

HYDROCEPHALUS

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DEFINITION

• Hydrocephalus is derived from a combination of two Greek words: (hydro) which means water and (cephalous) which means head. So, the term means water in the head. But actually, it is not water, but it is CSF and it is not in the head but within the ventricular system. So, the term means dilatation of the ventricular system.

ANATOMY

 The ventricular system is made of four ventricles; two lateral ventricles, situated one in each cerebral hemisphere, and a midline situated third ventricle. Each lateral ventricle is connected to the third ventricle by a foramen of Monro. The third ventricle has one exit to the aqueduct of Sylvius, which runs all the way through the brain stem until it opens into the roof of the fourth ventricle. The fourth ventricle is situated in the midline in the posterior fossa. It has a floor and three exits; one actual foramen in the midline posteriorly called the foramen of Magendie and two lateral meshes of openings termed loosely as the foramina of Luschka.

PHYSIOLOGY

- The CSF is produced by the choroid plexus, which is located mostly in the lateral ventricle, and in the 4th ventricle. A small amount is produced by the ependymal lining of the ventricles themselves, and still some of the CSF in the spinal theca is produced by the dura of the nerve root sleeves.
- Once the CSF exits the fourth foramen it enters the subarachnoid space (SAS), circulates around the whole brain surface to gain access to the arachnoid granulations where it is absorbed into the superior sagittal sinus (SSS). Some CSF enters the spinal theca and circulates around the spinal cord. Still a smaller amount enters the central canal of the spinal cord itself.

PHYSIOLOGY

 The amount of the CSF in the body is about 150 mls. The ventricles contain 25 mls, the rest (125 mls), is in the subarachnoid space. CSF is produced at the rate of almost 500 mls. per day. The CSF pressure is taken as the intracranial pressure and measured in mms of CSF; it ranges between 150 and 180. However, this pressure varies and it is almost zero in the newborn

Hydrocephalus will result from either one of three conditions:

- Over production as seen in **choroid plexus papilloma**.
- Obstruction within the ventricular system
- Diminished absorption at the arachnoid granulations

 The over production of CSF is a rare condition in which the benign tumor of papilloma of the choroid plexus produces large amounts of CSF resulting in dilatation of the ventricular system. Excising the papilloma usually treats the condition

- **Obstruction** within the ventricular system could result from:
- Congenital causes:
 - Usually in the area of the aqueduct of Sylvius as in the cases of aqueduct stenosis due to **gliosis** or **forking**, or
 - In the area of the exits of the 4th ventricle, as in atresia of the foramina, called (Dandy Walker malformation).
 - Chiari type1 and Chiari type 2 malformations.
- Non-congenital causes:
 - Masses, as **in colloid cyst** of the 3rd. ventricle, or **medulloblastoma** in the posterior fossa.
 - Arteriovenous malformations.

- **Diminished absorption** may result from:
- Fibrinous deposits due to meningitis.
- Fibrinous deposits due to subarachnoid hemorrhage (SAH).

TYPES

 Communicating hydrocephalus, when the problem lies at the level of the arachnoid granulation, and the ventricular system and its exits are free from obstructing agent. Here all the four chambers of the ventricular system are dilated and communicate with each other.



TYPES

• Non-communicating; when there is an obstruction within the ventricular system or at its exits into the subarachnoid space. Here the dilatation occurs only proximal to the site of obstruction, the rest of the ventricular system remains normal.



NORMAL PRESSURE HYDROCEPHALUS

- Occurs usually in people over 50 years of age, producing symptoms mimicking those of dementia, i.e.: memory disturbance, gait disturbance and loss of bladder control. It may occur after head trauma, sub arachnoid hemorrhage or occasionally infection.
- The differential diagnosis of this entity is dementia and chronic subdural hematoma, because all three produce the triad of memory problems, gait disturbance and loss of control over urination.
- Usually the diagnosis of dilatation of ventricles is confirmed by CT. Improvements of symptoms may result following withdrawal of CSF via a spinal tap.
- Treated by a normal pressure valve shunting.

CLINICAL PRESENTATION

• BEFORE CLOSURE OF SUTURES

- Enlargement of the head, as evidenced by increase in its circumference.
- Wide, tense bulging fontanel.
- Shiny stretched scalp.
- Dilated scalp veins.
- Frontal bossing
- Abnormal face to head ratio (Ape face)
- Limitation of upward gaze (sunset appearance).



THE CHARTS TO FOLLOW THE HC



HYDROCEPHALUS

• AFTER CLOSURE OF SUTURES

- Signs and symptoms of increased ICP (headache, vomiting and visual manifestations).
- Signs and symptoms of the causative disease (meningitis, SAH, etc.).
- Signs and symptoms of ventricular enlargement (memory problems, difficulty walking and urinary disturbance).

IMAGING

• Computerized tomography (CT) or magnetic resonance (MRI) is the mainstay of diagnosis. Either will reveal the presence of the hydrocephalus itself, and may reveal the cause in a large percentage of cases especially when there is obstruction by a mass, or a congenital anomaly. One has to take care in using CT in children due to the untoward effects of radiation on their developing brain in these cases ultrasound could'be used, but requires experience.



MANAGEMENT

- Once the diagnosis has been made, attention should be directed to solving the problem of the ICP rise. If the causative pathology could be removed (excision of a tumor and re-establishing the CSF pathway), then, no further management is needed. But when, the obstructing lesion could not be removed, or removed partially, and in cases of congenital hydrocephalus, then a diversion procedure should be performed.
- Diversion procedures are called shunts. They aim at shunting the CSF from a location proximal to the site of obstruction, if there was any, to an absorbing site outside the brain (external shunt). However, and in cases of aqueduct stenosis a more direct approach is used, and the obstruction is bypassed internally.

SHUNTING

• Ventricular CSF is usually shunted using a pressure-operated valve, with anti-syphon device and containing a sampling chamber. The shunt is composed of the valve, a proximal (ventricular) catheter and a distal catheter, which are assembled during surgery.

THE SUNTING SYSTEM



THE SHUNTING SYSTEM





SHUNTING

• The proximal catheter is usually inserted via a burr hole in the right posterior parietal region into the posterior horn of the lateral ventricle. It can also be inserted into the frontal horn. The ventricular catheter is then attached to the valve outside the skull. Once this is done, the distal catheter is tunneled under the skin to its destination. Its proximal end is connected to the free end of the valve and its distal end is inserted in the required destination

SHUNTING

 The destination of the distal end will indicate the type of the shunting procedure, namely (ventriculo-peritoneal) if the shunting was to the peritoneal cavity and this is the most commonly used type, or (ventriculo-atrial), if it is to the right atrium, or (ventriculo-jugular), if it is to the jugular vein, and lastly (ventriculo-pleural), when it is to the pleural cavity. This last location is not favored, due to the negative pressure inside the pleural cavity, which may "suck" CSF resulting in low ICP and consequent hemorrhage.

THIRD VENTRICULOSTOMY

 In cases of aqueduct stenosis an operation called "third ventriculostomy" is performed in which a hole is made using the endoscope into the floor of the 3rd ventricle allowing the obstructed CSF to go directly into the subarachnoid space circumventing the obstruction site.

COMPLICATIONS OF SHUNTING

- INFECTION
- MALFUNCTIONING
- OTHER COMPLICATIONS:
 - Fracture or disconnection of the shunt
 - Volvulus, intestinal obstruction and intestinal perforation (ventriculo-peritoneal shunts)
 - Ascites, hydrocele, hernia (ventriculo-peritoneal shunts)
 - Migration into the vascular compartment (atrial and jugular shunts).
 - Peritoneal cysts (ventriculo-peritoneal shunts)
 - Need to lengthen the shunt (all types).
 - Over drainage (all types).

EXTERNAL VENTRICULAR DRAINAGE

