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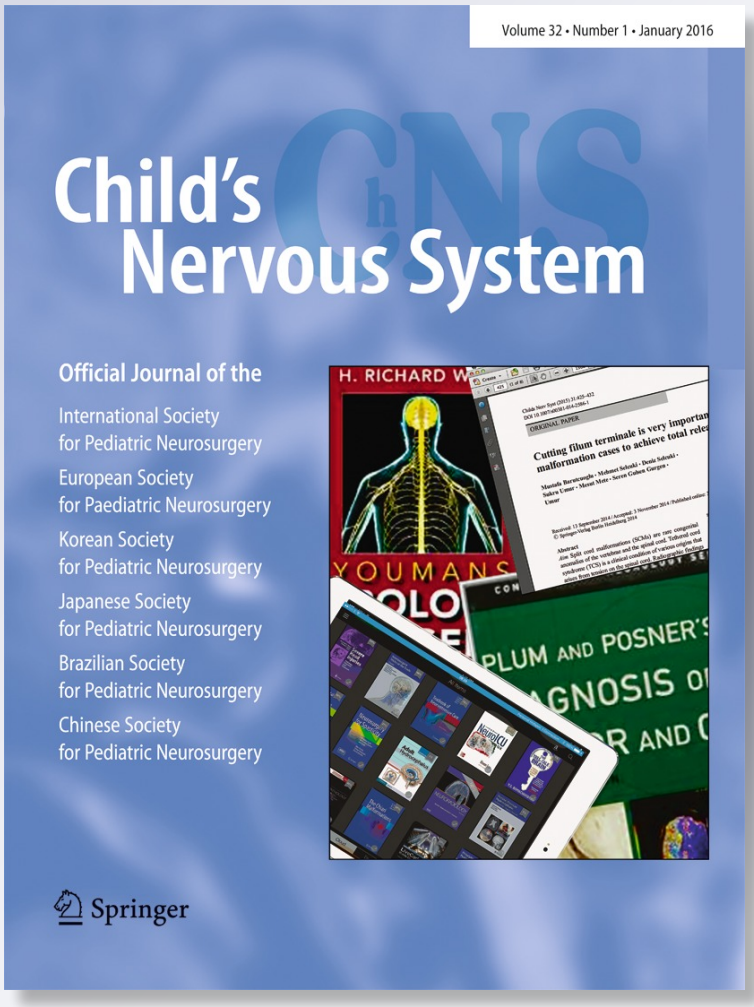

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CASE REPORT

Split cerebral aqueduct: a neuroendoscopic illustration

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Abstract
Purpose Forking of the cerebral aqueduct is a developmental malformation that is infrequently encountered by neurosurgeons as a rare cause of hydrocephalus, sometimes with a delayed onset. The etiology of an apparently forked aqueduct might be different. However, neuroendoscopy can often be the optimal treatment. The purpose of this study was to review the literature by analyzing the anatomical, functional, diagnostic, and therapeutic features of this unusual condition and adding our personal cases.
Methods We present a case of forking of the cerebral aqueduct that was detected in vivo and treated with a flexible scope. A thorough review of the pertinent literature is also discussed. In the past years, diagnosis of forked aqueduct was possible only postmortem.
Results A forked aqueduct is occasionally encountered in patients when a delayed hydrocephalic decompensation occurs.
Conclusions Flexible neuroendoscopy enables for a direct, in vivo diagnosis and immediate treatment through a third ventriculostomy.

Keywords Aqueductal stenosis · Cerebral aqueduct · Forking · Hydrocephalus

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Introduction

Although some authors have described the forking of the cerebral aqueduct (CA) as a common cause of hydrocephalus, a PubMed search of this malformation revealed only a few reported cases, all recognized postmortem. Moreover, the terms forking, atresia, duplication, and obstruction have been inconsistently used to illustrate different anatomical variants. We reviewed the literature to clarify this intriguing aqueductal abnormality, its pathogenesis, and its importance in explaining some types of delayed hydrocephalus. Usually, a forked or split aqueduct is not clearly detected on magnetic resonance imaging (MRI) and represents an example of a patent but nonfunctional aqueduct. We also discuss a case directly observed in vivo during our 25-year experience after more than 700 neuroendoscopic procedures, where the adytum of the aqueduct was explored in about 2/3 of the cases.

Case report

A young adult presented to our clinics complaining only of episodic headache. MRI revealed a triventricular hydrocephalus with indirect signs of tensive dilatation of the third ventricle without evidence of aqueductal septation. A neuroendoscopic procedure was scheduled. No history of intraventricular hemorrhage or infection was reported. During the exploration of the posterior third ventricle, a thick septum was noticed dividing the aqueduct. It appeared as a bridge of ependyma stretched in the middle of the aqueduct. We decided to cut off the bridge in order to restore a normal aqueductal anatomy and CSF flow. However, the scaffold of the bridge did not collapse, remaining rigidly in the middle of the

aqueduct and interfering with the CSF flow (Fig. 1). This observation prompted us to proceed with a standard endoscopic third ventriculostomy (ETV). After surgery, the patient fully recovered and was discharged with no headache and without any evidence of additional neurological deficit.

Discussion

The cerebral aqueduct, or aqueduct of Sylvius, is a narrow channel that connects the third and the fourth ventricle at the level of the dorsal midbrain. Its deep position and its tiny structure do not make its exploration easy. Historically, the description of the anatomy and the pathological variants of the cerebral aqueduct had been possible only through postmortem examination and, more recently, through MRI. However, technical limits prevent the systematical and precise analysis of this minute canal. In the last years, the wide diffusion of neuroendoscopy, particularly the flexible neuroendoscopy, allowed a direct in vivo inspection not only of the adytum but also of the lumen in selected cases [13–16]. The precise definition of the cause of aqueduct obstruction is therefore almost always possible during surgery, just by bending the tip of the fiberscope backwards after entering the third ventricle through the foramen of Monro.

Forking of the aqueduct is considered one of the causes of aqueductal stenosis. It must be distinguished from other causes of CA defective patency, such as atresia (complete absence of the aqueduct), membrane formation (the lumen is completely or partially obstructed by a thin transversal membrane, consequence of inflammatory or mechanical distortions), simple stenosis (the aqueduct is histologically normal but abnormally narrow due to a developmental error), gliosis (marked proliferation of subependymal fibrillary glia in the absence of inflammation, phagocytic cells, or tumor), inflammatory occlusion (following meningitis), and tumors (directly or indirectly occluding the aqueduct). Although they are two different conditions, the term atresia has been frequently associated with forking. The forking of the floor of the CA results from incomplete fusion of the median fissure and, according to some authors, is biologically not significant [6, 9]. As they both result from developmental anomalies of the median fissure, it is possible that forking and atresia are the consequence of a similar developmental malformation with different severity. During embryological life, after the closure of the neural tube, the aqueduct is relatively large in the first phases of normal fetal development, becoming narrower later, and this process may go too far [20]. This progression has been compared to the physiological closing of the central canal of the spinal cord. Notably, forking can occur also in the central medullary canal. It has been shown that 45 % of healthy children have major forking of the canal in the conus, and 31 % have minor forking [12]. However, Foltz and Shurtleff hypothesized a different mechanism of forking development, occurring after CSF diversion. During active hydrocephalus, the ependymal cells become flattened from the effects of ventricular enlargement. When the dilating force of the CSF is relieved by an effective CSF shunt from the ventricles, ventricles and aqueduct wall collapse. The redundant wall of the aqueduct may actually fold in so that the scarring effect created by the interstices between the ependymal cells pinches off part of the invaginating wall to produce an apparent forking (acquired forking). In the same way, opposite walls no longer held open by pulsatile CSF flow may come in contact with each other and obliteration of the lumen occurs through fibrosis of the interspaces between the ependymal cells [5].

Moreover, the caudal third of the floor of the CA usually presents a deep median slit. The walls of the slit may be fused in its dorsal portion, forming a narrow channel projecting in a cranial direction. Mac Farlane and Maloney proposed to call this physiologic ventral channel of the normal CA "simple" forking, to distinguish it from "complex" forking with two or more intertwining channels in the midsagittal plane [17]. The latter form is not common and only seen in CNS malformations, while the dorsal forking has not been reported in otherwise normal individuals [9]. CA forking is sometimes associated with rhombencephalosynapsis, fusion of the colliculi (mesencephalosynapsis), atresia of the third ventricle (diencephalosynapsis), and corpus callosum abnormalities [1].

Russell provided the first clear description of pathologic aqueductal forking in 1949 [22]. She described it as "two

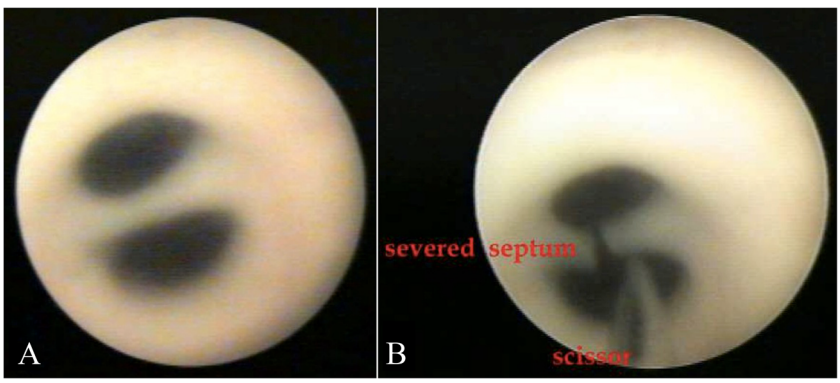

Fig. 1 **a** The aqueduct is forked by a bridge of tissue. **b** The bridge of tissue does not collapse after cutting it with microscissors

Table 1 Forking of the cerebral aqueduct: review of the literature

Author	Patients	Age, sex	Onset	Aqueduct	Diagnosis	Treatment	Outcome
Russell 1949 [22]	10						
Drachman and Richardson 1961 [4]	1	72 years, F	6 months feverishness, weight loss; 25 years headaches, sleepy feeling, legs weakness	3 channels	Postmortem	Steroids	Died (leukemia, spleen rupture)
Foltz and Shurtleff 1966 [5]	1	Adult, M	History: SAH, VP shunt	Acquired forking	Postmortem		Died 6 years after shunt
Kepes et al. 1969 [10]	1	6 months, M	Enlarging head, spastic quadriplegia, bilateral optic atrophy	Areas with 2 channels, areas with multiple channels	Postmortem; fused thalamic mass; quadrigeminal plate replaced by single dorsal protuberance; cerebellar hemispheres fused, one centrally located dentate nucleus		Died
Oberson and Gessaga 1972 [19]	1	56 years, M	Consciousness disturbance and progressive dementia after head trauma	2 channels fusing caudally	Neuroradiological, confirmed postmortem	VP shunt	Died 62 days after trauma from infection
McMillan and Williams 1977 [18]	1	32 years, F	10-year history of occipital pain; 3 weeks history of drowsiness, papilledema, neck stiffness		Postmortem		Died
Hori et al. 1984 [7]	1	1 year	Multiple aqueductal diverticula				
Conover and Roessmann 1990 [3]	1	0 year, M	Complex malformations of CNS, 35-week gestation		Postmortem		Died
Kim et al. 1990 [11]	1	0 year, F	Complex malformations and aprosencephaly, 39-week gestation	Many channels	Postmortem		Died
Pasquier et al. 2009 [21]	13	Fetuses	Various degrees of mesencephalic malformations (dentate fusion, colliculi fusion…)		Postmortem		
Adle-Biassette et al. 2013 [1]	Mixed with atresia of CA	Fetuses	Various degrees of mesencephalic malformations		Postmortem		

distinct channels situated in the midsagittal plane and separated from one another by normal nervous tissue. The ventral channel is usually a simple slit in the dorsoventral plane. The dorsal channel is considerably branched and the neighbouring tissue contains many groups of displaced ependymal cells, many of which form tubules". The morphology of the two channels is variable. Either the ventral or the dorsal channel may unite the ventricles. The other one either shrinks and disappears or it joins the first one leading to a normal lumen.

According to Russell, the definitive diagnosis of developmental-based forking requires histopathological evidence of ependymal-lined channels. Of course, such demonstration is not possible in our case, which was inspected in vivo. However, regardless the etiopathogenesis (developmental error, or acquired fusion of a bridge from one ependymal surface to the another), in our experience, a split aqueduct is an uncommon finding during neuroendoscopy, which can lead to a surgically treatable hydrocephalus. In our case, the forking appears not to significantly reduce the lumen of the aqueduct. However, the simple presence of a thin obstacle in the narrow lumen of the aqueduct is enough to disrupt the laminar CSF flow, which becomes turbulent. Computer modeling of the CSF flow dynamics has shown that narrowing caused by forking changes the geometry of the channel, increasing the pressures needed to drive the flow. Some stenoses result in pressures, which are significant and potentially measurable within the intracranial pressure (ICP), although they may not contribute significantly to the total ICP [8]. This may explain the cases of patients affected by forking of the aqueduct who develop symptoms only in adulthood, as in our case, more likely because of the loss of an instable balance between CSF pressure and aqueduct diameter. A functional decompensation of a preexisting congenital narrowing of a forked aqueduct has also been described after head trauma [19]. Moreover, aqueduct forking increases wall shear stresses, which may result in ongoing damage to the aqueduct, further leading to CSF flow decompensation. More severe degrees of forking, where the aqueduct is reduced to a series of ductules, may result in pressure differences significant enough to cause a detectable increase in ICP. The developing of a pressure gradient between the supratentorial and infratentorial compartments may lead to typical anatomical deformations of the third ventricle and to symptoms specific of hydrocephalus secondary to aqueductal stenosis [2]. Interestingly, flow velocities through a forked aqueduct are also increased, potentially causing a flow void on MRI. This phenomenon can lead to the misdiagnosis of a patent aqueduct [8]. The decision to cut off the bridge of tissue in our case was probably a wrong choice, as its rigid scaffold prevented a complete restoration of the aqueductal lumen. Moreover, the channels have been reported to be potentially separated by normal nervous tissue. Although this tissue could be not completely functional, as our case would suggest, any attempt to restore a single passage from the third to the fourth ventricle should be avoided.

According to some authors, forking of the CA is a fairly common condition, which has been observed in both hydrocephalic and non-hydrocephalic subjects [9]. However, a thorough review of the pertinent literature revealed only 31 cases of forking of the aqueduct and only 14 in adult patients (Table 1) [1, 3–5, 7, 10, 11, 18, 19, 21, 22]. Although forking of the CA might be a relatively common finding in fetuses that died because of more complex malformations, it appears quite uncommon in young adults. There are many possible reasons to explain this uncommonness. Simple types of forking may be clinically silent and lead to clinically relevant hydrocephalus only at a later stage. Moreover, some cases may be misdiagnosed as simple stenosis if flexible neuroendoscopy is not performed to assess the anatomy of the CA. Actually all the reported cases were diagnosed postmortem, besides the case reported by Oberson and Gessaga, where a brilliant neuroradiological diagnosis was made [19].

When a forked aqueduct is diagnosed during neuroendoscopy, ETV is the safest and most effective treatment.

Conclusions

Forking of the CA is commonly seen in association with other developmental malformations. However, it is rarely diagnosed in adulthood, when hydrocephalus-related symptoms may appear because external events (traumatic) or intrinsic changes (damage to the aqueduct due to shear stress, change in CSF production rates, …) decompensate the fragile equilibrium between CSF pressures and CA-altered anatomy. Flexible neuroendoscopy is the only way to assess the correct diagnosis and allows a safe and effective treatment through ETV.

Conflict of interest The authors declare no conflict of interest. No funding nor financial support was received for this work. The authors declare no industry affiliation.

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