Fourth ventricle outlet obstruction



The fourth ventricle outlet obstruction (FVOO) is a rare but well-established cause of obstructive tetraventricular hydrocephalus, characterizing with dilatation or large cerebrospinal fluid collection of the foramen of Magendie and foramen of Luschka.

Hydrocephalus is classified as noncommunicating and communicating based on whether all ventricular and subarachnoid spaces are communicating. Although the diagnosis between the two different states is crucial, it is difficult in certain conditions. In particular, communicating hydrocephalus and noncommunicating hydrocephalus owing to fourth ventricle outlet obstruction are highly misdiagnosed.

In FVOO, cerebrospinal fluid (CSF) is blocked at the fourth ventricle outlets by a membranous structure in the absence of any additional obstructive organic pathologies. Various terms for referring to FVOO have been used in previous reports, such as fourth ventricle/ventricular outlet obstruction ^{1) 2)} ^{3) 4)}, fourth ventricular outflow obstruction ⁵⁾, membranous obstruction of the fourth ventricle outlet ⁶⁾, obstruction of Magendie's and Luschka's foramina ⁷⁾, obstruction of fourth ventricular exit ⁸⁾ and primary obstruction of the fourth ventricle outlets ⁹⁾. Far distal obstructive hydrocephalus is a term that includes Dandy Walker or Arnold Chiari malformation, membranous obstruction or fourth ventricle and intercisternal external obstruction of the CSF ¹⁰⁾.

Etiology

The etiology and pathogenesis of FVOO are unclear, although some cases present with a history of meningitis or intraventricular hemorrhage.

In children, it is usually the consequence of posterior cerebral fossa malformations; while in adult, the occlusion is rather acquired than congenital, mostly linked to an inflammatory process, intraventricular hemorrhage, head trauma, brain tumors or Arnold-Chiari malformation. However,

idiopathic FVOO is extremely rare, and only 6 such cases have been reported in the English literature.

Bai et al., described an extraordinarily rare case of idiopathic FVOO in a 15-year-old patient successfully treated with direct microsurgical excision of the obstruction membrane. Furthermore, the clinical characteristics and treatment for this rare disease were investigated and reviewed ¹¹.

Diagnosis

CT ventriculography in infants, and CT cisternography in elder children, may assist to differentiate between FVOO and communicating hydrocephalus. The importance of these tests is for children with MRI suggestive of FVOO related hydrocephalus, but with no clear demonstration of the obstruction site. The implication of this differentiation may be for deciding between treatment of hydrocephalus with a ventriculoperitoneal shunt or with an endoscopic third ventriculostomy ¹²⁾.

Treatment

Third ventricle-fourth ventriculostomy is by far the most frequently used technique for cannulation of the aqueduct in a trapped fourth ventricle. In reported cases of , they have introduced a silicone tube stent from below after accessing the fourth ventricle through a small suboccipital craniectomy, ascending it on the aqueduct in order to reach the third ventricle. Management of this infrequently isolated fourth ventricle, but communicated with the rest of ventricular system, remains a challenge for neurosurgeons. Lack of knowledge of the pathophysiology makes it difficult to treat a problem that we do not understand ¹³⁾.

ETV is a viable option for treatment of patients with FVOO. The high failure rate in infants younger than 6 months of age suggests that ventriculoperitoneal shunting is a favorable option in this age group, rather than ETV. Isolated fourth ventricle is uncommon after ETV in hydrocephalus attributable to FVOO ¹⁴.

Suehiro et al., reported the use of neuroendoscopic third ventriculostomy to treat successfully both hydrocephalus and syringomyelia associated with fourth ventricle outlet obstruction. A 27-year-old woman presented with dizziness, headache, and nausea. Magnetic resonance (MR) imaging demonstrated dilation of all ventricles, downward displacement of the third ventricular floor, obliteration of the retrocerebellar cerebrospinal fluid (CSF) space, funnellike enlargement of the entrance of the central canal in the fourth ventricle, and syringomyelia involving mainly the cervical spinal cord. Cine-MR imaging indicated patency of the aqueduct and an absent CSF flow signal in the area of the cistema magna, which indicated obstruction of the outlets of the fourth ventricle. Although results of radioisotope cisternography indicated failure of CSF absorption, neuroendoscopic third ventriculostomy completely resolved all symptoms as well as the ventricular and spinal cord abnormalities evident on MR images. Neuroendoscopic third ventriculostomy is an important option for treating hydrocephalus in patients with fourth ventricle outlet obstruction ¹⁵.

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Case series

Three patients who were 21, 53, and 68 years of age presented with either headaches (isolated or associated with raised intracranial pressure) or vertigo, or a combination of gait disorders, sphincter disorders, and disorders of higher functions. In each case, magnetic resonance (MR) imaging demonstrated hydrocephalus involving the four ventricles (mean transverse diameter of third ventricle 14.15 mm; mean sagittal diameter of fourth ventricle 23.13 mm; and mean ventricular volume 123.92 ml) with no signs of a Chiari Type I malformation (normal posterior fossa dimensions, no herniation of cerebellar tonsils). The diagnosis of obstruction was confirmed using ventriculography (in two patients) and/or MR flow images (in two patients). All patients presented with marked dilation of the foramen of Luschka that herniated into the cisterna pontis. All patients were treated using ETV. No complications were observed. All three patients became asymptomatic during the weeks following the surgical procedure and remained stable at a mean follow-up interval of 36 months. Postoperative MR images demonstrated regression of the hydrocephalus (mean transverse diameter of third ventricle 7.01 mm; mean sagittal diameter of fourth ventricle 16.6 mm; and mean ventricular volume 79.95 ml), resolution of dilation of the foramen of Luschka, and good patency of the ventriculostomy (flow sequences). These results confirm the existence of hydrocephalus caused by idiopathic fourth ventricle outflow obstruction without an associated Chiari Type I malformation, and the efficacy of ETV for this rare indication ¹⁶.

Case reports

Duran D, Hadzipasic M, Kahle KT. Mystery Case: Acute hydrocephalus caused by radiographically occult fourth ventricular outlet obstruction. Neurology. 2017 Jan 31;88(5):e36-e37. doi: 10.1212/WNL.00000000003555. PubMed PMID: 28138085¹⁷⁾.

A 66-year-old woman with gait disturbance and incontinence caused by hydrocephalus underwent ventriculoperitoneal shunt surgery. After 9 months, her fourth ventricle became enlarged and could not be controlled by lowering the shunt pressure. Magnetic resonance imaging (MRI) demonstrated obstruction at the foramen of Magendie, foramina of Luschka, and the cerebral aqueduct. Endoscopic surgery for aqueduct plasty with third ventriculostomy was planned. Because the aqueduct was observed to open spontaneously, only the standard third ventriculostomy was performed. When MRI findings were reviewed retrospectively, an unnatural space was observed between the lower cranial nerves and cerebellar hemisphere that grew along with the fourth ventricular enlargement. This space was determined by MRI cisternography to be the cystic membrane ballooning out from the foramen of Luschka. The primary hydrocephalus likely resulted from fourth ventricle outlet obstruction.

Enlargement of the whole ventricular system with an expanded space between the lower cranial nerves and cerebellar hemisphere can be caused by fourth ventricle outlet obstruction. In such cases, preoperative evaluation of anatomic architecture and cerebrospinal fluid obstruction using MRI cisternography is essential and leads to a successful endoscopic strategy ¹⁸⁾.

A 3-year old boy without any remarkable medical history presented with a headache and vomiting. Computed tomography (CT) images, which had incidentally been taken 2 years previously due to a minor head injury, showed no abnormality. Magnetic resonance imaging on admission showed tetraventricular hydrocephalus associated with the dilatation of the fourth ventricle outlets, without any obstructive lesions. However, CT ventriculography, involving contrast medium injection through a ventricular catheter, suggested mechanical obstruction of the cerebrospinal fluid (CSF) at the fourth ventricle outlets. Thus, the patient was diagnosed with FVOO and ETV was performed; the hydrocephalus was subsequently resolved. Although hydrocephalus recurred 1 year postoperatively, re-ETV for the highly stenosed fenestration successfully resolved this condition.

ETV should be considered for FVOO treatment, particularly in idiopathic cases without CSF malabsorption 19 .

A 15-year-old girl with amenorrhea and a several-week history of headache. After the diagnosis of membranous obstruction of the foramen of Magendie suggested by MRI, suboccipital craniotomy for removal of the membrane was carried out. The patient made an excellent postoperative recovery, and postoperative phase-contrast MRI demonstrated patent cerebrospinal fluid (CSF) pathways at the level of the foramina of Magendie and Luschka. We believe that this case is of interest because of the unequivocal evidence on MRI studies of the occlusion of the foramen of Magendie preoperatively, and because of the dramatic postoperative MRI findings demonstrating the effectiveness of the surgical procedure both in terms of ventricular size and CSF flow characterization ²⁰.

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