

Case Report

An Asymptomatic Child with a Large Dandy–Walker Cyst despite the Presence of a Ventriculoperitoneal Shunt

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Keywords

• Dandy–Walker syndrome; Hydrocephalus; Fourth ventricle; Ventriculoperitoneal shunt

Abstract

Introduction: Hydrocephalus occurs in 80% of patients with Dandy–Walker (DW) syndrome. According to the literature, when permeability of the cerebral aqueduct is demonstrated, it is possible to use a single shunt, either ventriculoperitoneal or cystoperitoneal. The case of an asymptomatic child with a large Dandy–Walker cyst despite the presence of a ventriculoperitoneal shunt is reported herein.

Case report: A 1-year-old female child was brought to an emergency department (General State Hospital) with a clinical picture of macrocephaly and signs of intracranial hypertension (headache, vomiting, and papilledema). A CT scan of the head revealed hydrocephalus and other changes, such as a cyst in the fourth ventricle and hypoplasia of the cerebellar vermis, which were compatible with Dandy–Walker malformation (DWM). It was decided to place a ventriculoperitoneal shunt. After six years of follow-up, the child still had the cyst in the fourth ventricle, which caused compression of the brainstem and hypoplasia of the cerebellar vermis, but there was no hydrocephalus. Nevertheless, the child remains asymptomatic, with normal motor development.

Discussion/Conclusion: The treatment of hydrocephalus and DWM remains complex and therefore controversial. However, some children may benefit from a ventriculoperitoneal shunt and remain asymptomatic despite the persistence of Dandy–Walker cysts.

ABBREVIATIONS

CP: Cystoperitoneal; DW: Dandy–Walker; DWM: Dandy–Walker Malformation; ETV: Endoscopic Third Ventriculostomy; VP: Ventriculoperitoneal

INTRODUCTION

The first report of a patient with hypoplasia, a cyst in the fourth ventricle, and hydrocephalus was published by Setton in 1887 [1]. In 1914, Walter Dandy and Kenneth Blackfan described the case of a 13-month-old child with similar findings and absence of the foramina of Luschka and Magendie [2]. In 1942, Taggart and Walker described three children with the same triad and confirmed that the absence of the foramina of Luschka and Magendie contributes to the formation of a cyst in the fourth ventricle [3]. Benda, in 1954, was the first to use the term “Dandy–Walker (DW) syndrome” and also the first to report that a malformation in the fourth ventricle region would not result merely in atresia of the foramina of Luschka and Magendie [4]. Hydrocephalus occurs in over 80% patients with DW malformation (DWM), most often within the first three months of life [5,6]. The present case report describes an asymptomatic child with a DW cyst that did not disappear after hydrocephalus was treated only with a ventriculoperitoneal shunt.

CASE PRESENTATION

A one-year-old female child was brought to an emergency

department with a clinical picture of macrocephaly and signs of intracranial hypertension (headache, vomiting, and papilledema). A CT scan of the head revealed hydrocephalus and other changes, such as a cyst in the fourth ventricle and hypoplasia of the cerebellar vermis, which were compatible with DWS. Because of the signs of intracranial hypertension, it was decided to only place a ventriculoperitoneal shunt (Figure 1). The child showed good progress after the procedure, with significant improvement of the hydrocephalus and no complications (infections or impairments). After six years of follow-up, the child still had the large cyst in the fourth ventricle, which caused compression of the brainstem (Figure 2); however, the child remained asymptomatic (Figure 3), with normal psychomotor development.

DISCUSSION

Some of the hypotheses for the pathophysiology of hydrocephalus in DWM are: 1) atresia of the foramina of Luschka and Magendie, which is questionable in some cases in which the foramina are observed to be permeable and hydrocephalus is not present at birth [7,8]; 2) aqueductal stenosis, despite the fact that in typical DWM, the aqueduct is clearly permeable, with free communication between the third and fourth ventricles [9,10]; 3) arachnoiditis, in which the obstruction of the cerebrospinal fluid (CSF) flow may also be distal to the outlets of the fourth ventricle. Basal arachnoiditis may cause CSF flow blocks in the cisterna magna [11,12], the premedullary cistern [13], or the incisura [9].

DWM treatment options are. 1) Excision of the cyst membrane was the first surgical technique to be proposed [2,3], but it should no longer be considered as a treatment option, owing to its morbidity and mortality risks [14]. 2) Shunting, either ventriculoperitoneal (VP), cystoperitoneal (CP), or combined (VP/CP), should be considered as the treatment of choice. Aqueduct permeability is a prerequisite for a single shunt (VP or CP) [15]. VP shunts are easier to insert than CP shunts and

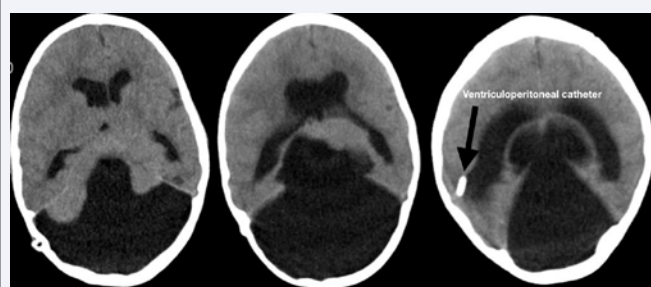


Figure 1 One Year old female with Ventriculoperitoneal shunt and Dandy-Walker Cyst.

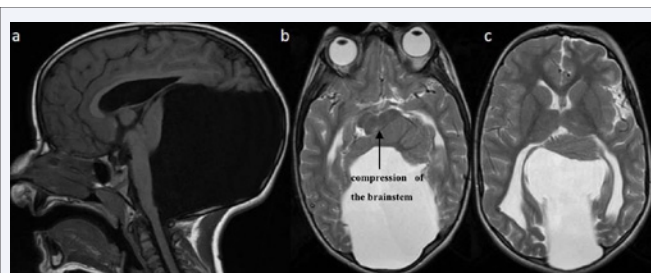


Figure 2 Saggital T1 MRI (a) and Axial T2 MRI (b,c) with compression of Brainstem.

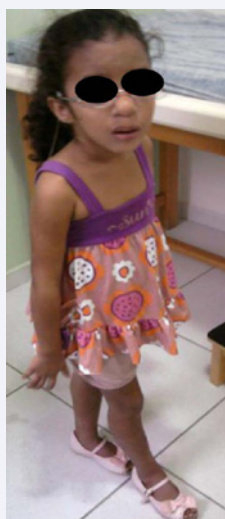


Figure 3 Asymptomatic Child.

also have a lower incidence of migration. Some authors prefer a combination of shunts to equalize the pressure across the tentorium [14]. 3) Neuroendoscopy, namely endoscopic third ventriculostomy (ETV), is a good treatment option for elderly patients and patients with shunt dysfunction [15].

The treatment of hydrocephalus and DWM remains complex, therefore controversial. The therapeutic strategy of using a single shunt, particularly the VP shunt, appears to be a good option because it is easier to insert and is associated with lower rates of complications than the CP shunt or the VP/CP combination.

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