Neurosurg Rev (2011) 34:417–432
DOI 10.1007/s10143-011-0327-4

REVIEW

Benign external hydrocephalus: a review, with emphasis on management

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

Received: 4 October 2010 / Revised: 18 April 2011 / Accepted: 1 May 2011 / Published online: 7 June 2011

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

S. M. Zahl · K. Wester
Department of Neurosurgery, Haukeland University Hospital, Bergen, Norway

S. M. Zahl (✉) · K. Wester
Section for Neurosurgery, Department of Surgical Sciences, University of Bergen,
5021 Bergen, Norway
e-mail: sverre.zahl@gmail.com

A. Egge
Department of Neurosurgery,
Oslo University Hospital—Rikshospitalet,
Oslo, Norway

E. Helseth
Department of Neurosurgery, Oslo University Hospital—Ullevål,
Oslo, Norway

E. Helseth
Faculty of Medicine, University of Oslo,
Oslo, Norway

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 "clinicoradiological 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 1 [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].

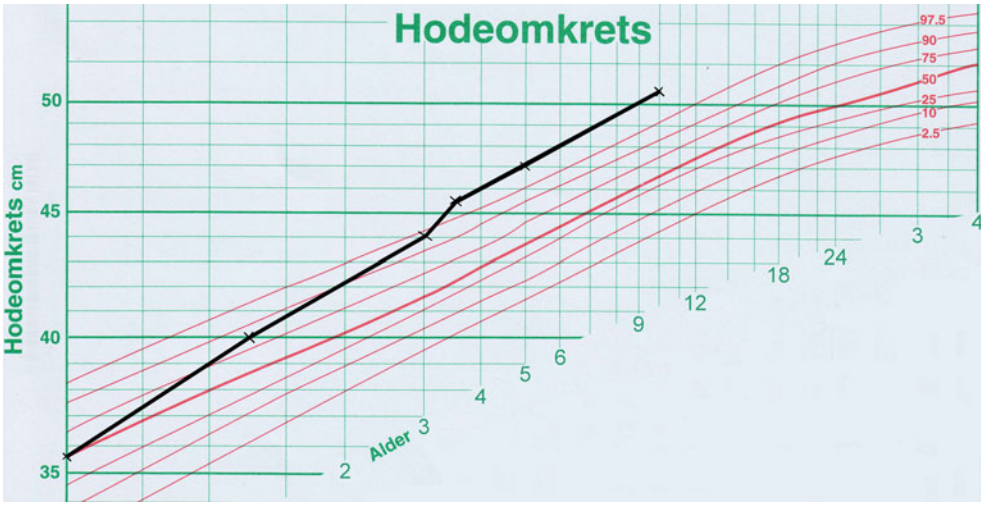

Fig. 1 This Norwegian head circumference registration sheet [90] shows the head growth of an infant boy admitted because of increasing head circumference. *Hodeomkrets*=head circumference, which is shown (in cm) along the *Y* axis. The *X* axis shows the age in months. The head circumference gradually increased after birth, with a rapid growth around the age of 3 months. Thereafter, it stabilized above, but parallel to, the upper percentile. The boy had an apparently normal psychomotor development

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, 152, 156, 159, 165], 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 interhemisferic 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

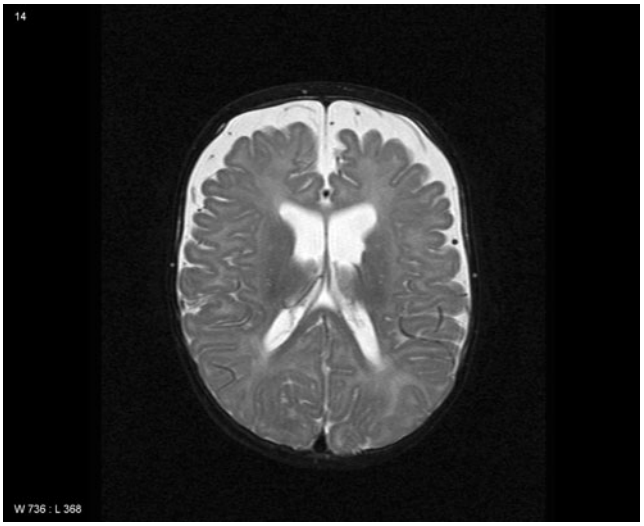

Fig. 2 MRI of the boy in Fig. 1 at 6.5 months of age The frontal subarachnoid space is enlarged, and there is slight ventriculomegaly. Vessels can be seen traversing the subarachnoid space

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 at 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_{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_{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/hygroma. 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 retract 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 to 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 consideration, 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? Animal 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 more 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 craniocortical 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 hydrocephalus 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 defined 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 deterioration 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 considered 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 management 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.

Acknowledgement Sverre Morten Zahl was supported by a Ph.D. grant from the Western Norway Regional Health Authority.

References

1. Akaboshi I, Ikeda T, Yoshioka S (1996) Benign external hydrocephalus in a boy with autosomal dominant microcephaly. Clin Genet 49:160–162
2. al-Saedi SA, Lemke RP, Debooy VD, Casiro O (1996) Subarachnoid fluid collections: a cause of macrocrania in preterm infants. J Pediatr 128:234–236
3. Alper G, Ekinci G, Yilmaz Y, Arikan C, Telyar G, Erzen C (1999) Magnetic resonance imaging characteristics of benign macrocephaly in children. J Child Neurol 14:678–682
4. Alvarez LA, Maytal J, Shinnar S (1986) Idiopathic external hydrocephalus: natural history and relationship to benign familial macrocephaly. Pediatrics 77:901–907
5. Amodio J, Spektor V, Pramanik B, Rivera R, Pinkney L, Fefferman N (2005) Spontaneous development of bilateral subdural hematomas in an infant with benign infantile hydrocephalus: color Doppler assessment of vessels traversing extra-axial spaces. Pediatr Radiol 35:1113–1117
6. Andersson H, Elfverson J, Svendsen P (1984) External hydrocephalus in infants. Childs Brain 11:398–402
7. Ando S, Otani M, Moritake K (1997) Usefulness of spinal drainage for post-traumatic external hydrocephalus: report of two cases. J Clin Neurosci 4:236–240

8. Aoki N (1994) Extracerebral fluid collections in infancy: role of magnetic resonance imaging in differentiation between subdural effusion and subarachnoid space enlargement. J Neurosurg 81:20–23
9. Arbour L, Watters GV, Hall JG, Fraser FC (1996) Multifactorial inheritance of non-syndromic macrocephaly. Clin Genet 50:57–62
10. Arling GL, Harlow HF (1967) Effects of social deprivation on maternal behavior of rhesus monkeys. J Comp Physiol Psychol 64:371–377
11. Asch AJ, Myers GJ (1976) Benign familial macrocephaly: report of a family and review of the literature. Pediatrics 57:535–539
12. Azais M, Echenne B (1992) Idiopathic pericerebral swelling (external hydrocephalus) of infants. Ann Pediatr (Paris) 39:550–558
13. Babcock DS, Han BK, Dine MS (1988) Sonographic findings in infants with macrocrania. AJR Am J Roentgenol 150:1359–1365
14. Barlow CF (1984) CSF dynamics in hydrocephalus—with special attention to external hydrocephalus. Brain Dev 6:119–127
15. Bateman GA, Smith RL, Siddique SH (2007) Idiopathic hydrocephalus in children and idiopathic intracranial hypertension in adults: two manifestations of the same pathophysiological process? J Neurosurg 107:439–444
16. Beni-Adani L, Biani N, Ben-Sirah L, Constantini S (2006) The occurrence of obstructive vs absorptive hydrocephalus in newborns and infants: relevance to treatment choices. Childs Nerv Syst 22:1543–1563
17. Bhasker B, Raghupathy P, Nair TM, Ahmed SR, deSilva V, Bhuyan BC, Al Khusaiby SM (1999) External hydrocephalus in primary hypomagnesaemia: a new finding. Arch Dis Child 81:505–507
18. Blakemore C, Van Sluyters RC (1974) Reversal of the physiological effects of monocular deprivation in kittens: further evidence for a sensitive period. J Physiol 237:195–216
19. Blount JP, Campbell JA, Haines SJ (1993) Complications in ventricular cerebrospinal fluid shunting. Neurosurg Clin N Am 4:633–656
20. Boaz JC, Edwards-Brown MK (1999) Hydrocephalus in children: neurosurgical and neuroimaging concerns. Neuroimaging Clin N Am 9:73–91
21. Bodensteiner JB (2000) Benign macrocephaly: a common cause of big heads in the first year. J Child Neurol 15:630–631
22. Bosnjak V, Besenski N, Marusic-Della MB, Kogler A (1989) Cranial ultrasonography in the evaluation of macrocrania in infancy. Dev Med Child Neurol 31:66–75
23. Boulton M, Armstrong D, Flessner M, Hay J, Szalai JP, Johnston M (1998) Raised intracranial pressure increases CSF drainage through arachnoid villi and extracranial lymphatics. Am J Physiol 275:R889–R896
24. Bradbury MW, Cole DF (1980) The role of the lymphatic system in drainage of cerebrospinal fluid and aqueous humour. J Physiol 299:353–365
25. Bradley WG Jr, Bahl G, Alksne JF (2006) Idiopathic normal pressure hydrocephalus may be a "two hit" disease: benign external hydrocephalus in infancy followed by deep white matter ischemia in late adulthood. J Magn Reson Imaging 24:747–755
26. Bradley WG, Safar FG, Furtado C, Ord J, Alksne JF (2004) Increased intracranial volume: a clue to the etiology of idiopathic normal-pressure hydrocephalus? AJNR Am J Neuroradiol 25:1479–1484
27. Brant-Zawadzki M, Kelly W, Kjos B, Newton TH, Norman D, Dillon W, Sobel D (1985) Magnetic resonance imaging and characterization of normal and abnormal intracranial cerebrospinal fluid (CSF) spaces. Neuroradiology 27:3–8
28. Briner S, Bodensteiner J (1981) Benign subdural collections of infancy. Pediatrics 67:802–804
29. Burstein J, Papile LA, Burstein R (1979) Intraventricular hemorrhage and hydrocephalus in premature newborns: a prospective study with CT. AJR Am J Roentgenol 132:631–635
30. Caldarelli M, Di RC, Romani R (2002) Surgical treatment of chronic subdural hygromas in infants and children. Acta Neurochir (Wien) 144:581–588
31. Caldarelli M, Di RC, Rossi GF (1979) Lumbar subarachnoid infusion test in paediatric neurosurgery. Dev Med Child Neurol 21:71–82
32. Camerota AJ, Rash FC (1994) External hydrocephalus in a set of triplets. Clin Pediatr (Phila) 33:255–256
33. Carolan PL, McLaurin RL, Towbin RB, Towbin JA, Egelhoff JC (1985) Benign extra-axial collections of infancy. Pediatr Neurosci 12:140–144
34. Castro-Gago M, Perez-Gomez C, Novo-Rodriguez MI, Blanco-Barca O, onso-Martin A, Eiris-Punal J (2005) Benign idiopathic external hydrocephalus (benign subdural collection) in 39 children: natural history and relationship to familial macrocephaly. Rev Neurol 40:513–517
35. Chadduck WM, Chadduck JB, Boop FA (1992) The subarachnoid spaces in craniosynostosis. Neurosurgery 30:867–871
36. Chazal J, Tanguy A, Irthum B, Janny P, Vanneuville G (1985) Dilatation of the subarachnoid pericerebral space and absorption of cerebrospinal fluid in the infant. Anat Clin 7:61–66
37. Chen CY, Chou TY, Zimmerman RA, Lee CC, Chen FH, Faro SH (1996) Pericerebral fluid collection: differentiation of enlarged subarachnoid spaces from subdural collections with color Doppler US. Radiology 201:389–392
38. Chumas P, Tyagi A, Livingston J (2001) Hydrocephalus—what's new? Arch Dis Child Fetal Neonatal Ed 85:F149–F154
39. Cole TR, Hughes HE (1991) Autosomal dominant macrocephaly: benign familial macrocephaly or a new syndrome? Am J Med Genet 41:115–124
40. Compen-Kong R, Landeras L (1991) The neuroevolutionary profile of the nursing infant with macrocephaly and benign enlargement of the subarachnoid space. Bol Méd Hosp Infant Méx 48:440–444
41. Cowan JA, McGirt MJ, Woodworth G, Rigamonti D, Williams MA (2005) The syndrome of hydrocephalus in young and middle-aged adults (SHYMA). Neurol Res 27:540–547
42. Cundall DB, Lamb JT, Roussounis SH (1989) Identical twins with idiopathic external hydrocephalus. Dev Med Child Neurol 31:678–681
43. da Silva MC, Michowicz S, Drake JM, Chumas PD, Tuor UI (1995) Reduced local cerebral blood flow in periventricular white matter in experimental neonatal hydrocephalus—restoration with CSF shunting. J Cereb Blood Flow Metab 15:1057–1065
44. Dandy WE (1918) Extirpation of the choroid plexus of the lateral ventricles in communicating hydrocephalus. Ann Surg 68:569–579
45. Dandy WE (1946) Treatment of an unusual subdural hydroma (external hydrocephalus). Arch Surg 52:421–428
46. Dandy WE, Blackfan KD (1914) Internal hydrocephalus: an experimental, clinical and pathological study. Am J Dis Child 8:406–482
47. Day RE, Schutt WH (1979) Normal children with large heads—benign familial megalencephaly. Arch Dis Child 54:512–517
48. Di Rocco C, Caldarelli M, Mangiola A, Milani A (1988) The lumbar subarachnoid infusion test in infants. Childs Nerv Syst 4:16–21
49. Dillon T, Berman W Jr, Yabek SM, Seigel R, Akl B, Wernly J (1986) Communicating hydrocephalus: a reversible complication

of the Mustard operation with serial hemodynamics and long-term follow-up. Ann Thorac Surg 41:146–149
50. Eide PK, Due-Tonnessen B, Helseth E, Lundar T (2001) Assessment of intracranial pressure volume relationships in childhood: the lumbar infusion test versus intracranial pressure monitoring. Childs Nerv Syst 17:382–390
51. Eide PK, Due-Tonnessen B, Helseth E, Lundar T (2002) Differences in quantitative characteristics of intracranial pressure in hydrocephalic children treated surgically or conservatively. Pediatr Neurosurg 36:304–313
52. Eide PK, Egge A, Due-Tonnessen BJ, Helseth E (2007) Is intracranial pressure waveform analysis useful in the management of pediatric neurosurgical patients? Pediatr Neurosurg 43:472–481
53. Eidlitz-Markus T, Shuper A, Constantini S (2003) Short-term subarachnoid space drainage: a potential treatment for extraventricular hydrocephalus. Childs Nerv Syst 19:367–370
54. Enzmann DR, Lane B (1978) Enlargement of subarachnoid spaces and lateral ventricles in pediatric patients undergoing chemotherapy. J Pediatr 92:535–539
55. Erdincler P, Dashti R, Kaynar MY, Canbaz B, Ciplak N, Kuday C (1997) Hydrocephalus and chronically increased intracranial pressure in achondroplasia. Childs Nerv Syst 13:345–348
56. Fessell DP, Frankel DA, Wolfson WP (2000) Sonography of extraaxial fluid in neurologically normal infants with head circumference greater than or equal to the 95th percentile for age. J Ultrasound Med 19:443–447
57. Fouyas IP, Casey AT, Thompson D, Harkness WF, Hayward RD (1996) Use of intracranial pressure monitoring in the management of childhood hydrocephalus and shunt-related problems. Neurosurgery 38:726–731
58. Frankel DA, Fessell DP, Wolfson WP (1998) High resolution sonographic determination of the normal dimensions of the intracranial extraaxial compartment in the newborn infant. J Ultrasound Med 17:411–415
59. Fukuyama Y, Miyao M, Ishizu T, Maruyama H (1979) Developmental changes in normal cranial measurements by computed tomography. Dev Med Child Neurol 21:425–432
60. Gherpelli JL, Scaramuzzi V, Manreza ML, Diament AJ (1992) Follow-up study of macrocephalic children with enlargement of the subarachnoid space. Arq Neuropsiquiatr 50:156–162
61. Gideon P, Sorensen PS, Thomsen C, Stahlberg F, Gjerris F, Henriksen O (1995) Increased brain water self-diffusion in patients with idiopathic intracranial hypertension. AJNR Am J Neuroradiol 16:381–387
62. Girard N, Gire C, Sigaudy S, Porcu G, d'Ercole C, Figarella-Branger D, Raybaud C, Confort-Gouny S (2003) MR imaging of acquired fetal brain disorders. Childs Nerv Syst 19:490–500
63. Girard NJ, Raybaud CA (2001) Ventriculomegaly and pericerebral CSF collection in the fetus: early stage of benign external hydrocephalus? Childs Nerv Syst 17:239–245
64. Gomez DG, DiBenedetto AT, Pavese AM, Firpo A, Hershan DB, Potts DG (1982) Development of arachnoid villi and granulations in man. Acta Anat (Basel) 111:247–258
65. Gooskens RH, Willemse J, Faber JA, Verdonck AF (1989) Macrocephalies—a differentiated approach. Neuropediatrics 20:164–169
66. Gordon N (1980) Apparent cerebral atrophy in patients on treatment with steroids. Dev Med Child Neurol 22:502–506
67. Gout A, Gautier I, Bellaiche M, Pinard JM, Tremon M, Rodriguez D, Foucaud P (1997) Idiopathic peri-cerebral enlargement in infants: simple anatomical variant or hemorrhagic risk factor? Arch Pediatr 4:983–987
68. Govaert P, Oostra A, Matthys D, Vanhaesebrouck P, Leroy J (1991) How idiopathic is idiopathic external hydrocephalus? Dev Med Child Neurol 33:274–276
69. Govaert P, Pauwels W, Vanhaesebrouck P, De PC, Afschrift M (1989) Ultrasound measurement of the subarachnoid space in infants. Eur J Pediatr 148:412–413
70. Greenberg MS (2006) Hydrocephalus. In: Greenberg MS (ed) Handbook of neurosurgery. Thieme, New York, pp 180–207
71. Greitz D (2004) Radiological assessment of hydrocephalus: new theories and implications for therapy. Neurosurg Rev 27:145–165
72. Grossman ML (1975) Early child development in the context of mothering experiences. Child Psychiatry Hum Dev 5:216–223
73. Gupta SN, Belay B (2008) Intracranial incidental findings on brain MR images in a pediatric neurology practice: a retrospective study. J Neurol Sci 264:34–37
74. Hamza M, Bodensteiner JB, Noorani PA, Barnes PD (1987) Benign extracerebral fluid collections: a cause of macrocrania in infancy. Pediatr Neurol 3:218–221
75. Handique SK, Das RR, Barua N, Medhi N, Saharia B (2002) External hydrocephalus in children. Indian J Radiol Imaging 12:197–200
76. Hanlo PW, Gooskens RJ, van Schooneveld M, Tulleken CA, van der Knaap MS, Faber JA, Willemse J (1997) The effect of intracranial pressure on myelination and the relationship with neurodevelopment in infantile hydrocephalus. Dev Med Child Neurol 39:286–291
77. Rekate HL (2003) Hydrocephalus in children. In: Winn HR, Youmans JR (eds) Youmans neurological Surgery. Saunders, St. Louis, pp 3387–3404
78. Hellbusch LC (2007) Benign extracerebral fluid collections in infancy: clinical presentation and long-term follow-up. J Neurosurg 107:119–125
79. Hobbs C, Childs AM, Wynne J, Livingston J, Seal A (2005) Subdural haematoma and effusion in infancy: an epidemiological study. Arch Dis Child 90:952–955
80. Hoppe-Hirsch E, Sainte RC, Renier D, Hirsch JF (1987) Pericerebral collections after shunting. Childs Nerv Syst 3:97–102
81. Hubel DH, Wiesel TN (1970) The period of susceptibility to the physiological effects of unilateral eye closure in kittens. J Physiol 206:419–436
82. International PHVD Drug Trial Group (1998) International randomised controlled trial of acetazolamide and furosemide in posthaemorrhagic ventricular dilatation in infancy. Lancet 352:433–440
83. Kapila A, Trice J, Spies WG, Siegel BA, Gado MH (1982) Enlarged cerebrospinal fluid spaces in infants with subdural hematomas. Radiology 142:669–672
84. Kapoor KG, Katz SE, Grzybowski DM, Lubow M (2008) Cerebrospinal fluid outflow: an evolving perspective. Brain Res Bull 77:327–334
85. Karantanas AH, Bakratsi E (2002) Benign enlargement of peripheral subarachnoid spaces in twins. Comput Med Imaging Graph 26:47–48
86. Karmazyn B, Dagan O, Vidne BA, Horev G, Kornreich L (2002) Neuroimaging findings in neonates and infants from superior vena cava obstruction after cardiac operation. Pediatr Radiol 32:806–810
87. Kendall B, Holland I (1981) Benign communicating hydrocephalus in children. Neuroradiology 21:93–96
88. Kennedy CR, Ayers S, Campbell MJ, Elbourne D, Hope P, Johnson A (2001) Randomized, controlled trial of acetazolamide and furosemide in posthemorrhagic ventricular dilation in infancy: follow-up at 1 year. Pediatrics 108:597–607

89. Kleinman PK, Zito JL, Davidson RI, Raptopoulos V (1983) The subarachnoid spaces in children: normal variations in size. Radiology 147:455–457
90. Knudtzon J, Waaler PE, Skjaerven R, Solberg LK, Steen J (1988) New Norwegian percentage charts for height, weight and head circumference for age groups 0–17 years. Tidsskr Nor Laegeforen 108:2125–2135
91. Kumar R (2006) External hydrocephalus in small children. Childs Nerv Syst 22:1237–1241
92. Kumar R, Singhal N, Mahapatra AK (2008) Traumatic subdural effusions in children following minor head injury. Childs Nerv Syst 24:1391–1396
93. Kuzma BB, Goodman JM (1998) Differentiating external hydrocephalus from chronic subdural hematoma. Surg Neurol 50:86–88
94. Lam WW, Ai VH, Wong V, Leong LL (2001) Ultrasonographic measurement of subarachnoid space in normal infants and children. Pediatr Neurol 25:380–384
95. Laubscher B, Deonna T, Uske A, van Melle G (1990) Primitive megalencephaly in children: natural history, medium term prognosis with special reference to external hydrocephalus. Eur J Pediatr 149:502–507
96. le Gros Clark WE (1920) On the pacchionian bodies. J Anat 55:40–48
97. Leliefeld PH, Gooskens RH, Braun KP, Ramos LM, Uiterwaal CS, Regli LP, Tulleken CA, Kappelle LJ, Hanlo PW (2009) Longitudinal diffusion-weighted imaging in infants with hydrocephalus: decrease in tissue water diffusion after cerebrospinal fluid diversion. J Neurosurg Pediatr 4:56–63
98. Libenson MH, Kaye EM, Rosman NP, Gilmore HE (1999) Acetazolamide and furosemide for posthemorrhagic hydrocephalus of the newborn. Pediatr Neurol 20:185–191
99. Libicher M, Troger J (1992) US measurement of the subarachnoid space in infants: normal values. Radiology 184:749–751
100. Lim JJ, Yoon SH (2008) The first neurosurgical analysis of 8 Korean children with Sotos syndrome. J Korean Neurosurg Soc 44:240–244
101. Lorber J, Bhat US (1974) Posthaemorrhagic hydrocephalus. Diagnosis, differential diagnosis, treatment, and long-term results. Arch Dis Child 49:751–762
102. Lorber J, Priestley BL (1981) Children with large heads: a practical approach to diagnosis in 557 children, with special reference to 109 children with megalencephaly. Dev Med Child Neurol 23:494–504
103. Lorch SA, D'Agostino JA, Zimmerman R, Bernbaum J (2004) "Benign" extra-axial fluid in survivors of neonatal intensive care. Arch Pediatr Adolesc Med 158:178–182
104. Lumenta CB, Skotarczak U (1995) Long-term follow-up in 233 patients with congenital hydrocephalus. Childs Nerv Syst 11:173–175
105. Machado MAD, Vieira NA, de Matos PE, Matos HD, Barbosa VA, Puglio N, Bacelar A (1999) External hydrocephalus—a review of 15 cases. International Journal of Neuroradiology 5:266–270
106. Macmahon B, Pugh TF, Ingalls TH (1953) Anencephalus, spina bifida, and hydrocephalus incidence related to sex, race, and season of birth, and incidence in siblings. Br J Prev Soc Med 7:211–219
107. Mandel H, Braun J, el-Peleg O, Christensen E, Berant M (1991) Glutaric aciduria type I. Brain CT features and a diagnostic pitfall. Neuroradiology 33:75–78
108. Martinez-Lage JF, Casas C, Fernandez MA, Puche A, Rodriguez CT, Poza M (1994) Macrocephaly, dystonia, and bilateral temporal arachnoid cysts: glutaric aciduria type 1. Childs Nerv Syst 10:198–203
109. Mataro M, Poca MA, Salgado-Pineda P, Castell-Conesa J, Sahuquillo J, Diez-Castro MJ, Aguade-Bruix S, Vendrell P, del Mar MM, Junque C (2003) Postsurgical cerebral perfusion changes in idiopathic normal pressure hydrocephalus: a statistical parametric mapping study of SPECT images. J Nucl Med 44:1884–1889
110. Maytal J, Alvarez LA, Elkin CM, Shinnar S (1987) External hydrocephalus: radiologic spectrum and differentiation from cerebral atrophy. AJR Am J Roentgenol 148:1223–1230
111. McArdle CB, Richardson CJ, Nicholas DA, Mirfakhraee M, Hayden CK, Amparo EG (1987) Developmental features of the neonatal brain: MR imaging. Part II. Ventricular size and extracerebral space. Radiology 162:230–234
112. McLaughlin JF, Loeser JD, Roberts TS (1997) Acquired hydrocephalus associated with superior vena cava syndrome in infants. Childs Nerv Syst 13:59–63
113. McNeely PD, Atkinson JD, Saigal G, O'Gorman AM, Farmer JP (2006) Subdural hematomas in infants with benign enlargement of the subarachnoid spaces are not pathognomonic for child abuse. AJNR Am J Neuroradiol 27:1725–1728
114. Medina LS, Frawley K, Zurakowski D, Buttros D, DeGrauw AJ, Crone KR (2001) Children with macrocrania: clinical and imaging predictors of disorders requiring surgery. AJNR Am J Neuroradiol 22:564–570
115. Ment LR, Duncan CC, Geehr R (1981) Benign enlargement of the subarachnoid spaces in the infant. J Neurosurg 54:504–508
116. Modic MT, Kaufman B, Bonstelle CT, Tomsick TA, Weinstein MA (1981) Megalocephaly and hypodense extracerebral fluid collections. Radiology 141:93–100
117. Mori K, Sakamoto T, Nishimura K, Fujiwara K (1993) Subarachnoid fluid collection in infants complicated by subdural hematoma. Childs Nerv Syst 9:282–284
118. Muenchberger H, Assaad N, Joy P, Brunsdon R, Shores EA (2006) Idiopathic macrocephaly in the infant: long-term neurological and neuropsychological outcome. Childs Nerv Syst 22:1242–1248
119. Munch TN, Bech-Azeddine R, Boegeskov L, Gjerris F, Juhler M (2007) Evaluation of the lumbar and ventricular infusion test in the diagnostic strategy of pediatric hydrocephalus and the therapeutic implications. Childs Nerv Syst 23:67–71
120. Narli N, Soyupak S, Yildizdas HY, Tutak E, Ozcan K, Sertdemir Y, Satar M (2006) Ultrasonographic measurement of subarachnoid space in normal term newborns. Eur J Radiol 58:110–112
121. Neveling EA, Truex RC Jr (1983) External obstructive hydrocephalus: a study of clinical and developmental aspects in ten children. J Neurosurg Nurs 15:255–260
122. Nickel RE, Gallenstein JS (1987) Developmental prognosis for infants with benign enlargement of the subarachnoid spaces. Dev Med Child Neurol 29:181–186
123. Nishimura K, Mori K, Sakamoto T, Fujiwara K (1996) Management of subarachnoid fluid collection in infants based on a long-term follow-up study. Acta Neurochir (Wien) 138:179–184
124. Nogueira GJ, Zaglul HF (1991) Hypodense extracerebral images on computed tomography in children. "External hydrocephalus": a misnomer? Childs Nerv Syst 7:336–341
125. Odita JC (1992) The widened frontal subarachnoid space. A CT comparative study between macrocephalic, microcephalic, and normocephalic infants and children. Childs Nerv Syst 8:36–39
126. Orrison WW, Robertson WC, Sackett JF (1978) Computerized tomography in chronic subdural hematomas (effusions) of infancy. Neuroradiology 16:79–81
127. Paciorkowski AR, Greenstein RM (2007) When is enlargement of the subarachnoid spaces not benign? A genetic perspective. Pediatr Neurol 37:1–7
128. Palencia LR, Aldana GJ, Tresierra UF (1992) Idiopathic external hydrocephaly and familial macrocephaly in infancy. An Esp Pediatr 36:186–188

129. Papasian NC, Frim DM (2000) A theoretical model of benign external hydrocephalus that predicts a predisposition towards extra-axial hemorrhage after minor head trauma. Pediatr Neurosurg 33:188–193
130. Pascual-Castroviejo I, Pascual-Pascual SI, Velazquez-Fragua R (2004) A study and follow-up of ten cases of benign enlargement of the subarachnoid spaces. Rev Neurol 39:701–706
131. Pedersen H, Gyldensted M, Gyldensted C (1979) Measurement of the normal ventricular system and supratentorial subarachnoid space in children with computed tomography. Neuroradiology 17:231–237
132. Pettit RE, Kilroy AW, Allen JH (1980) Macrocephaly with head growth parallel to normal growth pattern: neurological, developmental, and computerized tomography findings in full-term infants. Arch Neurol 37:518–521
133. Pfluger T, Weil S, Muntau A, Willemsen UF, Hahn K (1997) Glutaric aciduria type I: a serious pitfall if diagnosed too late. Eur Radiol 7:1264–1266
134. Piatt JH Jr (2001) Monozygotic twins discordant for external hydrocephalus. Pediatr Neurosurg 35:211–215
135. Pietila TA, Palleske H, Distelmaier PM (1992) Subdural effusions: determination of contrast medium influx from CSF to the fluid accumulation by computed tomography as an aid to the indications for management. Acta Neurochir (Wien) 118:103–107
136. Pittman T (2003) Significance of a subdural hematoma in a child with external hydrocephalus. Pediatr Neurosurg 39:57–59
137. Pouplard F, Pineau P (1990) Use of acetazolamide in external hydrocephalus in infants. Ann Pediatr (Paris) 37:310–312
138. Prassopoulos P, Cavouras D (1994) CT evaluation of normal CSF spaces in children: relationship to age, gender and cranial size. Eur J Radiol 18:22–25
139. Prassopoulos P, Cavouras D, Golfinopoulos S, Nezi M (1995) The size of the intra- and extraventricular cerebrospinal fluid compartments in children with idiopathic benign widening of the frontal subarachnoid space. Neuroradiology 37:418–421
140. Raimondi AJ (1994) A unifying theory for the definition and classification of hydrocephalus. Childs Nerv Syst 10:2–12
141. Ravid S, Maytal J (2003) External hydrocephalus: a probable cause for subdural hematoma in infancy. Pediatr Neurol 28:139–141
142. Robertson WC Jr, Chun RW, Orrison WW, Sackett JF (1979) Benign subdural collections of infancy. J Pediatr 94:382–386
143. Robertson WC Jr, Gomez MR (1978) External hydrocephalus. Early finding in congenital communicating hydrocephalus. Arch Neurol 35:541–544
144. Roshan K, Elizabeth C, Chacko A, Rajendra J, Gururaj A, Dilip S (1998) External hydrocephalus—a report of 16 cases from Oman. J Trop Pediatr 44:153–156
145. Rosman NP, Shands KN (1978) Hydrocephalus caused by increased intracranial venous pressure: a clinicopathological study. Ann Neurol 3:445–450
146. Rubinstein M, Denays R, Ham HR, Piepsz A, VanPachterbeke T, Haumont D, Noel P (1989) Functional imaging of brain maturation in humans using iodine-123 iodoamphetamine and SPECT. J Nucl Med 30:1982–1985
147. Sahar A (1978) Pseudohydrocephalus–megalocephaly, increased intracranial pressure and widened subarachnoid space. Neuropadiatrie 9:131–139
148. Sainte-Rose C, LaCombe J, Pierre-Kahn A, Renier D, Hirsch JF (1984) Intracranial venous sinus hypertension: cause or consequence of hydrocephalus in infants? J Neurosurg 60:727–736
149. Sakai N, Nokura H, Deguchi K, Decarlini E, Futamura A, Yamada H (1990) Surgical indications for infantile subdural effusion. Childs Nerv Syst 6:447–450
150. Sasaki H, Ishii K, Kono AK, Miyamoto N, Fukuda T, Shimada K, Ohkawa S, Kawaguchi T, Mori E (2007) Cerebral perfusion pattern of idiopathic normal pressure hydrocephalus studied by SPECT and statistical brain mapping. Ann Nucl Med 21:39–45
151. Sawin PD, Muhonen MG, Menezes AH (1996) Quantitative analysis of cerebrospinal fluid spaces in children with occipital plagiocephaly. J Neurosurg 85:428–434
152. Segal-Kuperschmit D, Cozacov C, Luder A (1995) Idiopathic external hydrocephalus. Harefuah 128(150–2):199
153. Shen WC, Yang CF, Chang T (1986) Benign hydrocephalus in infants. A computed tomographic and clinical correlative study. Acta Radiol Suppl 369:689–691
154. Shinnar S, Gammon K, Bergman EW Jr, Epstein M, Freeman JM (1985) Management of hydrocephalus in infancy: use of acetazolamide and furosemide to avoid cerebrospinal fluid shunts. J Pediatr 107:31–37
155. Shirane R, Sato S, Sato K, Kameyama M, Ogawa A, Yoshimoto T, Hatazawa J, Ito M (1992) Cerebral blood flow and oxygen metabolism in infants with hydrocephalus. Childs Nerv Syst 8:118–123
156. Smith R, Leonidas JC, Maytal J (1998) The value of head ultrasound in infants with macrocephaly. Pediatr Radiol 28:143–146
157. Snyder RD (1979) Benign "subdural" collections. J Pediatr 95:499–500
158. Stroobandt G, Evrard P, Thauvoy C, Laterre C (1981) Subdural or sub-arachnoid pericerebral effusions in the infant with subdural or subarachnoid localisation (author's transl). Neurochirurgie 27:49–57
159. Suara RO, Trouth AJ, Collins M (2001) Benign subarachnoid space enlargement of infancy. J Natl Med Assoc 93:70–73
160. Trounce JQ, De VL, Levene MI (1985) External hydrocephalus—diagnosis by ultrasound. Br J Radiol 58:415–417
161. Tsubokawa T, Nakamura S, Satoh K (1984) Effect of temporary subdural–peritoneal shunt on subdural effusion with subarachnoid effusion. Childs Brain 11:47–59
162. Upton ML, Weller RO (1985) The morphology of cerebrospinal fluid drainage pathways in human arachnoid granulations. J Neurosurg 63:867–875
163. Vertinsky AT, Barnes PD (2007) Macrocephaly, increased intracranial pressure, and hydrocephalus in the infant and young child. Top Magn Reson Imaging 18:31–51
164. Veyrac C, Couture A, Baud C (1990) Pericerebral fluid collections and ultrasound. Pediatr Radiol 20:236–240
165. Wachi A, Sato K (1997) Anatomical and biomechanical similarity in intracranial environment in identical twins with external hydrocephalus. Childs Nerv Syst 13:633–635
166. Williams CA, Dagli A, Battaglia A (2008) Genetic disorders associated with macrocephaly. Am J Med Genet A 146A:2023–2037
167. Wilms G, Vanderschueren G, Demaerel PH, Smet MH, Van CF, Plets C, Goffin J, Casaer P (1993) CT and MR in infants with pericerebral collections and macrocephaly: benign enlargement of the subarachnoid spaces versus subdural collections. AJNR Am J Neuroradiol 14:855–860
168. Wilson RK, Williams MA (2007) Evidence that congenital hydrocephalus is a precursor to idiopathic normal pressure hydrocephalus in only a subset of patients. J Neurol Neurosurg Psychiatry 78:508–511
169. Yasuda T, Tomita T, McLone DG, Donovan M (2002) Measurement of cerebrospinal fluid output through external ventricular drainage in one hundred infants and children: correlation with cerebrospinal fluid production. Pediatr Neurosurg 36:22–28

170. Zahl SM, Wester K (2008) Routine measurement of head circumference as a tool for detecting intracranial expansion in infants: what is the gain? A nationwide survey. Pediatrics 121: e416–e420

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.