Endoscopic Transaqueductal Placement of a Single-Catheter Cyst-Ventriculoperitoneal Shunt in a Neonate With Dandy-Walker Malformation-Associated Hydrocephalus

—Case Report—

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Abstract

A neonate with hydrocephalus associated with Dandy-Walker malformation was successfully treated with an endoscopic placement of a transaqueductal ventricular single catheter. The modified catheter was provided with additional fenestration on its proximal side to allow simultaneous drainage from both the supra- and infratentorial compartments. This technique is well known for isolated fourth ventricles, but has not been applied to hydrocephalus associated with Dandy-Walker malformation. The cyst-ventriculoperitoneal shunt effectively drained both compartments. The patient was doing well 18 months after the surgical procedure. Endoscopic transaqueductal shunt placement can be considered, especially in patients with aqueductal patency.

Key words: Dandy-Walker malformation, hydrocephalus, neuroendoscopy, ventriculoperitoneal shunt, pediatric neurosurgery

Introduction

Controversy remains over the optimal treatment for hydrocephalus associated with Dandy-Walker malformation (DWM). The surgical management of this disorder has shifted from craniectomy and excision or fenestration of the cyst wall to the placement of various shunts.1,5,9,11–14,22) Lateral ventriculoperitoneal (VP), cyst-peritoneal (CP), and combined VP and CP shunts have been placed and endoscopic third ventriculostomy (ETV) has been performed in the past decade.14) Transaqueductal placement of a multiperforated shunt catheter has been used to treat isolated fourth ventricle,20,21) but not hydrocephalus associated with DWM. We report a pediatric patient with hydrocephalus associated with DWM who was successfully treated with this technique.

Case Report

A male infant was born at 37 weeks of gestation after an uneventful pregnancy; his weight and head circumference at birth were 1,984 g and 30.5 cm. Prenatal ultrasonography demonstrated hypoplastic cerebellum and amniocentesis revealed chromosomal abnormality (46,XY,t(2;3)(q34;q24)). Cranial echography from the anterior fontanel and computed tomography (CT) revealed hypoplastic cerebellum, dilation of the fourth ventricle, and very mild hydrocephalus. A diagnosis of DWM was made. He manifested dysphasia due to micrognathia and cleft soft palate. Magnetic resonance (MR) imaging performed 15 days after birth disclosed intraventricular hemorrhage in the left lateral ventricle, but no worsening of hydrocephalus occurred. At 21 days of age he was transferred to our institution because of CT evidence of marked progression of hydrocephalus.

On the day of admission we placed an Ommaya reservoir at the right frontal horn. Ventriculocisternography performed from the Ommaya reservoir revealed that the posterior fossa cyst was continuous with the fourth ventricle and showed delayed wash-out of contrast material. MR imaging demonstrated the classic features of DWM: triventricular hydrocephalus, high insertion of the tentorium, hypoplasia of the cerebellar vermis, cystic dilation of the fourth ventricle, and anterolaterally displaced cerebellum. No aqueductal stenosis was detected. The brainstem was displaced forward and flattened against the clivus (Fig. 1A–C).

When the patient was 47 days old we placed a cyst-ventriculoperitoneal shunt. A semi-rigid neuroendoscope (NeuroPEN; Medtronic PS Medical, Goleta, CA, USA), 155 mm in length and 1.1 mm in diameter, was connected to the light source and video system. The Ommaya reservoir...
Fig. 1  A–C: Preoperative axial T₁-weighted (A, B) and sagittal T₂-weighted (C) magnetic resonance (MR) images demonstrating supratentorial hydrocephalus, high insertion of the tentorium, and a large posterior fossa cyst with a patent aqueduct. D–F: Postoperative axial T₁-weighted (D, E) and sagittal T₂-weighted (F) MR images 6 months after surgery showing significant decompression of both the supratentorial and infratentorial compartments. The arrowhead indicates the shunt catheter.

Fig. 2  A: Skull radiograph. Arrows indicate the additional fenestrations on the proximal side of the ventricular catheter. B: Schematic drawing of the modified ventricular catheter.

was removed and the neuroendoscope was introduced into the right frontal horn through the same tract. The third ventricle was entered via the foramen of Monro and the aqueduct was visualized beneath the massa intermedia. The ventricular catheter (Innervision; Medtronic PS Medical), 1.3 mm in inner and 2.5 mm in outer diameter, and 23 cm in length, was conjointed with the stylet-type neuroendoscope in its tunnel. The modified ventricular catheter featured additional fenestrations on the proximal side to allow simultaneous drainage (Fig. 2). An arteriotomy side punch tool and/or a bone rongeur forceps were used to make additional holes corresponding to the third and lateral ventricle drainage points which were decided based on preoperative sagittal MR imaging. The ventricular catheter was inserted carefully under neuroendoscopic visualization into the floor of the infratentorial cyst through the aqueduct and released. The proximal catheter was then connected to a shunting system with an adjustable valve draining distally into the peritoneum. Postoperative MR imaging revealed decreased sizes of the supratentorial ventricle and posterior fossa cyst (Fig. 1D–F). The patient was doing well at the 18-month follow-up examination.

Discussion

The goal of treatment in patients with hydrocephalus associated with DWM is the simultaneous drainage of the supra- and infratentorial compartments to preserve the transtentorial pressure equilibrium.¹¹,¹²,¹³ The presence of aqueductal patency is thought to be the deciding factor for the selection of the initial type of shunt for insertion¹¹,¹²,¹³; if the aqueduct is patent, only the insertion of either a VP or CP shunt is required. However, the very high incidence of delayed secondary aqueductal stenosis and isolation of the fourth ventricle after VP shunt placement has necessitated the placement of an additional CP shunt.¹⁰,¹²,¹³,¹⁴ In addition, upward transtentorial herniation due to transtentorial pressure disequilibrium has been reported.¹⁰ Some patients required placement of an additional VP shunt after the insertion of a CP shunt because of persistent ventricular dilation⁶ and chronic downward transincisural herniation.¹⁰ Moreover, CP shunt-associated complications are severe and include brainstem tethering,⁸ brainstem injury,⁹ and posterior fossa hematomas.²,⁵ Frequent disconnection and migration,¹⁰ the need for frequent revisions,¹⁰,¹⁷ and increased incidence of cerebrospinal fluid (CSF) leakage and subsequent infection have been reported in patients treated by only CP shunt placement.⁵,¹⁴,¹⁷ ETV is an alternative treatment, but is controversial in children younger than 2 years because success rates vary widely and ETV carries the risk of secondary aqueductal stenosis.²,⁴,⁷ If shunt malfunction occurs in patients older than 2 years, as in the present case, we would choose shunt revision to avoid the risk of secondary aqueductal stenosis. However, we should consider ETV after the assessment of the CSF reabsorption if frequent shunt malfunctions occur. After ETV, we should continue to monitor the patient for secondary aqueductal stenosis.

Based on these considerations, combined VP and CP shunt placement has been recommended as a first treatment.¹⁰ However, this carries a higher risk for the development of secondary aqueductal stenosis attributable to the great reduction in CSF flow through the aqueduct.¹,¹² The obstruction of one component of the two-shunt system may result in the development of a dangerous transtentorial pressure disequilibrium.²,¹²,¹⁵,¹⁷ Moreover, shunt malfunction attributable to the complexity of the shunt systems has been reported.³ Various new techniques involving the placement of a modified single catheter have been reported. CSF can be drained from the infratentorial cyst via the original distal pore and from the supratentorial ventricle via the additional fenestrated proximal pore. Under stereotactic im-

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age guidance, a multiperforated single ventricular catheter was inserted from a frontal burr hole into both the supra- and infratentorial compartments through the tentorium. This maneuver minimizes the complexity associated with the placement of combined VP and CP shunts, avoids CP shunt-associated complications, and facilitates transtentorial ventricular pressure equalization. However, sophisticated instrumentation and relatively complicated maneuvers are required. A multiperforated single catheter was placed under ultrasonic guidance via a frontal burr hole into both compartments through the dorsal third ventricular wall. Good results were obtained with readily available equipment, but the inability to visualize the intraventricular systems carries the risk of injury to intraventricular vessels. Nonetheless, placement of the modified single catheter was reportedly safe and effective.

Our procedure is relatively simple and feasible. Using a neuroendoscope, the catheter can be inserted under visualization to the floor of the fourth ventricle. The problem with this technique is the post-procedural morbidity of the transferring aqueduct, e.g. internuclear ophthalmoplegia, sixth and seventh cranial nerve palsies, and sybian aqueduct syndrome. A series of 54 patients underwent transaqueductal endoscopic navigation of the fourth ventricle using a flexible endoscope and encountered 2 small ependymal contusions of the opening of the cerebral aqueduct. The only neurological complication in the series was mild diplopia attributable to aqueductoplasty rather than the transaqueductal passage of the endoscope. Therefore, transaqueductal exploration of the fourth ventricle is feasible and reasonably safe. The endoscope was 4 mm in width; ours is a stylet-type endoscope with width of 2.5 mm, the diameter of a shunt tube. Placement of the burr hole must take into account the position of the massa intermedia and insertion of the catheter, especially through the aqueduct, must be performed carefully. Most DWM patients do not present with aqueductal stenosis. However, if aqueductal stenosis is detected before surgery, another procedure should be considered. Aqueductoplasty or stent placement may be necessary in patients with aqueductal stenosis, but this carries a higher risk of injury to the periaqueductal structures. ETV was performed with placement of a stent from the third ventricle to the cyst cavity via the postero-inferior wall of the aqueduct in patients with DWM and aqueduct obstruction. Although repositioning of one stent and one subsequent placement of a VP shunt became necessary in 3 patients, they did not suffer neurological deterioration. Four patients underwent transaqueductal placement of a multiperforated shunt catheter with aqueductoplasty for an isolated fourth ventricle under a 2.5-mm flexible endoscope introduced into the third ventricle, and no perioperative morbidity was encountered. Eleven patients with isolated fourth ventricle were treated with aqueduct plasty and shunts were placed via the cerebral aqueduct in the fourth ventricle with endoscopic assistance. No surgical complications occurred although the endoscope passed through the cerebral aqueduct with shunt catheters in some patients.

These results indicate that transaqueductal shunt placement using a small-diameter stylet-type endoscope is reasonably safe and can be considered, especially in patients with aqueductal patency confirmed by preoperative MR imaging. The accumulation of a large series of patients with hydrocephalus associated with DWM is needed to confirm the efficacy and safety of this technique.

References


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