External Hydrocephalus: Radiologic Spectrum and Differentiation from Cerebral Atrophy

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External Hydrocephalus: Radiologic Spectrum and Differentiation from Cerebral Atrophy

Joseph Maytal1
Luis A. Alvarez1
Charles M. Elkin2
Shlomo Shinnar1,3

External hydrocephalus (EH) is a condition in which infants with rapidly enlarging heads are found to have a CT scan that shows widening of the subarachnoid space with mild or no ventricular dilation. In this study, 74 infants with EH associated with a variety of conditions were examined clinically and with CT scans to identify the radiologic features of the condition as well as to understand its clinical and radiologic evolution. Some of these CT scans were compared with those of patients with cerebral atrophy. The characteristic CT findings in EH were bifrontal widening of the subarachnoid space and widened interhemispheric fissure frontally with only mild enlargement of the rest of the subarachnoid space. Occasional mild to moderate ventriculomegaly was also present. In most cases the radiologic findings could be distinguished from those seen in cerebral atrophy, even without the clinical history of an enlarging head. The cases of cerebral atrophy showed prominent sulci throughout without disproportionate bifrontal widening of the subarachnoid space. Two major groups of infants are identified. The largest group (47) consisted of infants with primary megalencephaly. Megalencephaly was idiopathic in 40 infants, many of whom (19) met the criteria for benign familial macrocephaly. Others (7) had a clearly defined genetic syndrome. The radiologic abnormalities in these infants usually disappear by 2 years of age. The other major group (14) consisted of infants who sustained CNS or systemic insults such as subdural hematomas (8) or meningitis (3), which cause impaired CSF absorption. Thirteen premature infants with EH were also identified. It is concluded that EH is an age-related self-limited condition occurring in infants with open cranial sutures and that usually resolves without intervention by 2-3 years of age.

Excessive head growth is a common problem in infancy. Studies in the pre-CT era suggested that the most frequent cause of macrocephaly is communicating or noncommunicating hydrocephalus [1]. With CT, however, another entity is now frequently seen during the evaluation of infants with macrocephaly, namely external hydrocephalus (EH) or the enlargement of the subarachnoid spaces with mild to moderate or no ventricular dilation [2]. In the last few years over 100 cases of EH in infants have been reported [2-12]. Nonetheless, the condition remains poorly understood and is still frequently unrecognized or misdiagnosed.

The multiple terms used to describe external hydrocephalus, such as “pseudohydrocephalus-megalomégencephalie” [4], “benign subdural collections of infancy” [6], and “extraventricular obstructive hydrocephalus” [7] demonstrate the confusion that has surrounded the entity. The difficulty with diagnosing and recognizing EH is due in part to the frequency with which it is associated with other conditions, such as prematurity [9, 11], subdural hematomas [6, 10], intraventricular hemorrhages [9], and genetic syndromes [2, 13]. Another problem is the radiologic similarities between EH and cerebral atrophy. In fact, according to many authors, the differentiation between these two entities is very difficult without the history of an enlarging head [2, 10].

The present study summarizes the radiologic and clinical findings of 74 infants with EH seen over the last several years. It extends the results of a previous study...
on idiopathic EH [14]. In spite of its frequent association with a number of dissimilar conditions, EH appears to be a distinct radiologic entity that can clearly be differentiated from cerebral atrophy.

**Materials and Methods**

The CT scans of all infants evaluated for macrocephaly at Montefiore Medical Center and Bronx Municipal Hospital Center during the period January 1975 through December 1983 were studied retrospectively. The 41 who met the criteria for EH were identified and their medical records reviewed. During the period January 1984 to June 1985, 33 infants with EH were identified prospectively. The criteria used to define external hydrocephalus [2, 11, 14] were (1) maximal occipitofrontal head circumference (OFC) greater than 95th percentile on the standard growth curve [15] or head circumference below or at the 95th percentile but that had crossed at least two percentile lines on the standard growth curves, and (2) radiographic evidence on CT scan of unusually prominent subarachnoid space with or without some degree of ventricular dilation present during the time of head growth (Fig. 1).

Infants were sedated for the scan with a dose of 50–75 mg/kg of chloral hydrate or a combination of 2 mg/kg of meperidine with 1 mg of hydroxyzine. There were no complications. CT scans were performed without contrast enhancement using GE models 8800 and 9800 and an EMI 5005 scanner.

A total of 74 patients with EH were identified and studied. These patients were grouped according to their history and each group was studied separately. The term prematurity was used to identify all infants with an estimated gestational age of less than 36 weeks at birth. Those born with an estimated gestational age ≥36 weeks who did not meet the criteria for a recognized genetic syndrome and who

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Fig. 1.—Radiologic spectrum of external hydrocephalus (EH). All cases are noncontrast CT scans.

A, Idiopathic EH in 4-month-old boy. CT scan shows prominent widened subarachnoid space in frontal and frontoparietal regions and enlarged interhemispheric fissure frontally. Frontal horns are also enlarged.

B, Idiopathic EH in 8-month-old boy with rapidly increasing head circumference. Note prominent widened subarachnoid space frontally and enlarged interhemispheric fissure. Mild ventricular enlargement is present. While sulci directly adjacent to wide subarachnoid space in frontal and frontoparietal regions are prominent, the parietal and occipital sulci are normal.

C, EH occurring in 9-month-old infant with Weaver syndrome. Radiologic findings are similar to those in B, but more severe and asymmetrical.

D, EH occurring in 4-month-old girl who had been born prematurely after 32 weeks gestation and who presented with rapid head growth. CT scan shows widened subarachnoid space frontally and prominent interhemispheric fissure without evidence of sulci enlargement or ventricular dilation.
did not have a history of an identifiable CNS insult immediately before developing EH were labeled idiopathic.

Eight of the patients with EH were selected at random and their CT scans compared with those of eight infants with cerebral atrophy [16]. Cerebral atrophy was defined as radiologic evidence of diffuse loss of brain tissue (i.e., prominence of the cortical sulci out of proportion to the degree of ventricular dilation with a pattern of ventricular dilation that tends to be more prominent frontally) [17] in patients who were either microcephalic (OFC <5th percentile on standard growth curve [15]) or had an OFC above the 5th percentile with arrested head growth after an insult. All 16 scans were then reviewed by two of the investigators, who were blinded to the clinical diagnosis of each scan.

Results

Study Population

A total of 74 CT scans of patients with EH were identified. EH was associated with a variety of conditions as summarized in Table 1. The largest identifiable group consisted of 40 infants with idiopathic EH. These infants had a uniformly benign prognosis. In 22 infants, development was mildly delayed in the first year of life but was normal in all but one by age 2 years. In addition, family members with benign familial macrocephaly [18] were identified in 85% of the cases in which a family history was available. Thus, this entity probably represents the early and transient radiologic findings of benign familial macrocephaly. The clinical and radiologic findings in 36 of these 40 infants with idiopathic EH have been previously reported [14].

The remaining 34 infants had a variety of associated conditions, such as known genetic syndromes (i.e., achondroplasia [13], Beckwith [19], Soto [20], Goldenhar [21], and Weaver [22]) (7), prematurity (13), CNS insults (11), and systemic insults (3). The clinical course in these patients did not vary from that of infants with similar conditions who did not have EH. All infants presented in the first year of life. The cranial sutures were open in all infants with EH.

Radiologic Criteria and Differentiation from Cerebral Atrophy

The CT scans of patients with EH differed in severity but were qualitatively similar in all cases, regardless of etiology. The main findings consisted of prominent widening of the subarachnoid space in the frontal and frontoparietal regions and of the interhemispheric fissure frontally. Some degree of widening, but to a much lesser extent, was also present in the rest of the subarachnoid space. These patients had normal (64%) or mild to moderately dilated (36%) ventricles. The basal cisterns were enlarged in 68% of the cases. The sulci often appeared prominent in the frontal and frontoparietal regions but were of normal appearance in most other areas. The spectrum of EH due to a variety of causes is shown in Figure 1.

In the CT scans of eight patients with cerebral atrophy the main radiologic finding was diffuse prominence of the sulci distributed throughout the entire cerebrum (Fig. 2). The ventricles and basal cisterns were dilated proportionately to the subarachnoid dilation. Ventricular dilation was more prominent than in the cases with EH. Prominence of the cerebellar sulci was also occasionally present (38%). The interhemispheric fissure was usually normal and, when widened (38%), was prominent throughout and not just in the frontal areas. In no case was there prominent widening of the subarachnoid space in the frontal and frontoparietal region that was disproportionate to the rest of the subarachnoid space. When these CT scans were compared with those of patients with EH by observers who were unaware of the clinical diagnosis in each

<table>
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<th>TABLE 1: Conditions Associated with External Hydrocephalus</th>
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<td><strong>No. of Cases</strong></td>
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<td>Idiopathic</td>
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<td>Benign familial macrocephaly</td>
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<td>Genetic syndromes</td>
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<tr>
<td>Achondroplasia</td>
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<td>Beckwith syndrome</td>
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<td>Cerebral gigantism (Soto syndrome)</td>
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<td>Goldenhar syndrome</td>
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<td>Weaver syndrome</td>
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<td>Prematurity</td>
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<td>CNS insults</td>
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<td>Subdural hematoma</td>
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case, there was no difficulty distinguishing between the two entities, and the correct diagnosis was made in all cases.

In one case the radiologic picture of a patient with acute subdural hematomas correlated with changing clinical findings. Soon after the development of the subdural hematoma the OFC rapidly increased and a repeat CT scan showed the characteristic features of EH. These findings persisted through the period of increased head growth. Subsequently, there was arrest of the head growth, at which time the CT scan showed the characteristic findings of cerebral atrophy. The sequential CT scans and their correlation with the rate of head growth are shown in Figure 3.

Fig. 3.—EH developing after a subdural hematoma.
A, 2-month-old boy with acute subdural hematoma in right posterior interhemispheric fissure. Increased density in posterior limb of left internal capsule is artifactual.
B, CT scan performed 5 days later, a time of rapidly increasing head circumference (see Fig. 3E). Widened subarachnoid space in frontal region has developed while interhemispheric subdural hematoma has resolved.
C, CT scan 1 month after initial insult shows typical findings of EH with prominent widening of subarachnoid space frontally (left more than right) and widened interhemispheric fissure. Head circumference was still increasing rapidly.
D, Posttraumatic atrophy. 8 months later the disproportionate widening of subarachnoid space frontally has disappeared and external hydrocephalus has resolved. However, cortical sulci are large, especially in parietal lobes, suggesting residual atrophy. Head growth had arrested approximately 7 months before this CT.
E, Graph correlating head circumference growth rate with CT scans (Figs. 3A-3D). The three curves in growth chart represent 5th, 50th, and 95th percentiles, respectively [15].
Radiologic Evolution of EH

1. Idiopathic and genetic syndromes.

The evolution of the CT scans of patients with idiopathic external hydrocephalus and those of patients with EH associated with genetic syndromes was very similar. The first area that appeared to enlarge was the interhemispheric fissure frontally, followed by the subarachnoid space in the frontal and frontoparietal regions. Subsequently, the basal cisterns enlarged, and, finally, ventricular dilation, when it occurred, was a late finding. The time of onset of the EH was difficult to assess. Since the reason for referral was to evaluate an already enlarging head, EH was already present in the first CT scan of all patients. The CT picture of EH and the rapid head growth, which were usually present by age 3 months, appeared to stabilize at approximately 15–18 months of age. The radiologic findings of EH resolved by 2–2.5 years of age, after which the CT was that of a megaloecephalic child with only occasional minimal widening of the interhemispheric fissure frontally (Fig. 4). None of the patients developed significant (internal) hydrocephalus or received any form of medical or surgical therapy for the EH.

2. CNS insults.

The patients with subdural hematomas developed the CT scan of EH soon after the onset of the hematomas, usually within 3–4 days (Fig. 3). In all cases the CT picture resembled that of the patients with idiopathic external hydrocephalus except for the presence of the subdural hematomas and occasional blood in the subarachnoid space. In contrast to the infants with idiopathic EH, in this group of infants once the CT picture of EH started to resolve, the OFC returned to normal. In three cases, however, it was followed by development of microcephaly and a CT picture of cerebral atrophy, as illustrated in Figure 3. Progressive hydrocephalus requiring a shunt did not develop in any of these infants.

In the patients with meningitis, EH also developed soon after the onset of the infection. Although difficult to differentiate from subdural effusion, the diagnosis of EH was made by the prominence of the cerebral sulci under the enlarged (subarachnoid) space and the prominent interhemispheric fissure. The sulci tend to be obliterated in the cases of the subdural effusions. There has been no resolution of the CT picture in the two infants who are still under the age of 18 months. A picture of mild cerebral atrophy developed in the third and older patient, who is now microcephalic.

3. Prematurity.

There were 13 premature infants with EH. Eight also had intraventricular hemorrhage, a complication frequently found in premature infants [23]. The radiographic picture and evolution of EH in these 13 infants (Fig. 1D) was similar to that of the infants with idiopathic EH.

Discussion

The term external hydrocephalus was first used by Dandy in 1917 to describe enlargement of the subarachnoid space in the presence of increased intracranial pressure [24]. It was only recently that Robertson and Gomez [3] reintroduced the term to describe a condition in which children with enlarging heads have a CT scan of enlarged subarachnoid spaces with mild to moderate or no ventricular dilation. There appears to be an excess of normal CSF in the subarachnoid space. This was demonstrated by Andersson et al. [12] who performed craniotomies on four patients with idiopathic EH and found an enlarged subarachnoid space without other abnormalities.

EH has frequently been associated with a number of conditions (Table 1). In addition to the conditions found in our study it has also been reported in association with vitamin A deficiency [25], intraventricular hemorrhage [2, 9], and sub-
arachnoid hemorrhage [9]. The pattern of radiologic findings in all these patients, regardless of the presence or absence of associated conditions, appears to be identical. Thus, the accurate diagnosis of EH should not be overlooked because of the associated condition. The possibility of EH should be considered in any infant with a rapidly enlarging head.

In some specific instances, particularly in infants with subdural hematomas, the recognition of EH may be difficult, in spite of the history of an enlarging head. Zimmerman et al. [26] reported a large number of abused infants who developed “cerebral atrophy” immediately after the development of subdural hematomas. This “cerebral atrophy” resolved in some of the children and persisted in others. EH is a common finding soon after the development of subdural hematomas [2, 10, 11] (Fig. 4). This phenomenon, which probably results from mechanical obstruction of the arachnoid villi by the subdural hematomas, usually disappears once the hematomas resolve. At that point, however, the brain may either return to normal or, because of significant damage from the original insult, become atrophic. One would assume that at least some of the infants with transient “cerebral atrophy” reported by Zimmerman et al. [26] may have had EH. Although the diagnosis in this particular group of infants can be difficult, when one studies the radiologic findings in serial CT scan evaluations together with the concurrent OFC growth rate, an accurate differentiation between EH and cerebral atrophy can usually be made. Such a differentiation is particularly important in this group of patients because of the medical-legal implications often present.

In the absence of a clinical history and OFC growth curve, differentiating between EH and cerebral atrophy, because of their radiographic similarities, has been difficult. Many of our patients with EH were originally diagnosed as having “cerebral atrophy.” However, we found certain radiologic differences between EH and cerebral atrophy that when present allow a radiologic differentiation between these two entities in most cases, even with little knowledge of the clinical history. EH is invariably associated with prominent widening of the subarachnoid space in the frontal and frontoparietal region disproportionate to the rest of the subarachnoid space and widening of the interhemispheric fissure in the frontal region. Atrophy, on the other hand, is almost always associated with prominence of the cerebral sulci throughout and does not exhibit the two main features of EH, both of which should be present in order to make the diagnosis of EH. To determine whether the frontal fluid collection could be due to the infant’s position, we attempted to scan several infants in the brow-down position. This failed because the infants invariably either woke up or had signs of respiratory compromise. However, we believe that the location of the fluid is truly frontal and not an artifact of position. Support for this comes from the fact that in CT scans of infants with atrophy (Fig. 2) there are equally prominent sulci and fluid frontally and occipitally. In addition, the neurosurgical data cited earlier [12] found enlarged subarachnoid fluid spaces frontally. The size of the basal cisterns and the degree of ventricular dilation cannot be used to differentiate the two conditions.

It is important to distinguish between EH and cerebral atrophy because the two conditions have very different clinical outcomes. Idiopathic EH appears to be a benign self-limited condition that resolves spontaneously without sequelae by 2–3 years of age [14]. On the other hand, the presence of cerebral atrophy is often associated with a poor neurologic prognosis. In the cases of patients with CNS insults (i.e., subdural hematomas and meningitis) and EH, the outcome is variable and the radiologic EH may either resolve, evolve to a picture of cerebral atrophy (Fig. 3), or lead to the development of progressive hydrocephalus. In this particular group it appears that the severity of the original insult, not the EH, dictates the eventual outcome.

While in premature infants the radiologic evaluation of EH is similar to that of idiopathic EH, with resolution by 2–3 years of age, the neurologic outcomes were more variable. Prematurity is frequently associated with CNS insults, such as intraventricular and subarachnoid hemorrhages, which often occur without symptomatology [23, 27]. Since detailed radiologic studies in the perinatal period to distinguish between the two possibilities (which clearly can affect neurologic outcome) were not available, the premature infants were classified separately.

The finding of EH in a variety of infants with recognized genetic syndromes is particularly interesting. Primary megalencephaly appears to be a common feature in these disorders as well as in benign familiar macrocephaly. We could not find any report of EH in these genetic syndromes. However, the syndromes are relatively rare and most published scans are of children over 2 years of age. Perhaps EH is an early finding in all primary megalencephalic syndromes. Further work on defining the primary and secondary megalencephalic syndromes is currently in progress.

The size of the subarachnoid spaces in children is variable, and radiologic findings qualitatively similar to those seen in EH are occasionally seen in normocephalic patients [28]. Kleinman et al. [29] found that some degree of subarachnoid enlargement in normal patients is not uncommon during infancy. However, in this group of children the widening of the subarachnoid space was of a much lesser degree than that of children with EH. Unfortunately, in that study, OFC growth curves immediately before or during the CT scan evaluation were not reported. This could have helped distinguish normal infants from patients who may have had early onset of EH or cerebral atrophy. Nonetheless, due to the benign nature of idiopathic external hydrocephalus [14], the differentiation between EH and the mildly enlarged subarachnoid space occasionally found in normal infants is not as critical as the differentiation between EH and cerebral atrophy.

A few other conditions must be considered when evaluating infants with an enlarging head and CT findings of a widened subarachnoid space with minimal or no ventricular dilation. These include malnutrition with “catch-up growth” [30], steroid therapy, and chemotherapy [31]. These conditions are all capable of producing reversible brain shrinkage in patients who may exhibit subsequent rapid head growth. Another group of patients that can present with a CT picture resem-
bling EH are infants with neurodegenerative diseases such as Canavan’s and Alexander’s disease [32, 33]. These patients originally present with an enlarging head and CT picture of megalooencephaly, but may develop cerebral atrophy at a later age when they already have macrocranium. In this last group of patients, if the CT picture is obtained at a later age and the head circumference measurement is obtained without looking at the growth curve, the diagnosis of external hydrocephalus can be made erroneously. In all these cases, however, even though there may be an earlier history of a “large head” or of an enlarging head, at the time of the radiologic similarities the head growth has arrested and the CT appearance is that of cerebral atrophy.

The pathophysiology of external hydrocephalus is poorly understood. The most widely accepted hypothesis is that it is caused by a defect of CSF absorption at the level of the arachnoid villi in the presence of open cranial sutures [2, 11]. An elegant study of CSF absorption in infants with idiopathic EH showed delayed absorption of CSF in the infants by means of isotope cisternography [34]. The isotope pattern, which included delayed absorption and CSF reflux into the fourth and lateral ventricles, was similar to that seen in communicating hydrocephalus, which led the authors to call this condition “external obstructive hydrocephalus.” Although this hypothesis may fully explain EH resulting from insults affecting the CSF pressure gradient at the level of the arachnoid villi, it does not fully explain why EH is often present in infants with primary megalooencephaly. When one combines the finding of this study with those of other reports [2–12], it appears that EH occurs in two major groups of patients. The first and largest group consists of infants with primary megalooencephaly, either in isolation or in the context of a known genetic syndrome affecting multiple systems. The second group consists of infants in whom a CNS (e.g., subdural hematoma and meningitis) or a systemic condition (e.g., increased venous pressure) results in decreased CSF absorption. An intriguing hypothesis would be that the syndromes associated with primary megalooencephaly had an associated delay in the maturation of the arachnoid villi, thus causing EH.

Subdural hematomas and meningitis lead to the impairment of CSF absorption due to mechanical or inflammatory changes at the level of the arachnoid villi. While in some infants, and especially in older children with closed fontanelles, a communicating hydrocephalus may develop after these insults [2, 11], in the young infant with open sutures EH is a more common finding. In infants, increased venous pressure causing changes in CSF pressure gradient can result in both (internal) communicating hydrocephalus [35] and EH [2]. In adults, however, this same condition will usually cause pseudotumor cerebri [11, 35]. Interestingly, EH does not occur in children with closed fontanelles, and pseudotumor cerebri is not reported in children under 1 year old. Thus it appears that EH is an age-related phenomenon closely related to both (internal) communicating hydrocephalus and pseudotumor cerebri, and that the presence of open sutures is necessary for its development. Further studies, involving a much larger population and a longer follow-up, may still be necessary to define the full spectrum of EH and to determine the developmental and anatomic factors that lead to its occurrence in this population.

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