Hydrocephalus

- Author: Alberto J Espay, MD; Chief Editor: Howard A Crystal, MD  more...

Updated: Apr 27, 2010

Background

Hydrocephalus can be defined broadly as a disturbance of formation, flow, or absorption of cerebrospinal fluid (CSF) that leads to an increase in volume occupied by this fluid in the CNS.[1] This condition also could be termed a hydrodynamic disorder of CSF. Acute hydrocephalus occurs over days, subacute hydrocephalus occurs over weeks, and chronic hydrocephalus occurs over months or years. Conditions such as cerebral atrophy and focal destructive lesions also lead to an abnormal increase of CSF in CNS. In these situations, loss of cerebral tissue leaves a vacant space that is filled passively with CSF. Such conditions are not the result of a hydrodynamic disorder and therefore are not classified as hydrocephalus. An older misnomer used to describe these conditions was hydrocephalus ex vacuo.

Normal pressure hydrocephalus (NPH) describes a condition that rarely occurs in patients younger than 60 years.[2] Enlarged ventricles and normal CSF pressure at lumbar puncture (LP) in the absence of papilledema led to the term NPH. However, intermittent intracranial hypertension has been noted during monitoring of patients in whom NPH is suspected, usually at night. The classic Hakim triad of symptoms includes gait apraxia, incontinence, and dementia. Headache is not a typical symptom in NPH.

Benign external hydrocephalus is a self-limiting absorption deficiency of infancy and early childhood with raised intracranial pressure (ICP) and enlarged subarachnoid spaces. The ventricles usually are not enlarged significantly, and resolution within 1 year is the rule.

Communicating hydrocephalus occurs when full communication occurs between the ventricles and subarachnoid space. It is caused by overproduction of CSF (rarely), defective absorption of CSF (most often), or venous drainage insufficiency (occasionally).

Communicating hydrocephalus with surrounding "atrophy" and increased periventricular and deep white matter signal on fluid-attenuated inversion recovery (FLAIR) sequences. Note that apical cuts (lower row) do not show enlargement of the sulci, as is expected in generalized atrophy. Pathological evaluation of this brain demonstrated hydrocephalus with no microvascular pathology corresponding with the signal abnormality (which likely reflects transependymal exudate) and normal brain weight (indicating that the sulci enlargement was due to increased subarachnoid cerebrospinal fluid [CSF] conveying a pseudoatrophic brain pattern).

Noncommunicating hydrocephalus occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space, resulting in impairment of the CSF from the ventricular to the subarachnoid space. The most common form of noncommunicating hydrocephalus is obstructive and is caused by intraventricular or extraventricular mass-occupying lesions that disrupt the ventricular anatomy.[3]
Noncommunicating obstructive hydrocephalus caused by obstruction of the foramina of Luschka and Magendie. This MRI sagittal image demonstrates dilatation of lateral ventricles with stretching of corpus callosum and dilatation of the fourth ventricle.

Noncommunicating obstructive hydrocephalus caused by obstruction of foramina of Luschka and Magendie. This MRI axial image demonstrates dilatation of the lateral ventricles.

Noncommunicating obstructive hydrocephalus caused by obstruction of foramina of Luschka and Magendie. This MRI axial image demonstrates fourth ventricle dilatation.

Congenital hydrocephalus applies to the ventriculomegaly that develops in the fetal and infancy periods, often associated with macrocephaly. The most common causes of congenital hydrocephalus are obstruction of the cerebral aqueduct flow, Arnold-Chiari malformation or Dandy–Walker malformation. These patients may stabilize in later years due to compensatory mechanisms but may decompensate, especially following minor head injuries. During these decompensations, determining the extent to which any new neurological deficits may be due to the new acute event, compared with hydrocephalus that may have gone unnoticed for many years, is difficult.

Pathophysiology

Normal CSF production is 0.20-0.35 mL/min; most CSF is produced by the choroid plexus, which is located within the ventricular system, mainly the lateral and fourth ventricles. The capacity of the lateral and third ventricles in a healthy person is 20 mL. Total volume of CSF in an adult is 120 mL.

Normal route of CSF from production to clearance is the following: From the choroid plexus, the CSF flows to the lateral ventricle, then to the interventricular foramen of Monro, the third ventricle, the cerebral aqueduct of Sylvius, the fourth ventricle, the 2 lateral foramina of Luschka and 1 medial foramen of Magendie, the subarachnoid space, the arachnoid granulations, the dural sinus, and finally into the venous drainage.

ICP rises if production of CSF exceeds absorption. This occurs if CSF is overproduced, resistance to CSF flow is
increased, or venous sinus pressure is increased. CSF production falls as ICP rises. Compensation may occur through transventricular absorption of CSF and also by absorption along nerve root sleeves. Temporal and frontal horns dilate first, often asymmetrically. This may result in elevation of the corpus callosum, stretching or perforation of the septum pellucidum, thinning of the cerebral mantle, or enlargement of the third ventricle downward into the pituitary fossa (which may cause pituitary dysfunction).

The mechanism of NPH has not been elucidated completely. Current theories include increased resistance to flow of CSF within the ventricular system or subarachnoid villi; intermittently elevated CSF pressure, usually at night; and ventricular enlargement caused by an initial rise in CSF pressure; the enlargement is maintained despite normal pressure because of the Laplace law. Although pressure is normal, the enlarged ventricular area reflects increased force on the ventricular wall.

**Epidemiology**

**Frequency**

**United States**

The incidence of congenital hydrocephalus is 3 per 1,000 live births; the incidence of acquired hydrocephalus is not known exactly due to the variety of disorders that may cause it.

**International**

Incidence of acquired hydrocephalus is unknown. About 100,000 shunts are implanted each year in the developed countries, but little information is available for other countries.

**Mortality/Morbidity**

In untreated hydrocephalus, death may occur by tonsillar herniation secondary to raised ICP with compression of the brain stem and subsequent respiratory arrest.

Shunt dependence occurs in 75% of all cases of treated hydrocephalus and in 50% of children with communicating hydrocephalus. Patients are hospitalized for scheduled shunt revisions or for treatment of shunt complications or shunt failure. Poor development of cognitive function in infants and children, or loss of cognitive function in adults, can complicate untreated hydrocephalus. It may persist after treatment. Visual loss can complicate untreated hydrocephalus and may persist after treatment.

**Sex**

Generally, incidence is equal in males and females. The exception is Bickers-Adams syndrome, an X-linked hydrocephalus transmitted by females and manifested in males. NPH has a slight male preponderance.

**Age**

Incidence of human hydrocephalus presents a bimodal age curve. One peak occurs in infancy and is related to the various forms of congenital malformations. Another peak occurs in adulthood, mostly resulting from NPH. Adult hydrocephalus represents approximately 40% of total cases of hydrocephalus.

**Contributor Information and Disclosures**

**Author**

Alberto J Espay, MD  Assistant Professor, Department of Neurology, Gardner Family Center for Parkinson's Disease and Movement Disorders, Director of Clinical Research, University of Cincinnati

Alberto J Espay, MD is a member of the following medical societies: American Academy of Neurology and Movement Disorders Society

Disclosure: Boehringer-Ingelheim Consulting fee Board membership; Codman Grant/research funds Other; Medtronic Grant/research funds Other; Medtronic Grant/research funds Other; Allergan Grant/research funds Other; UCB-Schwarz Pharm Honoraria Speaking and teaching; Novartis Speaking and teaching; Solvay Consulting fee Board membership
Specialty Editor Board

**Anthony M Murro, MD**  Laboratory Director, Professor, Department of Neurology, Medical College of Georgia

Anthony M Murro, MD is a member of the following medical societies: American Academy of Neurology and American Epilepsy Society

Disclosure: Nothing to disclose.

**Francisco Talavera, PharmD, PhD**  Senior Pharmacy Editor, eMedicine

Disclosure: eMedicine Salary Employment

**Richard J Caselli, MD**  Professor, Department of Neurology, Mayo Medical School, Rochester, MN; Chair, Department of Neurology, Mayo Clinic of Scottsdale

Richard J Caselli, MD is a member of the following medical societies: American Academy of Neurology, American Association of Neuromuscular and Electrodiagnostic Medicine, American Medical Association, American Neurological Association, and Sigma Xi

Disclosure: Nothing to disclose.

**Selim R Benbadis, MD**  Professor, Director of Comprehensive Epilepsy Program, Departments of Neurology and Neurosurgery, University of South Florida School of Medicine, Tampa General Hospital

Selim R Benbadis, MD is a member of the following medical societies: American Academy of Neurology, American Academy of Sleep Medicine, American Clinical Neurophysiology Society, American Epilepsy Society, and American Medical Association

Disclosure: UCB Pharma Honoraria Speaking, consulting; Lundbeck Honoraria Speaking, consulting; Cyberonics Honoraria Speaking, consulting; Glaxo Smith Kline Honoraria Speaking, consulting; Ortho McNeil Honoraria Speaking, consulting; Pfizer Honoraria Speaking, consulting; Sleepmed/DigiTrace Speaking, consulting

Chief Editor

**Howard A Crystal, MD**  Professor, Departments of Neurology and Pathology, State University of New York Downstate; Consulting Staff, Department of Neurology, University Hospital and Kings County Hospital Center

Howard A Crystal, MD is a member of the following medical societies: American Academy of Neurology and American Neurological Association

Disclosure: Accera Pharmaceuticals Honoraria Consulting

References


http://emedicine.medscape.com/article/1135286-overview