External hydrocephalus

Key concepts

- enlarged subarachnoid spaces over the frontal poles in the first year of life
- ventricles are normal or minimally enlarged
- may be distinguished from subdural hematoma by the “cortical vein sign”
- usually resolves spontaneously by 2 years of age

Enlarged subarachnoid space (usually over the cortical sulci of the frontal poles) seen in infancy (primarily in the first year of life), usually accompanied by abnormally increasing head circumference with normal or mildly dilated ventricles. There are often enlarged basal cisterns and widening of the anterior interhemispheric fissure. No other symptoms or signs should be present (although there may be slight delay only in motor milestones due to the large head). Etiology is unclear, but a defect in CSF resorption is postulated. External hydrocephalus (EH) may be a variant of communicating hydrocephalus. No predisposing factor may be found in some cases, although EH may be associated with some craniosynostoses (especially plagiocephaly), or it may follow intraventricular hemorrhage or superior vena cava obstruction.

External hydrocephalus (EH) is a rapid increase in head circumference, combined with enlarged subarachnoid spaces especially overlying the frontal lobes and normal or only moderately enlarged ventricles.

Many other terms have been used for the same or similar conditions, for instance, “subdural hygroma”, “subdural effusion”, “benign subdural collections”, “extraventricular obstructive hydrocephalus”, “idiopathic/benign hydrocephalus”, “primitive megalencephaly”, “enlargement of the subarachnoid spaces”, or even “chronic subdural hematoma”.

The anatomical substrate, whether this is subdural fluid or CSF in the subarachnoid space, has been subject to disagreement

Etiology

The cause is unknown, but many patients have a family history consistent with autosomal dominant inheritance.

Several pathogenic hypotheses have been proposed but the most accredited theory seems to be delayed maturation of the arachnoid villi. There is a consensus that this is a benign entity, correlated to a familial predisposition and, in some cases, inheritance.

Secondary cases of external hydrocephalus are associated with hemorrhage, meningitis, and elevated venous pressure.

Elevated venous pressure has been shown to be a much more common cause of communicating hydrocephalus in children than previously thought.

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The absorption of CSF in infants is into the capillary bed of the deep white matter rather than the arachnoid granulations. Absorption into a capillary bed depends on hydrostatic pressure. Similar to older children with communicating hydrocephalus, the infants in this cohort with external hydrocephalus showed evidence of an elevation in venous pressure. Elevated venous pressure may be a much more common cause of external hydrocephalus than previously recognized.\(^8\)

The hypothesis that encephalocranial disproportion is the basic underlying entity for the CT images was proposed.\(^9\)

Transplacental acquisition of anti-Ro antibodies has been associated with external hydrocephalus.\(^10\)

**Diagnosis**

Posterior mild ventricular dilatation and prominent subarachnoid spaces in a posterior distribution can be considered an early stage of benign external hydrocephalus that is nicely illustrated by MRI.\(^11\)

CT and MRI can provide a highly accurate diagnosis in these patients, allowing a preliminary assessment of the prognosis, particularly regarding the enlarged subarachnoid space limits and the "cortical vein" sign which can predict a further complication. These results are obtained with the same examination performed in a standard CT or MRI study of the brain and no injection of contrast medium is needed.\(^12\)

**Ultrasound**

Trounce et al., describe five infants with the appearance of external hydrocephalus diagnosed by real-time cranial ultrasound. The indication for scanning in all cases was a head circumference crossing the 90th centile. The interhemispheric fissure is widened with the falx usually visible and the cortical surface can be seen beneath the anterior fontanelle. There is minimal, if any, ventricular dilatation and none of the children went on to develop internal hydrocephalus. Two children had minor motor problems but there were no other neurodevelopmental sequelae.\(^13\)

**Differential Diagnosis**

EH is probably distinct from benign subdural collections (or extra-axial fluid) of infancy.

★ EH must be distinguished from symptomatic chronic extra-axial fluid collections (or chronic subdural hematoma), which may be accompanied by seizures, vomiting, headache... and maybe the result of child abuse. With EH, MRI or CT may demonstrate cortical veins extending from the surface of the brain to the inner table of the skull coursing through the fluid collection ("cortical vein sign"), whereas the collections in subdural hematomas compress the subarachnoid space, which apposes the veins to the surface to the brain.

**Treatment**

Considering the few studies that have dealt with the effect of treatment of external hydrocephalus, it is obvious that more knowledge is needed. For now, the apparent diversity in results and opinions probably reflects a similar variety in clinical courses and patients, this again reflecting the different etiologies and partial inheritance often seen as well as the differences in what is regarded as "normal." A good way to answer some of these questions is to carry out a larger population-based (retrospective) study, comparing treated (shunted) and untreated children with external hydrocephalus and focusing on developmental outcome on long-term follow-up, including the use of standardized neuropsychological tests. By doing this, it may be possible to reveal subtypes/subgroups...
of patients with different outcome prognoses, hence in need of different initial managements. Surgical indication could, for instance, be determined by the initial radiological presentation (width of subarachnoid space, diffusion-weighted MRI), by a thorough neuropsychological investigation, or maybe by a combination of all signs and tests available (ICP, CSF flow, etc.) \(^{(14)}\).

**Medical Therapy**

Several studies describe temporary acetazolamide treatment lasting for 1–2 months, resulting in a gradual reduction of excessive head growth. Furthermore, Roshan et al. used acetazolamide combined with mannitol in four patients who presented with vomiting, irritability, and a bulging fontanel. The patients responded well.

Acetazolamide and **furosemide** have been recommended for mild hydrocephalus of the newborn and in infants, but based on large, randomized trials it is not recommended for the treatment of posthemorrhagic ventricular dilatation in infancy \(^{(15)}\).

**Exploratory craniotomy**

7 patients were subjected to exploratory craniotomy which disclosed a deep arachnoid space. 2 patients were shunted. All follow-up CT examinations showed normal conditions.

Andersson et al., suggest that infants with clinical signs of hydrocephalus and CT picture of external hydrocephalus should not be treated with shunt. The widening of the subarachnoidal space will normalize. The rate of headgrowth will also normalize \(^{(16)}\).

**Outcome**

It occurs mainly during infancy, and the subarachnoid space enlargement gradually decreases and disappears over the next years \(^{(17)}\) \(^{(18)}\) \(^{(19)}\).

In sixteen patients with this condition, 7 of which had a family history of megalo-encephaly and 4 had delayed motor development, although it was transient in 3 of the cases. The cranial circumference was normal at about 18 months of age, but 4 patients had megalo-encephaly after 3 years of age. The radiological images were normal in all cases between 24 and 48 months of age without treatment \(^{(20)}\).

Idiopathic external hydrocephalus is a relatively benign, self-limited condition that resolves without treatment and is closely related to benign familial macrocephaly \(^{(21)}\).

In 99 patients, 5-12 years old (55% males). Twenty were born prematurely, 12 with <33 weeks gestation. Children presented at an average age of 9 ± 4.8 months (mean ± SD). The presenting complaint was macrocephaly in 65 cases. Other presenting findings were positional head shape deformity and torticollis; 10% had a family history of macrocephaly. Developmental delay was present in 21% of patients (4% verbal, 20% gross motor, 4% fine motor delay). Four patients had small subdural hematomas, none with suspicion of a non-accidental trauma. During clinical reassessment over a mean follow-up of 13 months, the average head percentile was stable and none of the patients developed new subdural hematomas. Gross motor delay resolved in 15/20 and fine motor delay in 4/4 patients. Verbal delay resolved in 2/4 patients, but interestingly, was newly detected in 6 other children. None of the patients required cerebrospinal fluid shunting. The response rate to the HOQ was 25% (median age 7 years, 74% females). The average overall HOQ score was 0.75 ± 0.24 versus 0.68 ± 0.19 for a previously published cohort of shunted hydrocephalic children.

In this series generally saw resolution of presenting motor developmental delays; however, new
verbal delays were detected in a non-trivial number of patients. Quality of life measurements suggest some reduction in health status, but less so than is seen with shunted hydrocephalus 22).


