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HYDROCEPHALUS: OVERVIEW AND PERSPECTIVES

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ABSTRACT

Cerebrospinal fluid (CSF) is produced by choroid plexus, circulates in the subarachnoid space and the resorbtion occurs at the level of arachnoid granulation. Ventricular enlargement appears because of: 1exces of CSF production, 2. altered CSF flow or resorbtion, 3. secondary post-traumatic, and 4. post-aneurysmal SAH, with or without symptoms or neurologic impairment. Hydrocephalus (HY) (count in infants 3-5 / 1.000 new-born) represents the excess of CSF in ventricles or subarachnoidal space and is commonly associated with meningomyelocele (MMC) & acqueductal stenosis. The most often clinical syndrome which occurs in hydrocephalus is Intracranian Hypertension syndrome, which consists in headache, vomitting and papilary edema. The diagnostic protocol is represented by neurological examination, ophtalmological examination, CT scan and MRI. Purposes of surgical procedures are to decrease ICP, to re-expand the brain and to prevent neuronal destruction. The main surgical procedures are endoscopic third ventriculostomy (ETV), ventriculoperitoneal shunt and ventriculoatrial shunt.

Key words: hydrocephalus, CT scan, endoscopic third ventriculostomy (ETV), ventriculoperitoneal shunt; child

HISTORICAL DATA

Hippocrates (460–370 BC) used the term "hydrocephalus" for the first time and Albucasis (AD 936–1013), Cordoba, first described hydrocephalic newborns. Vesalius, (1514-1564) made the first scientific description of internal hydrocephalus and Thomas Willis in 1664 introduced the concept that choroid plexuses produced cerebrospinal fluid (CSF).

In 1914 Dandy and Blackfan published their classic paper, confirming the production of CSF by the choroid plexus and diffuse absorption by the pia-arachnoid veins. In 1922, Dandy W.E. described an original surgical procedure for hydrocephalus: Endoscopic plexectomy, 1922.

INTRODUCTION

Cerebrospinal fluid (CSF) is produced by choroid plexus, circulates in the subarachnoid space and the resorbtion occurs at the level of arachnoid granulation. Hydrocephalus represents a progressive disease caused by the abnormal rise in CSF volume and, usually, pressure, that results from an imbalance of CSF production and absortion.

The incidence of hydrocephalus (HY) counts in infants approximately 3-5 cases per 1.000 new-born. Commonly HY associates with meningomyelocele (MMC) and acqueductal stenosis (post hemorrhagic and post meningitis). In 10-30% cases, the HY are of unknown etiology. The exact incidence of acquired types of hydrocephalus is unknown.

The etiology of hydrocephalus in congenital cases is unknown. Few cases, less than 2%, are inherited (X-linked hydrocephalus). The most common causes of hydrocephalus in acquired cases are tumor obstruction, trauma, intracranial hemorrhage and infection.

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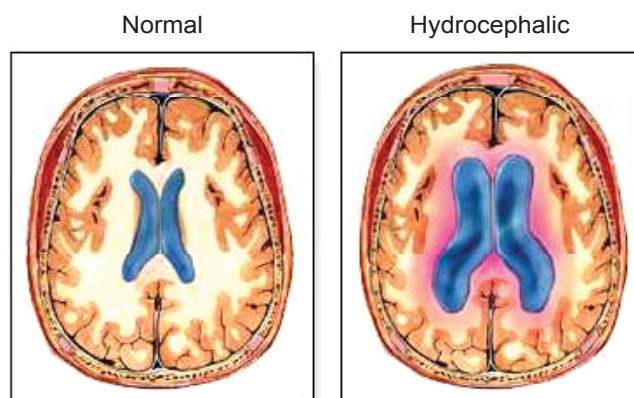
FIGURE 1. *Schematic drawing of normal ventricles versus hydrocephalic ventricles.*

PATHOPHYSIOLOGY AND HYDROCEPHALUS CLASSIFICATION

Hydrocephalus can be divided in the following forms:

- Disorders of CSF circulation. This form results from obstruction of the pathways of CSF circulation. This can occur at the ventricles or any other point within the CSF space up to the arachnoid villi. Tumors, hemorrhages, congenital malformations, such as aqueductal stenosis, and infections can obstruct at either point in the pathways.
- Disorders of CSF absorbtion. Conditions, such as cerebral sinus thrombosis, but also the superior vena cava syndrome, meningitis, and subarachnoid hemorrhage can interfere with CSF absorbtion.
- Disorders of CSF production. This is the rarest form of hydrocephalus. Choroid plexus papillomas and choroid plexus carcinomas can secrete CSF in excess of its absorption capacity.
- Unclassified or not well understood disorders of CSF dynamics. Some forms of hydrocephalus cannot be classified clearly. This group includes e.g. normal pressure hydrocephalus. Besides enlargement of the ventricles, disturbed CSF dynamics can also lead to other diseases like idiopathic intracranial hypertension, syringomyelia and Tarlov's cysts.

CLINICAL SYMPTOMATOLOGY

The various types of hydrocephalus will present differently in different age groups. Acute forms of hydrocephalus typically presents with headache, gait disturbance, vomiting, and visual changes in older children and adults. However, in infants, irritability and poor head control can be the only

and early signs of hydrocephalus. When the third ventricle dilates, the patient can present with Parinaud syndrome- upgaze palsy with a normal vertical Doll response- or the setting sun sign- Parinaud syndrome with lid retraction and increased tonic downgaze. Occasionally, a focal deficit, such as sixth nerve palsy, can be the presenting sign. Papilledema is often present, although it may lag behind symptomatology. Infants present with bulging fontanelles, dilated scalp veins, and increasing head circumference. When advanced, hydrocephalus presents with brainstem signs, coma, and hemodynamic instability.

Normal pressure hydrocephalus has a very distinct symptomatology. The patient is older and presents with progressive gait apraxia, incontinence, and dementia. Although this triad of symptoms usually defines normal pressure hydrocephalus, in many cases it remains difficult to unequivocally diagnose NPH clinically, based upon this triad of symptoms or even with imaging studies.

FIGURE 2. *Child with severe hydrocephalus*

NEUROIMAGING STUDIES

1. CT scan of the head delineates the degree of ventriculomegaly and, in many cases, the etiology. When performed with contrast, it can show infection and tumors that cause obstruction. It also helps with operative planning. Ventricles are usually dilated proximal to the point of obstruction. In idiopathic intracranial hypertension, the CT scan findigs are usually normal.

2. Perform MRI scan of the head in most, if not all, congenital cases of hydrocephalus. This delineates the extent of associated brain anomalies such as corpus callosum agenesis, Chiari malformation, disorders of neuronal migration, and vascular malformations. Some tumors, for example the midbrain tecal gliomas, only can be detected with this study. T2-weighted images can show transependymal egress of CSF.

3. Fetal and neonatal cranial ultrasound is a good study for monitoring ventricular size and intraventricular hemorrhage in the neonatal ICU setting. Certainly, prior to treatement, one should perform other imaging studies.
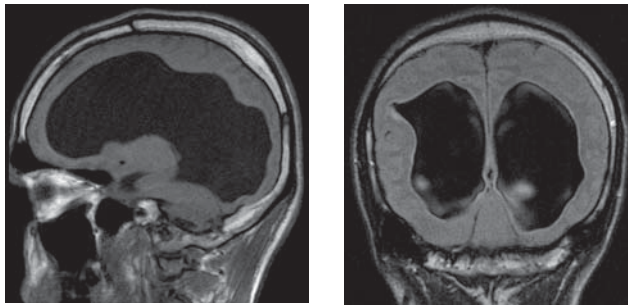

FIGURE 3. *MR aspect of hydrocephalus*

DIAGNOSTIC PROCEDURES

Lumbar puncture can be used to measure intracranial pressure, but it should only be performed after imaging studies rule out an obstruction. A diagnostic high-volume lumbar puncture in normal pressure hydrocephalus can assist in making decisions regarding shunting. Spinal fluid can show the type and severity of infection.

TREATMENT

Most cases of symptomatic hydrocephalus need to be treated before permanent neurologic deficits result or neurologic deficits progress.

When an etiologic factor is known, hydrocephalus can be treated with temporary measures while the underling condition is treated. Examples of temporary treatment measures are ventriculostomy until a posterior fossa tumor is resected or lumbar puncture in a neonate with intraventricular hemorrhage until the blood is resorbed and normal CSF absorption resumes.

Medical Therapy

Medical therapy is usually a temporizing measure. In transient conditions, such as sinus occlusion, meningitis, or neonatal intraventricular hemorrhage, medical therapy can be effective.

- Acetazolamide 25mg/kg/d in 3 dose. Careful monitoring of respiratory status and electrolytes is crucial. Treatment beyond 6 month is not recommended.
- Furosemide 1 mg/kg/d in 3 doses. Again, electrolyte balance and fluid balance need to be monitored carefully.
- Lumbar punctures. In neonates recovering from intraventricular hemorrhage, serial lumbar punctures can, in some cases, resolve hydrocephalus. If possible, this is the preferred method of treatment.
- Removal of the underlying cause usually resolves hydrocephalus.

Surgical Therapy

As performance of CSF diversion- most often ventriculoperitoneal shunting- has increased in frequency, so has awareness of pitfalls of the procedure. Recently, a resurgence of interest in third ventriculostomies has occurred.

Preoperative Details

Make every effort to identify the cause of hydrocephalus prior to considering a diversion procedure. Do not consider an indwelling distal catheter in patient with active infection or high CSF protein > 150 mg/dl. Obtain some idea of brain compliance in order to select the optimum valve pressure and decide if the pressure- programmable valve should be used. Use one dose of preoperative prophylactic antibiotics during induction of anesthesia.

Intraoperative Details

Endoscopic Third Ventriculostomy (ETV). Reserve this procedure for obstructive cases in patients who have normal or near-normal spinal fluid absorptive capacity. Use a blunt instrument to penetrate the floor of the third ventricle. Sharp instruments or lasers can cause vascular injury. Leaving a clamped drain in place postoperatively is still a matter of debate but might be prudent in some cases.The burr hole is placed just in front of the coronal suture which allows a straight trajectory to the foramen of Monro. Stereotactic guidance is not needed if endoscopic techniques are used and the ventricles are enlarged. Other indications to use neuro-endoscopy in case of hydrocephalus include:

- dilating or stenting an obstructed aqueduct
- direct positioning of a proximal shunt catheter
- fenestration of intraventricular membranes
- opening of the septum pellucidum in unilateral obstruction of the foramen of Monro
- biopsy or excision of intraventricular tumours
- fenestration of intra- and paraventricular arachnoid cysts, and marsupialisation of neuro-epthelial cysts.

But certainly indications are still evolving.

Ventriculoperitoneal shunting. This procedure is still by far the most common procedure for CSF

diversion. The abdomen should be able to absorb the excess spinal fluid. Either 1 of 2 major locations for the burr holes are typically used. The ventricular catheter can be placed more reliably from the (right) frontal approach. Some surgeons still prefer parietooccipital catheters. The proximal catheter tip should lie anterior to the choroid plexus in the frontal horn of the lateral ventricle when the parietooccipital approach is used. In difficult cases, placement under neuronavigation guidance should be consider whenever avaible.

Ventriculoatrial shunting. This procedure is usually the first choice for patients who are unable to have distal abdominal catheters (eg, multiple operations, recent abdominal sepsis, known malabsorbative peritoneal cavity, abdominal pseudocyst). The procedure carries more risk. Long-term complications are more serious (eg, renal failure, vena cava thrombosis). Fluoroscopic guidance is necessary to prevent catheter thrombosis (short distal catheter) or cardiac arrhythmias (long distal catheter).

Ventriculopleural shunting. Reserve this procedure for patients with failed peritoneal and atrial shunts.

Torkildsen shunts or internal shunts are straight tubes that communicate to CSF spaces without a valve, usually from the occipital horn to the cistern magna. Their effectiveness and long-term efficacy are not proven.

Lumboperitoneal shunts are used in communicating hydrocephalus, especially if ventricles are small. Idiopathic intracranial hypertension (IIH) is the classical indication of this method of shunting. A positional valve is helpful because it turns off the flow of CSF when the pacient is upright, thereby preventig overdrainage headache.

FIGURE 4.
Schematic drawing of ventriculoperitoneal and ventriculoatrial shunt
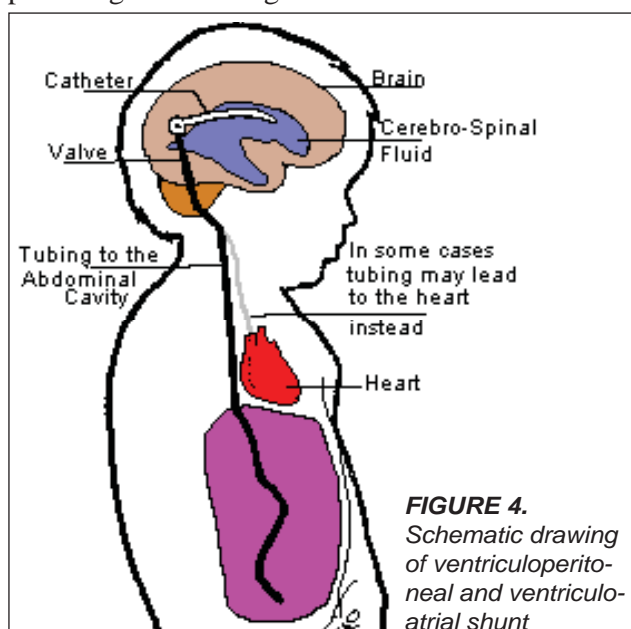
Contraindication

Cases in which treatment should not be implemented include the following:
- Children with hydranencephaly
- Ventriculomegaly of senescence; the patient who does not have the symptom triad
- Ex vacuo hydrocephalus which represent the replacement of lost cerebral tissue with cerebrospinal fluid
- Arrested hydrocephalus is defined as a rare condition in which the neurological status of the patient is stable in the presence of stable ventriculomegaly.
- Benign hydrocephalus of infancy is found in neonates and young infants. The children are asymptomatic, and head grow is normal. CT scan shows mildly enlarged ventricules and subarachnoid spaces.

Postoperative follow-up

Plain radiographs of the entire hardware system confirm good position and serve as excellent baseline studies for the future. Immediate postoperative CT scan is used to document ventricular size, and then performs CT scan for base-line at 2-4 weeks after shunting. In cases with third ventriculostomy, the MRI is performed for base-line at 8-12 weeks after surgery. Children with shunt or third ventriculostomy should be monitored every 6-12 months. Carefully monitor head growth in infants. Check distal tubing length with plain radiographs when child grows.

COMPLICATIONS

The complications depend mainly on the kind of procedure and the underlying pathophysiology. Bleeding is the most feared complication of ETV, although very rare. If it is due to damage to the basilar artery, this is usually fatal. Venous bleeding from puncturing the ependyma or damaging the choroid plexus will eventually stop with prolonged rising. Other complications include damage to the hypothalamus and infundibulum, subdural hematoma and CSF leakage or pseudomeningocele formation.

In the case of shunting in young age, infection is the most feared complication. The majority of infections occur within 6 months of the original procedure. Early infections occur more frequently in neonates and are associated with more virulent bacteria such as Escherichia Coli. Infected shunts need to be removed, CSF needs to be sterilized and

a new shunt needs to be replaced. Subdural hematomas occur almost exclusivelly in adults and children with completed head growth. The treatment is drainage and may require temporary oclusion of the shunt.

Shunt failure is mostly due to suboptimal proximal catheter placement and less frequent due to distal tube failure.

Overdrainge is more common in lumboperitoneal shunts and manifests with hedache in the upright position. In most overdrainage is a self-limiting process, in other cases the revision to a higher-pressure valve may be required.

Slit ventricle syndrome is a rare condition in which brain compliance is unusually low. It mostly occurs in the setting of prior ventriculitis or shunt infection. The slit ventricle syndrome does not imply overdrainage, and the symptoms usually are those of high pressure rather than low pressure. ETV has been described as a treatment for slit-ventricles but it is technically much more difficult because of small size of the ventricles.

FUTURE IN HYDROCEPHALUS

Hydrocephalus research has advanced in the last decades. Examples are the development of new shunt materials and, more recently, programmable and gravitational valve technology. Current research categories include the following:

- Transplantation of tissue, such as vascularized omentum to reestablish the normal CSF circulation
- Endoscopic third ventriculostomy have eliminated the need for shunting in noncomunicating cases of hydrocephalus in older children and adults. New optics and smaller scopes have expanded this field in the last decade
- Characterisation of multiple transgenic mouse models has highlighted the importance of the secretory ependymal cells of the subcommisural organ as the – key factor in development of congenital hydrocephalus and many more areas are now under research

REFERENCES

1. **Arseni C, Lenke Horvath, Ciurea AV** – Hidrocefalia sugarului În: Lichidul cefalo-rahidian, sub redacția Arseni C, Chimion D, Editura Didactică şi Pedagogică, Bucureşti, 1979
2. **Ciurea AV, Coman TC, Mircea D** – Postinfectious hydrocephalus (Cap 15). In: Cinalli G, Maixner WJ, Sainte-Rose C (eds) Pediatric Hydrocephalus, Springer Verlag, Milano, Berlin, Heidelberg, New York, Hong Kong, London, Paris, Tokyo. pp:201-218, 2004
3. **Ciurea AV, Constantinovici A** – Patologia neurochirurgicală. În: Tratat de patologie chirurgicală, sub redacția N. Angelescu, Editura Medicală, Bucureşti, vol. II, pp.: 2439 – 2615, 2001
4. **Ciurea AV** – Hidrocefalia sugarului şi copilului. În: Tratat de Pediatrie, sub redacția Prof. Dr. E.P. Ciofu, Dr. Carmen Ciofu, Ed. Medicală, 2001
5. **Dandy WE** – An operative procedure for hydrocephalus John Hopkins Hosp Bull 33:189-196; 1922 Dandy WE
6. **Fritsch MJ, Mehdorn M** – Endoscopic intraventricular surgery for treatment of hydrocephalus and loculated CSF space in children less than one year of age. *Pediatr Neurosurg.* 36:183-188, 2002
7. **Horvath Lenke, Ciurea AV** – Malformaţiile congenitale ale sistemului nervos În: Tratat de Neurologie, vol. III, sub redacția Arseni C, Editura Medicală, Bucureşti, 1981
8. **Matson DD** – Hydrocephalus. *Clin Neurosurg* 13:324, 1965
9. **Matson DD** – A new operation for the treatment of communicating hydrocephalus. Report of a case secondary to generalized meningitis. *J Neurosurg,* 6:238-247, 1949
10. **World Federation of Neurosurgery,** Essential in Neurosurgery, 2010