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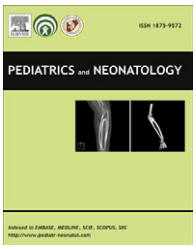

CASE REPORT

Successful Treatment of Dandy—Walker Syndrome by Endoscopic Third Ventriculostomy in a 6-Month-Old Girl With Progressive Hydrocephalus: A Case Report and Literature Review

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Dandy—Walker syndrome (DWS) is a congenital brain malformation involving the cerebellum and fourth ventricle. We report a 6-month-old girl with DWS presenting an initially normal ventricular system and mild cyst-like lesion over the posterior fossa as assessed by postnatal brain sonography. However, symptoms and signs of increased intracranial cerebral pressure in terms of frequent vomiting and tense anterior fontanel developed, and these were associated with mild hypotonia and poor neck support, and upward-gaze palsy at the age of 6 months. Magnetic resonance imaging revealed a huge cystic lesion of the fourth ventricle, which filled the posterior fossa and ventricular dilatation. The tentorium was progressively displaced upward by the cyst. A nearly complete agenesis of the cerebellar vermis was also confirmed. After a successful endoscopic third ventriculostomy, a series of brain magnetic resonance imaging scans, taken during a follow-up survey, showed normal lateral and third ventricles. Consequently, symptoms of intracranial cerebral pressure resolved, and a developmental milestone was achieved. In conclusion, DWS can be confirmed postpartum, and endoscopic third ventriculostomy was found to be a preferential operative procedure for DWS with hydrocephalus. It may be effective for patients younger than 1 year.

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1. Introduction

Dandy—Walker syndrome (DWS) is characterized by a large posterior cranial fossa cyst, partial or complete agenesis of the cerebellar vermis, and high position of the tentorium.[1] Hydrocephalus may cause increased intracranial cerebral pressure, progressive enlargement of the head, convulsions, mental disability, and even death. Although hydrocephalus is not an absolute criterion for the diagnosis of DWS, complications from it are common, and 5—10% of congenital hydrocephalus cases are found in DWS.[2]

Traditionally, patients with DWS-related hydrocephalus required combinations of shunting systems to effectively drain the supratentorial ventricles and posterior fossa cyst.[2] However, frequent shunt dependency and malfunction have been constant problems. With the growing popularity of neuroendoscopy in managing obstructive hydrocephalus, endoscopic procedures have become more widely favored than shunt placement,[3,4] but using endoscopic third ventriculostomy (ETV) in DWS with hydrocephalus is still not standardized.

Here, we report a 6-month-old girl with DWS complicated by hydrocephalus. She made marked improvement after an ETV. Early surgical intervention, as a result of clinical suspicion, may account for her good recovery.

2. Case Report

A 6-month-old girl presented with an accelerated enlargement of head circumference (HC) since the age of 4 months. She was preterm, with a gestational age of 34 weeks and 5 days, and was delivered by means of Cesarean section because of preeclampsia and fetal distress. We had performed brain sonography after her birth, and only a vague small cyst in the posterior fossa was revealed (Figure 1). In addition, she was small for gestational age and had neonatal hypoglycemia and patent ductus arteriosus. The growth curve of HC was 25th percentile at birth, 90th percentile at 4 months, and 95th percentile at 6 months of age. Subsequently, this rapid increase of HC led to increased intracranial cerebral pressure signs in terms of frequent vomiting, tense anterior fontanel, and hypotonia. She was then brought to hospital for help, where brain magnetic resonance imaging (MRI) showed a large cyst in the posterior fossa, connecting with the opening of the fourth ventricle. This was associated with atrophic change of the inferior cerebellar vermis and was compatible with the diagnosis of Dandy—Walker malformation (Figure 2).

Grade III heart murmur was found from auscultation, and patent ductus arteriosus was detected by echocardiogram. Neurological examination revealed a bulging forehead, widening anterior fontanel (more than 2 fingers' width), mild hypotonia with poor neck support, and upward gaze palsy. The patient underwent ETV on the 10th day of admission. ETV was performed through a precoronal burr hole and transfrontal approach. All procedures were smooth, and the patient recovered uneventfully.

In the following year, she received brain MRI every 6 months (Figure 3). The lateral ventricles and HC size (25th percentile at 13 months) were normalized after surgery, but the posterior fossa cyst persisted. Consequently, vomiting subsided and gross development seemed acceptable other than language delay and a mild unsteady gait in clinical presentation for more than 1 year's follow-up post-ETV.

3. Discussion

By definition, DWS is a congenital brain anomaly, and more than 85% of cases are found before the age of 1 year.[5] In several earlier publications, investigators have stated that the incidence of hydrocephalus was around 86—94%.[6] Some

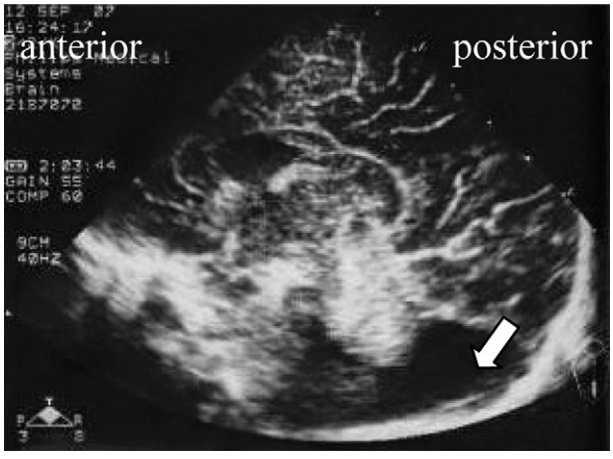

Figure 1 Sagittal view of brain sonography at the neonatal stage. Only a vague small cyst (arrow) at the posterior fossa was noted.

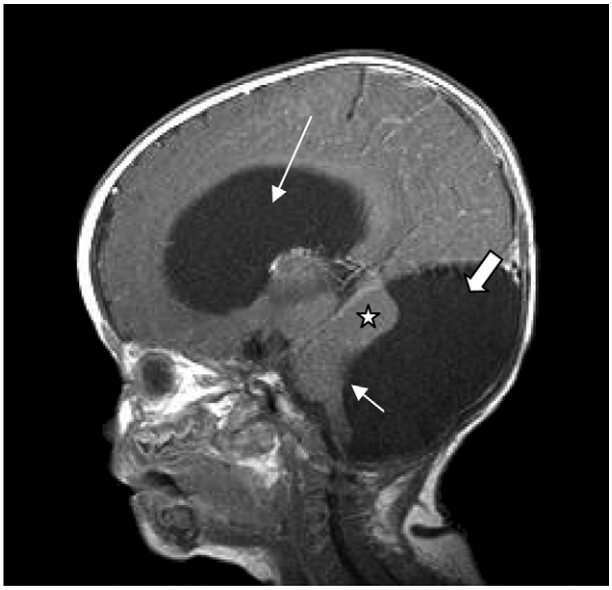

Figure 2 Magnetic resonance imaging before surgery. A large cyst (boldface arrow) in the posterior fossa, connecting with the opening of the fourth ventricle (short arrow), associated with compressed change of the inferior cerebellar vermis (star) and marked hydrocephalus (long arrow), is compatible with the diagnosis of Dandy—Walker malformation.

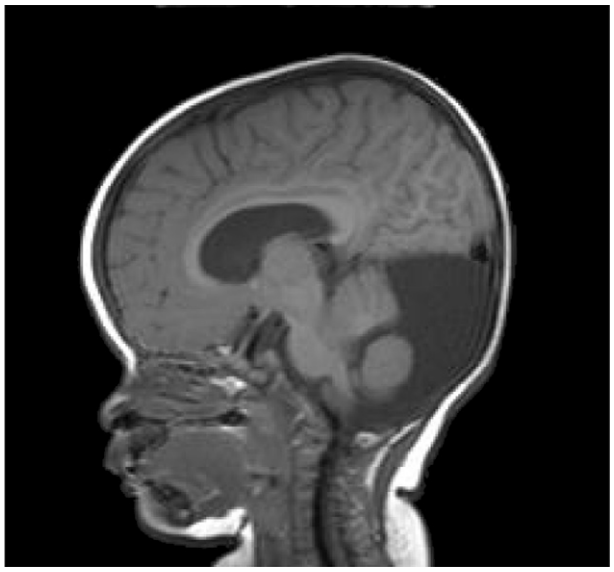

Figure 3 Magnetic resonance imaging 6 mo postsurgery follow-up. The hydrocephalus had improved and cyst had shrunk compared with those in Figure 2.

obstetricians have found this malformation by means of prenatal routine sonography during the antenatal period, especially during the later prenatal period.[7] However, in other studies, there was no evidence of hydrocephalus in 80% of patients with DWS after prenatal ultrasonographic studies, suggesting the probable development of hydrocephalus during the neonatal period.[8] Correspondingly, the case we reported presented similar clinical manifestations; thus, an early diagnosis and management are both important for such cases.

In the late 1980s, ETV gained an established place in the surgical treatment of obstructive hydrocephalus. It is a surgical procedure in which an opening is created in the floor of the third ventricle using an endoscope placed within the ventricular system through a burr hole.

This allows the cerebrospinal fluid to flow directly to the basal cisterns, thereby shortcutting any obstruction. It is used to treat certain forms of obstructive hydrocephalus, such as aqueductal stenosis. Complications, which include subarachnoid hemorrhage, basilar artery injury, and hypothalamic or pituitary injury, may be reduced or avoided by a highly skilled surgeon.[9]

Compared with ETV, traditional shunt insertion in treating patients with DWS has been well established. However, infection, malposition, migration, and dysfunction are always matters of concern. ETV seems to be less invasive and closer to physiological conditions.[10]

A current review article revealed very low success rates of ETV in the first months of life, which raises the question as to whether or not this procedure should be offered at all to very young patients and, hence, possibly be reserved for infants beyond the first or second year of life.[11] Alternatively, some opposing opinions were proposed suggesting that the success of ETV is related to the etiology rather than to age. This suggests that there are no grounds for denying children younger than 2 years this chance for a shunt-free life.[10] As the case with DWS we are reporting had successfully undergone ETV at her age of 6 months, this supports the point of view of treatment with ETV being more etiologically related rather than being age dependent. Nevertheless, more evidence and more cases should be studied to clarify this controversy.

According to a report illustrating that only 16—33% of patients had smaller ventricles after this procedure, ventricle size often does not change after ETV.[12] Findings, such as resolution of papilledema, normalization of ICP, relaxation of the fontanel, and stable HC are considered valuable for assessing the success or failure of ETV.[9] Effectively, this girl received ETV at the age of 6 months, and the subsequent brain MRI showed adequate drainage of ventricular cerebrospinal fluid to the cisterna magna, reduced size of the lateral ventricles, and relatively normalized HC development. Moreover, some neurosurgeons suggest that the routine insertion of a ventricular reservoir allows life-saving emergency therapeutic cerebrospinal fluid aspiration for sudden neurological deterioration caused by ventriculostomy failure. It also assists in the diagnosis of ETV failure and subsequent emergency ventricular access.[13]

Another noteworthy issue is the need for extensive follow-up of DWS patients. One study showed that the patency rate in patients who had undergone ETV decreased over time to 44% at 10 years postsurgery.[14] Thus, long-term radiological follow-up should be done in these cases, and the procedure should be repeated if necessary.

In conclusion, ETV may be considered as a safe surgery for DWS patients with hydrocephalus, even in a patient younger than 1 year, as shown in this case. In addition, the insertion of a shunting system may not be suggested unless the use of ETV fails. This procedure of ETV seems to have significant advantages over shunting when used in appropriate cases.

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