

# Hydrocephalus During Infancy

Hydrocephalus is a congenital or acquired disorder characterized by the abnormal accumulation of cerebrospinal fluid within the cavities of the brain, called ventricles. The accumulation of cerebrospinal fluid, the clear fluid surrounding the brain and spinal cord, causes an abnormal widening of the ventricles. The widening creates potentially harmful pressure on the tissues of the brain that can result in brain damage or death. The most obvious sign of hydrocephalus is the rapid increase in head circumference or an unusual large head size due to the accumulation of cerebrospinal fluid in the brain. In infants, hydrocephalus can be caused by congenital factors such as malformations of the brain, or acquired factors such as tumors, cysts, meningitis, or bleeding. Treatment after the infant is born can lead to normal cognitive and physical development with few limitations.

Medical documents dating back to Hippocrates, a Greek physician during the Age of Pericles, and Galen, a Greek physician and philosopher in the Roman Empire, as well as early and medieval Arabian physicians, describe the enlarged head common to hydrocephalus in infancy. All of those early physicians claimed that the cause of hydrocephalus originated from the accumulation of water within the brain. In the tenth century, the Arab physician and medieval surgeon Abulkassim Al Zahrawi even described the removal of superficial intracranial fluid in hydrocephalic children.

However, few doctors operated to correct hydrocephalus prior to the 1900s, when medical technology enabled for the condition and the brain to be visualized. In 1914, Walter Dandy and Kenneth Blackfan first described the clinical aspects and pathology of hydrocephalus at Johns Hopkins University in Baltimore, Maryland. They reported on a thirteen-month-old female observed with an enlargement of the fourth ventricle. The patient died two months after their observation. By 1919, Dandy conducted experiments using animal models to study how hydrocephalus arose and to create potential treatments for the fatal condition. He injected colored dye into the ventricles of living animals to visualize the flow of cerebrospinal fluid and detect possible hindrances of the flow. Dandy found that cerebrospinal fluid originated within a network of blood vessels called the choroid plexus.

From that research, Dandy classified hydrocephalus into two types. He defined the first type, communicating hydrocephalus, as the form of hydrocephalus in which he could recover the injected dye from the ventricle because the obstruction did not occur within the ventricles. The second type, non-communicating hydrocephalus, he defined as the form in which the dye did not reach the ventricle indicating that an obstruction within the ventricular system prevented the dye from proper flow.

Dandy attempted to treat hydrocephalus by removing the nerve networks in which cerebrospinal fluid developed, called the choroid plexuses. Physicians used that technique until the 1950s when upon review researchers found that the majority of patients with their choroid plexuses removed demonstrated progressive enlargement of the ventricles at the same rate or increased rate as observed before surgery. The poor outcome of the review led physicians to discontinue the technique.

Between 1898 and 1925, surgeons developed various techniques to insert a drainage system into the brain, called a shunt, all of which had a high failure rate. In 1950, Donald Darrow Matson, a pediatric neurosurgeon, showed that the use of the shunt successfully treated some forms of hydrocephalus. In 1952, researchers developed the valve-regulated shunt, a surgically implanted device that diverted cerebrospinal fluid to an internal delivery site such as the abdomen or the heart. Unlike previous shunts, the valve-regulated shunt used a spring and ball mechanism to regulate the flow and pressure of cerebrospinal fluid. That internal shunt permitted cerebrospinal fluid under pressure an easier avenue of escape, diverting it to the abdomen or heart, which both contain membranous tissue capable of absorbing the incoming fluid.

In 1960, Joseph Ransohoff conducted experiments that led to rename Dandy's non-communicating hydrocephalus as obstructive hydrocephalus. Ransohoff showed that non-communicating hydrocephalus involved the obstruction of cerebrospinal fluid flow between the point of production in the ventricular system and the point of absorption into the systemic circulation. With the flow of fluid obstructed, Ransohoff suggested renaming non-communicating hydrocephalus as obstructive hydrocephalus, a reclassification still in use as of 2017.

Surgeons in the late 1990s commonly treated hydrocephalus using neuroendoscopy, in which a small scope with a light and camera is passed through a manufactured hole in the skull into the brain allowing images to be projected onto a screen or monitor. Such a technique led to the development of endoscopic third ventriculostomy, a procedure in which an endoscope is used to make a small perforation in the floor of the third ventricle, allowing cerebrospinal fluid to escape from the blocked ventricular system. The objective of the procedure was to normalize pressure on the brain without using a shunt. Endoscopic third ventriculostomy is an alternative treatment for hydrocephalus rather than a cure. The procedure is often used for the treatment of obstructive hydrocephalus in infants older than one year due to the ability of cerebrospinal fluid to bypass the obstruction through the perforation in the third ventricle. Infants younger than one year show a high mortality rate with endoscopic third ventriculostomy. However, as of 2017 shunts are still the common treatment form of hydrocephalus due to the high risks of complication, infection, and death associated with endoscopic ventriculostomy.

Hydrocephalus is typically classified according to the location of the obstruction of cerebrospinal fluid flow. The obstruction can occur any place along the passageways of cerebrospinal fluid within the brain's four ventricles. Ventricle one and two are called lateral ventricles and are located along the midline of the brain, toward the front of the head, one on the left and the other on the right. The two lateral ventricles connect to the third ventricle in the middle of the ventricular system that connects and communicates with the two lateral ventricles on each side of it, and the fourth ventricle below it. The fourth ventricle extends from the midbrain to the central canal of the upper end of the spinal cord. Cerebrospinal fluid originates in the lateral ventricles and flows into the third midline ventricle. The fluid then drains via the canal that connects the ventricles, called the cerebral aqueduct into the fourth ventricle and out into the subarachnoid space between the brain and the skull. Cerebrospinal fluid is absorbed in the pouches of the subarachnoid space, a process that prevents excess fluid from accumulating and leading to fatal brain damage.

The communicating type of hydrocephalus is one in which the flow of cerebrospinal fluid is blocked after it exits the ventricles. The blockage may result from prior bleeding or meningitis and causes the thickening of the protective membranes of the brain leading to the obstruction of the return-flow channels of cerebrospinal fluid. By contrast, obstructive hydrocephalus is the one in which the lesion is obstructing the flow of cerebrospinal fluid before it enters the subarachnoid space. The distinction is useful for determining the cause of hydrocephalus and the available treatment options.

The cause of hydrocephalus largely depends on the patient's age. Premature infants are susceptible to brain bleeding, which can lead to hydrocephalus due to the impairment of cerebrospinal fluid reabsorption leading to the accumulation of fluid in the brain. Hydrocephalus caused by a brain bleed is treated conservatively with less than twenty-five percent of cases requiring surgical intervention and cerebrospinal fluid drainage because the swollen protective membranes in the subarachnoid space often return to normal allowing the subarachnoid space to reabsorb cerebrospinal fluid. Infants born at term are likely to have hydrocephalus arising from congenital malformation. One of the congenital malformations includes the narrowing of the canal connecting the ventricles, called aqueduct stenosis. The Dandy-Walker malformation, an expansion of the fourth ventricle caused by a cyst, is another form of a congenital malformation that may occur. Lastly, congenital malformations may stem from hindbrain abnormalities, called the Chiari malformation named after Hans Chiari, who studied the condition in Europe in the nineteenth century.

The presentation of hydrocephalus depends on the age of the patient and the rate of progression of the hydrocephalus, specifically before the closure of the soft, cartilaginous fontanelles, composed of a flexible, fibrous tissue that hold the bones of an infant's skull together before the cranial sutures are formed. As the fontanelles are flexible to a degree they can allow the cranium to expand in the

presence of excess fluid, leading to the enlarged head at times noted with hydrocephalus. During infancy, hydrocephalus typically presents with increased head circumference called macrocephaly, the down-deviation of the eyes colloquially called sun setting, distended scalp veins, developmental delays or regressions, and poor appetite.

Once doctors observe the clinical presentation of hydrocephalus, the diagnosis can be confirmed with imaging, which may also indicate the underlying cause. In utero, and in infants with fontanelles, brain ultrasound scanning is useful. Physicians measure ventricular size and detect bleeding within the ventricles. Computerized tomography scans, or CT scans, are helpful in assessing ventricular size and morphology, and can be used to determine the level of obstruction to the cerebrospinal fluid flow. CT scans may also show acute bleeding and the majority of tumors. Magnetic resonance imaging, or MRI scans, provide the most detailed anatomical pictures and are helpful in detecting lesions. The images from any scanning technology may be used to reveal the type of hydrocephalus, communicating or obstructive. When a scan reveals communicating hydrocephalus, physicians order a lumbar puncture to remove cerebrospinal fluid from the lower region of the spinal canal, which may be useful to measure the pressure of cerebrospinal fluid and determine treatment options.

The treatment of hydrocephalus depends on the cause, the age of the patient, and the how fast symptoms manifest. In an emergent situation, the quickest treatment is an external ventricular drain. That procedure releases intracranial pressure by draining cerebrospinal fluid with a shunt. Doctors pass a catheter through a burr hole made in the front of the skull, then through the front of the brain into the lateral ventricle and into an external collection device. The device is designed to drain the cerebrospinal fluid when the intracranial pressure exceeds the normal pressure range.

The most common form of treatment in non-emergent situations is a ventriculoperitoneal shunt, which is very similar to the external ventricular drain. Surgeons pass the catheter through a burr hole into the lateral ventricle but rather than draining externally, the catheter is connected to the valve and a second catheter tunneled to reach the peritoneal cavity within the abdomen where the cerebrospinal fluid is reabsorbed. When the peritoneal cavity is not favorable, the ventricles can be drained to other cavities such as the heart and the lungs. Many risks are associated with the ventriculoperitoneal shunt and the shunt is typically needed for the duration of the patient's life.

An alternative procedure for treating hydrocephalus is to use an endoscope to make a new drainage route out of the ventricular system, called the endoscopic third ventriculostomy. The procedure is suitable for patients with obstructive hydrocephalus from lesions, such as aqueduct stenosis, the narrowing of the connecting ventricular canal, or from tumors. The benefits of the procedure include the reduction of infection associated with shunt placement, as well as the long-term risks of shunts. However, endoscopic third ventriculostomy is not always successful in infants and the surgery may have to be repeated to maintain adequate opening and drainage. Fetal surgery is the most recent treatment option for hydrocephalus that is detected very early during development, although it poses a high risk of death or damage to both the fetus and the pregnant woman, and it is not practiced in the US.

The detection and treatment of hydrocephalus are important for the cognitive and physical development of the infant. The early detection and quick treatment may remarkably improve the development and quality of life for the affected infant. However, doctors are unable to predict the extent of recovery. When left untreated, the condition typically results in death of the infant. In contrast, when treated effectively there is minimal to no cognitive and developmental impairment.

## Sources

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