Benign external hydrocephalus: a review, with emphasis on management

Sverre Morten Zahl · Arild Egge · Eirik Helseth · Knut Wester

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Abstract Benign external hydrocephalus in infants, characterized by macrocephaly and typical neuroimaging findings, is considered as a self-limiting condition and is therefore rarely treated. This review concerns all aspects of this condition: etiology, neuroimaging, symptoms and clinical findings, treatment, and outcome, with emphasis on management. The review is based on a systematic search in the Pubmed and Web of Science databases. The search covered various forms of hydrocephalus, extracerebral fluid, and macrocephaly. Studies reporting small children with idiopathic external hydrocephalus were included, mostly focusing on the studies reporting a long-term outcome. A total of 147 studies are included, the majority however with a limited methodological quality. Several theories regarding pathophysiology and various symptoms, signs, and clinical findings underscore the heterogeneity of the condition. Neuroimaging is important in the differentiation between external hydrocephalus and similar conditions. A transient delay of psychomotor development is commonly seen during childhood. A long-term outcome is scarcely reported, and the results are varying. Although most children with external hydrocephalus seem to do well both initially and in the long term, a substantial number of patients show temporary or permanent psychomotor delay. To verify that this truly is a benign condition, we suggest that future research on external hydrocephalus should focus on the long-term effects of surgical treatment as opposed to conservative management.

Keywords Communicating hydrocephalus · Outcome studies · Macrocephaly · Subarachnoid space · Intracranial pressure

Introduction

Hydrocephalus is a relatively common neuropediatric condition, with an incidence of about 0.9 per 1,000 births [106, 170]. It is defined as the abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles and/or subarachnoid spaces, leading to an increase in intracranial pressure (ICP) [77]. Raimondi defined it as an increase in CSF volume [140].

The subtype “external hydrocephalus” is usually defined as a rapid increase in head circumference, combined with enlarged subarachnoid spaces as seen on neuroimaging—especially overlying the frontal lobes—and normal or only moderately enlarged ventricles [4, 91, 105, 118, 140, 143]. It occurs mainly during infancy, and the subarachnoid space
enlargement gradually decreases and disappears over the next years [91, 110, 118].

Many other terms have been used for the same or similar conditions, for instance, “subdural hygroma” [30], “subdural effusion” [92], “benign subdural collections” [142], “extraventricular obstructive hydrocephalus” [132], “idiopathic/benign hydrocephalus” [4, 118], “primitive megalencephaly” [95], “enlargement of the subarachnoid spaces” [115], or even “chronic subdural hematoma” [93]. As some of these names clearly are used for totally different conditions, they will not be a part of this review.

The many terms probably reflect the different views on etiology and outcome (see the following text discussions) and the often difficult neuroimaging differentiation between these conditions. Even more, the anatomical substrate, whether this is subdural fluid or CSF in the subarachnoid space, has been subject to disagreement [3, 21].

The word “benign” is often used together with “external hydrocephalus,” reflecting the common view that this is a self-limiting condition occurring during infancy, resolving spontaneously during childhood [6, 20, 77, 125, 129, 163]. Hence, most patients are probably not treated.

The aim of this study is to provide a complete review of the literature, focusing on all aspects of external hydrocephalus: etiology, neuroimaging, symptoms, treatment, and outcome.

Materials and methods

As mentioned above, many different terms have been used concerning benign external hydrocephalus or similar conditions. In order to obtain all relevant information, we therefore included these terms in our search. However, when reviewing the literature, we used this definition of benign external hydrocephalus: an idiopathic condition in infants characterized by a large or rapidly increasing head circumference and radiologically confirmed enlarged frontal subarachnoid spaces.

In the beginning of the era of computed tomography (CT), the differentiation between subdural and subarachnoid fluid collections was difficult, not to say impossible. This fact may of course confound our review, which is why some of the earliest articles have been excluded where there is doubt about the origin of the radiologically detected fluid.

Review of the literature

The following review is based on a systematic search in the PubMed and Web of Science databases. The terms used in the search were “hydrocephalus” combined with any of the following words: external, benign, extraventricular, extracerebral, or idiopathic. Other search terms were: “idiopathic/familial megalencephaly,” “idiopathic/familial macrocephaly,” “subdural effusion,” “benign subdural collections,” “subdural hygroma,” “extraventricular obstructive hydrocephalus,” “subdural/extracerebral/extraaxial/subarachnoid/pericerebral fluid collections,” and “benign communicating hydrocephalus.”

The review includes all original articles written in English or in other languages with an informative English abstract that report cases or larger groups of children with benign external hydrocephalus as defined above. Cases with a known cause of hydrocephalus or with accompanying conditions possibly affecting a long-term neurodevelopmental outcome, such as prematurity, are excluded.

Results

A total of 1,871 articles were identified by the search (March 3, 2010). Of these, only 147 studies and case reports dealt with this condition and were therefore included. These articles are discussed separately under “What is benign external hydrocephalus?”, “Neuroimaging”, etc. in the following subsections. Hence, 1,724 articles were excluded as they dealt with non-idiopathic conditions or with adult patients or only mentioned the search words but did not contribute any new information.

What is benign external hydrocephalus?

Definition

Before the CT era, the condition was hardly seen. However, Dandy defined external hydrocephalus as increased ICP combined with dilated subarachnoid spaces in infants, but he questioned whether it existed as a primary condition or instead was a subtype of internal hydrocephalus [44, 46].

Today, external hydrocephalus is commonly defined as a large or rapidly growing head circumference in infants combined with enlarged subarachnoid spaces and no or only moderate ventricular enlargement as seen on neuroimaging (see below) [4, 70, 132, 143, 147]. Kumar added the absence of “clinico-radiological features of raised intracranial pressure,” e.g., ventriculomegaly without periventricular lucency, and non-tense fontanels as criteria [91].

Epidemiology

No studies seem to report the incidence or prevalence of external hydrocephalus in the normal population nor did we find figures for the relative amount of hydrocephalic children diagnosed with this subtype of hydrocephalus. It
seems that most studies are too small and selective to yield information about the incidence or prevalence of external hydrocephalus.

While idiopathic external hydrocephalus seems to be the most common cause of macrocephaly in infants [4, 74], many patients have a history of prematurity [2, 4]. A review of incidental findings in a tertiary pediatric neurology center showed that 0.6% of the children were found to have external hydrocephalus [73].

It seems that about two thirds of children with external hydrocephalus are boys [3, 33, 34, 78, 121–123, 126, 130, 132, 142, 147, 149], which is about the same gender distribution as in hydrocephalus [106, 170].

Etiology

In most reported cases, there is no obvious cause of the external hydrocephalus, and it is therefore classified as idiopathic. However, it has been reported after numerous situations and conditions such as prematurity and intraventricular hemorrhage [78, 87, 101, 115, 160], meningitis [77, 87], metabolic disorders [17], steroid therapy [66], chemotherapy [54], neurosurgery [80], and trauma [77, 87].

A complicating fact is that intraventricular and subarachnoid hemorrhages in premature infants often occur without symptoms [29], thus making it difficult to know if idiopathic hydrocephalus really is idiopathic or simply caused by such silent, clinical events [68].

External and communicating hydrocephalus is described in children with raised venous pressure [87], e.g., following various thoracic/cardiac conditions [49, 86, 112, 145].

Heredity

Some patients with external hydrocephalus seem to have a familial form as one or more close relatives are macrocephalic.

Most studies report that around 40% of children with external hydrocephalus have at least one close relative with a large head (usually defined as a head circumference above the 95th to 98th percentile) [3, 6, 34, 122, 124, 128, 167]; however, this coherence was found to be as high as 80–90% in two reports [4, 152].

Case reports of twins and triplets also suggest some heredity [32, 42, 85, 165].

An autosomal dominant mode of transmission has been assumed [4, 11, 39, 47], although a multifactorial model of inheritance is the most recent proposal [9]. The dominant inheritance might be due to a single gene exhibiting a major effect as part of a multifactorial phenomenon in some families [166], probably during a limited time of susceptibility in fetal development [134]. Maytal et al. suggested that the primary phenotype merely was the delayed maturation of the arachnoid villi [110].

External hydrocephalus in infants seems to be closely linked to “familial macrocephaly/megalencephaly” in the literature. This term is commonly defined as children born with head circumferences in the upper normal range, which increase beyond the 98th percentile during the first year of life [102]. A number of underlying causes were described [11].

Pathophysiology

There are probably mechanisms common for both ordinary hydrocephalus and external hydrocephalus, but here we will focus on the latter. As most reported cases of external hydrocephalus seem to be idiopathic, various theories regarding the underlying pathophysiology have been presented.

The most common theory suggests that external hydrocephalus is caused by immature arachnoid villi not able to absorb the CSF that is produced continuously [14]. The accumulated CSF then expands the ventricles and the subarachnoid space inside the compliant and growing skull of an infant, thus avoiding a marked increase in intracranial pressure [87]. The arachnoid villi mature at about 18 months of age, ending the CSF accumulation and thus the widening of the subarachnoid space. Why the arachnoid villi do not mature remains unknown, but some heredity has been described (see above).

Other theories have been suggested, such as an arachnoid membrane tear creating a one-way valve [45], CSF becoming “loculated” [157], or subdural fluid obstructing CSF reabsorption [142].

Some believe that external hydrocephalus is only a step towards internal hydrocephalus in children with communicating hydrocephalus, i.e., if the arachnoid villi cannot absorb the CSF, it will first accumulate in the nearby subarachnoid space, thereafter gradually involving the ventricular system [123, 143]. The term arrested hydrocephalus is associated with this theory; some suggest that there may be a difference between the delayed maturation of arachnoid villi leading to a regression of the pericerebral dilatation and agenesis of the villi corresponding to cases requiring a shunt [36, 64].

Some have even suggested that the skull is growing faster than the brain for some time, giving a transient subarachnoid CSF accumulation [124, 134].

The understandings of CSF dynamics all in all seem incomplete and up to this time a subject of debate [38, 71].

Cerebrospinal fluid outflow

External hydrocephalus is commonly classified as a communicating hydrocephalus [14, 132]. A recent review
summarizes the current knowledge on the physiology of CSF outflow [84]. In brief, three pathways are recognized: the arachnoid granulations, the lymphatic capillaries, and the transependymal passage.

The arachnoid granulations (or villi) become visible between 6 and 18 months of age, developing gradually in terms of size and number over the next years [96, 162]. The lymphatic pathway is thought to be CSF flowing in the subarachnoid space enclosed by perineural sheaths of cranial and spinal nerves, escaping into lymphatic capillaries mainly in the nasopharyngeal area [84]. Animal studies have shown that 10–50% of CSF is drained by lymphatics [23, 24]. The transependymal passage of CSF probably occurs only when the intracranial pressure exceeds a limit [61, 97].

External hydrocephalus in the fetus

Neuroimaging of fetuses gives additional information about the development of the subarachnoid spaces and external hydrocephalus.

It has been found that human fetuses who were diagnosed with external hydrocephalus as infants had prominent subarachnoid spaces with a posterior fluid distribution prenatally [63]. This is thought to reflect the development of the subarachnoid space, which is seen as a cavitation of the primitive meninges spreading from the ventral to the dorsal portion of the neural tube.

The same authors reported that 19% of human fetuses that had mild ventriculomegaly and prominent subarachnoid spaces developed an external hydrocephalus after birth [62].

External hydrocephalus as a risk factor

Several studies have shown an increased risk of subdural hematomas in children with external hydrocephalus after minimal or no known head trauma [12, 67, 78, 83, 95, 113, 117, 129, 136, 141].

Association with other conditions

External hydrocephalus may coexist with a series of conditions, such as some types of craniosynostoses [35, 125, 151], achondroplasia [55, 127], Sotos syndrome [100, 110, 127], and glutaric aciduria type I [107, 108, 127, 133]. A case of external hydrocephalus in a microcephalic infant has also been reported [1]. The hydrocephalus in craniosynostosis and achondroplasia is supposedly caused by a rigid venous outflow obstruction [148].

Clinical symptoms and signs

The large or enlarging head appear indistinguishable from those seen in other hydrocephalus cases [3, 4, 14, 32, 33, 68, 78, 115, 118, 123, 132, 134, 141, 143, 160, 167]. A relatively common sign is a tense anterior fontanel [6, 14, 75, 123, 124, 143, 144, 147, 165]. Other early symptoms and signs have also been reported occasionally: dilated scalp veins [65, 143, 147], frontal bossing (an unusually prominent forehead) [95], irritability [33, 53, 91, 124, 144, 165], hypotonia [12, 36, 42, 65, 75, 130, 139, 152], vomiting [78, 91, 124, 144], gross motor delay [12, 34, 42, 53, 65, 75, 115, 118, 121, 123, 124, 128, 139, 147, 152, 167], ataxia [91, 144, 147], poor head control [91, 121, 122], seizures [3, 68, 75, 78, 124, 144, 147], fever [75, 78], and mental retardation [87]. We have not found any articles reporting sunset gaze.

Head circumference

Infants with external hydrocephalus usually show a rapid increase in head circumference (Fig. 1) [4, 12, 14, 132, 143], which appears to be the most common symptom in all children developing hydrocephalus during their first year of life [170]. Most of the increase in head circumference occurs around the age of 6 months [4, 130, 147]. It seems that the head circumference usually stabilizes before the age of 18 months [2, 33]. Measurements afterwards typically lie above but parallel to the upper (95th to 98th) percentile [2, 6, 14, 22, 115, 121, 147]. The amount of children ending up with macrocephaly varies considerably from 11% to 87% on long-term follow-up [3, 34, 60, 118].

The natural history of untreated external hydrocephalus

Short-term outcome—transient delay of development

A developmental delay is commonly seen during some time of infancy [4, 22, 95, 118, 122, 123].

Muenchberger et al. reported that out of their 15 patients, who were followed until adulthood, two had transient motor delay and two had speech delay at a mean 27 months of age [118]. Alvarez et al. found that about half of the 32 children were delayed in motor or language development at 5 months of age, but by 15–18 months of age all but one were found to be normal [4]. Nickel and Gallenstein reported seven out of nine infants with delayed gross motor development during the first year of life, with four of them described as normal after 2–3 years of age [122].

Similar results are found in several surveys, reporting delayed gross motor development and to a lesser extent delayed language development that decrease and disappear within 1–4 years [2, 13, 22, 91, 95, 123, 128, 152].

In two studies, hypotonia was reported during the first year of life, but with normal findings on later examinations [132, 160].
Lorch et al. performed a survey on macrocephalic survivors of neonatal intensive care [103]. The children with benign extraaxial fluid were compared with those without; it was found that the presence of extraaxial fluid was associated with an increased risk of developmental delay and cerebral palsy (followed up to 24 months of age).

**Long-term outcome**

We have found a total of 37 articles that report outcome after some time [3, 4, 6, 13, 14, 22, 32, 33, 40, 42, 60, 74, 75, 78, 85, 87, 91, 95, 115, 116, 118, 121–124, 128, 132, 142–144, 147, 152, 153, 156, 159, 165, 167]. Scientific strength however is low; only one article [95] was considered as level 3 (Levels of Evidence, http://www.cebm.net/). The studies report in general normal physical and neurological findings. Some conclude that all children are normal on last follow-up [6, 14, 22, 32, 33, 40, 42, 74, 85, 91, 115, 142, 144, 145, 152, 156, 159, 165, 167], while the remaining articles describe developmental delay among some of their patients [3, 4, 13, 60, 75, 78, 87, 95, 116, 118, 121–124, 128, 132, 143, 147, 153, 167].

The bulk of long-term affected children show failure to reach developmental milestones [87, 116, 124, 143, 147, 167], especially in gross motor function [13, 75, 78, 121, 122, 128, 132]. Speech or language delays are also quite common [3, 13, 78, 122], while mental retardation seems relatively rare [95]. The symptoms related to increased intracranial pressure, which often can be seen initially, all appear to be absent at follow-up.

Only two studies have followed the children up to school age: Muenchberger at al. did a long-term follow-up of nine patients with external hydrocephalus (plus six who would not do the psychological tests) [118]. At final follow-up (mean 19 years), all nine were considered as neurologically normal and the neuropsychological assessment showed an intellectual ability within the normal range. Nevertheless, reduced performance was noted in several of the patients on two tests associated with attention, and the two patients who had speech delay at the age of 2 years performed at below-average levels in most psychological tests at long-term follow-up. Furthermore, as many as ten of the 15 patients reported specific learning problems in reading and mathematics or had been diagnosed with a psychiatric disease. Eight of the children had to repeat grades or go to special classes. One of these eight children also had been diagnosed with a psychiatric disease and so had another two children without specific learning problems.

Laubscher et al. investigated 22 megalencephalic children with “dilated pericerebral subarachnoid spaces” [95]. Twelve of them were developmentally delayed (type of delay and age not specified). Eleven of 12 children who had reached school age at the time that the study ended had a normal school outcome. The children were compared with 22 children with normal pericerebral subarachnoid space, looking at psychomotor development and school outcome, with findings not significantly different between the groups.

Several studies have followed children with external hydrocephalus for 2–5 years [13, 14, 34, 60, 78, 87, 122, 123, 147, 152]. About 17% of the 196 children included in these publications were described as having an abnormal psychomotor development at last follow-up.

**Neuropsychological testing**

While most studies seem to base their evaluation of outcome on clinical and neurological examination, some use standardized neuropsychological tests as well: the Denver Developmental Screening test [4, 121, 124], the Milani Comparetti (gross motor assessment study) [121], the Denver II screening test and Peabody Picture—
Vocabulary test [3], the developmental scales of Brunet and Lezine [22], and the Revised Gesell Developmental Schedules and the Movement Assessment of Infants [122]. Below we review articles using standardized neuropsychological tests, some analyzing short-term outcome and some analyzing long-term ones.

Studying ten infants with external hydrocephalus, Neveling and Truex used the Denver Developmental Screening Test focusing on four areas: personal–social, fine motor–adaptive, language, and gross motor skills [121]. All of the areas concerned were above or equal to the Denver 50th percentile, indicating normality, except for the gross motor skills. They therefore continued with the Milani Comparetti Screening Test, which found that the infants were lacking in crawling and sitting skills and displayed an abnormal developmental pattern (e.g., walking prior to belly crawling). The authors assumed that the abnormal developmental progression was caused by the increased head size.

The Denver Developmental Screening Test was used at least once in all 36 patients reported by Alvarez et al. [4]. As mentioned above, a transient developmental delay was seen in many of the children. Fourteen were found to be delayed in gross motor development and five were found to have delayed language development, with only one remaining globally delayed at last follow-up (30 months of age).

Nogueira and Zaglul also used the Denver Developmental Screening Test [124]. They reported that 14 out of 58 children showed “abnormal development” at follow-up, without further specification.

Alper et al. found two out of 13 children with fine motor delay using the Denver II screening test [3]. Performing Peabody Picture–Vocabulary testing for seven children older than 2.5 years, they found two with expressive language delay.

Bosnjak et al. reported nine patients with external hydrocephalus, all assessed developmentally by a psychologist using the developmental scale of Brunet and Lezine [22]. Six of the nine had abnormal neurodevelopmental findings at presentation: four of these however had normalized at follow-up and the other two were not available for follow-up. Further details about development were not described.

Nickel and Gallenstein reported nine patients investigated with the Revised Gesell Developmental Schedules and the Movement Assessment of Infants [122]. Seven of them showed gross motor delay during the first year of life, while only one remained delayed at last follow-up. Three children had speech/language delay at last follow-up.

Furthermore, Muenchberger et al. utilized several neuropsychological tests suitable for adults in their thorough survey (described above) [118].

### Neuroimaging

#### Normal range of the subarachnoid space

As no consensus exists, the definition of a normal subarachnoid space width varies in the literature: in infants (below 1 year of age) the upper limits of normal craniocortical width range from 4 to 10 mm [56, 59, 94, 99, 138] and in neonates from 3.3 to 5 mm [58, 111, 120]. The defined upper limit of the normal interhemispheric fissure width ranges from 6 to 8.5 mm, while the similar spectrum for sinocortical width is 2 to 10 mm [56, 59, 69, 94, 99, 131]. Sinocortical width is defined as the distance from the lateral wall of the superior sagittal sinus to the surface of the cerebral cortex [56, 99].

Lam et al. found that the width of the normal subarachnoid spaces increased from birth up to about 7 months of age, after which a gradual decline was observed [94]. Other studies confirm this decrease in fluid volume, as the normal subarachnoid spaces are smaller between 1 and 2 years of age [59] and essentially absent after this age [89]. There seems to be no significant difference in size between the genders [58, 94, 120].

The studies concerning external hydrocephalus use different standards; hence, the limits of inclusion differ among the surveys. Less accurate, subjective grading in, e.g., normal, mild, and moderate subarachnoid space enlargement has also been used [33].

#### Neuroimaging characteristics of external hydrocephalus

The neuroimaging characteristics of external hydrocephalus are frontal subarachnoid spaces that are enlarged beyond the upper limit together with normal to moderately enlarged ventricles (Fig. 2). A concurrent finding is often a wide interhemispheric fissure and sometimes enlarged third ventricle and basal cisterns [14, 42, 83, 91, 110, 115, 121, 139, 161].

Among the surveys reporting the size of the ventricular system, from none to all patients with external hydrocephalus showed some degree of ventricular dilatation [3, 6, 14, 33, 34, 60, 91, 95, 110, 114, 118, 139, 142]. These reports, however, do not give exact measurements. Prassopoulos et al. found that the degree of dilatation of the lateral ventricles was roughly proportional to the width of the frontal subarachnoid space [139]. Maytal et al. observed that the first area that appeared to enlarge was the frontal interhemispheric fissure, followed by the subarachnoid space over the frontal and frontoparietal convexities. Enlarged basal cisterns and ventricular dilation, when it occurred, was a late finding [110].

#### Neuroimaging differentiation

External hydrocephalus must be differentiated from conditions such as subdural fluid collections and cerebral
atrophy. The latter differs from external hydrocephalus in the global widening of cerebral sulci (not only in the frontal region and interhemisferic fissure); neither is cerebral atrophy associated with an increasing head circumference [110].

Modern neuroimaging techniques are used to distinguish external hydrocephalus from a subdural fluid collection (e.g., chronic subdural hematoma) [8, 27, 167], e.g., looking for the “cortical vein sign” on magnetic resonance imaging (MRI) [93] or cranial (Doppler) ultrasound [37, 164]. The cortical vein sign is defined as the visualization of cortical veins within fluid collections at the cerebral convexities. A positive sign suggests that the fluid collection is caused by an enlarged subarachnoid space and not a subdural collection which would compress the subarachnoid space and the veins traversing it.

The immediate influx of a contrast medium from CSF into a fluid collection suggests external hydrocephalus, whereas no influx indicates a subdural effusion [135]. Ment et al. observed that the enlargement of the basal cisterns often were seen in external hydrocephalus but not in subdural hematomas [115]. Finally, when using MRI, differentiation can be made based on the intensity of the fluid relative to CSF [8].

Studies of CSF flow

Neuroimaging investigation of CSF flow can be done by injecting an isotope or a contrast medium intrathecally (cisternography), which has been done in several studies of external hydrocephalus. Such studies usually report signs of slow flow/stasis or no flow at all over the cerebral convexities [6, 28, 36, 83, 122, 124, 142, 147]. Ventricular reflux is also reported [121]. On the other hand, Modic et al. reported three patients whose radionuclide cisternograms were all normal [116].

Neuroimaging outcome

The frontal subarachnoid enlargement in external hydrocephalus seems to decrease and disappear spontaneously within 2–3 years of age in most patients [34, 75, 91, 105, 110, 118, 123, 125, 128, 130, 144, 152, 167]. However, three surveys found that most of their patients had essentially static CT appearances beyond 2 years of follow-up [60, 87, 124].

The longest follow-up was described by Muenchberger et al. who found that all of the nine patients investigated (mean 19 years old) appeared normal on MRI [118]. Nishimura et al. support this finding: none of their patients had a recurrence of subarachnoid fluid once it resolved [123].

Other investigations

Fluid characteristics

Some of the studies dealing with external hydrocephalus report the composition of the subarachnoid fluid. Findings vary considerably from normal CSF [4, 60, 91, 121, 124, 144, 167] to xanthochromic fluid with protein concentrations up to 12 g/L [28, 36]. However, some studies report difficulty in extracting any fluid at all [14, 115, 121, 124].

In a case report describing two patients with external hydrocephalus, Chazal et al. found a considerably higher protein concentration in the CSF withdrawn from over the cerebral convexities than in ventricular and lumbar CSF [36]. The authors suggest that this difference is related to a “stagnation” of CSF over the convexities.

Intracranial pressure measurements

There is no consensus as to what is a normal ICP in young children, but values of more than 15 mmHg are usually considered raised [51, 57]. Few studies have reported ICP measurements in children with external hydrocephalus, and we found only three studies with a total of 11 patients reporting exact pressures [36, 147, 165]. They show normal to slightly increased ICP, ranging from 6 to 16 mmHg.

Lumbar or ventricular infusion tests are sometimes used in the evaluation of hydrocephalic children [31, 48]. Resistance to CSF outflow ($R_{\text{out}}$) is calculated and believed to express the CSF absorption capacity. However, investigations in children with hydrocephalus have not been able to find a correlation neither between $R_{\text{out}}$ and the
continuously monitored ICP [50] nor between $R_{out}$ and the need for shunting [119].

ICP wave investigations have shown that mean wave amplitude may be a better predictor than mean ICP when considering shunting or not [52].

**Electroencephalography**

Seizures have been reported in several studies of children with external hydrocephalus (see above). However, only a few have reported electroencephalography findings, which often proved abnormal [36, 124, 144, 147]. The abnormality has most often been described as a non-specific slowing.

**Treatment**

Studies that compare the treatment and non-treatment of external hydrocephalus do not exist. Most children seem to be managed conservatively, which usually means observation only. The reported treatment options were shunting, other CSF diverting procedures, or medical therapy.

**Shunting**

The following studies report patients with external hydrocephalus that underwent shunting procedures [36, 78, 118, 123, 143, 161, 165, 167]. Ventriculoperitoneal or subduroperitoneal shunts seem favored. Symptoms and signs of increased ICP are the most common causes leading to shunting, while no studies reported delayed development as a treatment indication alone. In general, it is difficult to find a common indication for surgery in the studies that are included. Information about outcome is referred below when this is mentioned in the studies.

Robertson and Gomez treated two out of six patients with shunts (one lumboperitoneal and one ventriculoperitoneal) because of excessive head growth, ventricular dilatation, and other signs of increased intracranial pressure [143]. One of them was followed for 7 years and developed normally.

Hellbusch reported three out of 39 patients requiring shunt [78]. The first received a subduroperitoneal shunt because of macrocrania and the development of subdural hematoma/hygrroma. The second also had macrocrania, along with some vomiting, and underwent an insertion of a subduroperitoneal shunt. The third received a ventriculoperitoneal shunt because of enlarged ventricles together with a large head.

Chazal et al. described shunting in both of their patients [36]. One was referred with a large head, bulging anterior fontanel, and hypotonia. She received a ventriculoatrial shunt and had rapid clinical improvement. The other underwent a shunting procedure because of a large, growing head and the persistence of psychomotor retardation; the development normalized afterwards.

Wachi and Sato described a pair of identical twins who developed external hydrocephalus during the first few months of life [165]. Irritability and bulging of the anterior fontanel developed; they therefore underwent shunt surgery at 9 months of age with satisfying findings 6 months later.

Nishimura et al. reported three out of 20 patients who were in need of surgery because of subdural hematomas complicating the subarachnoid fluid collections [123]. Burr hole and irrigation were performed in two and one underwent subduroperitoneal shunt insertion.

Ten out of the 14 patients reported by Tsubokawa et al. had macrocephaly and bulging fontanels [161]. All ten underwent surgery with temporary subduroperitoneal shunt insertion. At 4–6 months after surgery, neuroimaging normalization was seen, although the ventricle enlargement seemed to retrace slower. Seven of the ten children operated had a developmental quotient (DQ) of more than 100 at follow-up, indicating normal development, while two of the four non-operated patients had a DQ of less than 39.

Other studies report shunting of some of the patients without further information regarding indications, outcome, etc. [118, 167].

**Other CSF diverting procedures—external drainage**

Eidlitz-Markus et al. reported a case of a 6-month-old girl with external hydrocephalus and developmental delay [53]. She was treated for 48 h with temporary bilateral drainage of the frontal subarachnoid spaces via burr holes, draining 300 ml of CSF. The head circumference and psychomotor development normalized within a few months and remained so at the last follow-up at 2 years of age. CT showed a modest reduction in the size of the CSF spaces 2 months after surgery. Similarly, Stroobandt et al. suggested treatment with external drainage for 1 week, thereafter inserting a shunt if the effusion had not “dried up” by this time [158]. Treatment of posttraumatic external hydrocephalus with temporary spinal drainage is described in adults [7].

Andersson et al. performed exploratory craniotomy in seven of their nine patients [6]. They reported widened and deep subarachnoid spaces. Three patients needed a ventriculoperitoneal shunt in order to control postoperative CSF leakage.

**Medical therapy**

Several studies describe temporary acetazolamide treatment lasting for 1–2 months, resulting in a gradual reduction of excessive head growth [14, 91, 137]. Furthermore, Roshan...
et al. used acetazolamide combined with mannitol in four patients who presented with vomiting, irritability, and a bulging fontanel [144]. The patients responded well.

Acetazolamide and furosemide have been recommended for mild hydrocephalus of the newborn and in infants [98, 154], but based on large, randomized trials it is not recommended for the treatment of posthemorrhagic ventricular dilatation in infancy [82, 88].

Discussion

What is benign external hydrocephalus?

External hydrocephalus is defined as a rapid increase in head circumference in an infant combined with enlarged frontal subarachnoid spaces as seen on CT, MRI, or cranial ultrasound and with normal or slightly enlarged ventricles.

The underlying mechanism for the formation of external hydrocephalus is poorly understood, although several theories exist. The familial macrocephaly associated with some of the cases indicates that heredity may play a role. CSF flow studies have shown reduced flow over the cerebral convexities; an impairment of CSF absorption through the arachnoid villi therefore seems intuitive. In normal children, it has been shown that the arachnoid villi are not fully mature at birth but that they gradually become so during infancy. This lack of maturation in combination with the pronounced increase in CSF production during the first year of life [169] may be the underlying mechanism and may also explain why the head starts to grow at around 6 months of age in most cases. This may not be a problem in most children, as their draining capacity through the villi or other draining pathways is balanced against the CSF production. In children with external hydrocephalus, on the other hand, there may be a misbalance because of either delayed maturation or excessive CSF production.

The pronounced increase in CSF production during the first year of life may also explain why external hydrocephalus rarely is described in newborns. The finding that CSF production in boys is greater than in girls may also partly explain the unequal gender distribution.

The delayed maturation theory does not contradict the belief that external hydrocephalus may be an arrested form of internal communicating hydrocephalus. The finding by Maytal et al. about the order in which the CSF-containing compartments dilate supports this view [110]. Mechanisms believed to cause ordinary hydrocephalus may therefore play a role in the formation of external hydrocephalus, e.g., altered venous sinus pressures [15] or restriction of arterial pulsation [71].

In sum, the etiology of external hydrocephalus is most likely multifactorial and, if so, the condition may develop in several ways.

Clinical presentation

By our definition, increased head circumference is found in all patients with external hydrocephalus. In most cases, the head circumference increases disproportionally only during the first year of life, an observation that may support the delayed maturation theory as discussed above. However, as the cranial sutures close between 1 and 2 years of age, it is difficult to exclude a persistently increased ICP. Many children end up with large heads, i.e., they do not normalize, signifying a continued growth stimulus beyond infant age.

Many patients are found to have a delay in gross motor development, although only a few surveys have tested the children using valid neuropsychological test batteries. Reports of children with hypotonia, seizures, vomiting, etc. also indicate that the brain may be under marked strain during one phase of the condition.

The natural history of untreated external hydrocephalus

It seems evident that external hydrocephalus in some cases is associated with delayed psychomotor development. The important questions are whether this delayed development is caused by an increased ICP and whether this increased pressure can interfere with the individual’s acquisition of motor, cognitive, emotional, and social skills in the critical phases of the brain’s development, thus hampering the future motor and mental functions of the affected child.

The transient delay of development seen up to 4 years of age supports the idea that the lack of increase in head circumference seen in the older children merely is caused by the closing of sutures rather than the actual reduction of a slightly increased ICP.

The majority of patients are described as physically, neurologically, and developmentally normal on follow-up. However, this may only be because the outcome has been evaluated by the relatively coarse methods used in the majority of the studies. Most studies did not use valid developmental tests; subtle psychomotor impairments may therefore have passed as normal. This assumption is supported by the fact that a considerable amount of patients show some forms of developmental delay, including the two studies where children were followed up to school age [95, 118]. Unfortunately, no studies were designed to show if the patients who ended up with a developmental delay could have been revealed at an earlier stage.

The studies show remarkably varying results for long-term outcome. This makes it hard to conclude and may reflect the heterogeneity that probably exists. Taking the
presenting symptoms and additional findings into consider-
aton, together with the motor delay seen in some patients
for some time, the statement that this is a benign condition
seems questionable.

As discussed above: could the temporary and mild
“insult” at a critical age lead to a permanent damage? An-
imal studies have shown that the development of the
young brain occurs step-wise, i.e., specific functions
develop within a limited time span, a “critical period”
when the brain is ready to learn that developmental task
[10, 18, 81]. Deprivation of stimuli during this critical time
may cause deficits, although the process is not entirely
irreversible [72]. It is reasonable to assume, however, that
the learning after the closure of this “time window” is much
difficult than when the neural network of the
developing brain is still susceptible to new impulses.

Theoretically, the pressure exerted on the brain tissue by
the excess CSF in the subarachnoid space during infancy
may be high enough to provide imperfect conditions during
a critical time of development, thereby giving rise to
permanent, irreversible learning difficulties and other
problems. The strict sequence of regional perfusion as
discussed under “Neuroimaging” could perhaps be seen as
the vascular basis for these critical periods.

Hanlo et al. showed in a study of hydrocephalic
infants that raised ICP is related to developmental
outcome through the process of myelination as seen
on MRI [76]. Moreover, most children with severely
delayed preoperative myelination showed at least a partial
recovery following CSF diversion. The importance of
myelination is supported by an animal study finding that
white matter blood flow seems vulnerable in hydrocephalic
kittens [43].

Neuroimaging

It is difficult to define the limit between a normal and an
enlarged subarachnoid space as the definitions used vary as
does the subarachnoid space with age. However, a
cranio cortical width above 10 mm appears to be an absolute
sign of pathology. The degree of ventricular dilatation is
usually described as “minimal” or “moderate” without more
specific measures: this probably explains the variation in
incidence of patients with this finding.

Neuroimaging differentiation between external hydro-
cephalus and subdural hygroma/effusion certainly became
easier after the introduction of MRI, and the tools presented
are useful. With the addition of CT cisternography, and
ultrasound in the youngest, a correct diagnosis should be
achieved in most patients. Subdural effusion could be
declared as a collection of protein-rich fluid of greater
density than the CSF [79], hence making the differentiation
easier both radiologically and biochemically.

Cortical hypoperfusion is seen in some infants and
should be further investigated. Studies of adult normal
pressure hydrocephalus (NPH) patients have found the
hypoperfusion to be more dominant in the frontal areas and
that it seems to improve after shunt surgery [109, 150]. A
survey in normal children showed that the distribution of
regional cortical blood flow followed a strict sequence in
time, matching the behavioral evolution occurring during
infancy [146]. Frontal activity, for instance, remained
scarcely recognizable until the second month, after which
it rose to present an adult-like pattern at the beginning of
the second year. Furthermore, observations using positron
emission tomography scan indicate that metabolic deterio-
ration occurs in the cortex surrounding the lateral ventricles
in infants with hydrocephalus [155]. Such features and
techniques may dominate the future neuroimaging analysis
of this and related conditions.

Surgical treatment and clinical outcome

Since external hydrocephalus can be anatomically con-
sidered a communicating hydrocephalus, insertion of a
ventriculoperitoneal shunt should be the appropriate
surgical method [16]. Shunting in itself carries some risk
[19, 38, 104], and whether this will equalize the possible
benefits of treating external hydrocephalus remains uncertain.

To our knowledge, no systematic studies that compare
the effect of surgical treatment and conservative manage-
ment in external hydrocephalus have been performed.
Furthermore, only a few report on the effect of surgical
treatment with information about the long-term effects. It
seems as if the cases described in the literature were treated
because of the presence of obvious signs of increased ICP,
not because of fear of the potentially long-term negative
effects on psychomotor development. The prevailing view
emerging from the existing literature seems to be that
external hydrocephalus in its most common form is a
benign, self-limiting condition that should be handled
conservatively [4, 6, 91, 122]. By “most common” we
mean macrocephalic children without other symptoms and
with the typical neuroimaging features. Given the results
discussed under “The natural history of untreated external
hydrocephalus”, we question this belief.

In cases where external hydrocephalus is combined with
subdural fluid collection, treatment alternatives such as
subduroperitoneal shunting, needle aspiration, or burr hole
evacuation should be considered.

Only a few studies have reported the outcome after the
surgical treatment of external hydrocephalus. As presented
under “Results”, they mainly reported good outcomes of
shunting. However, the value is limited as the studies are
not easy to compare and the cases are highly selected.
Some studies report medical therapy as an effective
treatment, but only short-term improvement on symptoms of increased ICP is published.

A detailed analysis of ICP pressure waves seems to yield useful additional information regarding which patients should be treated or not [52].

Associated conditions

The risk of developing subdural hematoma after minimal or no head injury is reportedly increased in children with external hydrocephalus. The proposed cause is stretching of the bridging veins traversing the enlarged subarachnoid space [5, 83, 141].

A relatively new, most interesting theory is whether there might be a connection between external hydrocephalus in childhood and the development of idiopathic NPH in the elderly. Bradley et al. found that patients with NPH have significantly larger intracranial volumes than control subjects as studied on MRI [26]. The authors suggested that these patients may have had external hydrocephalus as children and that they had remained asymptomatic until their later years, when a proposed deep white matter ischemia would occur and yield symptoms [25]. Wilson and Williams had the same finding as Bradley et al. and reported that about 20% of NPH patients had head circumferences above the 90th percentile, suggesting that external hydrocephalus may be responsible for some, but not all, patients with NPH [168]. A link between external hydrocephalus and NPH may be the recently described syndrome of hydrocephalus in young and middle-aged adults that appear asymptomatic or with a series of only slight and subtle symptoms which improve after shunt surgery [41].

Such a possible connection between external hydrocephalus and NPH gives new perspectives to the question of early surgical treatment in these children.

Benign external hydrocephalus—what to do?

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.” We think that 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.).

Conclusion

In this literature survey, we have found a relatively large number of untreated external hydrocephalus patients with temporary or permanent affection of mental functions. We therefore question the validity of the traditional view that this is a benign condition that does not need treatment. The level of evidence in most of the studies that are included in this survey is very low; there is in fact no evidence at level 2 or above when it comes to treatment. No studies that can rule out the possibility of a long-term negative effect of an increased ICP on psychomotor development were found; on the contrary, several studies indicate that external hydrocephalus may be harmful, at least in some children. Future research should focus on this, comparing the outcome of surgical treatment and conservative management of external hydrocephalus.

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Comments

Hartmut Collmann, Würzburg, Germany

This is a diligently compiled review on a fairly common yet still obscure condition of infancy known under a variety of terms such as “benign macrocephaly,” “benign subarachnoid enlargement,” or “external hydrocephalus”. It is characterized by transient acceleration of head growth, some signs of mild intracranial hypertension, and, morphologically, distinct enlargement of the subarachnoid space, often combined with mild ventricular dilation. In their comprehensive review of the available literature, the authors address all major aspects of this condition, i.e., considerations concerning etiology, pathogenesis, clinical and radiological diagnosis, and prognosis. As to the pathogenic factors, a disproportion between a rapidly increasing CSF secretion rate and delayed maturation of the arachnoid villi is the most commonly held theory. Little attention has been paid as yet to the venous system, and one is wondering if there is any relationship to the pseudotumor cerebri. While the authors underline the transient nature of the abnormal head growth and CSF accumulation, they challenge its completely benign character as a substantial proportion of patients appear to exhibit persistently retarded psychomotor skills. Consequently, they suggest a larger population-based study comparing the outcome of treated and untreated children.

Dieter Hellwig, Hannover, Germany

Benign or “idiopathic” external hydrocephalus is a rare entity in childhood and mostly resolves in the first 2 years after birth. It is characterized by an increased head circumference and neuroimaging shows a subarachnoidal fluid collection over the frontal hemispheres. In most cases, this pathology is asymptomatic and resolves without treatment; however, it is not clear if in some cases it can cause delay in mental, motor, and speech development.

In their review, Zahl et al. included a total of 147 studies. They described several theories about the etiology of benign external hydrocephalus, which seems to be still unclear. They emphasize that the main clinical sign is the rapid increase of head circumference and a tense anterior fontanelle. The final diagnosis is established by CCT or MRI.

The crucial question is if there is a need for treatment either with drugs like acetazolamide or by surgery using shunting procedures.

In accordance with the authors, who stress that there are no controlled studies about the long-term outcome of children with benign external hydrocephalus, I would like to recommend treating this form of communicating hydrocephalus by the insertion of a CSF shunt to prevent psycho-motor defects. In conclusion, there is still a lack in understanding the pathophysiology of this kind of hydrocephalus and controlled trials to evaluate the short- and long-term outcomes are urgently needed.