Hydrocephalus

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Common Names for Disorder

Hydrocephalus; “Water on the Brain”; The word “Hydrocephalus” in Greek literally means “watery head.”

It is a condition in which the primary characteristic is excessive accumulation of fluid in the brain. The "water" is actually cerebrospinal fluid (CSF) — a clear fluid surrounding the brain and spinal cord. The excessive accumulation of CSF results in an abnormal dilation of the spaces in the brain called ventricles. This dilation causes potentially harmful pressure on the tissues of the brain.

The balance between production and absorption of CSF is critically important. Ideally, the fluid is almost completely absorbed into the bloodstream as it circulates; however, there are circumstances which, when present, will prevent or disturb the production or absorption of CSF, or which will inhibit its normal flow. When this balance is disturbed, hydrocephalus is the result.

Causes / Etiology

Congenital / Acquired

Hydrocephalus may be congenital or acquired. Congenital hydrocephalus is present at birth and may be caused by either environmental influences during fetal development or genetic predisposition. Acquired hydrocephalus develops at the time of birth or at some point afterward. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease.

Communicating / Non-Communicating

Hydrocephalus may also be communicating or non-communicating. Communicating hydrocephalus occurs when the flow of CSF is blocked after it exits from the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open. Non-communicating hydrocephalus - also called "obstructive" hydrocephalus - occurs when the flow of CSF is blocked along one or more of the narrow pathways connecting the ventricles. One of the most common causes of hydrocephalus is "aqueductal stenosis." In this case, hydrocephalus results from a narrowing of the aqueduct of Sylvius, a small passageway between the third and fourth ventricles in the middle of the brain.

Ex-Vacuo / Normal Pressure

There are two other forms of hydrocephalus which do not fit distinctly into the categories mentioned above and primarily affect adults: hydrocephalus ex-vacuo and normal pressure hydrocephalus.

Hydrocephalus ex-vacuo occurs when there is damage to the brain caused by stroke or traumatic injury. In these cases, there may be actual shrinkage (atrophy or wasting) of brain tissue. Normal pressure hydrocephalus can occur in people of any age, but it is most common in the elderly population. It may result from a subarachnoid hemorrhage, head trauma, infection, tumor, or complications of surgery. However, many people develop normal pressure hydrocephalus even when none of these factors are present. In these cases the cause of the disorder is unknown.

The causes of hydrocephalus are not all well understood. Hydrocephalus may result from genetic inheritance (aqueductal stenosis) or developmental disorders such as those associated with neural tube defects including spina bifida and encephalocele. Other possible causes include complications of premature birth such as intraventricular hemorrhage, diseases such as meningitis, tumors, traumatic head injury, or subarachnoid hemorrhage.

Other

Premature infants have an increased risk of intraventricular hemorrhage in which severe bleeding within the ventricles of the brain can lead to hydrocephalus. Other problems that can occur during pregnancy may increase an infant's risk of developing hydrocephalus, including intrauterine infection or a disorder involving incomplete closure of an infant's spinal column (myelomeningocele).

Congenital or developmental defects can increase older children's risk of hydrocephalus. Lesions or tumors of the brain or spinal cord, central nervous system infections, bleeding in the brain, and severe head injury also can increase the risk of hydrocephalus.

Incidence

Incidence and prevalence data are difficult to establish as there is no existing national registry or database of people with hydrocephalus and closely associated disorders; however, hydrocephalus is believed to affect approximately 1 in every 500 children. At present, most of these cases are diagnosed prenatally, at the time of delivery, or in early childhood. Advances in diagnostic imaging technology allow more accurate diagnoses in individuals with atypical presentations, including adults with conditions such as normal pressure hydrocephalus.

- In the United States, a little over 1 in 1000 births are affected by hydrocephalus.
- Hydrocephalus is one of the most common "birth defects" and afflicts in excess of 10,000 babies each year.
- Studies by the World Health Organization show that one birth in every 2,000 result in hydrocephalus.
- There are 70,000 discharges a year from hospitals in the United States with a diagnosis of hydrocephalus.
- More than 50% of hydrocephalus cases are congenital.
- As many as 75% of children with hydrocephalus will have some form of motor disability.
- Over the past 25 years, death rates associated with hydrocephalus have decreased from 54% to 5%; intellectual disabilitity has decreased from 62% to 30%.
- About 80% of hydrocephalus patients are born with other defects. Other medical conditions usually associated with Hydrocephalus include:
  - Arachnoid Cysts.
  - Brain Injury.
  - Dandy-Walker Syndrome.
  - Head Trauma.
  - Meninigitis.
  - Porencephaly.
  - Tumors.
  - Spina Bifida.
    - Hydrocephalus occurs in 70 to 90% of children with the most severe form of Spina Bifida.

Characteristics

Symptoms of hydrocephalus vary with age, disease progression, and individual differences in tolerance to CSF. For example, an infant's ability to tolerate CSF pressure differs from an adult's. The infant skull can expand to accommodate the buildup of CSF because the sutures (the fibrous joints that connect the bones of the skull) have not yet closed.

In infancy, the most obvious indication of hydrocephalus is often the rapid increase in head circumference or an unusually large head size. Other symptoms may include vomiting, sleepiness and irritability, downward deviation of the eyes (also called “sunsetting”), seizures, and developmental delays.

Older children and adults may experience different symptoms because their skulls cannot expand to accommodate the buildup of CSF. In older children or adults, symptoms may include headache followed by vomiting, nausea, papilledema (swelling of the optic disk which is part of the optic nerve), blurred vision, diplopia (double vision), sunsetting of the eyes, problems with balance, poor coordination, gait disturbance, urinary incontinence, slowing or loss of development, lethargy, drowsiness, irritability, or other changes in personality or cognition including memory loss.

Symptoms of normal pressure hydrocephalus include progressive mental impairment and dementia, problems with walking, and impaired bladder control leading to urinary frequency and/or incontinence. The person also may have a general slowing of movements or may complain that his or her feet feel "stuck." Because these symptoms are similar to those of other disorders such as Alzheimer's disease, Parkinson's disease, and Creutzfeldt-Jakob disease, the disorder is often misdiagnosed. Many cases go unrecognized and are never properly treated. Doctors may use a variety of tests, including brain scans (CT and/or MRI), a spinal tap or lumbar catheter, intracranial pressure monitoring, and neuropsychological tests, to help them diagnose normal pressure hydrocephalus and rule out other conditions.

The symptoms described in this section account for the most typical ways in which progressive hydrocephalus manifests itself. It is, however, important to remember that symptoms vary significantly from individual to individual.

IDEA Category / DSM Category

IDEA: Other Health Impaired

DSM:

Long term Developmental Outcomes

The prognosis for patients diagnosed with hydrocephalus is difficult to predict, although there is some correlation between the specific cause of the hydrocephalus and the patient's outcome. Prognosis is further complicated by the presence of associated disorders, the timeliness of diagnosis, and the success of treatment. The degree to which decompression (relief of CSF pressure or buildup) following shunt surgery can minimize or reverse damage to the brain is not well understood.

Affected individuals and their families should be aware that hydrocephalus poses risks to both cognitive and physical development. However, many children diagnosed with the disorder benefit from rehabilitation therapies and educational interventions and go on to lead normal lives with few limitations. Treatment by an interdisciplinary team of medical professionals, rehabilitation specialists, and educational experts is critical to a positive outcome. Left untreated, progressive hydrocephalus is, with rare exceptions, fatal.

The symptoms of normal pressure hydrocephalus usually get worse over time if the condition is not treated, although some people may experience temporary improvements. While the success of treatment with shunts varies from person to person, some people recover almost completely after treatment and have a good quality of life. Early diagnosis and treatment improves the chance of a good recovery.

Assessment Approaches (Clinical Interview, Qualitative, Quantitative)

Hydrocephalus is diagnosed through clinical neurological evaluation and by using cranial imaging techniques such as ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), or pressure-monitoring techniques. A physician selects the appropriate diagnostic tool based on the patient's age, clinical presentation, and the presence of known or suspected abnormalities of the brain or spinal cord.

Interventions and Treatments

Hydrocephalus is most often treated with the surgical placement of a shunt system. This system diverts the flow of CSF from a site within the central nervous system (CNS) to another area of the body where it can be absorbed as part of the circulatory process.

A shunt is a flexible but sturdy silastic tube. A shunt system consists of the shunt, a catheter, and a valve. One end of the catheter is placed in the CNS - most usually within a ventricle inside the brain, but also potentially within a cyst or in a site close to the spinal cord. The other end of the catheter is commonly placed within the abdominal cavity, but may also be placed at other sites within the body such as a chamber of the heart or a cavity in the lung where the CSF can drain and be absorbed. A valve located along the catheter maintains one-way flow and regulates the rate of CSF flow.

A limited number of patients can be treated with an alternative procedure called third ventriculostomy. In this procedure, a neuroendoscope — a small camera designed to visualize small and difficult to reach surgical areas — allows a doctor to view the ventricular surface using fiber optic technology. The camera is guided into position so that a small hole can be made in the floor of the third ventricle, allowing the CSF to bypass the obstruction and flow toward the site of re-absorption around the surface of the brain.

Shunt systems are not perfect devices. Complications may include mechanical failure, infections, obstructions, and the need to lengthen or replace the catheter. Generally, shunt systems require monitoring and regular medical follow-up. When complications do occur, usually the shunt system will require some type of revision.

Some complications can lead to other problems such as over-draining or under-draining. Over-draining occurs when the shunt allows CSF to drain from the ventricles more quickly than it is produced. This over-draining can cause the ventricles to collapse, tearing blood vessels and causing headache, hemorrhage (subdural hematoma), or slit-like ventricles (slit ventricle syndrome). Under-draining occurs when CSF is not removed quickly enough and the symptoms of hydrocephalus recur.

Treatment of hydrocephalus for children or adults will likely involve a neurologist**,** neurosurgeon, obstetrician, pediatrician, and specialty nurses and physical therapists.

Contributions of the School Psychologist

Being aware of the possible academic difficulties that students with hydrocephalus may experience is a way that school psychologist may contribute to the educational success of these students. Most children with hydrocephalus are educated in mainstream education with other students, sometimes with extra help if they have any learning difficulties. Although most children with hydrocephalus are within the normal range of intelligence, most experience specific learning difficulties. Most hydrocephalus students appear interested and motivated to learn. However, they are easily distracted and find it difficult to sustain attention until the completion of work. Some have difficulty identifying the most salient aspect of a task and focusing their attention. Instead they tend to get distracted to less relevant aspects. Students with hydrocephalus are often described as sociable and talkative with good vocabulary skills. However, they may have difficulty monitoring what they say for logic, relevance or appropriateness. This poor comprehension may be difficult to identify when associated with articulate presentation. Some 'over talk', perhaps to compensate for their limited mobility or to conceal their inability to do what is asked, may be evident.

Immediate memory for auditory/verbal information may be intact, however, there is a rapid loss of information over time and difficulty in retrieving the appropriate bit of information from long term memory when it is needed. Students with hydrocephalus can learn effectively, but often take longer to learn and struggle with abstract concepts, for example, mathematics. Most students with hydrocephalus have difficulty with tasks requiring eye-hand coordination and motor planning skills. They may have difficulty with accurately interpreting what they see in terms of shape, size, space, distance and then correctly matching their movements (gross or fine). Some students may experience confusion differentiating between left and right. Complaints about slow and untidy handwriting are common and written tasks are often not fully completed. Most students with hydrocephalus find it hard to organize themselves, plan ahead and think flexibly. In addition, some may experience difficulty in understanding the passage of time or understand when matters are urgent. They may be unable to generate strategies for solving problems or to alter their approach if the first attempt is unsuccessful. They seem lost when confronted by a novel or multi-staged task and their work output falls off when they are expected to work independently.

Resources for parents, teachers and professionals

- Hydrocephalus Association www.hydroassoc.org
- The Hydrocephalus Foundation, Inc www.hydrocephalus.org
- National Hydrocephalus Foundation nhfonline.org
- MedlinePlus: Hydrocephalus www.nlm.nih.gov/medlineplus/hydrocephalus.html
- Mayo Clinic.com www.mayoclinic.com/health/hydrocephalus/DS00393
- National Institute of Neurological Disorders and Stroke
  www.ninds.nih.gov/disorders/hydrocephalus/hydrocephalus.htm