Pediatric Hydrocephalus Etiology

- MPDZ Pathogenic Variants Cause Obstructive Ventriculomegaly Related to Diencephalosynapsis and Third Ventricle Atresia
- Evaluation of the Endoscopic Third Ventriculostomy Success Score for pediatric hydrocephalus: experience from a Singapore children's hospital
- Volumetric predictors for shunt-dependency in pediatric posterior fossa tumors
- Rehabilitation in a child with Chiari II malformation, lumbosacral meningomyelocele, achondroplasia and impaired respiratory regulation - a case report and literature review
- Clinico-epidemiological profile of 75 cases of TB meningitis in children and adoloscents
- Genetically determined alterations in inflammation and infection-associated genes are associated with hydrocephalus in patients of African Ancestry
- Differences in brain development and need for CSF diversion based on MMC level: Comparison between prenatal and postnatal repair
- GLAPAL-H: Global, Local, And Parts Aware Learner for Hydrocephalus Infection Diagnosis in Low-Field MRI

---

see also Hydrocephalus Etiology.

---

see Congenital Hydrocephalus

Etiologies in one series of pediatric patients

1. congenital

a) Chiari Type 2 malformation and/or myelomeningocele (MM) (usually occur together)

b) Chiari Type 1 malformation: Hydrocephalus may occur with fourth ventricle outlet obstruction

c) Primary aqueductal stenosis (usually presents in infancy, rarely in adulthood)

d) Secondary aqueductal gliosis: due to intrauterine infection or germinal matrix hemorrhage

e) Dandy Walker malformation: atresia of foramina of Luschka & Magendie. The incidence of this in patients with HCP is 2.4%

f) X-linked inherited disorder: rare

Acquired

a) infectious (the most common cause of communicating HCP)

● post-meningitis; especially purulent and basal, including TB, cryptococcus

● cysticercosis

b) post-hemorrhagic (2nd most common cause of communicating HCP)

● post-SAH

● post-intraventricular hemorrhage (IVH): many will develop transient HCP. 20–50% of patients with large IVH develop permanent HCP, requiring a shunt

Congenital (without myelomeningocele) 38%

Congenital (with myelomeningocele) 29%

Perinatal hemorrhage 11%

Trauma/subarachnoid hemorrhage 4.7%

Tumor 11%

Previous infection 7.6%

c) secondary to masses

● non neoplastic: e.g. vascular malformation

● neoplastic: most produce obstructive hydrocephalus by blocking CSF pathways, especially tumors around aqueduct (e.g. medulloblastoma). A colloid cyst can block CSF flow at the foramen of Monro.

Pituitary tumor: suprasellar extension of tumor or expansion from pituitary apoplexy

d) post-op: 20% of pediatric patients develop permanent hydrocephalus (requiring shunt) following p-fossa tumor removal. May be delayed up to 1 yr

e) neurosarcoidosis

f) “constitutional ventriculomegaly”: asymptomatic. Needs no treatment

g) associated with spinal tumors: ? due to ↑ protein?, ↑ venous pressure?, previous hemorrhage in some?

Hydrocephalus has many causes:

Postinfectious hydrocephalus.

Postoperative hydrocephalus.

Posttraumatic hydrocephalus.

Posthemorrhagic hydrocephalus.

Trapped fourth ventricle.

Neurofibromatosis type 1 related hydrocephalus

Congenital hydrocephalus, most commonly involving aqueductal stenosis, has been linked to genes that regulate brain growth and development.

Newborn infants with germinal matrix hemorrhage.

Hydrocephalus can also be acquired, mostly from pathological processes that affect ventricular outflow, subarachnoid space function, or cerebral venous compliance.

Aneurysmal subarachnoid hemorrhage.

see Hydrocephalus after intraventricular hemorrhage

Meningitis

Hydrocephalus after decompressive craniectomy.

Terminal deletion of chromosome 6q is a rare chromosomal abnormality associated with intellectual disabilities and various structural brain abnormalities.

Iwamoto et al. presented a case of 6q terminal deletion syndrome with unusual magnetic resonance imaging (MRI) findings in a neonate.

The neonate, who was prenatally diagnosed with dilation of both lateral ventricles, was born at 38 weeks of gestation. MRI demonstrated abnormal membranous structure continuing to the hypertrophic massa intermedia in the third ventricle that had obscured the cerebrospinal fluid pathway, causing hydrocephalus. G-band analysis revealed a terminal deletion of 6q with the karyotype 46, XY, add(6)(q25.3) or del(6)(q26). He underwent ventriculoperitoneal shunt successfully, and his head circumference has been stable.

6q terminal deletion impacts the molecular pathway, which is an essential intracellular signaling cascade inducing neurological proliferation, migration, and differentiation during neuronal development. In patients with hydrocephalus in association with hypertrophy of the massa intermedia, this chromosomal abnormality should be taken into consideration. This case may offer an insight into the hydrocephalus etiology in this rare chromosomal abnormality [1)].

Molecular mechanisms

The underlying molecular mechanisms remain unknown. Ju et al. performed proteomics of cerebrospinal fluid (CSF) from 7 congenital hydrocephalus and 5 arachnoid cyst patients who underwent surgical treatment. Differential expression of proteins (DEPs) were identified by label-free Mass Spectrometry followed by differential expression analysis. The GO and GSEA enrichment analysis was performed to explore the cancer hallmark pathways and immune-related pathways affected by DEPs. Then, network analysis was applied to reveal the location of DEPs in the human protein-protein interactions (PPIs) network. Potential drugs for hydrocephalus were identified based on drug-target interaction. They identified 148 up-regulated proteins and 82 down-regulated proteins, which are potential biomarkers for clinical diagnosis of hydrocephalus and arachnoid cyst. Functional enrichment analysis revealed that the DEPs were significantly enriched in the cancer hallmark

pathways and immune-related pathways. In addition, network analysis uncovered that DEPs were more likely to be located in the central regions of the human PPIs network, suggesting DEPs may be proteins that play important roles in human PPIs. Finally, they calculated the overlap of drug targets and the DEPs based on drug-target interaction to identify the potential therapeutic drugs of hydrocephalus. The comprehensive proteomic analyses provided valuable resources for investigating the molecular pathways in hydrocephalus, and uncovered potential biomarkers for clinical diagnosis and therapy [2].

Posttraumatic hydrocephalus

Posttraumatic hydrocephalus

Hydrocephalus after Vertebrobasilar Dolichoectasia

Hydrocephalus after Vertebrobasilar Dolichoectasia.

Myelomeningocele-associated hydrocephalus

Myelomeningocele-associated hydrocephalus

1)
Iwamoto H, Muroi A, Sekine T, Tsurubuchi T, Ishikawa E, Matsumura A. Unusual Form of Obstructive Hydrocephalus in Association with 6q Terminal Deletion Syndrome: A Case Report and Literature Review. Pediatr Neurosurg. 2019 Oct 9:1-5. doi: 10.1159/000503108. [Epub ahead of print] PubMed PMID: 31597145.
2)
Ju Y, Wan Z, Zhang Q, Li S, Wang B, Qiu J, Zheng S, Gu S. Proteomic analyses reveal functional pathways and potential targets in pediatric hydrocephalus. Curr Gene Ther. 2023 Jun 13. doi: 10.2174/1566523223666230613144056. Epub ahead of print. PMID: 37317915.