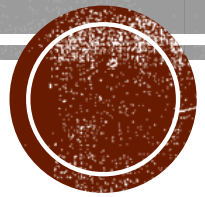
HYDROCEPHALUS

*acquired / congenital.

→ accumulation of CSF



- Communicating
  - ↓
  - no blockage in
- non-communicating
  - ↓
  - blockage in the

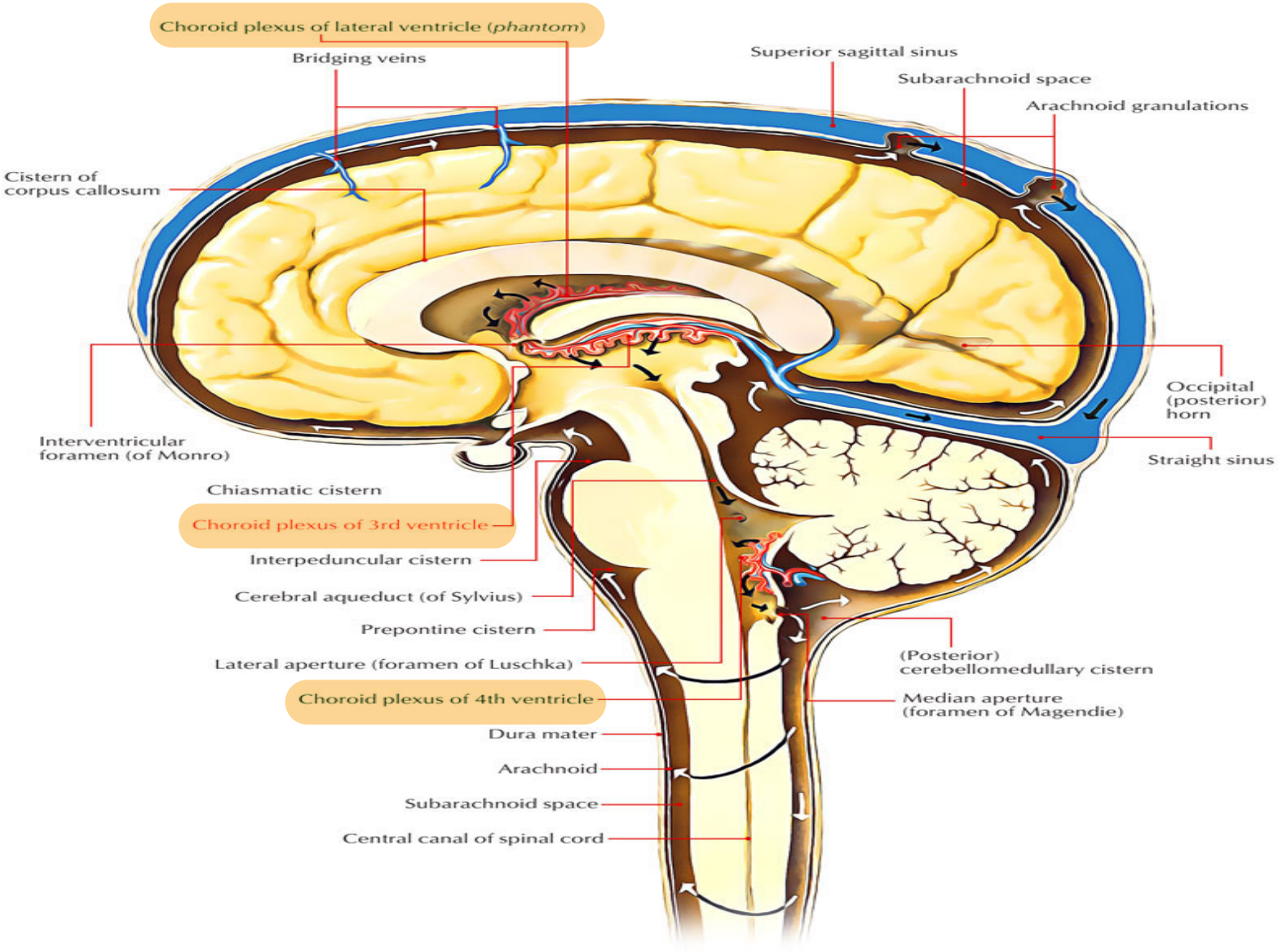
ventricular system

ventricular system [common cause]

- The **brain ventricles** are four cavities located within the brain that contain cerebral spinal fluid (CSF).
- Two lateral ventricles , the third ventricle andthe fourth ventricle
- It **protects** the brain by allowing it to "float" in a fluid bath and provides a shock absorber against head trauma. The CSF itself also helps to **provide nutrients** to the brain and to keep the brain in chemical balance.

⊛ CEREBROSPINAL FLUID CSF.

- The CSF is clear ,colorless fluid formed mainly by the choroid plexus , within the lateral vintricules, third and fourth ventricules of the brain . One third of CFS from brain ECF
- Normal values of CSF dynamics :
- Volume : 140 ml (120-150)
- Formation : 0.4 ml/ min (0.3-0.6)
- Turnover : 4 (4-6)
- ICP: <15 mmHg in adults (BTF recomends intervention when it exceeds 22)


Choroid plexus of lateral ventricle (*phantom*)
Bridging veins
Superior sagittal sinus
Subarachnoid space
Arachnoid granulations
Cistern of corpus callosum
Occipital (posterior) horn
Interventricular foramen (of Monro)
Straight sinus
Chiasmatic cistern
Choroid plexus of 3rd ventricle
Interpeduncular cistern
Cerebral aqueduct (of Sylvius)
Prepontine cistern
(Posterior) cerebellomedullary cistern
Lateral aperture (foramen of Luschka)
Choroid plexus of 4th ventricle
Median aperture (foramen of Magendie)
Dura mater
Arachnoid
Subarachnoid space
Central canal of spinal cord

* Lateral ventricle

↓ Foramen of Monro

* 3rd ventricle

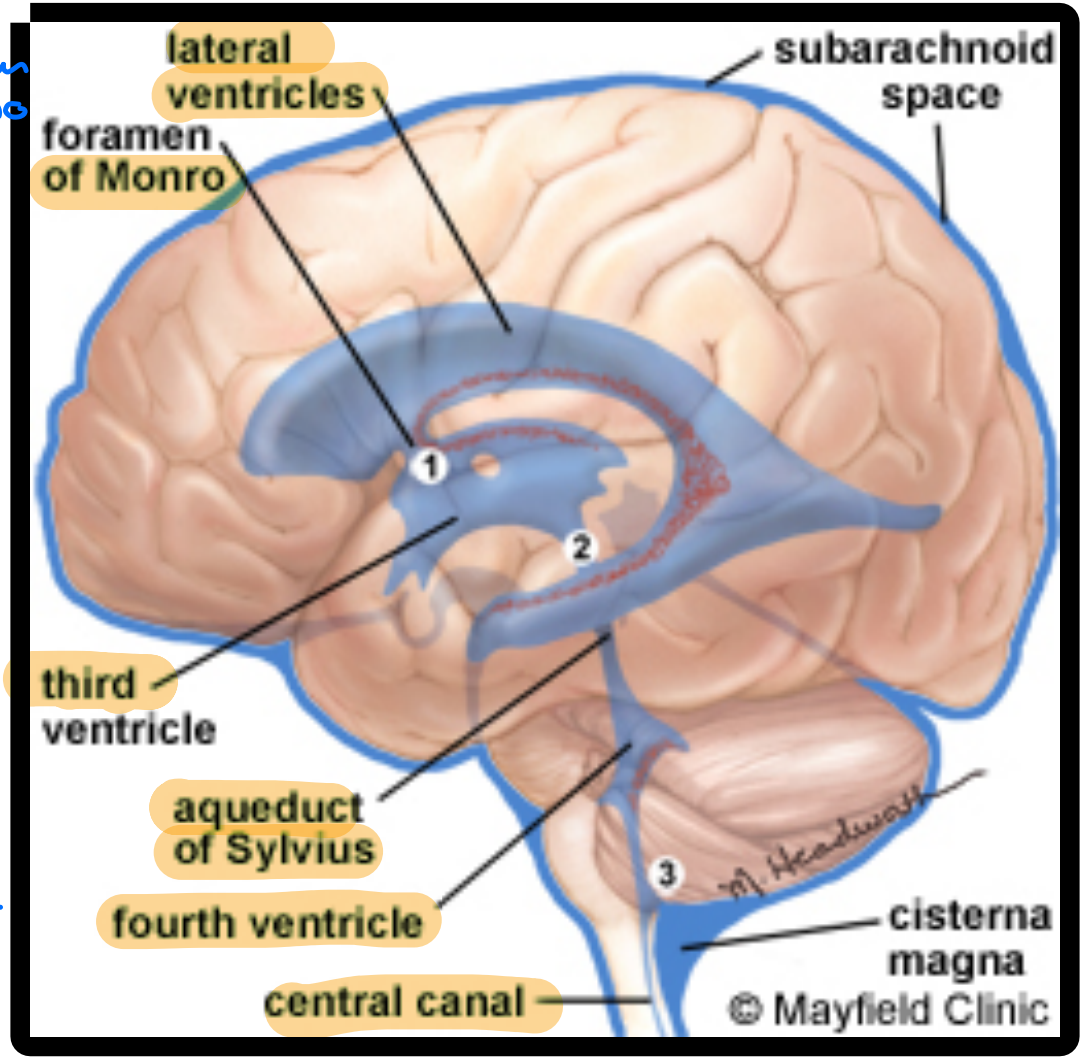
↓ aqueduct of Sylvis.

* 4th ventricle

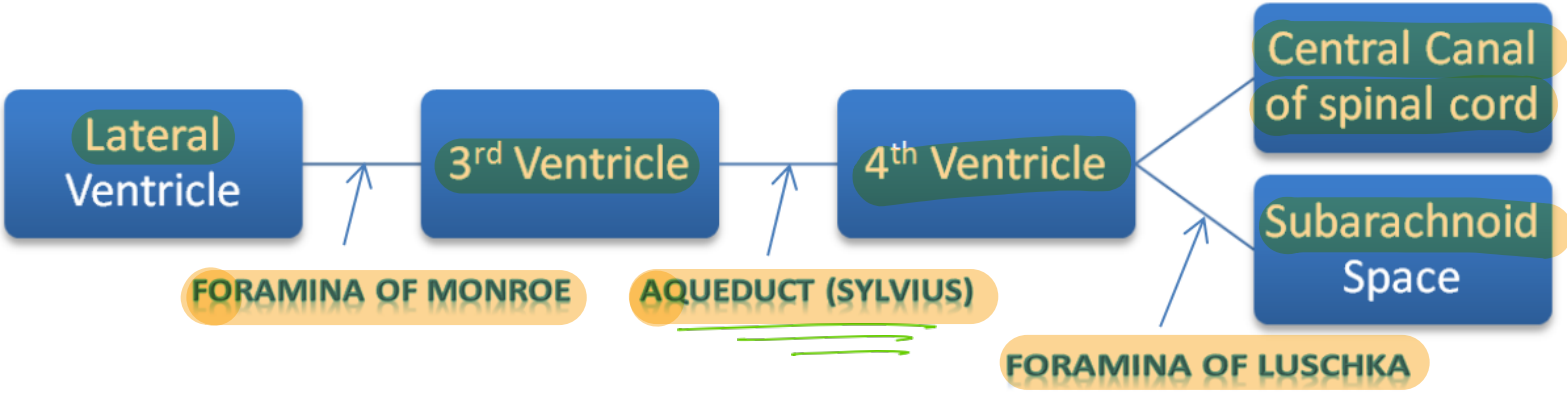
↓ Luscka.

↙ Central canal

↘ SA space

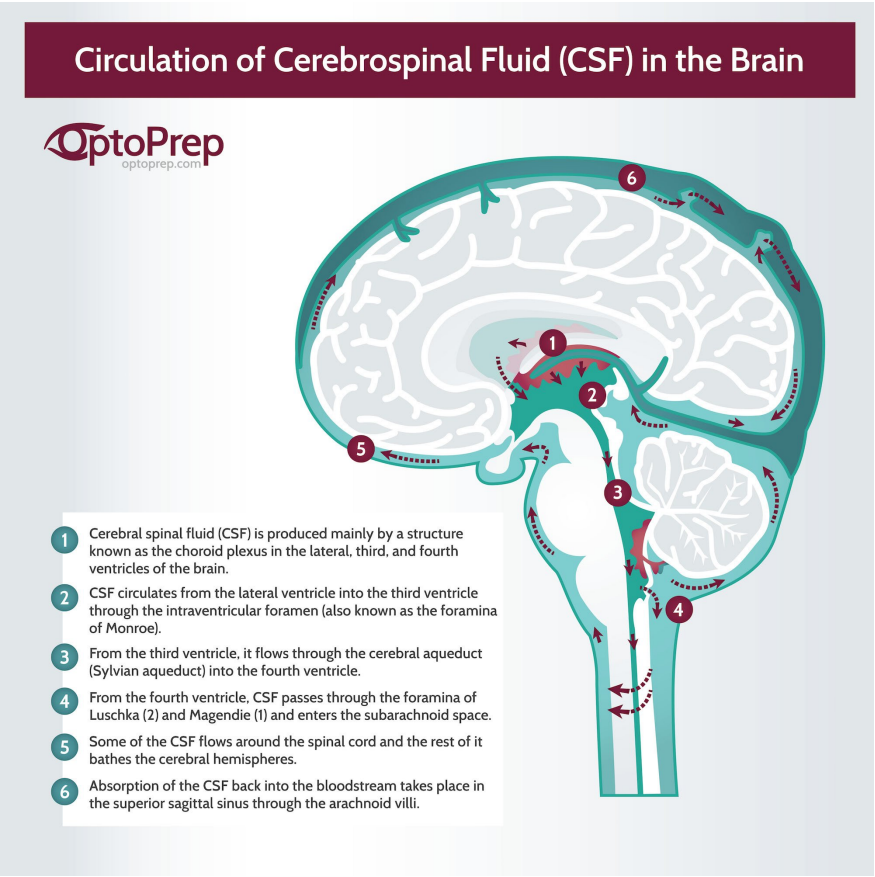


NORMAL CSF CIRCULATION

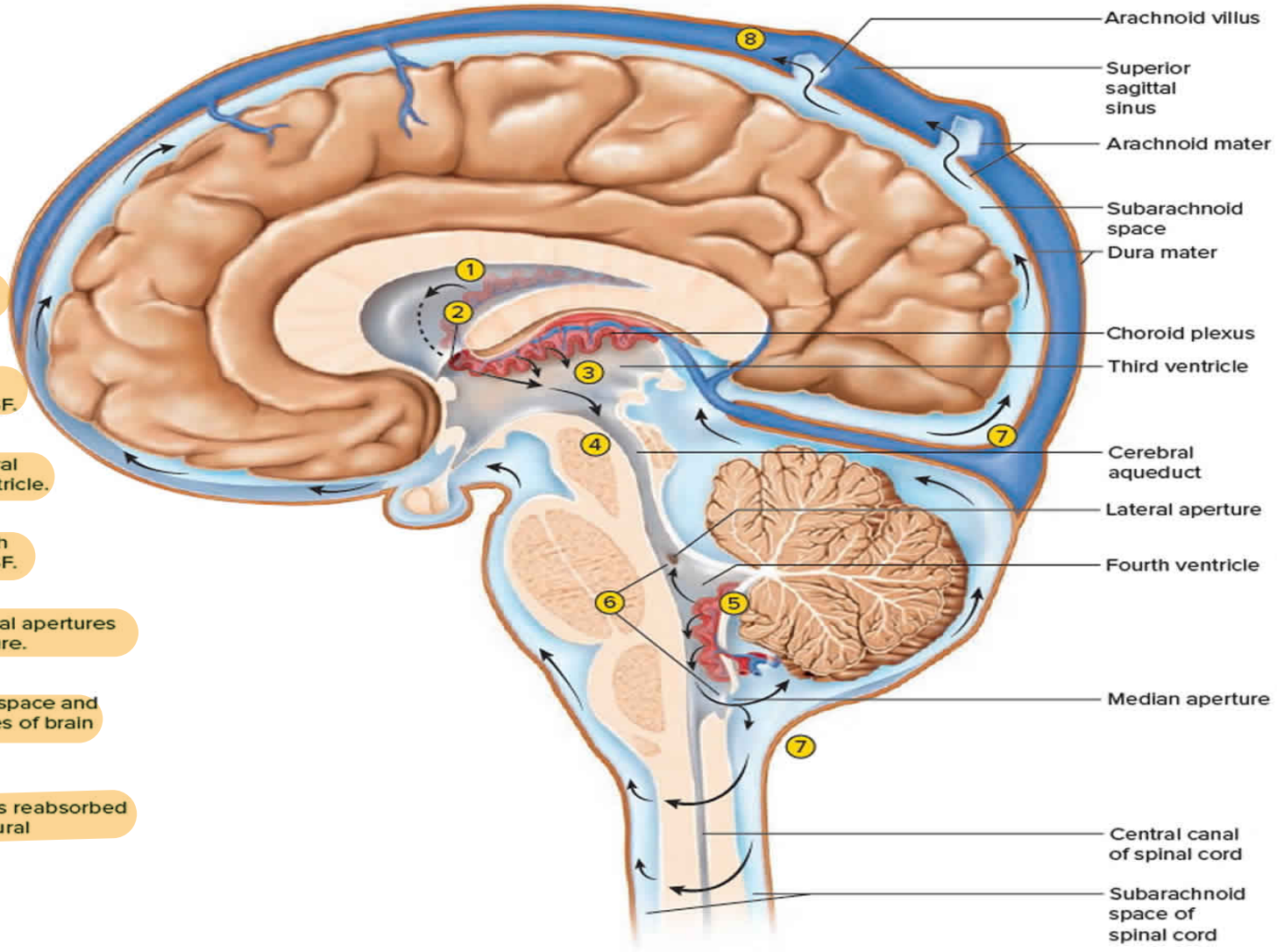


CSF CIRCULATION

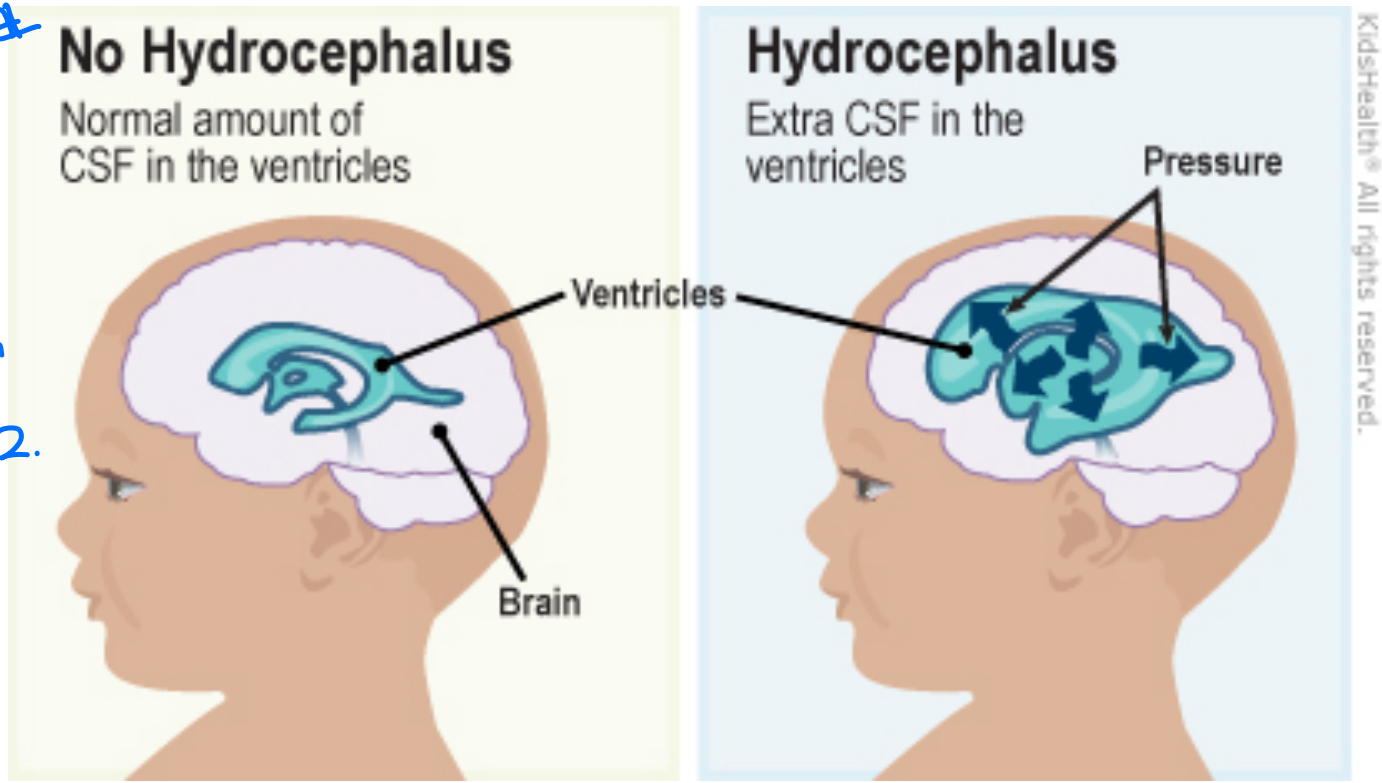
- The CSF flows from the lateral ventricles through the foramen of Monro into the 3rd ventricle, via the aqueduct of Sylvius into the 4th ventricle and then through the foramina of Magendie and Luschka into the subarachnoid space and basal cisterns.
- The CSF circulates throughout the spinal subarachnoid space and the basal cisterns up through the tentorial hiatus. It flows over the cerebral hemispheres and is largely absorbed by the arachnoid villi of the dural sinuses.



1. CSF is secreted by choroid plexus in each lateral ventricle.
2. CSF flows through interventricular foramina into third ventricle.
3. Choroid plexus in third ventricle adds more CSF.
4. CSF flows down cerebral aqueduct to fourth ventricle.
5. Choroid plexus in fourth ventricle adds more CSF.
6. CSF flows out two lateral apertures and one median aperture.
7. CSF fills subarachnoid space and bathes external surfaces of brain and spinal cord.
8. At arachnoid villi, CSF is reabsorbed into venous blood of dural venous sinuses.



Hydrocephalus is an abnormal enlargement of the ventricles due to an excessive accumulation of CSF resulting from a disturbance of its **flow**, **absorption** or, uncommonly, **secretion**.



communicating (no obstruction).

non communicating (obstruction in ventricles)

CLASSIFICATION OF HYDROCEPHALUS

* ***Obstructive hydrocephalus (non-comunicating)***—when there is an obstruction to the flow of CSF through the ventricular system. [normal production].

* ***Communicating hydrocephalus***—when there is no obstruction to the flow of CSF within the ventricular system but the hydrocephalus is due either to obstruction to CSF flow **outside the ventricular system** or to **failure of absorption** of CSF by the arachnoid villi.

some books classified it according to etiology

* non communicating hydrocephalus

congenital

acquired

ETIOLOGY OF OBSTRUCTIVE HYDROCEPHALUS

- **Congenital** : Aquedectal stenosis (most common).
  - Chiari malformation
  - Dandy walker malformation
  - Intrauterine infection (TORCH)
  - Colloid cyst obstructing the intervertibular foramen
- **Aquired** : brain tumer obtructiong the flow of CSF (espicially medulloblastomas, pinealoma, ependymomas, and astrocystoma)

COMMUNICATING HYDROCEPHALUS

↓ absorption

↑ production

- Decrease Apsorption : (damage to arachnoid villi) → يتربط عند ال subarachnoid.
  1. Infectious disease of the central nervous system (post-meningitis and cystricosis)
  2. Post hemorrhagic (post subarachnoid hemorrhage or post-intraventricular hemorrhage)
- Increase production :

  choroid plexus papilloma or carcinoma or even inflammation of choroid plexus

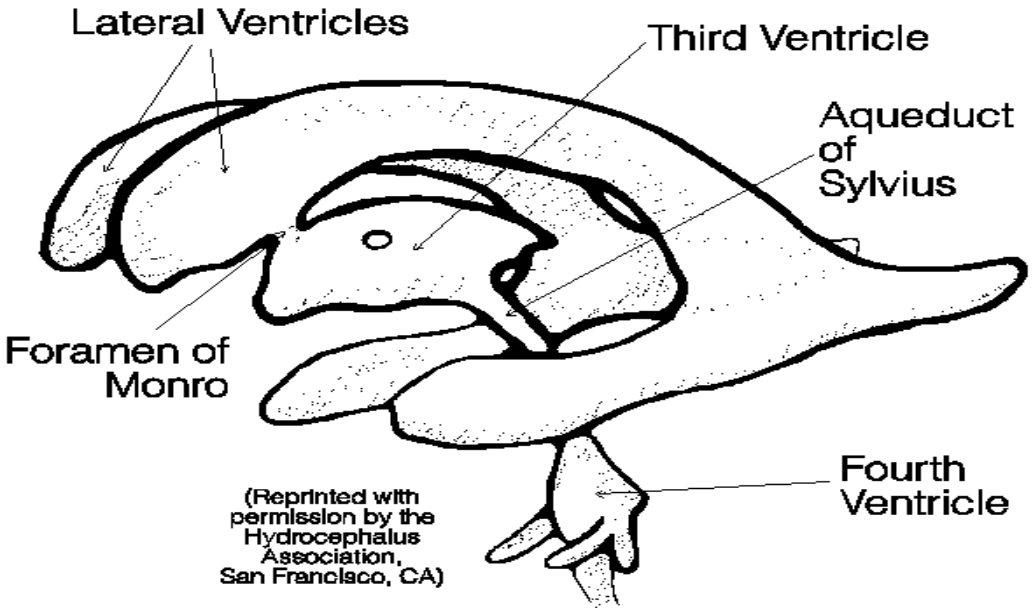
- The most common **congenital cause** is **stenosis of the aqueduct of Sylvius**. This is a major cause of hydrocephalus in children with spina bifida and myelomeningocele .

- The **acquired forms** of hydrocephalus occur most frequently after **intracranial bleeding**, in meningitis and because of **tumours**.

most common congenital cause.

AQUEDUCTAL STENOSIS

- The normal aqueduct measures about 1 mm in diameter, and is about 11 mm in length.



AQUDUCTAL STENOSIS

- Is the **most common cause** of congenital hydrocephalus (43%).
  - ↳ with Spina Bifida + myelomeningiocele.
- ✓ Aqueduct develops about the **6th week** of gestation .
- → **Obstruction** between **3rd and 4th ventricules**
- **M:F = 2:1** .
- Other congenital anomalies (16%): **thumb deformities**.
- Prognosis: **11-30% mortality** .

intrinsic [in infancy]

extrinsic [in adult].

ETIOLOGY OF AQUDUCTAL STENOSIS

Intrinsic Pathology of the Aqueduct (in infancy)

(inflammation)

1. **Gliosis of the Aqueduct (peri-aqueductal gliosis):** Usually of infectious origin showing a marked gliofibrillary response. The lumen is clear of ependyma. (proliferation of astrocyte)
2. **Stenosis of the Aqueduct (true stenosis):** Narrowed aqueduct without evidence of gliosis (aqueduct histologically normal). This may have hereditary basis.
3. **Forking of the Aqueduct:** Multiple chanells (often narrowed) with normal epithelial lining that do not meet, separated by normal nervous tissue unable to handle CSF volume. Most often seen with **spina bifida**.

حتى الصرفاية ، كثير تحكوب فيها.

4. **Septum or Membrane Formation**: A thin membrane of neuroglia may occlude the aqueduct. There may be a primary developmental defect or it may follow granular ependymitis from intrauterine infections. This is **the rarest** of the types of narrowing.

ETIOLOGY OF AQUDUCTAL STENOSIS

Extrinsic Pathology of the Aqueduct (in adult) :

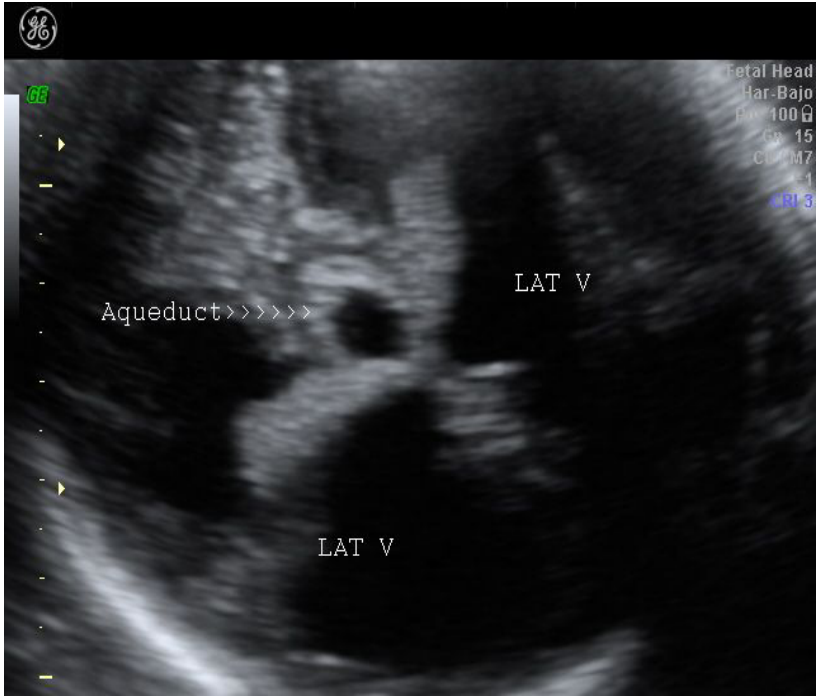
1. Infectious: Abscesses.
2. Neoplastic: Pineal tumors, brainstem gliomas, medulloblastoma, ependymoma.
3. Vascular: AVM, aneurysm.
4. Developmental: Arachnoid cysts.

CLINICAL FEATURES OF AQUEDUCTAL STENOSIS

- **Obstructive hydrocephalus**: presents with macrocephaly and/or intracranial hypertension.
- **Parinaud's syndrome**: Inability to elevate eyes.
- **Collier's sign**: Retraction of the eyelids.

IMAGING OF AQUEDUCTAL STENOSIS

- Ultrasonography can detect aqueductal stenosis in utero.

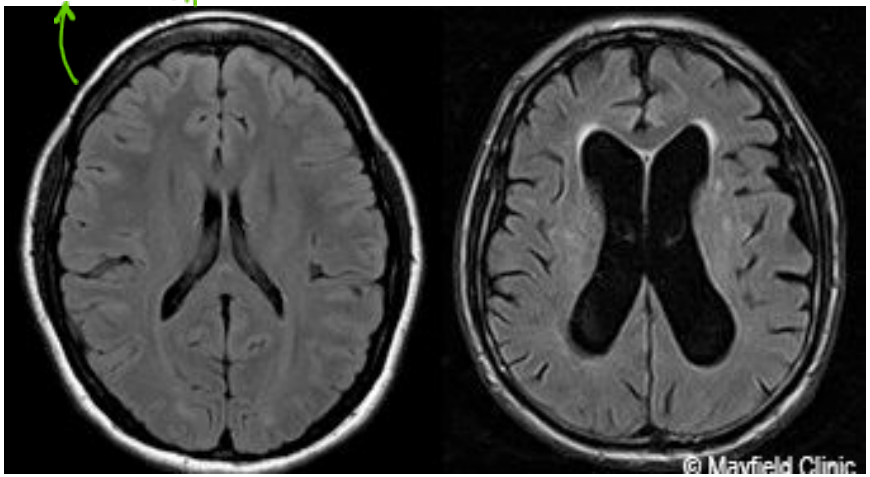


IMAGING OF AQUEDUCTAL STENOSIS

- CT and MRI. MRI is essential if third ventriculostomy is to be considered.

normal

no compression



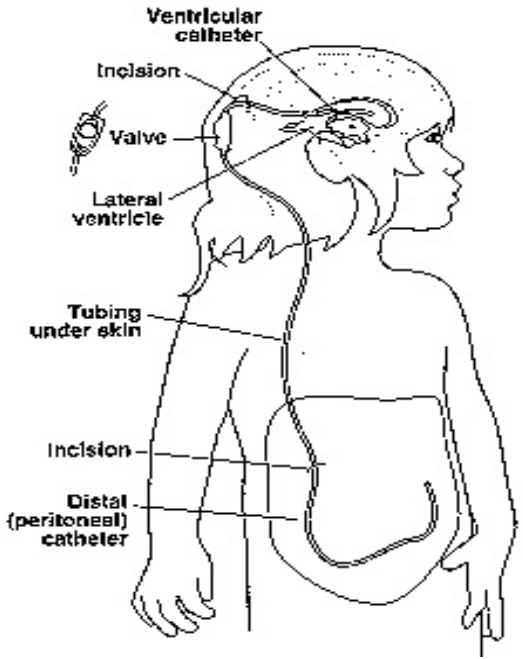
↳ Compression

T2

Fig. 15.4 Aqueductal stenosis in an adult.
Sagittal T2 MRI showing a web-like obstruction of the Sylvian aqueduct in a patient with dilated third and lateral ventricles with normal-sized 4th ventricle. The aqueduct proximal to the obstruction is also dilated, but the aqueduct connecting to the 4th ventricle (distal to the obstruction) is not dilated.
Inset depicts the location in the sagittal T2 brain MRI from where the detail is obtained.

TREATMENT OF AQUEDUCTAL STENOSIS

- Remove underlying cause of obstruction if possible.
- Third ventriculostomy as initial treatment of choice.
- VP shunt if technical reasons do not allow third ventriculostomy or if the child fails after ventriculostomy.
- Aqueductal stent can be placed if technically feasible. Usually rarely done due to risk of upper brain stem injury.



Treat the hydroceph + shunt.

PRESENTING FEATURES HYDROCEPHALUS IN INFANTS

- failure to thrive.
- increased skull circumference (compared with normal growth curves).
- tense anterior fontanelle.
- ‘cracked pot’ sound on skull percussion. (hyper-resonance)
- transillumination of cranial cavity with strong light.
- severe, impaired conscious level and Vomiting .
- ‘setting sun’ appearance due to lid retraction and impaired upward gaze from 3rd ventricular pressure on the midbrain tectum.
- thin scalp with dilated veins.

PRESENTING FEATURES HYDROCEPHALUS IN ADULT

- **acute** onset and deterioration

or

- **gradual** onset and slowly progressive deterioration.

* ACUTE-ONSET ADULT HYDROCEPHALUS

- This type of presentation occurs particularly in patients with tumours causing obstructive hydrocephalus, although it may occur with any of the causes of hydrocephalus and an acute rapid neurological deterioration may occur in patients who have had long-standing chronic hydrocephalus.

- The major presenting features are due to the signs and symptoms of raised intracranial pressure :

✓ - headache [morning headache].

✓ - vomiting (↑ ICP)

✓ - papilledema

✓ - deterioration of consciousness .

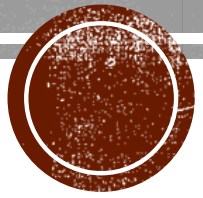
✓ - Upgaze will often be impaired due to pressure of the dilated 3rd ventricle on the superior colliculus of the tectum.

* GRADUAL ONSET ADULT HYDROCEPHALUS

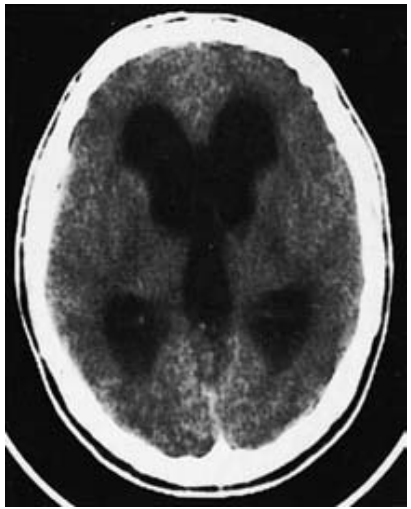
- this type of onset occurs less frequently than the previous type. The symptoms of raised intracranial pressure are only very gradually progressive and late diagnosis is common.
- Early features in the adolescent involve deteriorating school performance as a result of headaches, failing mental function, memory loss and behavioural disturbances.
- Endocrine abnormalities such as infantilism and precocious puberty can occur in association with chronic hydrocephalus in older children and adolescents due to disturbance of the hypothalamus and possible compression of the pituitary gland.
- If the condition is unrecognized progressive visual failure will occur, secondary to papilloedema and optic atrophy. acute decompensation may occur and the patient may suddenly develop a rapid deterioration of conscious state.

RADIOLOGICAL INVESTIGATION

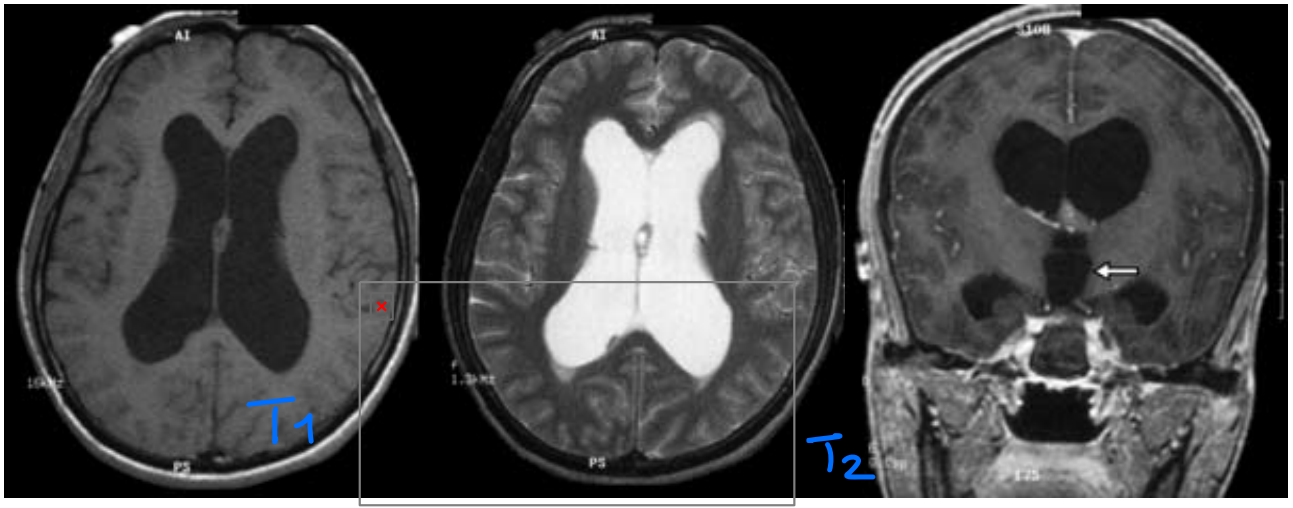
The most important investigation is either a CT scan or MRI of the brain which will show which ventricles are dilated.



CT



MRI

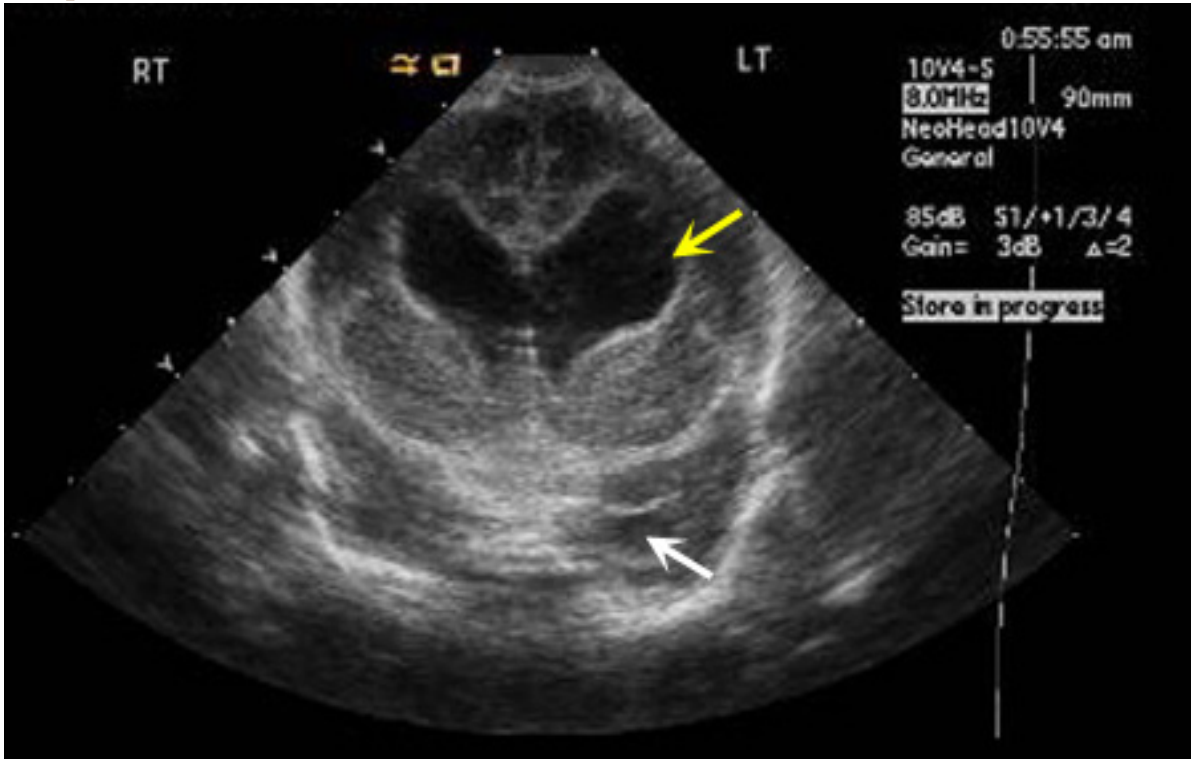


- If the lateral ventricles and 3rd ventricle are all very dilated, and the 4th ventricle is small, it is likely that the obstruction is at the level of the aqueduct of Sylvius.
- In a communicating hydrocephalus all the ventricles are dilated.

Ultrasonography

***Ultrasonography through the* open anterior fontanelle is useful in assessing ventricular size in infants and may obviate the need for repeated CT scans.**

Infant with hydrocephalus with dilatation of frontal horns of both lateral ventricles (yellow arrow) and temporal horn (white arrow).



TREATMENT

- Management of hydrocephalus will depend on the underlying cause, options include :
- Removing a causative mass lesion.
- Ventricular shunting.
- Third ventriculostomy.

MEDICAL MANAGEMENT

This can be tried in mild cases of hydrocephalus.

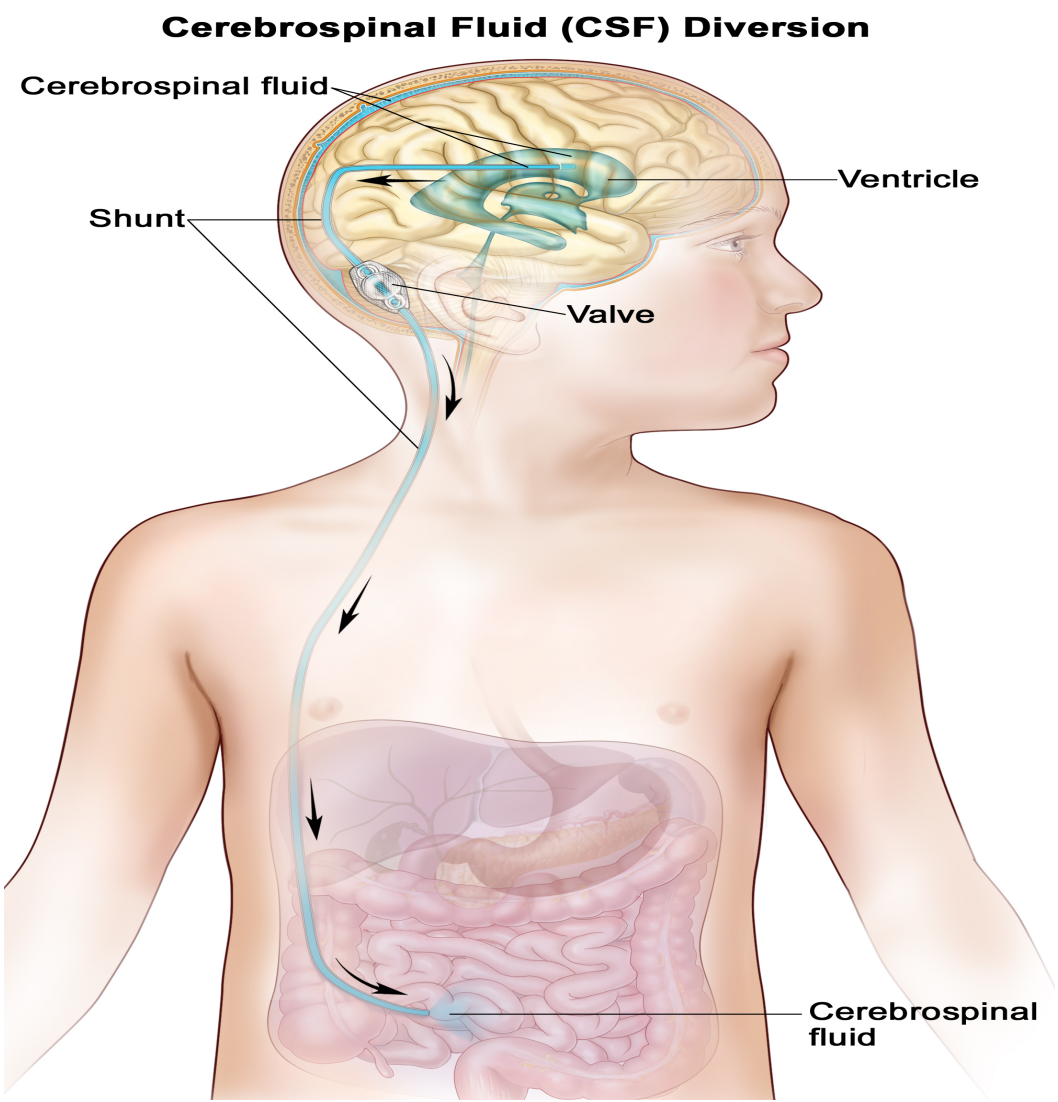
- **Acetozolamide**: dose of 50mg/kg/day dimnishes CSF production.
- **Oral glycerol** has also been used for the similar purpose.

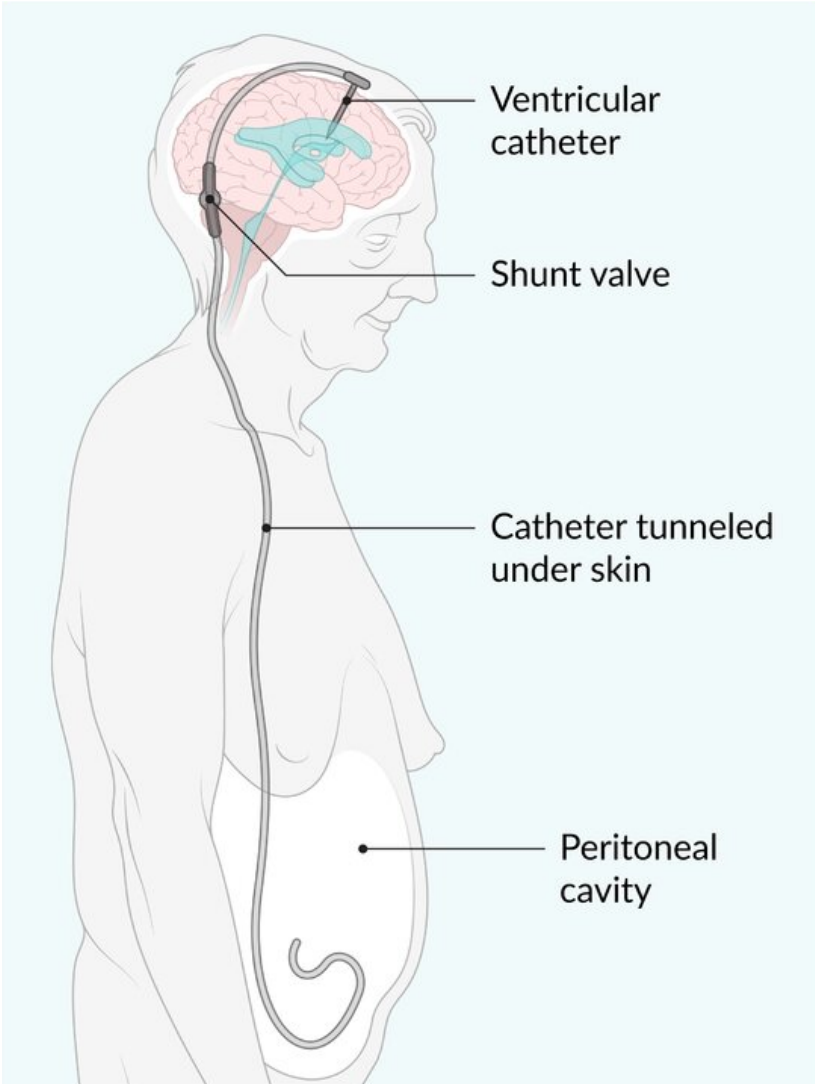
REMOVING A CAUSATIVE MASS LESION

- In some circumstances it may be appropriate to treat the hydrocephalus by tumour removal and decompression of the CSF pathways, perhaps with the insertion of an external ventricular drain (EVD) to cover the early postoperative period.

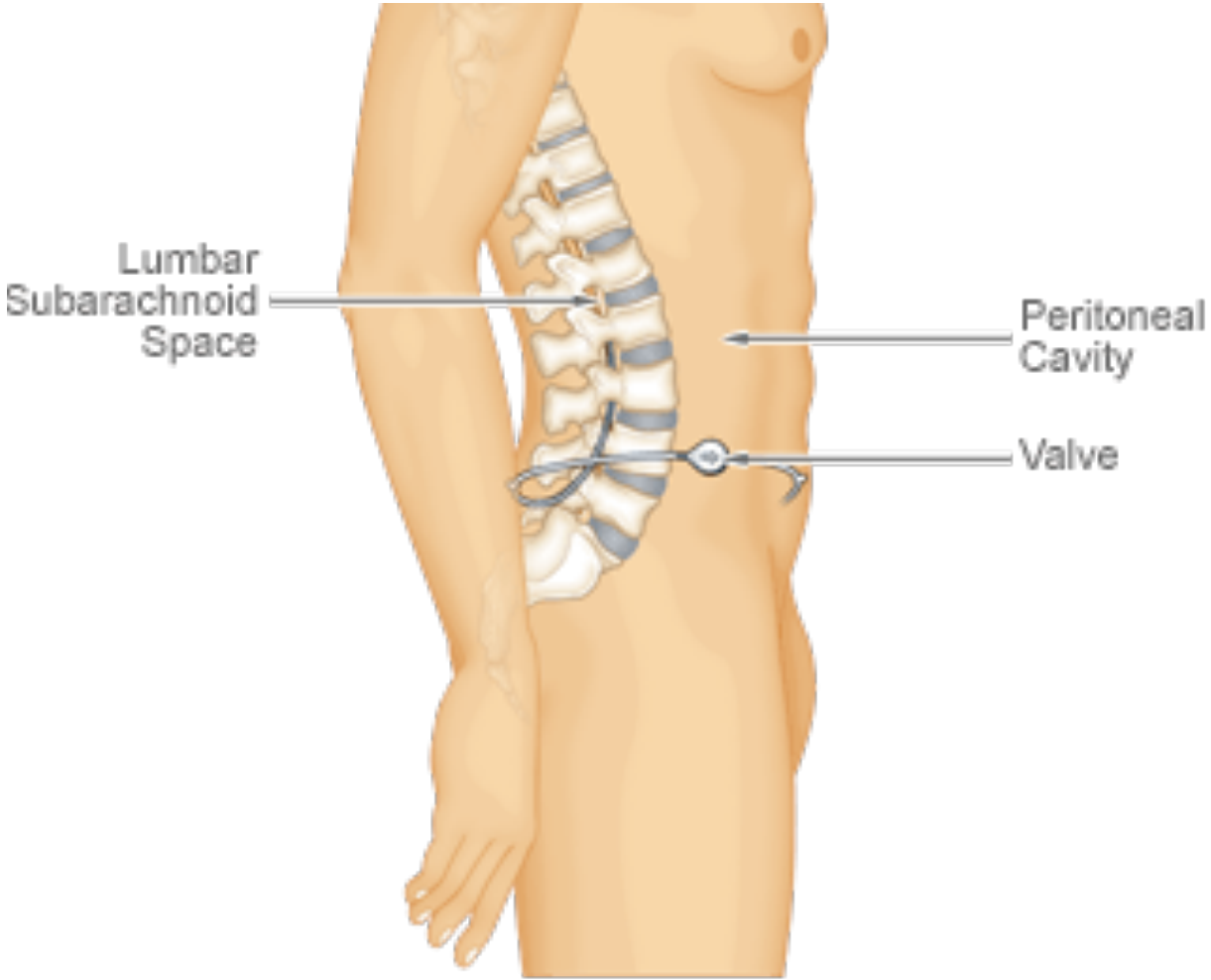
VENTRICULOPERITONEAL SHUNT

- involves the insertion of a catheter into the lateral ventricle. The catheter is then connected to a shunt valve under the scalp and finally to a distal catheter, which is tunneled subcutaneously down to the abdomen and inserted into the peritoneal cavity.
- If the CSF pressure exceeds the shunt valve pressure, then CSF will flow out of the distal catheter and be absorbed by the peritoneal lining.


Cerebrospinal Fluid (CSF) Diversion
Cerebrospinal fluid
Ventricle
Shunt
Valve
Cerebrospinal fluid


Ventricular catheter
Shunt valve
Catheter tunneled under skin
Peritoneal cavity

- Other options for distal catheter placement include:
- (ventriculo-atrial shunt): from the right atrium via the deep facial and jugular vein , it's necessary when there has been marked intraperitoneal sepsis or multiple abdominal operations.
- (lumboperitoneal shunt) : *The lumboperitoneal shunt involves drainage of the CSF from the lumbar theca rather than the ventricle. (pseudotumor cerebri)*
- LP shunts reduce the spinal CSF volume, while VP shunts keep the cranial and spinal CSF volume in the physiological range.



Lumbo-peritoneal shunt

⊛ infection indication.

↓ spinal CSF volume.

Febrile
↓ نشاط خف *

COMPLICATIONS OF THE SHUNT

1) Infection:

- Infection is the second most common cause of shunt malfunction ,with a approximately 8–15% among patients who undergo VPS placement.
- It's usually caused by skin commensals, such as *Staphylococcus epidermidis*. *Neonates are susceptible to Escherichia coli and haemolytic streptococcal infections*.
- Most infections become apparent clinically by 6 weeks and over 90% are apparent within 6 months.
- ***Risk factors for infection include :***
  - ***very young children.***
  - ***open myelomeningocele.***
  - ***longer operative time .***
  - ***excessive staff movement into and out of theatre.***

Treatment :

- admission, removal of the shunt, external CSF drainage and treatment of infection prior to re-insertion of the shunt at a different site.

- The introduction of antibiotic-impregnated catheters has resulted in a reduction in shunt infection rates.

irrigation with vanco.

COMPLICATIONS OF THE SHUNT

2) Shunt blockage : → high protein of CSF.

- **Shunt catheter obstruction is by far the most common cause shunt malfunction.**
- **Shunt blockage may affect the ventricular catheter, shunt ,valve or distal catheter.**
- **More than one-half of cases of shunt blockage are subsequently shown to be infected.**
- **Causes of blockage : choroid plexus adhesion, blood, cellular debris or misplacement of the distal catheter in the pre-peritoneal space.**

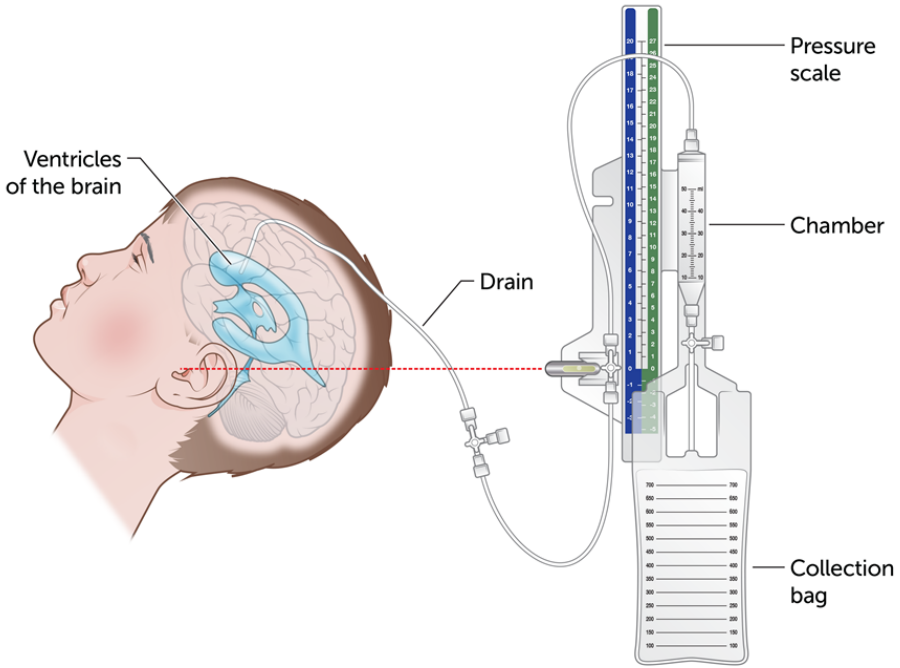
3) Intracranial hemorrhage :

- **Shunt systems may overdrain leading to subdural haemorrhage .**
- Other complications are common to intracranial surgery and include **seizures (5%), CSF leak, stroke and (< 1%) intracerebral haemorrhage**.

* EXTERNAL DRAINS

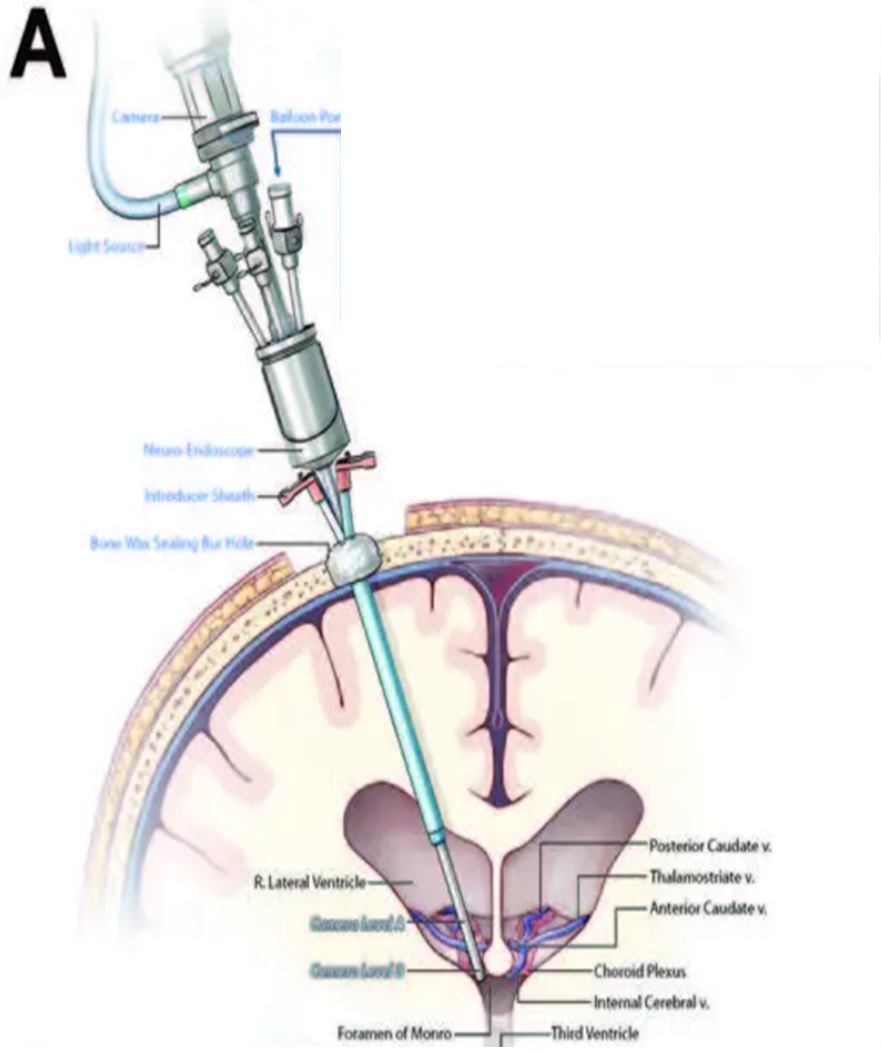
EVD
LD

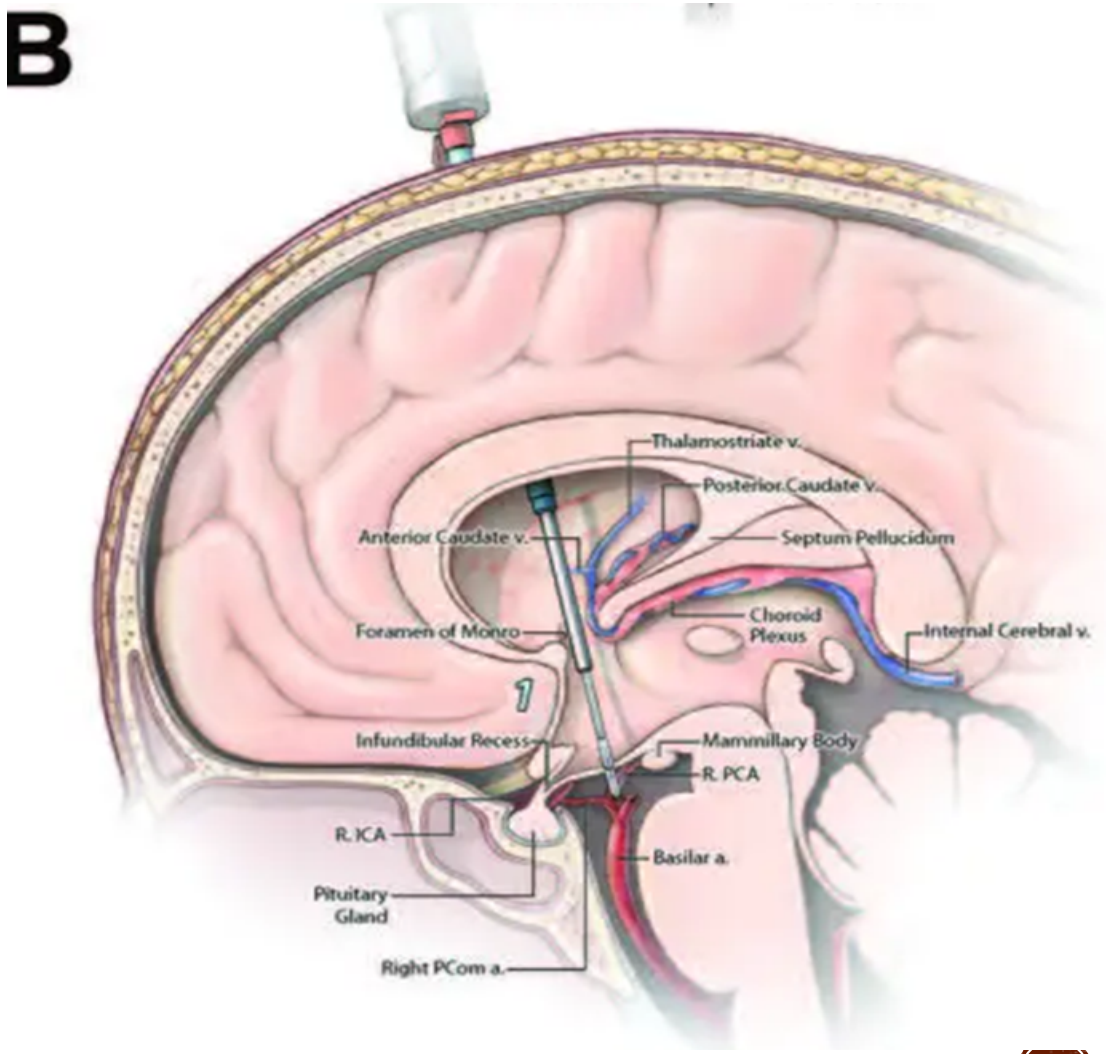
- External drains can be placed within the ventricle (EVD) or the lumbar thecal sac (lumbar drain).
- These are useful for temporary CSF drainage and can be used to administer intrathecal antibiotics to treat CSF infection., take sample and decrease ICP.



ENDOSCOPIC THIRD VENTRICULOSTOMY

- Involves the insertion of a neuro-endoscope into the *frontal horn of the lateral ventricle* and then into *the third ventricle* through the foramen of Munro. A stoma can be created in the floor of the third ventricle in between the mamillary bodies and infundibular (pituitary) recess.
- CSF can then communicate freely between the ventricular system and interpeduncular subarachnoid space.
- The technique is particularly useful when there is obstruction of the CSF pathways below the third ventricle such as with aqueduct stenosis or posterior fossa mass lesions.

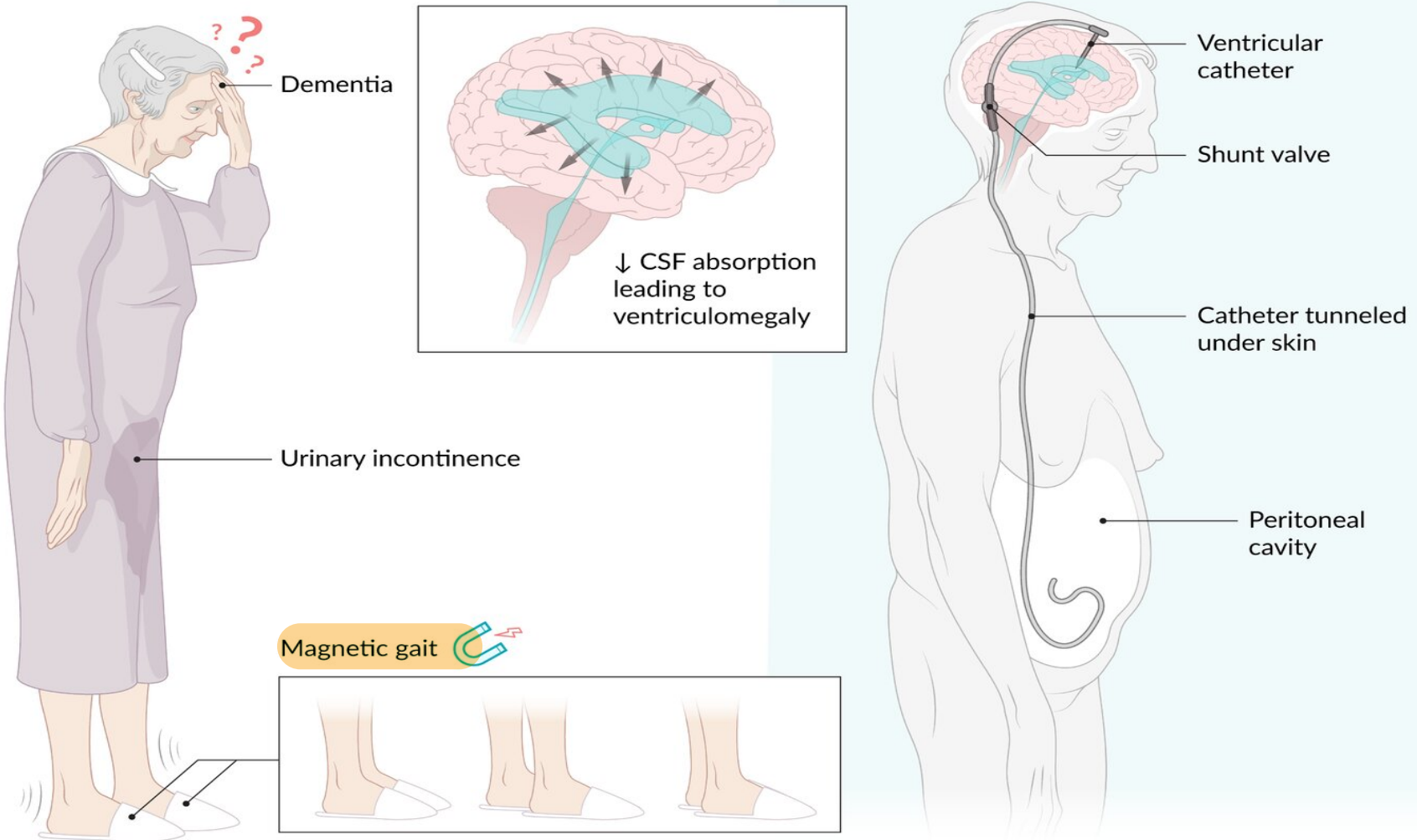

A
Camera
Light Source
Neuro-Endoscope
Introducer Sheath
Bone Wax Sealing Bur Hole
R. Lateral Ventricle
Posterior Caudate v.
Thalamostriate v.
Anterior Caudate v.
Choroid Plexus
Internal Cerebral v.
Foramen of Monro
Third Ventricle


B
Thalamostriate v.
Posterior Caudate v.
Anterior Caudate v.
Septum Pellucidum
Choroid Plexus
Foramen of Monro
Internal Cerebral v.
1
Infundibular Recess
Mammillary Body
R. PCA
R. ICA
Basilar a.
Pituitary Gland
Right PCom a.

- It has an advantage over shunting in that no tubing is left in the patient and therefore infection rates are lower.
- Rare, but serious, complications include basilar artery rupture or memory impairment from injury to the fornix.
- ETVs may block off, however, with about one-half of these patients ending up with a shunt.
- The procedure is less useful for communicating types of hydrocephalus or in infants of less than 6 months of age, but has a success rate of over 70% for accepted indications.

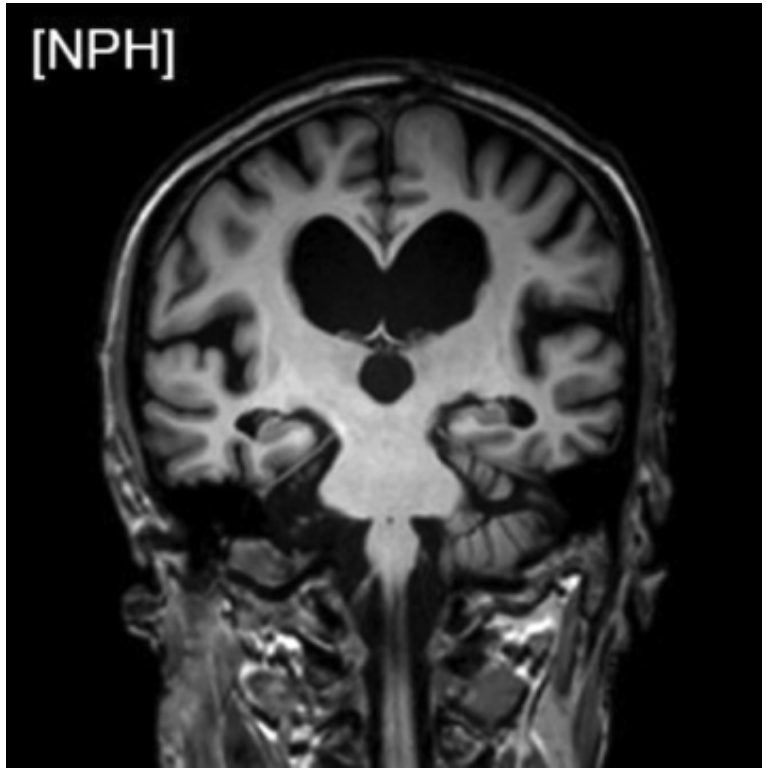
NORMAL-PRESSURE HYDROCEPHALUS

- Normal-pressure hydrocephalus a form of chronic communicating hydrocephalus that primarily affects elderly individuals (> 60 years) and is characterized by a distinct clinical triad (urinary incontinence, dementia, apraxic gait) , where the radiological studies showed hydrocephalus but the lumbar CSF pressure was normal.
- Etiology: in a large percentage the communicating hydrocephalus may have resulted from obliteration of the subarachnoid pathways in the basal cisterns following an episode of meningitis or subarachnoid haemorrhage, from either rupture of an aneurysm, arteriovenous malformation or following trauma.
- Although lumbar puncture pressure is within the normal range, continuous monitoring of the intracranial pressure in these patients will frequently reveal abnormal wave formation, especially at night.


Dementia
↓ CSF absorption leading to ventriculomegaly
Ventricular catheter
Shunt valve
Catheter tunneled under skin
Peritoneal cavity
Urinary incontinence
Magnetic gait

INVESTIGATION

- The CT scan or MRI will show dilated ventricles without significant cortical atrophy.
- The difficulty arises that normal-pressure hydrocephalus may occur in patients with a scan appearance of cortical atrophy, but in these patients the degree of ventricular dilation should be more than would be expected just to compensate for the degree of atrophic change.



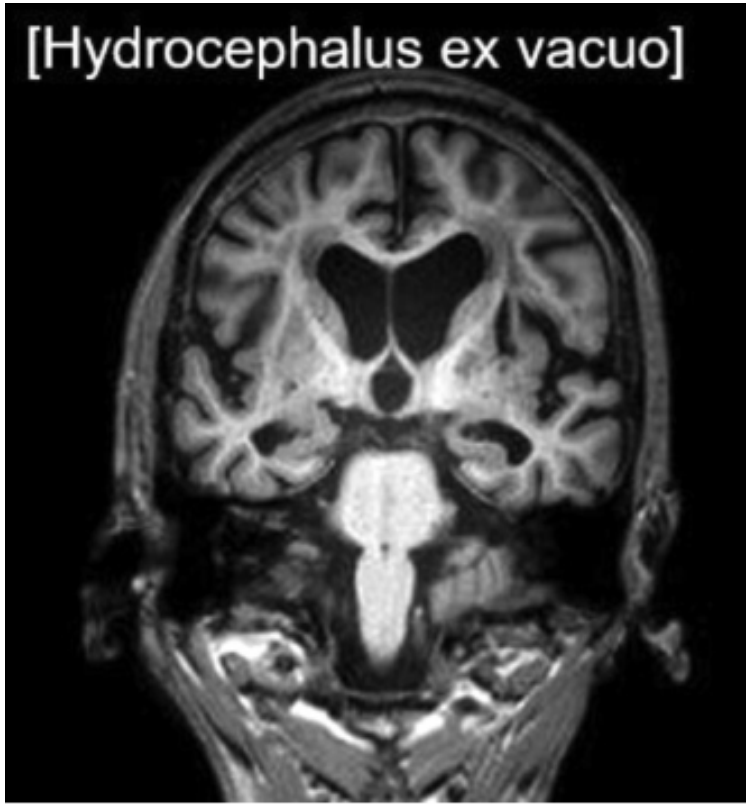
TREATMENT

- The following criteria can be used to assess the patients with the greatest chance of improvement following a shunt:
  - A clinical presentation of the classic triad, particularly if the features of gait disturbance predominate.
  - The CT scan or MRI showing marked hydrocephalus with minimal cortical atrophy.
  - A clearly defined cause for the hydrocephalus, such as a past episode of subarachnoid haemorrhage, trauma or meningitis.
  - Abnormal pressure waves on continuous intracranial pressure monitoring.

- Naturally, a patient who has all these positive criteria deserves a shunt and should make a good recovery following the operation.

HYDROCEPHALUS EX-VACUO

- The ventricles and subarachnoid space appear enlarged secondary to loss of brain tissue (an actual shrinkage of brain substance).
- Although there is more CSF than usual, intracranial pressure and flow of cerebrospinal fluid are normal.
- imaging: Enlarged CSF spaces, especially (lateral ventricles) and Cortical atrophy may be prominent.



EXTERNAL HYDROCEPHALUS

- A condition that occurs in infancy and early Childhood.
- characterised by: enlargement of the subarachnoid space, typically in the frontal areas and interhemispheric fissure with raised intracranial pressure without significantly enlarged ventricles.
- It is attributed to an absorption deficiency and typically resolves within a year.

Benign External Hydrocephalus