

Staged Neuroendoscopic Management of a Dandy Walker Malformation Case Presenting with Complex Hydrocephalus

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Abstract

Background: Dandy-Walker malformation is a rare congenital condition characterized by alteration of posterior fossa anatomy. Hydrocephalus is a common finding in these patients. VP-shunt placement remains the treatment of choice. Endoscopic Third Ventriculostomy (ETV) is an acceptable alternative for older children. Other techniques have also been proposed.

Case presentation: A full-term infant presented with signs of hydrocephalus. An endoscopic foraminoplasty of the posterior fossa cyst was conducted as a first stage. The cyst's volume was reduced, without any changes of the supratentorial ventricular dimensions. A month post-surgery signs of hydrocephalus reoccurred. An ETV was performed alongside a fenestration of the tentorium. The toddler was discharged in good condition with amelioration of her symptoms. One month later she presented with recurrent hydrocephalus and subdural hygromas. A shunt was placed in the posterior fossa under endoscopic guidance, and an additional one was introduced in the subdural space. The patient had 18 months followed-up without any further need for treatment. The patient had an 18 months follow-up.

Discussion: Lushka and Magendie foraminoplasty seem to be ineffective. ETV should be reserved for older children. The ventriculoscope can be employed for positioning the VP-shunt in a specific anatomical region. Subdural hygromas may present after neuroendoscopic treatment "VP-shunt placement remains the treatment of choice". A staged approach may be mandatory for complex hydrocephalus treatment.

Keywords: *Dandy Walker, hydrocephalus, endoscopic third ventriculostomy, tentorium fenestration, Ventriculo-Peritoneal shunt*

Introduction

Dandy-Walker malformation (DWM) is a rare congenital condition characterized by vermis hypoplasia/aplasia, cystic dilatation of the fourth ventricle and, upward displacement of tentorium and torcula. Common clinical manifestations include psychomotor development delay, ataxia, muscle weakness, seizures, apnea attacks and hydrocephalus. The treatment of the last is usually complex, frequently demanding a staged approach. We present the multi-phase neuroendoscopic treatment of a DWM case presenting with complex hydrocephalus.

Case presentation

A video presentation is available [see supplementary information Video 1. A video presentation of the case including imaging (MRI and CT) alongside intraoperative footage.]

Patient Characteristics

A full-term, African race infant was delivered from a first pregnancy with a Cesarean section. The first-trimester ultrasonographic screening revealed ventricular enlargement, absence of septum pellucidum, hypoplasia of choroid plexus, and a cystic dilatation of the posterior fossa with no detectable vermis.



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Apart from cephalomegaly other somatometric parameters were normal. Cardiopulmonary Resuscitation was performed upon delivery due to cyanosis and bradycardia. She was admitted to the Neonatal Intensive Care Unit. Neurologic assessment revealed lethargy, generalized hypotonia, a bulging fontanel, and a rapid increase in head circumference.

MRI Findings

A diffuse dilatation of the ventricular system, a fourth ventricle cystic enlargement, cerebellar and vermian hypoplasia, an anterograde brainstem and upward tentorium and torcula displacement were registered. The aqueduct was regarded patent and there was no detectable choroid plexus. Findings were associated with a "real" DWM¹.

Surgical plan

Hydrocephalus was associated with a fourth ventricle outlet obstruction, and an endoscopic foraminoplasty was attempted through a posterolateral approach.

Endoscopic Posterior Fossa Cyst Fenestration

Beneath the aqueduct of Sylvius the median sulcus and eminences were seen, alongside the inferior fovea and area postrema. The cyst walls and underlying membrane were fenestrated, initially to the left and right of the brainstem, in an attempt to create two artificial foramina of Luschka. The first cervical rootlet and the branches of the posterior inferior cerebellar artery were seen through the left stoma and vertebral artery and the hypoglossal nerve through the right. The pulsating movements and the optical confirmation of the fenestration of all membranes was considered as a sign of adequate cerebrospinal fluid movement. The stoma size was approximately 2mm (diameter of open Decq forceps). An additional fenestration in the anatomical location of the foramen of Magendie was performed. This part presented a challenge due to the close relationship between the neural structures and the cyst's wall. A biopsy clamp was used in order to grasp the membrane while moving away from the neural tissue during the pulsation cycles. This resulted in avulsion of a minor vascular vessel. Forced irrigation was employed in order to achieve hemostasis but this was not sufficient. Subsequently a diathermy electrode was used to apply direct compression to the vessel without cauterization. Hemostasis was finally achieved. The early post-surgery period was uneventful.

Follow-up

Post-surgery MRI after 3 weeks revealed a reduction of the cyst's total volume; with no significant change at the supratentorial

ventricular system. A month post-surgery the toddler presented with a gradual consciousness deterioration, thus an Endoscopic Third Ventriculostomy and a fenestration of the tentorium was planned.

ETV and Tentorium Fenestration

The third ventricle floor was thick, and the recognition of the exact borders of the mamillary bodies was challenging. The third ventricle floor was fenestrated using a Decq forceps revealing the hypophyseal tissue beneath, thus the stoma was widened toward the mamillary bodies. The underlying web-like Lilequist membrane was also fenestrated and the clivus was visualized, ensuring the successful communication of the ventricular system with the subarachnoid space. Pulsatile movements confirmed the successful communication between the spaces. A fenestration was placed also on the tentorium.

Follow-up

One-month post-surgery follow-up showed that the patient presented a rapid head circumference increase. The MRI confirmed the enlargement of the ventricular system and showed the presence of bilateral subdural collections.

Fenestration of tentorium and endoscopically assisted VP- shunt placement into the posterior fossa

A left Kochers' point burr hole was used. The previous tentorial fenestration could not be detected; thus, a new one was created but towards the tentorial hiatus this time, in proximity with the left internal cerebral vein. The posterior fossa was observed through the stoma in order to ensure communication between the compartments. A VP-shunt catheter was introduced under endoscopic guidance, as a stent through the new fenestration and into the posterior fossa cyst. A programmable valve was adjusted to an opening pressure of 8cmH₂O. Early post-surgery period was uneventful. Ultrasonographic evaluation revealed the collapse of the ventricular system and the subsequent enlargement of the subdural collections. The valve was re-adjusted to 12cmH₂O in an attempt to enlarge the ventricular system with subsequent obliteration of the subdural spaces. That proved inadequate and the next stage was to insert a right subdural catheter connected to a second valve and a second peritoneal catheter. The seconds' valve initial setting was set to zero. The child was discharged in good condition with a significant developmental delay and persistent hypotonia. The follow-up extended for an additional period of 18 months. There was no need for an additional treatment. Despite the subdural-peritoneal shunt there was no significant change in the diameter of the subdural hygromas.

Discussion

DWM presents a group of congenital posterior fossa malformations associated with conditions affecting the early brain development, including exposure to infectious and teratogenic agents² (4th-7th gestation week), and chromosomal abnormalities (mostly on chromosomes 3,9,13,18)³. The presented case had a negative TORCH and endocrinology screening, chromosome analysis was normal and the mothers' medical history was unremarkable.

There is a variety of cystic lesion located in the posterior fossa incl. DWM, arachnoid cysts, Blakes' pouch syst and a prominent cisterna magna. The presented malformation is classified as a Barkovich Type A or a "real", "true" DWM⁴. This is based on the presence of an evident communication between the posterior fossa CSF and the fourth ventricle, alongside the absence of a visible vermis on MRI/CT imaging. The diagnosis is ultrasonographical and may be established in utero since 14 gestation week⁵. Here the diagnosis was established on the first trimester ultrasound.

The therapeutic approach in these patients consists of treating the associated problems. Hydrocephalus is a common finding (>80% of patients), especially during the first 3 months post-partum⁶. This was mainly related to the obstruction of the fourth ventricle and the ventricle outlets⁴. However, literature has shown that this is a multifactorial entity, with some patients presenting a partial obstruction or no obstruction^{7,8}. We should highlight that CSF dynamics and pathophysiology of hydrocephalus is not yet completely understood⁹. Some have proposed the importance of venous hypertension, associated with direct pressure and lengthening of the venous sinuses due to the elevation of the anatomical structures, however this remains an unproven assumption¹. The introduction of a VP-shunt remains the typical management¹⁰. Commonly a ventricular catheter and a cyst catheter were connected with a Y connector and then with one valve and a distal catheter. Others have used two separate valve systems, one for the supratentorial ventricles and a second for the cysts. These multiple catheters, with connectors or dual valve systems have been proven to be more prone to dysfunction and subsequent revisions are more challenging¹¹. ETV is considered an acceptable alternative in older children⁴. Other approaches including the introduction of a ventriculo-cysto-peritoneal shunt alone¹⁰, or in combination with a VP-shunt¹², microsurgical exploration and foraminoplasty of outlet foramina¹³, or endoscopic trans-aqueductal approach¹⁴ may be found

in the literature. Cyst membrane excision is not employed routinely anymore and is reserved for cases where other treatments have been unsuccessful. In this case the Magendie and Lushka foraminoplasty, thus CSF-flow obstruction elimination, was selected as a more physiological approach. The endoscopic suboccipital approach has been described afore¹, however, according to our knowledge the suboccipital lateral approach and foraminoplasty of Lushka and Magendie, as described, has not been previously presented in the literature. The post-surgery total cyst volume reduction confirmed the efficacy of the technique. We cannot provide a definitive explanation regarding the persistent imaging findings regarding the supratentorial ventricular system. This finding could defy the patency of the aqueduct, could point toward a partial communication, or could present a novel disruption of CSF flow. One-month post-surgery the toddler presented with signs of hydrocephalus. An ETV and fenestration of the tentorium were performed. ETV has become a treatment of choice in selected patients, even though the anatomical understanding of the third ventricle floor is limited¹⁵ and the exact mechanism leading to resolution of symptoms after ETV has not been exactly defined⁹. Some authors have also proposed the additive effect of Choroid Plexus coagulation¹⁶. As the patency was brought into question, the tentorium was fenestrated in order to create a communication between the subarachnoid space, the ventricular system and the posterior fossa cystic formation. However, the employed technique provided also a communication with the subdural space, due to the fenestration of the tentorium and the conduction of the ETV, a fact that could provide an explanation to the formation of the persistent subdural hygromas on the following imaging investigation, especially in case of reabsorption failure through the subarachnoid space.

The last could be potentially associated with age related anatomical variations regarding the number and function of arachnoid villi, or due to hydrodynamic disbalance associated with hemorrhage or other factors associated with potential chemical disruption of CSF⁹. Other authors have proposed the endoscopically assisted placement of a stent through the aqueduct when the patency is defied¹⁷. A month post-surgery the patient presented with signs of hydrocephalus. A shunt catheter was placed in the posterior fossa under endoscopic guidance. Additional openings were created on its surface in order to achieve a stent like function ensuring the communication of the cyst with the ventricular system. The previous fenestration was not detected, a fact that could be potentially associated with hydrocephalus recurrence. An additional catheter was also placed in the

right subdural hygroma. The child was discharged in good condition, without any change in her neurological image. There was no need for reintervention during the 18-month follow-up.

Conclusion

VP-shunt placement remains the treatment of choice for hydrocephalus in DWM patients. ETV (+/- CPC) may be considered in older children. Foraminoplasty seems to be ineffective, thus we do not advise its' use. Artificial communication of the subarachnoid space, the ventricular system, the posterior fossa cyst and the subdural space may result in the formation of subdural hygromas. A staged approach may be necessary in complex hydrocephalus cases.

Conflicts of Interest Disclosure

The authors have no conflict of interest to disclose.

Statement of Ethics: Written informed consent regarding publication of data and images was acquired from both of the parents of the infant. The provided neuroradiological depictions were anonymized in order to secure patient identity with respect to the General Data Protection European Union Regulation 2016/679. The conduction of the study is in accordance to the World Medical Association Declaration of Helsinki. No Ethical Committee approval is required as this is an observational retrospective study.

Author Contributions

Marios Theologou: Visualization, Conceptualization, Draft Preparation, Literature Search, Multi-media file creation

Panagiotis Varoutis: Draft review, Literature Search

Vassilios Tsitoura: Supervision, Motivation, Draft Review

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