

Hydrocephalus

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❖ A 2 months old infant.

Big head!!!

❖ Head circumference
was 42 cm

1- Is the H.C. **normal**?

2- Differential diagnosis?

3-How to investigate?

4-How to treat?

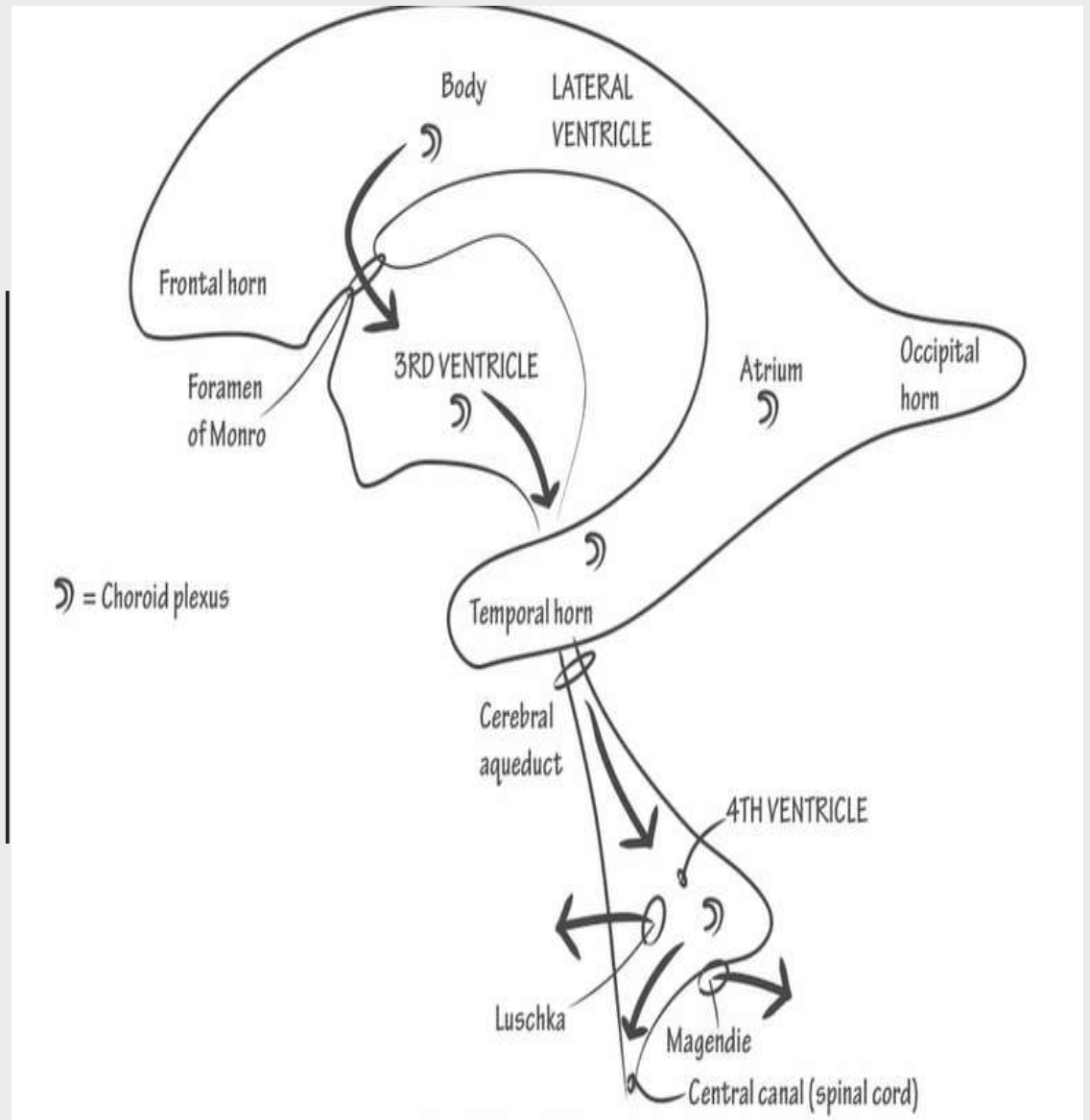
5-What is the outcome?



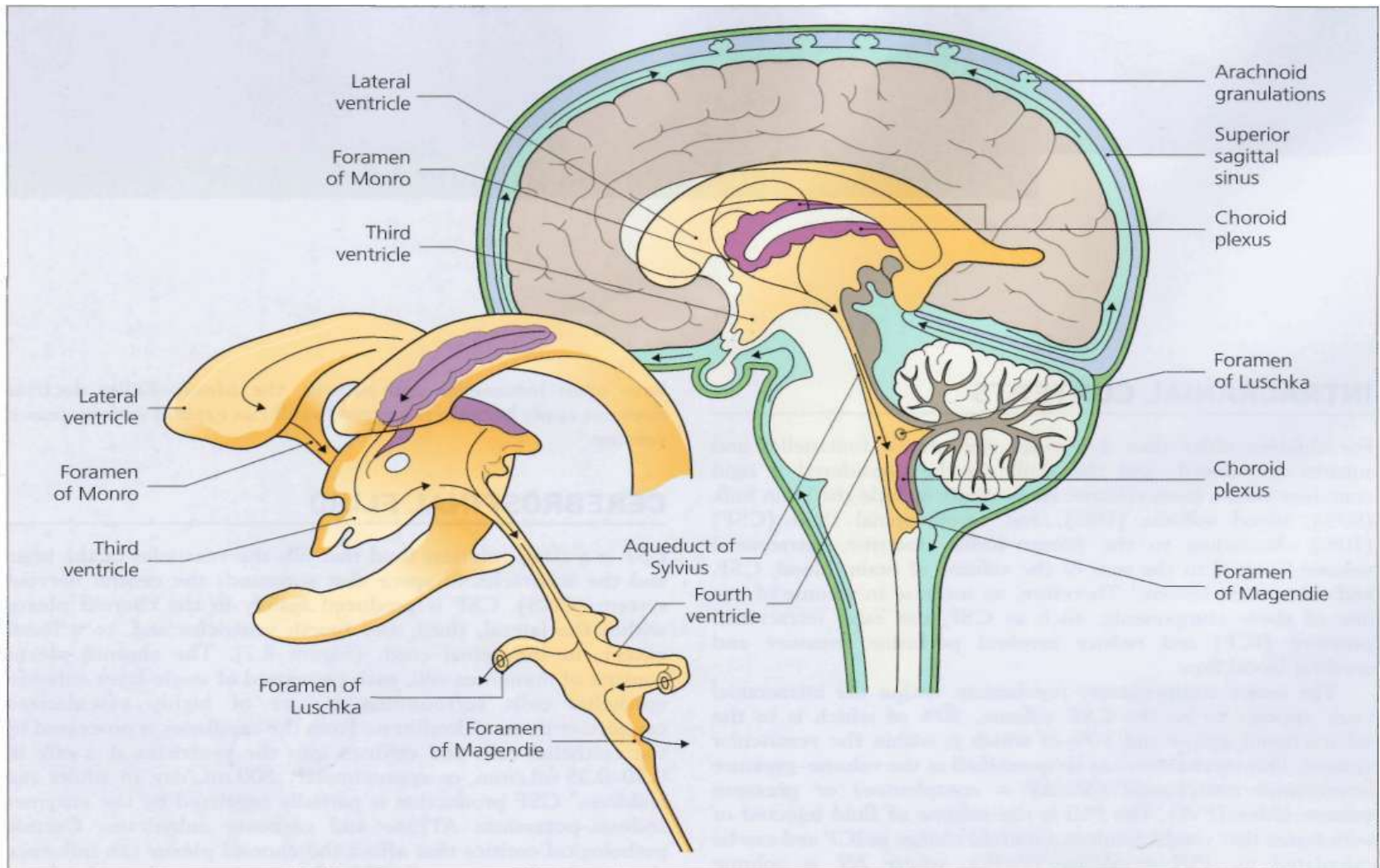
What is hydrocephalus?

- Is an accumulation of cerebrospinal fluid (CSF) within the ventricular space at an inappropriate pressure.
- Estimated prevalence is 1-1.05%
- Incidence of congenital cases is 0.9-1.8/1000 live birth.

Physiology of CSF circulation



The ventricular system



The cerebrospinal fluid (CSF)production

- CSF is produced mainly in the **choroid plexus**.
- Clear **colorless** fluid
- Total volume: **150mls**(40-50 ml in neonates and 65-140 ml in children).
- 25mls in ventricles + 125 mls in subarach.space
- Production: **500mls/day** and turnover: 3-4 times

CSF production is partially regulated by :
-sodium-potassium ATPase ...and
- carbonic anhydrase.

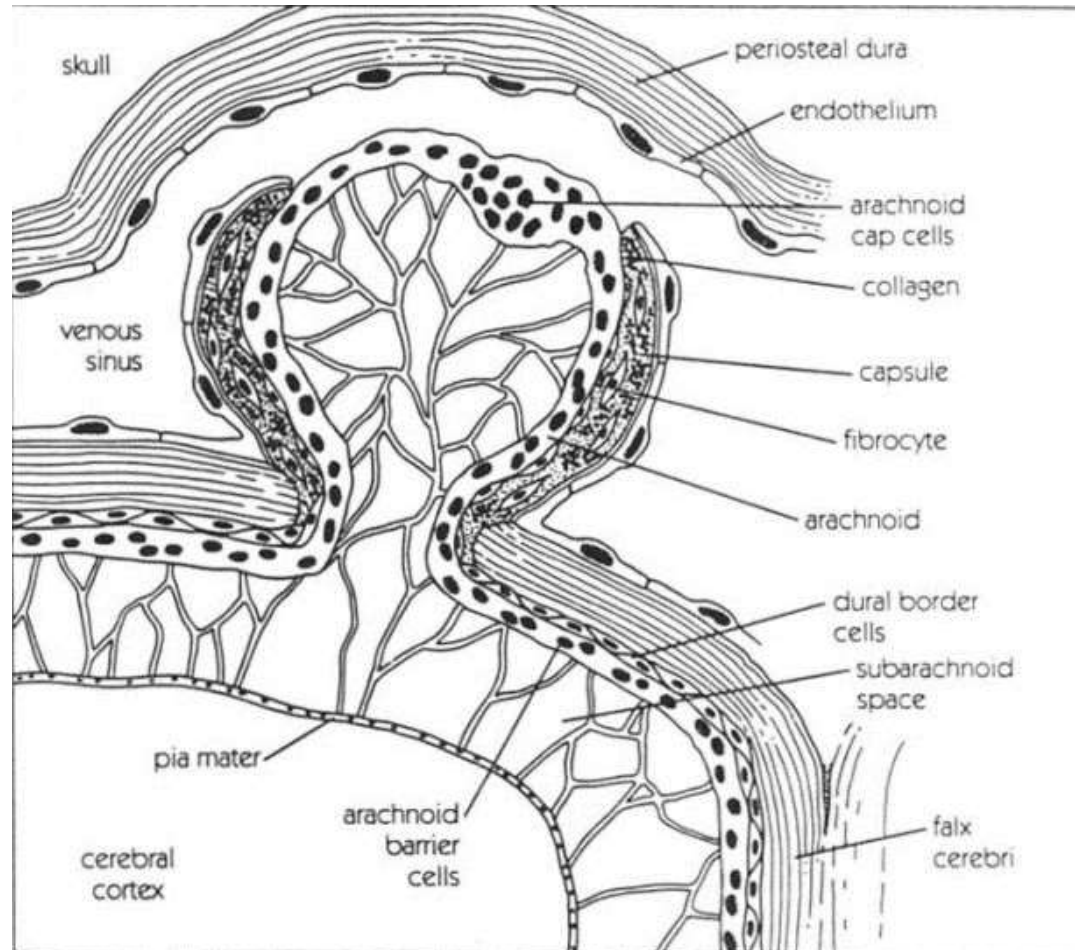
What is cerebrospinal fluid(CSF)?

● Contents:

1. **Acellular** (<5 lymphocytes, if higher:
Pleocytosis)
2. Similar Na⁺/ higher Cl⁻/ less K⁺ compared to
plasma
3. ≈15-40mg/dl of proteins depending on
site/age
4. 2/3 of sugar concentration in plasma

CSF is absorbed by ***arachnoid villi***, which are diverticula of arachnoid that invaginate within the sagittal sinus and nearby major cortical veins.

Clusters of arachnoid villi, called **arachnoid granulations**, are grossly visible.



IN GENERAL CSF WILL ACCUMULATE(**HYDROCEPHALUS**) DUE TO:

- ✓ Increase production
- ✓ Decrease absorption
- ✓ Obstruction to flow



PATHOPHYSIOLOGY OF HYDROCEPHALUS



- Clinically---intracranial hypertension
- If progressive ventricular dilatation separates ependymal cells lining the ventricles, interstitial cerebral edema will develop.

Normal ICP by age

Age	ICP range (mmHg)
Neonate	< 2
Infant	1.5–6
Young child	3–7
Adolescent (> 15 years)	< 15
Adult	< 15

(Modified from Greenberg, 2001.⁷⁰)

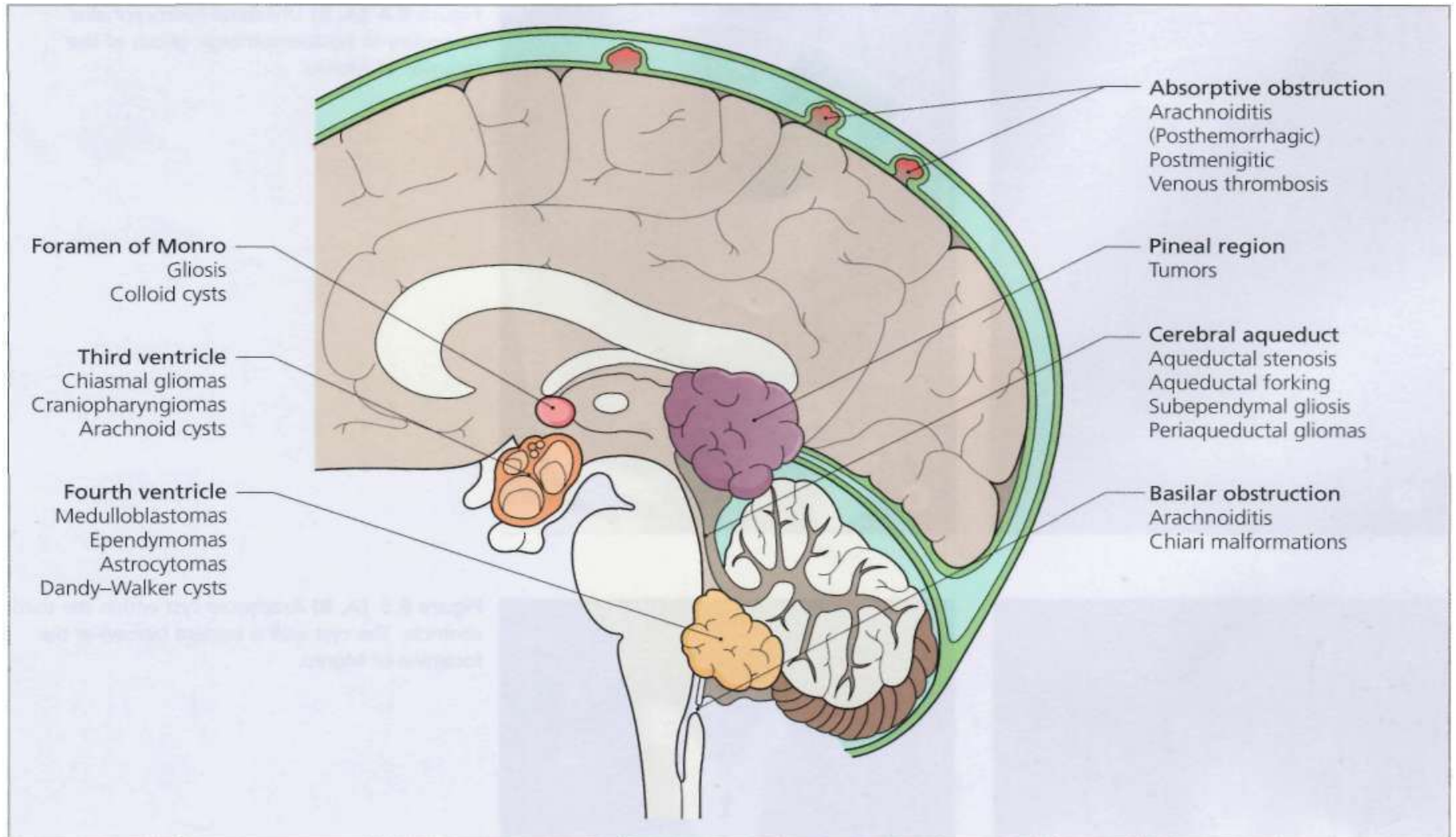
CLASSIFICATIONS

- **Communicating** or **Non-communicating**.
- **Congenital** versus **Acquired**.
- Its symptomatology as either **compensated** or **noncompensated**.
- Its chronicity: **acute versus chronic** hydrocephalus.
- **Internal** or **external**

CAUSES OF HYDROCEPHALUS:

- Hydrocephalus is almost always a result of an interruption of CSF flow and is rarely because of increased CSF production.

Common sites and causes of CSF obstruction.

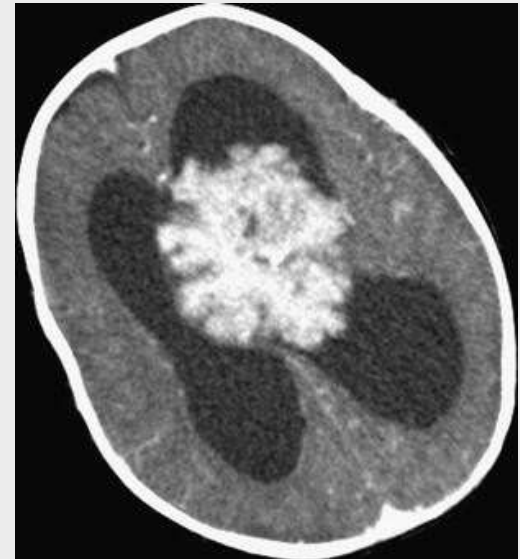


AETIOLOGY - SITE

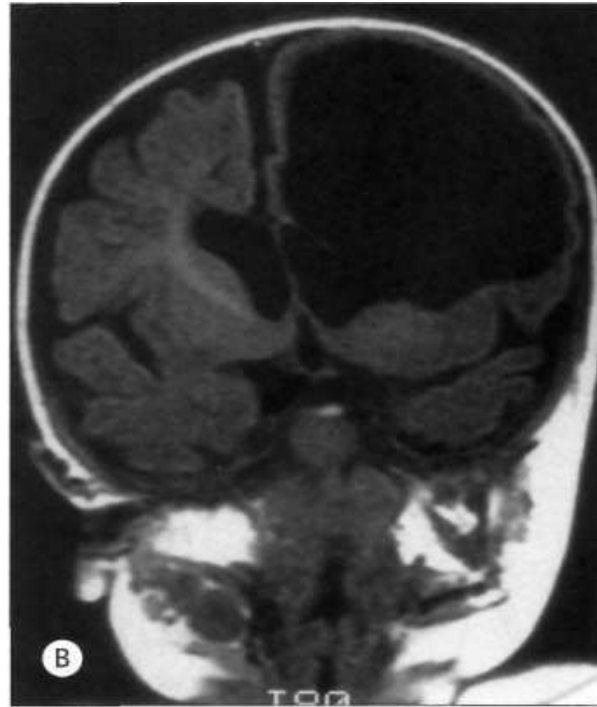
1. Lateral ventricles

Choroid plexus tumors:

- Rare in the general population(0.4 to 0.6% of all CNS tumors).
- With CSF production rates three to four times the normal.
- Endoscopic coagulation of the choroid plexus or surgical removal of the papilloma .



2-Foramina of Monro

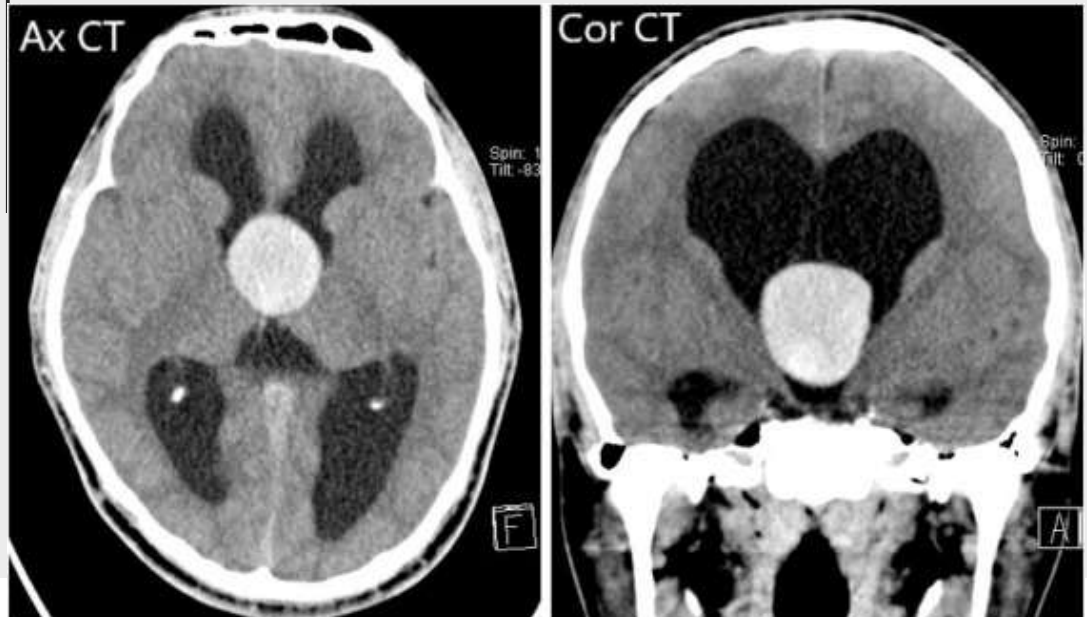


3-Third ventricle

Cysts and neoplasms within the third ventricle commonly cause hydrocephalus.

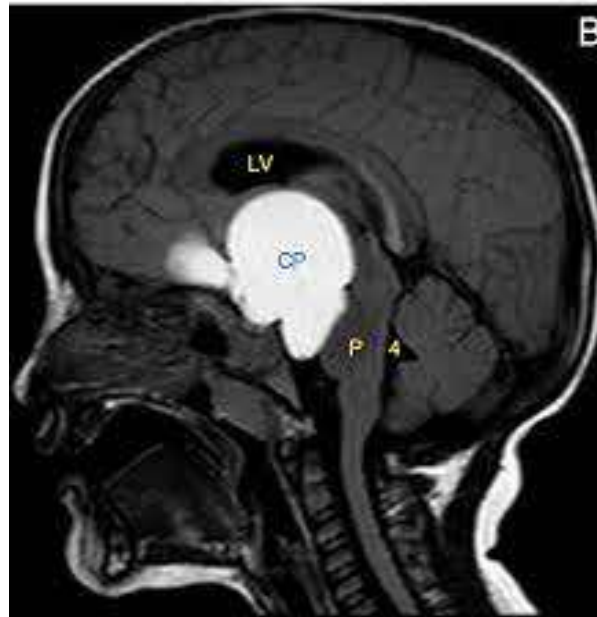
-**Colloid cysts** are uncommon neoplasms

-**Ependymal and arachnoid cysts** within the third ventricle
The endoscopic fenestration is a treatment option.



Third ventricle

- The most common pediatric neoplasms that obstruct the third ventricle are craniopharyngiomas and chiasmal-hypothalamic gliomas.

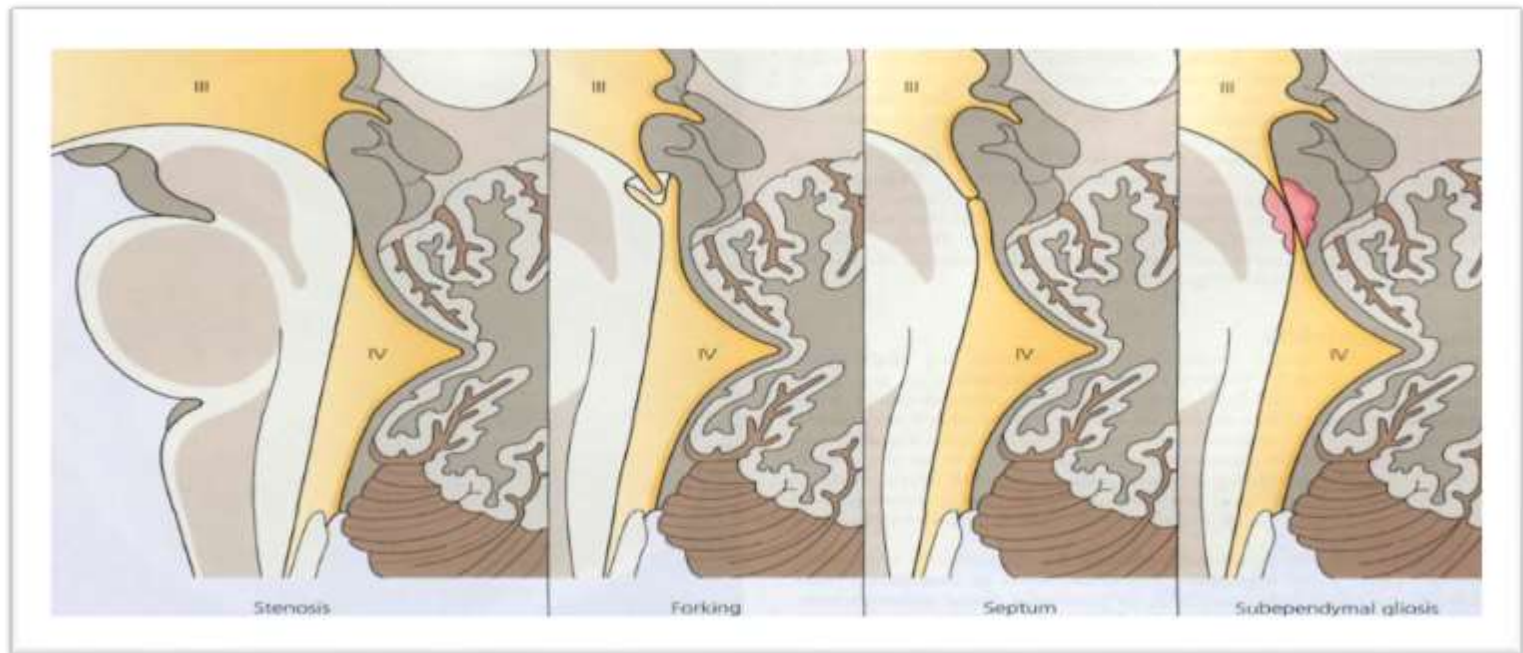


4-Sylvian aqueduct

The normal aqueduct of a neonate is 12-13 mm in length and only 0.2-0.5 cm in diameter,

Thus, it is prone to obstruction from a variety of lesions, including **congenital aqueductal stenosis** (classified as true stenosis, forking, septum, or subependymal gliosis)

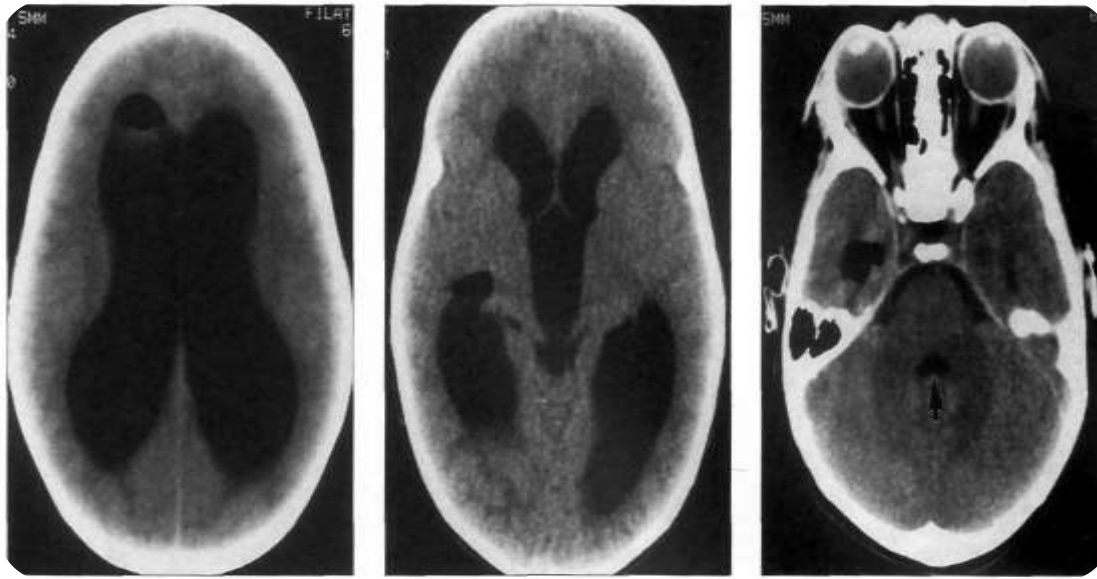
It may be secondary to *in utero infections (e.g. toxoplasmosis)*, *intraventricular hemorrhage*, or *mumps encephalitis*.



Aqueductal congenital malformations: (from left to right) stenosis, forking, septum, and subependymal gliosis.

Sylvian aqueduct stenosis

- Is the **commonest cause** of congenital hydrocephalus
- Less than 2% of cases -recessively inherited X-linked Bickers-Adams-Edwards syndrome, which is associated with flexion-adduction of the thumbs ("cortical thumbs").



CT scan of infant with aqueductal stenosis, demonstrating lateral and third ventricular distension, separation of the thalami, and compression of the cerebral hemispheres . The fourth ventricle (arrow) is normal.

Sylvain aqueduct

Pineal region neoplasms,

Arteriovenous malformations, and Periaqueductal neoplasms.

Low-grade astrocytomas are the most common periaqueductal pediatric neoplasms that cause hydrocephalus.



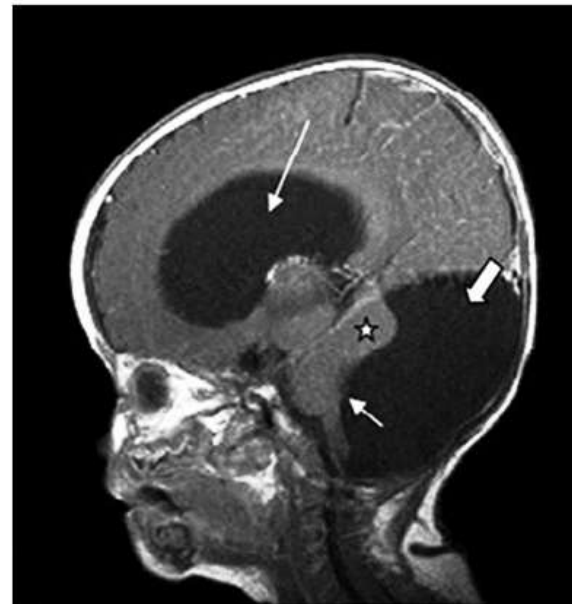
4-Fourth ventricle

In infants, the fourth ventricle is the location for obstruction secondary to Dandy-Walker cysts .

In older children, neoplasms are a common cause.

Dandy-Walker cysts are developmental abnormalities characterized by a large cyst in the fourth ventricle, hypoplasia of the cerebellar vermis, and atrophy of the cerebellar hemispheres.

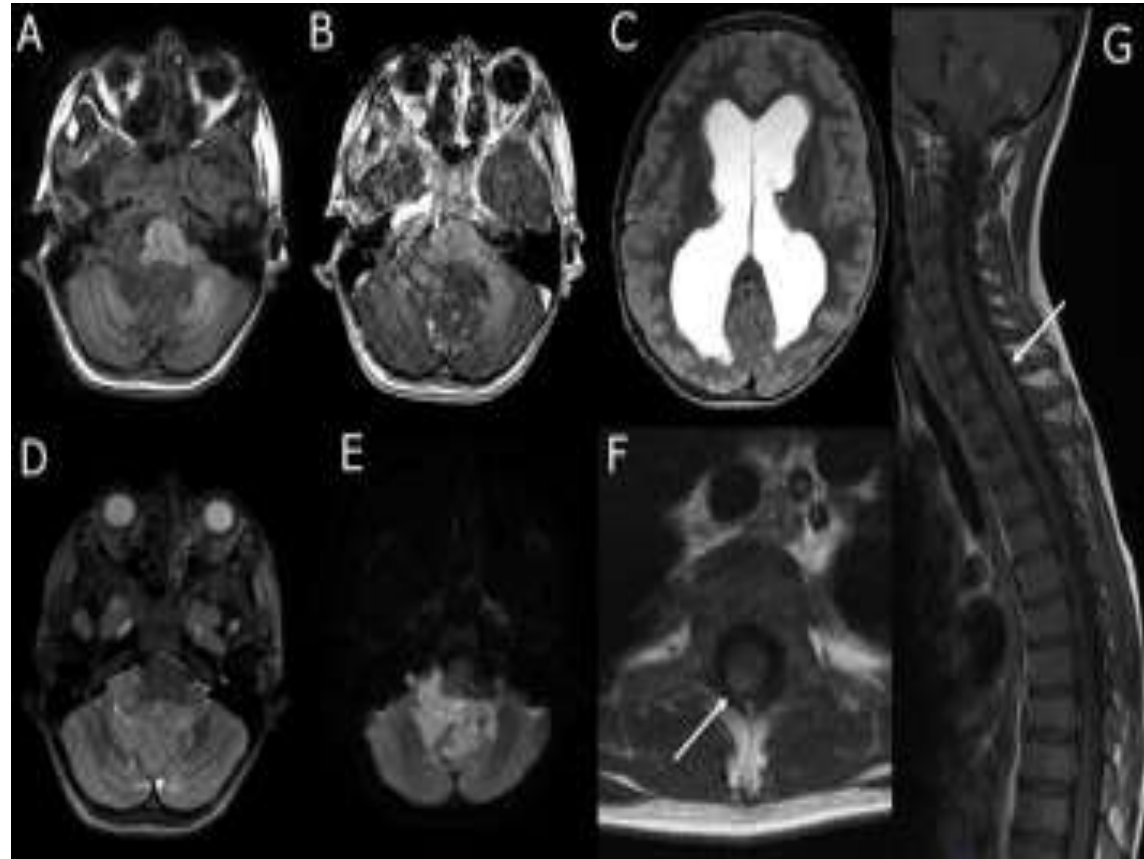
Over 85% of children with Dandy-Walker cysts have hydrocephalus.



4TH Ventricle tumors

Hydrocephalus is associated with

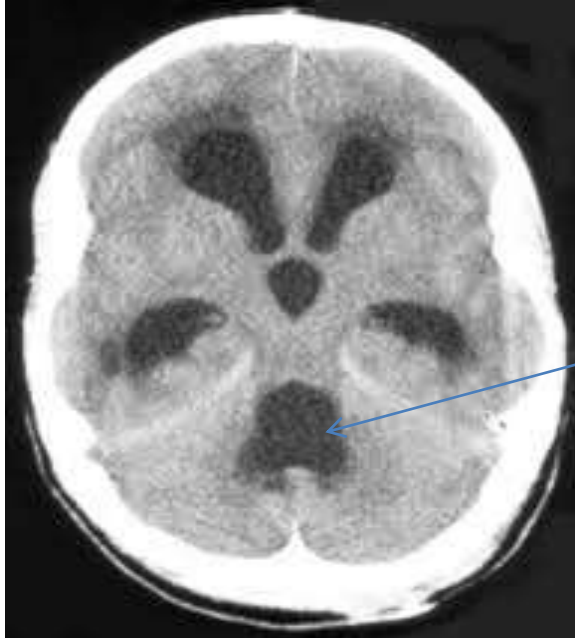
- 85% of **medulloblastomas**,
- 65% of posterior fossa **astrocytomas**,
- 75% of **ependymomas**,
- and 25% of **brainstem gliomas**.



5-Arachnoid granulations

Sclerosis or scarring of the arachnoid granulations can occur after meningitis, subarachnoid hemorrhage, or trauma.

Occasionally can be seen in cases with disseminated meningeal malignancies(meningeal carcinomatosis)

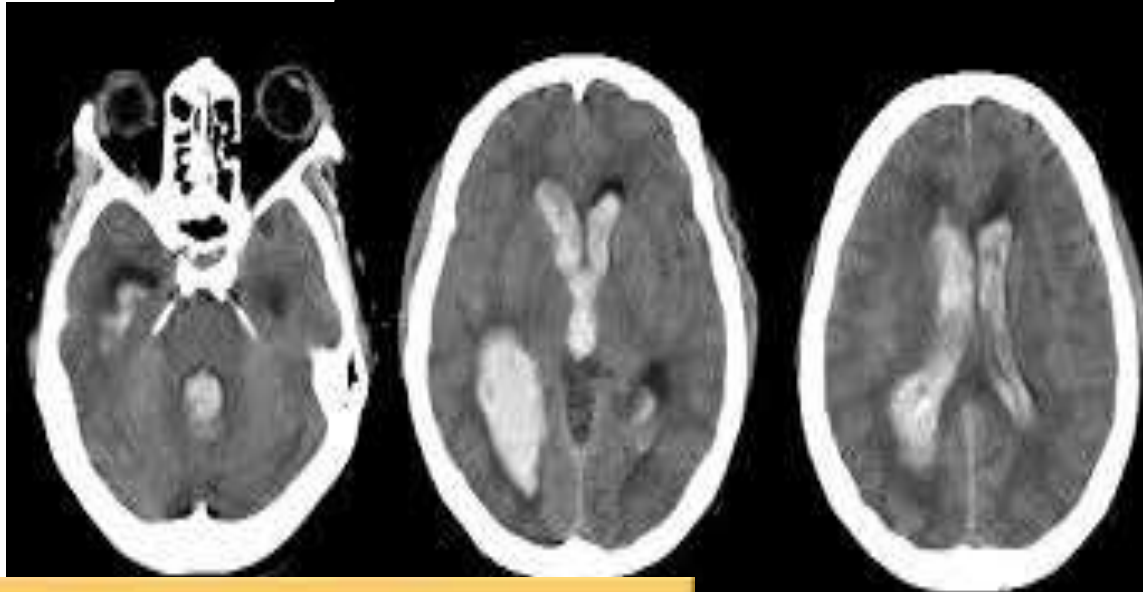


Dilated fourth ventricle

Etiology by age

1-Premature infants

Hydrocephalus in premature infants is predominantly caused by (intraventricular hemorrhage) IVH. The hemorrhage occurs in the germinal matrix,



Intracranial hemorrhage

Classification according to Papile

- Grade 1.** Hemorrhage limited to subependymal matrix
- Grade 2.** Hemorrhage extending into ventricular system, < 50%, without acute ventriculomegaly
- Grade 3.** Hemorrhage extending into ventricular system, with acute dilatation because of flooding of 50% or more of one or both lateral ventricles
- Grade 4.** Hemorrhage grade 1, 2 or 3 with extension into brain tissue

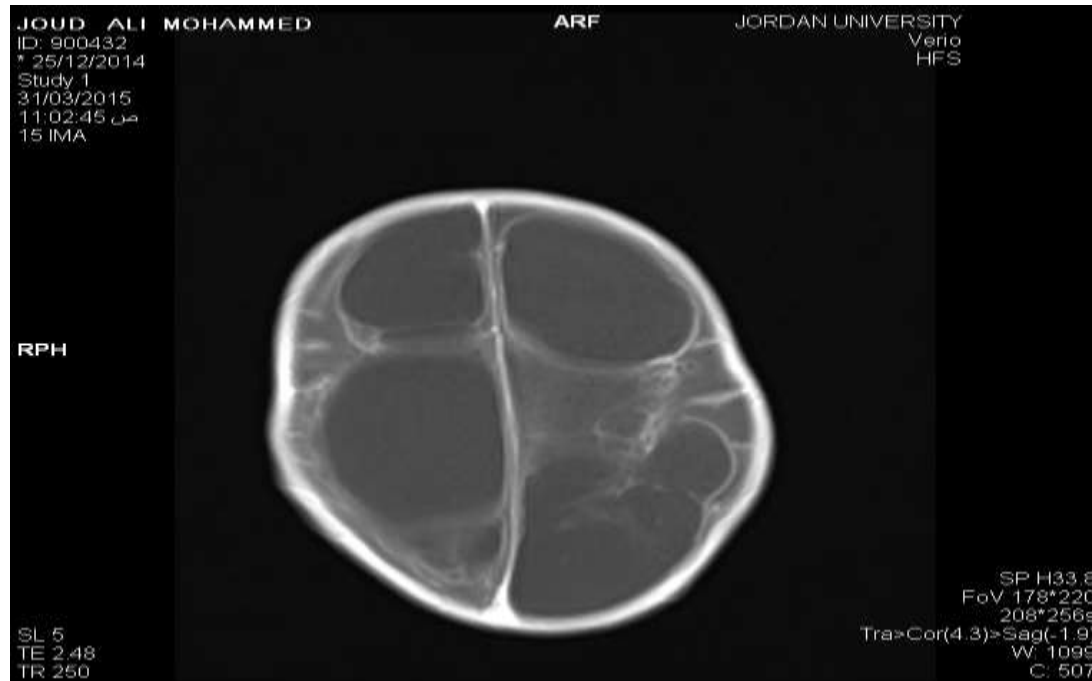
2-FULL TERM INFANT

- Aqueduct stenosis
- Chiari malformation
- Dandy walker malformation
- Cerebral malformation
- ..Encephalocele
- Neoplasm
- Vein of Galen aneurysm



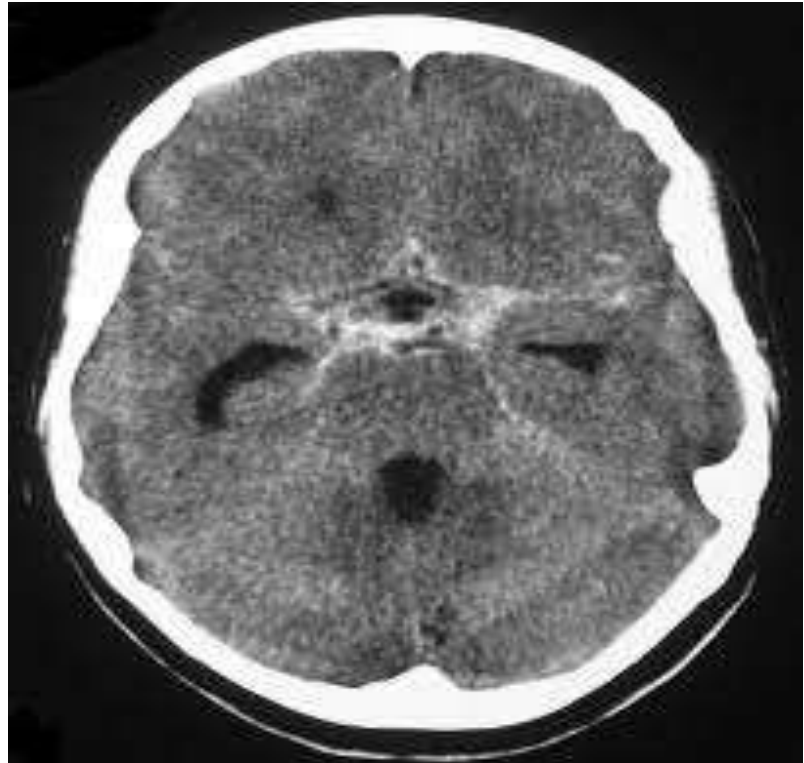
3-Older children

Hydrocephalus after infancy is usually secondary to trauma, meningitis or neoplasms.



4-Adults

- 30% idiopathic
- The rest :
 - Subarachnoid hemorrhage
 - Head injury
 - Brain tumors
 - Cranial surgery
 - Aqueduct stenosis
 - Meningitis



SIGNS AND SYMPTOMS

Premature infants	Infants	Toddlers and older
Apnea	Irritability	Headache
Bradycardia	Vomiting	Vomiting
Tense fontanelle	Drowsiness	Lethargy
Distended scalp veins	Macrocephaly	Diplopia
Globoid head shape	Distended scalp veins	Papilledema
Rapid head growth	Frontal bossing Macewen's sign Poor head control Lateral rectus palsy "Setting-sun" sign	Lateral rectus palsy Hyperreflexia/clonus



Full and tense
anterior fontanelle

Dilated scalp
veins

Suture
widening

Setting sun
eyes

Head circumference

- Normal head circumference for full-term infants is **33-36 cm** at birth.
- Head circumference increases by 2 cm/month during the first 3 months, by 1 cm/month from 4 to 6 months, and by 0.5 cm/month from 7 to 12 months.



Signs of hydrocephalus in children and adults

- Fundoscopic examination showing advanced optic disc swelling suggestive of frank papilloedema resulting from intracranial hypertension

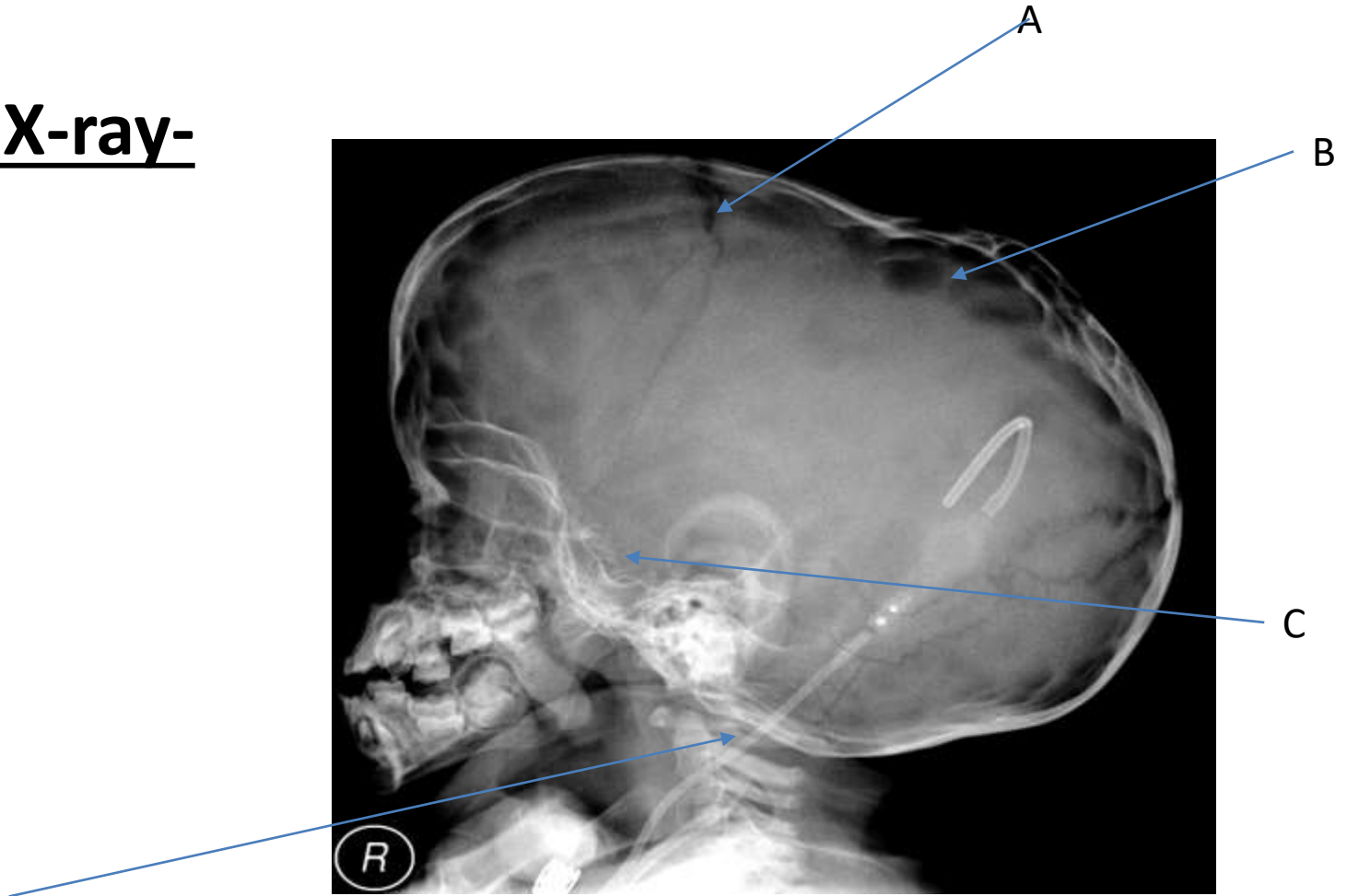


Right abducent nerve palsy



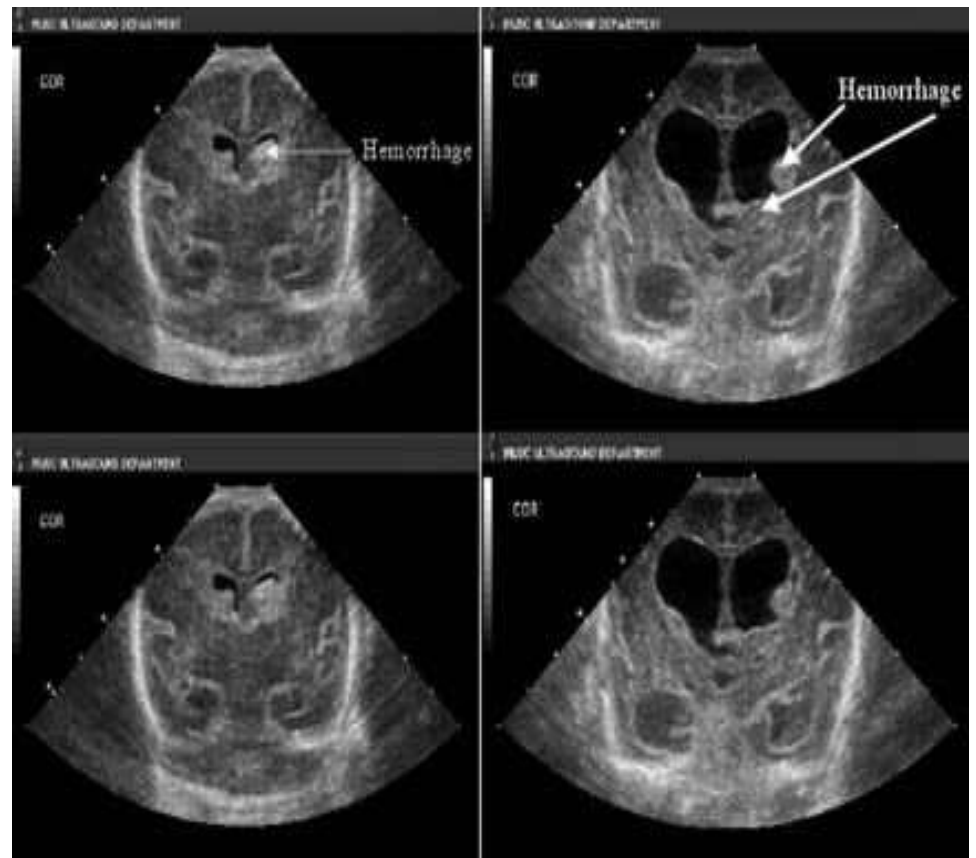
DIAGNOSTIC STUDIES

- Skull X-ray-



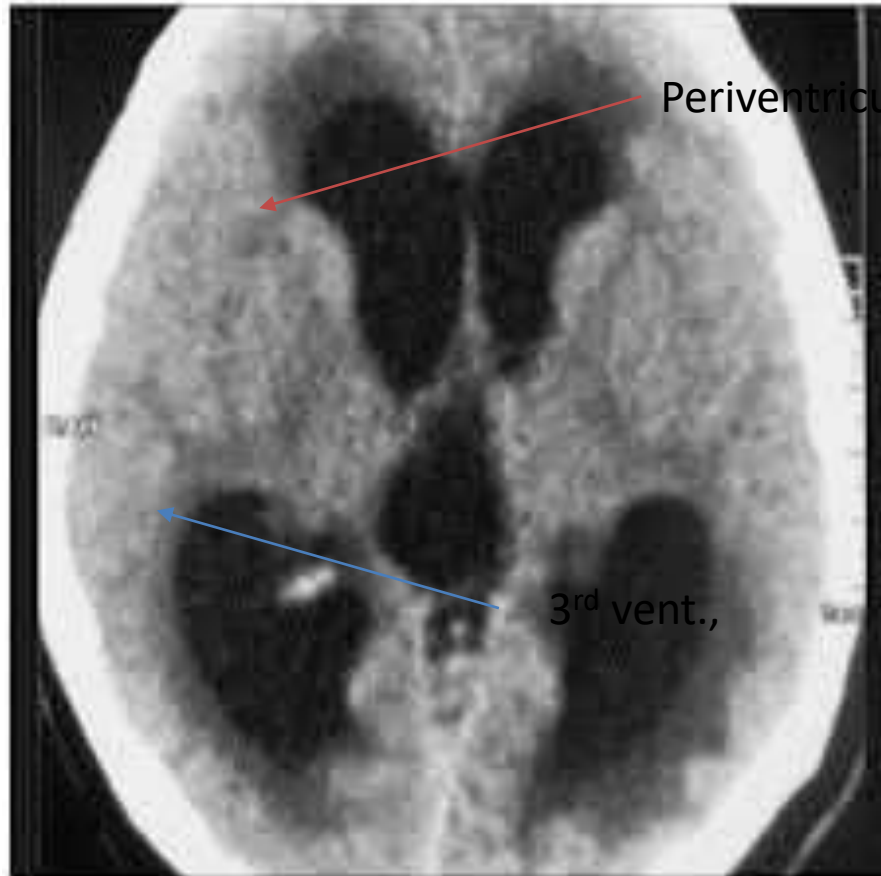
Cranial ultrasonography

- Evaluation of premature infants with IVH, & the detection and monitoring of ventricular size.



CT scanning

- From 1976 to 1986, CT scanning was the definitive method of diagnosing pediatric hydrocephalus.
- limitations: only in the **axial plane**,
Require **irradiation**, and have **less resolution** than MRI.



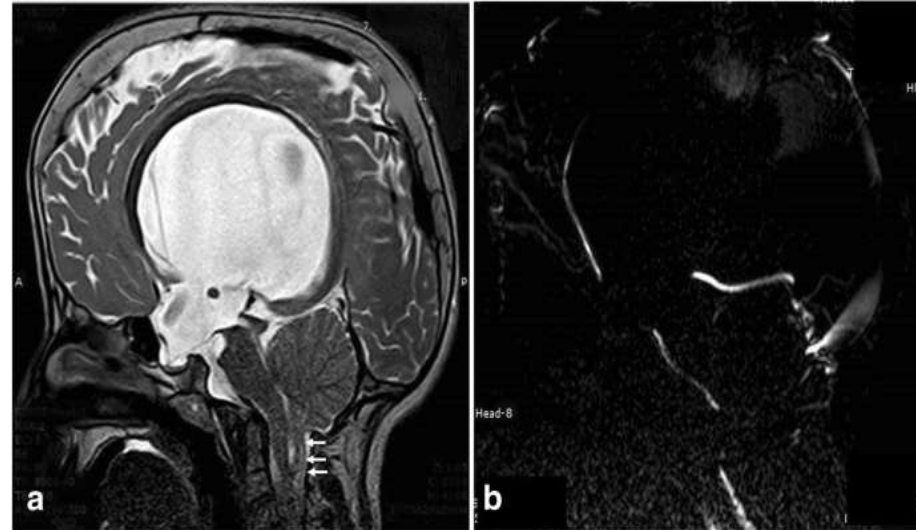
Periventricular fluids

3rd vent.,

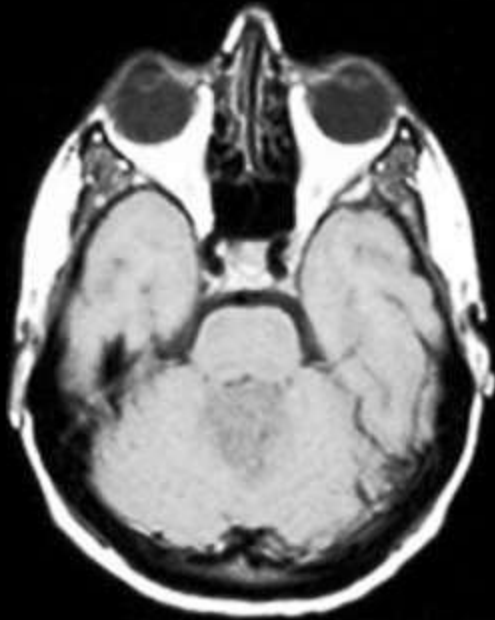
Brain MRI

Since 1986,

- can project the brain in axial, coronal, and sagittal projections.
- better soft tissue resolution
- determine CSF flow across the aqueduct.
- no radiation.



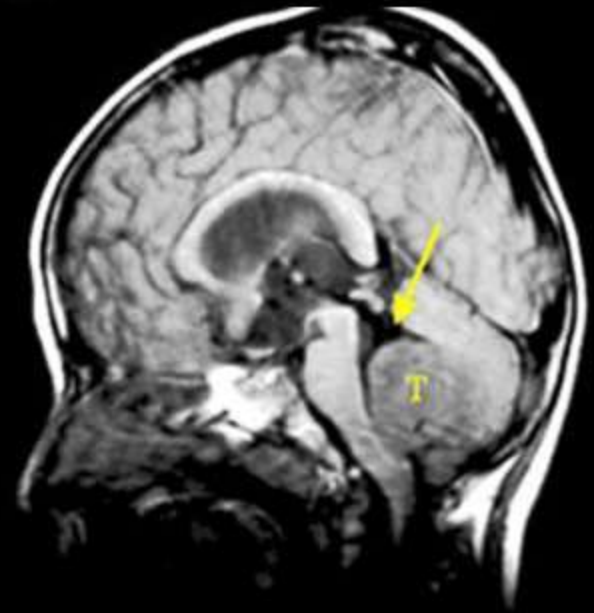
The same previous infant, MRI was done postoperatively after V-P shunt insertion. You can notice the reduction of the size of the ventricles with the posterior fossa tumor that caused the non communicating hydrocephalus



Pre-contrast axial T1 Wtd MRI
A



Post-contrast axial T1 Wtd MRI
B



Pre-contrast sagittal T1 Wtd MRI
C

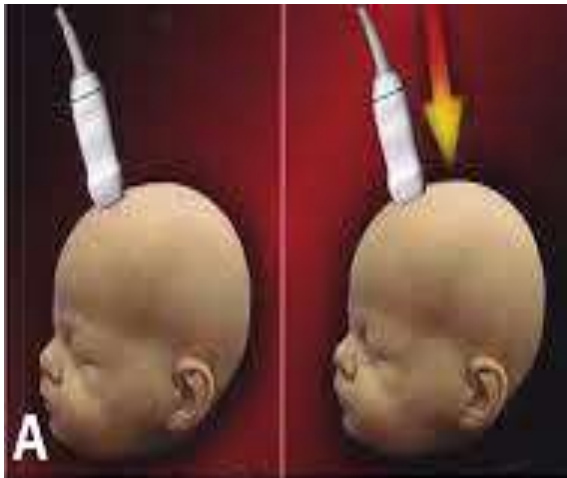
Treatment

- The goal is:
 1. Optimal neurological outcome
 2. Preservation of cosmesis
 - Normal sized ventricles should not be considered the goal
-
1. Non surgical
 2. Surgical
 - Shunting
 - Non shunting



Non surgical

- Not definitive treatment.
- Acetazolamide and frusamide
- Serial LP or ventricular tap
- Head rapping ,radioactive gold.....



Transfontanelle u/s
preparing for tapping
of the ventricles



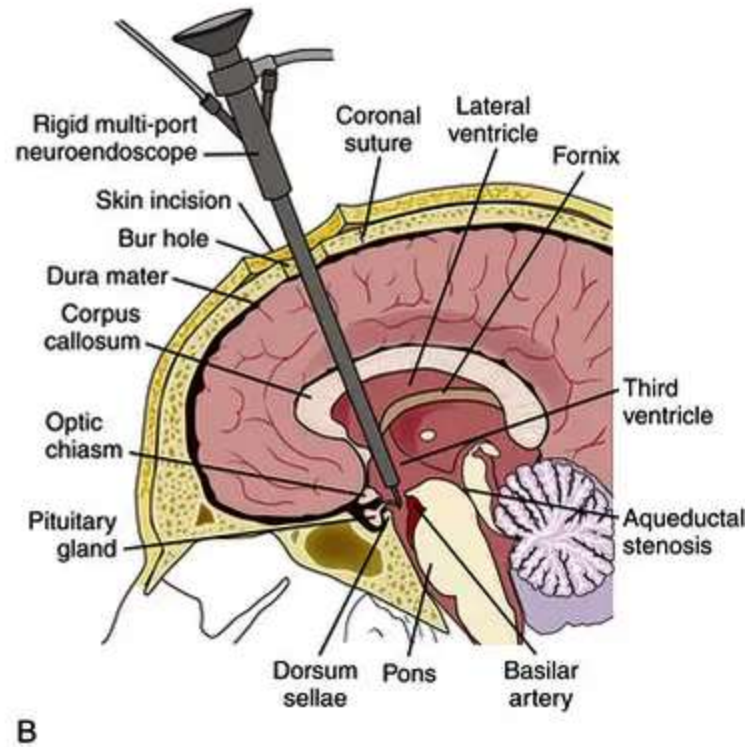
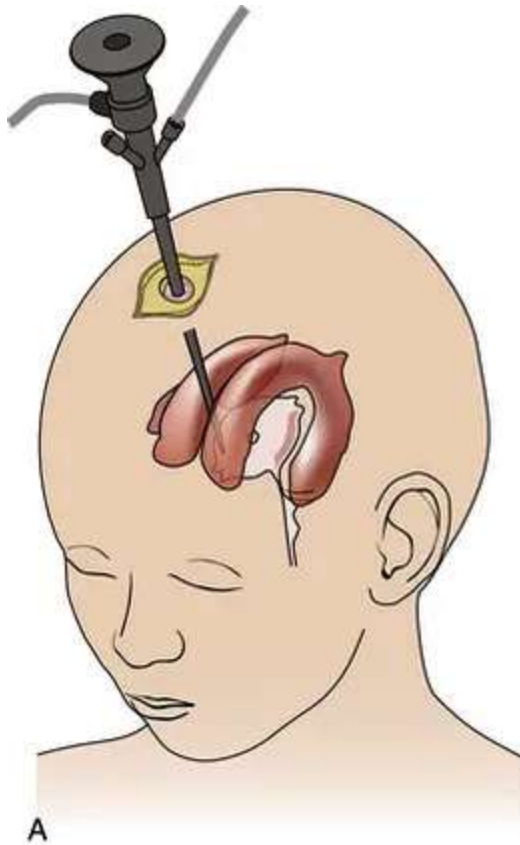
Lumbar puncture to
drain the CSF

Surgical – non- shunting options

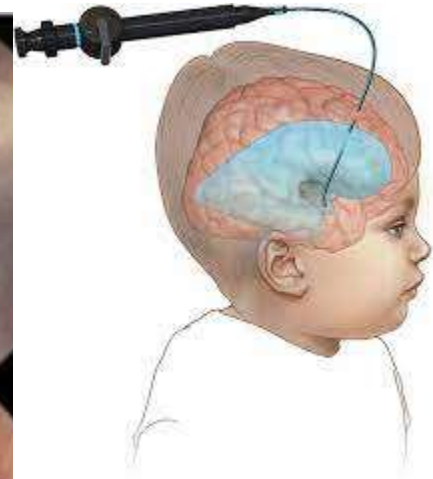
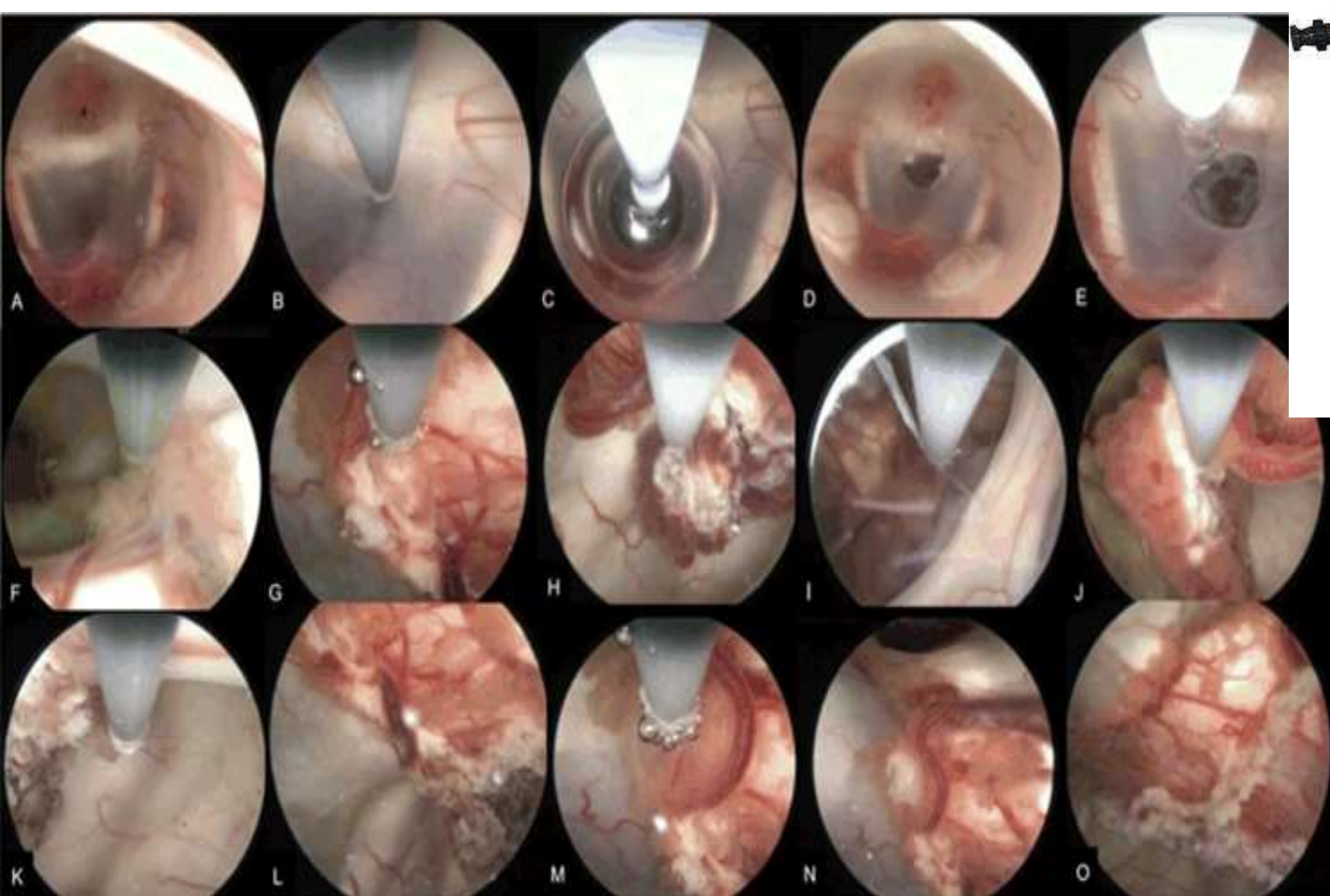
1-Whenever possible, the obstructing lesion that causes the hydrocephalus should be *surgically removed.*

2-For CSF obstruction at or distal to the aqueduct, a potential surgical treatment is the *endoscopic third ventriculostomy(ETV).*

Endoscopic Third Ventriculostomy(ETV)

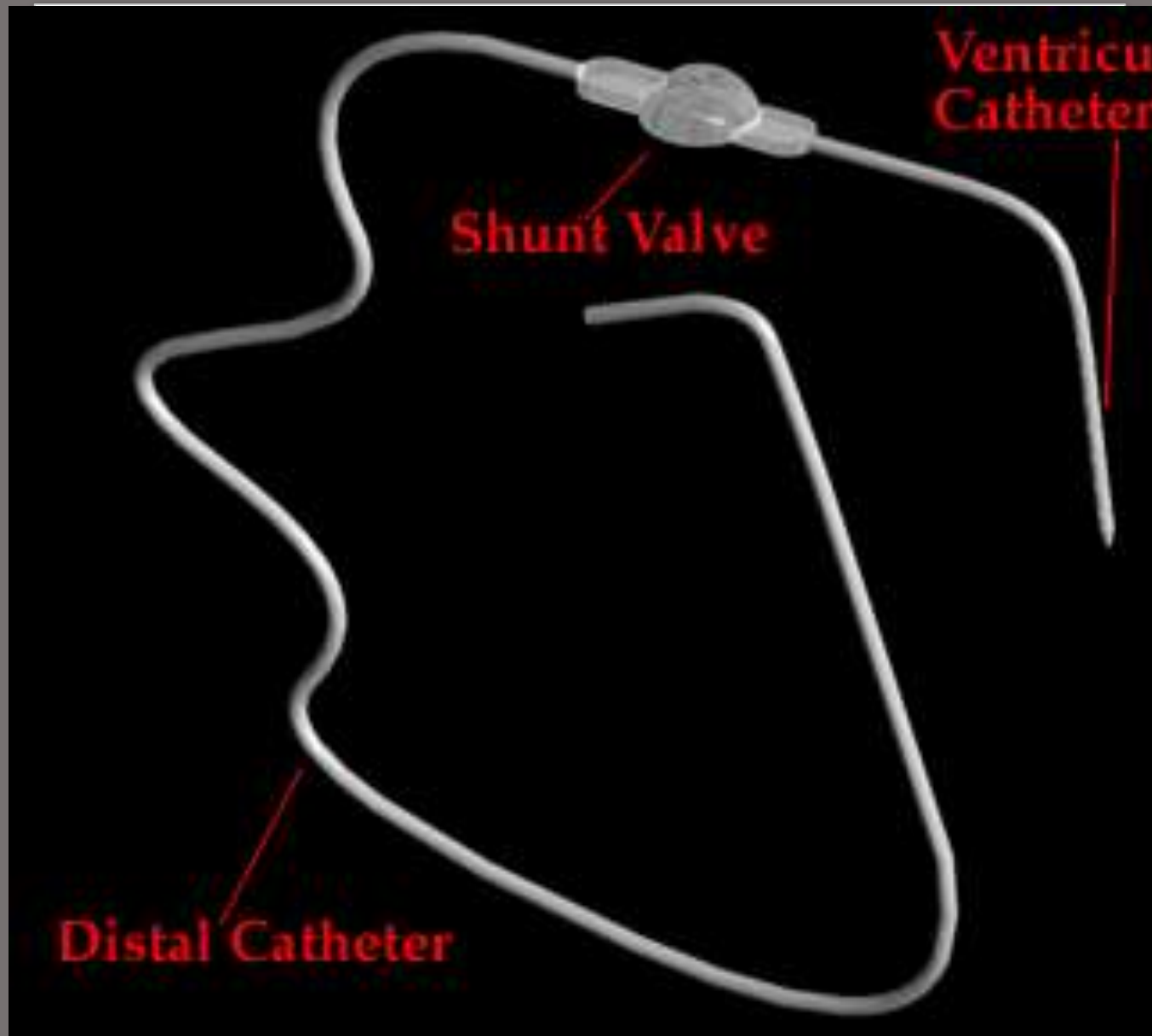


ETV-CPC-choroid plexus coagulation



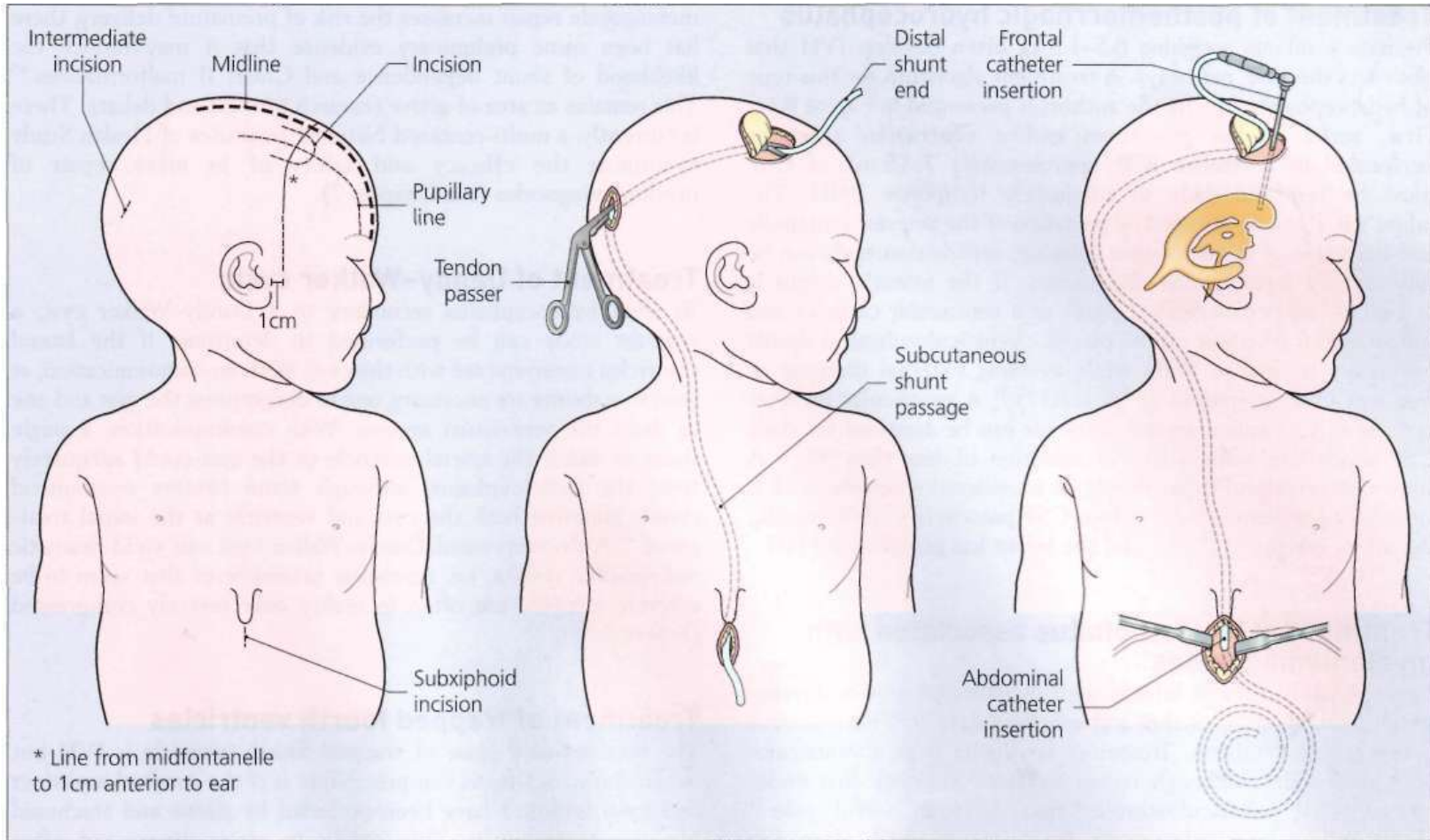
Surgical - CSF shunts

- CSF shunts are usually silastic tubes that divert CSF from the ventricles to other body cavities (i.e. peritoneal, atrium or pleural space), where normal physiologic processes can absorb the fluid.



Components
of the shunt

Surgical techniques



Placement of a frontal ventriculoperitoneal shunt. Patient positioned and coordinates marked; subcutaneous shunt passage; ventricular catheter insertion; peritoneal catheter insertion.

Shunt complications ?a tube ?a foreign body

Shunt complications and failures remain a significant problem in treating hydrocephalus.

- (1) mechanical failure of the device,
- (2) functional failure because of too much or too little flow of CSF(malfunction), and
- (3) infection of the CSF or the shunt device.

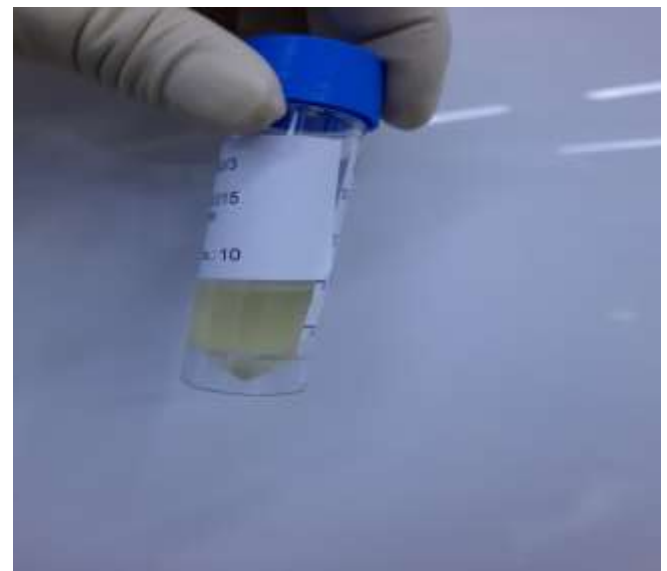
Shunt complications

Common complications	Uncommon complications			
	Cranial	Subcutaneous	Peritoneal	Atrial
Infection	Subdural hygroma	Shunt migration	Peritonitis	Endocarditis
Obstruction	Subdural hematoma	Shunt disconnection	Pseudocysts	Nephritis
Inadequate flow or overdrainage	Hemiparesis Hematoma	Shunt fracture	Perforation Hernias	

Shunt Infection

- In general, approximately 1-15% . Premature infants have an increased risk.
- Nearly 90% of all shunt infections are recognized within 12 months
- it is believed that most bacteria are introduced at the time of surgery.
- ***Staphylococcus epidermidis*** causes approximately 60% of shunt infections,
- ***Staphylococcus aureus*** is responsible for 30%,
- *and* coliform bacteria, propionibacteria, streptococci, or *Haemophilus influenzae* cause the remainder.

- **Diagnosis** is confirmed by CSF sampling from the shunt reservoir ;with the findings of leukocytosis and positive culture .
- **Treatment** usually involves
 - removal of the infected shunt
 - placement of an EVD.
 - appropriate intravenous antibiotics, based on culture and sensitivity results.
- When the infection is cleared, i.e. (1) at least 3 consecutive daily CSF cultures that are negative, (2) CSF white blood cell count < 50, and (3) CSF protein < 500 mg/dL, a new shunt system is implanted, and the EVD is removed.

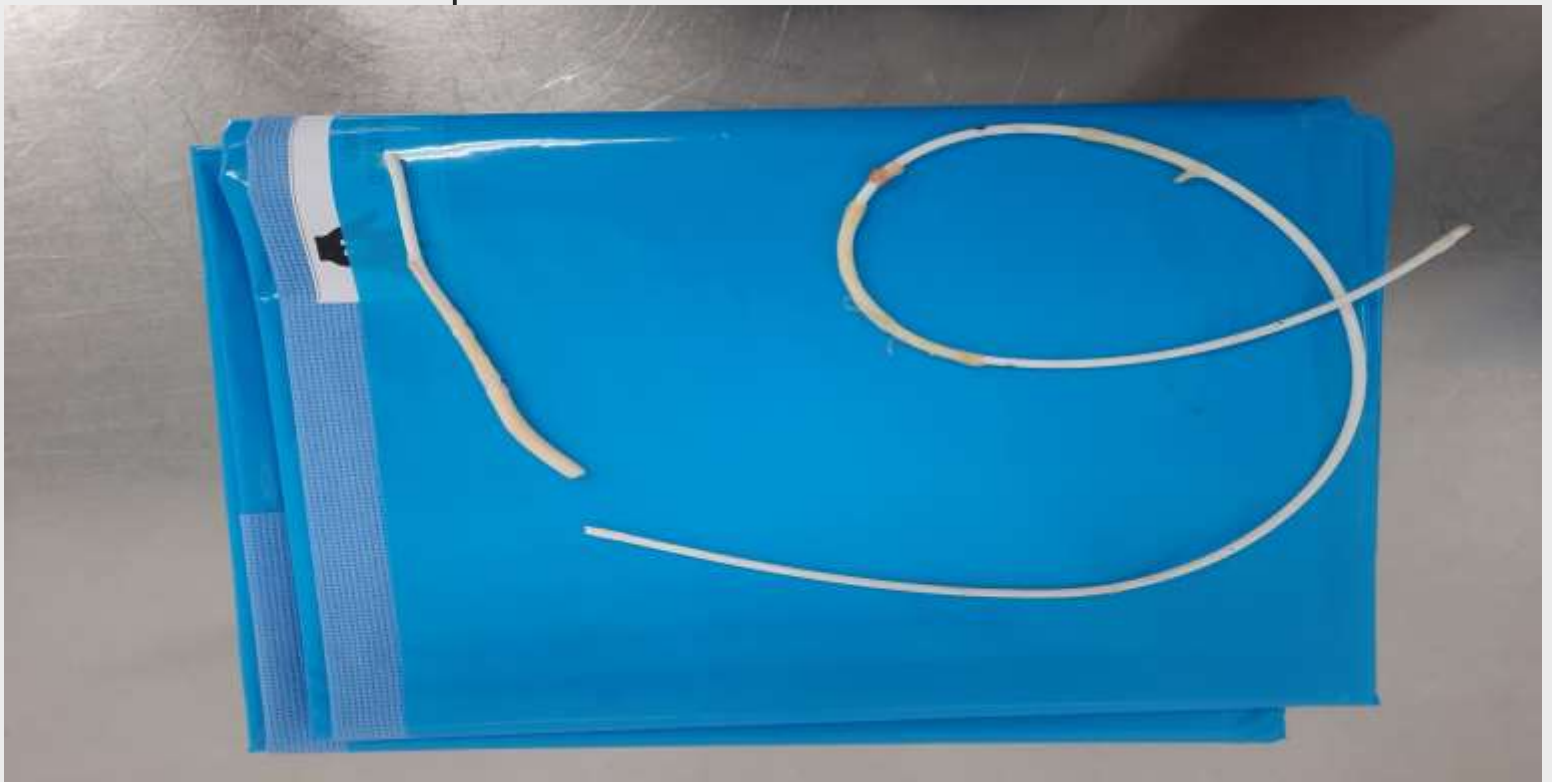


shunt obstruction

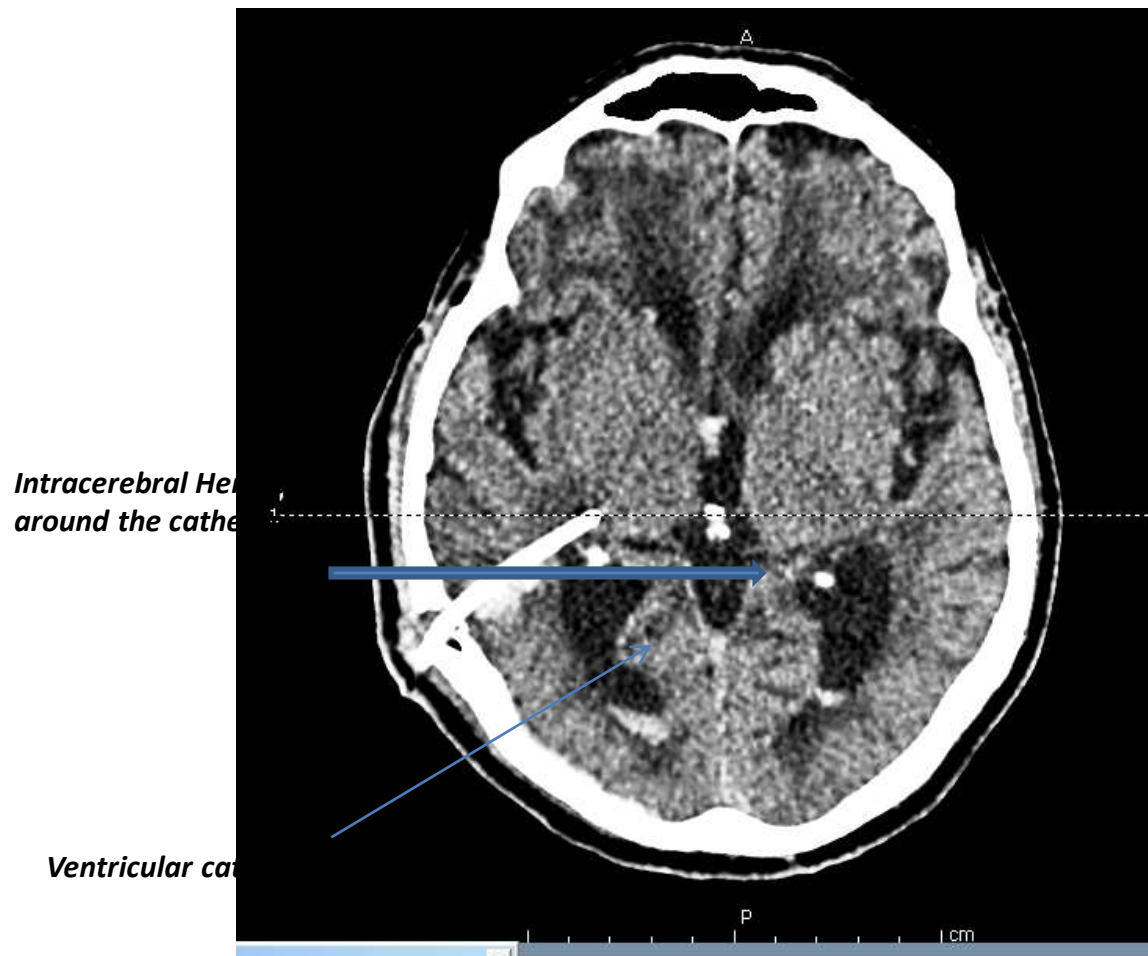
- A child with a shunt malfunction often presents with signs and symptoms of increased ICP.
- headache, irritability, lethargy, nausea, and/or vomiting.
- The shunt itself can be examined for evidence of obstruction..
- A head CT, as well as shunt survey Xrays

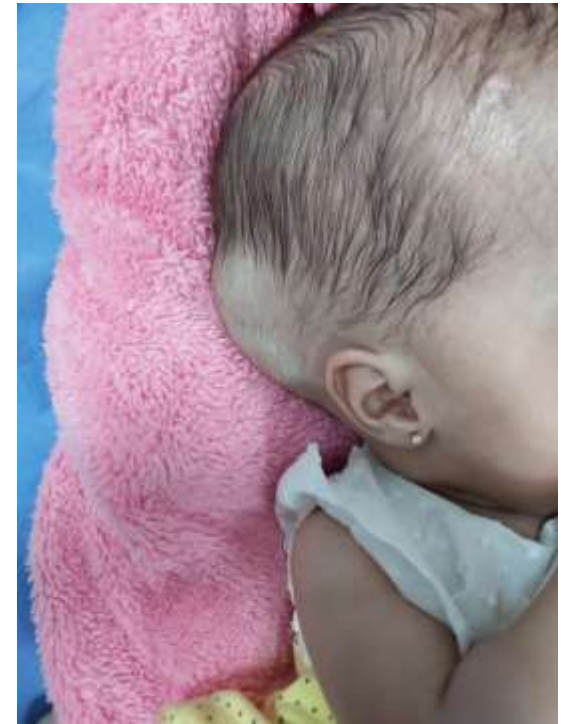
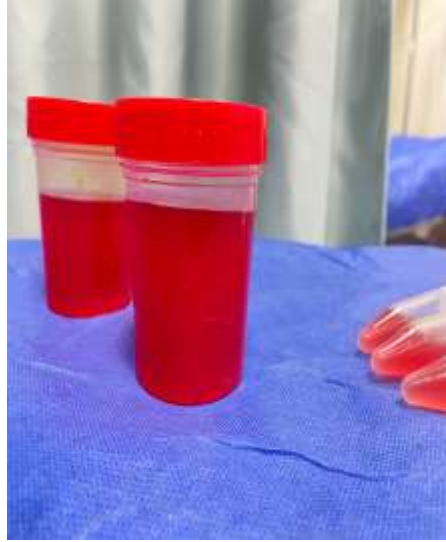


- Shunt obstructions/malfunctions are treated by replacing the occluded or nonfunctioning components, or by replacing the entire system.



Hematoma at sit of shunt insertion





Swelling around the valve of the shunt highly suggestive of shunt malfunction





Chronic or normal pressure hydrocephalus (NPH)

Chronic hydrocephalus of adulthood presents more insidiously, often weeks or even years after the inciting cause, sometimes without apparent cause at all.

- . The affected patient exhibits a combination of **motor** dysfunction, **urinary** incontinence, and **Dementia** the so called Hakim -Adams triad.

The early motor signs are most prominent in the lower extremities and are related to difficulty initiating walking, the so-called "magnet gait" phenomenon.

- . Advanced cases show frontal release signs, such as suck and grasp reflexes.

Parkinsonian and other dyskinesias have been described. In the early stages of hydrocephalus, the patient is well aware of the urge

to void, but urinary incontinence results from an uninhibited bladder and a gait-induced inability to reach the bathroom in time.