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Original Article

# Epidemiology of Benign External Hydrocephalus in Norway—A Population-Based Study



Ulrikke S. Wiig MD<sup>a</sup>, Sverre M. Zahl MD<sup>b,c</sup>, Arild Egge MD, PhD<sup>a</sup>, Eirik Helseth MD, PhD<sup>a,d</sup>, Knut Wester MD, PhD<sup>c,e,\*</sup>

- <sup>a</sup> Department of Neurosurgery, Oslo University Hospital, Oslo, Norway
- <sup>b</sup> Department of Ear, Nose and Throat, Aalesund Hospital, Aalesund, Norway
- <sup>c</sup> Department of Clinical Medicine K1, University of Bergen, Bergen, Norway
- <sup>d</sup> Faculty of Medicine, University of Oslo, Oslo, Norway
- <sup>e</sup> Department of Neurosurgery, Haukeland University Hospital, Bergen, Norway

#### **ABSTRACT**

**BACKGROUND:** Benign external hydrocephalus is defined as a rapidly increasing head circumference (occipitofrontal circumference) with characteristic radiological findings of increased subarachnoid cerebrospinal fluid spaces on neuroimaging. The incidence of benign external hydrocephalus has not been previously reported, and there is no available information on the ratio of benign external hydrocephalus in the population of hydrocephalic children. METHODS: This study is retrospective and population-based study, geographically covering two health regions in the southern half of Norway with a total mean population of 3.34 million in the ten-year study period, constituting approximately 75% of the Norwegian population. Children with a head circumference crossing two percentiles, or greater than the 97.5th percentile, and with typical imaging findings of enlarged frontal subarachnoid spaces with or without enlarged ventricles were included. Children were excluded if they had a history of head trauma, intracranial hemorrhage, central nervous system infection, other known causes of hydrocephalus, or were born preterm defined as birth before 37 weeks of gestation. RESULTS: A total of 176 children fitting the criteria were identified, giving an incidence of 0.4 per 1000 live births. One hundred fifty-two (86.4%) of the patients were male, and mean age at referral was 7.3 months. Increasing head circumference was the main reason for referral in 158 (89.8%) patients and the only finding in 60 (34.1%) patients. Thirty-seven (21%) children had normal ventricles on imaging; the remainder had increased ventricular size. The incidence of pediatric hydrocephalus in Norway is reported to be 0.75 per 1000 live births, thus benign external hydrocephalus accounts for approximately 50% of hydrocephalic conditions in this population. CONCLUSIONS: The incidence of benign external hydrocephalus was found to be 0.4 per 1000 live births in this population.

**Keywords:** benign external hydrocephalus, incidence, hydrocephalus, epidemiology, head circumference, macrocephaly Pediatr Neurol 2017; 73: 36-41

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#### Introduction

Hydrocephalus is a relatively common neuropediatric condition; the incidence is reported internationally as 0.36 to 0.75 per 1000 live births. In Norway the incidence has

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\* Communications should be addressed to: Dr. Wester; Department of Neurosurgery; Haukeland University Hospital; N 5021 Bergen, Norway. *E-mail address*: kgwe@helse-bergen.no

been found to be 0.75 per 1000 live births.<sup>1</sup> The most up-to-date definition of hydrocephalus was agreed upon internationally in 2010 and states that "Hydrocephalus is a condition characterised by a dynamic imbalance between the formation (production) and absorption of spinal fluid that results in an increase in the size of the fluid cavities within the brain and, in some situations, in an expansion of the spaces outside the brain, with or without an increase in the size of the ventricles."<sup>2</sup>

Benign external hydrocephalus (BEH) is a subgroup of hydrocephalus, which mainly occurs during infancy. It is defined as a rapid increase in head circumference (HC), measured as occipitofrontal circumference (OFC), combined with enlarged, usually frontal, subarachnoid cerebrospinal fluid spaces on neuroimaging and normal or only moderately enlarged ventricular system.<sup>3,4</sup> For a review of the condition, see Zahl et al.<sup>5</sup> A rapidly increasing HC or a large head a are most commonly what brings the infants to medical attention. Frontal bossing, dilated scalp veins, and a tense fontanel have also been described, as well as irritability, hypotonia, and developmental delay, most commonly gross motor delay; language delay is also seen. The developmental delay and hypotonia have been found to be generally transient, usually normalizing over a period of one to four years.<sup>4-7</sup> Neuroimaging findings generally also normalize over a few years. The disease has been regarded as benign and selflimiting and is rarely treated.<sup>9-1</sup>

The incidence of BEH has not been previously reported, and there is no available information on the ratio of BEH in the population of hydrocephalic children. We aim to determine the incidence of BEH in the general pediatric population. We will also discuss clinical and neuroimaging findings in the BEH population.

#### **Materials and Methods**

This study is a retrospective and population-based study, geographically covering two healthcare regions in the southern half of Norway with a total mean population of 3.34 million in the 10-year study period, constituting approximately 75% of Norway's mean population of 4.44 million during the same period. The annual average of live births in the health regions during the study period was 44,225. <sup>13</sup>

Norway is a sparsely populated country with a public three-level hierarchical hospital structure, with local community hospitals as the primary referral centers. Most counties have a central hospital with a pediatric department as a secondary referral center. At the top, there are four university clinics with a neurosurgical department, each serving a geographically well-defined health region consisting of several counties.

Within the Norwegian medicolegal system infants have to be seen at regular intervals at an outpatient mother-and-child health center. Instructions with the legal authority of law are given by the Norwegian health authorities; these regulate the activities of the health centers. Consequently, it is mandatory for the parents to bring the child to the local health center at certain intervals. Norwegian recommendations are that the HC should be measured routinely at each regular visit to the health center during the first year of life. According to these instructions, all children with a rapidly increasing HC should be referred to a specialist; for all practical purposes, all these children end up being referred to and evaluated by the collaborating pediatric and neurosurgical departments in the regional hospital.

Rapidly increasing HC is defined as crossing two percentile curves on the HC registration sheet, which is based on Norwegian reference values.

Diagnosis and treatment of the pediatric population in our two regions were undertaken in the two regional neurosurgical departments, Oslo University Hospital (Rikshospitalet) and Haukeland University Hospital in Bergen. These two departments were responsible for the pediatric neurosurgical service in the South-Eastern and Western regions, respectively. Medical records at the two centers were searched for relevant hydrocephalus diagnoses in the 10-year period from January 1, 1994 to December 31, 2003.

From the medical records information about age, gender, clinical symptoms and signs, HC, and neuroimaging findings were recorded for each patient.

Inclusion criteria included OFC crossing two percentiles or more, or OFC greater than 97.5th percentile in the first year of life, and typical neuroimaging findings. Children diagnosed after one year, but where diagnostic clinical information from primary care existed before age 12 months, were also included in the study population (seven children,

4%). All the included children had been examined with neuroimaging modalities allowing measurement of the subarachnoid/subdural space. For most children who were referred from a lower level institution, the neuroimaging was attached the referral documents and merged with the regional hospital's files.

Children were excluded if any of the following were identified: history of head trauma, intracranial hemorrhage, central nervous system infection, other known causes of hydrocephalus, or prematurity defined as birth before 37 weeks of gestation.

The project was approved by the Regional Ethics Committee, the Norwegian Social Science Data Service, and the Norwegian Directorate of Health.

#### Results

Overall epidemiological results

A total of 176 children with BEH were identified in the 10-year period in the two regions. This finding gives an incidence of 0.4 per 1000 live births (95% confidence interval, 0.34 to 0.46).<sup>13</sup> The incidence of pediatric hydrocephalus in Norway during the approximate same period was 0.75 per 1000 live births.<sup>1</sup> Thus the incidence of BEH is approximately half that of all primary hydrocephalus in a pediatric setting.

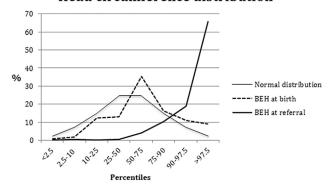
At birth, the patients had a slightly larger HC than in the normal distribution (Figure). At referral, this deviation was naturally much more marked, with most patients having an HC greater than the 97.5th percentile.

There was a marked male preponderance in the BEH population; 152 (86.4%) were boys. The corresponding figure for all hydrocephalic children is 74% in the reasonably matched population of Zahl et al. Approximately 51% of live births in Norway are boys. <sup>13</sup>

Symptoms and clinical findings

The mean age at referral for investigation by specialist care was 7.3 months (range 1.5 to 23 months, median 7 months). There was no difference in referral age between genders. The main reason for contact with the health service was a large and/or rapidly increasing OFC detected during the routine measurements at the public health

### Head circumference distribution



# FIGURE.

The graph shows that the head circumference (HC) for the study population did not deviate much from the normal distribution at birth and that the HC increase had occurred between birth and referral (mean age 7.3 months), BEH, benign external hydrocephalus.

clinics (158 patients, 89.8%). Another three patients had increasing OFC listed as an additional finding; thus a total of 91.5% of children were referred with increasing HC as one of the findings. In 68 (38.6%) children a large and/or rapidly increasing HC was the only finding at referral. Other symptoms and clinical findings are listed in Table 1. There was no gender difference with regards to symptoms or signs. Twenty-eight (15.9%) children had one or more findings that could be related to increased intracranial pressure (ICP) (sunset gaze, vomiting, lethargy, irritability, bulging/tense fontanels, and/or splaying of sutures).

#### **Neuroimaging**

As increased subarachnoid space was used as a diagnostic criterion, this was present in all patients. There was no grading used by reporting neuroradiologists.

A total of 37 (21%) children had normal ventricles according to reporting neuroradiologists; the remainder had some degree of ventricular enlargement. The degree of ventricular enlargement was subjectively graded as mild (39.6%) or moderate (11.5%), or was simply stated to be increased without any attempts at grading (48.9%). There was no statistically significant difference when clinical findings were compared with ventricular enlargement (Table 2).

#### Treatment of BEH

In total, 49 (27.8%) of the children with BEH were treated surgically. Ventriculoperitoneal shunting was the most common surgical procedure in 44 (89.8%) patients. Other treatment options were endoscopic third ventriculocisternostomy in three patients, whereas one patient each was treated with subduroperitoneal or

**TABLE 1.**Symptoms and Signs at Referral

Symptoms and signs*	Number (176)	%
Increased occipitofrontal	161	91.5
circumferenc <sup>†</sup>		
Frontal bossing	37	21
Delayed development	31	17.6
Distended veins	29	16.5
Large head <sup>†</sup>	20	11.4
Abnormal/asymmetric head shape	19	10.8
Large/bulging fontanel/suture	16	9.1
diastasis		
Hypotonia/head lag	13	7.4
Sunset gaze	10	5.7
Vomiting/retching	7	4
Other eye signs <sup>‡</sup>	6	3.4
Crying	5	2.8
Hypertonia/hyperreflexia	4	2.3
Seizures/seizure-like activity	3	1.7
Torticollis	2	1.1
Lethargy	2	1.1
Irritability	2	1.1
Reflux	1	0.6
Poor weight gain	1	0.6
Dysmorphic facial features	1	0.6

<sup>\*</sup> More than one per child.

**TABLE 2.**Ventricular Size and Clinical Findings at Referral

Ventricle Size	Increased intracranial pressure*	Other	None <sup>†</sup>	Sum
Normal	6 (16.2%)	18 (48.6%)	13 (35.1%)	37
Enlarged	22 (15.8%)	62 (44.6%)	55 (39.6%)	139
All patients	28 (15.9%)	80 (45.5%)	68 (38.6%)	176

<sup>\*</sup> Includes sunset gaze, vomiting, lethargy, irritability, bulging/tense fontanels, and splaying of sutures.

lumboperitoneal shunts. Of the 28 children who had one or more signs or symptoms that could be related to increased ICP, 14 children were surgically treated. This group of surgically treated patients constitute 28.6% of the 49 treated patients; thus 71.4% of the treated patients had no symptoms or signs of raised ICP. A slightly lower proportion of the surgically treated children had normal ventricular size (18.4%); however, this is not statistically significant (P = 0.69).

#### Discussion

In this population-based study, we found the incidence of BEH to be 0.4 per 1000 live births, constituting approximately 50% of all children diagnosed with hydrocephalus. The incidence of BEH in a population has not been previously reported. There are therefore no numbers for direct comparison with our findings. As will be discussed subsequently, establishing an exact, generally valid BEH incidence is hampered by several factors: methods for detecting infants at risk, as well as the differing clinical and radiological criteria from one study to another.

The patients in our study were identified through medical record searches in the two regional hospitals where children fitting the criteria of BEH should all be referred to following the national guidelines at local health centers. Thus we cannot be certain that some children may not have been identified or referred appropriately. However, with Norway having mandatory health center visits in infancy and firm guidelines for the referral of children fitting the criteria, we believe the numbers of missed children to be low, well within the range of the 95% confidence interval.

Previous attempts at establishing BEH incidence

Hamza et al.<sup>10</sup> found BEH in 13 of 81 patients (16%) with macrocrania in a group of children diagnosed with low-density fluid collections on computed tomography (CT). Kendall and Holland<sup>14</sup> also investigated CT images and found enlarged cerebrospinal fluid spaces of no known etiology in 14 of 500 CT image sets (2.8%). One retrospective review of incidental magnetic resonance imaging findings in a tertiary pediatric center found external hydrocephalus in 0.6% of imaged children. 15 This study and other studies 16-20 also include infants born prematurely (12% to 52%): in these studies, however, the definition of prematurity differs with a cutoff point of between 35 and 38 weeks of gestation. One study<sup>21</sup> excluded premature children, as we have done, but they did not include epidemiological data. Many publications do not mention whether preterm infants are included.<sup>6,11,17,22-24</sup>

<sup>†</sup> Subjective term from patient records.

<sup>‡</sup> Includes poor vision, nystagmus, strabismus, not fixing and following.

Increased occipitofrontal circumference and/or large head only.

One article<sup>16</sup> included only children with normal or only mildly dilated ventricles. Infants with signs of raised ICP,<sup>11,16,19</sup> abnormal neurological examination,<sup>25</sup> or developmental delay<sup>10,19</sup> are also sometimes excluded, which implies that the authors must have regarded BEH as a condition that cannot yield such symptoms. It also means that the studies cannot be easily and soundly compared.

#### Clinical criteria for the BEH detection and diagnosis

Mean referral age in our study is 7.3 months (range 1.5 to 23 months). This study compares well with other studies where referral age or age at diagnosis ranges from 6.5 to 8.9 months. 6,16,18,20 The age at which infants are diagnosed is determined by several factors. One such factor is the onset of clinical symptoms and signs. Many children present with an increase in HC, often with few, if any, other clinical findings. Early detection because of increased HC is probably facilitated by routine measurements. Thus one may expect such routines to influence the detection of the condition. The most commonly reported symptoms and signs apart from increased head OFC that lead to investigations and ultimately the diagnosis of BEH are seizures, <sup>26–28</sup> delayed psychomotor development, <sup>27,28</sup> and signs of increased ICP such as tense or large fontanels. <sup>27</sup>

Pediatric hydrocephalus is more common in the male population; this is even more so for the subgroup of hydrocephalic children with BEH. In various studies, the male preponderance ranges from 52% to 80%. 4.6,12,18-21 In our study, the gender distribution is even more skewed, with 86.4% boys. Study populations are, however, generally small compared with our study, some have less than 10 patients included, making direct comparison difficult. 6.7,29

From our results, we find that for those where OFC at birth was registered (153), the OFC was slightly higher than in the normal population at birth, with 71.9% having an HC greater than fiftieth percentile, 20.3% greater than the ninetieth percentile, and 9.2% greater than the 97.5th percentile. This finding compares well with Halevy et al. 16 who found an average at birth at the fifty-eighth percentile and the results of Hellbusch<sup>17</sup> who found that most had OFC between the fiftieth and the ninety-eighth percentiles. Laubscher et al. 18 found that 12 of 21 patients (57%) of their group with dilated pericerebral subarachnoid space had an HC of greater than the ninetieth percentile at birth. Thus there is a trend toward larger OFC at birth in children who later develop BEH, but still most of the children with BEH had HCs within the normal range at birth. At diagnosis the HC distribution had become much more skewered in the BEH population. In our study, 65.9% of children had an OFC greater than the 97.5th percentile (mean age 7.3 months). This finding compares well with 50% greater than the ninety-eighth percentile at a mean age seven months in Hellbusch's study and mean OFC at the 79.5th percentile at a mean age 5.8 months (Halevy et al.). The present study is the largest to date showing this relatively dramatic increase in HC from birth to diagnosis.

BEH can also be diagnosed due to excessive head growth alone, even if the OFC still is within the normal range. In Norway, rapidly increasing HC is defined as crossing at least two percentile curves on the national HC registration

charts.<sup>1</sup> There have also been published examples of BEH in microcephalic children.<sup>30</sup>

Throughout the world there are different percentile charts in use, our Norwegian population was studied using the growth charts that were introduced in the 1980s. 31,32 Many use the World Health Organization charts, which are based on data from Norway, Brazil, Ghana, India, Oman, and the United States. However, they have been shown to be at variance compared with national or regional OFC growth references. 33-36 As these studies have shown, the use of standard OFC charts that are not based on regional/national populations may cause variations in the registered incidence of hydrocephalus, including BEH.

In the studies of BEH, the cutoff value used in the diagnosis of macrocephaly varies from the ninetieth to the ninety-eighth percentile 4.10,14,17,18,25,37; in our study, we have used the 97.5th percentile as the cutoff point. As discussed previously, the use of different percentile charts and differing cutoff values will certainly have impact on the incidence of BEH in a population.

## Radiological criteria for the BEH diagnosis

In this study, the children were examined with ultrasound, CT, or magnetic resonance imaging. Many were investigated with more than one imaging modality. In most instances, the subarachnoid space is simply reported as increased by the neuroradiologist, with no exact measurements given. Several studies on the different imaging modalities have been done to evaluate what the normal range of subarachnoid space is in infants. The three most common measurements evaluated are sinocortical width (SCW), craniocortical width (CCW), and interhemispheric distance (IHD). SCW was introduced by Govaert et al.<sup>38</sup> and is defined as the shortest distance between the lateral wall of the triangular superior sagittal sinus and the surface of the adjacent cerebral cortex. The CCW is the shortest vertical distance between the calvarium and the surface of the cerebral cortex, whereas the IHD is defined as the widest horizontal IHD. Measurements are taken on coronal views, at the level of the foramen of Monro.<sup>39,40</sup> These distances vary with the infant's age with an increase in normal subarachnoid space during the first year of life, peaking at approximately seven months, with a gradual decrease thereafter. 40,41 Depending on the imaging modality chosen, the age of the child and the selection of study population with regards to OFC, the upper limit above which the CCW is likely to be abnormal, ranges from 4 mm to 10 mm. 4,23,39,41,42 The corresponding ranges for SCW are 2 mm to 10 mm and for IHD 6 mm to 8.5 mm. However, no validated normal values exist and thus the cutoff values may differ between radiologists. As the increased subarachnoid space is one of the diagnostic criteria for the diagnosis of BEH, this has implications for whether a child is diagnosed with BEH; thus the incidence in a population depends to some degree on the cutoff value used by radiologists. This lack of uniformity also applies to the present study, as the definitions of abnormal distances most probably varied between the describing radiologists.

Lateral ventricle size is generally defined as normal or only moderately enlarged in BEH. However, this definition, which is stated in many publications, does not seem to be supported by findings in those same publications where reported ventricle size ranges from normal to gross dilatation. The degree of dilatation of lateral ventricles has been found to be roughly proportional to the width of the frontal subarachnoid space.<sup>21</sup> Ventricular dilatation, when it occurs, also seems to be a later finding than enlarged subarachnoid spaces.<sup>43</sup>

# BEH versus idiopathic communicating hydrocephalus

BEH and other forms of idiopathic communicating hydrocephalus may very well be part of a spectrum of hydrocephalus, <sup>3,27</sup> and a clear distinction between the two is difficult to make in a retrospective study such as this one. One clear limitation of this study is that we cannot, at least in our group of treated children, be certain of this distinction between BEH and communicating hydrocephalus, as a proportion of the patients had signs and symptoms of raised ICP and/or enlarged ventricles. However, patients with communicating hydrocephalus are generally believed to be in need of surgical treatment, and most patients in our study with signs or symptoms of raised ICP and/or ventricular dilatation did not receive any surgical treatment.

The present study is the first to describe the incidence of BEH in a relatively large and well-defined population, with the limitations discussed previously. As most of the included children were detected by a mandatory regime of repeated routine HC measurements, we believe the figures reported here to be fairly representative.

#### **Conclusions**

BEH is the most common hydrocephalic condition in young children; it is also one of the least studied, which might be attributed to its assumed benign course. Our findings suggest that the incidence of BEH is approximately half the incidence of primary pediatric hydrocephalus in reasonably comparable populations. Because of the lack of studies of this condition, there is also no clear knowledge of diagnostic criteria or the correct treatment, if any. We suggest that the routine well-child clinic may help identify this group of patients. Any child found on routine follow-up to have a rapidly increasing OFC, or macrocephaly, should be referred on to the nearest pediatric department for clinical examination and imaging.

Data from other populations, and data including subgroups such as premature infants, would be helpful to validate our epidemiological findings in BEH.

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