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Original Article

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

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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

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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 been found to be 0.75 per 1000 live births.[1] 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."[2]

Benign external hydrocephalus (BEH) is a subgroup of hydrocephalus, which mainly occurs during infancy. It is

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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.[3,4] For a review of the condition, see Zahl et al.[5] 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.[4-7] Neuroimaging findings generally also normalize over a few years.[8] The disease has been regarded as benign and self-limiting and is rarely treated.[9-12]

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.[13]

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).[13] The incidence of pediatric hydrocephalus in Norway during the approximate same period was 0.75 per 1000 live births.[1] 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.[1] Approximately 51% of live births in Norway are boys.[13]

### *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

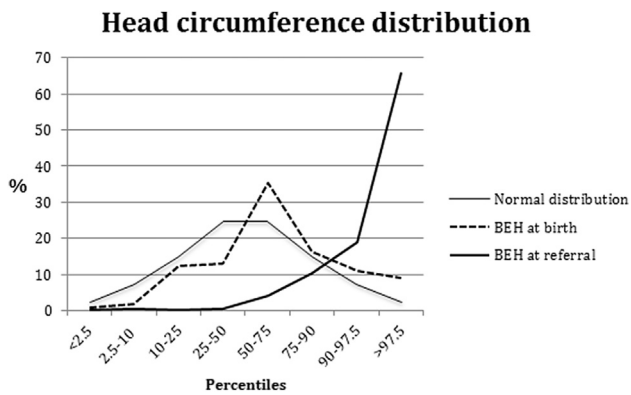


**FIGURE.**
The graph shows that the head circumference (HC) for the study population did not deviate much from the normal distribution at birth and that the HC increase had occurred between birth and referral (mean age 7.3 months). BEH, benign external hydrocephalus.