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CASE REPORT

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Short-term subarachnoid space drainage: a potential treatment for extraventricular hydrocephalus

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Abstract *Introduction:* Extraventricular hydrocephalus (EVOH), defined as the enlargement of all CSF compartments in the absence of an obstructing lesion, is usually associated with an increased head circumference and a relatively benign clinical course. Occasionally, because of concern about increased intracranial pressure (ICP), treatment with diuretics is initiated. In most cases, surgical drainage or diversion is not indicated. EVOH may follow a more alarming clinical course and be associated with developmental delay and/or other symptoms of increased ICP. *Case report:* We describe a 6-month-old girl with EVOH and developmental delay who was treated with temporary drainage of the subarachnoid space. Clinical response was immediate, with stabilization of the head circumference and improvement in motor performance. We propose that such a surgical procedure might be considered for more frequent use in selected cases.

Keywords Extraventricular hydrocephalus · Drainage · Infant

Introduction

External or extraventricular hydrocephalus (EVOH) is a form of communicating hydrocephalus in which the subarachnoid spaces are enlarged mainly at the convexity, (mostly frontal) and the inter-hemispheric spaces [4, 12]. EVOH is generally benign and self-resolving [9, 10], and may be associated with familial macrocephaly [14, 16]. Some affected infants present with psychomotor delay and accelerated increase in head circumference, raising the possibility of a causal relationship between the two [15]. When increased intracranial pressure (ICP) is suspected, treatment with oral acetazolamide and furosemide may be recommended to reduce cerebrospinal fluid (CSF) production and pressure. In most cases, surgical drainage is not required.

Although the name "benign" external hydrocephalus has been widely used as an alternative to EVOH, 10–15% of these children express some form of psychomotor delay of varying magnitude [6]. Therefore, more aggressive surgical options are occasionally discussed in cases with a significant clinical course.

We describe a child with EVOH in whom a different approach was applied, which led to an abrupt resolution of the clinical syndrome.

Case report

The child was born at 30 weeks' gestation with a birth weight of 1,590 g (appropriate for the gestational age). The parents were 25 years old, healthy, and unrelated. She was placed on mechanical ventilation for 3 days and discharged home at the gestational age 35 weeks. Physical and neurologic examinations at discharge revealed no abnormalities. Head circumference was in the 3rd percentile for the chronological age. Brain ultrasonography showed normal ventricular size and no ventricular hemorrhage.

At the age of 5 months, the child was referred to the ambulatory pediatric clinic at our hospital because of accelerated head growth velocity. Her head circumference at the time was at the 98th percentile (Fig. 1), corrected for the gestational age. Findings

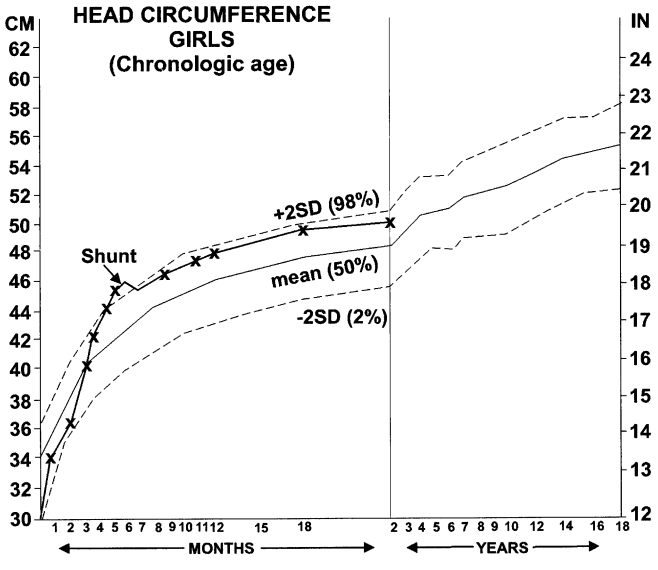

Fig. 1 Head growth chart with indication of drainage timing

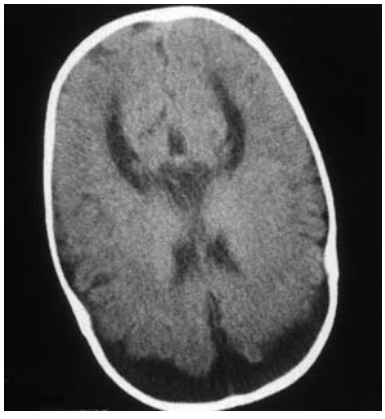

Fig. 2 Preoperative CT of the brain at 6 months of age. There is a widening of all CSF spaces, including a large amount of subarachnoid fluid collected around the frontal cortex

on physical and neurological examinations were normal. It was noted that the mother's head circumference was at the 98th percentile and the father's at the 50th. The child was irritable, and appeared to have sunset eyes. The parents reported developmental arrest in the last 2 months. Computed tomography of the head showed dilation of CSF spaces, with a large collection of fluid around the frontal cortex (Fig. 2). Treatment with acetazolamide was initiated, but proved ineffective, with a continued increase in head circumference.

The child was referred for neurosurgical consultation. A bilateral drainage tube was inserted via burr holes into the frontal subarachnoid space; after 48 h, 300 cc of clear CSF were drained. The tubes were then removed and the child was discharged. The parents described an immediate alleviation of her restlessness and an improved appetite.

Follow-up examination at the age of 8 months, 2 months following the procedure, showed that the head circumference had remained in the 98th percentile. The child had resumed psychomotor progress and was now able to roll and sit by herself. Computed tomography showed a modest reduction in the size of the CSF spaces. At the ages of 18 and 24 months, her head circumference was in the 90th percentile, and psychomotor development was normal for her age. There were no pathologic findings on neurological examination.

Discussion

Most patients with EVOH present between the ages of 2 and 10 months and there is a slight male predominance. The characteristic manifestation is macrocephaly. The head circumference is generally above the 90th percentile with normal or increasing growth velocity. The anterior fontanel feels normal or full [4, 14, 16]. The typical radiologic findings are mild to moderate ventricular enlargement, widened frontal subarachnoid space, interhemispheric and sylvian fissures, and enlargement of the basilar cisterns [4, 16]. The pathogenesis of EVOH remains controversial. Cundall et al. [5] suggested a genetic basis and Govaert et al. [6] proposed an undetected primary subarachnoid hemorrhage in the neonatal period caused by traumatic delivery.

The favorable prognosis for EVOH reported in earlier studies has been questioned by several authors [1, 3, 4, 7, 13, 16]. Most of the latter found that even the children with normal development by the age of 15–18 months had at least had a transient arrest in language and gross motor skills at the age of 5–12 months [1, 4, 16]. Azais and Echenne [3] reported muscular hypotony and motor delay in 30% of 41 infants with EVOH. Affected children were found to have poor performance in upper-limb tasks and poor visuomotor control, running speed, bilateral coordination, and visuomotor integration, in addition to minor neurologic indicators [13]. Nevertheless, it is not known whether treatment of any kind influences the neurodevelopmental course.

Extraventricular hydrocephalus is rarely complicated by subdural hemorrhage [3, 4, 12, 14], manifested by a full fontanel, emesis, or depressed level of consciousness. The presence of subdural hemorrhage or markedly enlarged ventricles may necessitate a diversion procedure [4, 11, 14]. Some authors suggest a temporary subdural peritoneal shunt for the subdural effusion [18].

For uncomplicated EVOH, treatment is mostly conservative. In the series of Andersson et al. [2], 7 patients underwent exploratory craniotomy and a shunt was placed in 2. The authors concluded that shunting was unnecessary. None of the 137 infants treated by Alvarez et al. [1] or Maytal et al. [8] received any medical or surgical intervention, and head growth stabilized spontaneously in all cases. Shinnar et al. [17] suggested medical therapy with acetazolamide and furosemide as an effective alternative to shunting, to halt the progression of hydrocephalus until spontaneous arrest occurs.

Although a conservative treatment approach to EVOH is generally adopted, concerns about the possi-

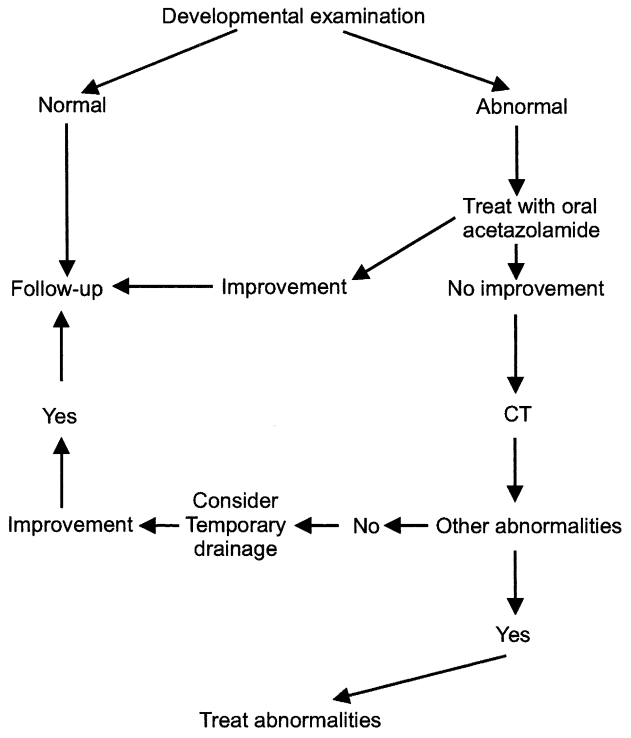

Fig. 3 Flow chart for the management of extraventricular hydrocephalus

ble deleterious effects of increased ICP have been raised, especially in the presence of an acute, or subacute, increase in head circumference. Physicians may consider a diversion procedure either in the presence of an acute increase in head circumference and when the intraventricular component is significantly enlarged, or when the extra-axial spaces in the brain convexities are very thick and seem to be under pressure. If the ventricles are enlarged, the situation may be better categorized under "communicating hydrocephalus" and not EVOH, and thereby require a standard ventriculo-peritoneal shunt. Subdural effusions may be difficult to differentiate from subarachnoid collections on imaging. Subdural collections (hematomas), a frequent occurrence in adults, are commonly treated with temporary drainage.

In our patient, accelerated head growth and developmental arrest continued despite treatment with acetazolamide. Drainage revealed a true subarachnoid collection. The CSF was clear, had a normal protein level, and contained no blood. The change in the clinical course of the disease was abrupt: there was an immediate reduction in tension in the fontanel and a reduction in head circumference (Fig. 1). The child showed less irritability, and soon afterward, an improvement in motor performance. In EVOH, the regular course of improvement is usually slow, terminating only after about 1 year of age.

A rapid change such as that seen in our patient almost never occurs spontaneously. We therefore assume that it was attributable to the drainage procedure.

To the best of our knowledge, this is the first documented report of the use of a temporary subarachnoid drain to treat EVOH. Although the procedure may be considered invasive, the response was immediate and persisted throughout the 24 months of follow-up.

A flow chart for the proposed management of EVOH is presented in Fig. 3. We suggest that temporary subarachnoid drainage be considered in patients with severe EVOH, accelerated head growth, and a worrisome neurological developmental course who do not respond to oral treatment.

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