Turkish Journal of Pediatric Disease

Original Article

Risk factors and outcomes of neonatal progressive hydrocephalus: A retrospective analysis

Kevser Elik[1], Cüneyt Tayman[2], Harun Demirci[3]

[1] Department of Pediatrics, Başkale State Hospital, Van, Türkiye, [2]Department of Neonatology, Ankara Bilkent City Hospital, Ankara, Türkiye, [3]Department of Neurosurgery, Ankara Bilkent City Hospital, Ankara, Türkiye

Correspondence Author: **Kevser Elik**, e-mail:kevserturann10@gmail.com

Received : 26.09.2025, Accepted : 15.01.2026 DOI: 10.12956/TJPD.2025.1248

ABSTRACT

Objective: Hydrocephalus is a complex neurological disorder affecting the central nervous system, which can lead to severe neurodevelopmental complications despite early diagnosis and treatment. Although advances in diagnostic and therapeutic methods have been achieved, managing hydrocephalus remains challenging. Neural tube defects are among the most common causes of hydrocephalus, highlighting the importance of prenatal diagnosis. Treatment often involves surgical intervention, making imaging techniques and timely surgical management critical. This study aimed to investigate the risk factors, etiologies, and treatment outcomes of neonates diagnosed with hydrocephalus.

Material and Methods: A retrospective study was conducted on neonates diagnosed with hydrocephalus and followed at Ankara Bilkent City Hospital Neonatal Intensive Care Unit between September 2019 and January 2023. Neonates who died in the delivery room or during transfer, whose records were incomplete, or whose families were unreachable were excluded. Clinical, demographic, and treatment data were obtained from hospital records and analyzed.

Results: Out of 115 patients screened, 104 met the inclusion criteria. Of these, 42% were born prematurely. The prevalence of congenital hydrocephalus was 74%, post-hemorrhagic hydrocephalus 24.1%, and post-infectious hydrocephalus 1.9%. Female infants comprised 58.7% of cases, and 87.5% were delivered via cesarean section. Among mothers, 42.5% reported regular folic acid intake during pregnancy. Parental consanguinity was noted in 24.5% of cases. Additional anomalies were present in 74% of patients, with 67.3% receiving an antenatal diagnosis. Epilepsy was observed in 36.5% of patients, and 39.9% of these were treated with antiepileptic drugs. Ventriculoperitoneal shunts were placed in many patients, with 44.7% requiring shunt revision, predominantly due to infection (33.7%). Referral cases accounted for 17.3%, mostly post-hemorrhagic hydrocephalus, with a 72% epilepsy rate and 25.3% antiepileptic treatment initiation in this subgroup.

Conclusion: This study provides comprehensive insights into the epidemiology, familial risk factors, etiological profiles, and treatment outcomes of progressive hydrocephalus diagnosed antenatally or postnatally. The findings offer valuable data to inform improved diagnostic and therapeutic strategies for neonates affected by hydrocephalus.

Keywords: Antenatal diagnosis, hydrocephalus, neonate, neural tube defects, risk factors, ventriculoperitoneal shunt

Introduction

Hydrocephalus is defined by the pathological accumulation of cerebrospinal fluid (CSF) within the cerebral ventricles and/or subarachnoid spaces due to impaired CSF circulation (1). The meninges—comprising the dura mater, arachnoid mater, and pia mater—together with CSF play a crucial role in protecting the central nervous system. CSF circulates continuously within the subarachnoid space, maintaining homeostasis and providing mechanical protection (2). Clinically, hydrocephalus is classified as obstructive (non-communicating) or communicating, depending on whether the impairment lies in CSF flow pathways or in the balance between CSF production and absorption. The condition is characterized by ventricular dilatation and increased intracranial pressure (ICP), resulting from complex pathophysiological mechanisms. Clinical manifestations are often non-specific and may occur irrespective of the underlying etiology (3).

The prevalence of congenital hydrocephalus in the United States and Europe is estimated at 0.5–0.8 per 1000 live and stillbirths. Approximately half of the cases are associated with meningomyelocele, although this proportion varies geographically (4). First described by Hakim and Adams in 1965, hydrocephalus has since been linked to potential genetic susceptibility, as suggested by its increased frequency in certain ethnic and regional populations. While no single causative gene has been identified, multiple genetic alterations are believed to contribute to its development (5,6). Several maternal risk factors have been implicated, including folic acid and zinc deficiency, fetal alcohol exposure, maternal obesity, antiepileptic drug use, and insulin-dependent diabetes mellitus. In the neonatal period, neural tube defects represent the most common etiology, with meningomyelocele being the predominant form. The extent of neurological impairment varies according to lesion level and is frequently permanent (7). Prenatal diagnosis, achievable in up to 90% of cases, allows for optimized delivery planning and early postnatal management. The primary goal in the management of meningomyelocele is to preserve neurological function at birth and prevent complications such as meningitis and sepsis. Early postnatal surgical repair of exposed neural tissue and closure of the defect remains the most effective intervention (8).

Diagnosis of hydrocephalus relies on imaging modalities such as ultrasonography, computed tomography, and magnetic resonance imaging. Treatment options include CSF diversion techniques, primarily ventriculoperitoneal shunting or endoscopic third ventriculostomy. Careful patient selection is essential, and differential diagnoses involving other neurodegenerative conditions must be excluded. Patients with shorter symptom duration and predominant lower extremity involvement are more likely to benefit from surgical intervention (9). Hydrocephalus remains a condition with significant morbidity despite advances in diagnosis and treatment. Early prenatal detection, identification of risk factors, and appropriate surgical management play a decisive role in improving outcomes. The aim of this study was to retrospectively evaluate risk factors, etiological causes, and treatment outcomes in newborns diagnosed with hydrocephalus and to propose recommendations for diagnostic, therapeutic, and follow-up strategies based on these findings.

Materials and Methods

This retrospective study analyzed a total of 104 newborns diagnosed with hydrocephalus at the Neonatal Intensive Care Units (NICU) of Ankara Bilkent City Hospital Children's Hospital between September 2019 and January 2023. Inclusion criteria comprised a diagnosis of hydrocephalus within the neonatal period (the first 28 days of life) confirmed via neuroimaging (ultrasonography, CT, or MRI), administration of medical or surgical treatment, and the availability of complete medical and follow-up records. Conversely, patients diagnosed after the neonatal period, those with insufficient imaging or incomplete follow-up data, and postmortem-diagnosed cases who did not receive treatment were excluded from the study. Demographic and clinical characteristics, laboratory findings, and follow-up data were retrospectively retrieved from the hospital's electronic medical record system.

The diagnosis of hydrocephalus was established based on neurological assessments and standard neuroimaging techniques, primarily including cranial ultrasonography (USG), magnetic resonance imaging (MRI), and computed tomography (CT). The presence of ventriculomegaly, elevated cerebrospinal fluid (CSF) pressure, and clinical symptoms—such as increased head circumference, persistent fontanelle patency, and neurological impairments—were considered in combination. Imaging findings were also used to classify the type of hydrocephalus as congenital, post-hemorrhagic, or post-infectious. Among the 104 patients included in the study, transfontanelle ultrasonography (TFUS) was performed in at least 88 patients (84.6%), while specific TFUS parameters were assessed in up to 95 patients (91.3%). The variation in patient numbers resulted from missing data or parameters that were not evaluated in all cases.

The decision to perform ventriculoperitoneal (VP) shunt placement was made by a multidisciplinary team, taking into account the patient's clinical status, imaging findings, and CSF pressure measurements. Indications for shunt surgery included elevated intracranial pressure, progressive ventricular dilatation, and worsening neurological symptoms. Post-treatment follow-up included regular assessment for complications, shunt revisions, and infection surveillance.

Statistical analysis

The data were analyzed using IBM SPSS Statistics for Windows (Version 24.0; IBM Corp., Armonk, NY, USA). As continuous variables did not show a normal distribution, they were summarized as median (interquartile range, Q1–Q3), minimum, and maximum values, while categorical variables were presented as counts and percentages. Comparisons between groups were performed using the Mann–Whitney U test for continuous variables and the chi-square test or Fisher's exact test, as appropriate. A p value of ≤ 0.050 was considered statistically significant.

Results

This retrospective study included 104 neonates diagnosed with hydrocephalus. Of these, 41.3% were male and 58.7% were female. Regarding the mode of delivery, 87.5% were born via cesarean section (CS), while 12.5% were delivered through normal spontaneous vaginal delivery (NSVD). A total of 42% of the infants were preterm births. Among the mothers, 75% had no chronic disease before or during pregnancy, whereas 22.1% had a chronic condition. According to maternal medical history, the most frequent chronic condition among the mothers was hypothyroidism, followed by hypertension, diabetes mellitus, and asthma.

Table I: Gender distribution and maternal pregnancy characteristics of patients (n=104)

Characteristic	n (%)
Gender	
Male	43 (41.3)
Female	61 (58.7)
Delivery mode	
Cesarean section	91 (87.5)
Normal spontaneous vaginal delivery	13 (12.5)
Maternal chronic disease (n=101)	
Present	23 (22.1)
Absent	78 (75.0)
Medications during pregnancy (n=19)	
Levothyroxine	8 (36.3)
Anticoagulant	3 (13.6)
Insulin	2 (9)
Antihypertensive	2 (9)
Inhaled corticosteroid	2 (9)
Antibacterial	2 (9)
Urinary antispasmodic	1 (4.5)
Antiretroviral	1 (4.5)
Phlebotonic	1 (4.5)
Folic acid supplementation (n=94)	
Regular	40 (42.5)
Irregular	31 (32.9)
None	23 (24.4)
History of abortion (n=104)	
Present	37 (35.1)
Absent	67 (64.9)

Among the 19 mothers who used medication during pregnancy, the distribution was as follows: 36.3% used levothyroxine, 13.6% anticoagulants, 9% insulin, 9% antihypertensives, 9% inhaled corticosteroids, 9% antibacterials, 4.5% urinary antispasmodics, 4.5% antiretrovirals, and 4.5% phlebotonics (Table I).

While 42.5% of the mothers regularly used folic acid throughout pregnancy, 32.9% used it irregularly, and 24.4% did not use it at all. Additionally, 35.1% of the mothers had a history of at least one abortion. Table I summarizes the gender distribution of the patients and the pregnancy characteristics of the mothers.

The mean maternal age was 27.48±6.22 years, the mean birth weight of the infants was 2569.80±882.97 g, and the mean gestational age was 35.65±4.30 weeks. The mean number of pregnancies among the mothers was 2.95±2.06. The mean APGAR scores were 5.69±1.87 at 1 minute and 7.68±1.48 at 5 minutes postpartum. The mean time to symptom onset was 3.32 ± 9.64 days (Table II).

Among the patients, 67.3% had received an antenatal diagnosis. The rates of ventriculomegaly, meningomyelocele, and encephalocele were 56.3%, 43.6%, and 10.8%, respectively. While 81.8% of the patients were born in the hospital, 17.3% were referred from external centers. The rate of parental consanguinity was 24.5%, and 11.1% of the mothers reported a history of previous pregnancies affected by hydrocephalus or spina bifida.

Regarding hydrocephalus types, 74% had congenital hydrocephalus, and 24.1% had posthemorrhagic hydrocephalus.

Table II: Demographic and clinical characteristics of the study population

Variable	mean±SD	min-max
Maternal age (years)	27.48±6.22	17.00-42.00
Birth weight (g)	2569.80±882.97	700.00-4880.00
Gestational age (weeks)	35.65±4.30	24.00-41.00
Gravidity	2.95±2.06	1.00-11.00
APGAR 1	5.69±1.87	1.00-8.00
APGAR 5	7.68±1.48	4.00-9.00
Antenatal diagnosis week	24.15±6.04	10.00-39.00
Symptom onset (postnatal day)	3.32±9.64	0-61

The rate of additional anomalies was 92.3%. Among the 32 patients with facial anomalies, 59.3% had dysmorphic facial features, and 12.5% had scaphocephaly.

Epilepsy was present in 36.5% of the patients, and antiepileptic treatment was initiated in 39.9%. Shunt revision was performed in 44.7% of the patients, and shunt infection developed in 33.7%. The rate of central nervous system (CNS) infection was 28.8%, and cerebrospinal fluid (CSF) culture was positive in 33.7% of the cases.

Discussion

Hydrocephalus is characterized by abnormal cerebrospinal fluid (CSF) accumulation resulting from impaired production, circulation, or absorption. Sustained intracranial pressure elevation and ventricular dilatation adversely affect neuronal development; therefore, early diagnosis and timely intervention are critical to reducing morbidity and mortality (10). In this context, our findings provide insight into both etiological distribution and outcome patterns in a tertiary referral neonatal population.

In our cohort, congenital hydrocephalus was the predominant etiology (74%), followed by posthemorrhagic (24.1%) and postinfectious forms (1.9%). The overall incidence of neonatal hydrocephalus (1.52%) is consistent with reports from developed countries. However, the relatively high proportion of posthemorrhagic hydrocephalus likely reflects our center's role as a referral unit for extremely preterm infants. Importantly, this distribution suggests that local neonatal care characteristics substantially influence etiological patterns, emphasizing the need for center-specific management strategies (11,12).

Contrary to reports of male predominance in pediatric hydrocephalus, female infants constituted the majority of our cohort (13). We believe this finding may be related to a higher proportion of prenatally diagnosed congenital cases and referral bias, rather than a true gender-related biological difference. This observation highlights the impact of prenatal screening practices on postnatal epidemiological profiles.

Birth weight was comparable to previous studies; however, lower gestational age and earlier prenatal diagnosis were

notable findings (14). Earlier detection likely reflects increased antenatal surveillance of high-risk pregnancies and may contribute to improved perinatal stabilization. At the same time, the lower gestational age may explain the substantial burden of posthemorrhagic hydrocephalus observed, reinforcing the close relationship between prematurity and CSF circulation disorders.

The very high cesarean section rate (87.5%) and relatively low first-minute APGAR scores mirror findings from similar healthcare settings (15). In our opinion, these findings reflect the complex perinatal management of prenatally diagnosed hydrocephalus cases rather than inadequate obstetric care, as most neonates showed improvement by the fifth minute. Maternal age did not differ from national data and does not appear to be an independent risk factor (13,16). However, maternal comorbidities were common (22.1%) and may have contributed indirectly to hydrocephalus development by increasing the risk of prematurity and intraventricular hemorrhage (17–19). This supports the concept that maternal health optimization may play a role in preventive strategies (20).

Medication exposure during pregnancy was documented in nearly one-fifth of cases. Although most medications were clinically justified, this finding underscores the importance of careful pharmacological counseling during pregnancy, particularly during the organogenesis period, when the fetal brain is highly vulnerable to teratogenic effects (21–23). Suboptimal folic acid supplementation remains a significant concern. Similar to previous reports, irregular or absent folic acid use was common (24). While maternal risk factors alone may not fully explain hydrocephalus development, our findings suggest that inadequate preventive care may exacerbate underlying genetic or environmental susceptibility (25,26).

The predominance of congenital hydrocephalus in our cohort contrasts with studies reporting posthemorrhagic etiologies as the leading cause (25). We believe this difference reflects referral of antenatally diagnosed and structurally complex cases. Additionally, emerging evidence on genetic and molecular mechanisms affecting neurodevelopment and CSF regulation supports the possibility that undiagnosed genetic factors contributed to our high congenital case rate (26). A striking finding of our study was the very high rate of associated anomalies (92.3%), far exceeding previously reported rates (14). This may be explained by lower pregnancy termination rates and referral of multisystem anomaly cases. Clinically, this finding underscores the necessity of comprehensive prenatal counseling and multidisciplinary postnatal care.

Long-term neurological morbidity was considerable. Neuromotor impairment and epilepsy were frequent, with epilepsy observed in 36.5% of patients, exceeding rates reported in European cohorts (27). Although epilepsy was less frequent in congenital hydrocephalus than in secondary forms, the overall neurological burden highlights the need for structured long-term follow-up and early neurodevelopmental intervention. Shunt-related complications and epilepsy remain major determinants of quality of life (28). Our findings support the growing emphasis on individualized treatment strategies, early risk stratification, and advances in prenatal diagnosis to reduce shunt dependency and improve long-term outcomes (29).

Conclusion

This study is limited by its retrospective, single-center design, incomplete records, and lack of standardized neurodevelopmental and genetic assessments. Nevertheless, it provides valuable real-world data reflecting the impact of referral patterns, prenatal diagnosis, and maternal factors on neonatal hydrocephalus. Future multicenter prospective studies incorporating genetic analyses and long-term follow-up are essential to further clarify disease mechanisms and optimize management.

Ethics committee approval

This study was conducted in accordance with the Helsinki Declaration Principles. The study was approved by Ankara Bilkent City Hospital (15.03.2023, reference number: E2-23-3297).

Contribution of the authors

EK, DH: Collected and recorded the patients' data, were responsible for literature research, TC, DH: Followed patients, TC, EK: Took the lead in writing the manuscript. All authors discussed the results and contributed to the final manuscript.

Source of funding

The authors declare the study received no funding.

Conflict of interest

The authors declare that there is no conflict of interest.

References

1. Haridas A, Tomita T. Hydrocephalus in children: Physiology, pathogenesis, and etiology. In: Nordli DR ed. UpToDate. (2022) Updated December 30 2024. Accessed August 2025. https://www.uptodate.com
2. Turhan M, Erbek S, Yılmaz DM. Hydrocephalus: Diagnosis, treatment and classification. Cukurova Med J. 2024;4(1):24-32.
3. Canaz H, Alataş İ, Batçık OE, Akdemir AO, Baydın S. Hydrocephalus in Early Childhood. Kafkas Medical Journal. 2013;(2):88-95. https://doi.org/10.5505/kjms.2013.41196
4. Hatiboglu MA, Abdallah A. Management of Acute Hydrocephalus. Tuzgen S, editor Emergency Approaches in Brain and Nerve Surgery. 1st ed. Ankara : Nobel Medical Bookstore, 2019. p.111-128.
5. Adams RD, Fisher CM, Hakim S, et al. Symptomatıc Occult Hydrocephalus Wıth "Normal" Cerebrospınal-Fluıd Pressure. A Treatable Syndrome. N Engl J Med. 1965;273:117-26. https://doi.org/10.1056/NEJM196507152730301
6. Zhang J, Williams MA, Rigamonti D. Genetics of human hydrocephalus. J Neurol. 2006;253(10):1255-66. https://doi.org/10.1007/s00415-006-0245-5
7. Serifoglu L, Isık S, Etli MU, Seckin MS, Onduc GG. Determining Hydrocephalus and V-P Shunt Requirements After Repair of Myelomeningocele Defects in Infants. Med J Bakirkoy. 2025 25;21(1):1-6. https://doi.org/10.4274/BMJ.galenos.2024.2024.4-11
8. Hakim S, Adams RD. The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure. Observations on cerebrospinal fluid hydrodynamics. J Neurol Sci. 1965;2(4):307-27. https://doi.org/10.1016/0022-510X(65)90016-X

9. Ozen A, Akar S. Retrospective evaluation of operated meningomyelocele cases. Adıyaman University Journal of Health Sciences. 2023;9(2):133-41. https://doi.org/10.30569/adiyamansaglik.1313886
10. Gocmen S, Colak A, Classification and Pathophysiology of Pediatric Hydrocephalus. Türkiye: Turkish Journal of Neurosurgery 2013;23(2): 174-9.
11. Cizmeci MN, Khalili N, Claessens NHP, et al. Assessment of Brain Injury and Brain Volumes after Posthemorrhagic Ventricular Dilatation: A Nested Substudy of the Randomized Controlled ELVIS Trial. J Pediatr. 2019;208:191-97.e2. https://doi.org/10.1016/j.jpeds.2018.12.062
12. Robinson S. Neonatal posthemorrhagic hydrocephalus from prematurity: pathophysiology and current treatment concepts. J Neurosurg Pediatr. 2012;9(3):242-58. https://doi.org/10.3171/2011.12.PEDS11136
13. Tully HM, Capote RT, Saltzman BS. Maternal and infant factors associated with infancy-onset hydrocephalus in Washington State. Pediatr Neurol. 2015;52(3):320-5. https://doi.org/10.1016/j.pediatrneurol.2014.10.030
14. Garne E, Loane M, Addor MC, Boyd PA, Barisic I, Dolk H. Congenital hydrocephalus--prevalence, prenatal diagnosis and outcome of pregnancy in four European regions. Eur J Paediatr Neurol. 2010;14(2):150-5. https://doi.org/10.1016/j.ejpn.2009.03.005
15. Dutra M, da Silva SC, Beggiora PDS, Santos MV, Machado HR, Lopes LDS. Epidemiology of hydrocephalus in Brazil. J Pediatr (Rio J). 2023;99(3):228-34. https://doi.org/10.1016/j.jped.2022.10.008
16. Tunc Z, Colak C, Ozdemir R. Classification of Hydrocephalus Disease and Determination of Associated Factors Using Machine Learning Methods. Ann Health Sci Res. 2018;7(2):14-20.
17. Van Landingham M, Nguyen TV, Roberts A, Parent AD, Zhang J. Risk factors of congenital hydrocephalus: a 10 year retrospective study. J Neurol Neurosurg Psychiatry. 2009;80(2):213-7. https://doi.org/10.1136/jnnp.2008.148932
18. Stothard KJ, Tennant PW, Bell R, Rankin J. Maternal overweight and obesity and the risk of congenital anomalies: a systematic review and meta-analysis. JAMA. 2009;301(6):636-50. https://doi.org/10.1001/jama.2009.113
19. Desticioğlu K, Malas M. Maternal factors affecting fetal growth. Med J SDU. 2009;13(2):47-54.
20. Ali TM, Elwy R, Abdelrazik B, Soliman MAR, Alsawy MF, Abdullah A,et al. Risk factors of congenital hydrocephalus: a case-control study in a lower-middle-income country (Egypt). J Neurosurg Pediatr. 2023;31(5):397-405. https://doi.org/10.3171/2022.12.PEDS22321
21. Munch TN, Rasmussen ML, Wohlfahrt J, Juhler M, Melbye M. Risk factors for congenital hydrocephalus: a nationwide, register-based, cohort study. J Neurol Neurosurg Psychiatry. 2014;85(11):1253-9. https://doi.org/10.1136/jnnp-2013-306941
22. Konak M, Unver E, Sündüs US, Yorulmaz A, Soylu H. Sociodemographic Characteristics of Neural Tube Defect Cases Monitored in the Neonatal Intensive Care Unit. J Contemp Med. 2018;8(3):218-22. https://doi.org/10.16899/gopctd.439737
23. Werler MM, Ahrens KA, Bosco JL, Mitchell AA, Anderka MT, Gilboa SM, et al. National Birth Defects Prevention Study. Use of antiepileptic medications in pregnancy in relation to risks of birth defects. Ann Epidemiol. 2011;21(11):842-50. https://doi.org/10.1016/j.annepidem.2011.08.002
24. Liu J, Li Z, Ye R, Ren A, Liu J. Folic acid supplementation and risk for congenital hydrocephalus in China. Public Health Nutr. 2021;24(13):4238-4244. https://doi.org/10.1017/S136898002100029X
25. Holwerda JC, van Lindert EJ, Buis DR, Hoving EW; Dutch Pediatric Neurosurgery Study Group. Surgical intervention for hydrocephalus in infancy; etiology, age and treatment data in a Dutch cohort. Childs Nerv Syst. 2020;36(3):577-82. https://doi.org/10.1007/s00381-019-04333-7
26. Liu XY, Song X, Czosnyka M, et al. Congenital hydrocephalus: a review of recent advances in genetic etiology and molecular mechanisms. Mil Med Res. 2024;11(1):54. https://doi.org/10.1186/s40779-024-00560-5
27. Vinchon, M., Rekate, H. & Kulkarni, A.V. Pediatric hydrocephalus outcomes: a review. Fluids Barriers CNS 2012;9(1):18. https://doi.org/10.1186/2045-8118-9-18
28. Noetzel MJ, Blake JN. Seizures in children with congenital hydrocephalus: long-term outcome. Neurology. 1992 ;42(7):1277-81. https://doi.org/10.1212/WNL.42.7.1277
29. Schubert-Bast S, Berghaus L, Filmann N, Freiman T, Strzelczyk A, Kieslich M. Risk and risk factors for epilepsy in shunt-treated children with hydrocephalus. Eur J Paediatr Neurol. 2019;23(6):819-826. https://doi.org/10.1016/j.ejpn.2019.09.004