated fourth ventricle.

Apart from that, there are relevant disease characteristics that were identified on preoperative imaging and were taken into consideration regarding the preoperative planning:

a. Length of aqueductal obstruction: when only a minor portion of the aqueduct of Sylvius is impaired, it seems that an aqueductal stent was the most appropriate option. On the other hand, patients with documented obstruction that refers to a great proportion of the aqueduct are generally offered the option of fourth ventriculoperitoneal shunt placement. Researchers[23] concluded that the existence of a limited, in length, stenosis of the aqueduct was suitable for both surgical alternatives, whereas a stenosis that incorporates the greater part of the aqueduct should be managed better with a shunting procedure.
b. Another, even though less important, parameter which should be taken into account in the selection of our treatment protocol is intimately related with the dimensions of the lateral ventricles. The presence of a supratentorial ventricular system without distension favors of initial externalization of the shunt. The main aim of this strategy is to achieve a gradual dilatation of the ventricular system to facilitate placement of a stent at a second stage.

However, a broad literature review was performed from our side and as the literature remains sparse, this issue is not satisfactorily addressed. It is common concept that most surgeons would consider operative intervention in the subpopulation of patients whose symptoms should be attributed exclusively to IFV.[26-29] The decompression of an asymptomatic, even progressively dilating, IFV is rarely adopted in clinical practice.[30] Based on that study, concerns are raised about the usefulness and utility of this approach. In a distinct subgroup of patients, i.e. infants and children with cerebral palsy and mental retardation, a progressively dilated IFV can cause significant brainstem dysfunction, although a definite association with recognizable symptoms is not easy to be established. Feeding difficulties, apathy, somnolence, and seizures often can be attributed to associated pathologic conditions, thus missing asymptomatic IFV remains possible. Similarly, a persistent dilation of the fourth ventricle with forward distortion of the brainstem is strongly indicative of the presence of significant elevation of the pressure within the fourth ventricle. Attempting a drainage of the IFV would potentially reduce such a concern. To enhance

our diagnostic accuracy and resolve diagnostic dilemmas, a thin section T2-weighted sagittal magnetic resonance imaging should be performed, to be able to assess the existence of aqueduct stenosis and brainstem distortion. Based on that, it seems to be logical that radiological documentation of progressive fourth ventricular enlargement and persistent brain stem compression, with or without distortion, could be a valuable and objective criterion for decompression of the IFV.

The armamentarium of applicable surgical alternatives for IFV can be broadly classified into the following subcategories:

- Suboccipital craniectomy and outlet fenestration
- Fourth ventricular shunting procedures
- Endoscopic procedures

For the past several decades, fourth ventricular shunts had constituted the mainstay of treatment for IFV.[1,26,30-32] Recently, a technical note has been published, which proposes a fourth ventricular-pleural shunt for diversion of an isolated fourth ventricle.[33] These shunts can be either inserted by a lateral transcerebellar route or by a midline transforaminal route.[1,23] Several complications appeared related with the transcerebellar route technique, most commonly involving injuries to the fourth ventricular floor during insertion of the shunt catheter, intracystic hemorrhage, and the appearance of frequent malfunctions.[1,23,30,31] A midline transforaminal approach, which adopts a catheter trajectory parallel to the brainstem along its long axis, avoids both these obstacles,[19] even though it requires intraoperative repositioning, from prone to supine position. Taking into consideration, the complications associated with the transcerebellar route and concern regarding the long-term functionality of these shunts, experts prefer placing most of their fourth ventriculoperitoneal shunt (FVPSs) by the trans-foramen of Magendie route.[23]

Another alternative, if catheter placement into the fourth ventricle is selected as the most appropriate treatment modality, is the stereotactically guided fourth ventriculoperitoneal shunting.[34,35] Proponents of this technique claim that this alternative is less invasive, as well as safer and more effective than the previously reported techniques, being able to avoid the vast majority of complications associated with them.

Reviewing the relevant literature, we have pointed out that there are several isolated "publications" (case reports), describing the occurrence of multiple cranial nerve paresis related to the treatment of isolated fourth ventricle. It is apparent that all of these patients were initially treated through fourth ventricular shunts for the IFV.[36-41] When the postoperative MRI was examined, it was evident that those patients shared in common posterior displacement of the brain stem, a fact that was compatible with shunt over-drainage.[37] A review of the relevant literature depicted that the procedures that were performed to overcome this complication included reprogramming of the shunt to a higher opening pressure,[39] inserting a shunt with an anti-siphon device,[39] removal of the shunt with a posterior fossa craniotomy and membrane excision in the region of the outlet foramina,[38] or conversion to a stent.[42]

When the open surgical option is considered for the treatment of ITV, a review of the literature reveals that there are several ways to treat an isolated fourth ventricle. More precisely, attempts have been made which were based on an open microsurgical fenestration, performed through the aid of a posterior fossa craniotomy.[1,28,43,44] Another suggested option is the insertion of a transcerebellar catheter, which may be added, through a Y connector, to an existing shunt system or, alternatively, diverted through its own distal valve and peritoneal catheter. An important drawback of that technique is that the revision and complication rate for the fourth ventricular catheters inserted with the help of anatomical landmarks only can exceed 40%.[30]

As far as the endoscopic approaches are considered, they can be broadly categorized into supratentorial and infratentorial approaches. The supratentorial approaches are further subdivided into the widely used trans-third ventricle trans-aqueduct approach and the trans-atrial approach. The most commonly performed transfrontal trans-third ventricular approaches rely on the coexistent ventriculomegaly to achieve safe navigation and visualization of the aqueduct, which is a prerequisite to execute the aqueductoplasty. An aqueductoplasty is followed by insertion of a stent to establish a communication between the fourth and the third ventricle. Another surgical alternative is to establish a communication between the dilated fourth ventricle and the atrium of the lateral ventricle, which could be feasible if the IFV extends above the tentorial incisura and has a relatively thin translucent wall.[45] The infratentorial endoscopic approach essentially is executed through the trans-foramen Magendie route, and a fenestration of the obstructed aqueduct is achieved from below through placement of a stent.[29,46]

Another important aspect of the debate regarding the utility of the endoscopic approaches and the selection of the most appropriate method is centered on the partial subversion of the evidence regarding the efficacy of these techniques. This was mainly due to the fact that although isolated aqueductoplasty was initially considered to be a very efficacious surgical strategy, this was somewhat revised. This is attributed to a novel observation which pointed out that most

aqueductoplasties become obstructed over a period of time, thus requiring reoperation.[27,29,47] This clinical scenario is especially referable to cases which have been implicated with postinfectious and posthemorrhagic conditions. There is wide acceptance of the fact that these situations display strong association with the development of intraventricular adhesions, along with isolation of several ventricular compartments. The main and widely accepted advantage of aqueductoplasty is the avoidance of implantation of any foreign material, which could be considered responsible for the development of infection and may be liable to migration and obstruction. Nevertheless, it is related with a high incidence of recurrence of scarring and subsequent obstruction of the communication. Taking into consideration all these data, placement of a transaqueductal stent (AST) appears to be the most feasible endoscopic option in IFV cases.

Apart from the management of the isolated fourth ventricle, another important consideration is the technique that should be applied to achieve drainage of the supratentorial compartment. Achievement of a communication of the fourth ventricle with the lateral and third ventricles through the aid of a stent is equivalent to equalization of the pressure in both (supra and infratentorial) compartments. Apart from the endoscopic technique, an additional CSF diversion procedure, which consists of the insertion of a lateral ventricle shunt or ETV, remains mandatory for the CSF drainage. In the subgroup of patients who undergo a simultaneous VP shunt insertion, the current trend is to insert a single system, consisting of a connection of the distal end of the CSF access reservoir to a shunt valve draining into the peritoneal cavity.[43,47,48] Another alternative that is proposed by another team of researchers[23] is the insertion of a completely discrete lateral ventricle shunt system which is not attached to the stent. The proposed advantage of this option is considered to be the opportunity provided to revise the malfunctioning shunt in case of an emergent occlusion, without having to deal with the stent, which would require endoscopic assistance for its replacement.

Endoscopic approaches have been extensively studied in the literature, regarding the treatment of IFV. These include endoscopic aqueductoplasty and interventriculostomy, endoscopic transaqueductal or interventricular stent placement, and endoscopic fourth ventriculostomy through suboccipital transaqueductal approach for fenestration of isolated fourth ventricle.[43,47,49-51] It is common concept, after reviewing all that studies (technical notes, case reports, research studies, retrospective studies) that neuro-endoscopy remains a feasible, effective, and safe technique, which should be included in our armamentarium for the treatment of ITV. However, we should never forget that the endoscopic approaches are inherently associated with complications. The most serious complication that is intimately related to aqueductoplasty, as well as stenting, is injury to upper brainstem structures, which is related to diplopia, ptosis, oculomotor paresis and less commonly, unresponsiveness, postoperative transient and permanent paresis of oculomotor function is reported in the literature in the range of 1.9% of cases.[48]

There are some points that deserve special mention. Image-guided stereotaxy and endoscopic approaches allow for a minimally invasive approach and aid to minimizing surgical morbidity. In selected cases, endoscopic management may offer the opportunity for permanent treatment, without the need for the implantation of foreign materials. On the contrary, if catheter placement into the fourth ventricle is selected or becomes mandatory, a trajectory parallel to the long axis of the brainstem, through the transtentorial hiatus, transventricular, or transforaminal (Magendie) route, is strongly advised, given the poor long-term results and morbidity of transcerebellar approaches.

When the efficacy of FVPS and AST is compared, they seem to be equally effective in the management of IFV. When the selection criteria for the decision of the most appropriate management option are considered, the extent of aqueductal obstruction and degree of ventriculomegaly are the most significant parameters. Another treatment alternative for isolated fourth ventricle is the combination of endoscopic placement of a transaqueductally or transcisternally guided stent, which is connected to a diverting shunt system. This seems to be an efficient procedure, able to reverse acute presenting clinical symptoms and probably capable to ameliorate longer-standing clinical symptoms. Apart from that, it is capable of demonstrating an objective decrease in the fourth ventricular size and associated mass effect. Detailed preoperative planning to precisely select the trajectory of stent placement and the length of the catheter is necessary, to perform this procedure safely with an uneventful outcome. When the complications are concerned, they were similar to described ones, associated with regular CSF-shunting procedures. Apart from them, no additional neurological adverse consequences resulting from the proposed procedure itself were stated.

We strongly believe that for the purpose of comparing the forms of surgical treatment, we have to discuss failure rates or efficacy of each one. To address that point, Table 2 is introduced. It lists the number of patients in each study analyzed, clinical presentation, treatment performed, and relevant results (success rate of various treatment modalities).

Conclusions

Considering the results of a broad literature review, we could adopt the assumption that treatment of the TFV remains a challenging, yet unresolved, clinical and radiological entity. Regarding the pathophysiologic substrate of this syndrome, it seems that the ventricular system is impaired by severe inflammation, along with relevant anatomic and physiological alterations. The clinical course of these patients is strongly unpredictable and because of that, careful follow-up is indicated for cases without evidence of clinical or radiographic progression. Apart from that, because of the high rate of surgical complications, careful clinical assessment is an option for a subgroup of patients who do not exhibit signs of deterioration.

Conversely, at the onset of clinical symptoms, and/or radiographic deterioration, prompt operative intervention is currently indicated. Regardless of the selected surgical approach, the surgical intervention that would be preferred should take into consideration the underlying cause of hydrocephalus, any other factors leading to the development of TFV, and previous treatment.

It seems that there is an agreement in the relevant literature studies that endoscopic treatment of an isolated fourth ventricle is technically feasible, and primary placement of a stent through the obtained fenestrations is efficacious, based on the results of recently published studies. It has been proposed that, especially in equivocal cases, the major determinants of the optimum management option are the existence of associated dilation of the lateral ventricular system, along with the degree, expressed as a percentage of its length, of aqueductal stenosis. In cases of patients harboring a functioning ventriculoperitoneal shunt combined with relatively small ventricles, our first option should be the acquisition of gradual ventricular dilation, before performing shunt placement. Stenting of the aqueduct remains an efficacious procedure and should not be abandoned because of its complications. The best preventive measure to avoid its complications is the determination of strict patient selection criteria.

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Conflicts of interest
There are no conflicts of interest.

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