Hydrocephalus

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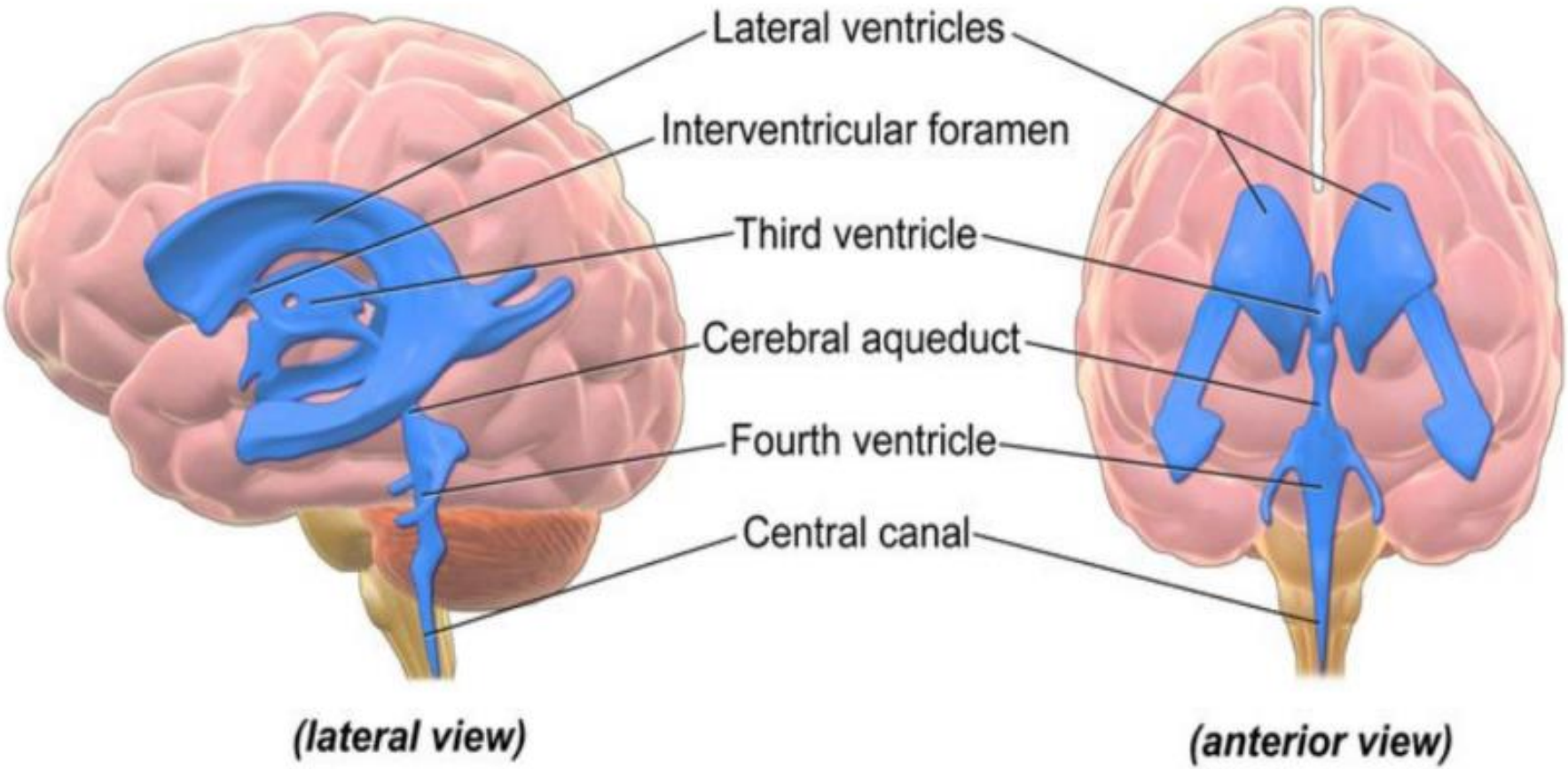
Outlines

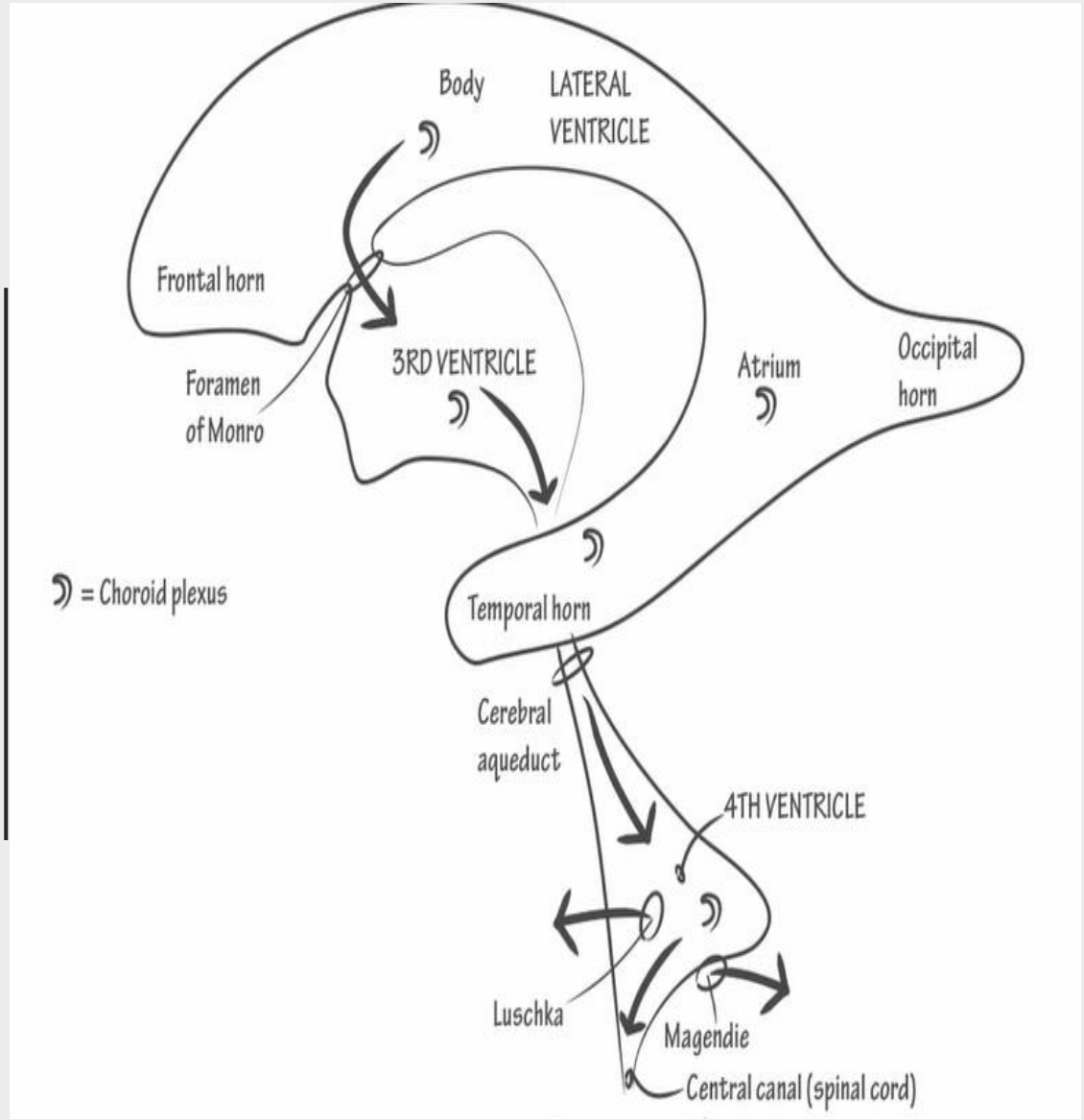
- Definition
- Pathophysiology
- Etiology
- Types
- Clinical presentation
- Diagnosis
- Treatment

What is hydrocephalus?

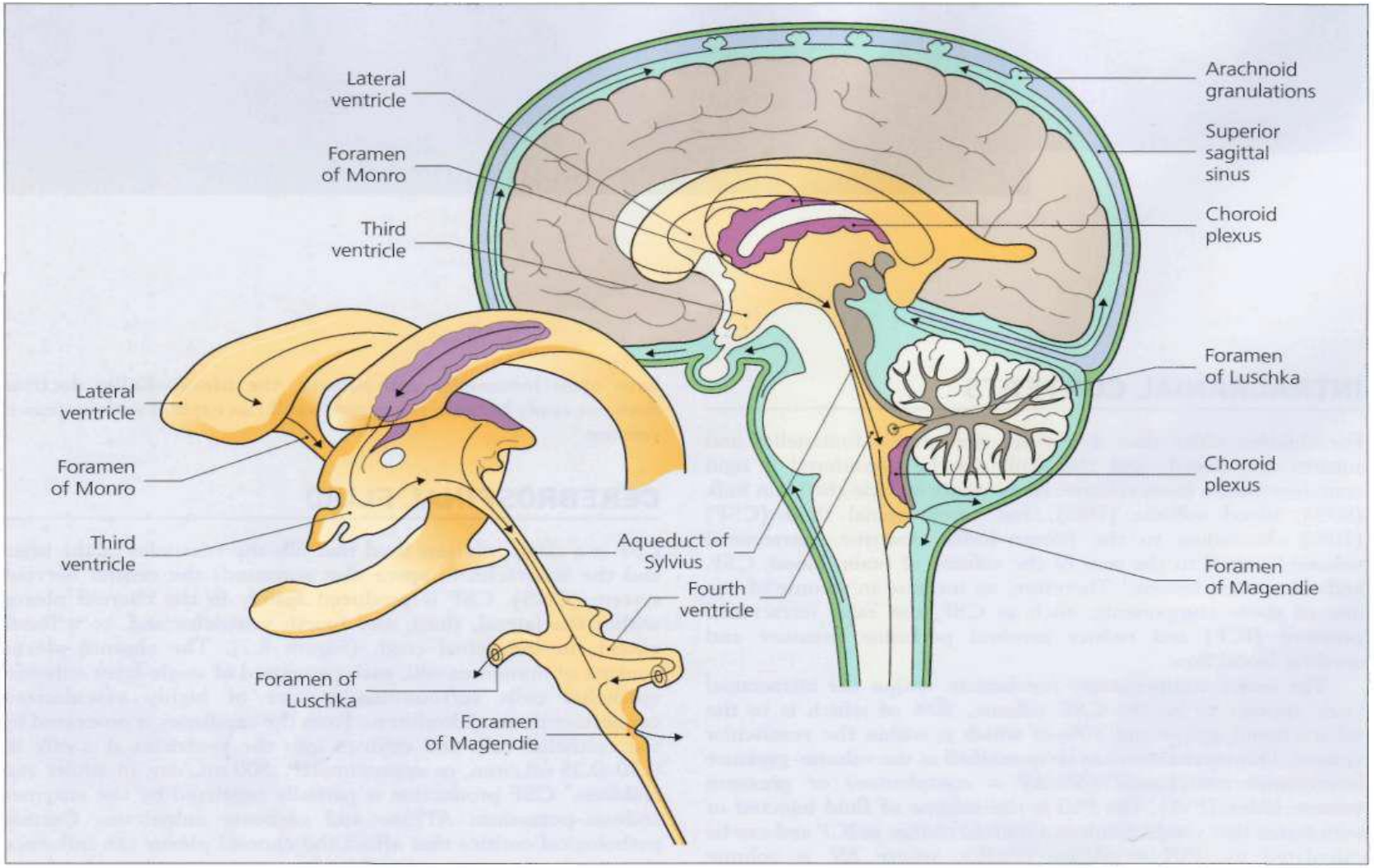
- Is an accumulation of cerebrospinal fluid (CSF) within the ventricular space at an inappropriate pressure.
- Estimated prevalence is 1-105%
- Incidence of congenital cases is 0.9-1.8/1000 live birth.


Lateral ventricles
Interventricular foramen
Third ventricle
Cerebral aqueduct
Fourth ventricle
Central canal
(lateral view)
(anterior view)

Physiology of CSF circulation



The ventricular system



The cerebrospinal fluid (CSF)production

- CSF is produced mainly in the choroid plexus.
- The choroid plexus=single-layer cuboidal epithelial cells surrounding a core of highly vascularized connective tissue.
- Rate o: 0.30-0.35 mL/min, or approximately 500 ml/day, in adults and children.
- CSF production is partially regulated by :

-sodium-potassium ATPase …and

- carbonic anhydrase.

What is cerebrospinal fluid(CSF)?

- Clear colorless fluid
- Total volume: 150mls(40-50 mL in neonates and 65-140 mL in children).
- 25mls in ventricles + 125mls in subarach.space
- Production: 500mls/day and turnover: 3-4 times
- **Contents:**

1.Acellular (<5 lymphocytes, if higher: Pleocytosis)

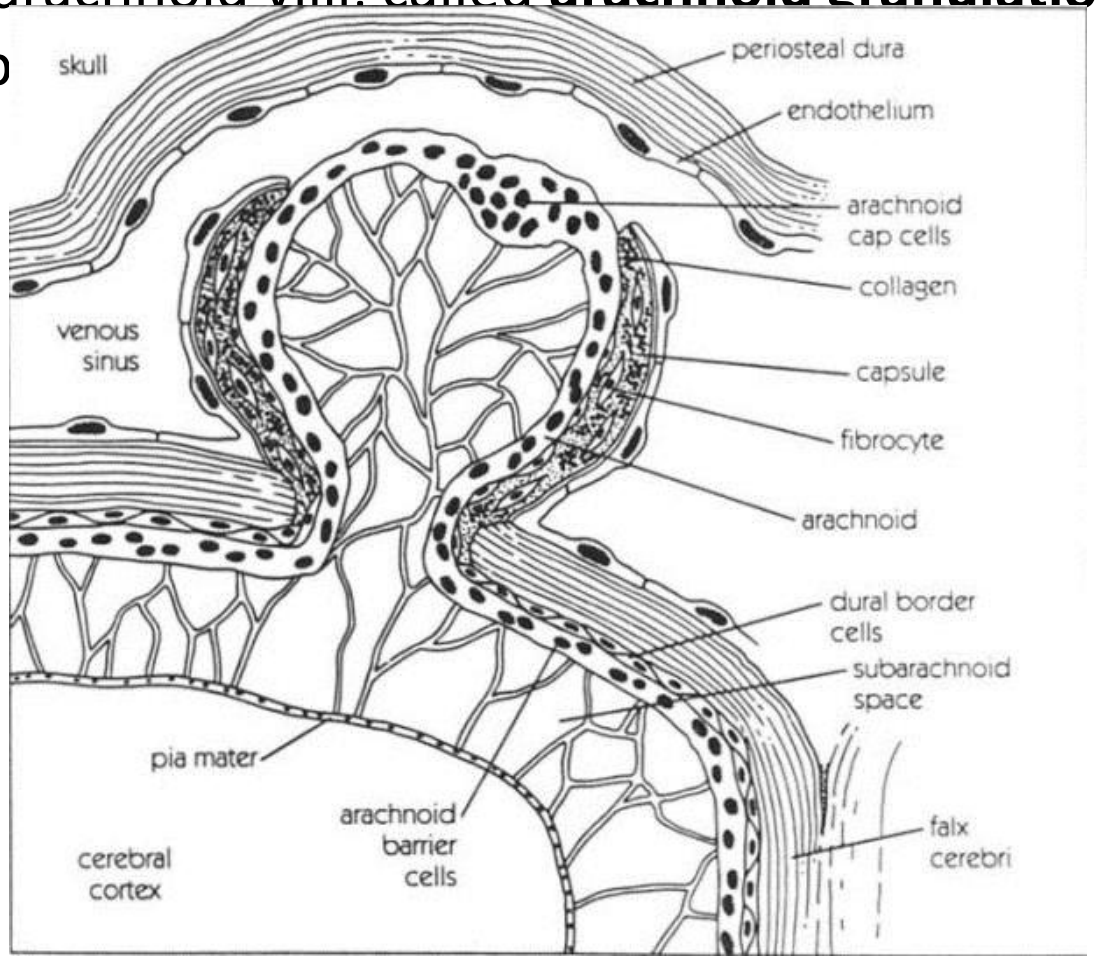
2. Similar Na+/ higher Cl-/ less K+ compared to plasma

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 visib



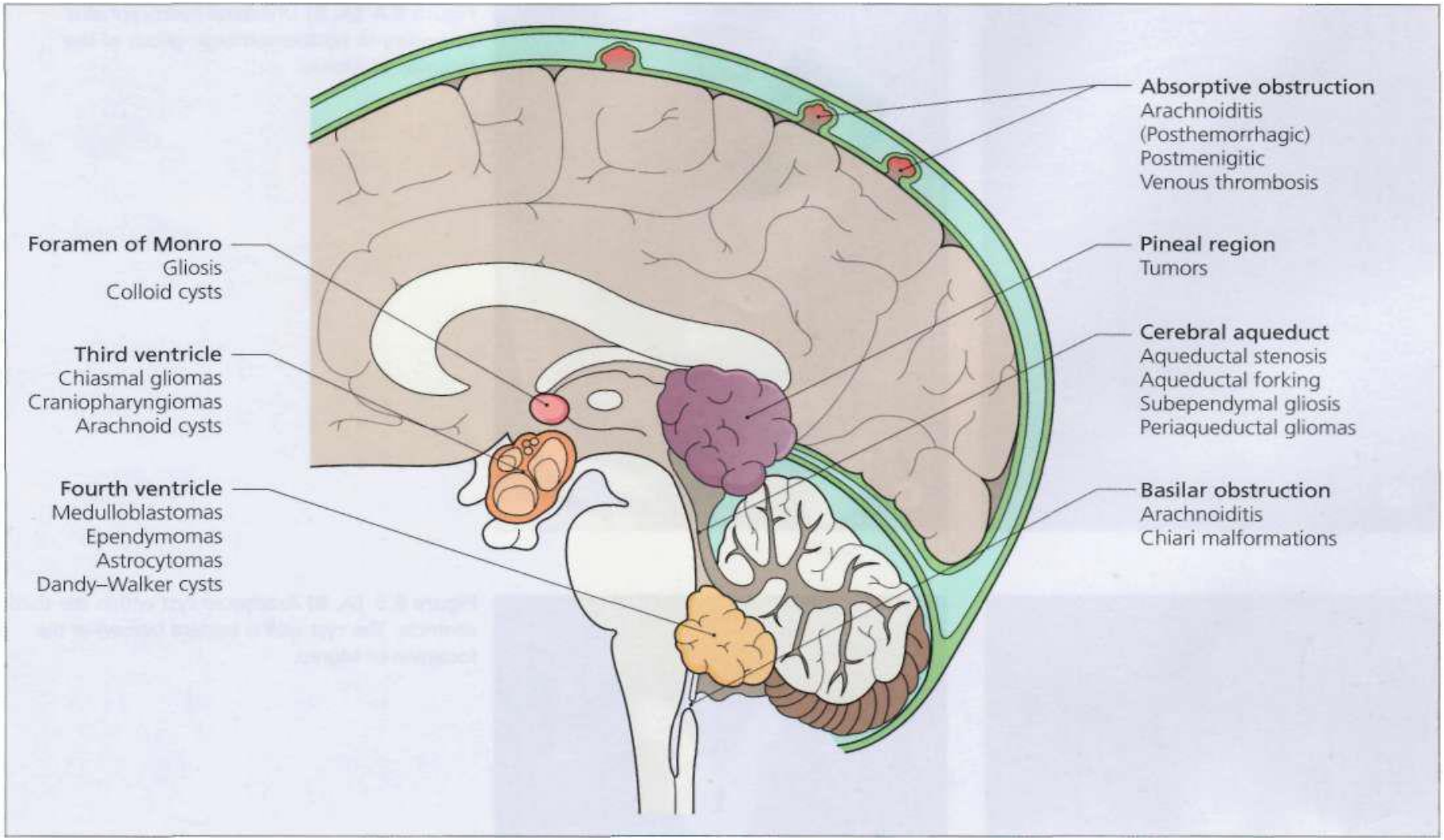
IN GENERAL CSF WILL ACCUMULATE(**HYDROCEPHALUS**) DUE TO:

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

CAUSES OF HYDROCEPHALUS:

- Hydrocephalus is often associated with dilatation of the ventricular system and increased ICP.
- Hydrocephalus is almost always a result of an interruption of CSF flow and is rarely because of increased CSF production.
- Common obstruction sites and etiologies are displayed in the next Figure.

Common sites and causes of CSF obstruction.



PATHOPHYSIOLOGY OF HYDROCEPHALUS

- As ICP rises, CSF absorption increases somewhat, but CSFproduction remains constant.
- If progressive ventricular dilatation separates ependymal cells lining the ventricles, interstitial cerebral edema will develop.
- The CSF will eventually enter the white matter of the brain via bulk flow through the ependymal cells lining the ventricles.

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

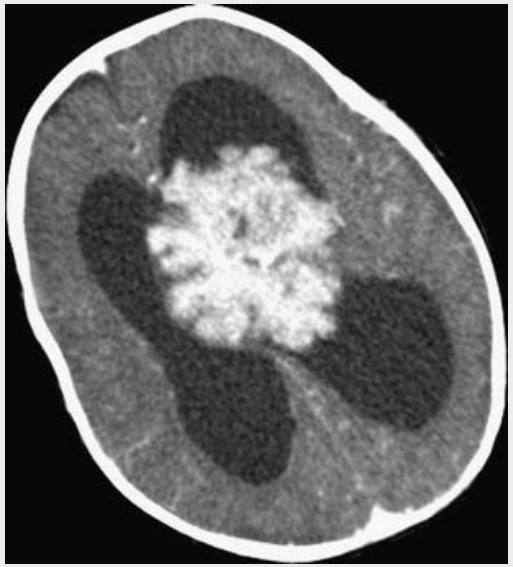
- A commonly used classification differentiates hydrocephalus between ***communicating*** *or* ***noncommunicating.***
- Its symptomatology as either **compensated** or ***noncompensated.***
- Its chronicity: ***acute versus chronic*** hydrocephalus.
- **Congenital** versus **acquired**.
- ***Internal*** *or* ***external***

AETIOLOGY - SITE

1. **Lateral ventricles**

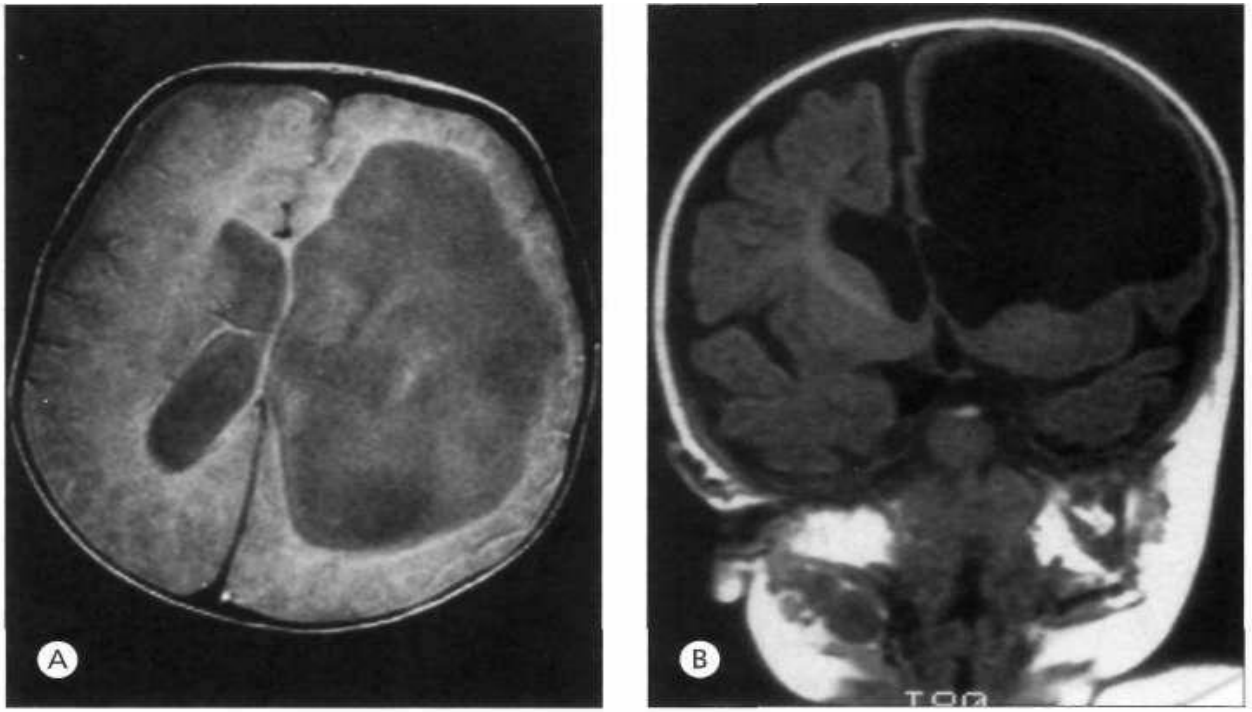
Choroid plexus tumors:

- 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 .

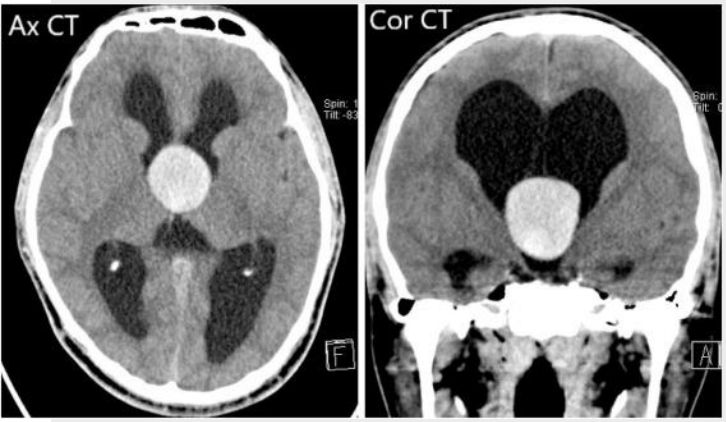


2-Foramina of Monro

- Occlusion of one foramen of Monro can occur secondary to a congenital membrane, atresia, or gliosis after intraventricular hemorrhage (IVH) or ventriculitis.



3-Third ventricle



Cysts and neoplasms within the third ventricle commonly cause hydrocephalus.

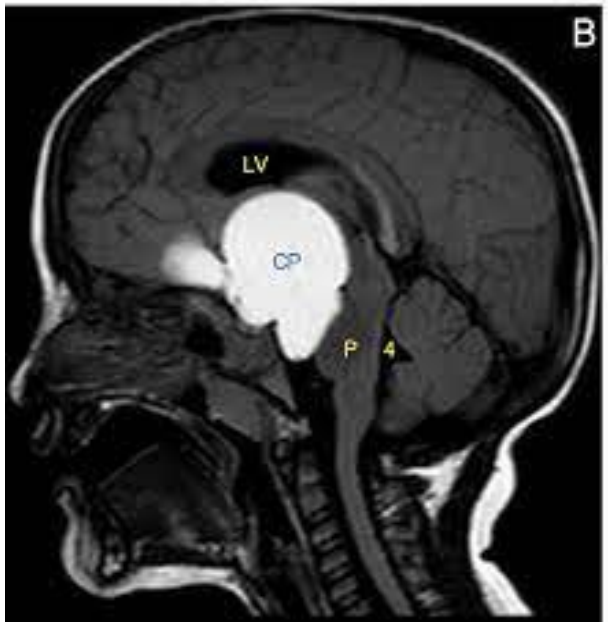
-Colloid cysts are uncommon neoplasms that present superiorly and anteriorly within the third ventricle,

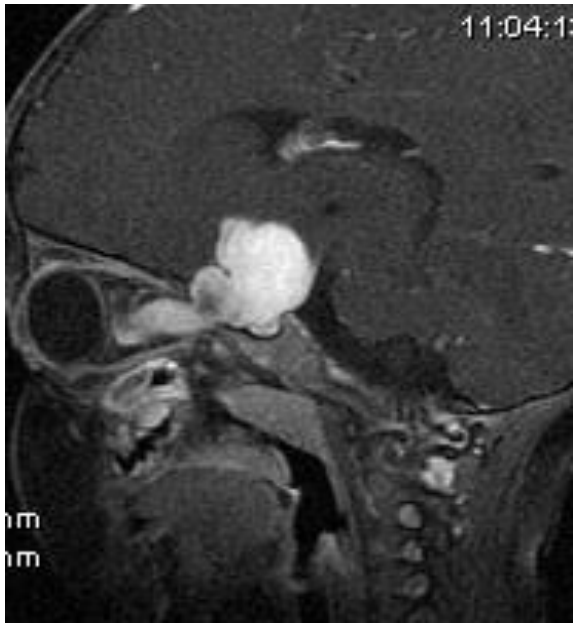
-**Ependymal and arachnoid cysts** within the third ventricle usually present with hydrocephalus in late childhood .Patients may present with bobble-head doll syndrome, a rhythmic head nodding at a frequency of two to three times per second.

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**.



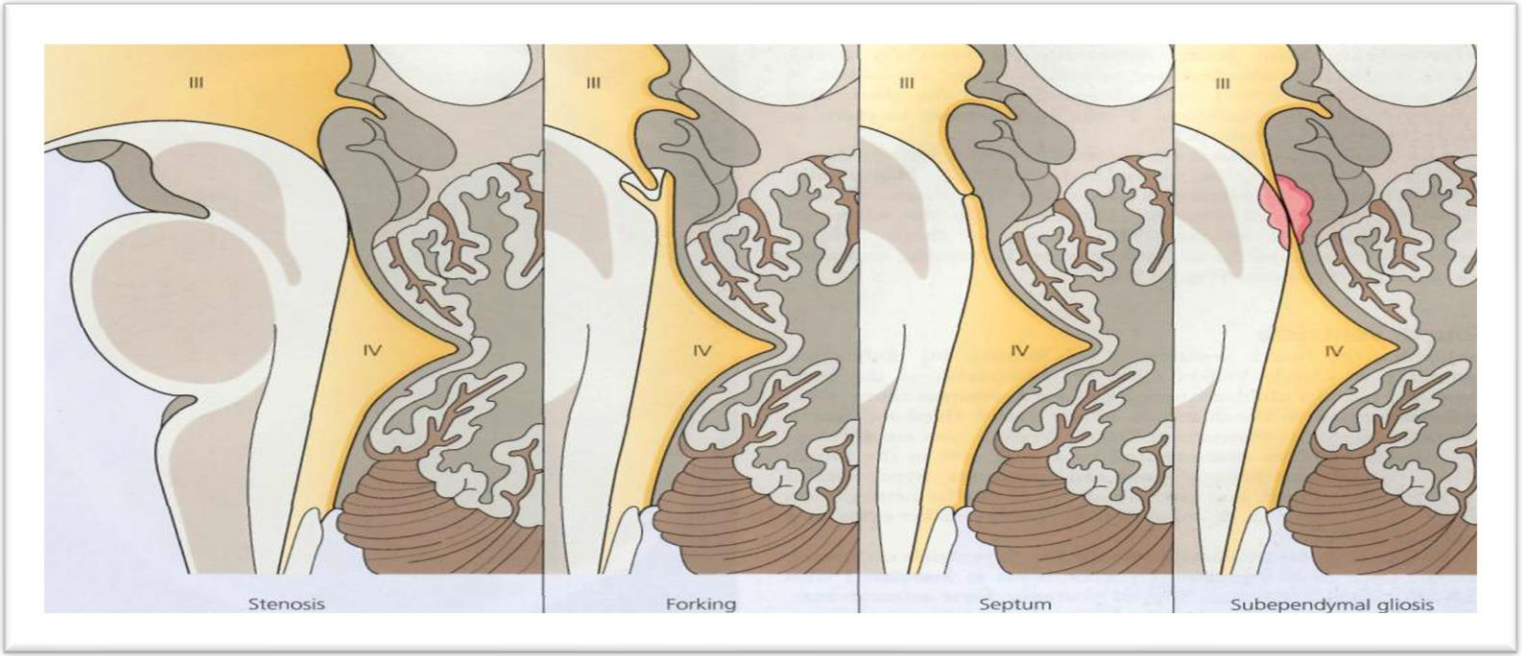


4-Sylvian aqueduct

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)

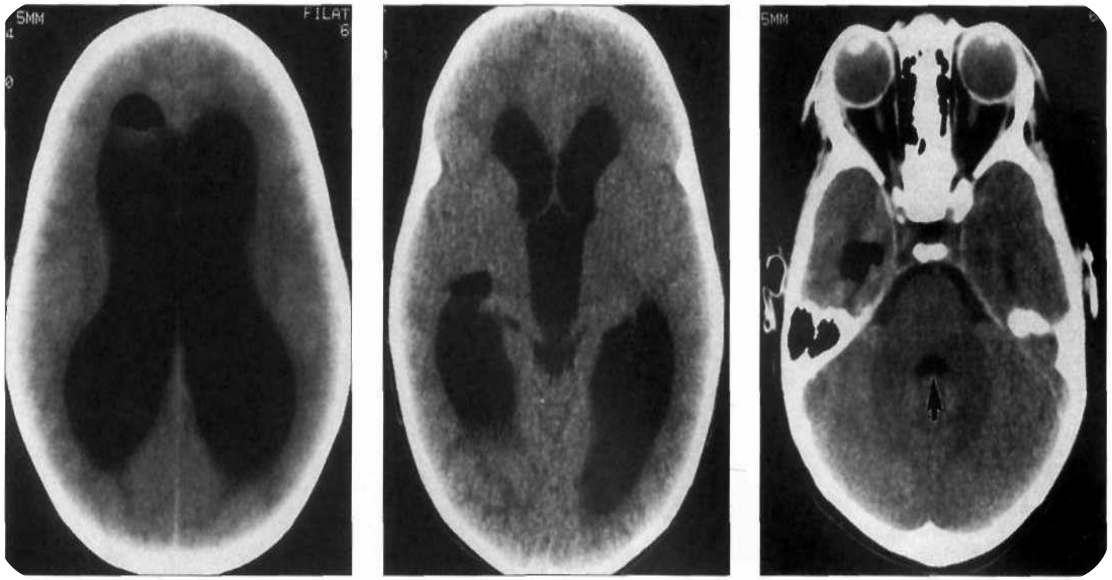
It may be secondary to *in utero infections (e.g. toxoplasmosis), intraventricular* hemorrhage, or mumps encephalitis.



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

Sylvian aqueductai 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.

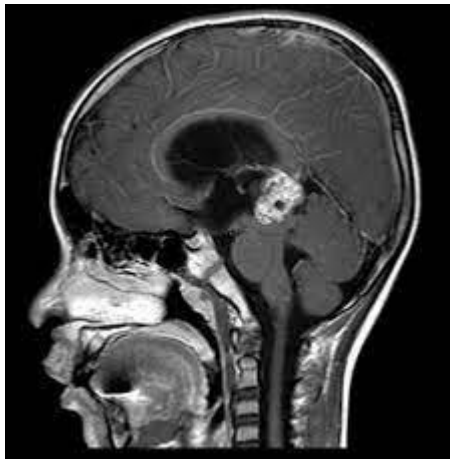
Sylvain aqueduct

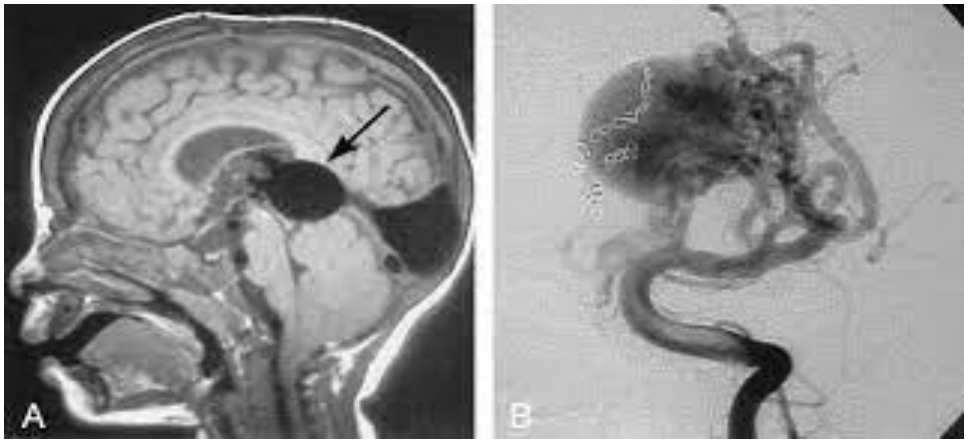
Pineal region neoplasms,

Arteriovenous malformations, and Periaqueductal neoplasms.

Any pineal mass can obstruct the aqueduct and produce hydrocephalus. Many pineal region tumors, especially germinomas, are highly radiosensitive; successful tumor irradiation, as well as surgical resection, may adequately treat the obstructive hydrocephalus.

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 or obliteration of the basal foramina.

In older children, neoplasms are a common cause.

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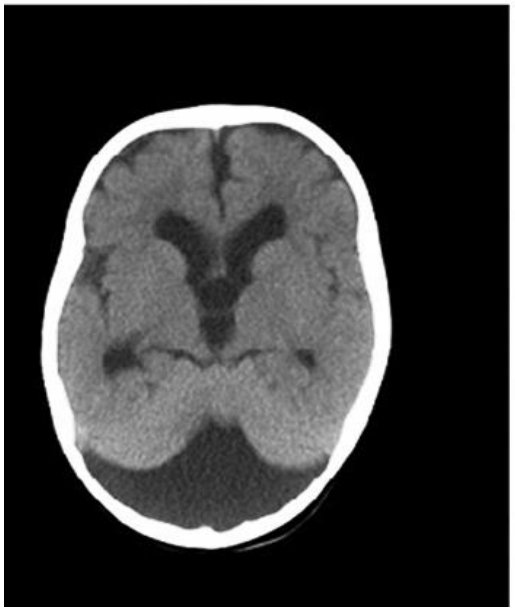
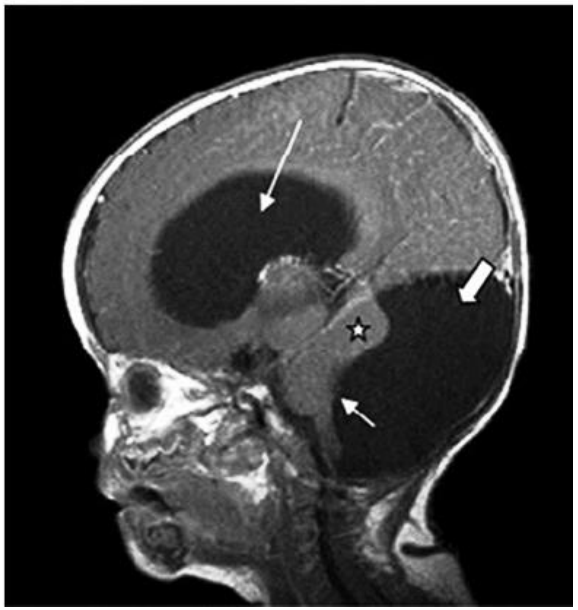
Dandy-Walker cysts are developmental abnormalities characterized by a large cyst in the fourth ventricle, hypoplasia of the cerebellar vermis, and atrophy of the cerebellar hemispheres.

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





4TH Ventricle tumors

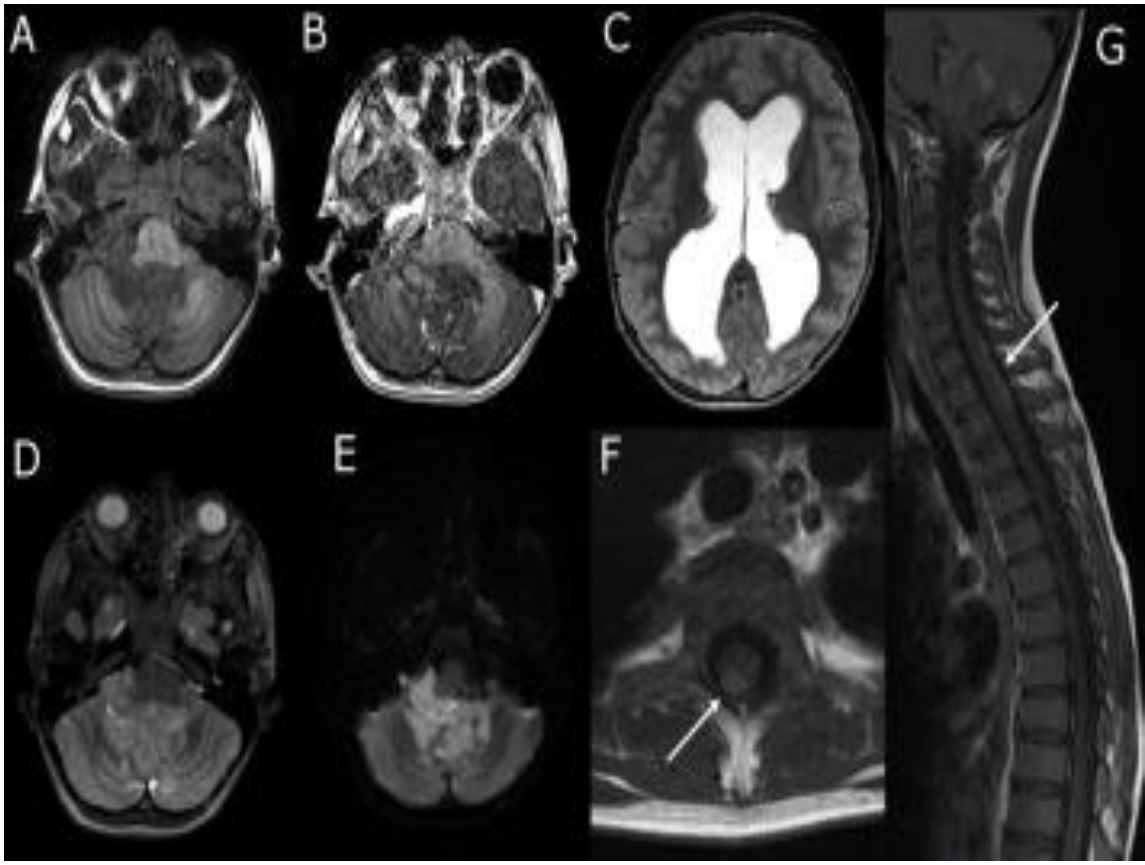
Hydrocephalus is associated with

85% of **medulloblastomas,**

65% of posterior fossa **astrocytomas,**

75% of **ependymomas**,

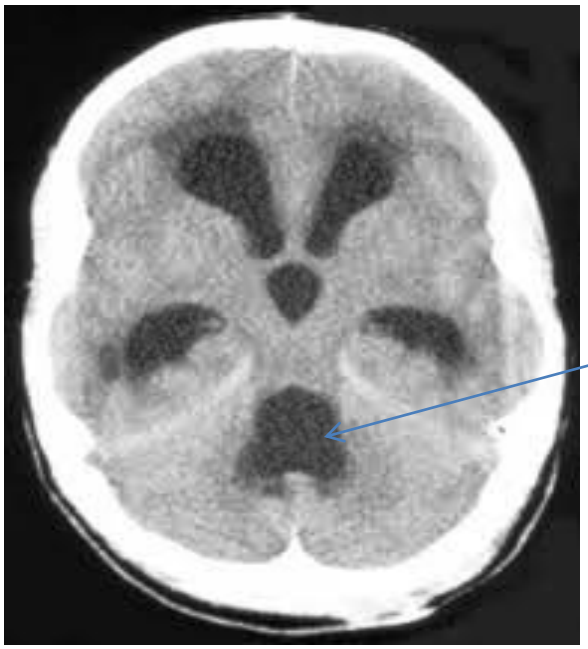
and 25% of **brainstem gliomas**.



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)**

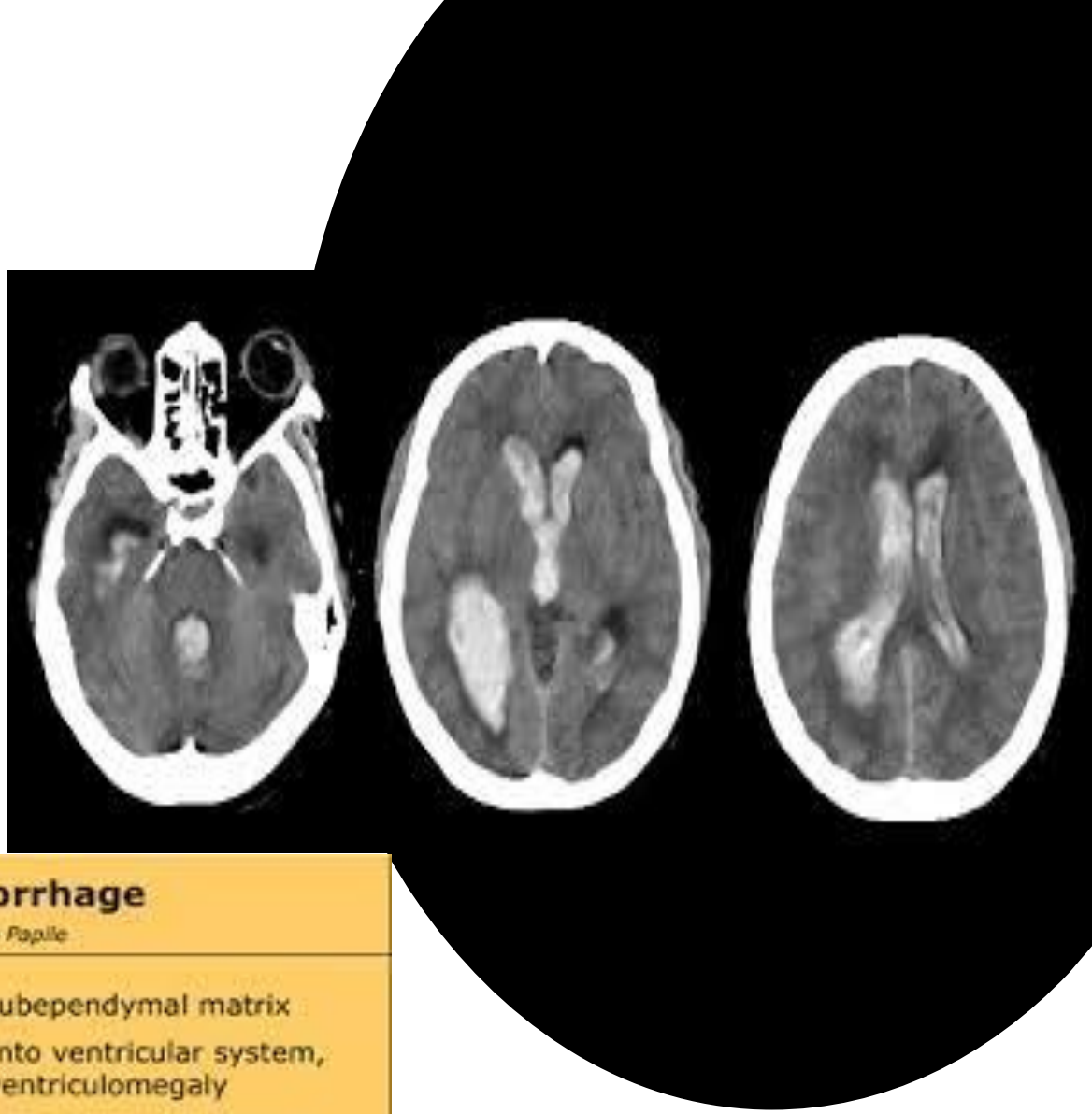


Dilated fourth ventricle

Etiology by age

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

Etiology by age

- **Full-term infants**

The common causes of hydrocephalus in full-term infants include

- **Aqueductal stenosis**,
- Chiari II malformation,
- Dandy-Walker syndrome,
- Cerebral malformations (e.g. encephaloceles, holoprosencephaly, and hydranencephaly), arachnoid cysts,
- Neoplasms,

and

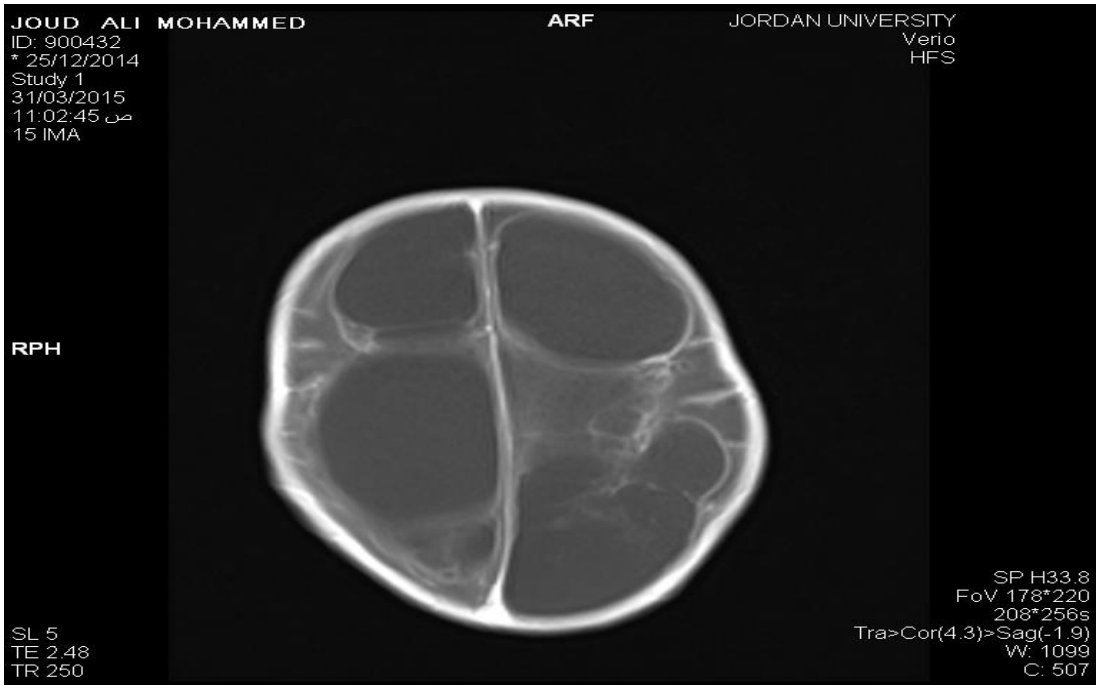
- Vein of Galen malformations.

FULL TERM INFANT

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

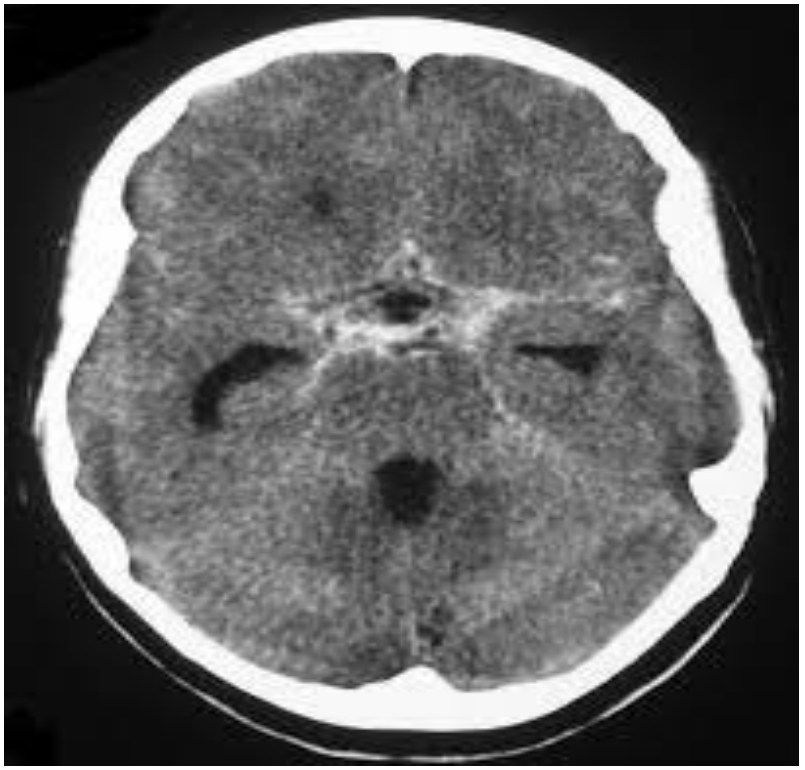
Older children

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



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 fonatanelle

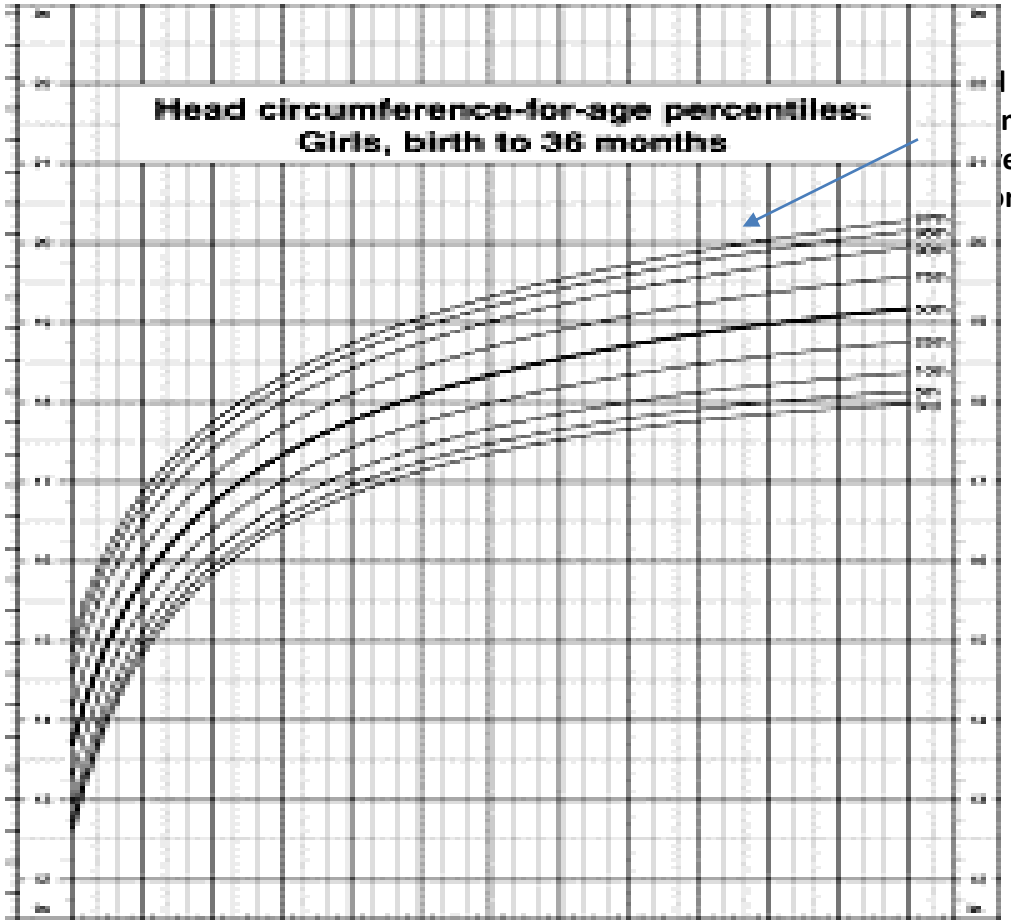
Dilated scalp veins

Suture widening

Setting sun eyes

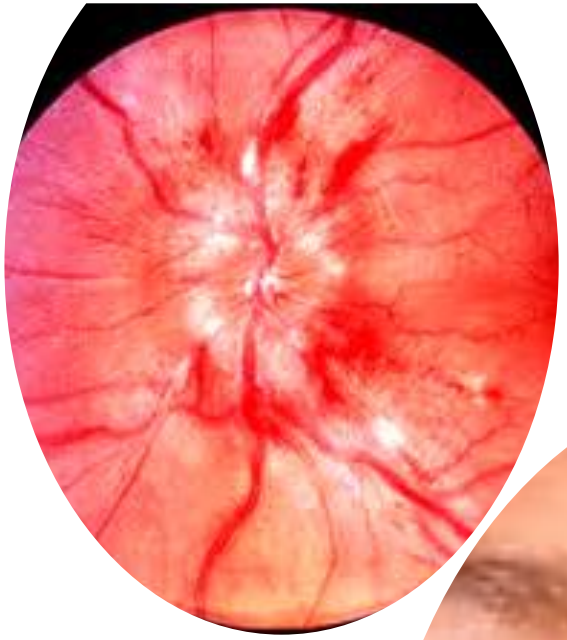
Head circumference

- 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.

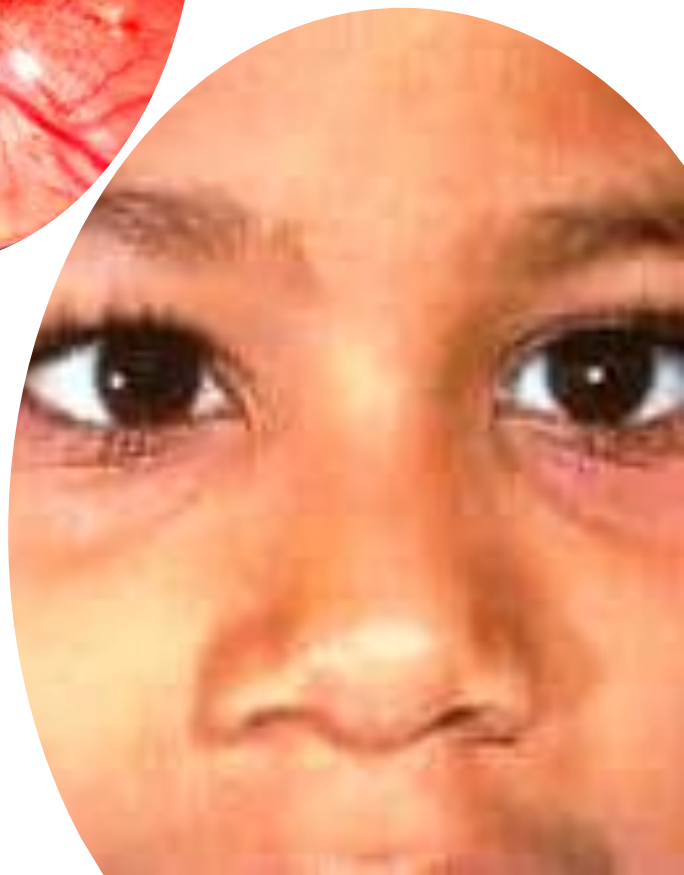


Signs of hydrocephalus in children and adults

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

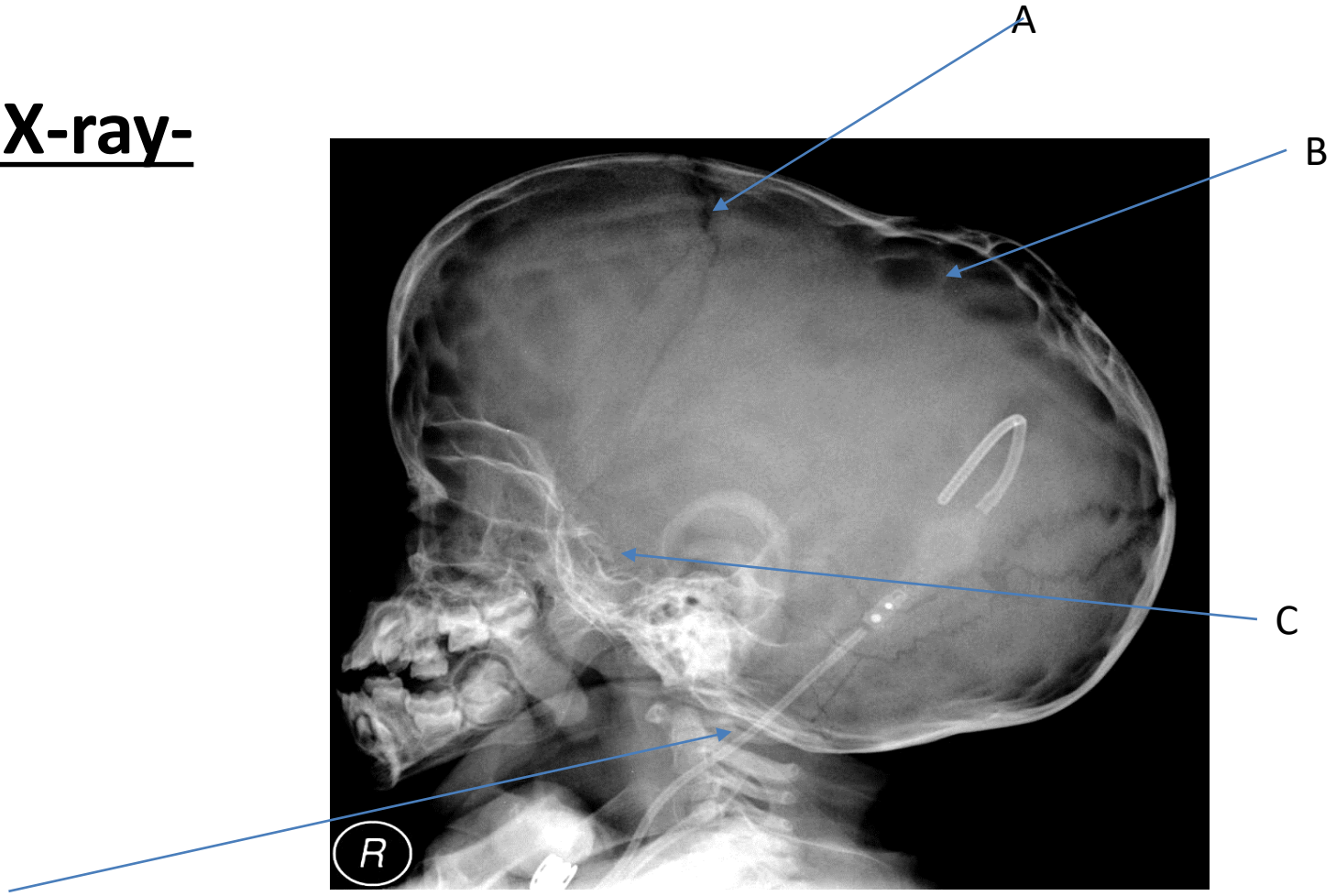


Right abducent nerve palsy



DIAGNOSTIC STUDIES

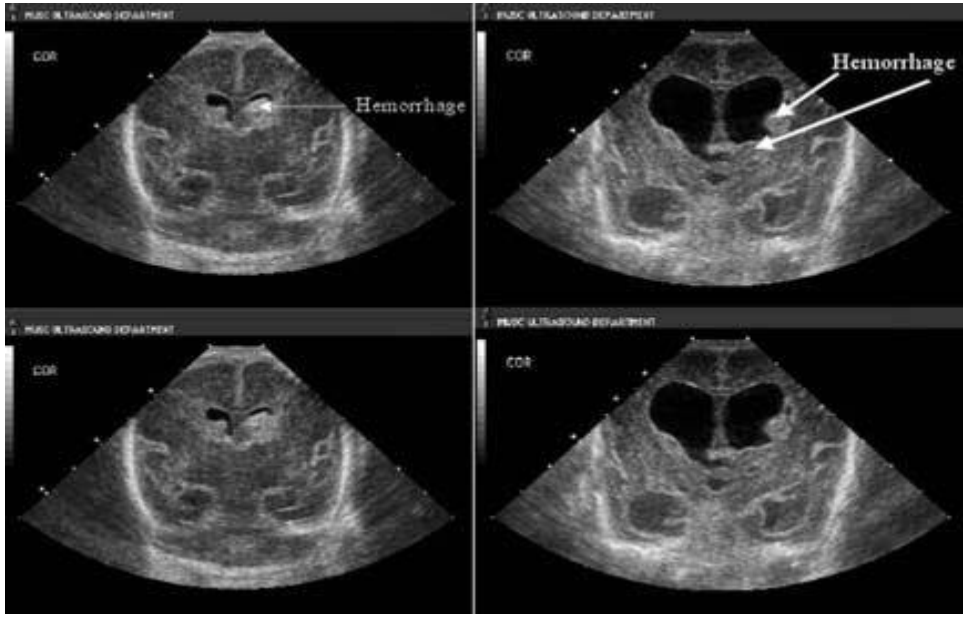
- **Skull X-ray-**



Cranial ultrasonography

- is particularly useful in the evaluation of premature infants with IVH, as well as the detection and monitoring

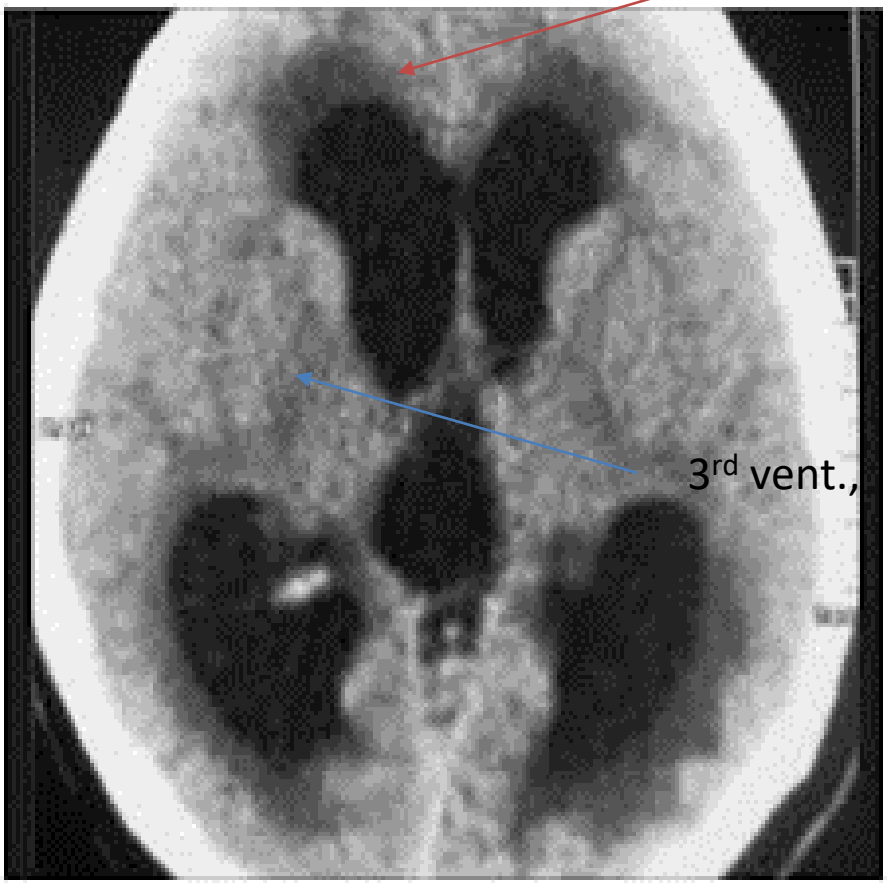
  ofventricular size.



CT scanning

- From 1976 to 1986, CT scanning was the definitive method of diagnosing pediatric hydrocephalus.
- limitations: only in the **axial plane**,

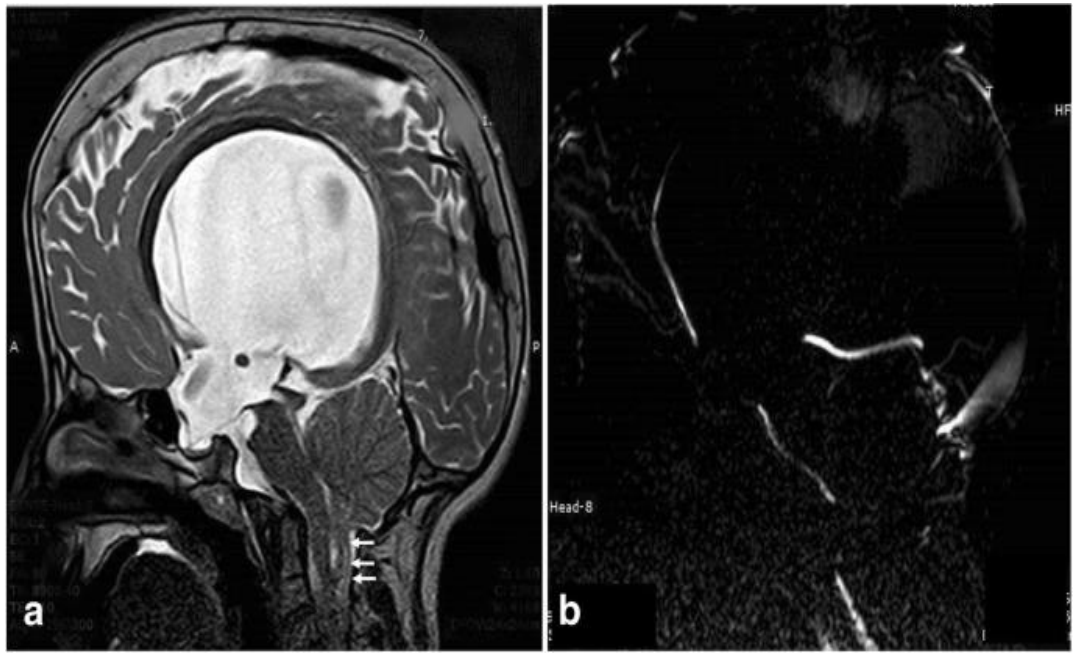
Require **irradiation,** and have **less resolution** than MRI.


Periventricular fluids
3rd vent.,

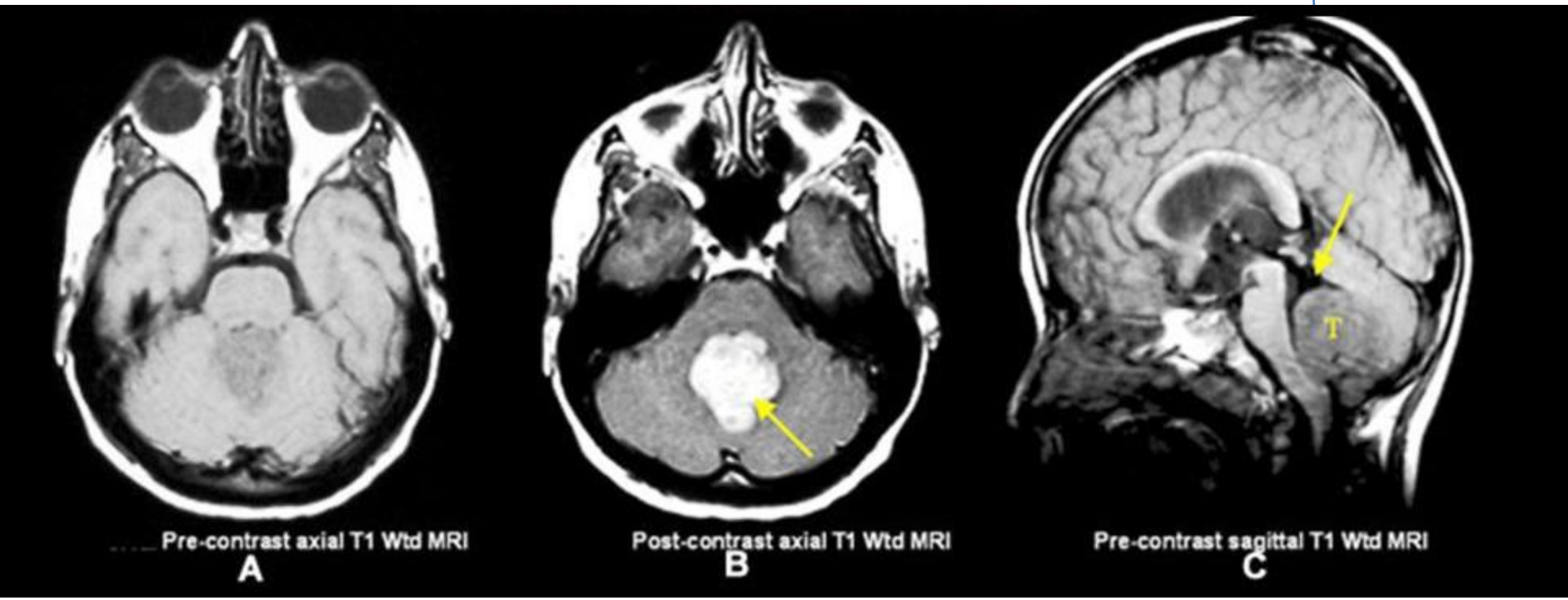
Brain MRI

Since 1986,

- can project the brain in axial,coronal, and sagittal projections.
- MRI can detect transependymal resorption and low-grade gliomas more clearly.
- as well as determine CSF flow across the aqueduct.



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



treatment

- The goal is:

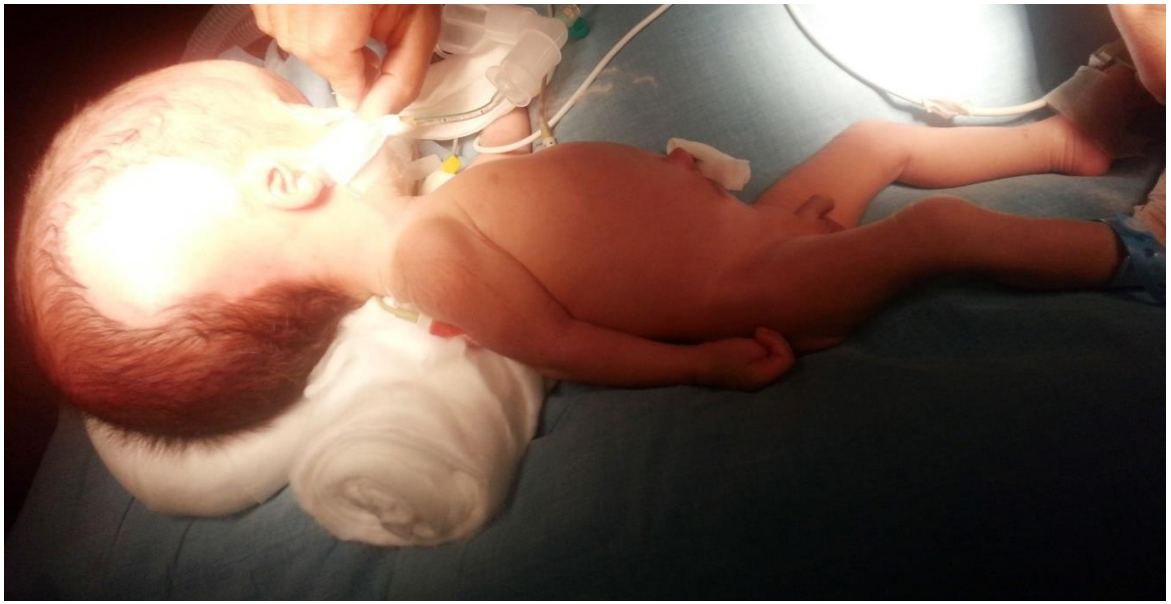
1. Optimal neurological outcome
2. Preservation of cosmosis

- Normal sized ventricles should not be considered the goal

1. Non surgical
2. Surgical

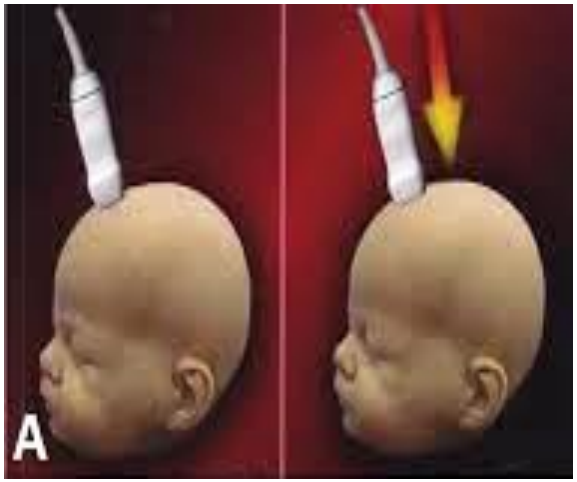
➢ Shunting

➢ Non shunting

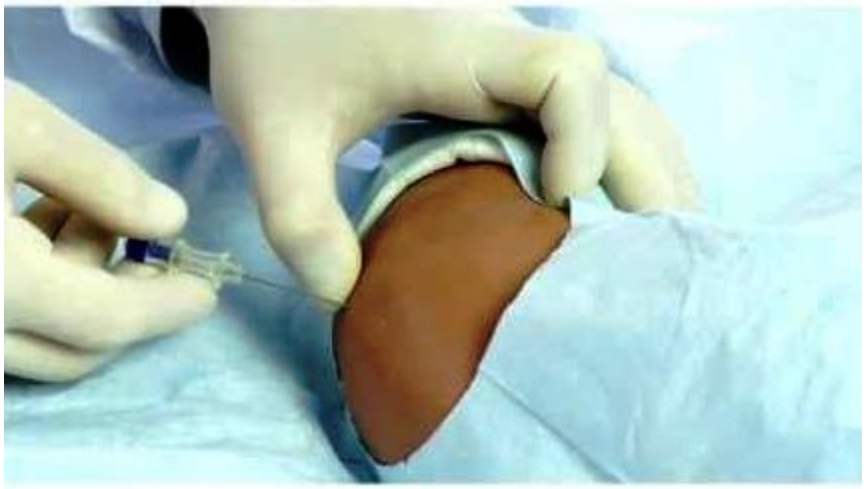


Non surgical

- Not defintive 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

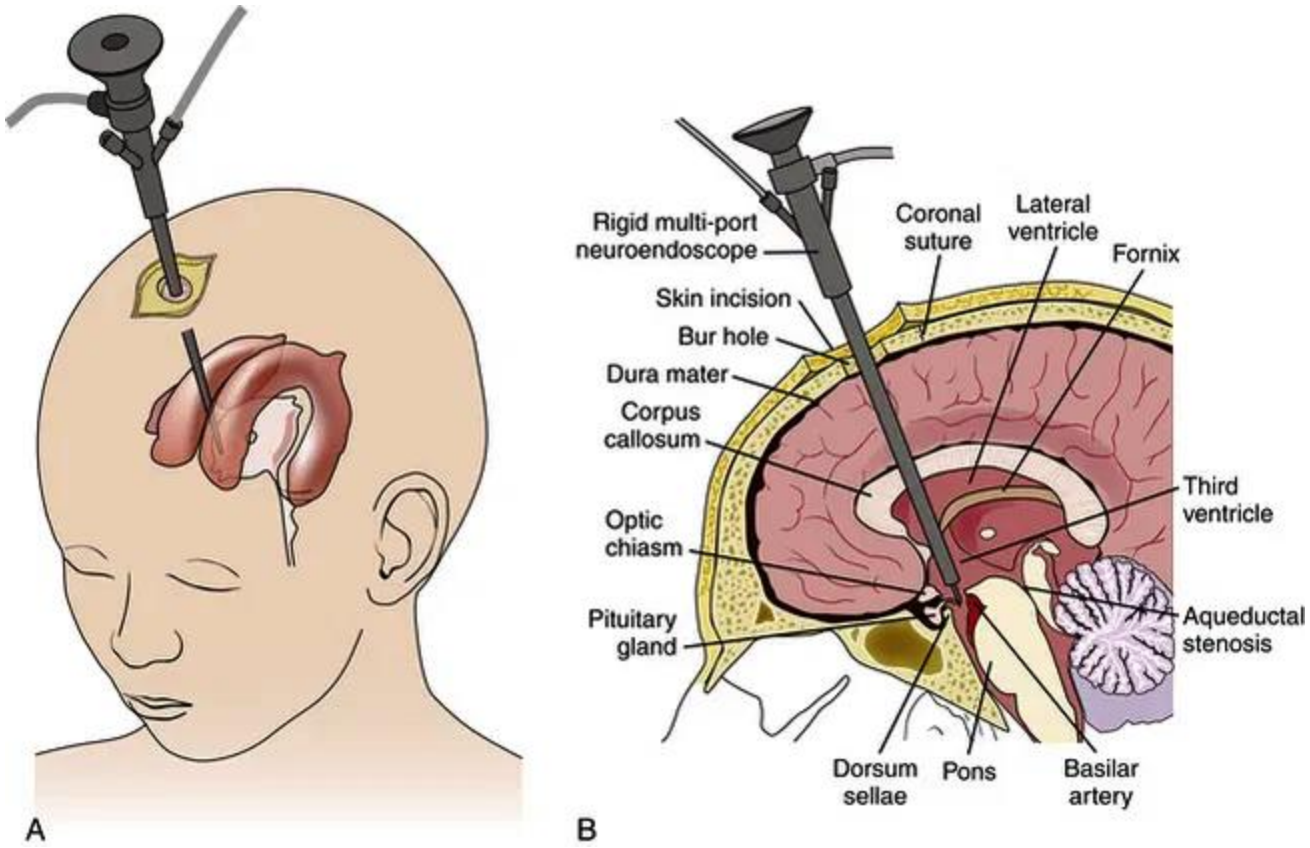
1-Whenever possible, the obstructing lesion that causes the

hydrocephalus should be ***surgically removed.***

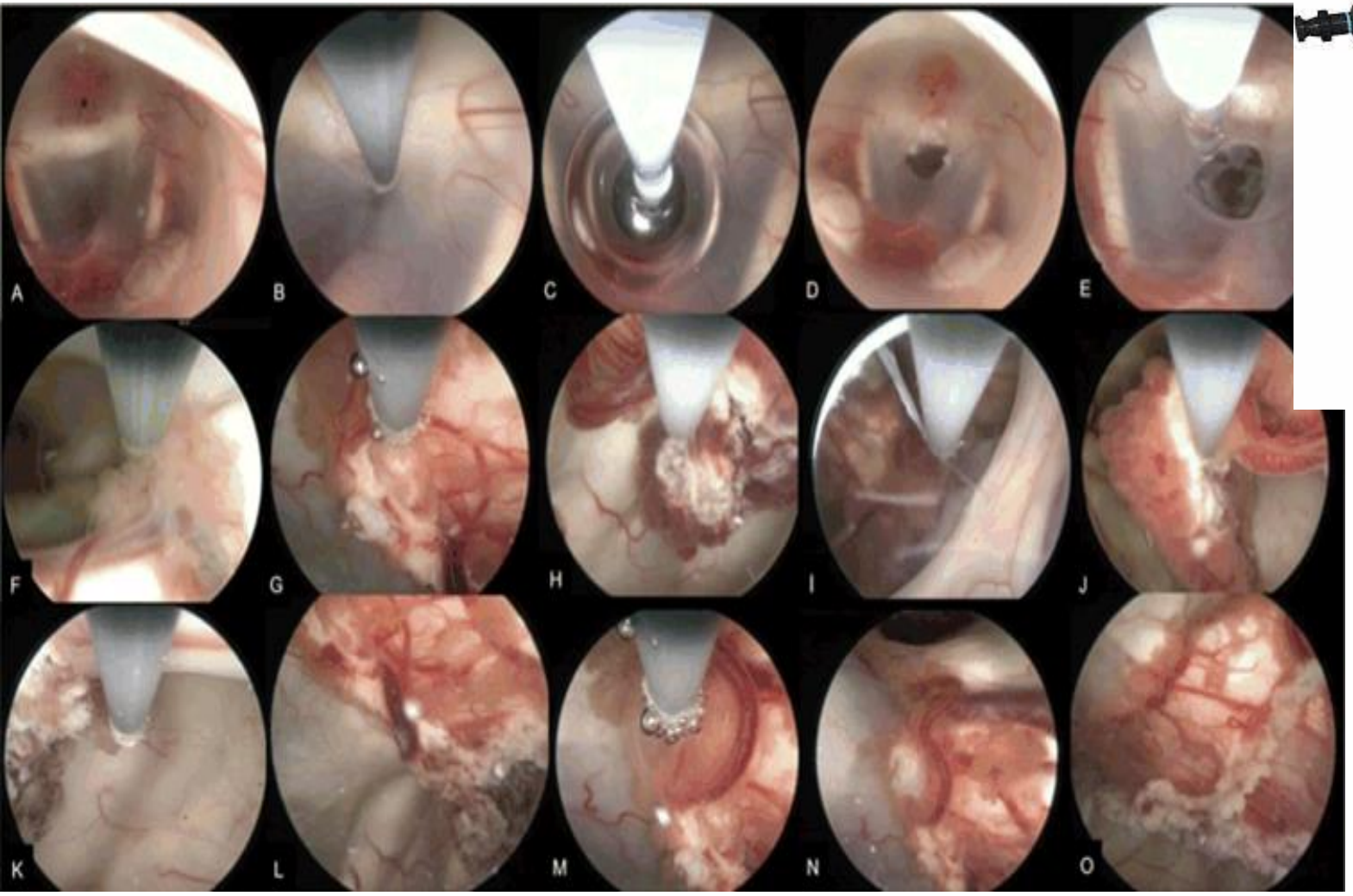
2-For CSF obstruction at or distal to the aqueduct, a potential surgical treatment is the ***endoscopic third ventriculostomy(ETV).***

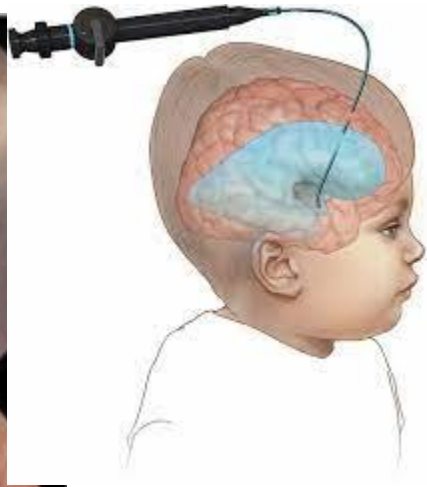
By surgically creating an opening at the floor of the third ventricle, CSF can be diverted without placing a shunt. Kamikawa and associates reported a 75% success rate for ETVamong 44 pediatric patients with hydrocephalus secondary to aqueductal stenosis; Other

Endoscopic Third Ventriculostomy(ETV)



ETV-CPC-choroid plexus coagulation

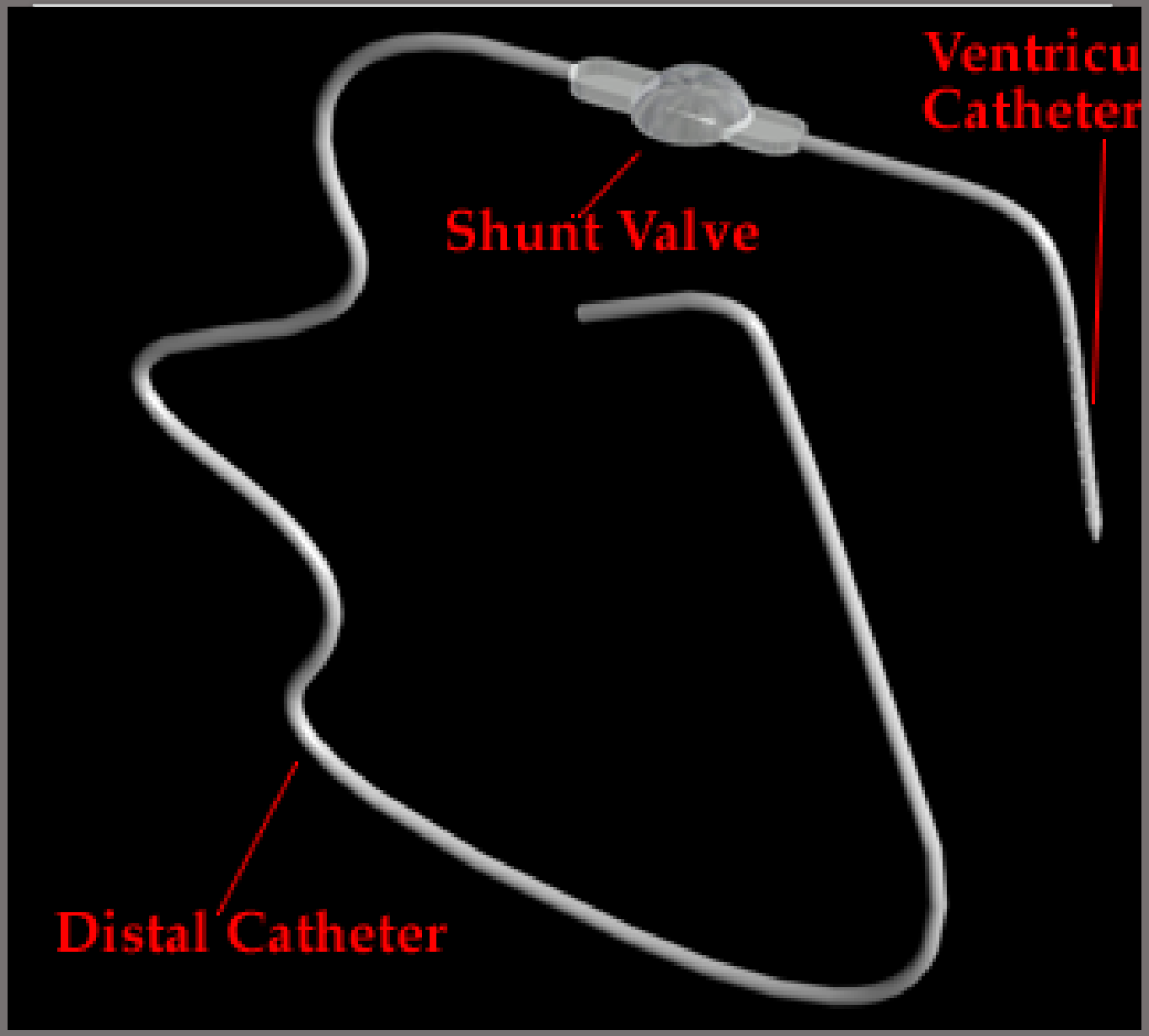




Surgical - CSF shunts

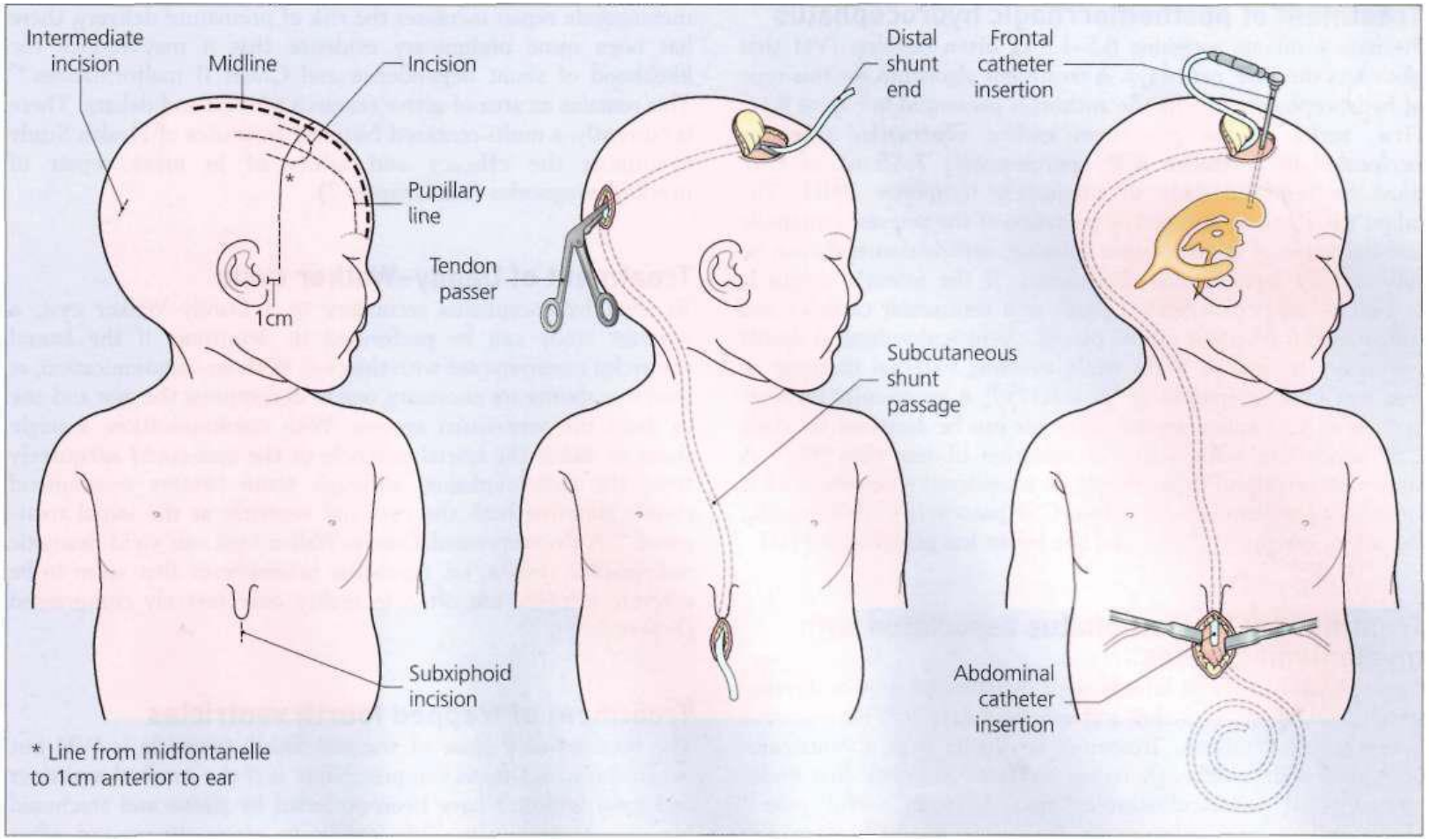
- CSF shunts are usually silastic tubes that divert CSF from the ventricles to other body cavities (i.e. peritoneal, atrium or pleural

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

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

	Uncommon complications			
Common 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	

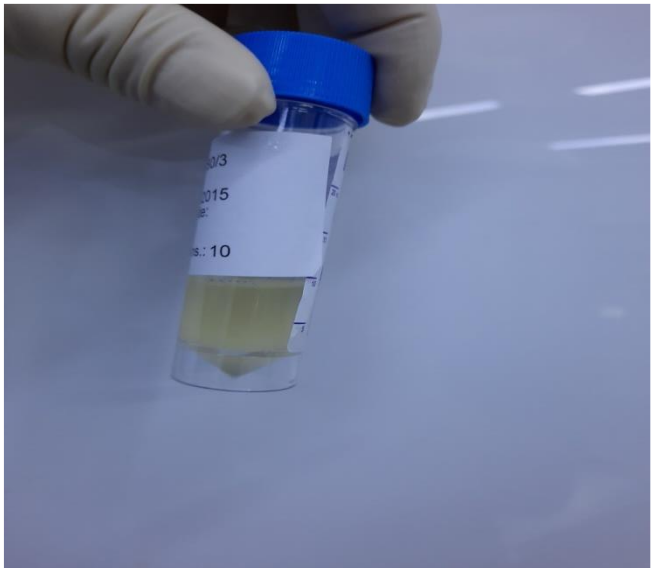
Shunt Infection

- In general, approximately 1-15% of all shunting procedures are complicated by infection.

Premature infants have an increased risk.

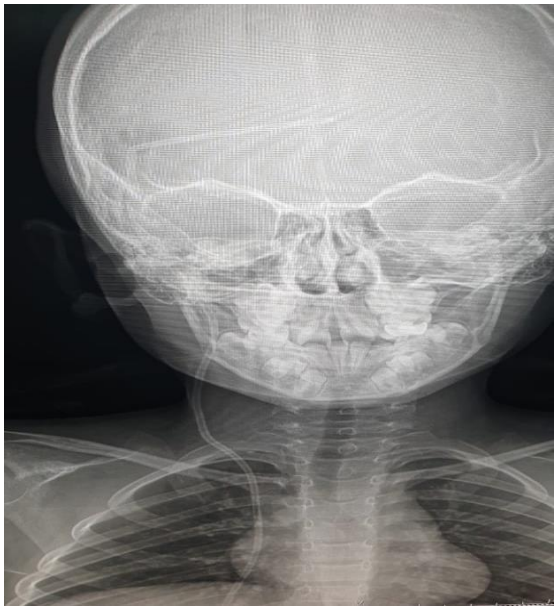
- Evident within 1 month of placement. Nearly 90% of all shunt infections are recognized within 12 months
- it is believed that most bacteria are introduced at the time of surgery.
- ***Staphylococcus epidermidis*** *causes approximately 60% of* shunt infections, ***Staphylococcus aureus*** *is responsible for 30%, and* coliform bacteria, propionibacteria, streptococci, or *Haemophilus influenzae cause the remainder.*

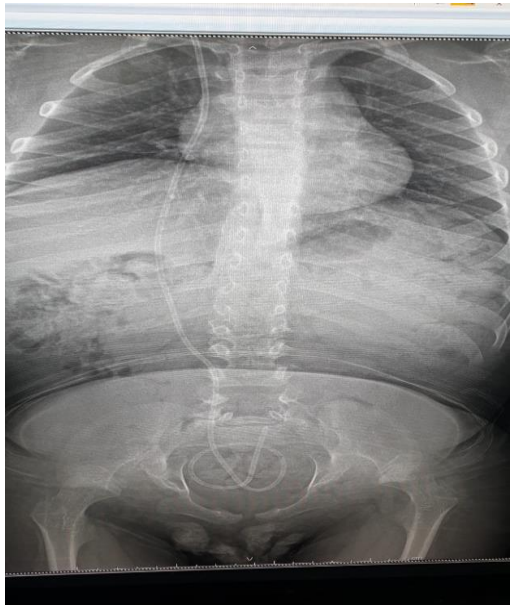
- **Diagnosis** is confirmed by CSF sampling from the shunt reservoir ;with the findings of leukocytosis and positive culture .
- **Treatment** usually involves
  - the removal of the infected shunt
  - and placement of an EVD. The patient is then treated
  - with the appropriate intravenous antibiotics, 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.



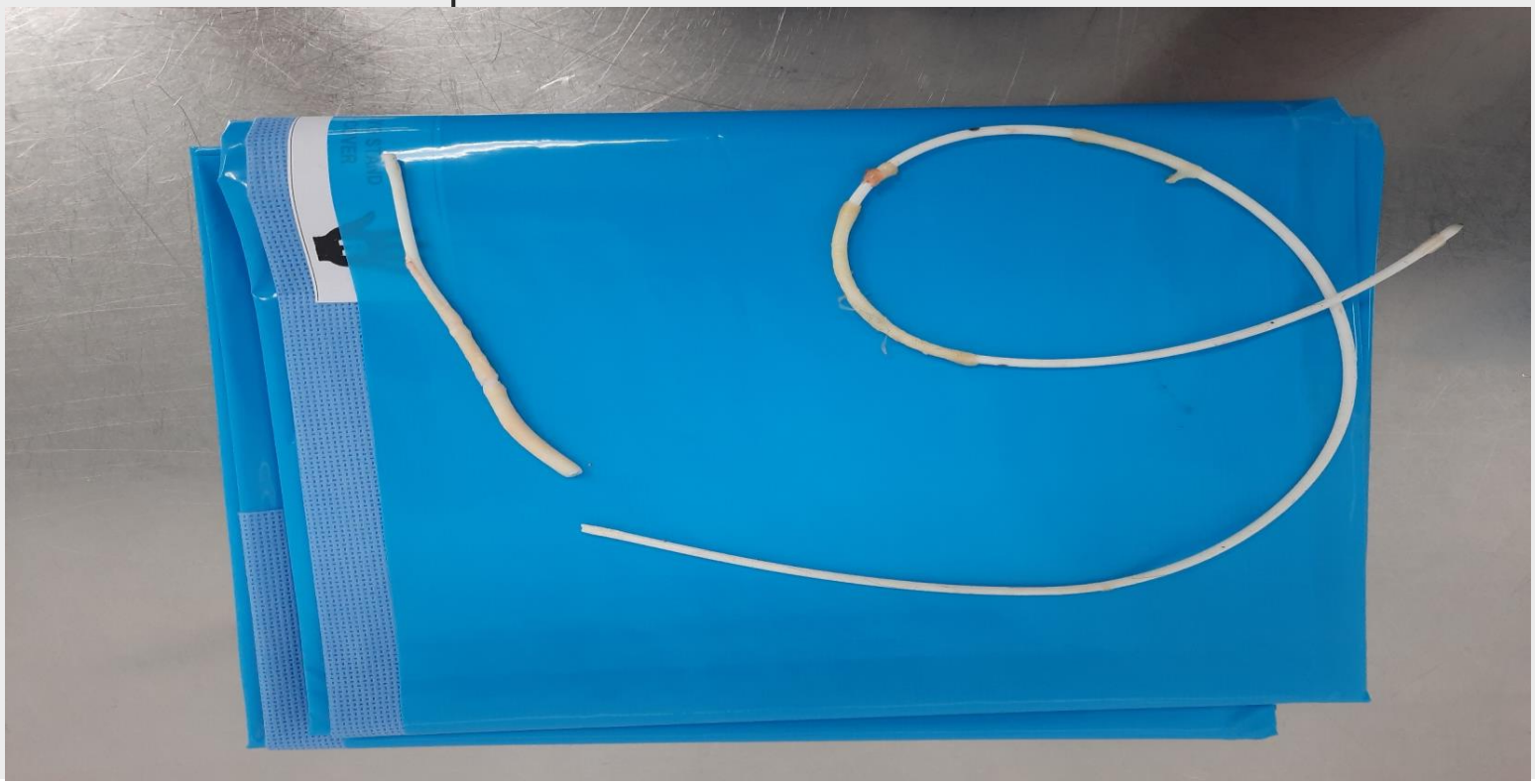
shunt obstruction

- A child with a shunt malfunction often presents with signs and symptoms of increased ICP.
- Children with a shunt malfunction usually present with 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

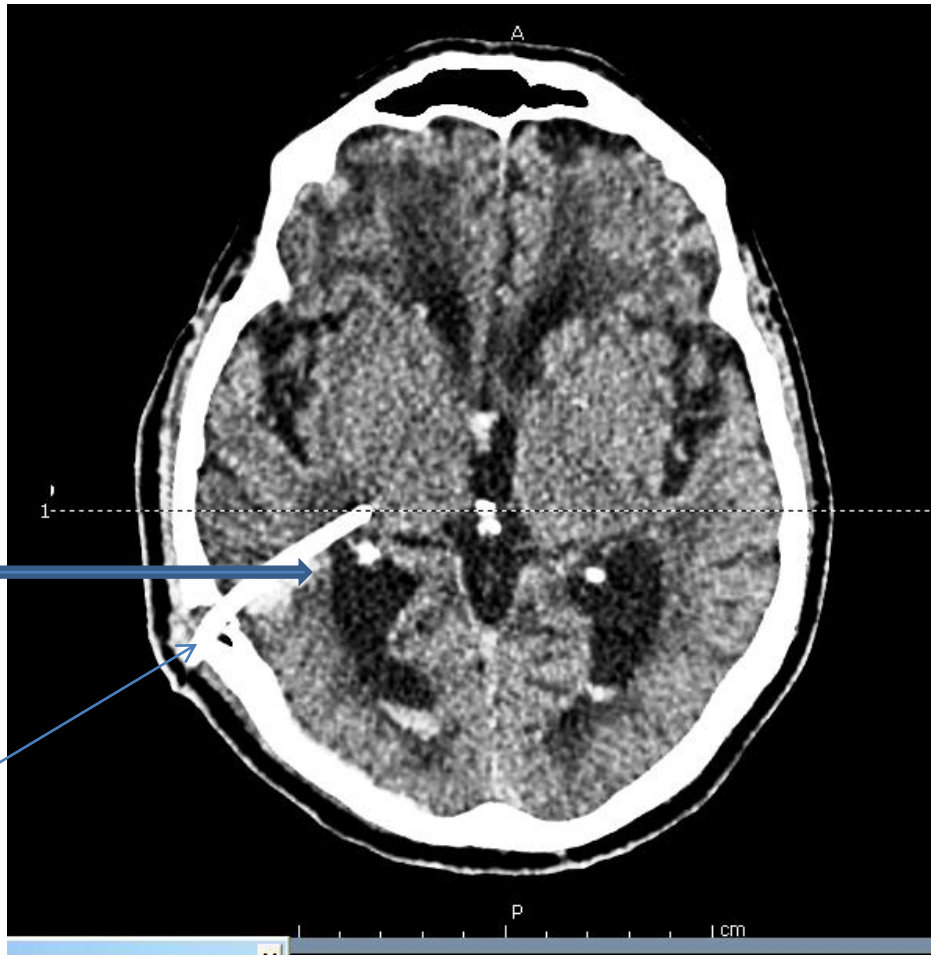




- Shunt obstructions/malfunctions are treated by replacing the occluded or nonfunctioning components, or by replacing the entire system.



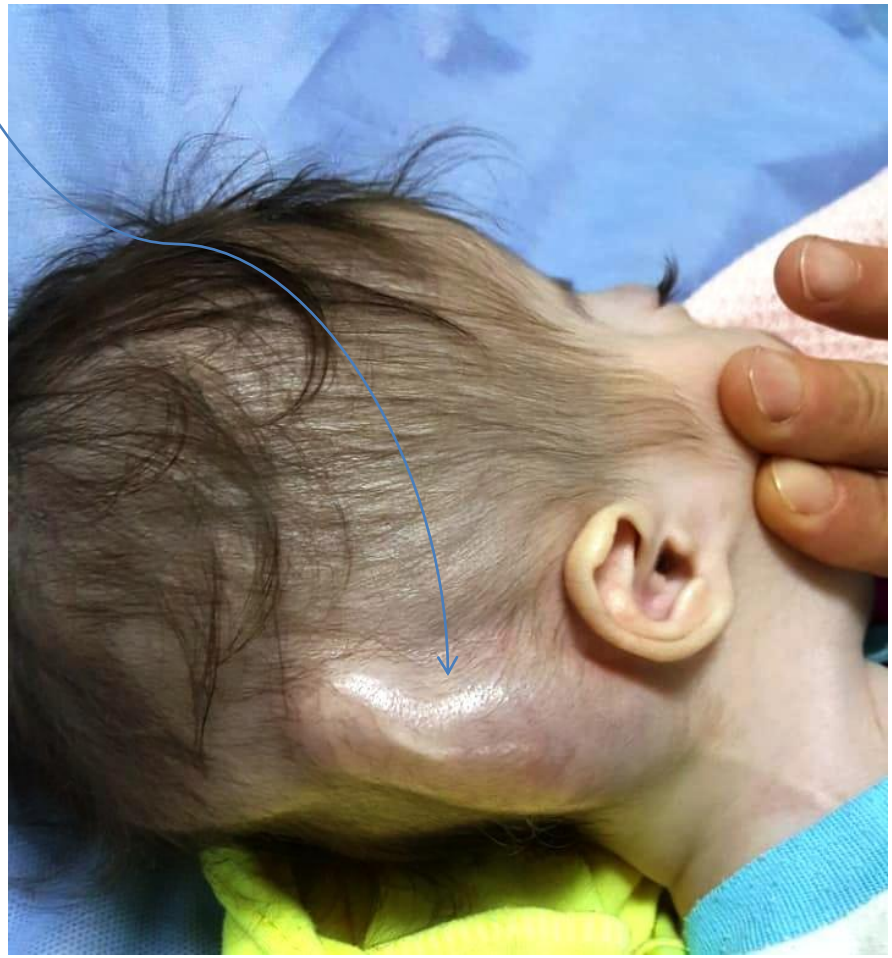
Hematoma at sit of shunt insertion



Intracerebral Hematoma around the catheter

Ventricular catheter

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.