Hydrocephalus

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Outlines

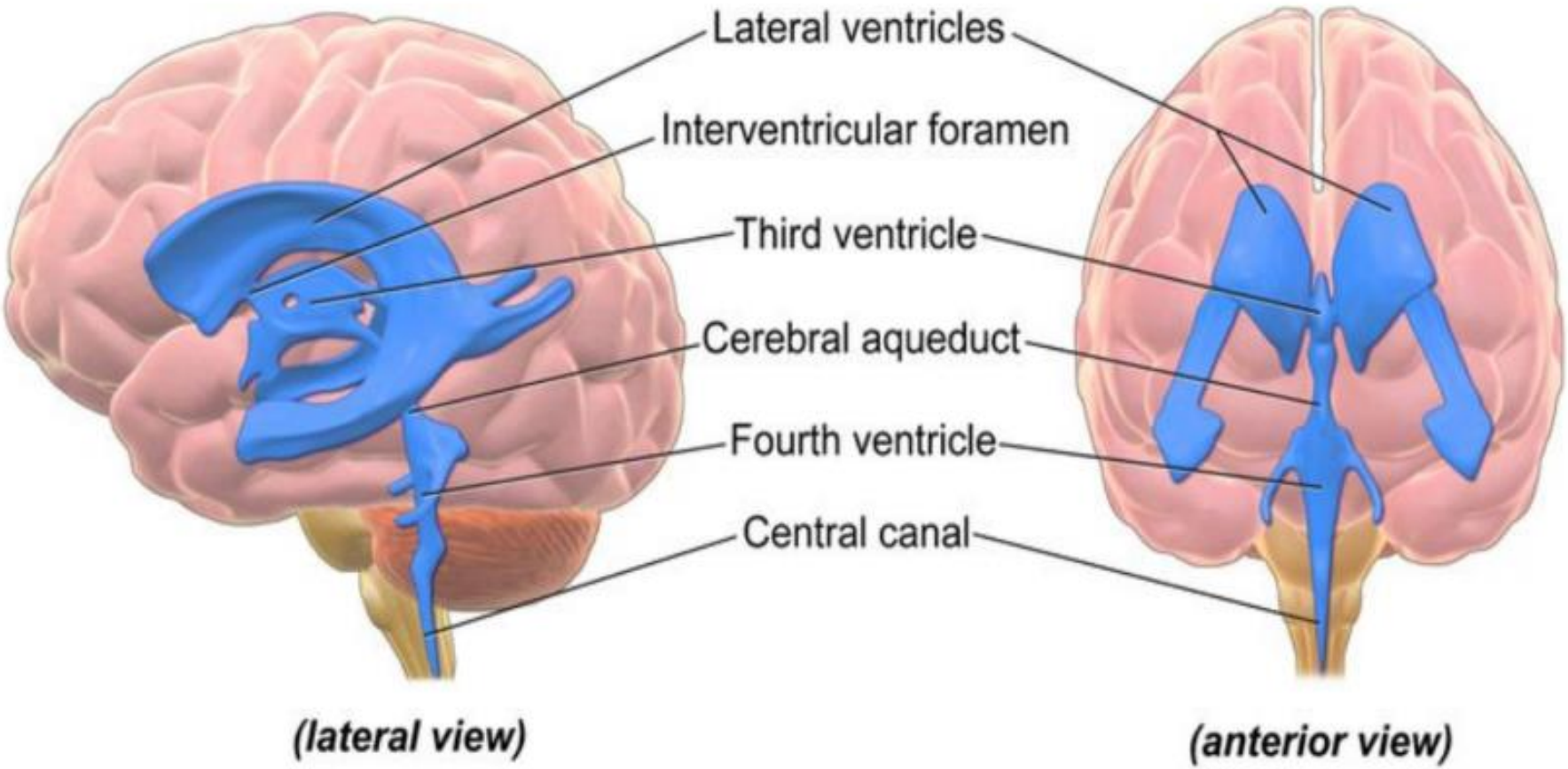
- Definition
- Pathophysiology
- Etiology
- Types
- Clinical presentation
- Diagnosis
- Treatment

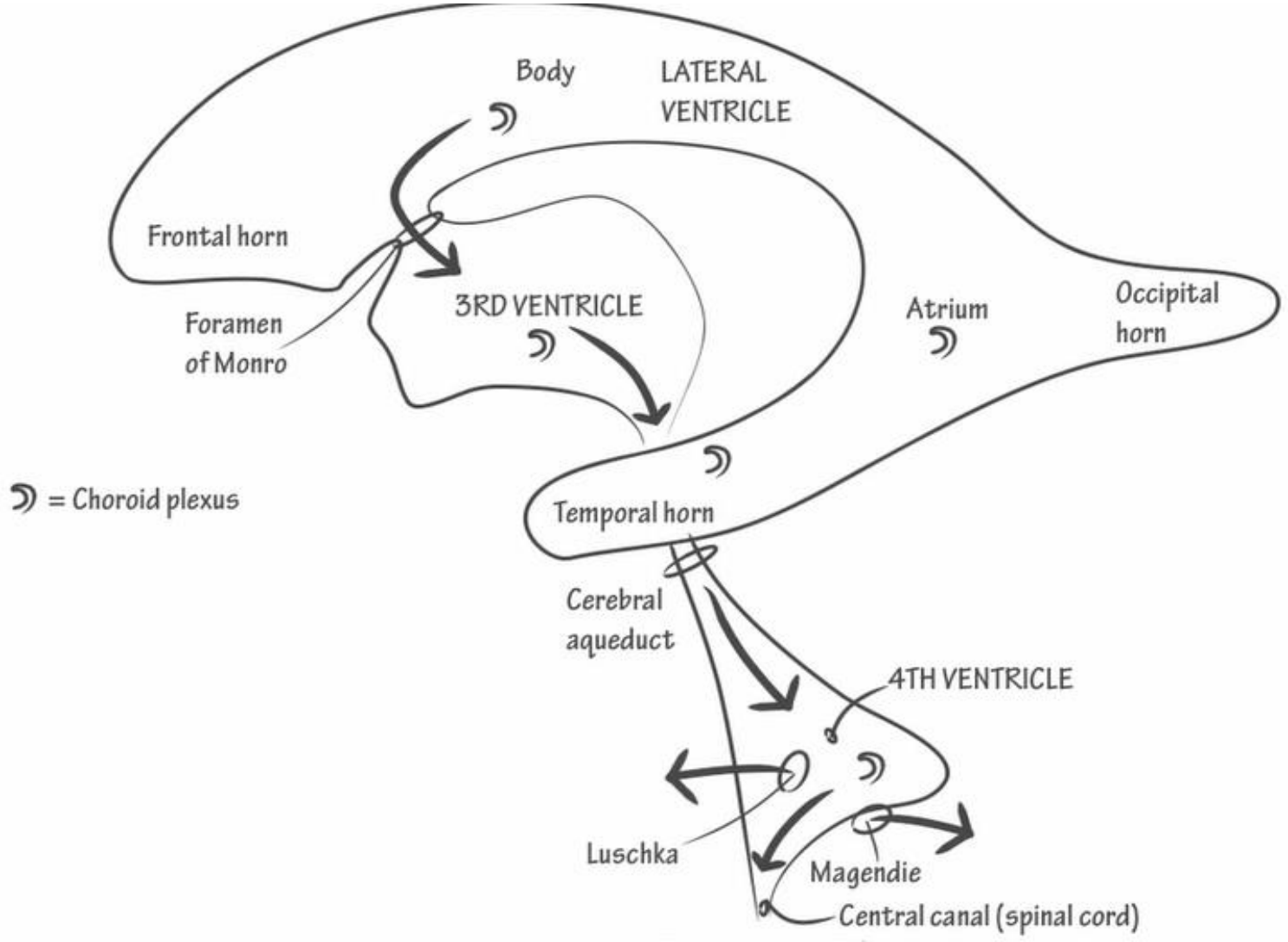
What is hydrocephalus?

- 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.

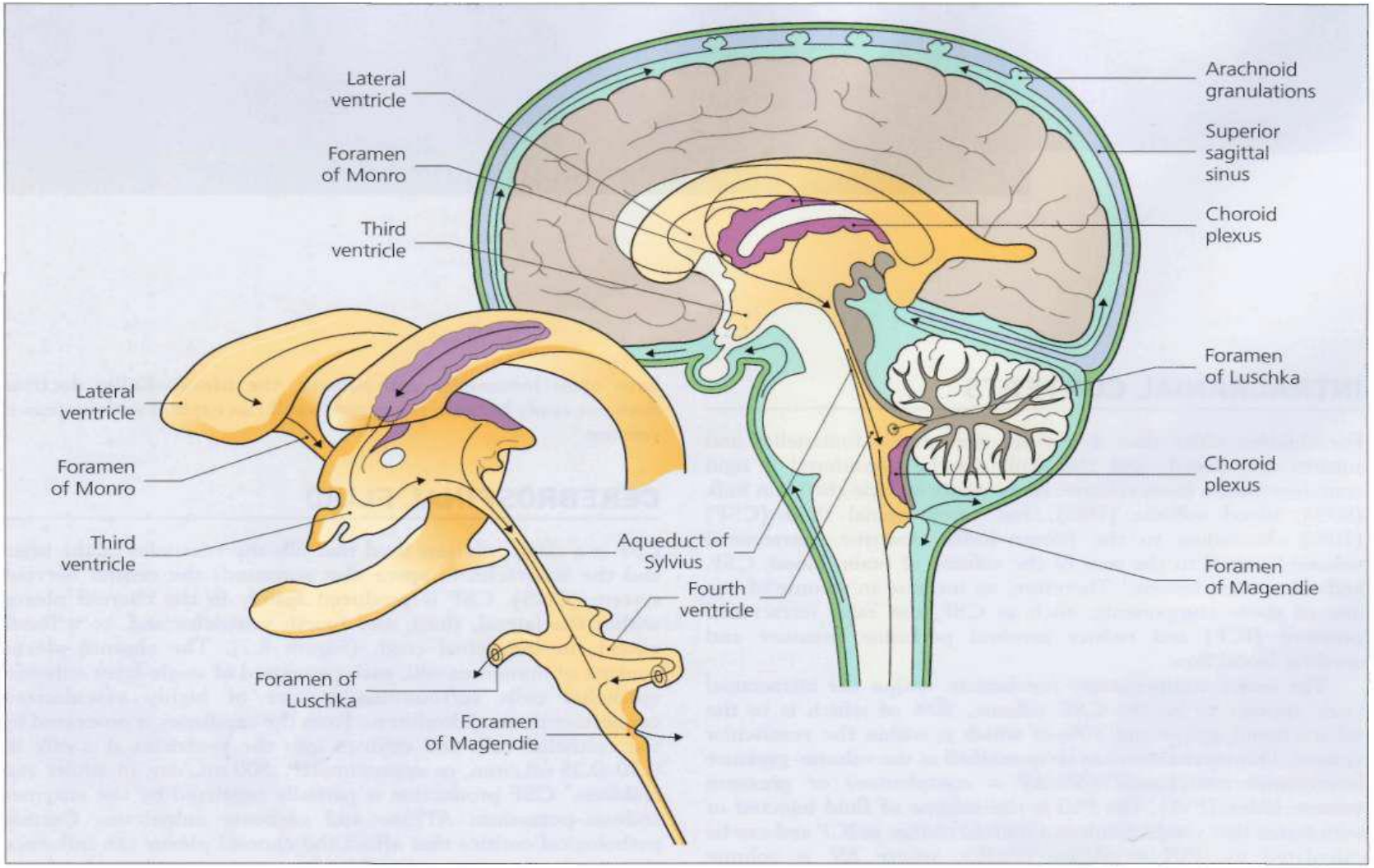
Physiology of CSF circulation

- The ventricular system consists of two lateral ventricles,a third ventricle, and a fourth ventricle.
- CSF is actively secreted in all four ventricles by choroid plexus through complex mechanisms.
- The CSF reabsorption surface is at the subarachnoid space arachnoid granulations, and along the cranial and spinal nerve roots.


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


Body
LATERAL VENTRICLE
Frontal horn
Foramen of Monro
3RD VENTRICLE
Atrium
Occipital horn
= Choroid plexus
Temporal horn
Cerebral aqueduct
4TH VENTRICLE
Luschka
Magendie
Central canal (spinal cord)

The ventricular system



The cerebrospinal fluid (CSF)

- CSF is produced mainly in the choroid plexus

within the lateral, third, and fourth ventricles and, to a lesser extent, in the spinal cord.

- The choroid plexus consists of numerous villi, each composed of single-layer cuboidal epithelial cells surrounding a core of highly vascularized connective tissue.
- Ultrafiltrate from the capillaries is processed by the epithelial cells and diffuses into the ventricles at a rate of 0.30-0.35 mL/min, or approximately 500 ml/day, in adults and children.
- CSF production is partially regulated by the enzymes sodium-potassium ATPase and carbonic anhydrase.

What is cerebrospinal fluid(CSF)

- Clear colorless fluid
- Total volume: 150mls
- 25mls in ventricles + 125mls in subarach.space
- Production: 500mls/day and turnover: 3-4 times
- Contents:

1. A-cellular (<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

The cerebrospinal fluid (CSF)

- Certain pathological entities that affect the choroid plexus can influence the production rate of CSF. For example, choroid plexus papillomas can increase CSF production, while ventriculitis can lead to sclerosis of the choroid plexus, which in turn will reduce production.
- The total CSF volume is approximately 40-50 mL in neonates and 65-140 mL in children.
- 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. A layer of endothelial cells lines the villi. Water and electrolytes freely traverse the endothelial layer into the systemic circulation, and proteins are actively transported out by pinocytosis.

Arachnoid villi

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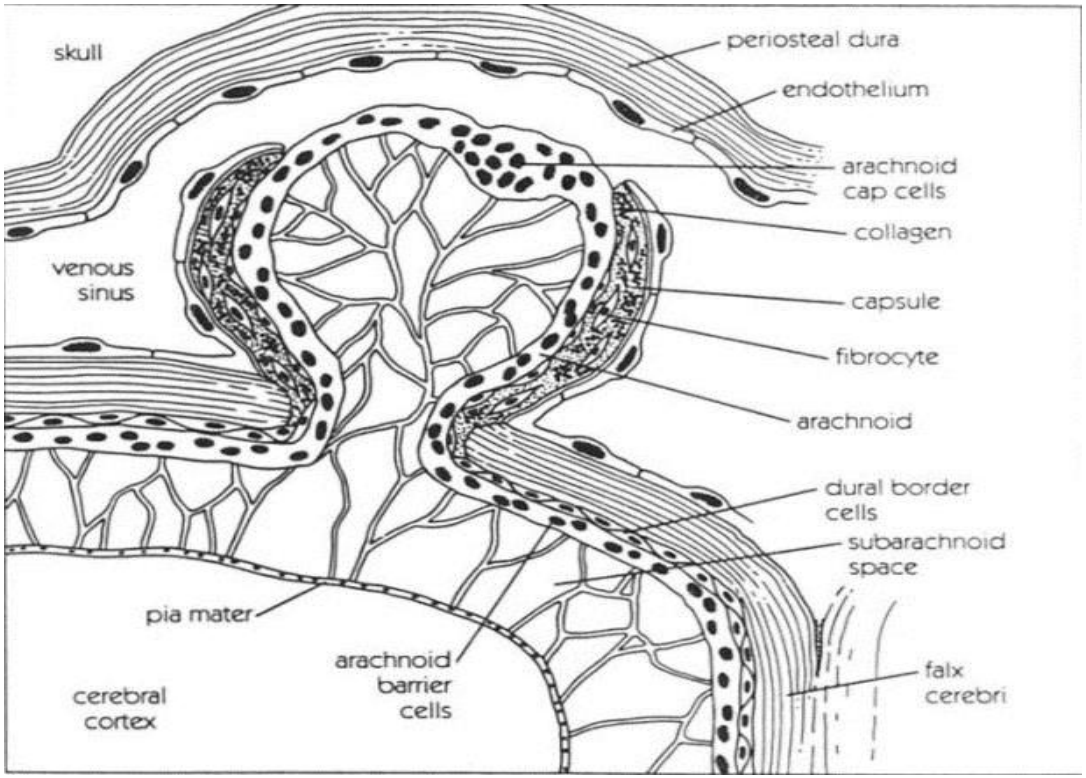


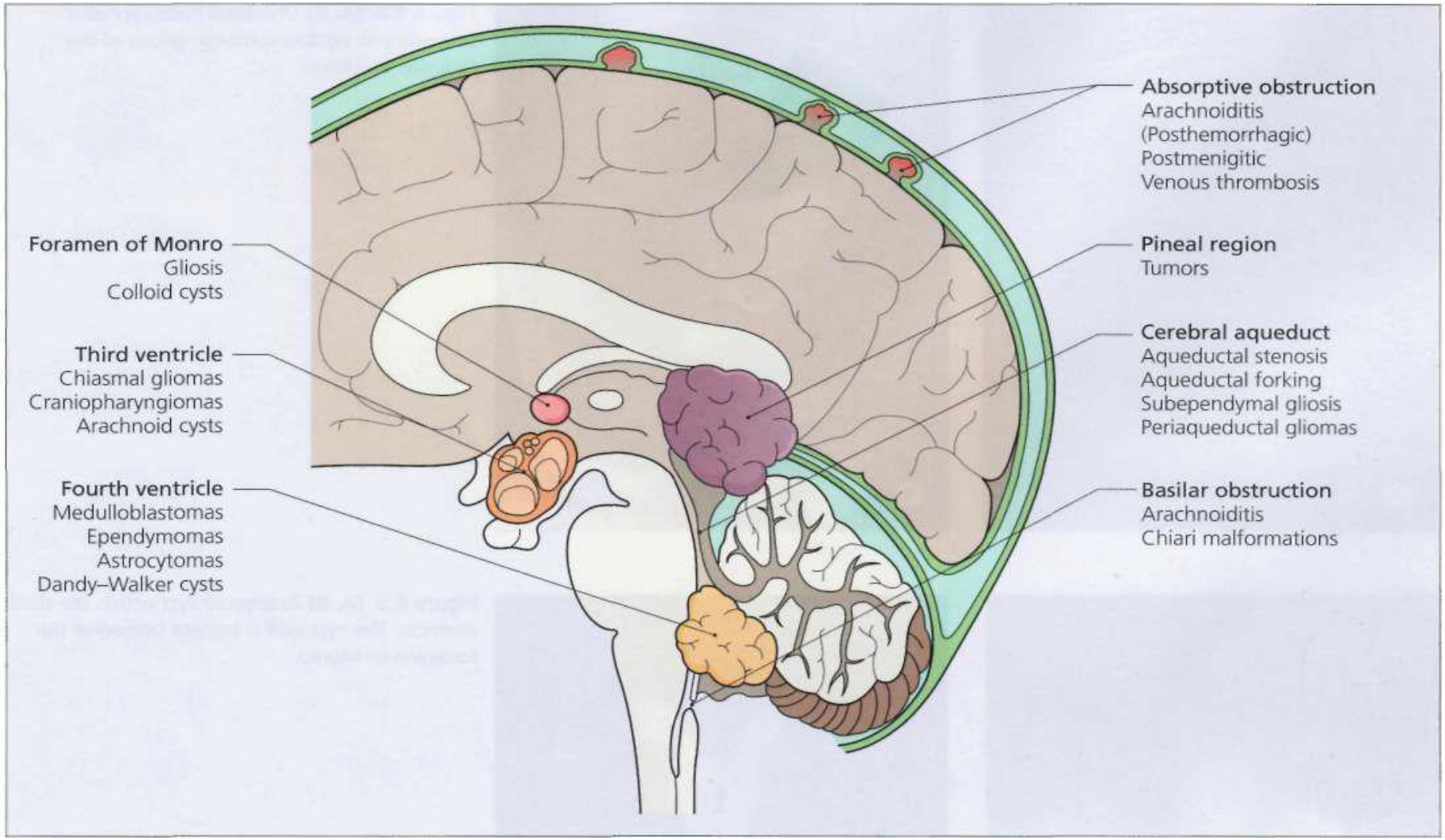
IN GENERAL CSF WILL ACCUMULATE DUE TO:

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

PATHOPHYSIOLOGY OF HYDROCEPHALUS

- Hydrocephalus is often associated with dilatation of the ventricular system and increased ICP.
- The incidence of pediatric hydrocephalus as an isolated congenital disorder is approximately *1/1000* live births. Pediatric hydrocephalusis often associated with numerous other conditions, such as spina bifida.
- 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.



- 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 intracranial pressure 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.***
- *Traditionally, this* classification was based on whether dye injected into the lateral ventricles could be detected in CSF extracted from a subsequent lumbar puncture.
- Currently, the term "noncommunicating hydrocephalus" refers to lesions that obstruct the ventricular system, either at the aqueduct of Sylvius or basal foramina(i.e. basal foramina of Luschka and Magendie).
- The term "communicating hydrocephalus" refers to lesions that obstruct at the level of the subarachnoid space and arachnoid villi.

- Another classification infrequently employed in the literature describes hydrocephalus as *internal or external, depending on whether the site of obstruction is proximal*or distal to the basal foramina.
- Hydrocephalus can also be classified by etiology and site of obstruction.

AETIOLOGY - SITE

1. **Lateral ventricles**

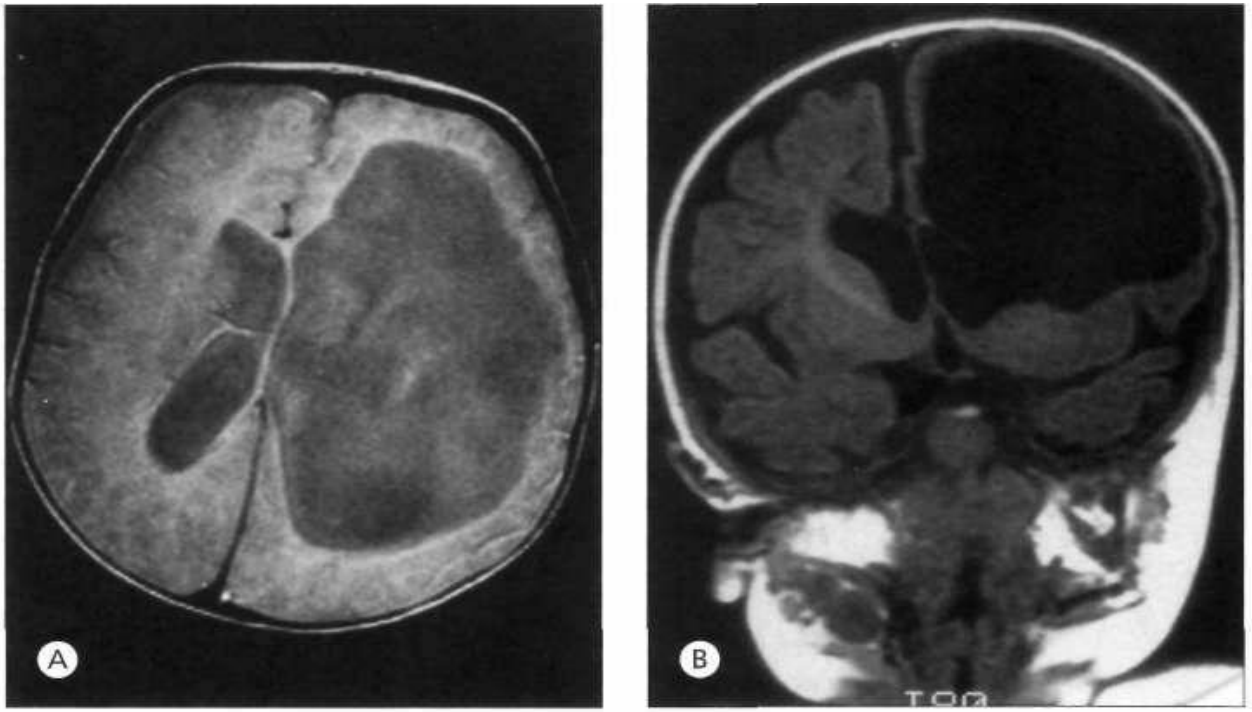
Choroid plexus tumors:

- Rare in the general population(0.4 to 0.6% of all CNS tumors).More In the pediatric population(1.5 to 3.9% of all pediatric CNS tumors).
- Most choroid plexus tumors are choroid plexus papillomas,.With CSF production rates three to four times the normal.
- Endoscopic coagulation of the choroid plexus or surgical removal of the papilloma has been used to

successfully treat the hydrocephalus.

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.
- The resulting unilateral ventriculomegaly is often occult until after age 6, and may enlarge the ipsilateral hemicalvarium (Figure).



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, and usually obstruct both foramina of Monro. Considered to becongenital lesions, they can become symptomatic at any age.

They can cause either intermittent, acute, life-threatening hydrocephalus or chronic hydrocephalus. They are customarily treated with stereotactic aspiration of the cyst, resection via craniotomy, or endoscopic resection .

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.

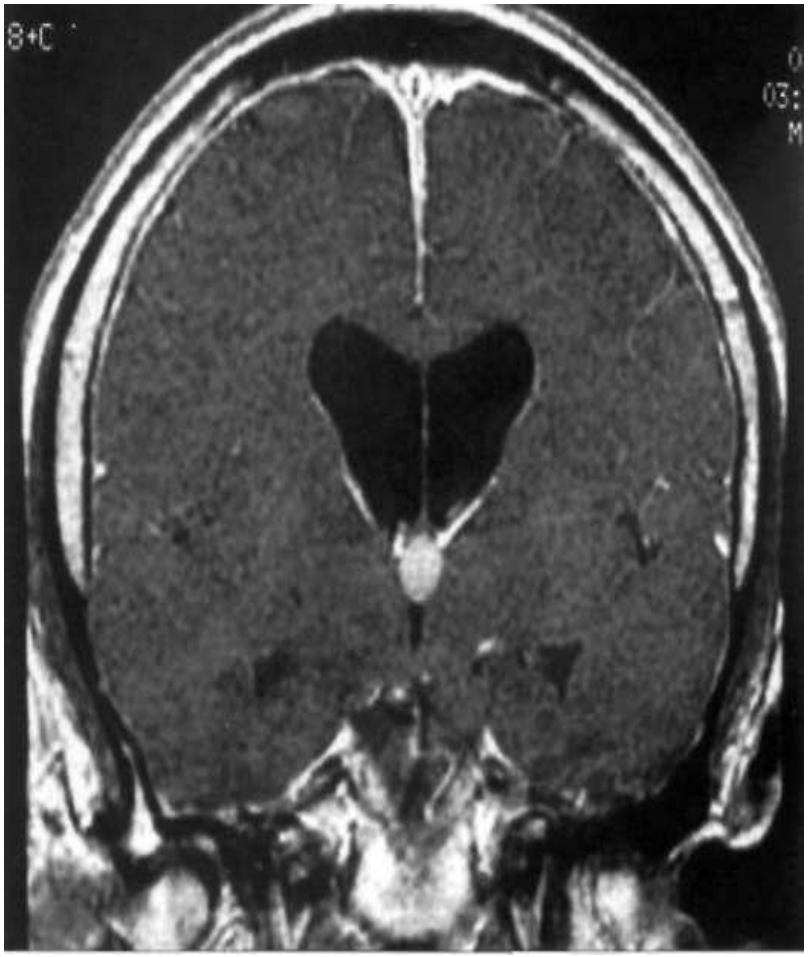
The endoscopic fenestration is a treatment option.

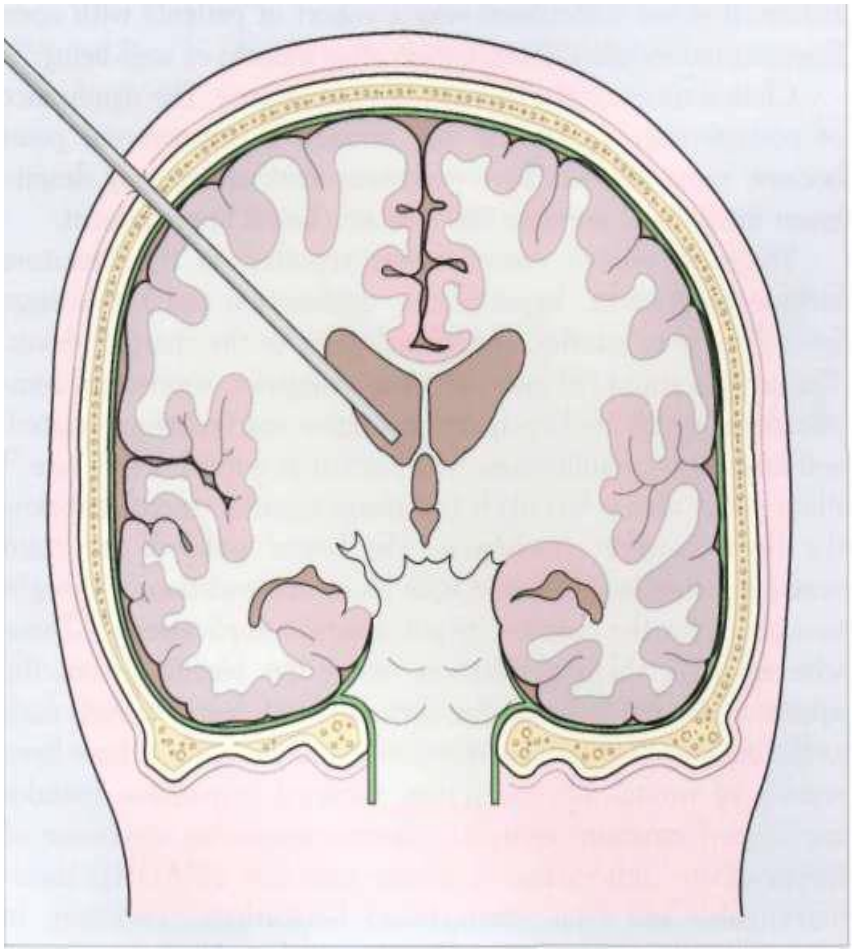
Third ventricle

- The most common pediatric neoplasms that obstruct the third ventricle are **craniopharyngiomas** and **chiasmal-hypothalamic gliomas**.
- Hydrocephalus secondary to craniopharyngiomas usually resolves after surgical resection of the tumor; hydrocephalus secondary to third ventricular gliomas usually does not resolve after surgical resection, and shunt placement is often necessary.

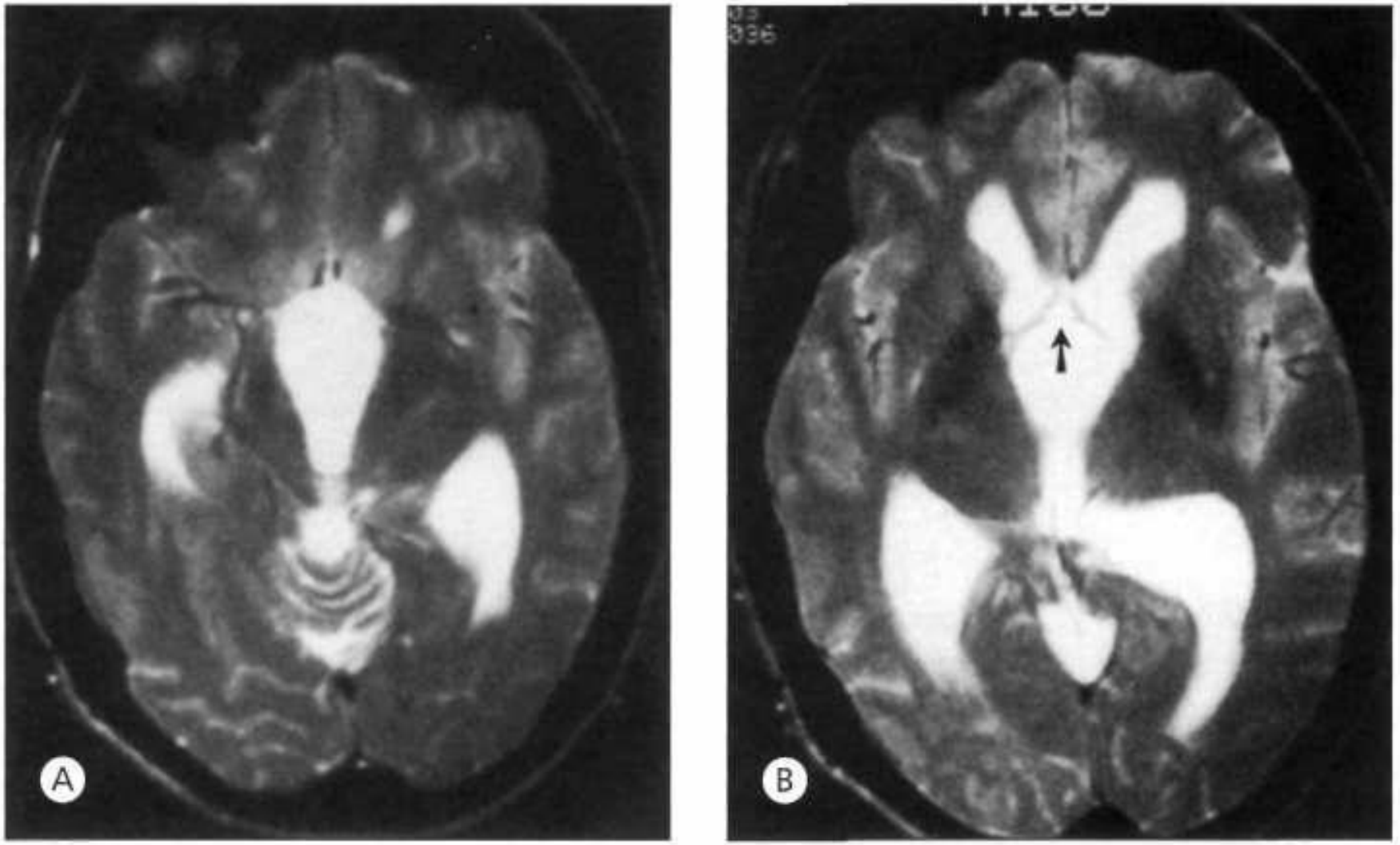
Endoscopic techniques are ideal for colloid cyst removal.

Coronal magnetic resonance image demonstrating enhancement of a colloid cyst





T2 axial MRI image: Arachnoid cyst within the third ventricle. The cyst wall is evident (arrow) at the foramina of Monro.

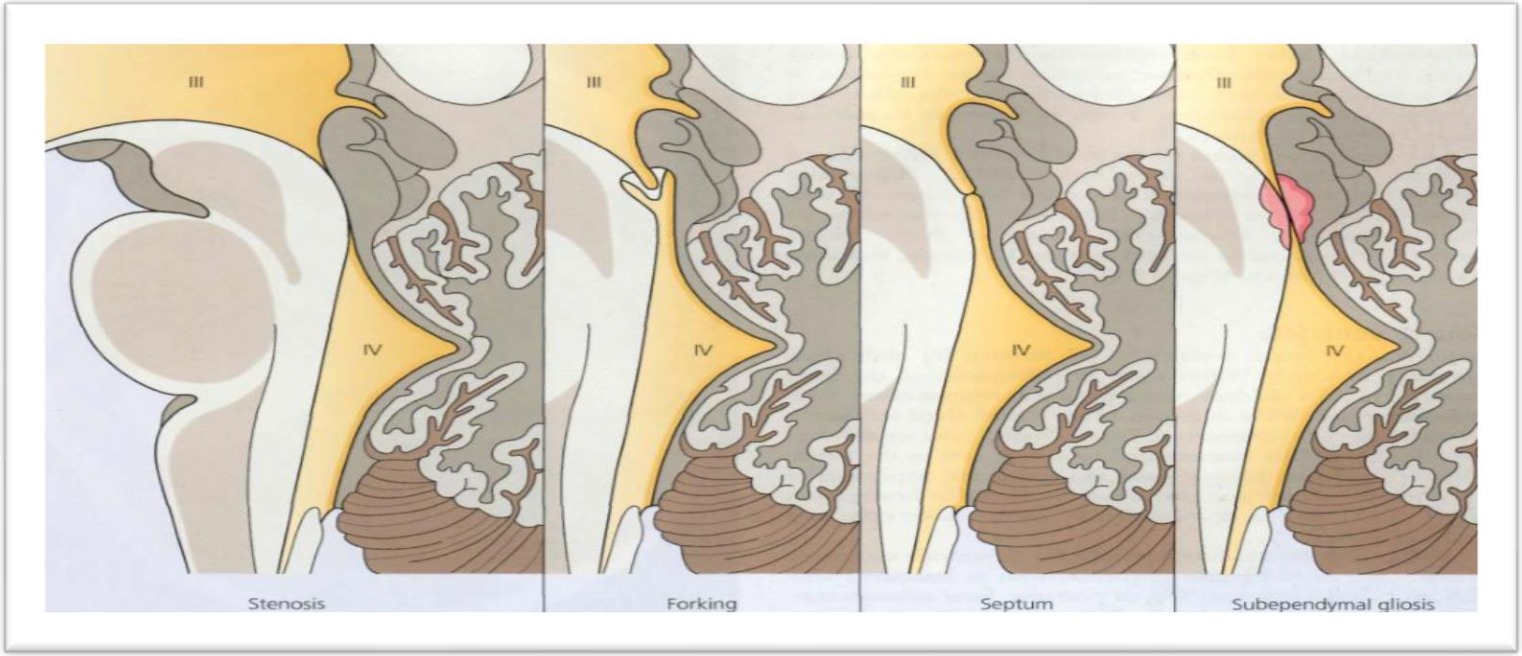


4-Sylvian aqueduct

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)

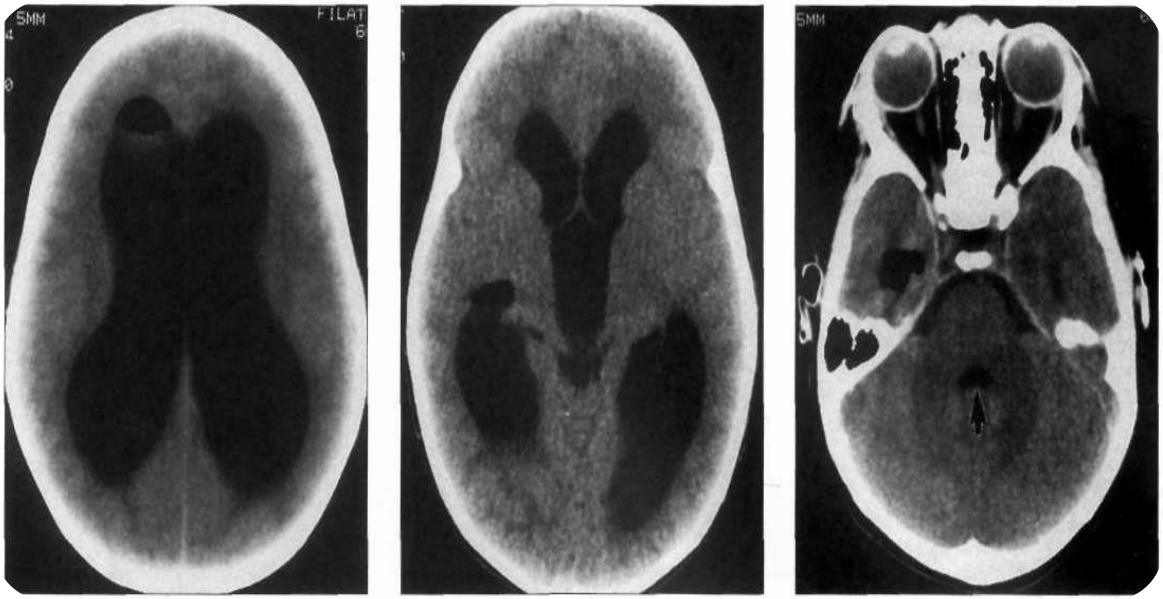
It may be secondary to *in utero infections (e.g. toxoplasmosis), intraventricular* hemorrhage, or mumps encephalitis.



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

Sylvian aqueduct

- Hydrocephalus secondary to aqueductal occlusion is generally severe and causes distension of the third and lateral venticles with small fourth ventricle and compression of the cerebral hemispheres (Figure).
- Less than 2% of cases of congenital aqueductal stenosis are the result of the 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.

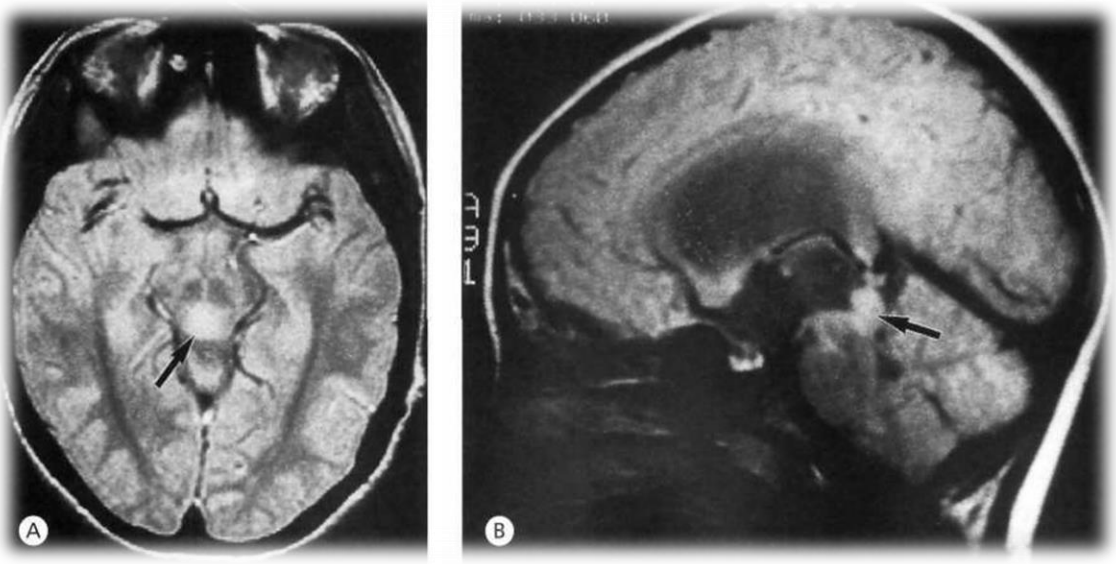
Sylvain aqueduct

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.

(A) Axial and (B) sagittal MR images demonstrating hydrocephalus secondary to a periaqueductal astrocytoma (arrow). The lesion was not visible on CT scans.



4-Fourth ventricle

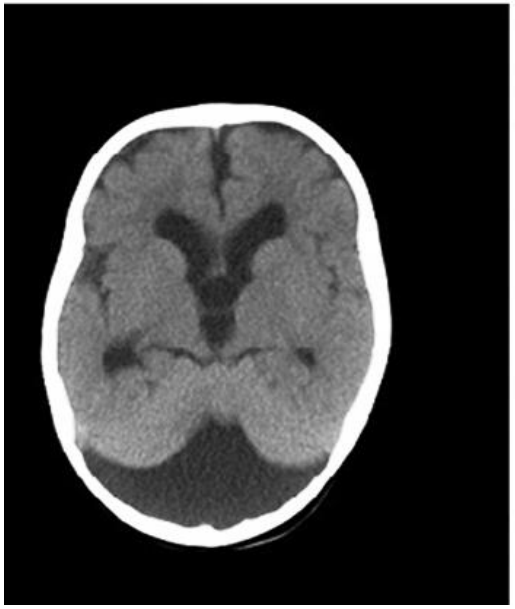
In infants, the fourth ventricle is the location for obstruction secondary to Dandy-Walker cysts or obliteration of the basal foramina.

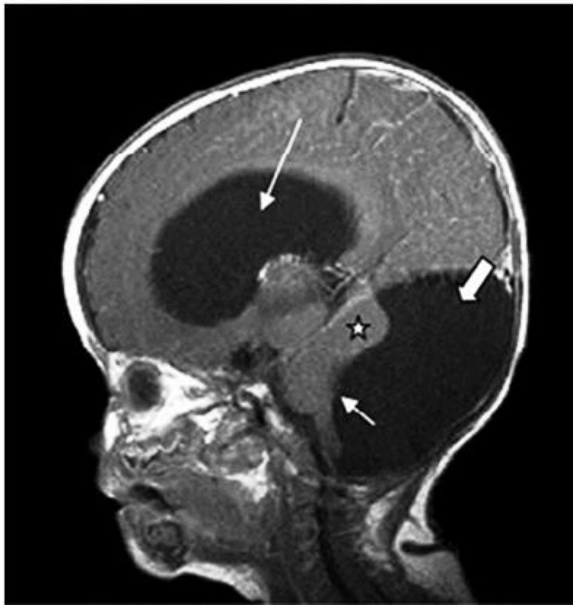
In older children, neoplasms are a common cause.

Such occlusions result in the dilatation of the lateral, third and fourth ventricles above the obstruction.

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.





Pediatric tumors associated with the fourth ventricle commonly

present with hydrocephalus.

Hydrocephalus is associated with 85% of **medulloblastomas,** 65% of posterior fossa **astrocytomas,** 75% of **ependymomas**, and 25% of **brainstem gliomas.**

Arachnoiditis secondary to either meningitis or subarachnoid

hemorrhage can occlude the basal foramina and cause obstructive

hydrocephalus.

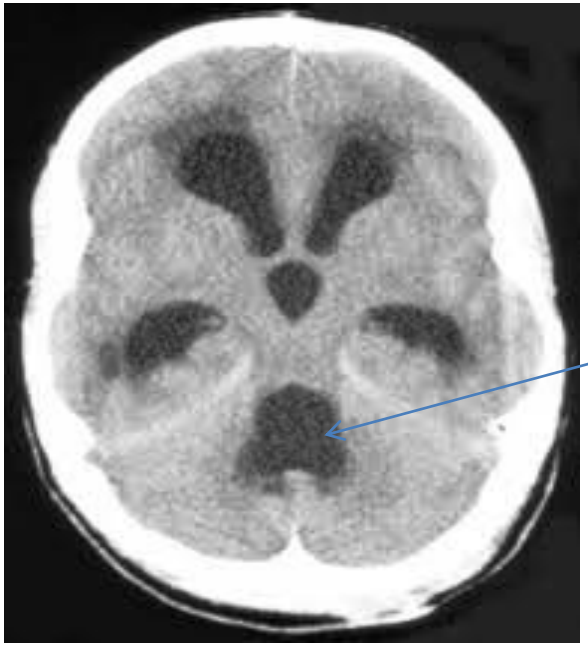
Infants with **Chiari II malformations** and myelomeningoceles have hydrocephalus secondary to blockage of CSF flow from basilar obstruction

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)

This will impair the CSF circulation in the subarachnoid space and CSF absorption causing pan ventricular dilatation-communicating hydrocephalus.

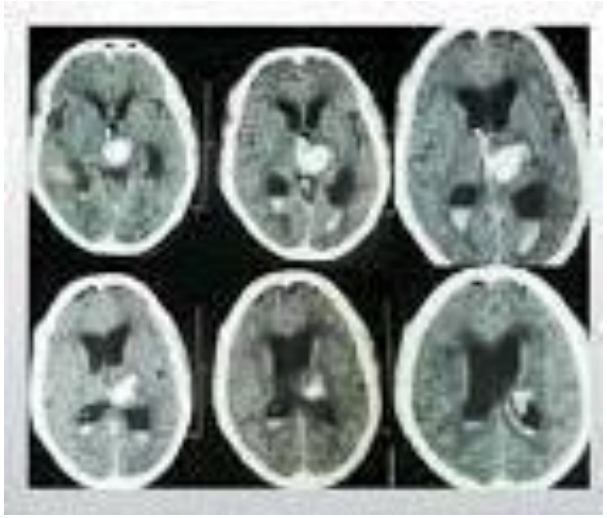


Dilated fourth ventricle

Etiology by age

- **Premature infants**

Hydrocephalus in premature infants is predominantly caused by(intraventricular hemorrhage) IVH. The hemorrhage occurs in the germinal matrix, and can extend into the ventricles and parenchyma depending on its severity. There is a general correlation between the amount of IVH and the likelihood of post hemorrhagic hydrocephalus



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.

Etiology by age

- **Older children**

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

Etiology by age

- **For adults,**

hydrocephalus can be classified to:

1. Acute and chronic,
2. Or primary (idiopathic)and secondary to a known pathology.

Among almost 1000 cases of adult hydrocephalus assembled 34% were idiopathic.

Causes among cases where etiology was determined included subarachnoid hemorrhage(51%), head injury (16%), brain tumor (9%), intracranial surgery

(7%), aqueduct stenosis (5%), and meningitis (5%).

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

IN PREMATURE INFANTS

- Poor feeding and vomiting are uncommon signs of hydrocephalus in premature infants.
- If ventriculomegaly progresses and ICP increases, the anterior fontanelle becomes convex, tense, and non pulsatile, and the scalp veins distend.
- As ventriculomegaly persists, the head develops a globoid shape, and the head circumference increases at a rapid, pathologic rate. Head circumference increases 0.5 cm/week in sick premature infants, 1 cm/week in healthy premature infants, and up to 2 cm/week in premature infants with PHH.
- Lateral rectus palsies and the "setting-sun" sign seen in older infants and children is not a common presentation because the neonate skull is so distensible and will initially expand to compensate for expanding ventricles.

Full-term infants

- **Symptoms** include: irritability, vomiting, and drowsiness.
- **Signs** include :

1. Macrocephaly(big head).
2. Convex and full anterior fontanelle,
3. Distended scalp veins,
4. Cranial suture splaying,
5. Frontal bossing,
6. Cracked pot sound on percussing over dilated ventricles (positive Macewen's sign),
7. Poor head control,
8. Lateral rectus palsies,
9. and the "setting-sun" sign, in which the eyes are inferiorly deviated. Paralysis of upgaze and Parinaud's sign herald dilatation of the suprapineal recess and compression of tectal part of midbrain.

Full and tense anterior fonatanelle

Dilated scalp veins

Suture widening

Setting sun eyes

Head circumference

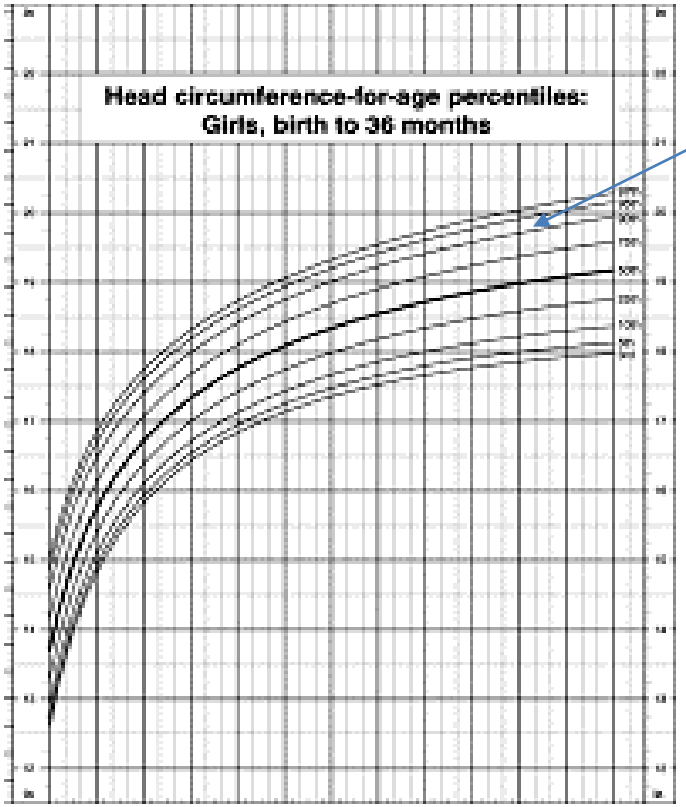
- Normal head circumference for full-term infants is **33-36 cm**at birth.
- Head circumference increases by 2 cm/month during the first 3 months, by 1 cm/month from 4 to 6 months, and by 0.5 cm/month from 7 to 12 months.
- Head circumference increases

that are progressive and rapid, crossing percentile curves on the head

growth chart are a stronger diagnostic indicator of hydrocephalus

than increases that are consistently above, but parallel to the 95%

percentile curve.



Head circumference above95%---abnormal

Older children

- Predominant symptom is usually a dull and steady **headache**, which typically occurs upon awakening.

It may be associated with

-**lethargy**,

-**vomiting**

-**blurred or double vision**.

-**decreased school performance** and

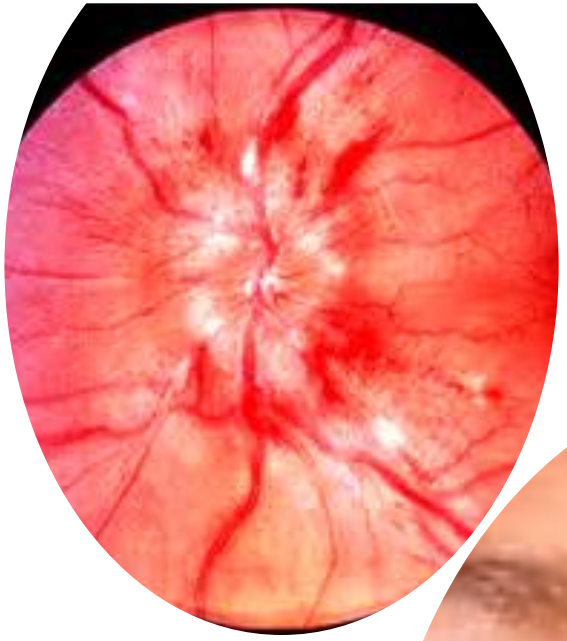
-**behavioral**disturbances

-**endocrinopathies** (e.g. precocious puberty, short stature, hypothyroidism).

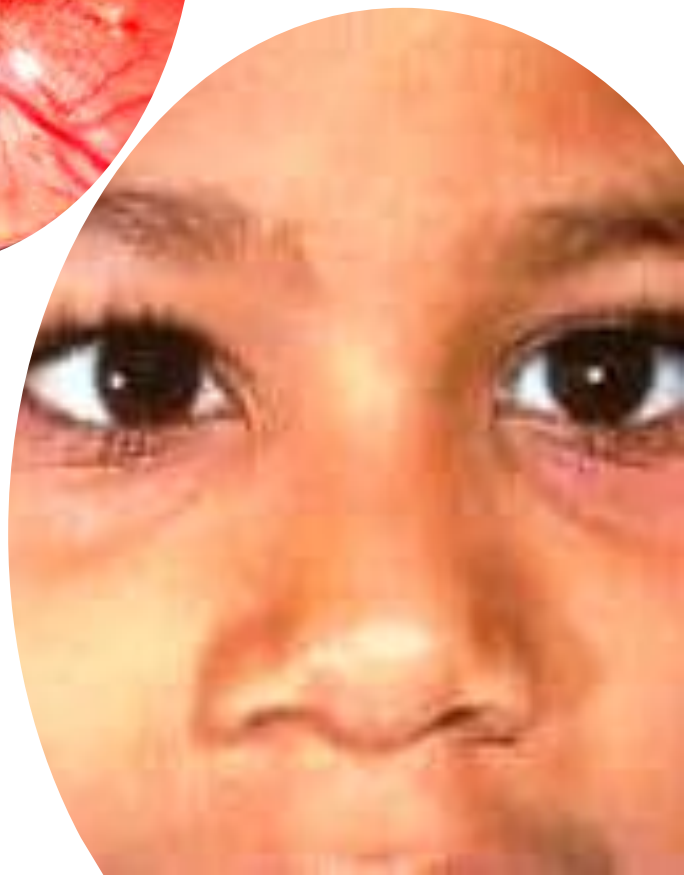
- Common signs include **papilledema and lateral rectus palsies** (unilateral or bilateral). **Hyperreflexia and clonus** are also seen.
- Rarely, children with hydrocephalus may experience transient or permanent blindness if the posterior cerebral arteries are compressed against the tentorium.
- If the hydrocephalus is severe, **Cushing's triad** of bradycardia, systemic hypertension, and irregular breathing patternsmay occur. This triad is rare and often denotes a very severe case of increased ICP requiring emergency treatment.

Signs of hydrocephalus in children and adults

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

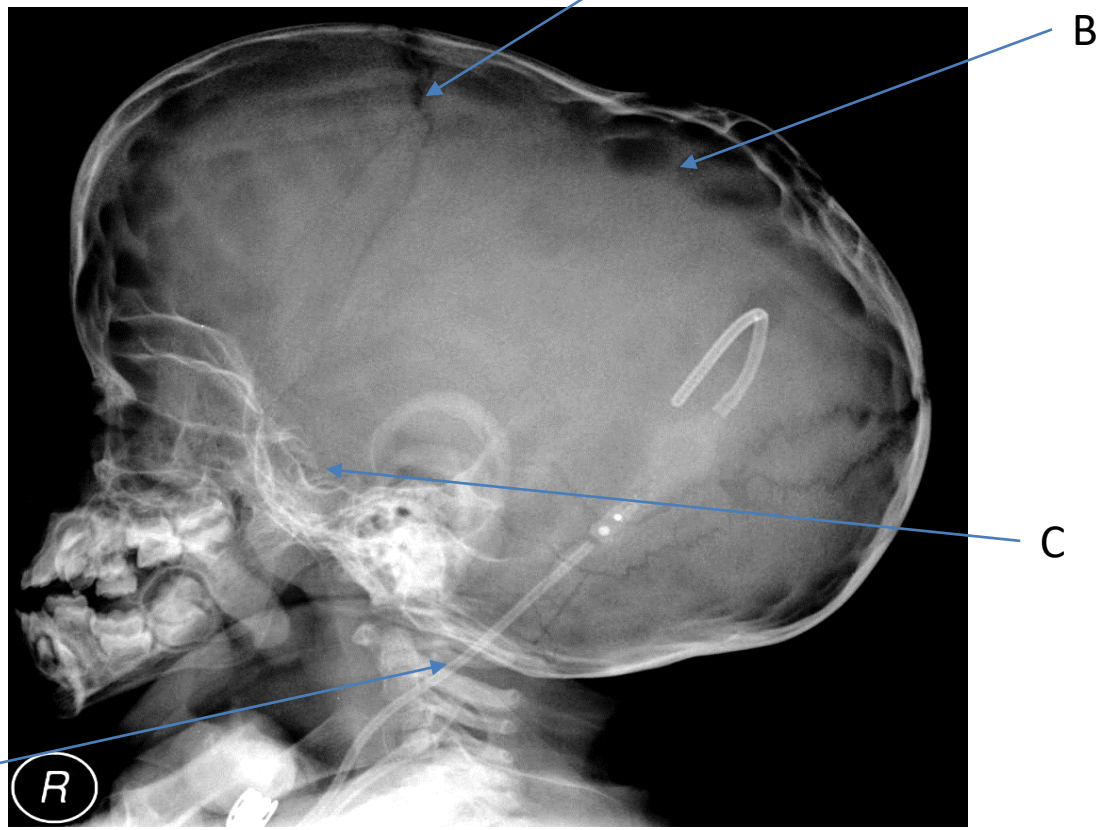


Right abducent nerve palsy



DIAGNOSTIC STUDIES

- **<u>Skull X-ray-</u>** demonstrate cranial suture separation in infants(A), beaten copper appearance (B)and enlarged sella (C)in older children and have since been supplanted by more modern imaging studies.
- N.B.,notice the shunt



Cranial ultrasonography

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

monitoring of ventricular size.

- By measuring through the anterior fontanelle of an infant, ultrasonography can demonstrate lateral and third ventricular morphology and intraventricular clots.

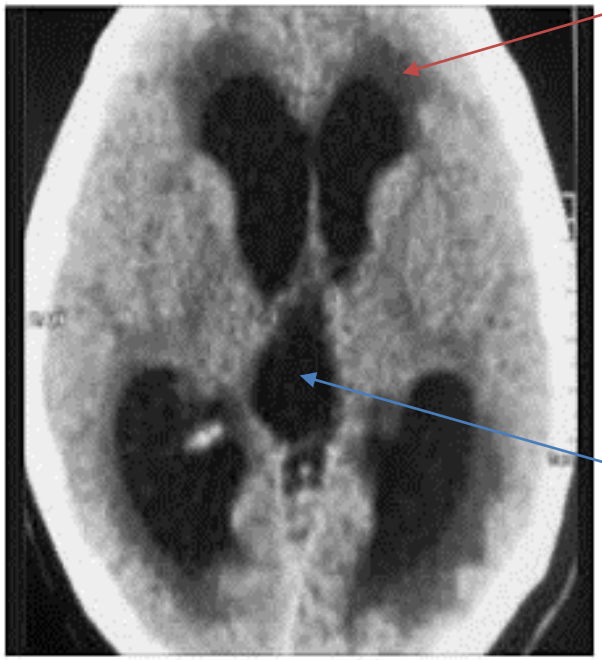
CT scanning

- From 1976 to 1986, CT scanning was the definitive method of

diagnosing pediatric hydrocephalus.

- CT images can accurately demonstrate ventricular size and shape, as well as the presence of blood and calcium deposits.
- Signs of increased ICP, such as compressed cerebral sulci, obliterated subarachnoid spaces over the convexity, and transependymal resorption of CSF into the white matter can all be detected on CT.
- With intravenouscontrast injections, CT scans can also reveal abnormalitiessuch as tumors and abscesses.
- CT scans are limited because they can only be performed predominantly in the **axial plane**, require

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



Periventricular fluids

3rd vent.,

A CT-scan of the head for an infant who presented with progressive irritability- you can see the dilated third ventricle ,both lateral ventricles and periventricular lucienies suggestive of active hydrocephalus

Brain MRI

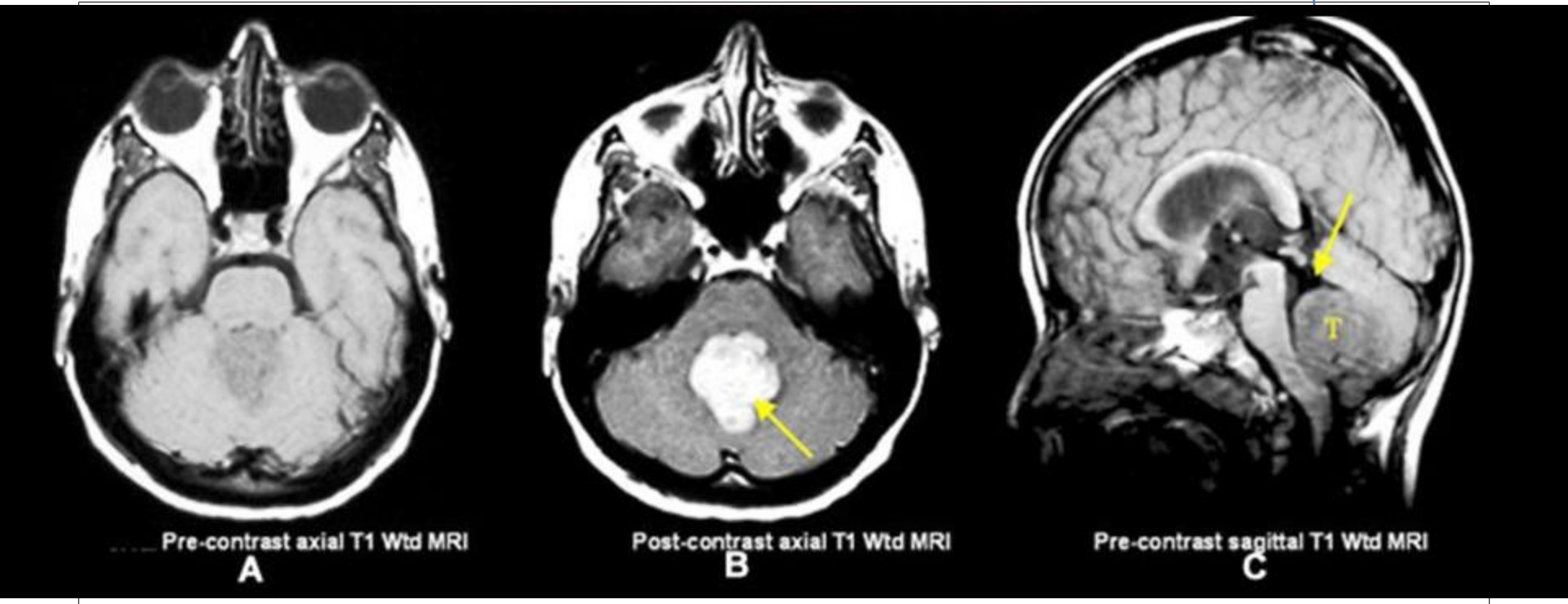
Since 1986, CT has been augmented by the introduction of commercially available MRI, which can project the brain in axial,coronal, and sagittal projections.

In addition, MRI can detect transependymal resorption and low-grade gliomas more clearly

than CT, as well as determine CSF flow across the aqueduct.

However, MRI does not supplant CT entirely because it does not demonstrate calcium as well.

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 treatment of hydrocephalus can be divided into

-Nonsurgical approaches and

-Surgical approaches, which in turn can be divided

into non-shunting or shunting procedures.

- The goals of any successful management of hydrocephalus are:

(1) optimal neurological outcome and

(2) preservation of cosmesis.

- The radiographic finding of normal-sized ventricles should not be considered the goal of any therapeutic modality.

Nonsurgical options

- There is no nonsurgical medical treatment that definitively treats

hydrocephalus effectively.

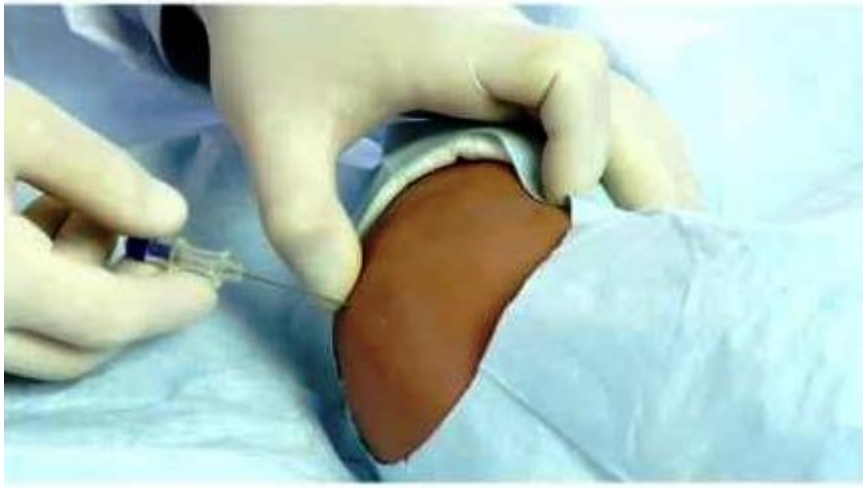
- Even if CSF production were to be reduced by 33%, ICP would only modestly decrease by 1.5 cmH2O.
- Historically, acetazolamide and furosemide have been

used to treat hydrocephalus. BUT they do not significantly reduce

ventriculomegaly.

- Other nonsurgical therapies tried, which have all been ineffective, include glycerol, isosorbide, radioactive gold, and serial head wraps.
- Serial lumbar or ventricular punctures to evacuate CSF are nonsurgical procedures to treat premature infants.
- The goal is to temporize the progressive ventriculomegaly with daily aspirations of fluid until the CSF protein levels drop to< 1000 mg/dL and the infant has increased in body size to tolerate operative intervention.

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.*** For example, the

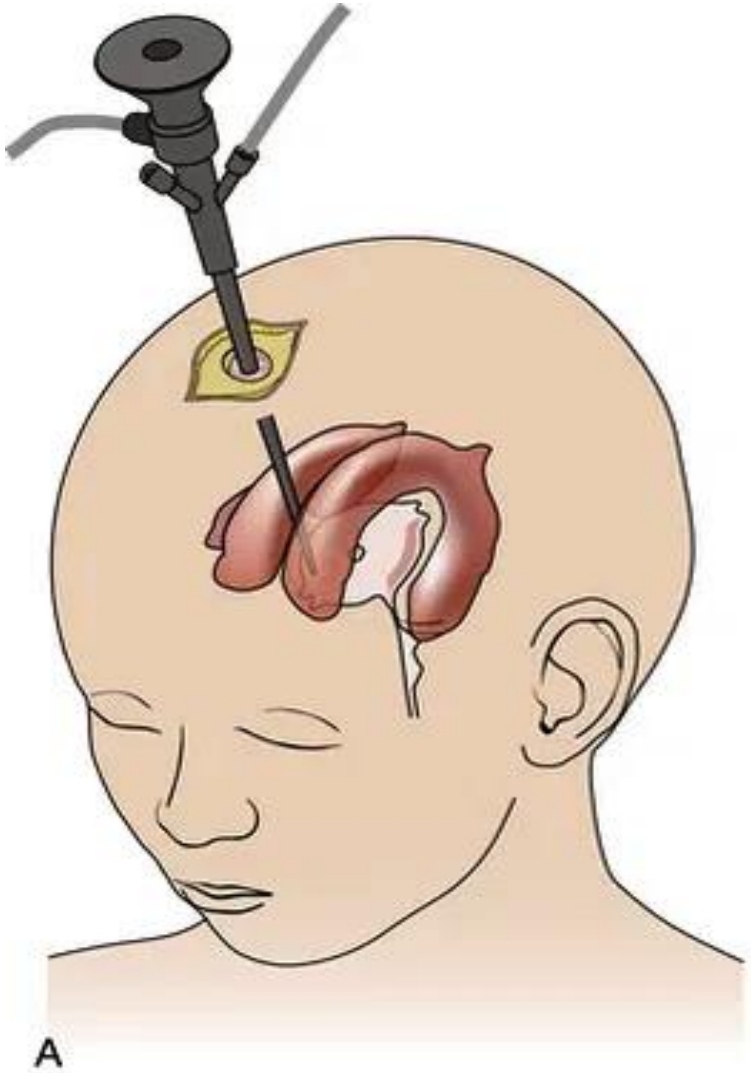
resection of tumors in the vicinity of the third and fourth ventricle often definitively treats the secondary hydrocephalus.

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

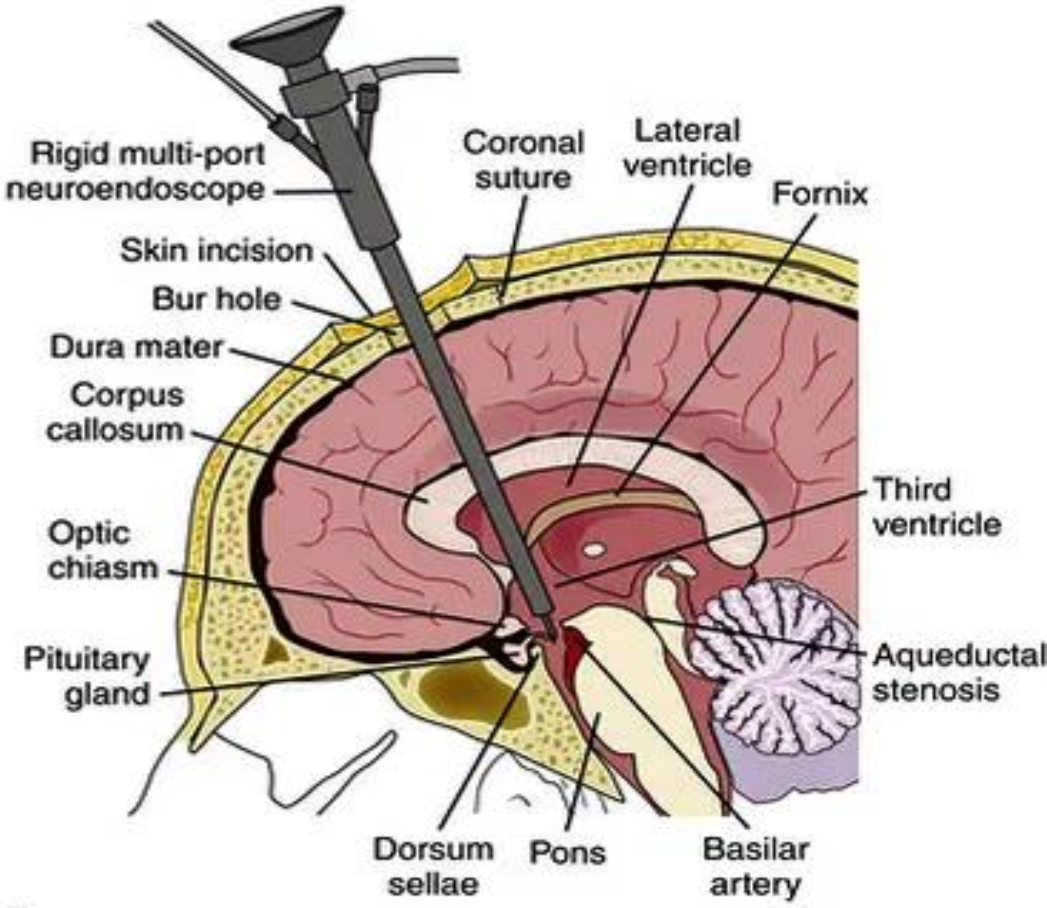
By surgically creating an opening at the floor of the third ventricle, CSF can be diverted without placing a shunt. Kamikawa and associates reported a 75% success rate for ETVamong 44 pediatric patients with hydrocephalus secondary to aqueductal stenosis; Other

- **Communicating hydrocephalus** is not an indication for a third ventriculostomy.

Endoscopic Third Ventriculostomy(ETV)



A



B

Surgical - CSF shunts

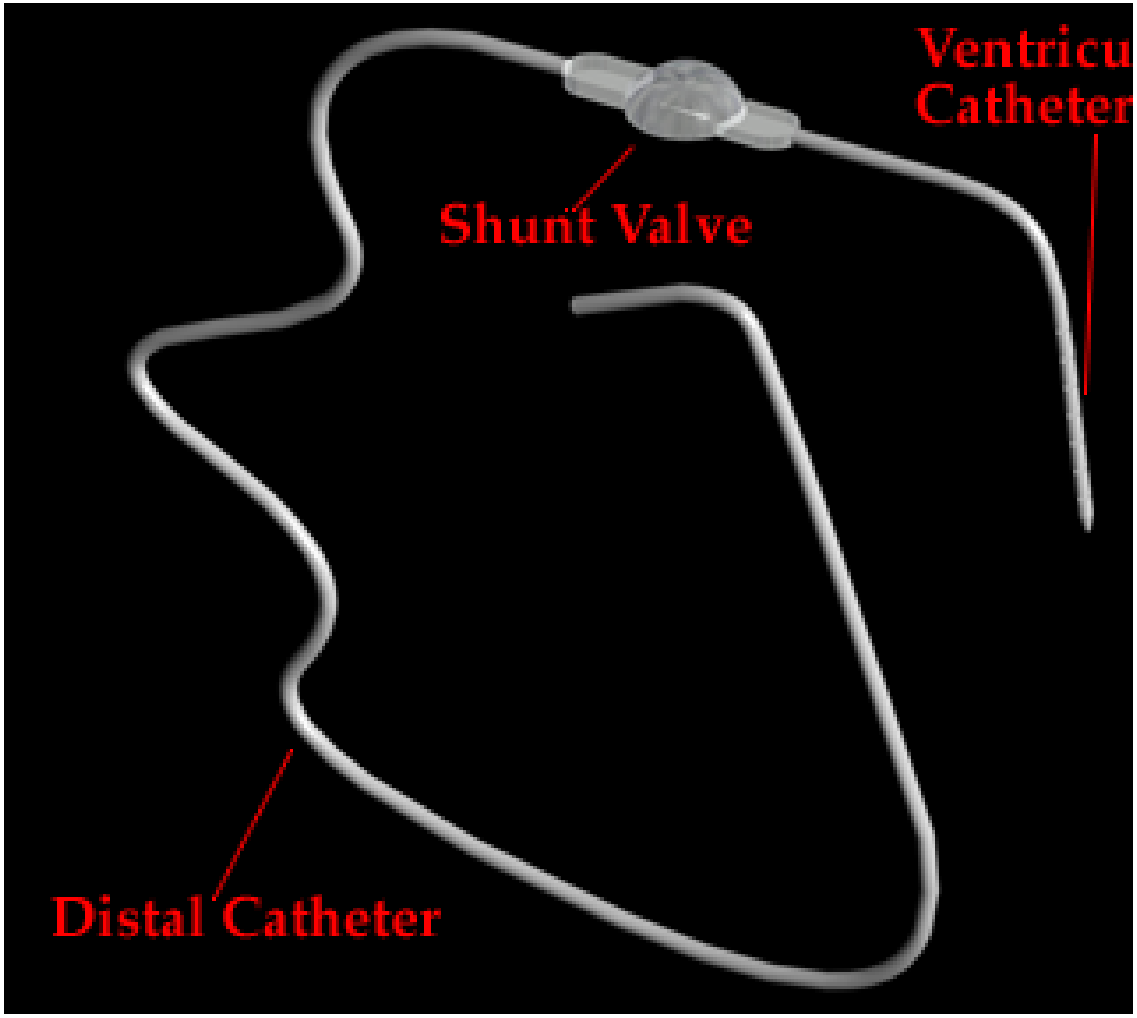
- CSF shunts are usually silastic tubes that divert CSF from the ventricles to other body cavities (i.e. peritoneal, atrium or pleural

space), where normal physiologic processes can absorb the fluid.

- Shunts typically have three components: **a proximal** (ventricular) catheter, a **one-way valve** that permits flow out of the ventricular

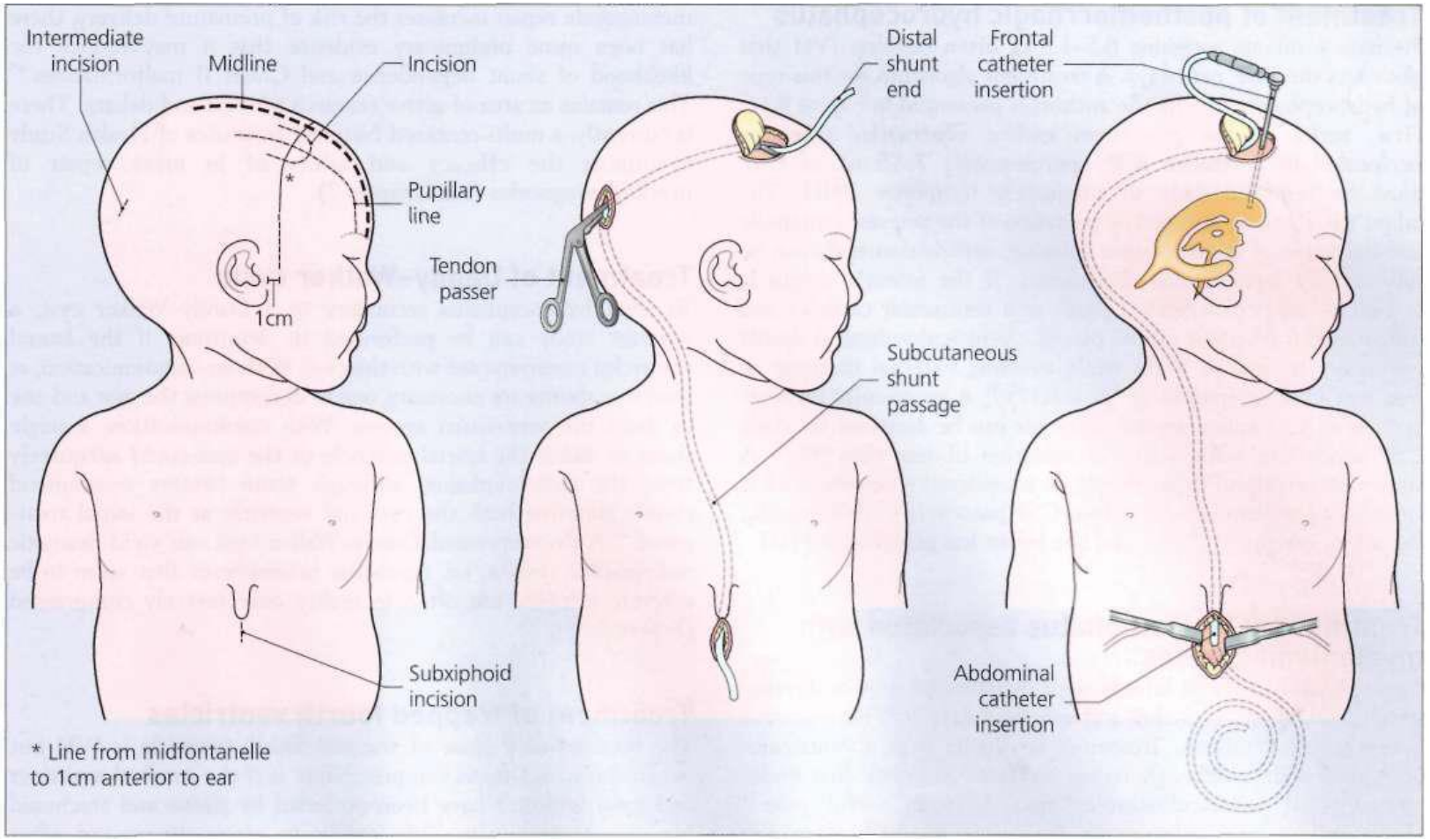
system, and a **distal catheter** that diverts the fluid to its eventual destination. Most shunts have built-in reservoirs that can be percutaneously aspirated for CSF.33

- Most shunt valves are pressure-differential valves, i.e. they are designed to open at designated pressures and remain open as long as the pressure differential across the valve is greater than the opening pressure.



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.

The goal in treatment of hydrocephalus

with a shunt is to decrease intracranial pressure and associated cerebral damage and simultaneously prevent complications

associated with the shunting procedure.

Shunt complications fall into three major categories:

(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.

- Approximately three quarters of all shunt infections become evident within 1 month of placement. Nearly 90% of all shunt infections are recognized within 12 months of the last shunt manipulation, as it is believed that most bacteria are introduced at the time of surgery.
- The offending organism is most often a member of the skin flora. *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.*

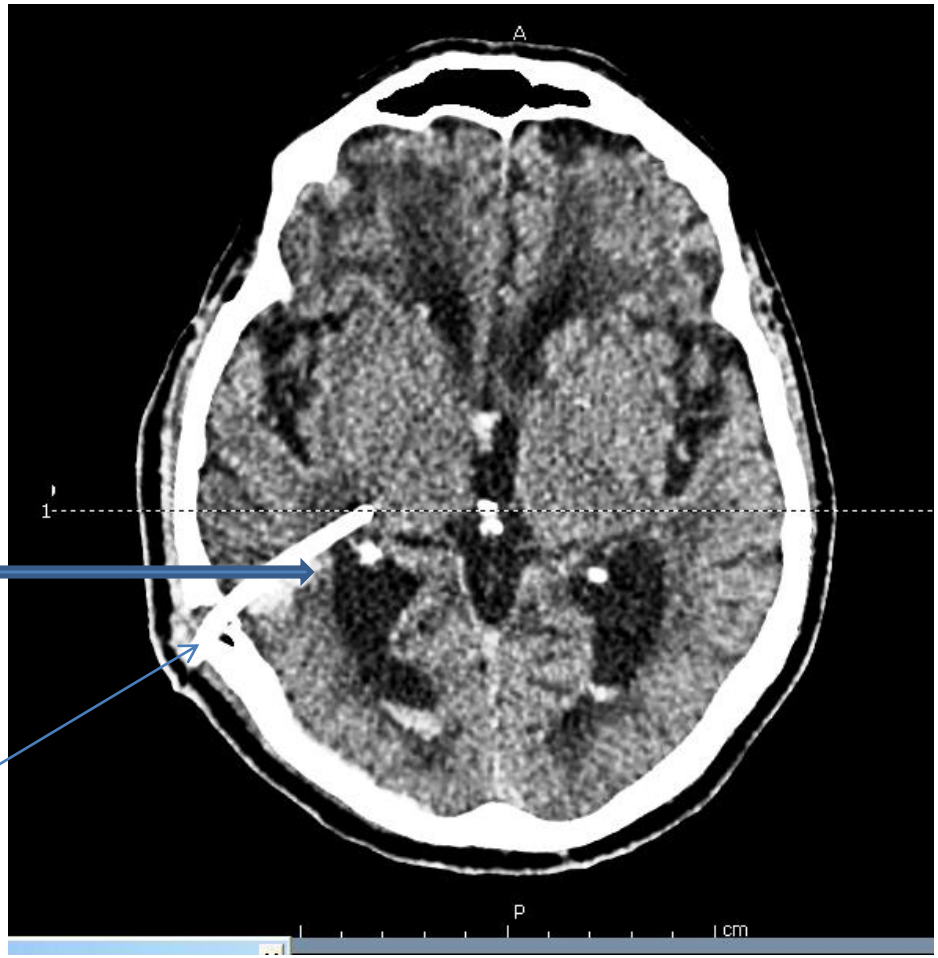
- **Common symptoms** include irritability and anorexia.
- **Common signs** include low-grade fever and elevated C-reactive protein. *Staphylococcus aureus infections often present with*erythema along the shunt track. Infected ventriculoatrial shuntsmay present with subacute bacterial endocarditis and shunt nephritis, an immune-complex disorder that resembles acute glomerulonephritis.
- **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. Infants with a shunt malfunction usually present with irritability, poor feeding, increased head circumference, and/or inappropriate sleepiness.
- 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.The presence of a fluid collection in the subcutaneous tissue in proximity to the shunt track is suggestive. The shunt valve can be"pumped," i.e. compressed several times against the skull, which may provide useful information.
- A head CT, as well as anteroposterior and lateral skull,chest, and abdominal radiographs, are obtained to evaluate for increased ventricular size and shunt hardware continuity.

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

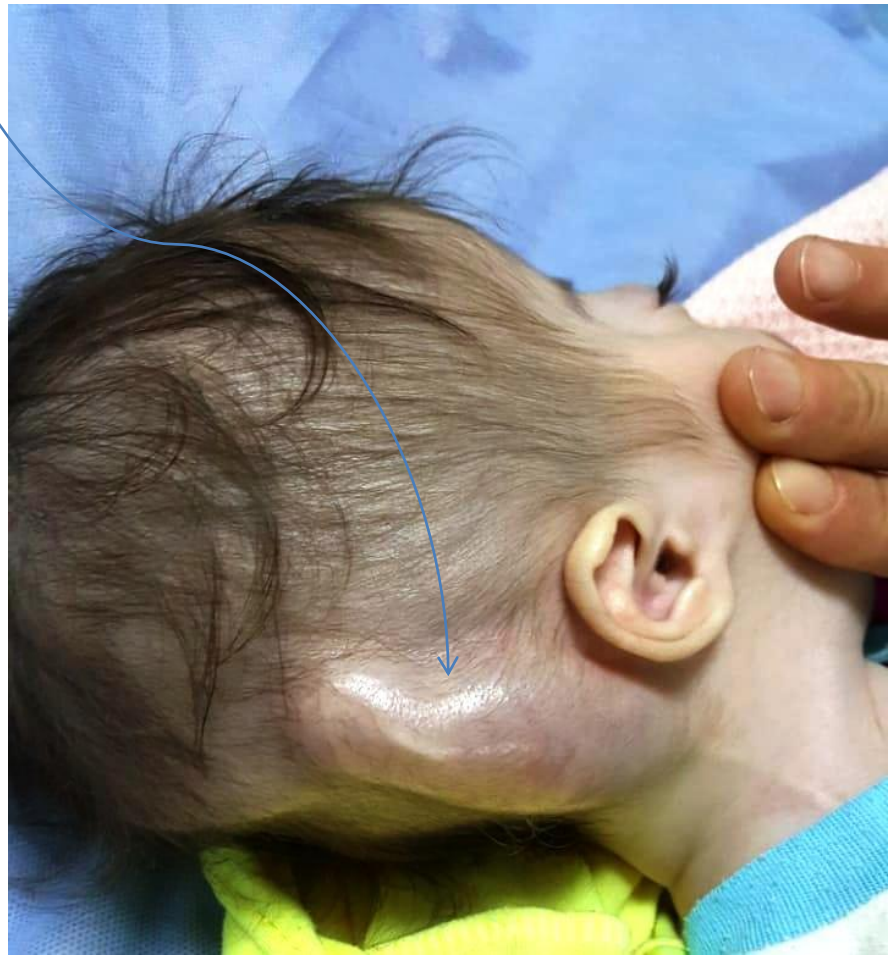
Hematoma at sit of shunt insertion



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.