Hydrocephalus in Children

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Abstract

The word "hydrocephalus" is derived from two Greek words: hydro meaning water; and cephalus meaning head; also known as "water on the brain". Historically it is believed to result from imbalance between CSF production and absorption, with net accumulation of fluid in the cranial cavity; characterized by increase in size of the cerebral ventricles. It is classified as: Communicating hydrocephalus, in which flow is not obstructed, but CSF is inadequately reabsorbed in the subarachnoid space and the Non-communicating hydrocephalus or the Obstructive type, in which flow of CSF from the ventricles to subarachnoid space is obstructed. This type may also be sub-classified into Congenital and Acquired. The overall incidence of hydrocephalus is not known. Approximately 55% of all hydrocephalus are congenital.

The etiology depends upon the age of the child. The clinical features are increase in the size of head, with wide anterior fontanelle, prominent scalp veins, sun-setting eyes, optic nerve atrophy, nystagmus and increased muscle tone in children upto 2 years. Children more than 2 years may present with these as hydrocephalus progresses; or if the fontanelles are closed, head size may be normal. These may present with optic atrophy or papilloedema, abnormal hypothalamic functions (short stature or gigantism, obesity, delayed puberty, primary amenorrhea or menstrual irregularity and diabetes inspidus) and spastic lower limbs. Performance IQ is worse than verbal IQ and learning problems are common.

The diagnostic procedures include measurement of head circumference, Plain X ray of head, Ventriculography, Pneumoencephalography, Ultrasonography, Computed Tomography and Magnetic Resonance Imaging. The management may be non-surgical and surgical.

Introduction

Hydrocephalus is a condition in which excess fluid accumulates in the brain. It is often referred to as "water on the brain," the "water" is actually cerebrospinal fluid (CSF) -- a clear fluid surrounding the brain and spinal cord.[1] Pathophysiologically, hydrocephalus is regarded as an imbalance in the formation and absorption of cerebrospinal fluid (CSF) to a sufficient magnitude producing accumulation of fluid leading to an elevation of intracranial pressure. Compensatory adjustments especially in very young and very old subjects may occur that may reduce prevailing CSF pressure to normal range. Thus, hydrocephalus must be carefully distinguished from cerebral atrophy, in which an excessive accumulation of CSF within the intracranial cavities is due to loss of cerebral substance rather than a primary defect in CSF formation or absorption.[2]

Cerebrospinal Fluid (CSF) formation

CSF is mainly formed within the ventricular system. Formation sites include choroid plexus, ependyma and parenchyma. Most of the CSF is formed by the choroids plexus of lateral ventricles. The rate of formation is 0.35-0.40 ml/min equivalent to 500 ml per day, the rate being same in paediatric and adult population.[3]

CSF Pathways

Lateral Ventricles -- Intraventricular foramen of Monro -- 3rd Ventricle -- Aqueduct of Sylivus -- 4th Ventricle -- Foramen of Luska and Magendie - subarachnoid space and Spinal Canal.[4]

Pathophysiology

The large head attributed to water on the brain has long attracted interest and speculation. Hypothetically, hydrocephalus can be subdivided into 3 forms:[5-7]

* Disorders of cerebrospinal fluid production: This is the rarest form of hydrocephalus. Choroid plexus papillomas and choroid plexus carcinomas can secrete cerebrospinal fluid in excess of its absorption.

* Disorders of cerebrospinal fluid circulation: This form of hydrocephalus results from obstruction of the pathways of cerebrospinal fluid circulation. This can occur at the ventricles or arachnoid villi. Tumors, hemorrhages, congenital malformations, and infections can cause obstruction at either point in the pathways.

* Disorders of cerebrospinal fluid absorption: Conditions, such as the superior vena cava syndrome and sinus thrombosis, can interfere with cerebrospinal fluid absorption. Some forms of hydrocephalus cannot be classified clearly. This group includes normal pressure hydrocephalus and pseudotumor cerebri.

Classification

In the very ancient times, Hippocrates recognized accumulation of water in the head and the cavities of brain as the cause of enlarged head. Two hundred years later, Magendie and Lusckha described the communication between ventricular system and subarachnoid spaces. Later, Key and Retzuis in 1872, Dandy and Blackfen in 1913 and 1914 and Weed in 1920, demonstrated that this can result due to obstruction of ventricular system. Thus, hydro-

cephalus was divided into two types: Obstructive due to blockage; and Communicating secondary to decreased absorption over the surface of brain.[8]

The classification being used presently is as follows:

1. Non-communicating or obstructive hydrocephalus in which flow of CSF from the ventricles to subarachnoid space is obstructed. Thus there is no communication between the ventricular system and the subarachnoid space. The commonest cause of this category is aqueduct blockage.

2. Communicating or non-obstructive hydrocephalus in which flow is not obstructed, but CSF is inadequately reabsorbed in the subarachnoid space. Thus there is communication between the ventricular system and the subarachnoid space. The commonest cause of this group is post-infective and post-haemorrhagic hydrocephalus.[9,10] This type may also be sub-classified into Congenital and Acquired. Approximately 55% of all hydrocephalus are congenital. The reported incidence of primarily congenital hydrocephalus is 0.9 to 1.5 per 1000 births and those occurring with spina bifida and myelomeningocele varies from 1.3 to 2.9 per 1000 per births.[11]

Etiology

The etiology differs in age group up to 2 and beyond 2 years. Table 1 presents etiologies according to age groups.[12-14]

Table 1. Etiology of Hydrocephalus.

0 to 2 Years

1) Intra uterine infections
    -Anoxic
    -Bacterial, granulomatous, parasitic
    -Traumatic perinatal heamorrhage
2) Neonatal, bacterial or viral meningio-encephalitis
3) Arachnoid cyst
4) Intracranial tumours
5) Arterivenous malformation of the galenic system
6) Post infectious (bacterial and granulomatous meningitis)
7) Development disorders
    -Aqueductal stenosis
    -Myelomeningiocoele
    -Dandy-walker cyst
    -Encephalocoele

More than 2 to 12 years

1) Mass lesion compressing the ventricular system
    -Craniopharyngioma
    -Pineal tumor
2) Posterior fossa tumor
    -Medulloblastoma
    -Astrocytoma

    -Ependymona
3) Deveplomental disorders
    -Aqueductal stenosis
    -Arnold-chiari malformation
4) Post infectious
    -Meningitis (bacterial and granulamatous)
5) Post haemorrhagic

Clinical features

0-2 years

Before 2 years, the head enlarges excessively because the cranial sutures are open. This enlargement is almost invariably the presenting sign.[12-14]

1) **Shape of Head:** An abnormal head shape may suggest the diagnosis. Occipital prominence is seen in Dandy walker malformation. A disproportionately large forehead is common with aqueductal stenosis. The enlarged cranial size is quite evident and helpful in diagnosis.

2) **Anterior Fontanelle:** Normally the anterior fontanelle is small, depressed in a relaxed sitting patient but with hydrocephalus, it is enlarged and full even when the infant is quiet and upright.

3) **Sutures:** palpation of spreading sutures also help in the diagnosis.

4) **Percussion Note:** The percussion note of a normal infant skull is of a "Crack pot", while with hydrocephalus it starts resembling a watermelon. This sound is particularly striking when examiner places his ear against the infant's skull while percussing.

5) **Scalp Veins:** The scalp veins are usually prominent, particularly in crying infants. The prominence is caused by compression of basal venous outlets by increased pressure, which results in shunting of blood through the valveless collateral system into easily distended scalp veins.

6) **The Eyes:** As the hydrocephalus progresses, the eyes are displaced downward by pressure on the thinned orbital roofs. This displacement of eyes causes the sclera to be visible above the iris, termed as the "setting sun" sign.[15]

7) **Cranial Nerves:** Optic atrophy is a common finding in advanced hydrocephalus due to the compression of the optic chiasma and the optic nerve by dilated anterior third ventricle and increased intracranial pressure (ICP). Abducent nerve paresis secondary to stretching is common. Nystagmus and random eye movement may be present. Vision can also be affected by damage to occipital cortex by grossly dilated occipital horns.[15]

8) **Muscle Tone and Deep Tendon Reflexes:** With progressive hydrocephalus, the deep tendon reflexes and muscle

tone in lower extremities are increased.

9) Growth Retardation: In hydrocephalic children, growth failure and delayed neurological development are common. Head and trunk control is particularly affected.[16]

More than 2 to 12 years

These patients fall into two groups differentiated by the presenting clinical features:[12-14]

1) The first group includes those children who have preexisting (infantile) unrecognized, progressive hydrocephalus with normal or retarded neurological development. They are usually diagnosed after an incidental head injury leading to rapid deterioration of neurological functions. They usually have slightly enlarged heads with optic atrophy or papilloedema, abnormal hypothalamic functions (short stature or gigantism, obesity, delayed puberty, primary amenorrhea or menstrual irregularity and diabetes inspidus) and spastic lower limbs.[15] Performance IQ is worse than verbal IQ and learning problems are common.[16]

2) The second group consists of children who develop hydrocephalus after closure of cranial sutures; therefore, head circumference is usually within normal limits. Papilloedema is a common finding due to raised ICP. Abducent nerve paresis and spasticity of lower extremities with hyper reflexia are also noted. Morning headaches and vomiting are common.[15]

Diagnostic Procedures

1) **Head Circumference:** The head size should be measured by taking maximal obtainable circumference with measuring tape. The circumference is plotted on a growth chart having head circumference column. This procedure is of greatest importance in demonstrating an excessive rate of growth by serial measurements.[12-14]

2) **Fusion of Sutures:** Progressive hydrocephalus beginning before closure of cranial sutures, prevents the fusion of sutures leading to continuous excessive head enlargement.[12-14]

3) **Plain X rays:** This modality is of great diagnostic value and will confirm many clinical findings, such as an enlarged head, crainofacial disproportion, wide spread sutures and large anterior fontanelle. Small posterior fossa with low position lumboid sutures in aqueductal stenosis or large posterior fossa in Dandy walker malformation may be found. In older children elongated inter digitations of suture line indicates increased ICP. There may be evidence of intracranial convolutional markings (silver beaten appearance) and demineralization of dorsum sella.[17]

4) **Ventriculography:** It is an invasive procedure in which the dye is injected into the ventricular system through lum-

bar puncture in order to see the size of ventricles and flow of dye on plain x-rays. It has been replaced by ultrasound and CT scan.

5) **Pneumoencephalography:** It is also an invasive technique similar to ventriculography, only difference is that here air is injected into the cerebral ventricles. This has also been replaced by non-invasive procedures.

6) **Ultrasonography:** It is a non-invasive procedure used only in patients in whom the anterior fontanelle is open.

7) **Computed Tomography:** CT scan superceded other invasive investigations like Ventriculography and Pneumoencephalography. It has a major role in accurate assessment of ventricular size, extracerebral spaces and site of obstruction.

8) **Magnetic Resonance Imaging (MRI):** This is also a noninvasive investigation.[17] It may also be used in antenatal diagnosis of Hydrocephalus.

Management

In early days when methods for diagnosis and treatment of hydrocephalus were unsatisfactory, a number of patients died due to inadequate treatment.

Non Surgical Management

Drugs such as Isosorbide which produce hyperosmotic diuresis and those such as acetazolamide which decrease the secretion of CSF may be treated as temporary management of clinical situation. Their brief action and side effects preclude their prolonged use in the definitive treatment of hydrocephalus. Wrapping of the infant's head was started as a treatment of hydrocephalus, the object being to force CSF into alternative absorptive pathways. This produces raised intracranial pressure and may cause progressive dilatation of central canal of spinal cord and resulting possibly in impairment of the cord function. It has now been abandoned.[18]

Surgical Management

The era of modern treatment of hydrocephalus began in early sixties, when Spitz, an engineer along with Holter, introduced pressure sensitive flow regulating valve for diversion of CSF from obstructed ventricles into the right atrium of the heart. These shunts have been subsequently refined and are now universally employed in the management of hydrocephalus.[19]

The surgical procedures used in modern times for relief of hydrocephalus have been exceedingly varied. Following procedures were tried before the definitive treatment the hydrocephalus evolved:[20]

Procedures designed to reduce CSF formation:

1) Excision of Choriod Plexus

2) Cauterization of Choriod plexus

Procedures for decompression of ventricles:

1) **Repeated Ventricular Puncture:** Keen is said to conclude the site of ventricular tap in present era. Later, Chipault, Chater and Grantham tried several alternatives for repeated ventricular punctures but this procedure failed due to a number of complications.

2) **Open Ventricular Drainage:** Wernicke in 18th century tried external ventricular drainage but due to high infection rate it was also abandoned.

3) **Closed Ventricular Drainage:** In order to avoid infection, many methods of internal drainage were devised. The modern surgical treatment had its origin in last decade of 19th and 1st decade of 20th century. Most of the procedures now in use such as peritoneal and circulatory shunting of CSF were conceived and tried at that time. Excellent reviews of surgical treatment of hydrocephalus have been published in journals by Dandy and Blackfan (1914)[21], Davidoff (1929)[20], Jackson (1951)[22], Scarff (1963)[23] and Balasubramanian (1967).[24] The sites for diversion of CSF from the cerebral ventricles is presented in Table 2.[20]

Table 2. Sites for diversion of CSF from the Cerebral Ventricles.

Structures of the Head and Neck:

* Subaponeurotic layers of the scalp
* Intracranial Venous Sinuses - Scalp and neck veins
* Subdural Space
* Mastiod Antrum
* Salivary Gland Ducts
* Thoracic Duct

Structures of the Spine:
* Spinal Epidural Space
* Bone Marrow

Structures of the Thorax:
* Plueral Cavity
* Right Atrium

Structures of the Abdomen:
* Peritoneal Cavity
* Omental Bursa
* Stomach
* Gallbladder
* Ileum
* Ureter
* Fallopian Tubes

Recent Techniques

Operation that bypass obstruction in the ventricular system:

* Third Ventriculostomy[25]

* Cannulation of the aqueduct[26]

Third ventriculostomy

This procedure is reserved for obstructive cases in patients who have normal or near normal spinal fluid absorptive capacity. A blunt instrument is used to penetrate the floor of the third ventricle. Sharp instruments or lasers can cause vascular injury. Leaving a clamped drain in place postoperatively might be prudent. The burr hole placed on the coronal suture allows a straight trajectory to the foramen of Monro. Stereotactic guidance is not needed if endoscopic techniques are used.[25]

Indications for Shunt Surgery

The indication of surgical intervention are related to intracranial hypertension, neurological dysfunction, evidence and degree of ventricular dilatation, the presence or absence of pathological lesion, the nature and the location of obstruction. Some surgeons insert the shunt in any patient with large ventricles while others restrict it to those with potentially reversible deficit or progressive deterioration. Progressive ventriculomegally on CT scan combined with observation of developmental deficits in infants or intellectual and motor disability in older children are a few criteria.[27]

Ventriculo peritoneal shunting

This is by far the most common procedure for cerebrospinal fluid diversion. The abdomen should be able to absorb fluid. The ventricular catheter can be placed more reliably from the coronal approach. Some surgeons still prefer parieto-occipital catheters. The proximal catheter tip should lie anterior to the choroid plexus in the frontal horn of the lateral ventricle.[19]

Ventriculo-atrial shunting

This procedure usually is the first choice for patients who are unable to have abdominal distal catheters (e.g. multiple operations, recent abdominal sepsis, and known malabsorptive peritoneal cavity). The procedure carries more risk and long-term complications are more serious (eg, renal failure, great vein thrombosis). Fluoroscopic guidance is necessary to prevent catheter thrombosis (short distal catheter) or cardiac arrhythmias (long distal catheter).[28,29]

Postoperative care

Postoperatively, patient is kept in horizontal position for 12-24 hours and then gradually elevation of head is allowed depending upon condition of anterior fontanelle. Oral

intake is initiated within 12 hours if there is no abdominal distension and peristalsis is audible. Follow up includes regular fortnightly visits for first three months and then after every month up to age of one year. From age of 1-3 years, 6 monthly visits and after that annual visits are required.[19]

Post-operative complications

The complications depend on the type of shunt and the underlying pathophysiology. Infection is the most feared complication in the young age group. The overwhelming majority of infections occur within 6 months of the original procedure. Treatment of infected shunts with antibiotics alone is not recommended because bacteria can be suppressed for extended periods and can resurface once antibiotics are stopped.[30-34]

Subdural haematomas occur almost exclusively in adults and children with completed head growth. Incidence of subdural hematomas can be reduced by slow postoperative mobilization. Shunt failure is mostly due to sub optimal proximal catheter placement. Occasionally, distal catheters fail. Headaches are caused by over draining. Peritoneal cyst (Excess fluid collection in peritoneal cavity) may occur because of poor absorption.[30-34]

Prognosis

Hydrocephalus is usually a lifelong disorder. Prognosis depends on a number of factors, including the underlying condition, its duration and degree, as well as response to treatment. The mortality rate in shunt-treated pediatric patients with hydrocephalus remains high, dependent on the underlying cause for shunt insertion and the subsequent development of infection and other complications related to shunt apparatus.[35,36]

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