

Hydrocephalus

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Definition

see [Hydrocephalus definition](#).

History

The earliest recorded mention of CSF appears in the [Edwin Smith Papyrus](#) ¹⁾.

[Hydrocephalus](#) cases were regularly described by [Hippocrates](#), [Galen](#), and early and medieval Arabian physicians, who believed that an extracerebral accumulation of water caused this disease. Operative procedures used in ancient times are neither proven by skull findings today nor clearly reported in the literature.

Evacuation of superficial intracranial fluid in [pediatric hydrocephalus](#) was first described in detail in the tenth century by Abulkassim [Al Zahrawi](#).

On 15th October 1744, the French surgeon Claude-Nicolas [Le Cat](#) (1700-1768) introduced a specially invented [canula](#) into the [lateral ventricle](#) of a newborn boy with [hydrocephalus](#). The canula was used as a [tap](#) and was left in place for 5 days, until the death of the child. This procedure should be seen as the first documented description of a device for repeated ventricular taps in the treatment of hydrocephalus ²⁾.

Effective therapy required aseptic surgery as well as pathophysiological knowledge—both unavailable before the late nineteenth century.

Key and Retzius put these previous studies together, proving that CSF is secreted by the choroid plexus, flows through the ventricular system, and is reabsorbed via subarachnoid villi and Pacchonian granulations ³⁾.

In 1881, a few years after the landmark study of Key and Retzius, Wernicke inaugurated sterile ventricular puncture and external CSF drainage. These were followed in 1891 by serial lumbar

punctures (Quincke) and, in 1893, by the first permanent ventriculo-subarachnoid-subgaleal shunt (Mikulicz), which was simultaneously a ventriculostomy and a drainage into an extrathecal low pressure compartment.

Between 1898 and 1925, lumboperitoneal, and ventriculoperitoneal, -venous, -pleural, and -ureteral shunts were invented, but these had a high failure rate due to insufficient implant materials in most cases. Ventriculostomy without implants (Anton 1908), with implants, and plexus coagulation initially had a very high operative mortality and were seldom successful in the long term, but gradually improved over the next decades.

In 1949, Nulsen and Spitz implanted a shunt successfully into the caval vein with a ball valve. Between 1955 and 1960, four independent groups invented distal slit, proximal slit, and diaphragm valves almost simultaneously. Around 1960, the combined invention of artificial valves and silicone led to a worldwide therapeutic breakthrough. After the first generation of simple differential pressure valves, which are unable to drain physiologically in all body positions, a second generation of adjustable, autoregulating, [antisiphon](#), and gravitational valves was developed, but their use is limited due to economical restrictions and still unsolved technical problems. At the moment, at least 127 different designs are available, with historical models and prototypes bringing the number to 190 valves, but most of these are only clones. In the 1990s, there has been a renaissance of endoscopic ventriculostomy, which is widely accepted as the method of first choice in adult patients with acquired or late-onset, occlusive hydrocephalus; in other cases the preference remains controversial. Both new methods, the second generation of valves as well as ventriculostomy, show massive deficits in evaluation. There is only one randomized study and no long-term evaluation ⁴⁾.

Walter Dandy, in collaboration with [Kenneth Blackfan](#), Department of Pediatrics, conducted experimental studies in dogs, which led him to conclude that the obstruction at the foramen of Monro, aqueduct of Sylvius, or around the brainstem, produce hydrocephalus and cause decreased absorption of [cerebrospinal fluid](#) (CSF).

Blackfan's research with Dandy involved an experimental model to produce hydrocephalus in dogs that helped establish the basis of our current understanding of cerebrospinal fluid physiology. This work was published in two classical papers in the American Journal of Diseases in Children, one in 1913 and the other in 1917 ⁵⁾.

The second paper was later reprinted in the Annals of Surgery ⁶⁾.

Blackfan's collaborative work with Dandy also expanded to the description of internal hydrocephalus in infants, the early recognition of hydrocephalus in children, the signs of cerebral venous thrombosis, and a landmark paper on the treatment of meningococcal meningitis.

Epidemiology

It is more common in infants, although it can occur in older adults.

The most common type of hydrocephalus in developing countries is [postinfectious hydrocephalus](#).

Classification

see [Hydrocephalus Classification](#).

Etiology

see [Hydrocephalus Etiology](#).

Pathophysiology

see [Hydrocephalus Pathophysiology](#).

Pathogenesis

see [Hydrocephalus Pathogenesis](#).

Clinical Features

[Hydrocephalus Clinical Features](#)

Diagnosis

[Hydrocephalus diagnosis](#).

Differential diagnosis

[Hydrocephalus differential diagnosis](#)

Treatment

see [Hydrocephalus treatment](#).

Outcome

Intracerebral hemorrhage with intraventricular extension and hydrocephalus may increase mortality or severe disability ⁷⁾.

Neurocognitive outcome

The evaluation of hydrocephalus remains focused on ventricular size, yet the goal of treatment is to allow for healthy brain development. It is likely that brain volume is more related to cognitive development than is fluid volume in children with hydrocephalus.

Hydrocephalus is treated by normalizing CSF, but normal brain development depends on brain growth. A combination of brain and CSF volumes appears to be significantly more powerful at predicting good versus poor neurocognitive outcomes in patients with hydrocephalus than either volume alone ⁸⁾.

In infants with hydrocephalus, a greater 1-year CSF diversion failure rate may occur after [endoscopic third ventriculostomy](#) (ETV) compared with [shunt](#) placement. This risk is most significant for procedures performed within the first 90 days of life. Further investigation of the need for multiple reoperations, cost, and impact of surgeon and hospital experience is necessary to distinguish which treatment is more effective in the long term ⁹⁾.

Case series

[Hydrocephalus case series.](#)

Case reports

see [Hydrocephalus case reports.](#)

Research

[Hydrocephalus research.](#)

¹⁾

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