

Original Article

Subtypes and shunt treatment course of paediatric hydrocephalus

Anna Duc Jensen^{1*}, Marveh Dokhi^{1*}, Frederikke Guldberg Nielsen¹, Elsebet Østergaard^{2, 3}, Carl Larsen¹, Marianne Juhler⁴ & Tina Nørgaard Munch^{1, 3, 5}

1) Department of Neurosurgery, Copenhagen University Hospital – Rigshospitalet, 2) Department of Genetics, Copenhagen University Hospital – Rigshospitalet, 3) Department of Clinical Medicine, University of Copenhagen, 4) Department of Neurosurgery, Aarhus University Hospital, 5) Department of Congenital Disorders, Statens Serum Institut, Denmark

Dan Med J 2026;73(11):A03260256. doi: 10.61409/A03260256

Abstract

INTRODUCTION. This study aimed to describe aetiologies and concurrent conditions of paediatric hydrocephalus, assess short-term symptoms by clinical improvement after ventriculoperitoneal shunting and estimate one-year risks of shunt revision and complications.

METHODS. This **retrospective, single-centre** study included children who underwent shunt insertion for hydrocephalus between 1 December 2016 and 31 December 2021. Data on disease course, surgical treatment and comorbidities were extracted from electronic records.

RESULTS. Among 71 children, 42 (59%) had obstructive hydrocephalus and 29 (41%) had communicating hydrocephalus. Median ages at postnatal diagnosis were 1.7 and 0.3 years, respectively. Neurological conditions occurred in 29% versus 28%. Genetic findings were identified in 15 out of the 36 patients referred for genetic testing. Increased head circumference (56%) and signs of raised intracranial pressure (49%) were frequent. Ventriculoperitoneal shunt failure, measured as worsening or lack of clinical improvement, occurred in 31% with obstructive hydrocephalus and 24% with communicating hydrocephalus. The one-year risk of shunt revision was 30%. Among revised patients, the most frequent causes of first revision were valve or proximal catheter obstruction (37%), disconnection (18%) and shunt displacement (16%).

CONCLUSIONS. The aetiological complexity and shunt revision risk highlight the relevance of cross-disciplinary collaboration among paediatricians, neurosurgeons and clinical geneticists.

FUNDING. None.

TRIAL REGISTRATION. Not relevant.

In Denmark, hydrocephalus occurs in 1.08/1,000 live-born children [1] and is a complex, heterogeneous condition often accompanied by comorbidities. Hydrocephalus is defined as ventriculomegaly with disturbed cerebrospinal fluid (CSF) formation, flow or absorption, potentially causing life-threatening raised intracranial pressure (ICP) [2].

Paediatric hydrocephalus can arise from congenital or acquired cause. Hydrocephalus is classified as obstructive or communicating, depending on whether a structural obstruction to CSF flow is present [3]. Common congenital causes include aqueductal stenosis and spina bifida [2]. Acquired causes include post-infectious and severe post-haemorrhagic events, such as grade III-IV intraventricular haemorrhage.

Our understanding of the genetic factors that predispose individuals to hydrocephalus is rapidly evolving [4],

including knowledge of these factors in the Danish population [5]. Thus, data on comorbidities and genetic findings among patients referred for genetic testing are considered essential to understanding hydrocephalus heterogeneity. Ventriculoperitoneal shunting is the most common surgical treatment for paediatric hydrocephalus, and it has been shown to improve survival. However, a ventriculoperitoneal shunt carries risks of obstruction, infection and mechanical failure [2, 6], with a cumulative one-year risk of shunt revision reaching 50% in children under two years [7], according to a previous Danish nationwide cohort study. Short-term complications, including post-haemorrhagic hydrocephalus, occur in 25-36% of severe intraventricular haemorrhage cases in infants [8, 9], with 72.6-90% of those requiring permanent surgical CSF diversion with a ventriculoperitoneal shunt [9, 10].

Shunt revision may not fully reflect clinical benefit, as the absence of revision does not necessarily indicate symptom improvement [11]. Reporting both clinical improvement and revision risk therefore provides a broader assessment of shunt outcomes.

We aimed to describe aetiologies and concurrent conditions in paediatric hydrocephalus, short-term clinical improvement after ventriculoperitoneal shunting and one-year risks of shunt revision and complications.

Methods

We conducted a retrospective single-centre cohort study of children treated for hydrocephalus at a Danish tertiary neurosurgical centre.

Study population

The cohort comprised children aged 0-18 years with imaging-verified hydrocephalus who underwent primary ventriculoperitoneal shunt insertion at the University Hospital of Copenhagen between 1 December 2016 and 31 December 2021. The hospital covers the Region of Eastern Denmark. Patients were identified through hospital surgical records, cross-checked against electronic medical records. Children without available clinical information from diagnosis to 12 months after their primary shunt insertion were excluded.

Data collection

Based on neuroimaging, hydrocephalus was classified as either obstructive or communicating hydrocephalus and further grouped by underlying aetiology. Obstructive hydrocephalus was defined by a visible mechanical obstruction of CSF pathways, whereas communicating hydrocephalus had no evident focal obstruction. As some aetiologies may cause either type, each case was classified individually according to radiological findings before shunt insertion.

In multifactorial cases, radiological evidence of focal obstruction at the time of shunt insertion was weighted most strongly, followed by the presumed underlying aetiology and clinical context. The available preoperative CTs and/or MRIs, as well as the radiological reports, were reviewed retrospectively by the first author under supervision of a neurosurgical specialist. In the few cases of doubt, a neuroradiologist was consulted. Preoperative symptoms, comorbidities, genetic findings and details of surgical treatment were extracted systematically from electronic medical records.

Outcomes

Early clinical improvement and subsequent shunt revision were recorded. Short-term outcome was defined as clinical improvement during primary admission. Long-term outcomes comprised complications or shunt revision during at least one year of follow-up.

Statistical analysis

Data were analysed descriptively. Categorical variables were compared using the chi-squared test or Fisher's exact test, as appropriate. Odds ratios with 95% CI were calculated. A $p < 0.05$ was considered statistically significant.

Ethics approval and consent to participate

The project was approved by the relevant regional authorities and conducted in accordance with Danish data protection regulations (H-22040623).

Trial registration: not relevant.

Results

A total of 121 children were identified and screened for eligibility. Fifty children were excluded because sufficient clinical information from diagnosis to 12 months after primary shunt insertion was unavailable, leaving 71 children for analysis. Among these, 42 had obstructive hydrocephalus and 29 had communicating hydrocephalus. Baseline demographic and clinical characteristics are summarised in **Table 1**.

TABLE 1 Summary of patient characteristics, including sex, prenatal diagnosis, postnatal age at diagnosis, aetiology, preoperative symptoms and comorbidities. Data are presented separately for obstructive and communicating hydrocephalus.

	Obstructive hydrocephalus (N _o = 42)	Communicating hydrocephalus (N _c = 29)
Female sex, n (%)	20 (48)	13 (45)
Prenatal diagnosis, n (%)	10 (24)	6 (21)
Postnatal age at diagnosis, median (range), yrs ^a	1.7 (0-15.8)	0.3 (0-5.0)
<i>Aetiology, n (%)^b</i>		
Tumour	22 (52)	0
Aqueductal stenosis	8 (19)	0
Intracranial cyst	4 (10)	0
Dandy-Walker malformation	4 (10)	0
Chiari malformation	2 (5)	0
Perinatal intraventricular haemorrhage ^c	2 (5)	9 (31)
Idiopathic	0	14 (48)
Myelomeningocele	0	6 (21)
<i>Preoperative symptoms, n (%)^{b, d}</i>		
Increased head circumference	20 (48)	20 (69)
Symptoms of raised intracranial pressure	24 (57)	11 (38)
Delayed motor development	8 (19)	7 (24)
Delayed cognitive development	2 (5)	3 (10)
Hearing impairment	1 (2)	0
Visual impairment	6 (14)	2 (7)
Failure to thrive	5 (12)	3 (10)
<i>Comorbidities, n (%)^{b, d}</i>		
Neurological conditions	12 (29)	8 (28)
Congenital malformations, syndromes or genetic conditions	7 (17)	10 (34)
Chronic cardiac, pulmonary or renal conditions	4 (10)	3 (10)
ADHD	2 (5)	2 (7)
Autism spectrum disorder	1 (2)	2 (7)
Bone and connective tissue diseases	1 (2)	1 (3)

a) Calculated among the 32 and 23 patients diagnosed after birth, respectively.

b) Percentages were calculated within each hydrocephalus subtype and may not total 100% due to rounding.

c) May result in either obstructive or communicating hydrocephalus; classification was based on the available preoperative CT and/or MRI findings in each patient.

d) Patients may have had > 1 preoperative symptom or comorbidity.

Aetiology

Tumour and aqueductal stenosis were the leading causes of obstructive hydrocephalus, whereas idiopathic hydrocephalus and perinatal intraventricular haemorrhage dominated in communicating hydrocephalus. In tumour-related hydrocephalus, improvement was expected only in signs of raised ICP, not focal neurological deficits caused by the tumour.

Sex and age

A slight male predominance was observed in both subtypes. Prenatal diagnosis occurred in ten children (24%)

with obstructive hydrocephalus and in six children (21%) with communicating hydrocephalus. Among patients diagnosed postnatally, the median age at diagnosis was 1.7 years (range: 0-15.8 years) in obstructive hydrocephalus and 0.3 years (range: 0-5.0 years) in communicating hydrocephalus. Prenatal diagnosis occurred in most aetiological groups.

Preoperative symptoms

Increased head circumference was the most frequent preoperative finding overall. Signs of raised ICP were more frequent in obstructive hydrocephalus than in communicating hydrocephalus.

Comorbidities

Neurological

Neurological comorbidities, mainly epilepsy and cerebral palsy, affected one third of the patients.

Non-neurological comorbidities

We found that 20 patients (28%, ten from each subgroup) had chronic non-neurological comorbidities, including cardiac, pulmonary and renal conditions

Genetic predispositions for hydrocephalus

Among the 71 patients, 36 (51%) underwent genetic testing based on a clinical suspicion of a monogenic or chromosomal cause. Among these, 15 patients (42%) presented with one or two genetic findings. Variants comprised chromosomal abnormalities, copy number changes and single-gene variants.

Pathogenic variants that were assessed as explanations for hydrocephalus were identified in *COL4A1* (n = 2), *FGFR2* (Crouzon syndrome), *NF1*, *NF2* (mosaic variant), *PLG* and *PTEN*. Furthermore, trisomy 8 mosaicism was found in one patient and a 2 Mb deletion involving *ZIC2* on chromosome 13 was found in another patient. In six patients, it was uncertain if the genetic abnormality could explain hydrocephalus (Table 2).

TABLE 2 Types of genetic findings were identified in 15 of 36 patients who underwent genetic testing for clinical indications. The remaining 35 patients in the cohort were not tested.

Variant type	n (%)	Findings
Chromosomal abnormality/ copy number variation	2 (13)	Trisomy 8 mosaicism, 13q deletion including <i>ZIC2</i>
Single gene variant	7 (47)	<i>COL4A1</i> (2 patients), <i>FGFR2</i> , <i>NF1</i> , <i>NF2</i> (mosaic), <i>PLG</i> , <i>PTEN</i>
Abnormal findings, uncertain if it explains phenotype	6 (40)	<i>SHOX</i> deletion, <i>PDGFB</i> variant, 3.2 Mb deletion on chromosome 3, <i>G6PD</i> variant, <i>FLNC</i> variant, 45,X/46,XY mosaicism
Total	15 (100)	-

FGFR = fibroblast growth factor receptor; NF = neurofibromin; PLG = plasminogen; PTEN = phosphatase and tensin homologue.

Table 3 presents the proportion of children with persistent symptoms and/or a need for additional surgery during their initial admission. This occurred in 31.0% of patients with obstructive hydrocephalus and 24.1% of patients with communicating hydrocephalus.

TABLE 3 Numbers and percentages of patients with unresolved symptoms and/or repeated surgery within the same hospital admission. Results are presented for patients with obstructive and communicating hydrocephalus.

Ventriculoperitoneal shunt	Total, n (%)	Unresolved symptoms/ repeated surgery, n (%)	OR (95% CI)	p value
<i>Hydrocephalus</i>				
Obstructive	42 (59)	13 (31)	1.41 (0.47-4.24)	0.53
Communicating	29 (41)	7 (24)		
Total	71 (100)	20 (28)		

The odds ratio for unresolved symptoms in obstructive hydrocephalus compared with communicating hydrocephalus was 1.41 (95% CI: 0.47-4.24; $p = 0.53$).

Long-term risks of shunt revision

All patients were followed for at least one year after their initial shunt placement. Within the first year, 21 patients (30%) underwent shunt revision: most underwent one or two revisions, whereas four had three or more. Overall, 38 patients (54%) required at least one shunt revision during the entire period of follow-up.

Causes of shunt revision are summarised in **Table 4**. The most frequent cause was obstruction of the valve or proximal catheter (36.8%), followed by disconnection (18.4%) and shunt displacement (15.8%). Infection accounted for 13.2% and overdrainage for 10.5% of revisions. Due to the limited number of infection-related revisions, no meaningful comparison by age group could be performed. Patients with overdrainage were treated by conversion to higher pressure valves.

TABLE 4 The causes of initial ventriculoperitoneal shunt revisions in the 38 patients who underwent shunt revision during follow-up.

Cause of shunt dysfunction	n (%)
Obstruction of the valve or proximal catheter	14 (37)
Disconnection	7 (18)
Displacement	6 (16)
Infection	5 (13)
Overdrainage	4 (11)
Other	2 (5)
Total	38 (100)

Discussion

This retrospective regional cohort study of 71 consecutive patients undergoing ventriculoperitoneal shunt placement demonstrates the heterogeneous aetiology of paediatric hydrocephalus and frequent genetic findings, identified in 21% of the total cohort.

Comorbidities and genetic conditions

Comorbidities were common. Neurological disorders, especially epilepsy, were the most frequent comorbidities. However neurological disorders also encompassed tetraplegia, dyskinetic cerebral palsy, paresis, dystonia and cranial nerve palsies. The high frequency of neurological and neurodevelopmental comorbidities supports the view of hydrocephalus being associated with impaired neurodevelopment and molecular genetic mechanisms [4, 5, 12].

Genetic findings were identified in 15 of 36 genetically tested patients (42%). The findings were heterogeneous and included chromosomal abnormalities, copy number variants and single-gene variants, some of which were considered potentially causative, whereas others had uncertain phenotypic relevance. Genetic contributions may also occur in non-syndromic hydrocephalus [4, 13].

Comorbidity burden may influence clinical improvement and may partly explain why one third of patients did not experience short-term improvement in preoperative symptoms. However, the present study was not designed to determine causal relationships between hydrocephalus subtype, genetic findings, comorbidities and surgical outcomes, which therefore remain incompletely understood. Genetic findings should primarily be interpreted descriptively and as a part of the striking clinical heterogeneity of paediatric hydrocephalus.

Ventriculoperitoneal shunt complications

Ventriculoperitoneal shunt carries risks, including malfunction, migration, overdrainage, infection, CSF leakage and abdominal complications [6, 14, 15]. Our observed one-year shunt revision rate of 30% was within the range reported in previous paediatric studies [7, 16, 17]. In our series, the predominant causes of shunt failure were mechanical, most often valve or proximal catheter obstruction, possibly related to patient-specific CSF composition (e.g., high protein or cellular content).

Catheter obstruction and overdrainage together accounted for approximately half (47%) of the shunt failures in our cohort. However, the present study did not have sufficient statistical power to investigate potential associations between specific treatment choices such as valve type, outlet pressure or other tailored shunt strategies, and no conclusions regarding optimal individualised management can therefore be drawn. Infection represents one of the leading and most costly causes of shunt failure [16-18] and has been associated with substantially higher costs than shunt obstruction [19]. Furthermore, younger age has previously been associated with increased infection risk [20]. The limited number of infections precluded reliable analysis of age-related infection risk, although younger age may represent a risk factor.

Our 2016-2021 data show lower one-year revision rates than the data previously published in a nationwide, register-based Danish cohort study based on 1996-2015 data, where 50% of patients underwent shunt revision within the first year after shunt placement. Since 2018, our institution has implemented antibiotic-impregnated shunt systems as standard practice, and the widely used adjustable shunts may have contributed to the lower observed revision rate.

Limitations and strengths

The study aimed to descriptively characterise a regional paediatric hydrocephalus cohort but did not have sufficient statistical power to evaluate treatment strategies or shunt configurations. The study therefore does not

propose a new surgical approach but documents the clinical and genetic heterogeneity of paediatric hydrocephalus within a Danish regional cohort. Distinguishing obstructive from communicating hydrocephalus may pose a challenge, particularly in cases such as myelomeningocele and perinatal intraventricular haemorrhage, where mixed CSF flow alterations may occur. CT and/or MRI were not acquired according to a standardised MRI protocol. As advanced MRI sequences allow for detailed assessment of aqueductal morphology and patency, variation in imaging modality may have introduced classification uncertainty, particularly in patients with suspected aqueductal stenosis [3]. To address this, all cases were retrospectively reviewed individually using available neuroimaging, radiological descriptions and clinical information. Nevertheless, some degree of classification uncertainty may persist in very few patients.

Although comorbidities were recorded descriptively, their specific impact on surgical outcomes was not analysed in detail and should therefore be interpreted descriptively.

Other limitations include the retrospective single-centre design and genetic testing restricted to selected patients with suspected genetic aetiology, which limits generalisability. The key strength lies in the detailed descriptions of aetiology, comorbidities and potential genetic mechanisms underlying paediatric hydrocephalus in a contemporary, unselected cohort. In addition, all patients were systematically followed for at least one year.

Conclusions

Paediatric hydrocephalus is a highly heterogeneous condition characterised by diverse aetiologies, comorbidities and genetic findings. The present study provides a descriptive overview of these characteristics and the shunt treatment course in a Danish regional cohort. Future studies are warranted to study potential associations between aetiologies, genetic predispositions, comorbidity burden and surgical outcomes.

Correspondence *Anna Duc Jensen*. E-mail: anna.duc.jensen@regionh.dk

*) Shared first-authorship

Accepted 8 September 2026

Published 15 October 2026

Conflicts of interest MJ reports financial support from or interest in Munksgaard and FADL. All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. These are available together with the article at ugeskriftet.dk/dmj

References can be found with the article at ugeskriftet.dk/dmj

Cite this as Dan Med J 2026;73(11):A03260256

doi 10.61409/A03260256

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