External hydrocephalus

The term “External Hydrocephalus” has also been used to describe the presence of extra ventricular cerebrospinal fluid (CSF) collections accompanied by hydrocephalus, particularly in cases of adults suffering from aneurysmal subarachnoid hemorrhage and severe head injuries. Several other terms have been used to describe this entity which has lead to confusion about this disease [1)]

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 [2) 3) 4) 5) 6) 7)].

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” [8)]

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

Etiology

External hydrocephalus etiology.

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 [9].

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 [10].

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 [11].

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

External hydrocephalus treatment.

Outcome

It occurs mainly during infancy, and the subarachnoid space enlargement gradually decreases and disappears over the next years [12] [13] [14]

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 [15].

Idiopathic external hydrocephalus is a relatively benign, self-limited condition that resolves without treatment and is closely related to benign familial macrocephaly [16].

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 [17].

1)
Tzerakis N, Orphanides G, Antoniou E, Sioutos PJ, Lafazanos S, Seretis A. Subdural effusions with hydrocephalus after severe head injury: successful treatment with ventriculoperitoneal shunt placement: report of 3 adult cases. Case Rep Med. 2010;2010:743784. doi: 10.1155/2010/743784. Epub 2010 Dec 12. PMID: 21209815; PMCID: PMC3014823.
2)
Alvarez LA, Maytal J, Shinnar S. Idiopathic external hydrocephalus: natural history and relationship to benign familial macrocephaly. Pediatrics. 1986 Jun;77(6):901-7. PubMed PMID: 3714384.
3)
Kumar R. External hydrocephalus in small children. Childs Nerv Syst. 2006;22:1237–1241
4)
Machado MAD, Vieira NA, Matos PE, Matos HD, Barbosa VA, Puglio N, Bacelar A. External hydrocephalus—a review of 15 cases. International Journal of Neuroradiology. 1999;5:266–270.
5) 14)
Muenchberger H, Assaad N, Joy P, Brunsdon R, Shores EA. Idiopathic macrocephaly in the infant: long-term neurological and neuropsychological outcome. Childs Nerv Syst. 2006;22:1242–1248.
6)
Raimondi AJ. A unifying theory for the definition and classification of hydrocephalus. Childs Nerv Syst. 1994;10:2–12.
7)
Robertson WC, Jr, Gomez MR. External hydrocephalus. Early finding in congenital communicating hydrocephalus. Arch Neurol. 1978;35:541–544.
8)
Zahl SM, Egge A, Helseth E, Wester K. Benign external hydrocephalus: a review, with emphasis on management. Neurosurg Rev. 2011 Oct;34(4):417-32. doi: 10.1007/s10143-011-0327-4. Epub 2011 Jun 7. Review. PubMed PMID: 21647596; PubMed Central PMCID: PMC3171652.
9)
Girard NJ, Raybaud CA. Ventriculomegaly and pericerebral CSF collection in the fetus: early stage of benign external hydrocephalus? Childs Nerv Syst. 2001 Apr;17(4-5):239-45. PubMed PMID: 11398943.
10)
Marino MA, Morabito R, Vinci S, Germanò A, Briguglio M, Alafaci C, Mormina E, Longo M, Granata F. Benign external hydrocephalus in infants. A single centre experience and literature review. Neuroradiol J. 2014 Apr 30;27(2):245-50. doi: 10.15274/NRJ-2014-10020. Epub 2014 Apr 18. PubMed

PMID: 24750715.
11)

Trounce JQ, De Vries L, Levene MI. External hydrocephalus–diagnosis by ultrasound. Br J Radiol. 1985 May;58(689):415-7. PubMed PMID: 3904903.
12)

Kumar R. External hydrocephalus in small children. Childs Nerv Syst. 2006;22:1237–1241.
13)

Maytal J, Alvarez LA, Elkin CM, Shinnar S. External hydrocephalus: radiologic spectrum and differentiation from cerebral atrophy. AJR Am J Roentgenol. 1987;148:1223–1230.
15)

Palencia Luaces R, Aldana Gómez J, Tresierra Unzaga F. [Idiopathic external hydrocephaly and familial macrocephaly in infancy]. An Esp Pediatr. 1992 Mar;36(3):186-8. Spanish. PubMed PMID: 1580426.
16)

Alvarez LA, Maytal J, Shinnar S. Idiopathic external hydrocephalus: natural history and relationship to benign familial macrocephaly. Pediatrics. 1986 Jun;77(6):901-7. PubMed PMID: 3714384.
17)

Yew AY, Maher CO, Muraszko KM, Garton HJ. Long-term health status in benign external hydrocephalus. Pediatr Neurosurg. 2011;47(1):1-6. doi: 10.1159/000322357. Epub 2011 Jul 21. PubMed PMID: 21778677.