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Hypodense extracerebral images on computed tomography in children

"External hydrocephalus": a misnomer?

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Abstract. External hydrocephalus (EH) was identified in 58 infants under 3 years of age during the period 1 June 1986 to 28 February 1990. Radiological images and clinical features were compared with 11 cases of cerebral atrophy (CA). Significant differences were found in delivery, head circumference, and the incidence of motor and developmental abnormalities. The population with EH was found to be quite heterogeneous, with a male preponderance. Intracranial pressure was normal in 15 cases in which lumbar puncture was done. The flow of cerebral spinal fluid was considered to be within the normal range in 6 cases. The results of a few cases examined with metrizamide cisternography are presented. The prognosis in our cases was not as benign as previously published, and use of the name EH is questioned. The less committal term "hypodense extracerebral images" is proposed when computed tomography (CT) is the only study done. The hypothesis that encephalocranial disproportion is the basic underlying entity for the CT images is proposed.

Key words: External hydrocephalus – Cerebral atrophy – Encephalocranial disproportion

The term "external hydrocephalus" (EH) was first introduced by Dandy between 1914–1918 [7, 8] to describe infants with increased intracranial pressure and dilated subarachnoid spaces. Since then, it has been used with increased frequency either in its original form or in any of the other denominations considered equivalent: pseudohydrocephalus-megalocephaly [29], benign subdural collections of infancy [28], extraventricular obstructive hydrocephalus [25], and benign enlargement of the subarachnoid spaces in the infant [19]. Correlations between radiological diagnosis of EH, clinical findings, and cerebrospinal fluid dynamics have been published [1, 2, 5, 11, 12, 19–21, 24, 25, 27, 28, 30].

Use of the term appeared confusing to us and prompted a review of all cases with a computed tomography (CT) diagnosis of EH and cerebral atrophy (CA) registered at Hamad General Hospital within the period 1 June 1986 through 28 February 1990 in order to evaluate: (a) the reliability of CT to identify EH as opposed to CA; (b) the correlation of CT with a well-defined population or clinical entity; (c) the correlation between the actual findings in our cases and the presumed physiopathology.

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Patients and methods

All CT scans with a diagnosis of EH and cerebral atrophy (CA) performed at Hamad General Hospital in infants less than 3 years old during the period 1 June 1986 to 28 February 1990 were reviewed retrospectively. The radiological review was made by two independent observers unaware of the previous diagnosis, clinical history, or purpose of the study. Patients were referred for CT studies from either the pediatric or neurology departments. The most common causes for requesting CT were seizures, abnormal development, abnormal head size or abnormal neurological examination. A total of 63 patients with EH and 11 with CA were identified and studied. Five cases of EH had to be omitted due to interobserver variation. In all 5 cases the discrepancy was whether the frontal subarachnoid space was enlarged or normal for the patient's age. The radiological criteria used for diagnosis of EH (Fig. 1) was the presence of a symmetrically enlarged "subarachnoid space" in the frontal area plus enlargement of the sylvian fissures, anterior interhemispheric fissure and variable enlargement of the basal cisterns and ventricles (0 to 2 in a scale 0 to 5). CA (Fig. 2) was diagnosed when the sulci enlargement was marked, widespread, or asymmetric, and ventricular and/or basal cistern enlargement surpassed 2 in the above scale, with or without hypodense areas in the parenchyma.

Cerebrospinal flow studies were performed in 6 cases, using simultaneous injection of metrizamide (3.5–4.0 cm^3/180 mg/ml) and < 500 Ci of technetium-99m diethyl triamine penta-acetic acid (^{99m}Tc-DTPA) in 3 cases, and only the isotope in the remaining 3. Patients were scanned immediately and at variable intervals up to 24 h.

Follow-up was accomplished in 49 cases of EH and in 9 of CA. Developmental assessment was done according to the guidelines of the Denver Development Screening Test [10]. The Chi-square analysis with Yates correction was used to compare appropriate variables between groups. A $P < 0.05$ was considered significant.

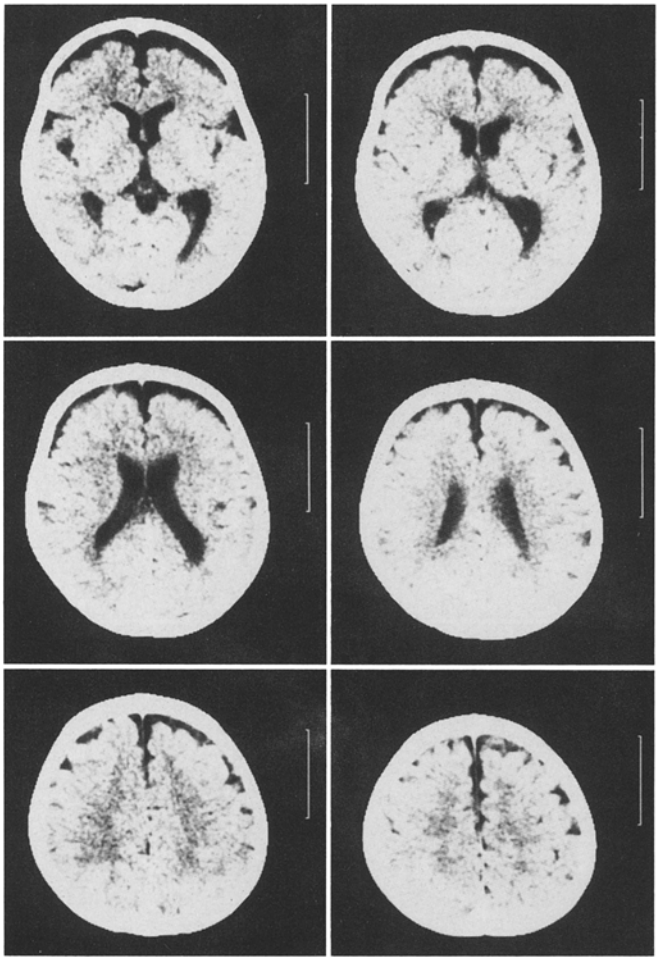

Fig. 1. Case HGH 858936, a 16-month-old male with "external hydrocephalus" (EH)

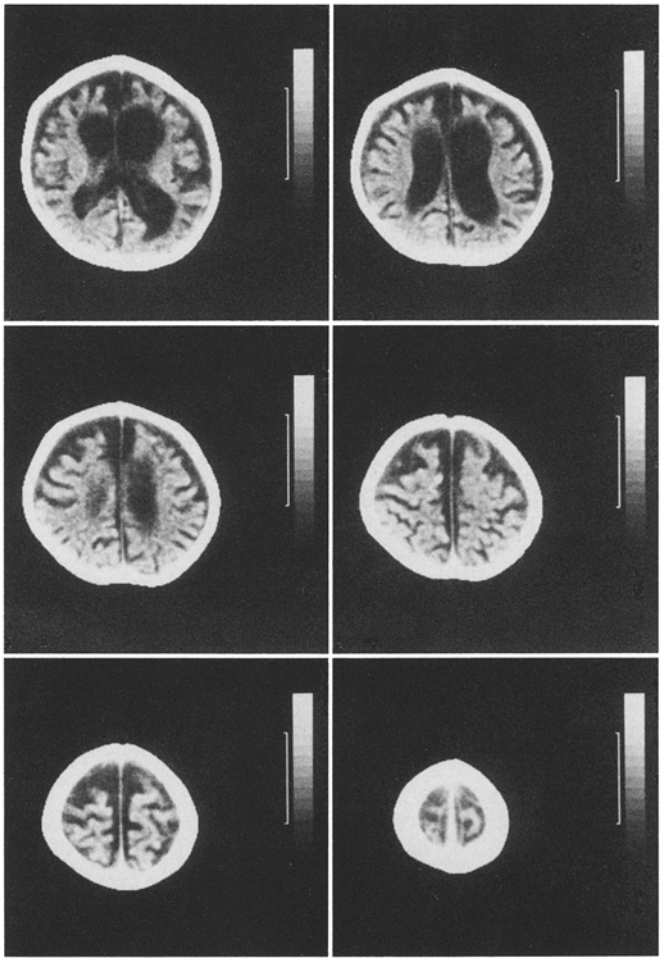

Fig. 2. Case HGH 854941, a 10-month-old male with cerebral atrophy (CA)

Results

Evaluation of the population characteristics of external hydrocephalus (Table 1) showed that most had a normal delivery at term. Birth weight and Apgar scores were also predominantly within the normal range. Of particular importance is the fact that the head circumference was also predominantly within the normal range with an almost equal distribution in the micro- and macrocephalic ranges. A positive family history and an abnormal neonatal period show a significant incidence in this group.

Seizures and abnormal development were the leading causes for evaluation, with head enlargement in third place and almost equal to a combination of several items (Table 2). This points to the heterogeneity of the population when CT findings are used as the selection criteria.

Examination of these children was consistent with the presenting signs and symptoms, as the neurological examination was predominantly abnormal (Table 3). EEG abnormalities were also often found.

Under no circumstances could increased intracranial pressure be proven when a spinal tap was performed. Flow studies were unable to demonstrate significant flow impairment, with the interesting finding that on metrizamide cisternography we were unable to visualize contrast

Table 1. Population characteristics of external hydrocephalus (EH) ($n=58$)

Age	8.84 months (1.5–26)
Sex (male/female)	44/13 1 undetermined
Gestational age	
Term	42
Preterm	8
Post-term	4
Unknown	4
Normal delivery	41
Apgar score 1′	8 (2–9)
Apgar score 5′	9.5 (2–10)
Birth weight	2976 (1100–4320) gm
Head circumference at birth	
Percentile	
<10	11
25–75	30
>90	7
Not recorded	10
Neonatal period	
Normal	24
Abnormal	33
Unknown	1
Family history	
Negative	35
Positive	23
Consanguinity	10

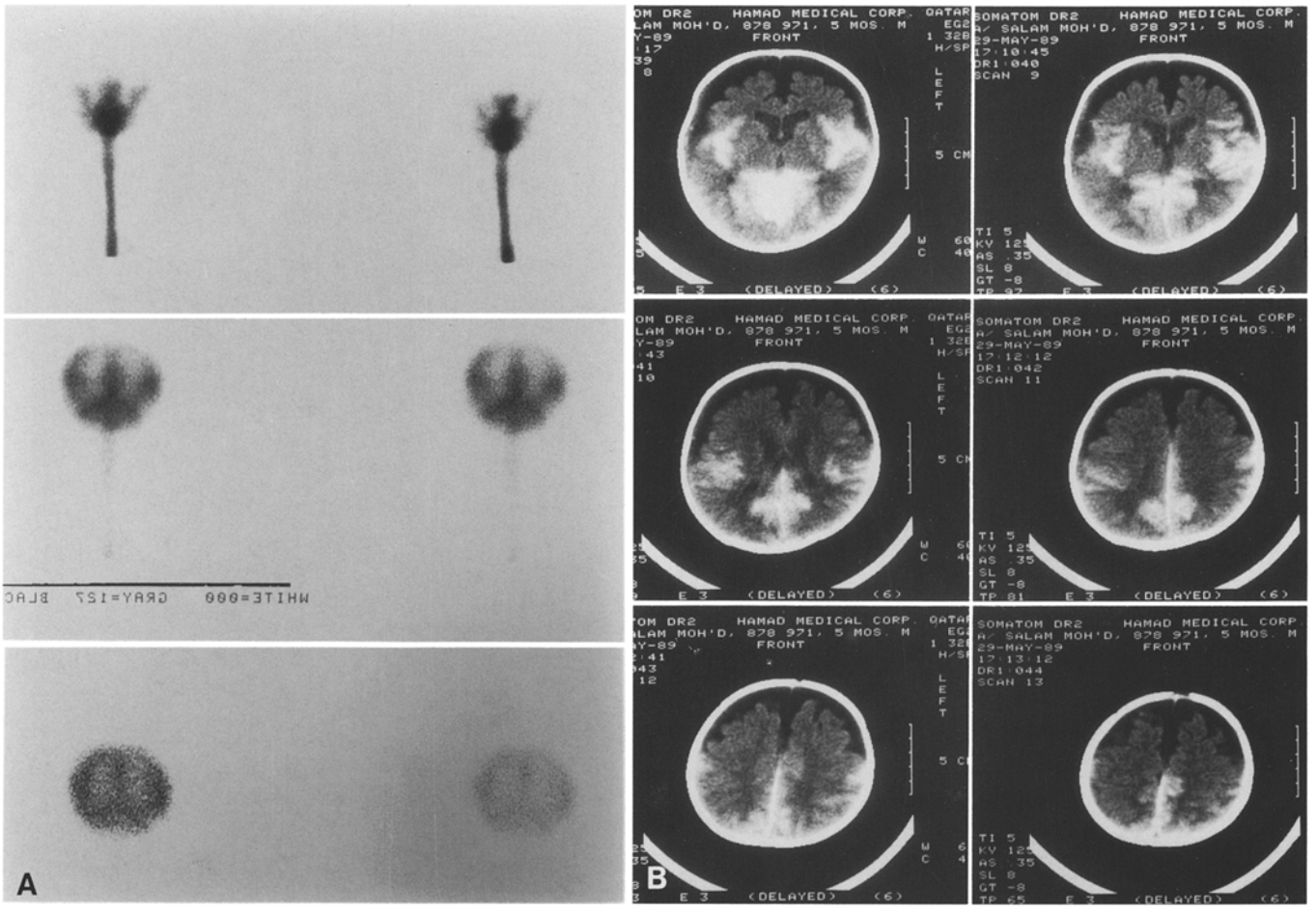

Fig. 3 A, B. Case HGH 878971, a 5-month-old male with EH. Bilateral subdural taps were negative on two separate occasions. Simultaneous isotope and metrizamide cisternography: isotope cisternography at 5, 10 and 20 h (**A**). Metrizamide cisternography at 8 h (**B**)

beyond the Silvian fissures, up to 12 h post-intrathecal injection (Fig. 3). Subdural taps, although performed in only four cases, were interesting, as all possible alternatives were found. We consider them significant, as the taps were repeated and bilateral and no discordant findings were seen in any case.

Follow-up (Tables 4, 5) shows that this condition, when diagnosed by the criteria used in this series, is not really benign and tends to be a rather stable or fixed condition, with a large proportion of children requiring treatment and remaining abnormal, at least within our observation period. On the other hand, head circumference did not show a preferential pattern towards macrocephaly. This is at variance with previous contentions of external hydrocephalus associated with macrocephaly and a benign course. However, when macrocephaly is present, it is usually an isolated finding (6/8). When interpreting these findings, one should remember that children with isolated macrocephaly may have CT findings similar to the whole group, but constitute a restricted population, probably representing encephalocranial disproportion.

The distribution of head circumferences (shown in Table 7) failed to show a high incidence of large head size outside the normal range at any stage. Finally, comparison with cases of atrophy demonstrated two definitely different populations, validating our radiological criteria in which we differentiate external hydrocephalus from cerebral atrophy (Table 6).

Discussion

The CT diagnosis of EH using criteria similar to those used in our series has been associated with different clinical findings, such as male preponderance, macrocephaly, family history of macrocephaly, rapid head growth, delayed motor development and benign prognosis [18, 24, 27]. It has also been related to prematurity, subdural

Table 2. Presenting signs and symptoms

Seizures	23
Abnormal development	17
Head enlargement	12
Abnormal neurologic examination	5
Vomiting	2
Trauma	2
Irritability	1
Drowsiness	1
Miscellaneous	7
Combination of above	10

Table 3. Examination at the time of CT

Abnormal development	22
Abnormal level of response	8
Motor abnormalities	4
Cranial nerve abnormalities	3
Suture splitting	2
Bulging fontanel	3
Head circumference	
Percentiles	
<10	15
25–75	24
>90	13
Not recorded	6
EEG ($n=26$)	
Normal	12
Abnormal	14
Lumbar puncture ($n=15$)	
Normal pressure and CSF	12
Abnormal 3 (1 "dry tap", 2 meningitis)	
Subdural taps ($n=4$)	
Negative	1
Normal CSF in small amount[a]	2
Xanthochromic fluid in small amount[a]	1
Metrizamide cisternography ($n=3$)	
All showed filling of the subarachnoid spaces up to the sylvian fissure, but no filling of the "enlarged frontal subarachnoid spaces" up to 12 h	
Isotope cisternography ($n=6$)	
All showed a pattern similar to type II of Velardi et al. [30]. Three had transient ventricular reflux. Two had asymmetric convexity flow	

[a] Few drops obtained spontaneously; no fluid obtained by siphoning or gentle suction

Table 4. Follow-up

Mean follow-up:	14 months (1–42)
Lost	9
>6 months of follow-up	35
Treatment	
Anticonvulsants	21 (prescribed) 15 (compliant)
Rehabilitation	6
None	25
Surgery	3 (occipital encephalocele repair, 2; craniosynostosis)
Miscellaneous	2
Follow-up CT	13
Normal	2
Improved	3
Unchanged	8
Mean age at follow-up CT: 20.7 months (range 4–36)	

hematomas, subarachnoid hemorrhages, intraventricular hemorrhages, vitamin A deficiency, genetic syndromes, agenesis of the corpus callosum, malnutrition, use of adrenocorticotrophic hormone or steroids, chemotherapy, and as an initial stage of communicating hydrocephalus [1, 9, 13, 14, 17, 19, 27].

Table 5. Condition at follow-up

Only cases with a follow-up of 6 months or longer ($n=35$)	
Abnormal development	14
Seizures	5
Head circumference	
<10	7
25–75	14
>90	8
Not recorded	6
Macrocephaly as the only abnormality (6/8)	
Microcephaly as the only abnormality (0/7)	
Died	1

Table 6. Differences between EH and CA

	EH ($n=58$)	$P<$	CA ($n=11$)
Abnormal delivery	17	0.05	7
Abnormal development	22	0.02	9
Head circumference: <10 percentile	10	0.01	6
Motor abnormalities	4	0.01	5
At follow-up	$n=35$		$n=9$
Abnormal development	17	0.05	8
Head circumference: <10 percentile	7	0.02	6

Table 7. Comparison of head circumference in EH at different stages

Percentile	Birth ($n=58$)	Time of diagnosis ($n=58$)	Follow-up ($n=35$)
<10	11	15	7
25–75	30	24	14
>90	7	13	8
Not recorded	10	6	6

The same variety is found when the intracranial pressure and CSF dynamics are considered. All possibilities have been described, including the often found normal pressure and normal or minimally delayed flow, actually at odds with the original description and name of EH [11, 12, 21, 29, 30]. From a clinical point of view it may be summarized that this entity has been reported in a pediatric population ranging from otherwise normal children with large heads to impaired children with small heads.

Our results show the same heterogeneity, but only two consistent findings: age when the diagnosis is made and male preponderance. According to our results and a review of previously published series, a heterogeneous population is to be expected when the radiological findings are used as the only criteria for case selection. Therefore, attempts to correlate an isolated clinical characteristic such as macrocephaly, benign prognosis, etc. with the radiological diagnosis of EH, represents an incorrect restriction related to selection criteria and not a unique, scientifically proven correlation.

Comparison of head circumferences of EH at different stages (Table 7) failed to show a definite correlation with a particular head size at any time. As a matter of fact, a head circumference within the normal range seemed to be

predominant, with a similar distribution of values above and below. This is against the assumption of a unique association of EH and macrocephaly.

CT diagnosis is not free from error. Difficulties in the differential diagnosis of subdural collections, enlarged subarachnoid spaces and atrophy have been acknowledged by a number of authors [6, 14, 15, 19, 20, 27, 28, 30]. The clinical differences between EH and CA showed that the radiological criteria used in our study were reasonably adequate to differentiate between these two clinical entities and may be interpreted as suggestive that our sample of EH was not unduly contaminated by cases of CA. This rules out one of the possible causes for having an heterogeneous clinical population. However, some of our cases highlight the diagnostic difficulties. The metrizamide and isotope cisternographies, as well as the subdural taps, failed consistently to show cerebrospinal fluid accumulation in the subarachnoid spaces, as expected according to the CT images. Similar findings have been reported previously [1, 9, 20, 27]. The follow-up results, either clinical or radiological (CT), showed that a considerable proportion of our patients remained abnormal. This is coincident with previous reports by Kendall and Hollan, Robertson et al. and Sahar [15, 27, 29], but at variance with those using the term "benign" to qualify this entity or suggesting a good prognosis [1, 2, 18–21, 24, 25]. As previously expressed, we consider the difference to be related to the selection criteria.

None of our cases in which a lumbar puncture was done showed evidence of increased intracranial pressure. When CSF dynamics studies were done, as in some of our cases, the results were compatible with normal or minimally impaired flow [11, 15, 20, 24, 29, 30]. Therefore, unrestricted use of the term EH seems inappropriate. The criteria for such a diagnosis is not fulfilled, as there is neither uniformly proven fluid accumulation in the subarachnoid space, under increased pressure, nor clearly demonstrated flow or absorption impairment.

Also, as pointed in another study [27], flow or absorption impairment still leaves unexplained the peculiarly apparent distribution of fluid accumulation. Metrizamide cisternography findings were interesting in this aspect, as all cases showed a consistent pattern of rapid flow up to the sylvian fissures and posterior subarachnoid spaces, but no filling of the presumably enlarged anterior subarachnoid spaces (Fig. 3). Whether the CT hypodense peripheral image represents a separate CSF compartment, a subdural collection, or simply an empty space remains open to further investigation. Much to our surprise, the negative subdural taps in the case of Fig. 3 suggest, at least for this case, the possibility of an empty space. This is in accordance with our proposal of encephalocranial disproportion as the underlying condition. Consequently, we consider that the term EH should not be used unless accurately demonstrated by different means in addition to CT. We prefer (and propose) the less committal but more operational term "hypodense extracerebral images" until the diagnosis of EH is proven.

We also propose, as a working hypothesis, that the isolated CT findings of hypodense extracerebral images in children under 2 years of age who are clinically asymptomatic may represent encephalocranial disproportion, that there is male preponderance, and that it will disappear with age. It may be considered a normal variant. Different data, such as the uncoupled inheritance of brain and skull size [22, 23], the significant male preponderance for macrocephaly and abnormal suture separation, the larger subarachnoid spaces in infants below 2 years [16], and the poor correlation between head circumference and intracranial volume [11, 12] lend support to this hypothesis. Our proposal is in agreement with Bode [4].

In order to prove it, the incidence of this radiologic finding in a large, unselected group of normal children should be known, as should the sex distribution and evolution. This information is hard to obtain, as normal children are seldom evaluated by CT.

All other conditions with a similar CT image, may represent one of three possible alternatives:

1. The disproportion is related to unusually fast growth of the skull, with normal brain growth.

2. The disproportion is related to delayed or impaired brain growth.

3. The disproportion is related to truly enlarged subarachnoid spaces, with either secondary skull enlargement or brain growth delay or impairment.

These three possibilities have different etiologies, clinical manifestations, prognosis, and treatment. They may also occur in a child of the normal variant group and therefore be considered as an intercurrent disease responsible for the clinical manifestations, which is otherwise not expected in such a group. The final diagnosis will depend on additional studies other than the CT. They are exemplified by the heterogeneity of our series, which used CT images as the selection criteria.

According to this analysis, the nature of the hypodense extracerebral area will vary, with several alternatives being equally possible: empty space, developmentally large subarachnoid space, ex-vacuo large subarachnoid space, and hypertensive CSF accumulation due to absorption or flow impairment. Our subdural taps and flow studies point in this direction. This condition in any of its forms (normal variant, alternatives 1, 2 and 3) may favor brain displacement as well as brain and skull distortion, therefore predisposing to the development of subdural hematomas, as suggested by Aoki [3].

Finally, we are convinced that the term EH, although not absolutely wrong in all circumstances, is not proven in most and is incorrect in some. In addition, it has many implications, such as the possibility of increased intracranial pressure or the need for surgical treatment, which are not warranted when not verified, as is the case when CT criteria alone are used. We would like to recommend the restriction of its use to only circumstances in which enlarged subarachnoid spaces are clearly demonstrated (surgery, metrizamide or isotope cisternography) and CSF flow or absorption impairment, as well as intracranial pressure, are documented by the appropriate methods. Correct use of the term is of more than pure academic interest.

Often the parents, the referring pediatrician, and the neurologist and neurosurgeon are faced with a situation in which this EH diagnosis, based only on a CT and loosely used, may either lead to an unwarranted optimistic prognosis or a more dangerous and aggressive approach, including surgery. In both cases, the important underlying pathology will be missed. We feel that a less committal term should act as a trigger for a more careful and judicious evaluation of such cases. This evaluation should start with the acknowledgement and incorporation of the concept of "brain fluid," with all its implications, as expressed by Raimondi [26] in his thorough analysis of hydrocephalus.

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References

1. Alvarez LA, Maytal J, Shinar S (1986) Idiopathic external hydrocephalus: natural history and relationship to benign familial macrocephaly. Pediatrics 77:901–907
2. Andersson H, Elfverson J, Svendsen P (1984) External hydrocephalus in infants. Child's Brain 11:398–402
3. Aoki N, Masuzawa H (1984) Infantile acute subdural hematoma. Clinical analysis of 26 cases. J Neurosurg 61:273–280
4. Bode H, Strassburg HM (1987) Craniocerebral disproportion. A contribution to the significance of extracerebral fluid collections in infancy. Klin Pädiatr 199:399–402
5. Briner S, Bodensteiner J (1980) Benign subdural collections of infancy. Pediatrics 67:802–804
6. Carnell S, Chiu LC, Christie JH (1978) Diagnosis of extracerebral fluid collections by computed tomography. Am J Roentgenol 131:107–110
7. Dandy WE (1918) Extirpation of the choroid plexus of the lateral ventricles in communicating hydrocephalus. Ann Surg 68:569–579
8. Dandy WE, Blackfan KD (1914) Internal hydrocephalus: an experimental clinical and pathological study. Am J Dis Child 8:406–482
9. Enzman DR, Lane B (1978) Enlargement of the subarachnoid spaces and lateral ventricles in patients undergoing chemotherapy. J Pediatr 92:535–539
10. Frankenburg WK, Dodds JB (1967) The Denver Development Screening Test. J Pediatr 71:181–191
11. Gooskens RHJM, Willemse J, Gielen CCAM (1985) Cerebrospinal fluids dynamics and cerebrospinal fluid infusion in children. II. Clinical application of lumbar cerebrospinal fluid infusion in children with macrocephaly and normal growth rate of the head circumference. Neuropediatrics 16:121–125
12. Gooskens RHJM, Gielen CCAM, Willemse J (1988) The value of estimating pressure-volume index in childhood macrocephaly. Child's Nerv Syst 4:233–236
13. Gordon N (1980) Apparent cerebral atrophy in patients on treatment with steroids. Dev Med Child Neurol 22:502–506
14. Kapila A, Trice J, Spies WG, Siegel BA, Gado MH (1982) Enlarged cerebrospinal fluid spaces in infants with subdural hematomas. Radiology 142:669–672
15. Kendall B, Hollan I (1981) Benign communicating hydrocephalus in children. Neuroradiology 21:93–96
16. Kleinman PK, Zito JL, Davidson RI, Raptopoulos V (1983) The subarachnoid space in children: normal variations in size. Radiology 147:455–457
17. Marks HG, Borns P, Steg NL, Stine SB, Stroud HH, Vates SS (1978) Catch-up growth demonstration by CAT scan. J Pediatr 93:254–256
18. Maytal J, Alvarez LA, Elkin CM, Shinnar S (1987) External hydrocephalus: radiologic spectrum and differentiation from cerebral atrophy. AJNR 8:271–278
19. Ment LR, Duncan CC, Geeghr R (1981) Benign enlargement of the subarachnoid space in the infant. J Neurosurg 54:504–508
20. Modic MT, Kafmann B, Bonstelle CT, Tomsick T, Weinstein MA (1981) Megalocephaly and hypodense extracerebral fluid collections. Radiology 141:93–100
21. Mori K, Handa H, Itoh M, Okuno T (1980) Benign subdural effusion in infants. J Comp Assist Tomogr 4:466–471
22. Mulke R (1972) Nervale Gesichtspunkte zur Pathogenese und Therapie der Kraniosynostose. Acta Neurochir (Wien) 26:191–250, 293–326
23. Myriantopoulos NC (1977) Epidemiology of central nervous system malformations. In: Vinken PJ, Bruyn GW (eds) Handbook of clinical neurology, vol 30. Elsevier, Amsterdam New York Oxford, pp 139–172
24. Nickel RE, Gallenstein JS (1987) Developmental prognosis for infants with benign enlargement of the subarachnoid spaces. Dev Med Child Neurol 29:181–186
25. Petit 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
26. Raimondi AJ (1987) Pediatric neurosurgery. Theoretic principles, art of surgical techniques. Springer, New York Berlin Heidelberg, pp 453–491
27. Robertson WC, Gomez MR (1978) External hydrocephalus: early finding in communicating hydrocephalus. Arch Neurol 35:541–544
28. Robertson WC, Chun RWN, Orrison W, Sackett JF (1979) Benign subdural collection of infancy. J Pediatr 94:382–385
29. Sahar A (1978) Pseudohydrocephalus-megalocephaly, increased intracranial pressure and widened subarachnoid space. Neuropädiatrie 9:131–139
30. Velardi F, Hoffman HJ, Ash JM, Hendrick B, Humphreys RP (1986) The value of CSF flow studies in infants with communicating hydrocephalus. Child's Nerv Syst 2:139–143