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Original Article

Clinical, Radiological, and Demographic Details of Benign External Hydrocephalus: A Population-Based Study

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ABSTRACT

Background: Benign external hydrocephalus has an incidence of about 0.4 per 1000 live births. It affects infants and is characterized by an increasing head circumference and typical neuroimaging findings. Previously published studies on benign external hydrocephalus often contain groups of few and selected patients.

Methods: This is a follow-up of a recently published article reporting the incidence of benign external hydrocephalus. This retrospective and population-based study covers two large health regions in Norway, over a 10-year period (1994 to 2003). Infants with increasing head circumference, combined with typical radiological findings of enlarged subarachnoid spaces, were included. Information about head circumference development, neuroimaging findings, and birth delivery methods, as well as demographic details, was retrieved from the hospital medical records.

Results: A total of 176 children with benign external hydrocephalus were included, 86.4% being boys. At birth, the head circumference was close to normal. Mean age for when the head circumference reached abnormal values, i.e., crossing two percentiles or reaching the 97.5 percentile, was 3.4 months; none was older than seven months. Around four of five children had dilated lateral ventricles in addition to enlarged subarachnoid spaces. The neuroimaging findings tended to normalize after age 12 months. About half of the patients ended up with head circumferences at or above the 97.5 percentile.

Conclusions: Most infants with benign external hydrocephalus are born with a normal head circumference that increases too fast and reaches abnormally high values before age six months. This age and gender distribution is very similar to that described for infant subdural hemorrhage.

Background

Benign external hydrocephalus (BEH) is a relatively common pediatric condition with an estimated incidence of about 0.4 per 1000 live births and with a marked male preponderance.[1] This condition occurs during infancy and is characterized by a rapidly increasing head circumference (HC) combined with typical neuroimaging findings of increased subarachnoid cerebrospinal fluid spaces—especially overlying the frontal lobes—and normal or enlarged ventricles.[2-7] For an extensive review of the condition, see Zahl et al.[2]

Many other terms have been used for this or similar conditions, such as "subdural effusion,"[8] "subdural hygroma,"[9] "extraventricular obstructive hydrocephalus,"[10] "benign subdural collections,"[11] "benign enlargement of the subarachnoid spaces,"[12,13] "primitive megalencephaly,"[14] and macrocephaly.[15] The condition is referred to as BEH in the following discussion.

The main sign leading to medical attention is increasing HC, although other symptoms and signs have been reported, such as a tense anterior fontanel,[16,17] dilated scalp veins,[3] irritability,[6,18] gross motor delay,[12,19] and seizures.[20,21]

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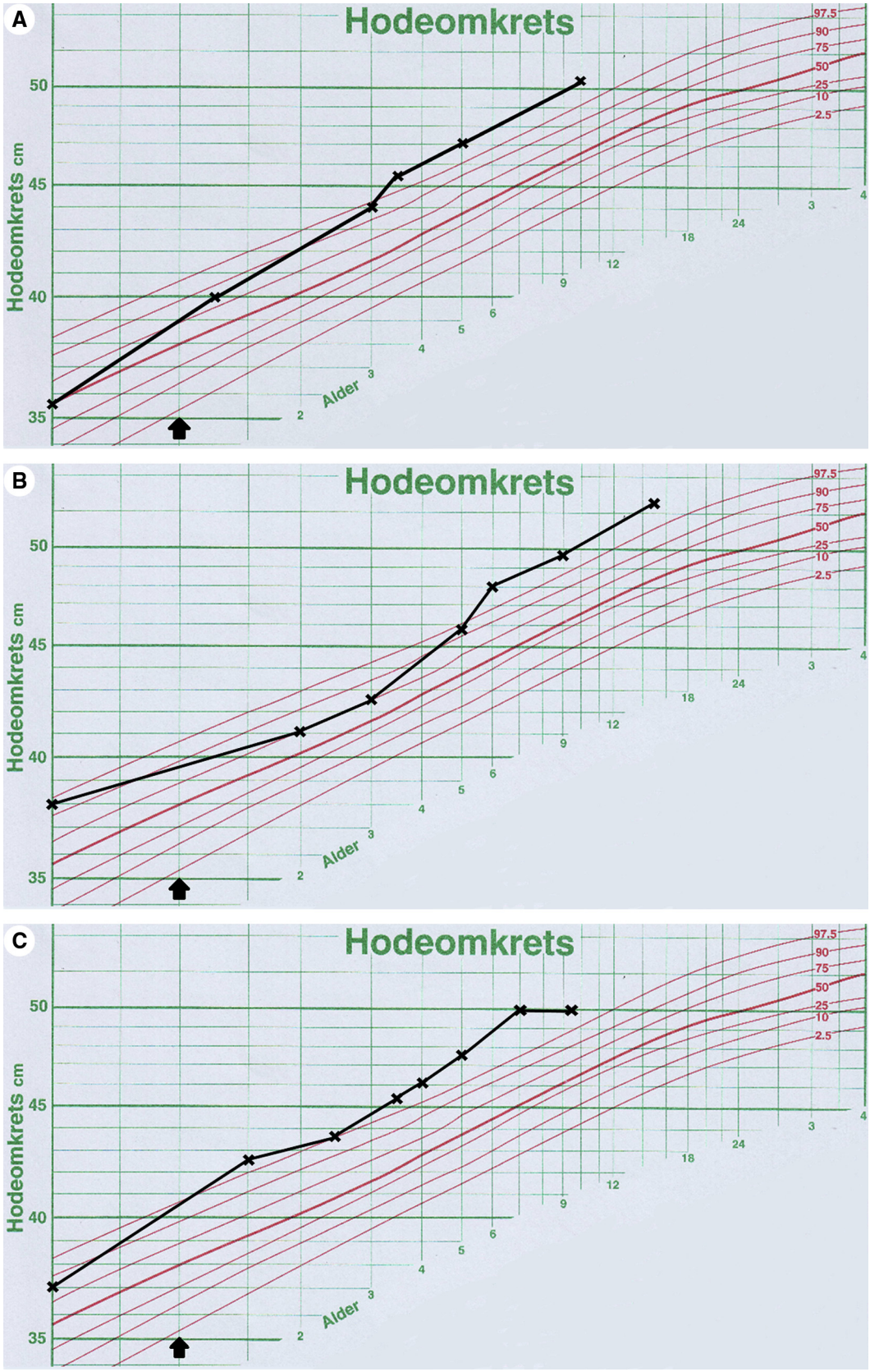

FIGURE 1. Norwegian head circumference registration sheets used at the time of the study. *Hodeomkrets* = head circumference, shown in centimeters along the *Y* axis. The *X* axis shows the age in months. Black arrows mark the age one month. (A) An infant boy with gradually increasing HC after birth, with rapid growth around age three months, thereafter stabilizing at a high percentile. (B) This boy had a fairly late HC growth spurt, most rapidly around the age five to six months. Of notice is also a significant decrease in HC in the first two months, possibly due to a temporary head swelling after birth. (C) Infant boy referred at age 5.5 months. The HC chart shows rapid growth even at an early age (before two months). The color version of this figure is available in the online edition.

In addition to reporting the incidence of BEH, our previous population-based study showed that the HC was close to normal at birth and that increased or enlarging HC was the main cause for referral.[1] We also found that approximately one-fifth of the children had normal ventricles on neuroimaging, whereas the remainder had increased ventricular size at diagnosis.

The aim of this study is to explore in detail when the head growth becomes abnormal, by collecting information from medical journals and HC growth charts. In addition, we report some information about radiological outcome.

Methods

This is a retrospective and population-based study, covering two well-defined health care regions in Norway with a mean total population of 3.34 million during the 10-year study period from 1994 to 2003; this constitutes about 75% of Norway's mean population during this period. Norway has a regionalized public health care system; within this system only two regional neurosurgical departments (in Oslo and Bergen) dealt with all pediatric neurosurgical conditions in these two regions. Medical records at these centers were searched for relevant hydrocephalus diagnoses. Information about age, gender, symptoms, clinical signs, neuroimaging, and HC development were collected for each patient. The radiological data are based on the radiologists' original reports; hence this is not a retrospective imaging study.

Inclusion criteria included HC above the 97.5th percentile or an HC crossing two or more percentiles during the first year of life, together with neuroimaging findings typical of BEH. Children with histories of head trauma, intracranial hemorrhage, central nervous system infection, other known causes of hydrocephalus, or prematurity (born before 37 weeks' gestation) were all excluded.

For further information about the study and selection methods, see Wiig et al.[1]

The study was approved by the Regional Committee for Medical Research Ethics.

Results

A total of 176 children (152 boys and 24 girls) matched the inclusion criteria for BEH during the study period; 44 (25 %) of the children required a ventriculoperitoneal shunt.[1]

Onset of sign was defined as the age at which the infant's HC curve crossed two percentiles or exceeded the 97.5 percentile. Detailed data for HC development were available for 107 children. Figure 1 shows the HC registration sheets for three infants.

Mean age of sign onset was 3.4 months (median 3.0 months, range 0 to 7.0 months). Mean age of onset for girls was 2.9 months ($n = 14$), and for boys, 3.5 months ($n = 93$). None of the 107 infants had sign debut after age seven months (Fig 2). The mean age for referral to our hospitals was 7.3 months, and the main reason for health service contact was a large or increasing HC.[1]

Twenty-one children (11.9 %) were delivered by Caesarean section, and two children had assisted deliveries (forceps and vacuum extraction). Thirteen children (7.4 %) were twins, but in no cases did the other twin develop BEH. Most of the twins (i.e., 11 of 13) were boys; unfortunately, we do not know the gender of the sibling twins.

For 28 of the 176 patients we had no information regarding the development of HC or radiology apart from what was found at the first hospital consultation. For the remaining children follow-up information existed to a varying degree.

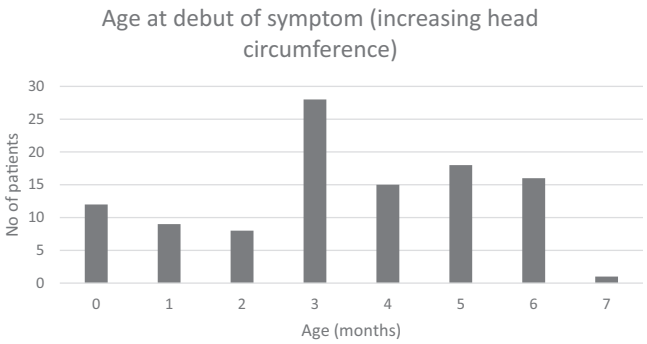

FIGURE 2. Bar graph showing sign onset, i.e., when the infants' HC became abnormally large. Onset was defined as the age at which the infant's HC crossed two percentiles or exceeded the 97.5 percentile. No patient had sign onset after age seven months. Please note that a small fraction of the infants showed an increased HC at birth or shortly thereafter. Detailed information about HC development was available for 107 of the patients ($n = 107$).

Neuroimaging: follow-up

Detailed information about radiology findings before age 12 months existed for 123 of the patients. They were all found to have excessive extracerebral fluid; 100 of these also had dilated ventricles (exact size not described). Figure 3 shows the magnetic resonance imaging of one of our patients with BEH.

For 77 infants we found reports on neuroimaging follow-up beyond age 12 months. In 57 of these, increased subarachnoid cerebrospinal fluid spaces were persisting, and 47 also had dilated lateral ventricles. Eight children had dilated lateral ventricles only, and in 12 patients the intracranial radiological findings were described as normal. Mean age at the final radiological examination for those who were followed-up beyond age 12 months was 21.7 months (median 18 months, range 12 to 104 months).

For 66 children we had information about neuroimaging both before and after age 12 months. Figure 4 gives a flow chart showing the development of radiological findings in these patients. As we

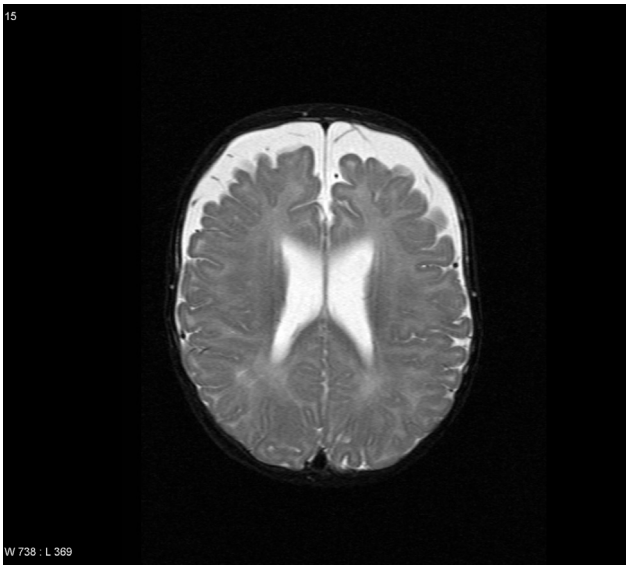

FIGURE 3. Magnetic resonance image of a 6.5-month-old boy (the same as in Fig 1A), referred because of increasing head circumference. The image shows enlarged frontal subarachnoid spaces, moderately increased lateral ventricles, and a widened frontal interhemispheric fissure.

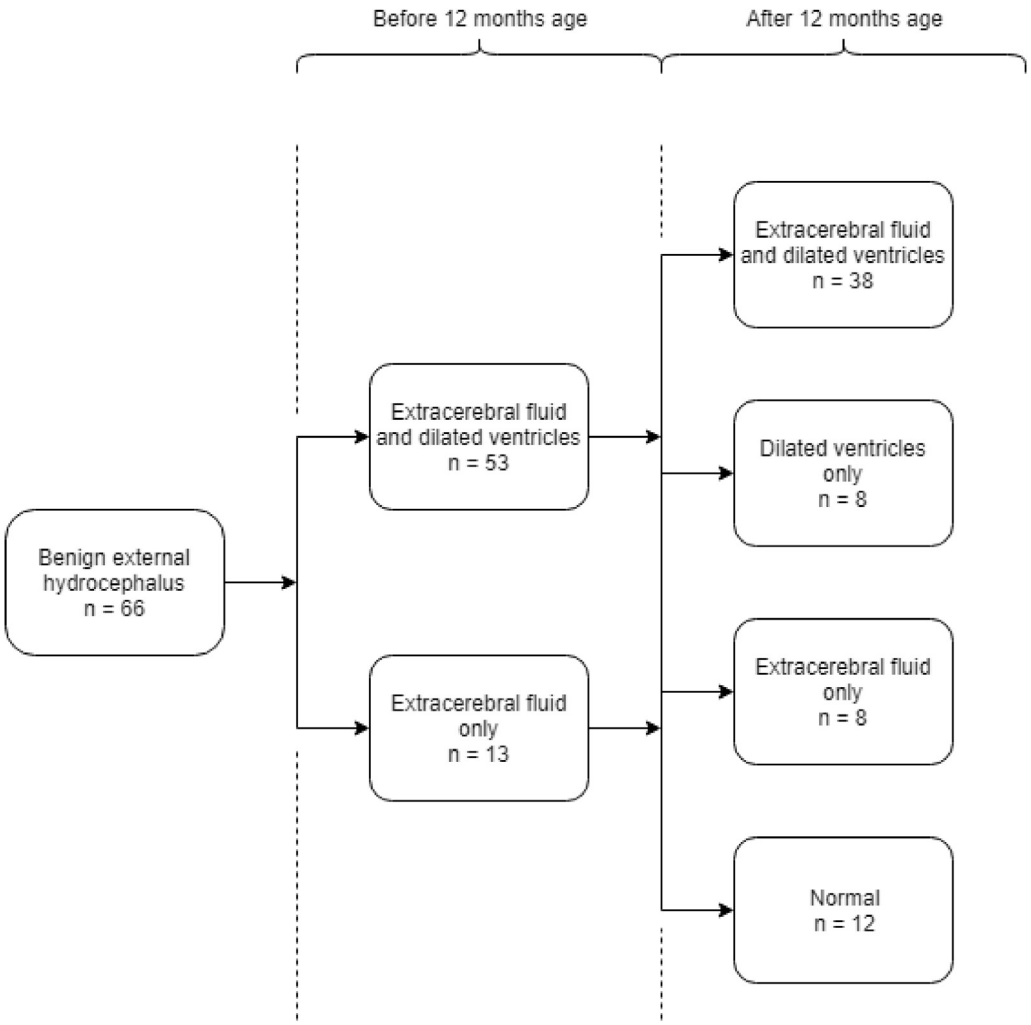

FIGURE 4. Flowchart that groups infants with neuroimaging both before and after age 12 months, according to the original radiologist's analysis. Number of patients mentioned below the description.

did not have access to the actual images—only the radiologists' interpretations—no exact numbers of size changes are available.

Head circumference: follow-up

For 106 children we had information about the HC development after age 12 months. Mean age at the final HC measurement for these children was 28 months. Of these children, 55 (52%) had a final HC at or above the 97.5 percentile. The mean age for the final measurement for these infants was 26 months. For the majority of these 55 children, the HC percentile stayed the same or decreased, whereas for 13 (24 %) the HC percentile continued to increase after age 12 months.

Discussion

This study is a follow-up and extension of our article from 2017,[1] based on the same population and study group.

The majority of patients had dilated lateral ventricles in addition to excessive extracerebral fluid on neuroimaging. This corresponds with earlier publications. Although we do not know when the excessive fluid begins to accumulate in each patient, it appears reasonable to assume that this coincides with the HC increase, i.e., sometime before age six months. For the long-term radiology, our results suggest that the excessive extracerebral fluid disappears earlier than the excessive intraventricular fluid.

In our selection of patients, we chose to exclude patients born prematurely or with a diagnosis of intracranial hemorrhage. We believe this was a mistake, as prematurity has been showed to be associated with BEH[22] and BEH is known to be complicated by subdural hematoma (SDH).[23,24] Thus these exclusions probably deprived us of valuable information about BEH in these groups of infants. Exact numbers for the excluded patients are not available.

Our results show that at least half of the patients will end up with large heads beyond infancy. We know from our previous study that HC at birth was close to normal.[1] As shown in Fig 2, the increase in HC typically occurs during the first six months of life, with a mean age of 3.4 months for sign onset. The mean age at referral for investigation by specialist was 7.3 months.[1] Thus the mean interval from sign debut to specialized medical evaluation was about four months.

This age distribution of BEH is strikingly similar to the distribution of infant SDH, which has a peak incidence during the first few months of life.[25-27] This also seems to be the case with the male preponderance (86.4% boys), a gender distribution similar to that of infant SDH.[25-28] These similarities in age and gender distribution may not be coincidental, as BEH is a known risk factor for developing SDH.[13,29,30] It is also somewhat intriguing that infants

diagnosed with abusive head trauma show an almost identical age and gender distribution to the one found for BEH.[31,32]

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