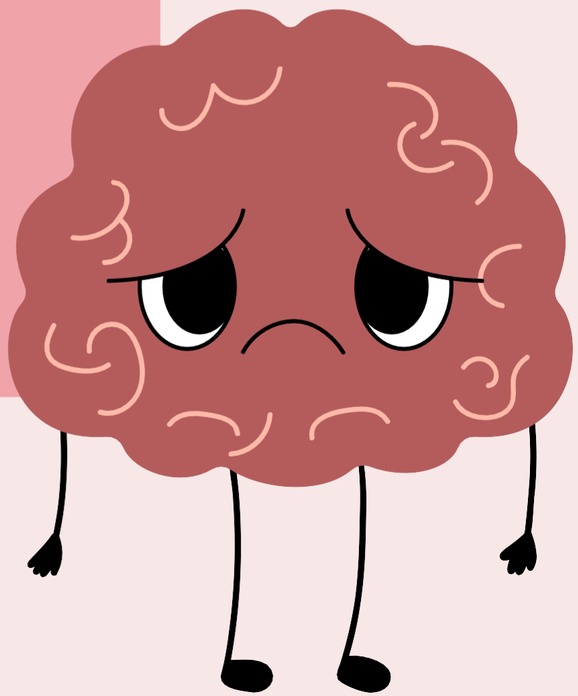
Congenital Anomalies of the brain



Presented by :

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Craniofacial development

Fontanelles

1.Anterior fontanelle

The largest fontanelle

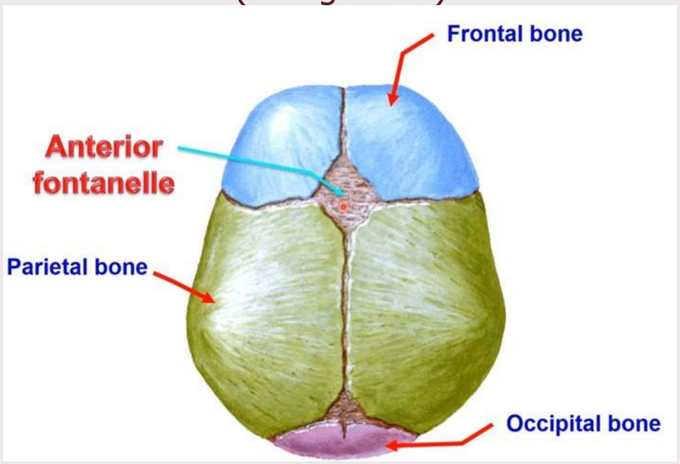
Diamond shaped

4cm (AP) × 2.5cm(transverse) at birth.

Normally closes by age of

18 months

(3 Finger rule)

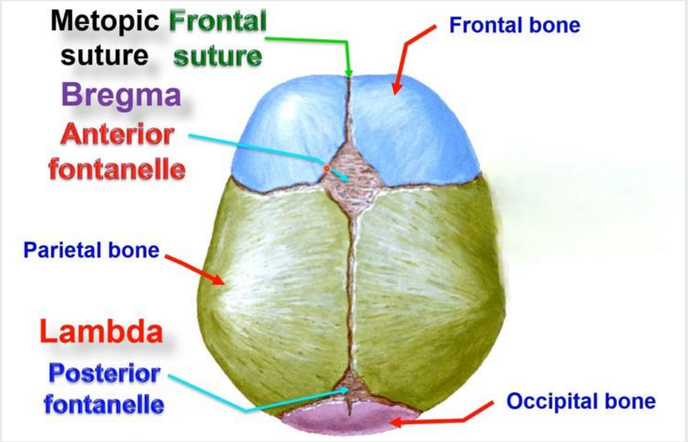


2.Posterior fontanelle

Triangular Shaped

Normally closes by age of

2-3 months



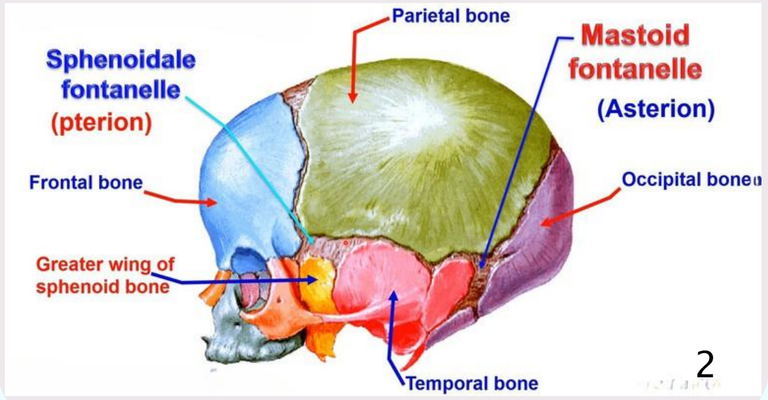
3.Sphenoid and mastoid fontanelles

Small , irregular

Normally closes by age of

Sphenoid:2-3 months

Mastoid :1 yr



Cranial valt development:

Growth :Growth is largely determined by brain development

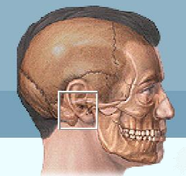
By 1 year: ~90% of adult head size is achieved
By 6 years: ~95% of adult head size is achieved

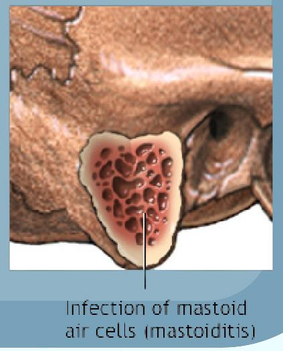
Growth essentially ceases around 7 years

By the end of the 2nd year, cranial bones interlock at the sutures; further growth occurs through accretion (deposition of new bone) and absorption (Remodeling of existing bone)
"Bone Remodeling"

Mastoid process Begins to develop around 2 years of age (Not present at birth !)

Air cells form by about 6 years, which is clinically important since this region can be affected by infections (mastoiditis).





Infection of mastoid air cells (mastoiditis)

Cranial valt development:

Bone Structure

At birth: skull is unilaminar (single bony layer).

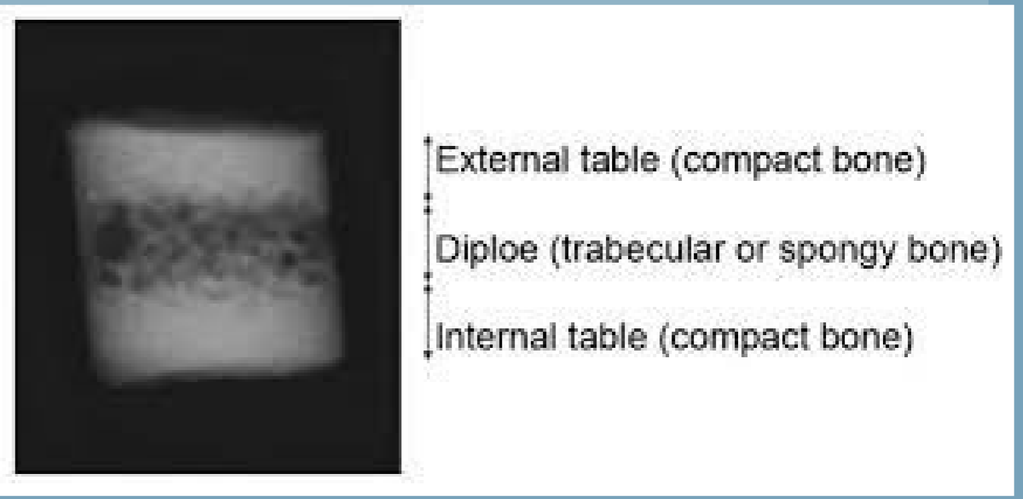
Diploë appears around 4 years of age

Diploë reaches maximum development by about 35 years, when diploic veins form.

Clinical Note

Infant skull: thin, fragile, and can be cut with a scalpel (soft)

Adult skull: thicker, denser, with Diploë → requires rougher surgical instruments



Craniosynostosis(CSO)

Old term: Craniostenosis

Definition: Premature ossification of one or more cranial sutures

Pathophysiology

Normally, the skull grows perpendicular to the suture lines

When a suture closes prematurely:

- Growth perpendicular to that suture stops.
- Compensatory growth continues parallel to the suture.
- This leads to characteristic skull deformities depending on which suture is affected.

Clinical Types & Deformities

Sagittal suture → Scaphocephaly (long, narrow skull).

Unilateral coronal suture → Plagiocephaly (asymmetrical skull).

Bilateral coronal sutures → Brachycephaly (broad, short skull).

Metopic suture → Trigonocephaly (triangular forehead).

Clinical significance

Cosmetic deformity (most common concern).

↑ Intracranial pressure in some cases (especially syndromic).

May be associated with genetic syndromes.

Epidemiology

Incidence: ≈0.6 per 1000 live births

May occur:

1) Isolated (most common).
2) Syndromic (part of genetic syndromes such as Crouzon, Apert).
3) Secondary (due to other diseases or intrauterine mechanical factors).

Craniosynostosis(CSO)

classification

Primary CSO → is usually a prenatal deformity. (syndromic)

Secondary CSO → Etiologies :

1. **metabolic (rickets,hyperthyroidism…),**
2. **toxic (drugs such as phenytoin, valproate, methotrexate…),**
3. **hematologic (sickle cell, thalassemia…)**
4. **structural (lack of brain growth due to e.g. microcephaly, lissencephaly,micropolygyria…).**

CSO is rarely associated with hydrocephalus (HCP).

CLINICAL PRESENTATION

1. craniofacial cosmetic deformities
2. increase ICP

Evidence of increased ICP in the newborn with craniosynostosis include:

1. radiographic signs (on plain skull X-ray or CT)

2: failure of calvarial growth (unlike the non-synostotic skull where increased ICP causes macrocrania in the newborn, here it is the synostosis that causes the increased ICP and lack of skull growth)

3. papilledema

4. developmental delay

How to diagnose?

01

Clinical Examination

02

X-ray

03

CT Scan

04

MRI

05

technetium bone scan

1) clinical Examination

1) palpation of a bony prominence over the suspected synostotic suture (exception: lambdoidal synostosis may produce a trough)

2) gentle firm pressure with the thumbs fails to cause relative movement of the bones on either side of the sutur

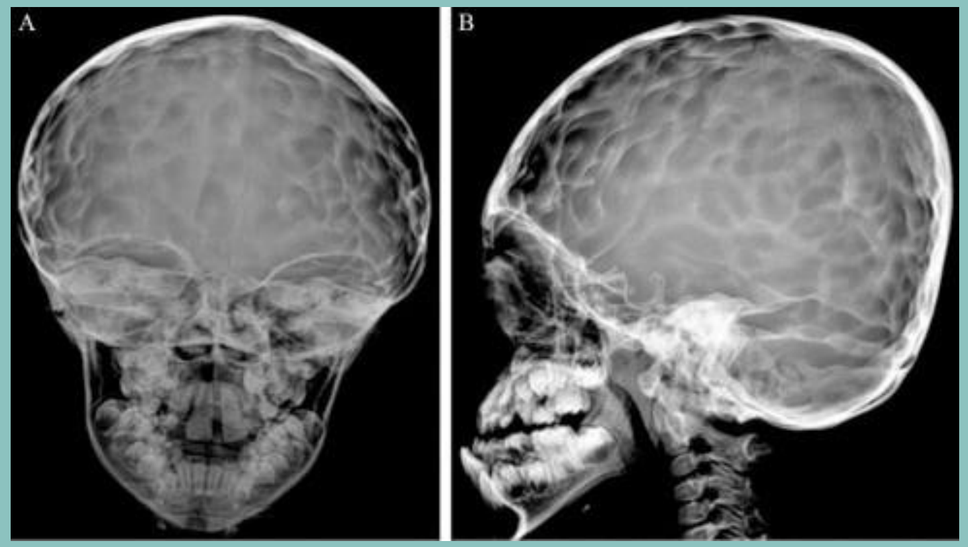
3) measurements, such as occipito-frontal-circumference may not be abnormal even in the face of a deformed skull shap

2) X-RAY

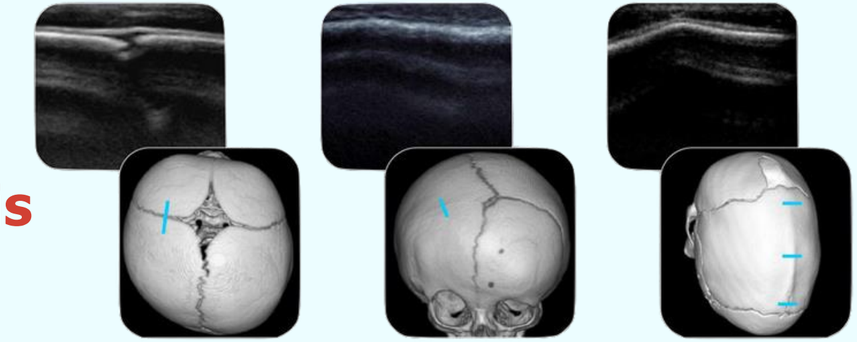
1) lack of normal lucency in center of suture.

Some cases with normal X-ray appearance of the suture (even on CT) may be due to focal bony spicule formation

2) beaten copper calvaria sutural diastasis and erosion of the sella may be seen in cases of increased ICP



3) CT Scan



1)helps demonstrate cranial contour
2) may show thickening and/or ridging at the site of synostosis
3) will demonstrate hydrocephalus if present
4) may show expansion of the frontal subarachnoid space
5) three-dimensional CT may help better visualize abnormalities

4)MRI rarely needed, only if brain/soft tissue anomalies suspected

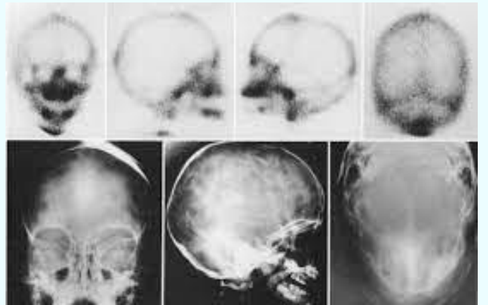
5) Technetium bone scan

Purpose:

Detects areas of increased osteoblastic activity → active bone formation at fused sutures.

Finding:

1)There is little isotope uptake by any of the cranial sutures in the first weeks of life
2) in prematurely closing sutures, increased activity compared to The other (normal) sutures will be demonstrated
3) in completely closed sutures, no uptake will be demonstrated



Treatment

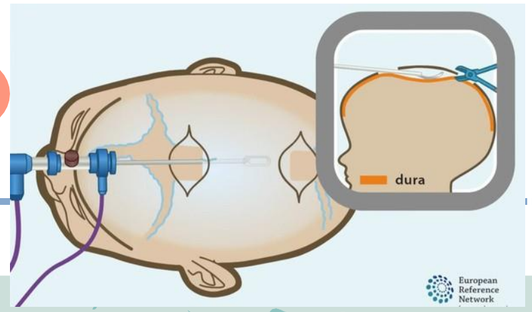
Surgery is the mainstay → to correct skull shape, prevent intracranial hypertension, allow normal brain growth

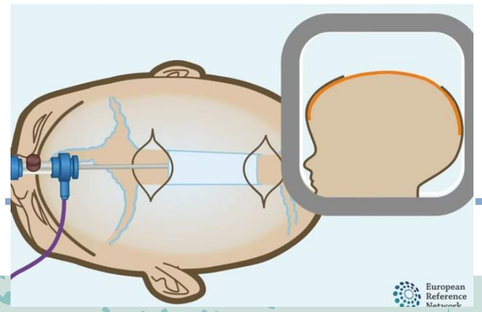
Timing : usually 6–12 months of age for single-suture cases; earlier if syndromic or multiple sutures.

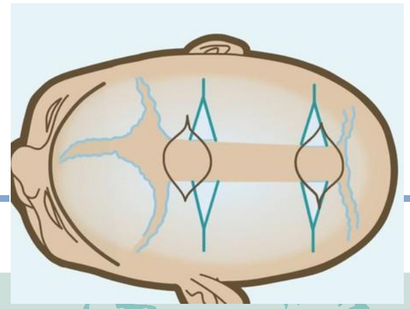
Surgical Options:

Cranial vault remodeling: open surgery → reshapes the skull, often used for complex or multiple sutures.

Endoscopic-assisted suturectomy: minimally invasive → best for single-suture craniosynostosis, typically followed by helmet therapy (best before 6 months)







Types of craniosynostosis

Diagnosis	Shape	Clinical features	Illustrative patient photographs
Sagittal synostosis 	Scaphocephaly Boat-shaped head	Frontal bossing Occipital bullet Biparietal narrowing Sagittal ridge Anteriorly positioned vertex Triangular anterior fontanelle Large head circumference	
Metopic synostosis 	Trigonocephaly Triangle-shaped head	Narrow forehead Bitemporal flattening Midline forehead vertical prominent ridge Biparietal widening Triangular anterior fontanelle	
Unicoronal synostosis 	Frontal plagiocephaly Oblique at front Trapezium-shaped head	Flattened forehead and brow ipsilaterally Ipsilateral wider eye Fullness to contralateral forehead Facial features curving away from affected side Shortened distance ear to lateral orbit on affected side	
Unilambdoid synostosis 	Posterior plagiocephaly Oblique at back Trapezium-shaped head	Flattening posterior unilaterally Mastoid bulging ipsilaterally Ear displaced posteriorly or inferiorly Wind-swept appearance (as if the skull has been blown over to the contralateral side)	
Positional plagiocephaly (no synostosis) 	Oblique front and back Parallelogram-shaped head	Flattening posteriorly unilaterally Corresponding ipsilateral frontal fullness Corresponding ipsilateral ear anteriorly displaced Parallelogram-shaped head No mastoid bulging	

1) Sagittal Synostosis (Scaphocephaly / Dolichocephaly)

Most common form of single-suture craniosynostosis

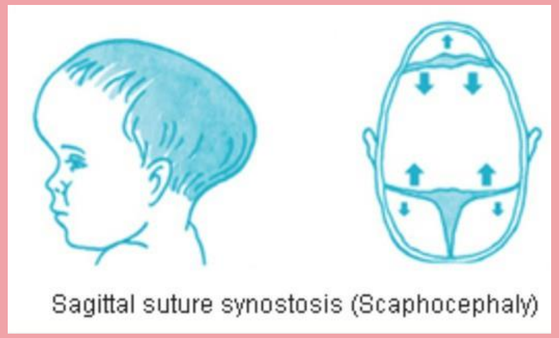
Sex ratio: about 80% male

Clinical features:

1) Palpable midline bony ridge along the sagittal suture.

2) Dolichocephaly: elongated, narrow skull with high forehead/frontal bossing.

3) Scaphocephaly: "boat-shaped skull" with prominent occiput.

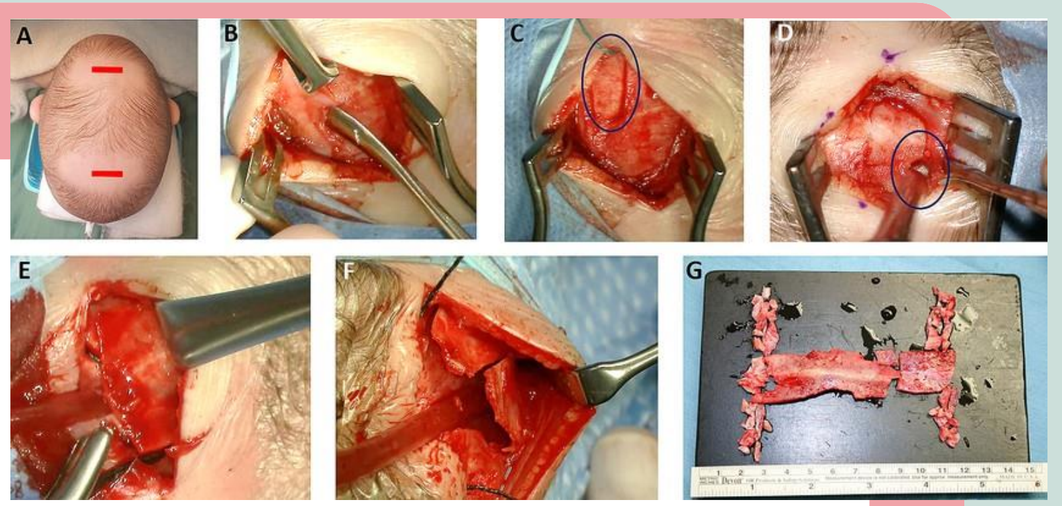

Sagittal suture synostosis (Scaphocephaly)

4) OFC (head circumference): usually normal.

5) Biparietal diameter: significantly reduced.

6) Intracranial pressure (ICP): elevated in up to 44% of nonsyndromic cases

OPEN SURGICAL TREATMENT



Procedure

Skin incision: longitudinal or transverse

Strip craniectomy → excision of the sagittal suture from coronal to lambdoid sutures. (Strip width should be at least 3 cm)

Ideal age:

surgery performed between 6 months and 1year of age (preferably early).

Precautions:

Avoid dural laceration or injury to the superior sagittal sinus

Follow-up:

- If re-fusion occurs before 6 months → reoperation may be necessary.
- After 1 year → usually requires cranial vault remodeling (more extensive surgery).

Note:

No evidence that placing artificial barriers (e.g., silastic sheeting) reduces recurrence.

2) Coronal Synostosis:

Represents about 18% of craniosynostosis (CSO)

More common in females

Can be unilateral or bilateral.

1) Unilateral Coronal Synostosis → Anterior Plagiocephaly

Forehead: flattened or even concave above the affected orbit.

Contralateral forehead: compensatory bossing (prominent)

Orbit: Supra-orbital rim higher on the affected side → Harlequin eye sign on X-ray/CT.

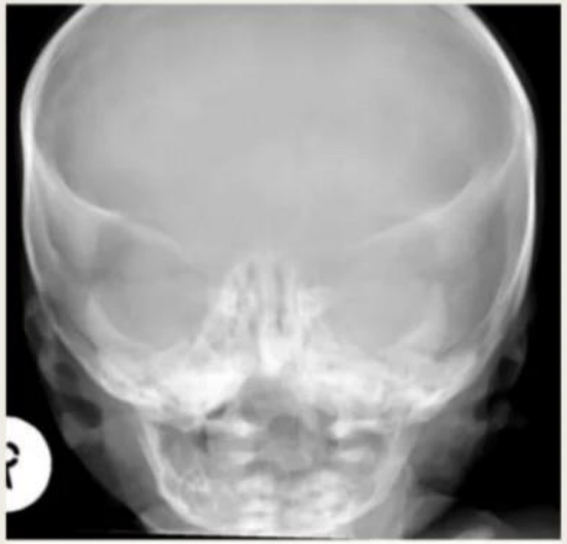
Orbit rotates outwards → risk of amblyopia (lazy eye)

Face:

Flattened cheek on affected side.

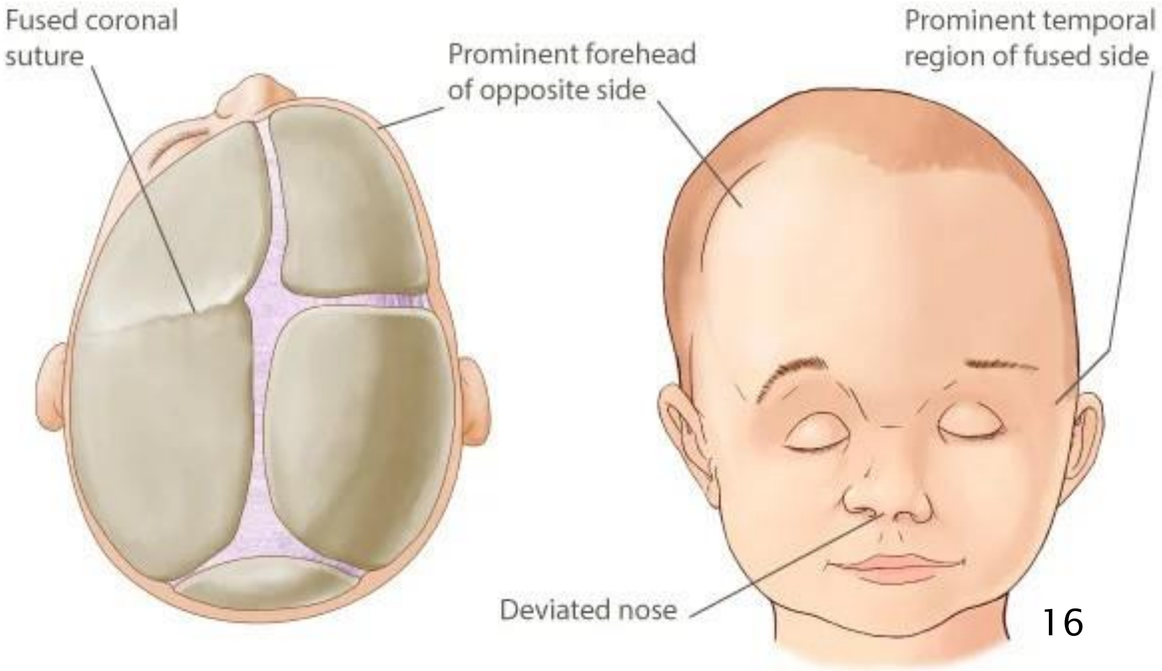
Nose deviates toward the normal side (root of nose rotates toward the deformity).

HARLEQUIN EYE SIGN



- The harlequin eye deformity may be seen in unilateral or bilateral coronal suture synostosis, and refers to the elevation of the superolateral corner of the orbit.

Unicoronal craniosynostosis



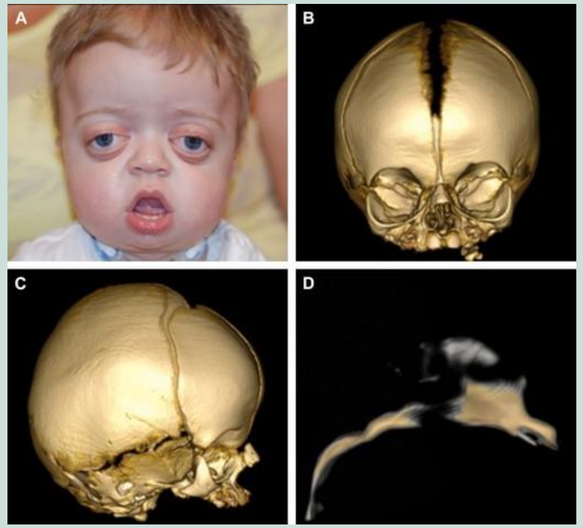
2) Bilateral Coronal Synostosis

- Often seen in syndromic cases (e.g., Apert syndrome)

- Produces brachycephaly:
  Broad, short head with flattened forehead.

- If severe → acrocephaly (tower skull)

- When associated with premature closure of frontosphenoidal + frontoethmoidal sutures → Foreshortened anterior cranial fossa Lead TO:



1) Midface hypoplasia (maxillary hypoplasia).
2) Shallow orbits → progressive proptosis.

Surgical Management in Coronal Synostosis

1.Simple Strip Craniectomy

- Removal of a narrow strip of bone along the affected suture
- Often produces excellent cosmetic results
- May be insufficient in some cases, especially if:

1) Synostosis is severe or multi-sutural.
2) Significant orbital or facial deformities are presen

2. Current Recommendation

- Frontal craniotomy (uni- or bilateral) → more extensive skull reshaping.
- Lateral canthal advancement:

Elevates the outer corner of the eye to improve orbital aesthetics.

- Orbital bar removal and remodeling:

Removal and reshaping of the orbital bar to correct anterior skull and facial contour.

Lambdoid synostosis:

Rare: 1–9% of craniosynostosis cases.

More common in males (4:1).

Right side affected in ~70% of cases.

Usually presents between 3–18 months, sometimes as early as 1–2 months.

Clinical Feature : Flattening of the occiput – can be unilateral or bilateral

Unilateral: → Posterior Plagiocephaly

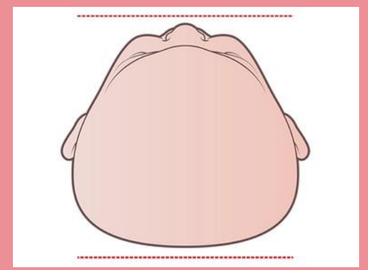
Ipsilateral ear displaced anteriorly and inferiorly.

Ipsilateral cheek may appear slightly bulging.

Indentation along the affected lambdoid suture (sometimes a ridge).

Bilateral: → Brachycephaly (broad, short head).

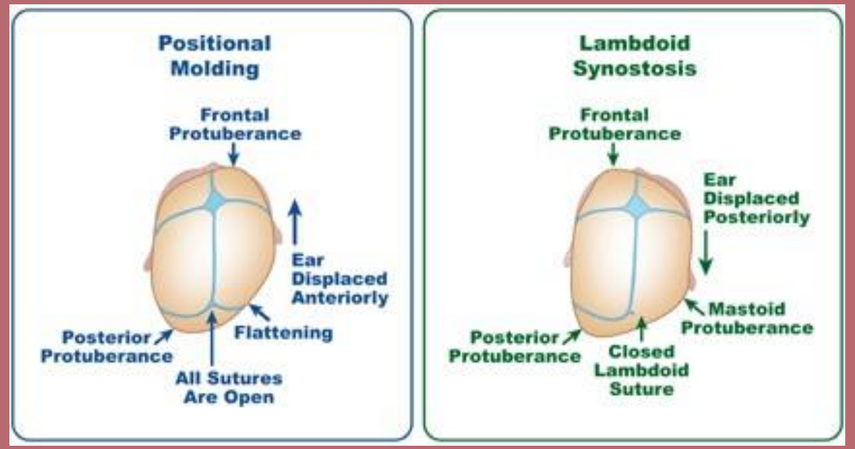
with both ears displaced anteriorly and Inferiorly



- Clinical Palpation
  - In sagittal or coronal synostosis → a palpable bony ridge is usually felt along the fused suture.
  - In lambdoid synostosis → instead of a ridge, there is often an indentation along the fused lambdoid suture.

Mimic: "Lazy Lambdoid" (Positional Flattening)

Definition: A condition that mimics lambdoid synostosis but is actually due to positional molding of the skull.



Features:

- **Flattening of the occiput on one side.**
- **Ipsilateral ear displaced anteriorly.**
- **Bulging of the ipsilateral cheek and forehead.**

Causes of Positional Flattening

1)Decreased mobility :Infants who lie supine with the head turned to one side.

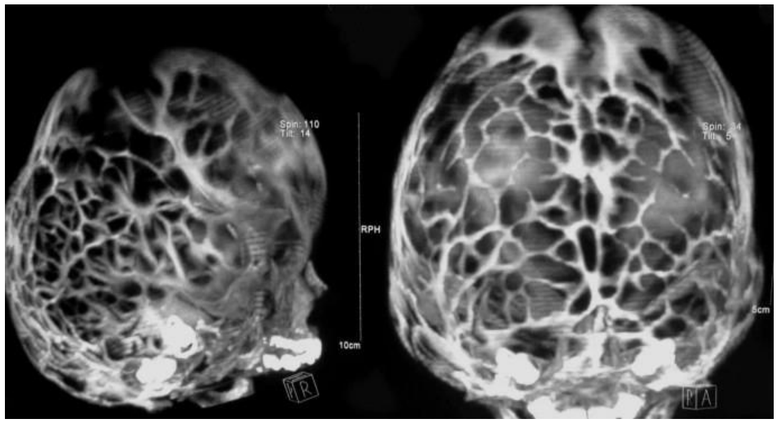
Examples: cerebral palsy, developmental delay, prematurity, chronic illness.

2) Abnormal postures :Congenital cervical spine disorders.

3) Intrauterine crowding (e.g., multiple gestation, large fetus) ,Uterine anomalies

4) Intentional positioning : sometimes worsened by using a foam wedge to tilt the child.

Diagnostic Evaluation



1) Physical exam: most important.

2) Skull X-ray:
   - Sclerotic margin along affected lambdoid suture (~70%).
   - May show beaten copper cranium (BCC) from localized increased ICP.

3) CT scan:
   - Bone windows: shows closed or thinned suture.
   - Brain windows: may reveal associated parenchymal abnormalities.

4) Bone scan:
   - Increased isotope uptake in the suture during the first year, peaking around 3 months.

TREATMENT :

Better results with earlier surgery

Indications for Early Surgical Treatment:

1)Severe craniofacial disfigurement.

2) Evidence of increased intracranial pressure (ICP).

Surgical treatment

- **Required in only ≈ 20% of cases.**
- **Ideal age: 6–18 months.**
- **Options:**
  - **Simple unilateral craniectomy of the suture.**
  - **More elaborate reconstruction by a craniofacial team.**

TREATMENT :

Non-surgical treatment

- **Observation:**
  - **Duration: 3–6 months.**
  - **Majority remain static or improve with time + simple nonsurgical interventions.**
  - **~15% continue to develop significant cosmetic deformity.**

- **Repositioning (effective in ≈ 85%):**
  - **Place infants on unaffected side or abdomen.**

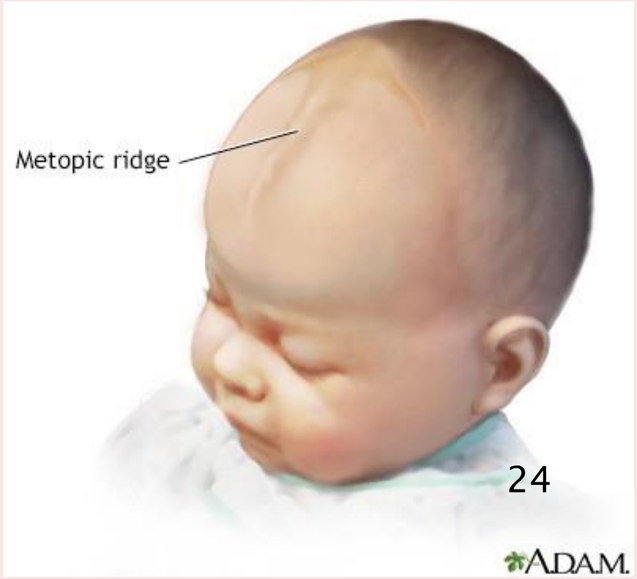
- **In occipital flattening from torticollis → aggressive physical therapy.**
  - **Resolution usually within 3–6 months.**

- **Molding Helmets:**
  - **May be tried in more severe involvement.**
  - **No controlled studies proving efficacy.**

Metopic synostosis

At birth, the frontal bone consists of two halves separated by the frontal or metopic suture.
Abnormal closure produces trigonocephaly (pointed or triangular shaped) forehead with a midline ridge and hypotelorism.

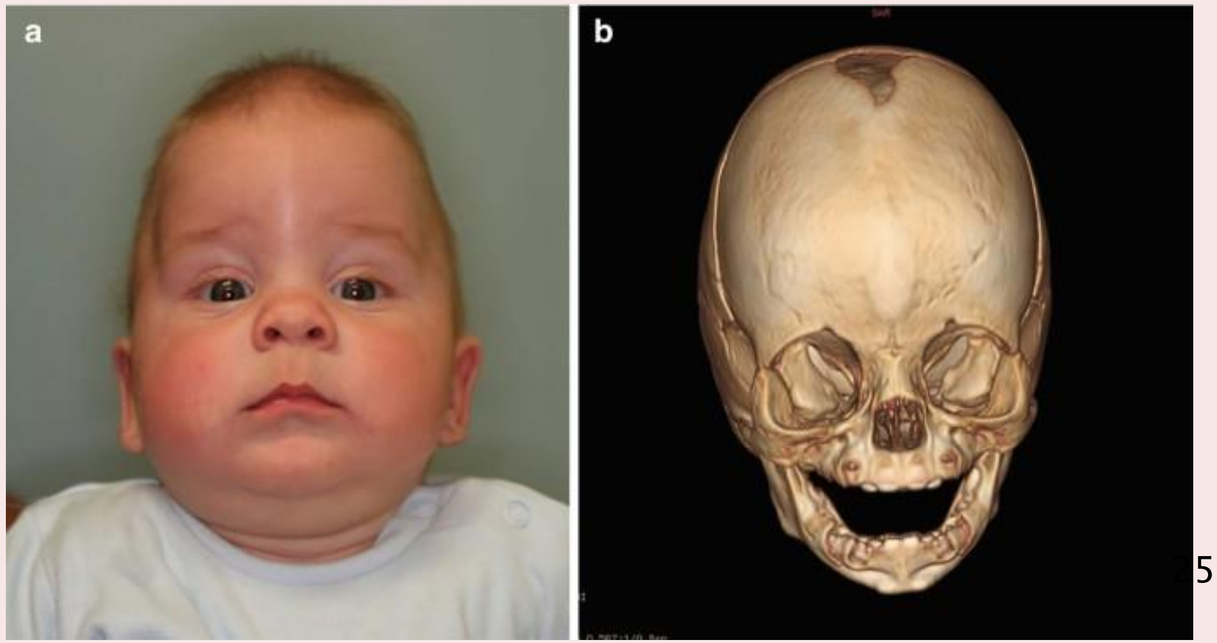
- **Incidence:** 1/15,000 live births.
- 75% are male.
- Many of these have a 19p chromosome abnormality and are mentally retarded.



Multiple synostoses

Fusion of many or all cranial sutures → **oxycephaly**
(tower skull with undeveloped sinuses and shallow orbits).

These patients have elevated ICP.



CRANIOFACIAL DYSMORPHIC SYNDROMES

Over 50 syndromes have been described ,number of craniosynostosis syndromes are due to mutations in the FGFR (fibroblast growth factor receptor) genes.

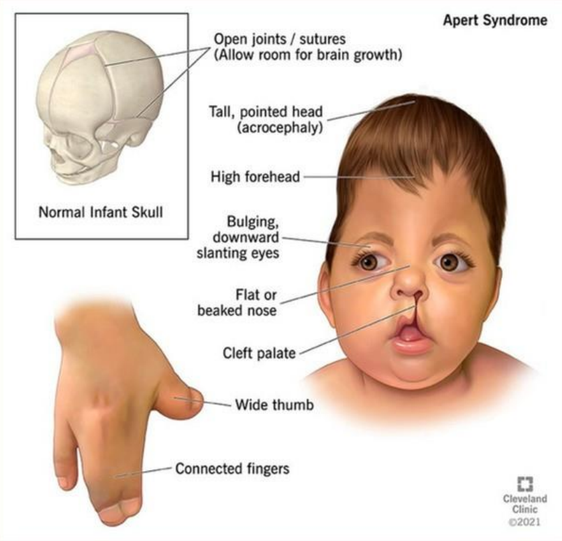
FGFR gene-related craniosynostosis syndromes include some classic syndromes (Apert, Crouzon, Pteiffer…)

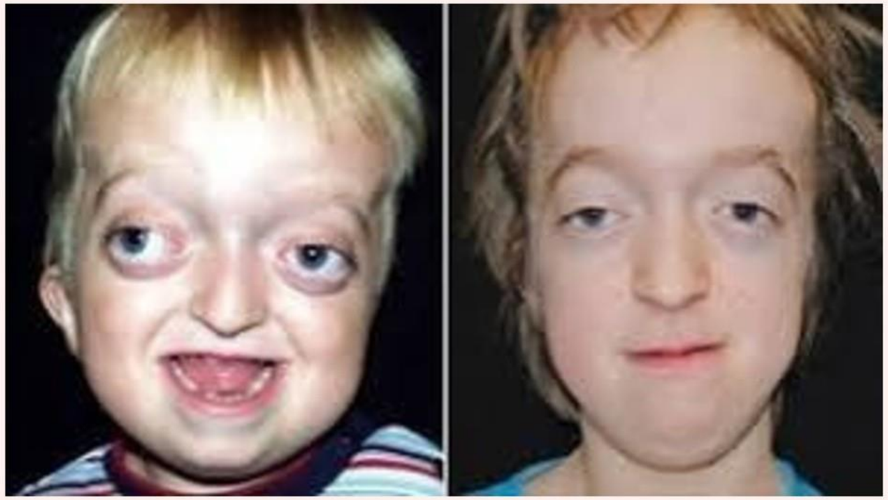
as well as several newer entities:

(BeareStevenson,Muenke,JacksonWeiss syndromes).

All exhibit autosomal dominant inheritance.

Syndrome	Genetics		Craniofacial findings	Associated findings
	Sporadic	Inherited		
Crouzon (craniofacial dysostosis)	yes (25%)	FGFR AD	CSO of coronal & basal skull sutures, maxillary hypoplasia, shallow orbits, proptosis	HCP rare
Apert (acrocephalosyndactyly)	yes (95%)	FGFR AD	same as Crouzon	syndactyly of digits 2,3,4; shortened UE, HCP common





Development of the Neural Tube

3 stages of the neurodevelopment :

1 **Neurulation** (making the Neural tube and separating from ectoderm)
2 **Neuronal proliferation** (Neuronal cells formation and multiplication)
3 **Neuronal Migration**

Begins at the 3rd week of gestation

originates from cranial end of embryo as thickening of ectoderm on either side sides of the midline

This thickening forms Neural plate

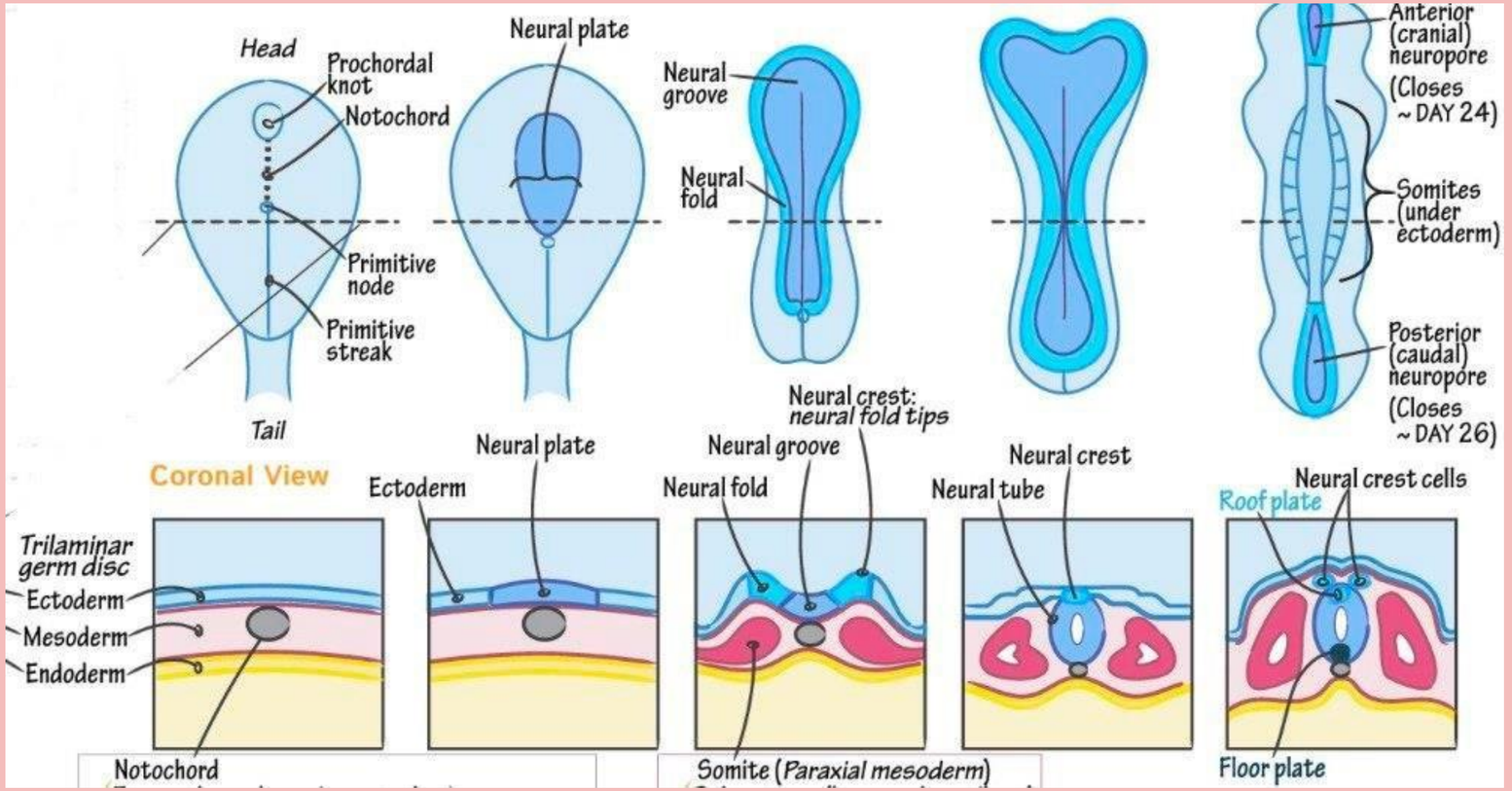
in the 4th week ,Neural folds form as a neural plate indentats and thickens in mesoderm

Then neural folds bend upwards forming neural tube with the Neural crest posterolateral and the Primitive Notochord Anterior to it

Neural tube forming : Brain and spinal cord
Neural crest forming : peripheral nerves, roots & ganglia of autonomic nervous system
primitive notocord forming: intervertebral disc

- As the neural tube closes, the neuroectoderm (which will form the CNS) separates from the cutaneous ectoderm in a process known as disjunction.
- Upon completion of disjunction, the cutaneous ectoderm fuses in the midline, dorsal to the closed neural tube.
- Neural tube closure probably begins at two or three levels in the middle of the embryo.
- Closure proceeds bidirectionally in a zipper-like fashion along the length of embryo.
- The cephalic and caudal ends of the neural tube (the so-called anterior and posterior neuropores) do not fuse until the twenty-fifth and twenty-seventh gestational days, respectively.

Neural crest cells : ectodermal cells that form during folding of neural plate , after the tube closes the neural crest cells detach (delaminate)

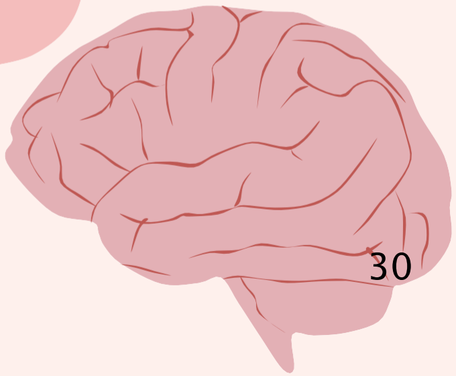


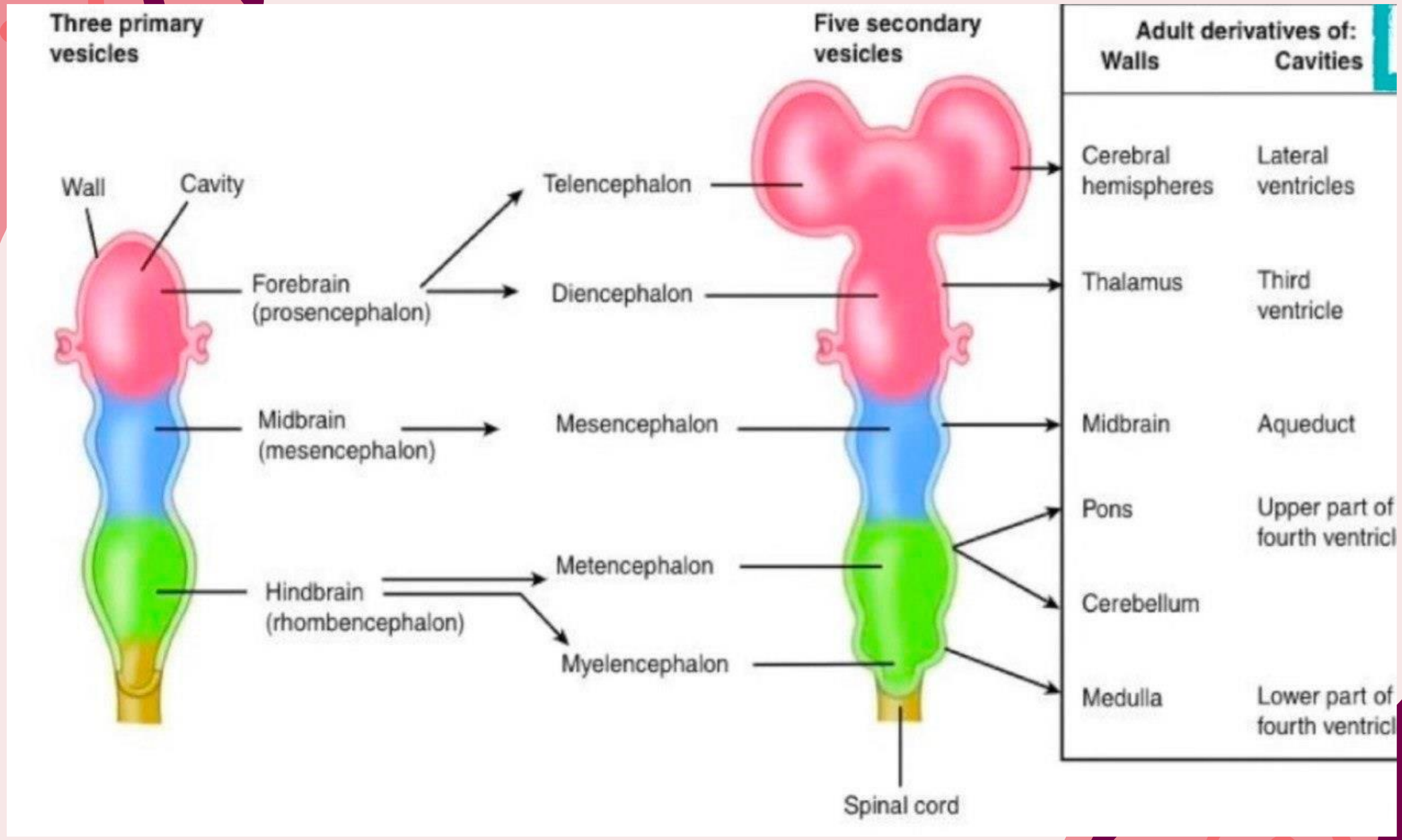
- **The Cephalic end of the Neural tube forms Forebrain and much of Midbrain while Caudal end of Neural tube forms Hindbrain**

- **This whole process is called Neurulation , any defect in this process causes Neurulation defects and any defect after this procedure is called Post-Neurulation defect**

- **Neural tube divides into 3 vesicles , and at the 4th week the embryonic brain grows rapidly and begins to bend, forming several flexures.**

- **at the 5th week each vesicle tends to divide to form its structures**




Three primary vesicles
Five secondary vesicles
Adult derivatives of:
Walls
Cavities
Wall
Cavity
Forebrain (prosencephalon)
Midbrain (mesencephalon)
Hindbrain (rhombencephalon)
Telencephalon
Diencephalon
Mesencephalon
Metencephalon
Myelencephalon
Spinal cord
Cerebral hemispheres
Lateral ventricles
Thalamus
Third ventricle
Midbrain
Aqueduct
Pons
Upper part of fourth ventricl
Cerebellum
Medulla
Lower part of fourth ventricl

Anencephaly

Anencephaly

- It is a serious NTD Caused by failure of closure of **cranial** neuropore in which:
  - -cranial vault are grossly malformed
  - the brain (cerebral hemispheres) is absent
  - the cerebellum is reduced/ absent
  - the hindbrain is present

* Leading to early fetal loss, stillbirth, neonatal death after few hours/ days
* baby born with anencephaly is usually blind, deaf, unaware of its surroundings and unable to feel pain

Epidemiology:

- -birth prevalence: 1/10.000
- pregnancy: 1/1000
- F>M
- survival period for such a patient is 5 months
- associated anomalies: cleft lip/palate,omphalocele.

Etiology

- Inadequatefolic acid or antagonists such as: valproic acid, methotrexate, carbamazepine, phenobarbital, trimethoprim
- Maternal diabetes mellitus (type 1)
- Maternal hyperthermia
- Genetics
- Amniotic band syndrome (constricting band syndrome)
- Alcohol consumption

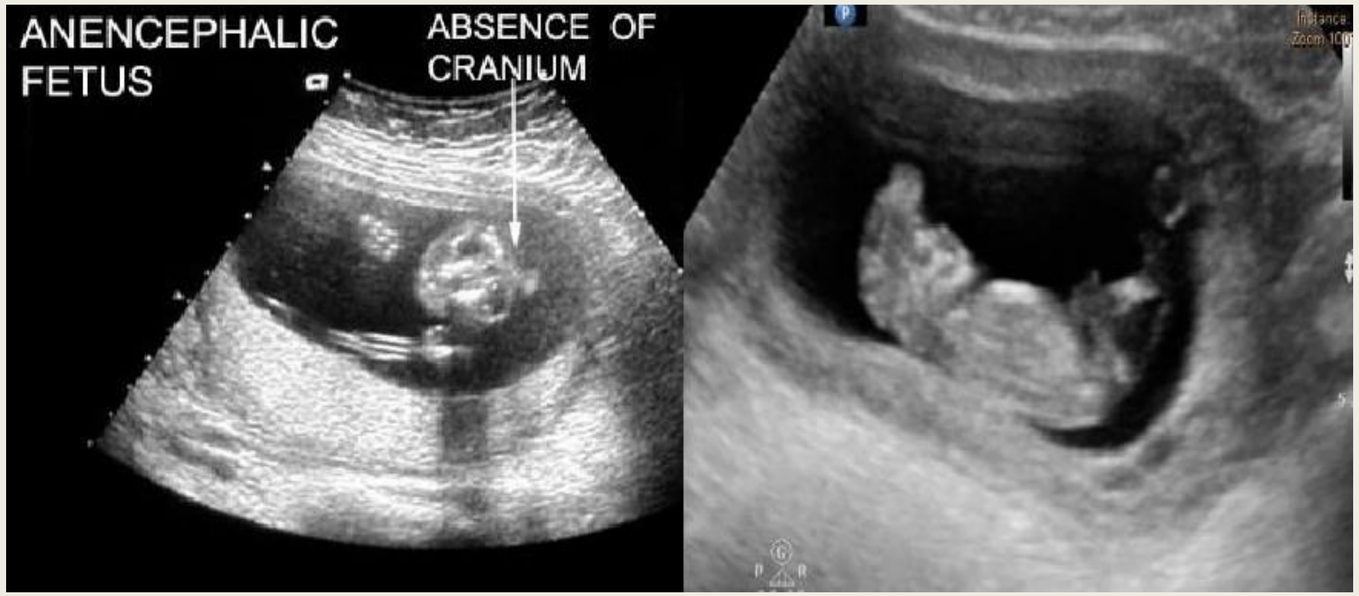
Diagnosis:

Lab studies: mainly during 2nd trimester

- ↑maternal serum AFP
- ↑ AF AFP
- ↑ acetylcholinesterase

Imaging studies: BY US (identified from 13 the week of gestation)

1) absence of brain & calvaria superior to the orbits (on coronal view of fetal head)
2) **↓crown –rump length**
3) **Later on: polyhydramnios**



Prevention:

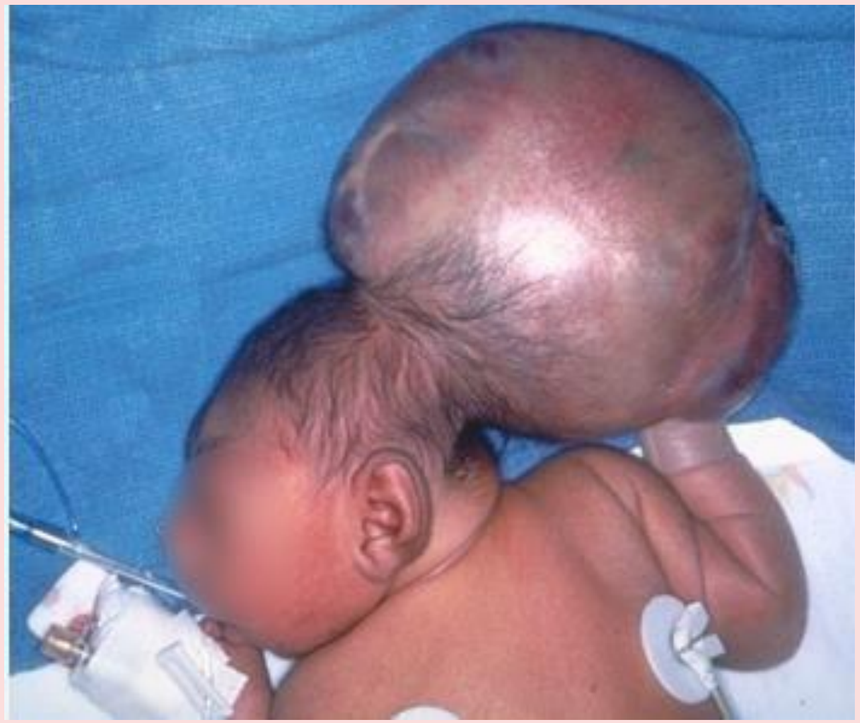
1) 4 mg of folic acid/ daily/ at least 3 months before pregnancy for women:
   - desire pregnancy
   - had child with an NTD
   - taking anticonvulsants
2) 0.4 mg/day of folic acid → all women who are pregnant or may become pregnant
3) Stop the folic acid antagonists at least in the 1st trimester
4) Control glucose level in diabetic patients

Cephalocele

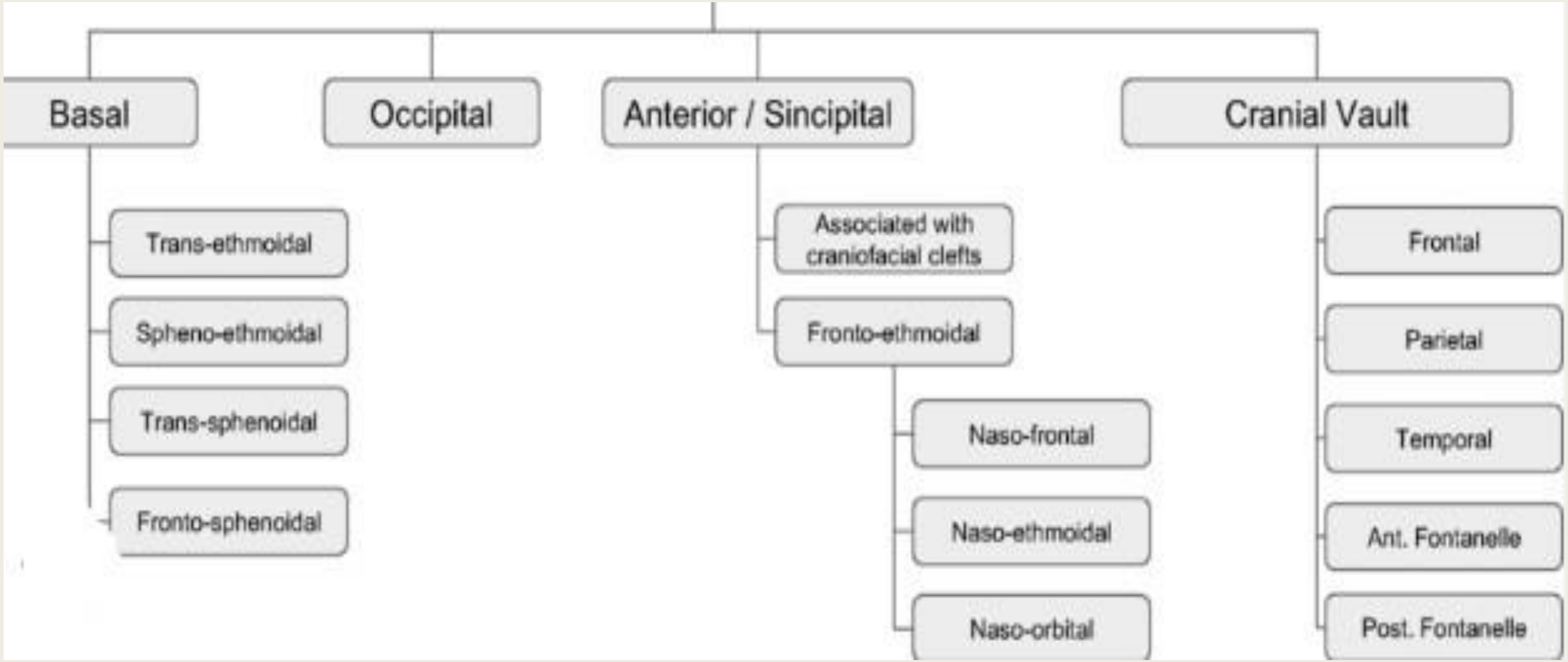
Failure of rostral/ cranial end closure → outwardherniationof CNS contents through a defect in the cranium

May associated with:

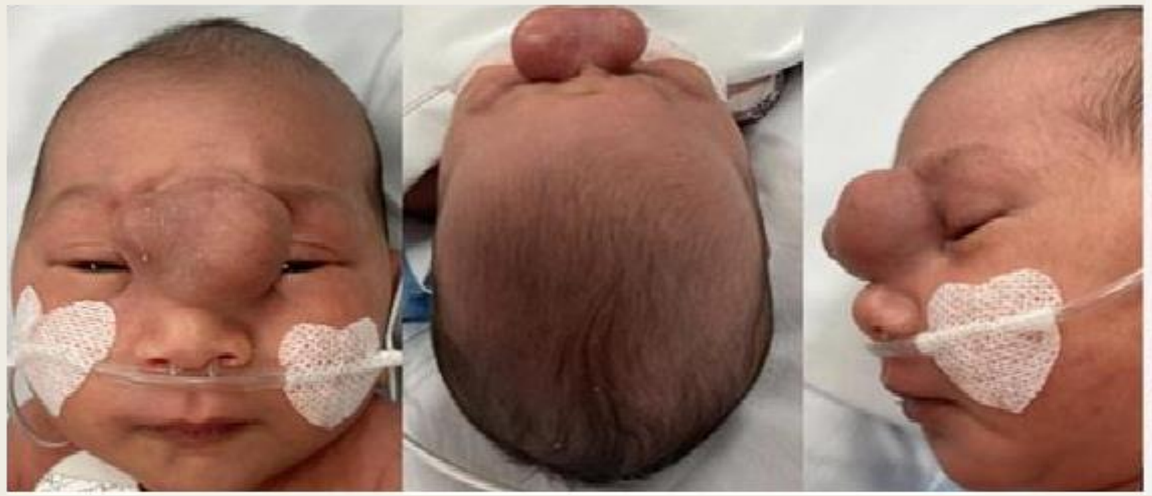
- Trisomy 13,18 Hydrocephalus,
- microcephalus
- Chiari III malformation , Meckel guber syndrome (occipital encephalocele, multiple renal cysts, postaxial polydactyly)



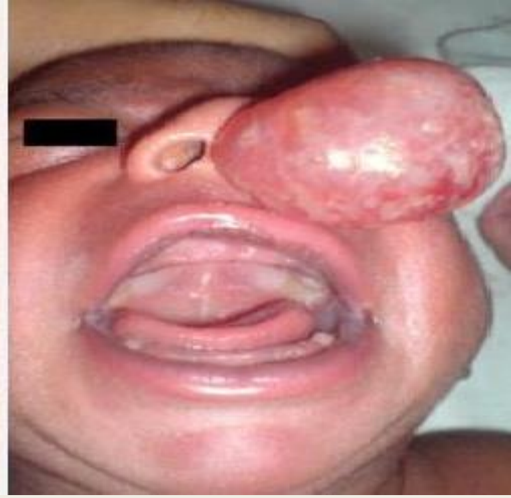
Classification according of location:



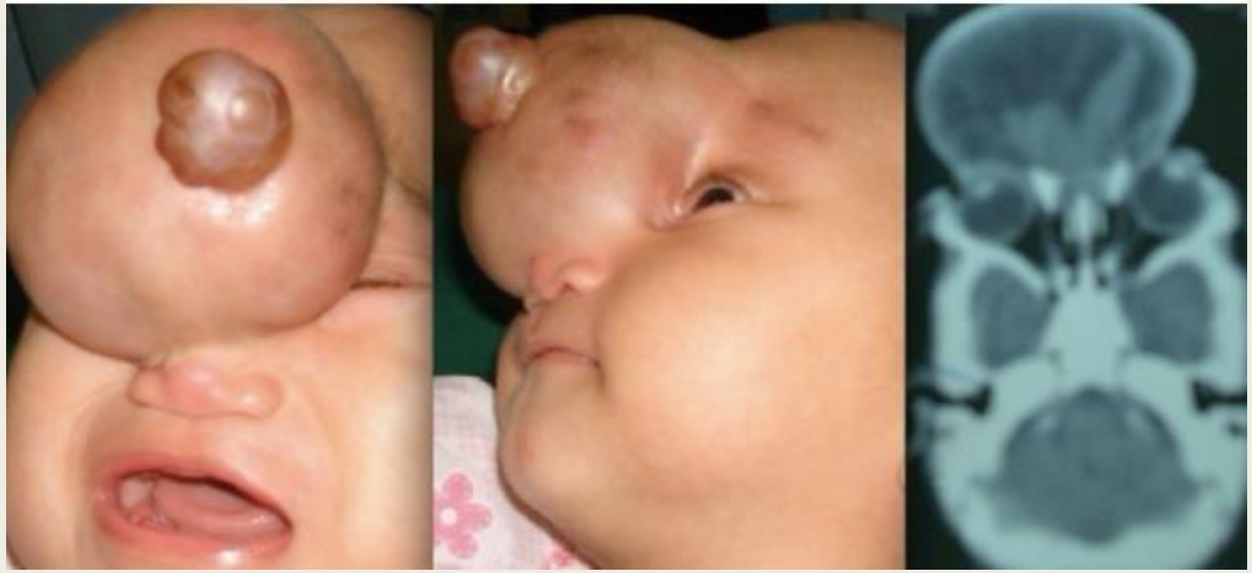
Occipital



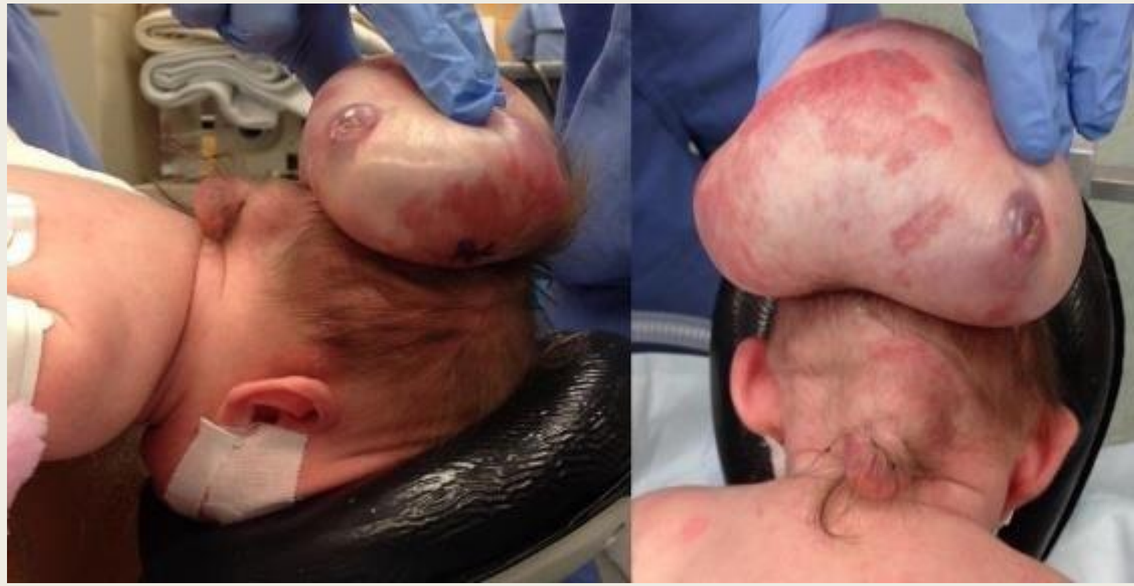
Frontal (nasofrontal)



Sphenoidal

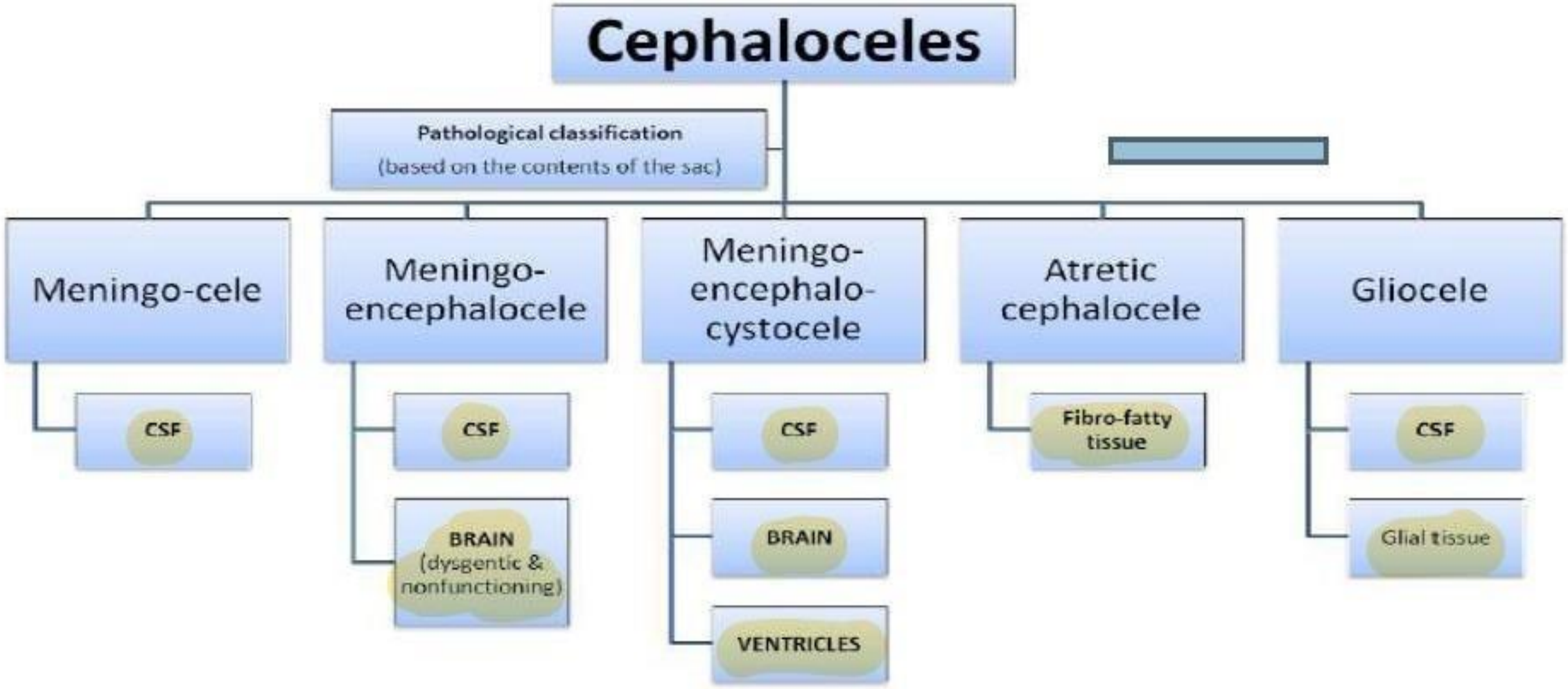


Ethmoidal



parital

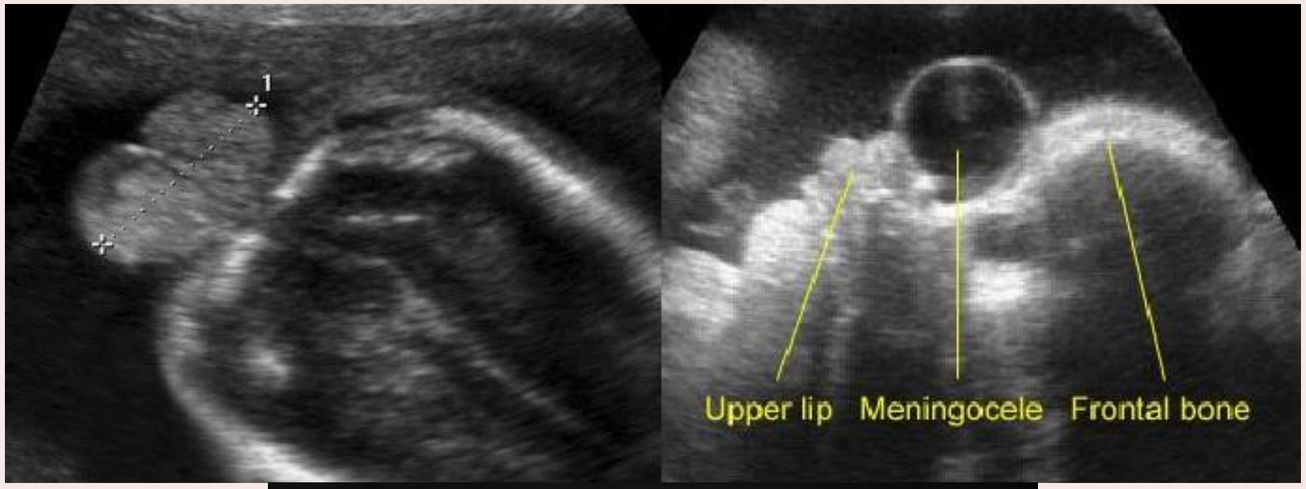
According contents

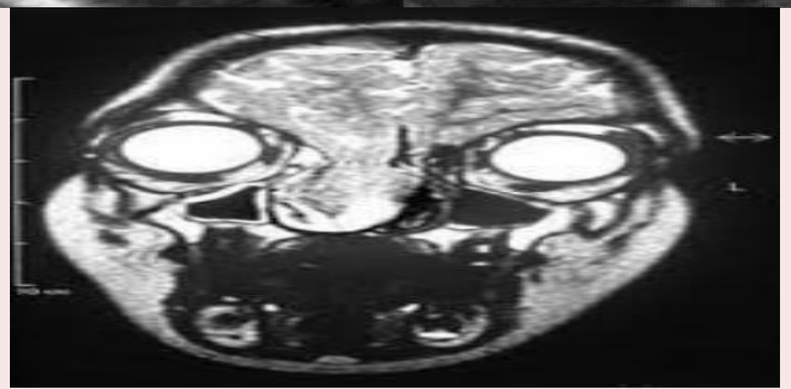


Diagnosis

Markers: ↑ MS AFP

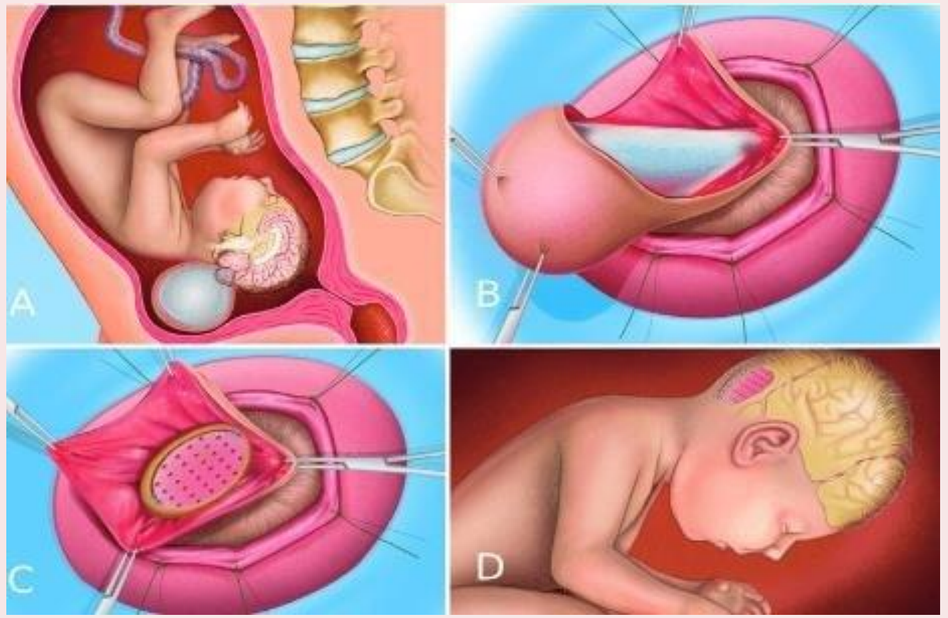
Imaging: US, MRI





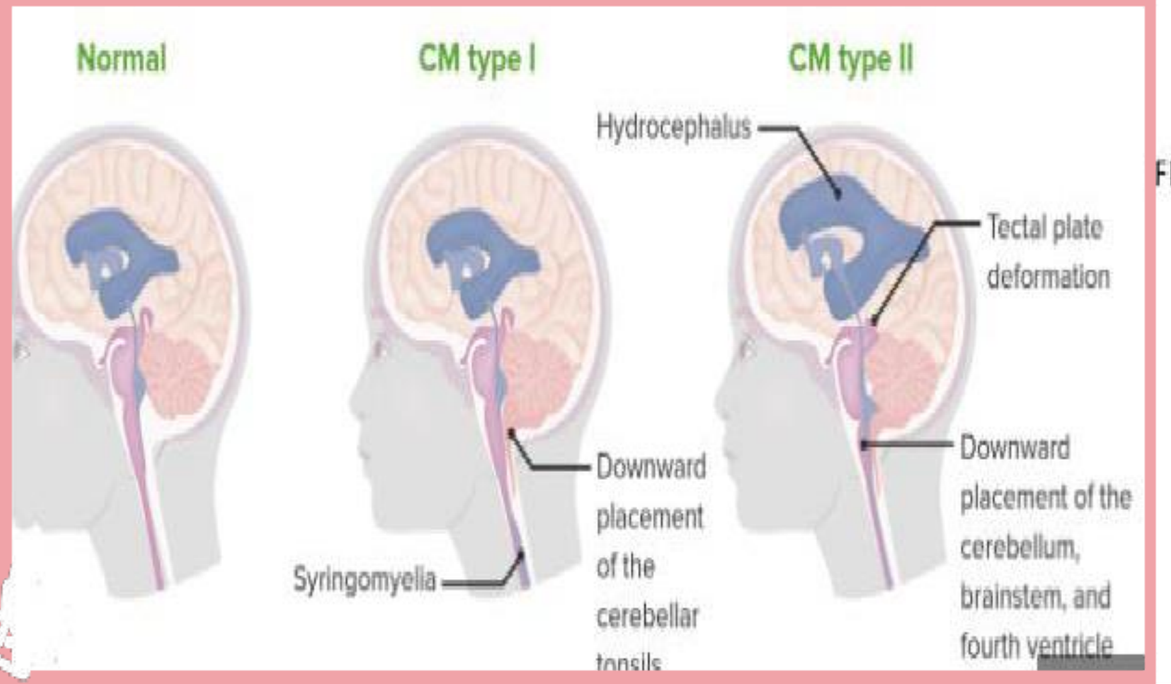
Treatment

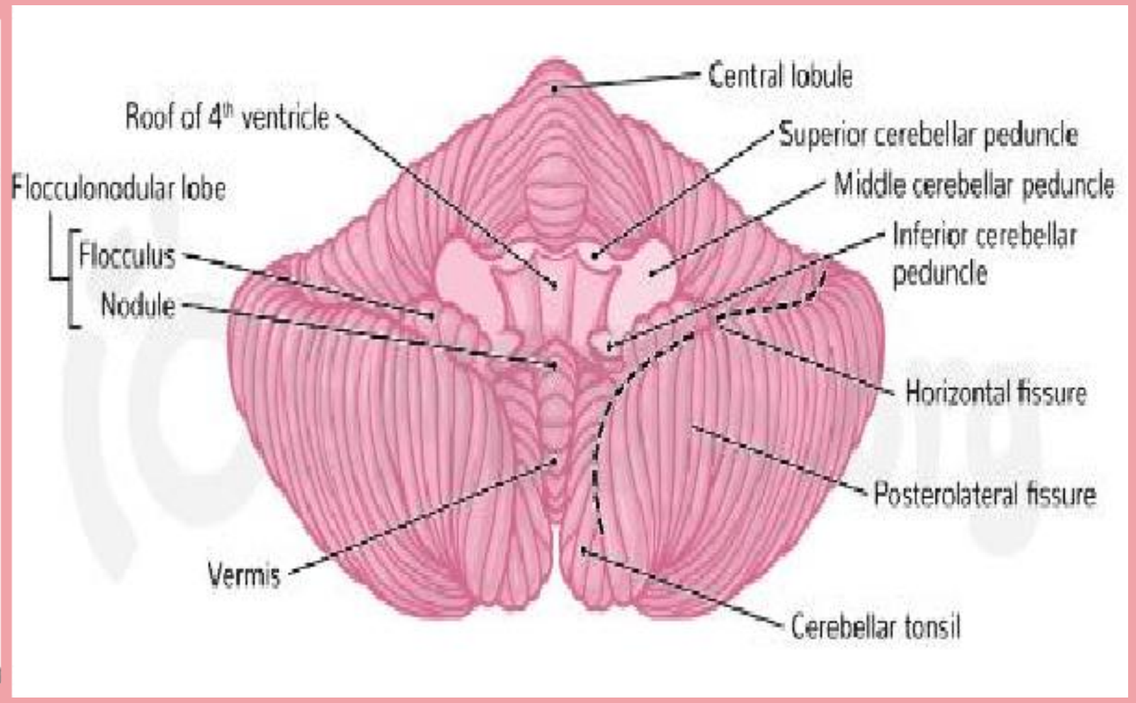
Excision of herniated sac followed by cranioraphy (repair the opening in the skull)



Chiari malformation

Congenital caudal displacement/protrusion of the cerebellum and lower brainstem

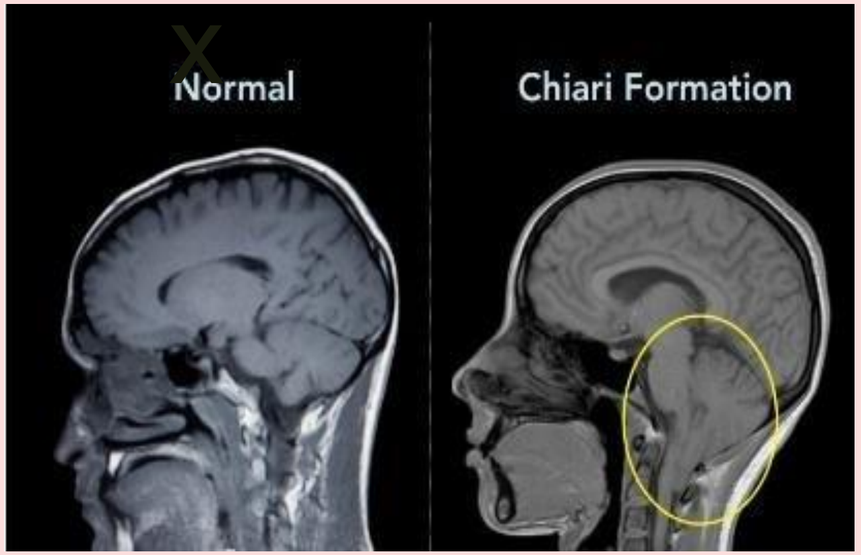


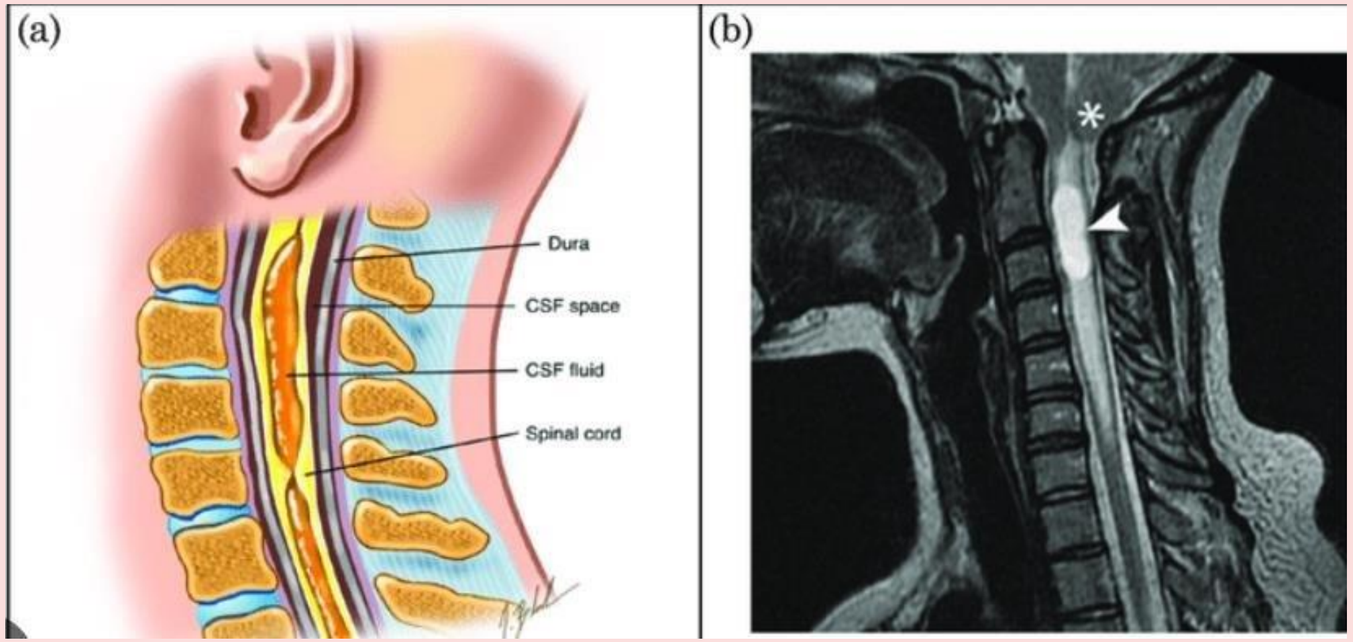


Type I CM	Type II CM	Type III CM	Type IV
- Most common, least severe - CD of cerebellar tonsils(no herniation of brainstem) below the foramen magnum >5mm into upper cervical canal - Maybe the result of formation of a small POST. Fossa → overcrowding→ herniation	- (Arnold – Chiari malformation) - CD of cerebellar vermis, 4th ventricle & medulla oblongata below foramen magnum	- Herniation of parts of cerebellum & brain stem through an abnormal opening in the occipital bone (occipital and/or high cervical encephalocele)	Cerebellar hypoplasia or agenesis
C/P: - Compression of upper cervical→ myelopathy - Compression of cerebellum → ataxia, dysmetria, intential tremor -disruption of CSF flow→↑ICP, suboccipitalheadache, neckpain, vomiting, visual defects, hydrocephalus (<10%) - syringomyelia	C/P: - Signs of brainstem dysfunction: swallowing/ feeding difficulties, stridor, apnea, respiratory depression, nystagmus, hydrocephalus, syringomyelia (less common) - Almost associated with myelomeningocele	Mostly fatal in early infancy	Not compatible with life Mostly associated with other brain anomalies (e.g. agenesis of corpus callosum)
Dx: by MRI (usually in young adulthood	Dx: by MRI (present in infancy)	Tonsillectomy if they reach C5 6	

.Tx: POST. Fossa decompression w/ or W/O Tonsillectomy → fails → secondary shunt duro-rrhaphy

Type I

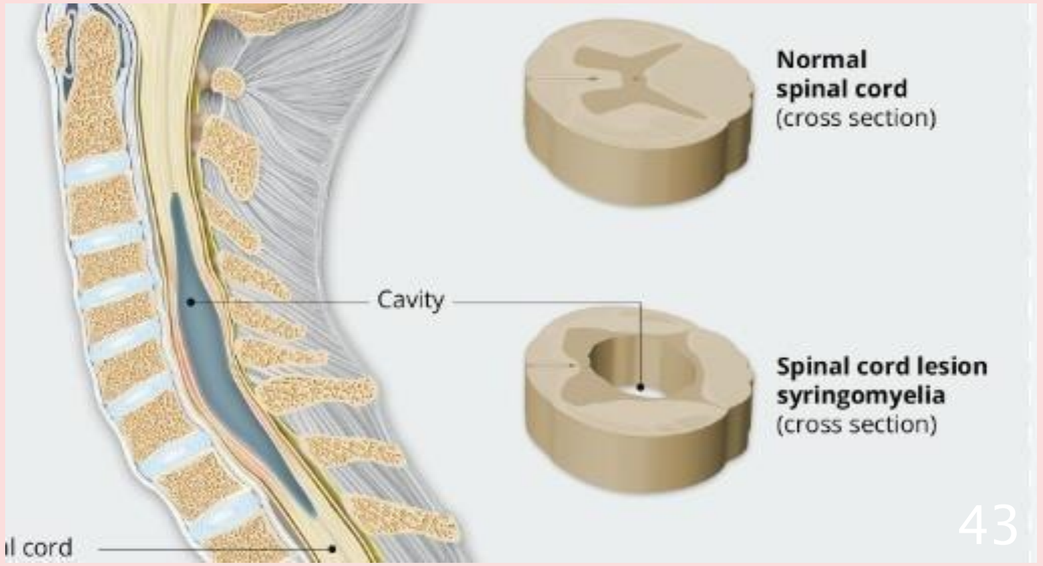


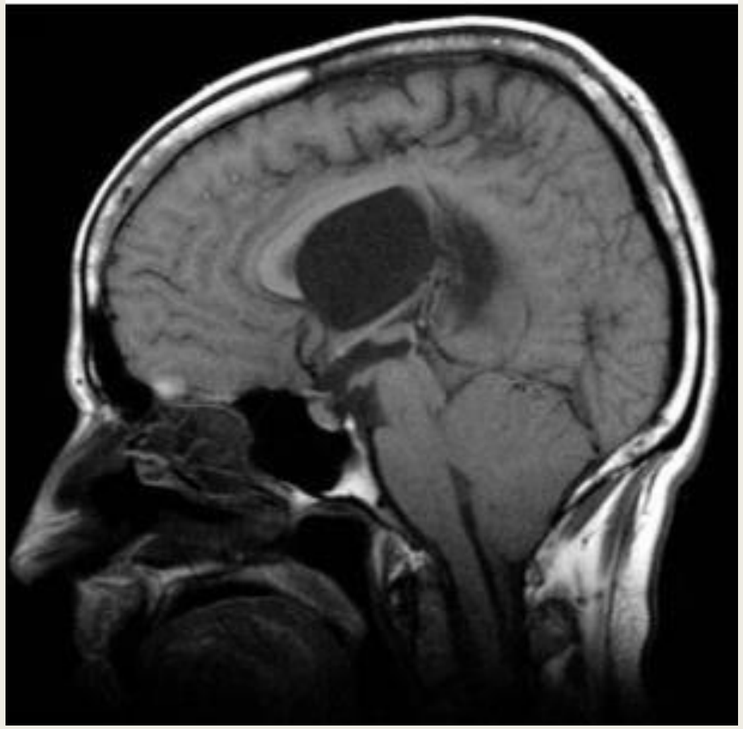


Syringomyelia :
cyst or cavity forms within the spinal cord, this cyst, called a syrinx, can expanded and elongate over time.

Related symptoms:

- cape like loss of pain and temperature sense.
- weakness and wasting of the small muscles of the hand.
- progressive motor deficit of upper & lower limbs.



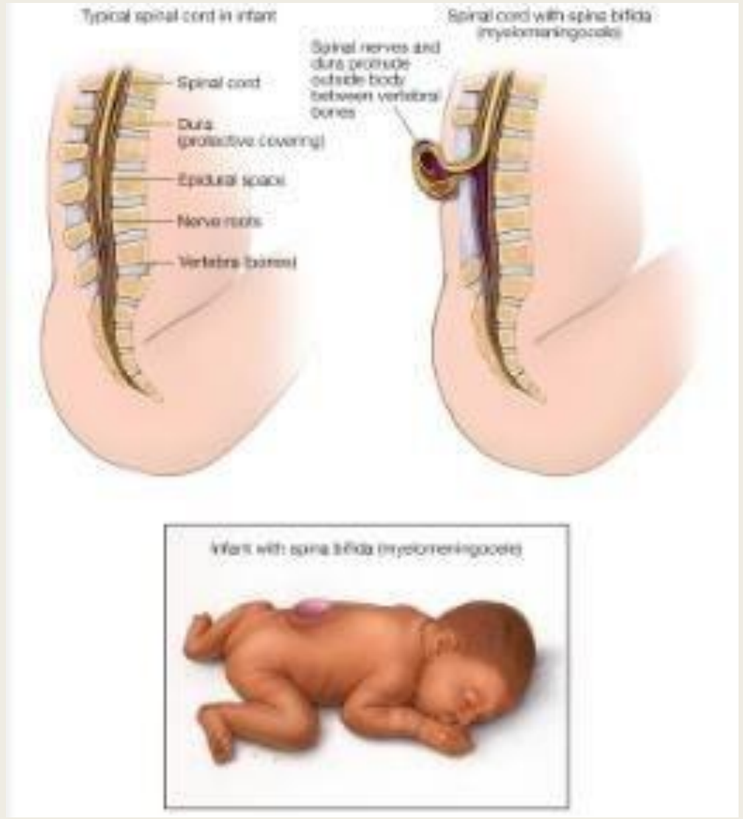


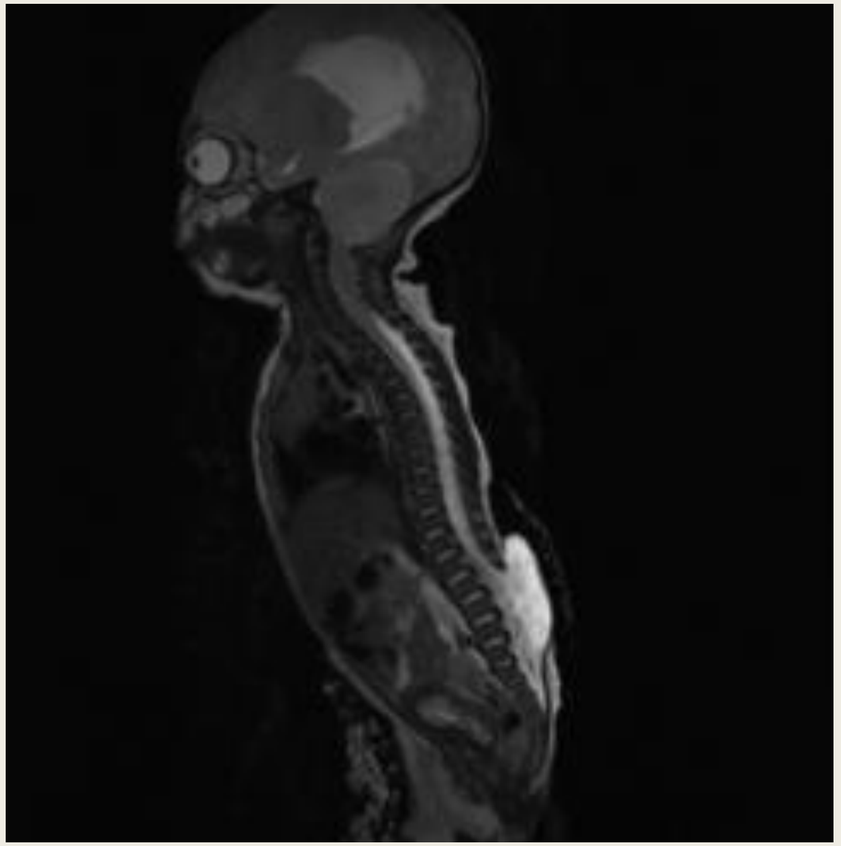
Type II

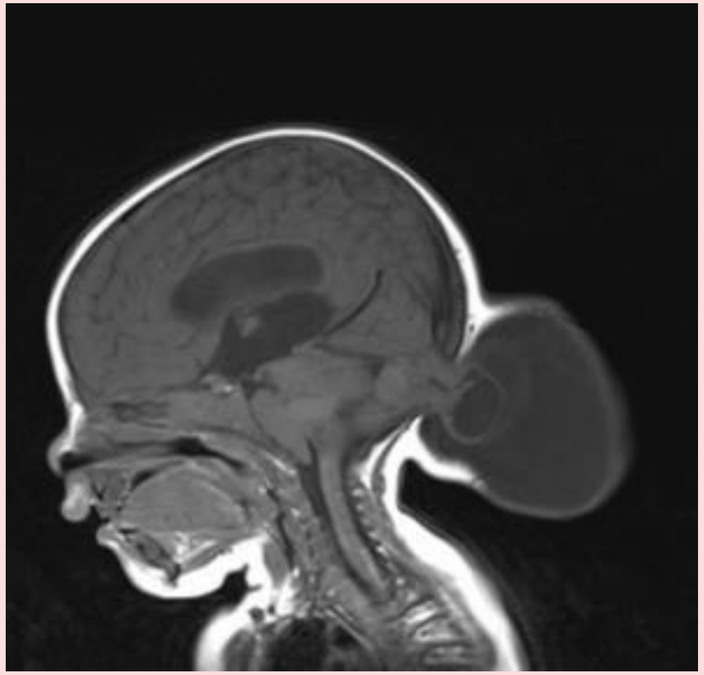
MYELOMENINGEOCELE:

Meninges and parts of the spinal cord herniate through the vertebral bone defect.

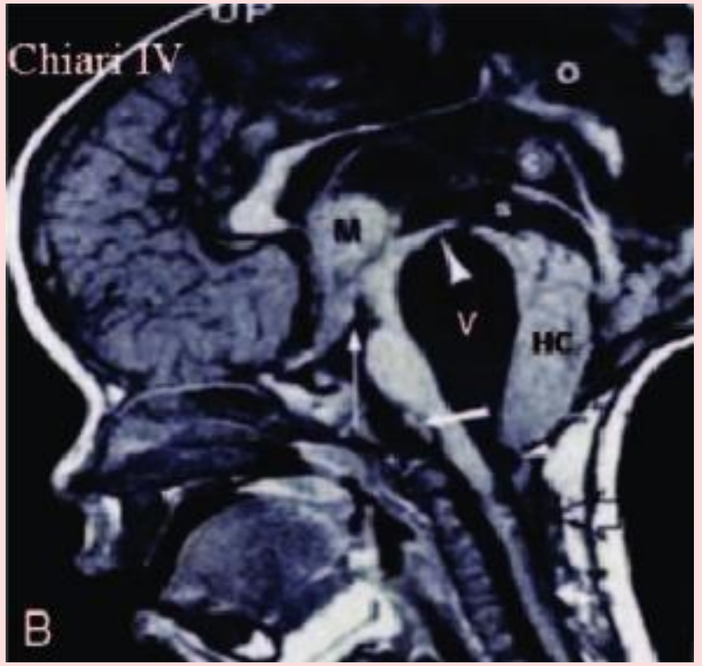
- ALL patients with myelomeningocele have Chiari II malformation



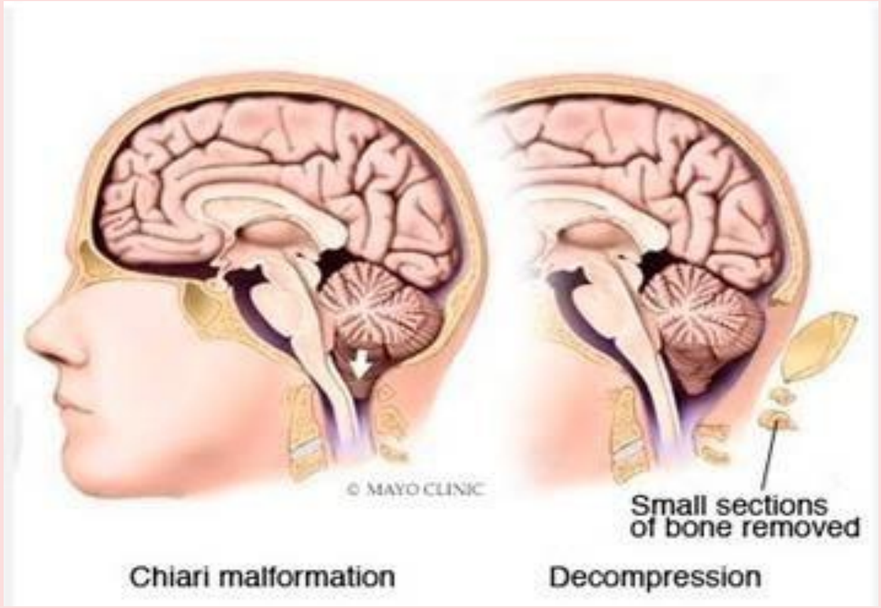




Type III



Type IV



Arachnoid Cysts

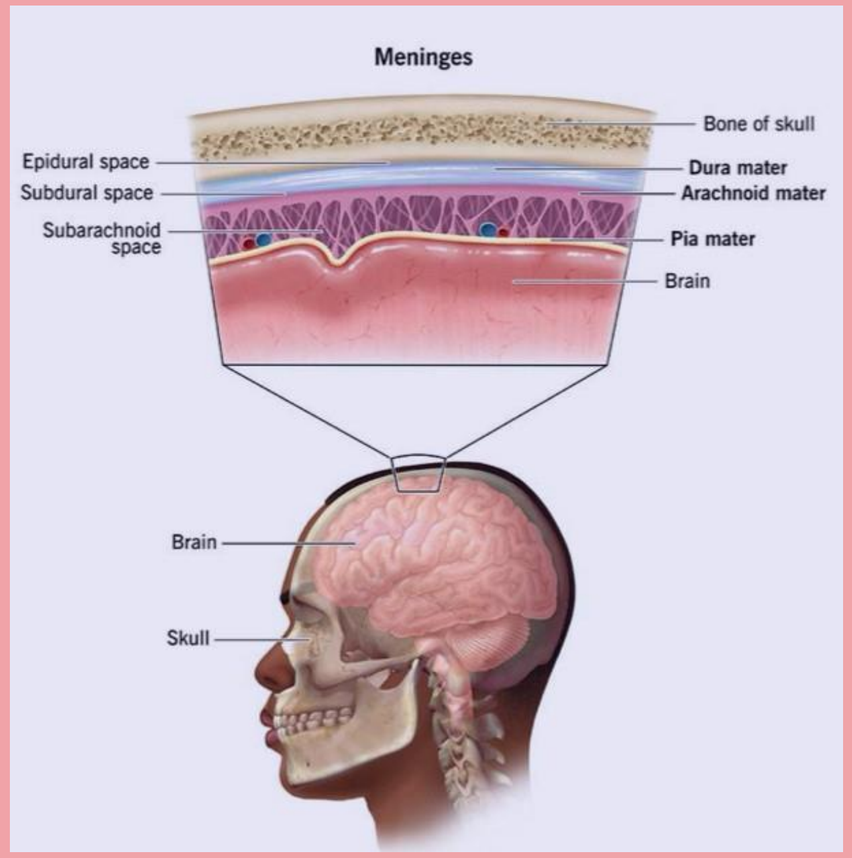
Arachnoid cysts are the most common type of brain cyst.
They are often benign asymptomatic lesion occurring in association with the central nervous system, both within the intracranial compartment (most common) as well as within the spinal canal. congenital, or present at birth (primary arachnoid cysts). Head injury or trauma can also result in a secondary arachnoid cyst.

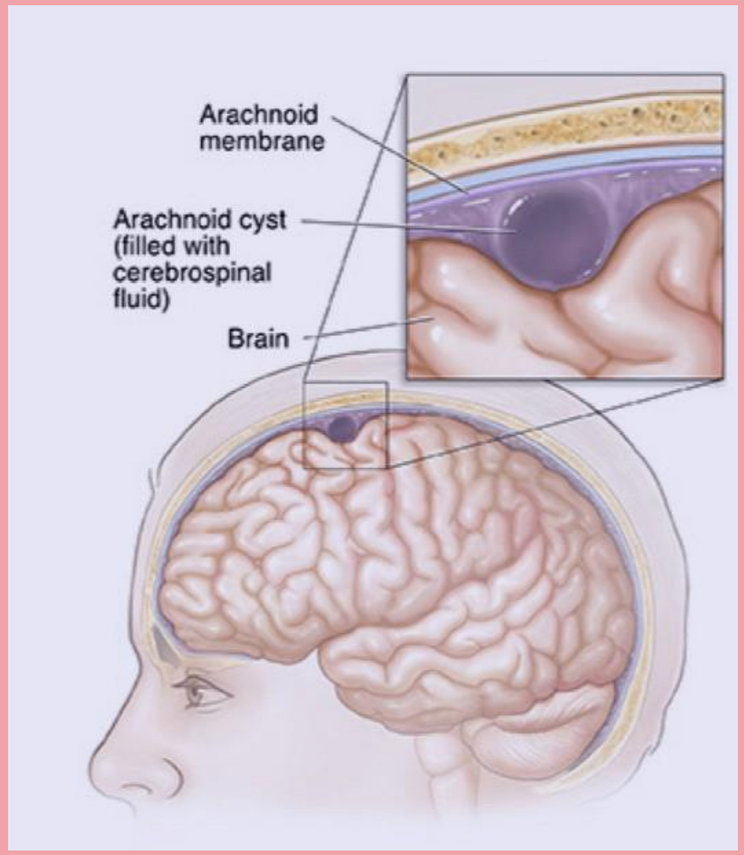
Epidemiology:
Incidence: 5 per 1000 in autopsy series.
Comprise = 1% of intracranial masses.
Male: female ratio is 4:1. More common on the left side.
Bilateral arachnoid cysts may occur in Hurler syndrome

Pathology :

There are three layers of tissue that surround the brain and spinal cord. The middle layer is called the arachnoid membrane, and this is where arachnoid cysts develop.
due to congenital splitting of the arachnoid layer with accumulation of CSF within this potential space (thus technically they are intra-arachnoid cyst).
The cyst wall is composed of flattened arachnoid cells forming a thin translucent membrane.
There is no solid component and no epithelial lining.

Pathology :


Meninges
Bone of skull
Epidural space
Dura mater
Subdural space
Arachnoid mater
Subarachnoid space
Pia mater
Brain
Brain
Skull


Arachnoid membrane
Arachnoid cyst (filled with cerebrospinal fluid)
Brain

Presentation:

Arachnoid cysts are typically asymptomatic, but they do present symptoms in some cases. These signs depend on the location and size of the cyst.

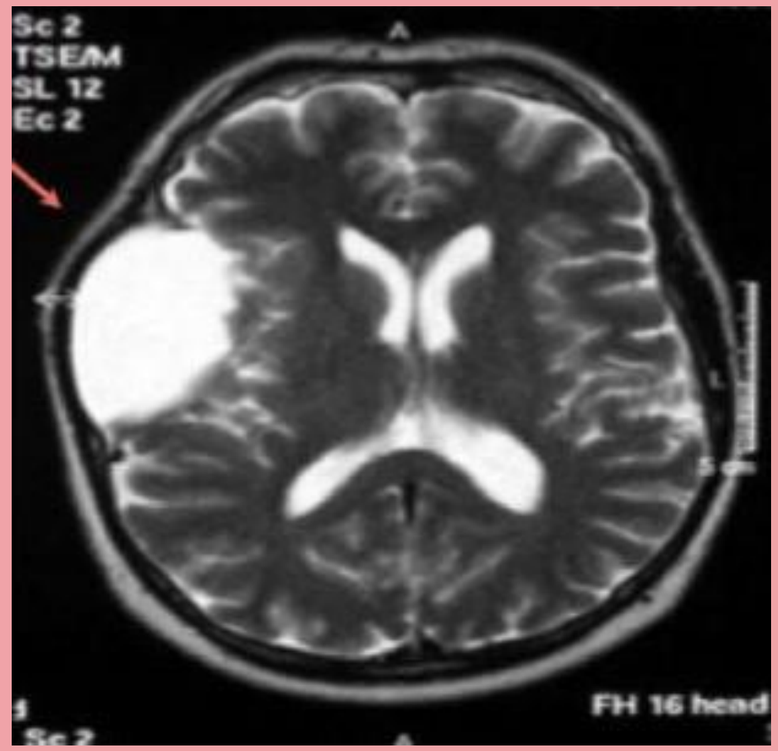
1. symptoms of intracranial hypertension (elevated ICP)
2. seizures
3. sudden deterioration:
    1. due to hemorrhage (into cyst or subdural compartment): middle fossa cysts are notorious for hemorrhage due to tearing of bridging veins.
    2. due to rupture of the cyst
    3. as a focal protrusion of the skull
    4. with focal signs/symptoms of a space occupying lesion
    5. incidental finding discovered during evaluation for unrelated condition

Locations :

- **Arachnoid cyst is mostly presented as incidental findings**

Sylvian fissure	49
CPA	11
supracollicular	10
vermian	9
sellar & suprasellar	9
interhemispheric	5
cerebral convexity	4
clival	3

Sylvian fissure



Most common

Bone remodeling

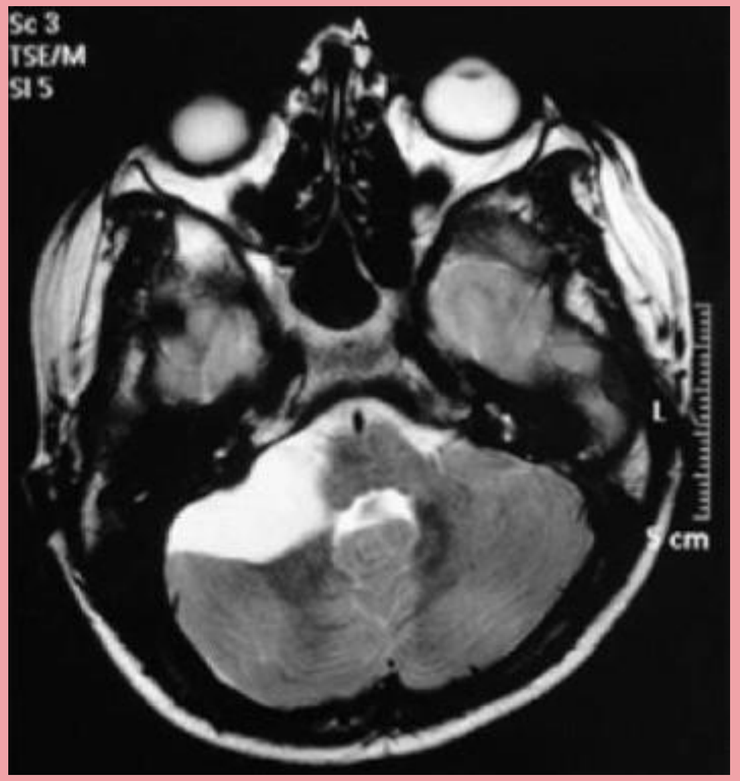
Clinical features :

- 1- ↑ICP : headache , Nausea ,Vomiting
- 2- Seizures
- 3- with minor head trauma > hrg > acute presentation

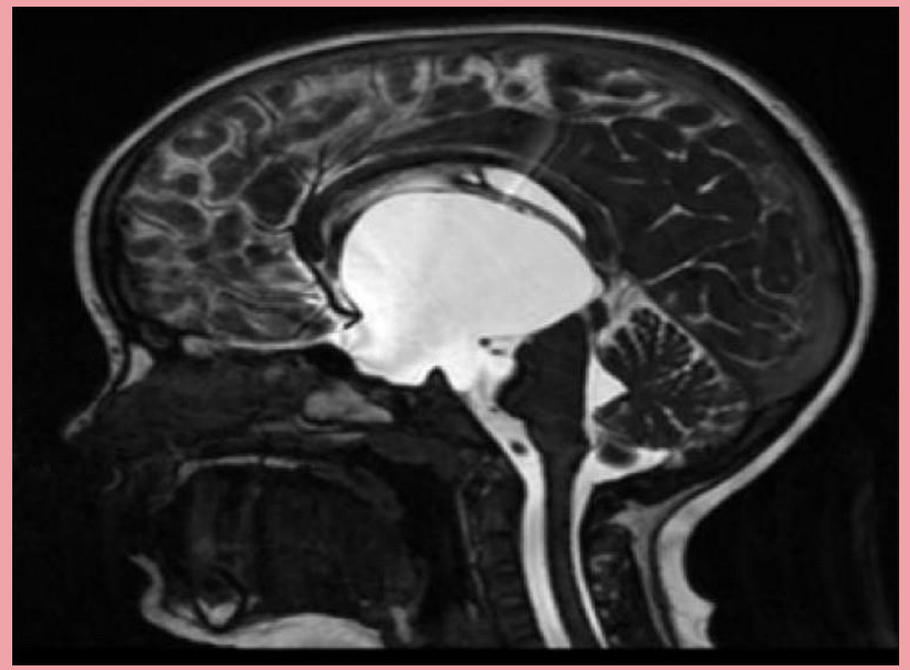
Cerebellopontine angle

Clinical features as in acoustic neuroma :

- - SN hearing loss
- - Tinnitus
- - Vertigo
- May cause compression on 5th CN



Suprasellar (the only extradural one)



- In children and adolescents .

Clinical features :

- 1- hydrocephalus .
- 2- visual impairment.
- 3- Endoc. Dysfunction as Precocious puberty .

Evaluation:

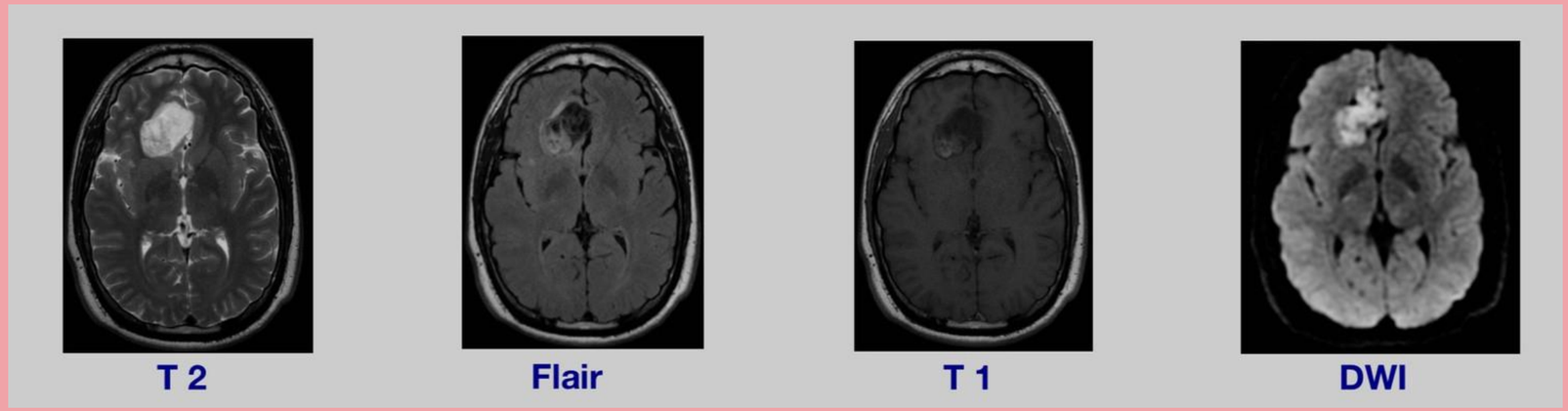
Diagnosis of an arachnoid cyst can come from imaging tests such as CT and MRI

flow studies (cisternograms, ventriculograms...) are only occasionally necessary for the diagnosis of midline suprasellar and posterior fossa lesions

Epidermoid cysts in the cerebellopontine angle (CPA) may mimic an .arachnoid cyst, but are high signal on DWI MRI

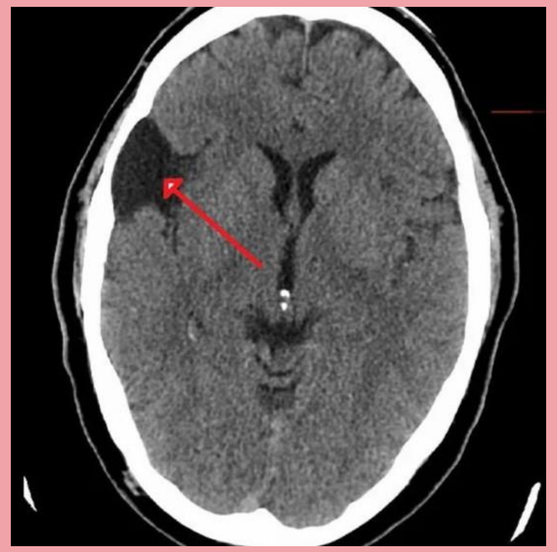
- This enables them to be distinguished from epidermoid cysts

often shows a heterogeneous/"dirty" signal on FLAIR
restricted diffusion
more lobulated
tend to engulf adjacent arteries and cranial nerves



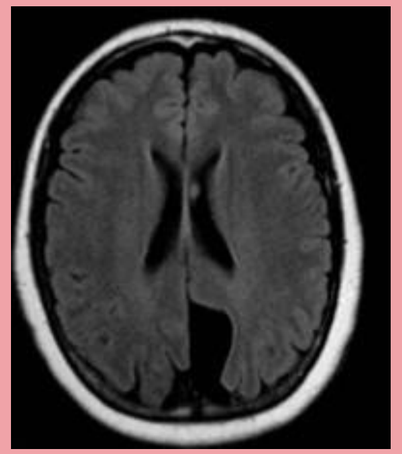
CT

smooth bordered non - calcified
extraparenchymal cystic mass
with density similar to CSF
and no enhancement with IV contrast

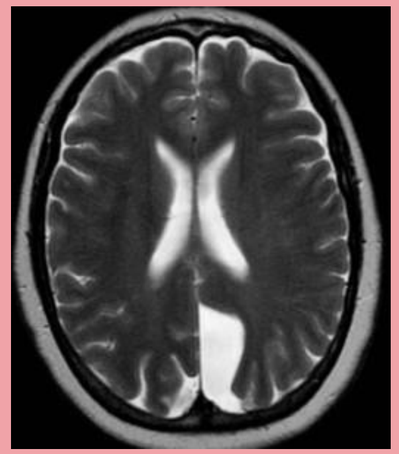


MRI

Better than CT in differentiating
the CSF contained in
arachnoid cysts from the fluid of
neoplastic cysts
o May also show cyst walls



Flair



T2

Management and Treatment :

Asymptomatic (no ventricular distortion, enlargement)

Follow up at regular intervals

symptomatic

drainage by needle aspiration or burr hole evacuation

craniotomy, excising cyst wall and fenestrating it into basal cisterns

endoscopic cyst fenestration through burr hole

shunting of cyst into peritoneum or into vascular system

Dandy Walker Malformation

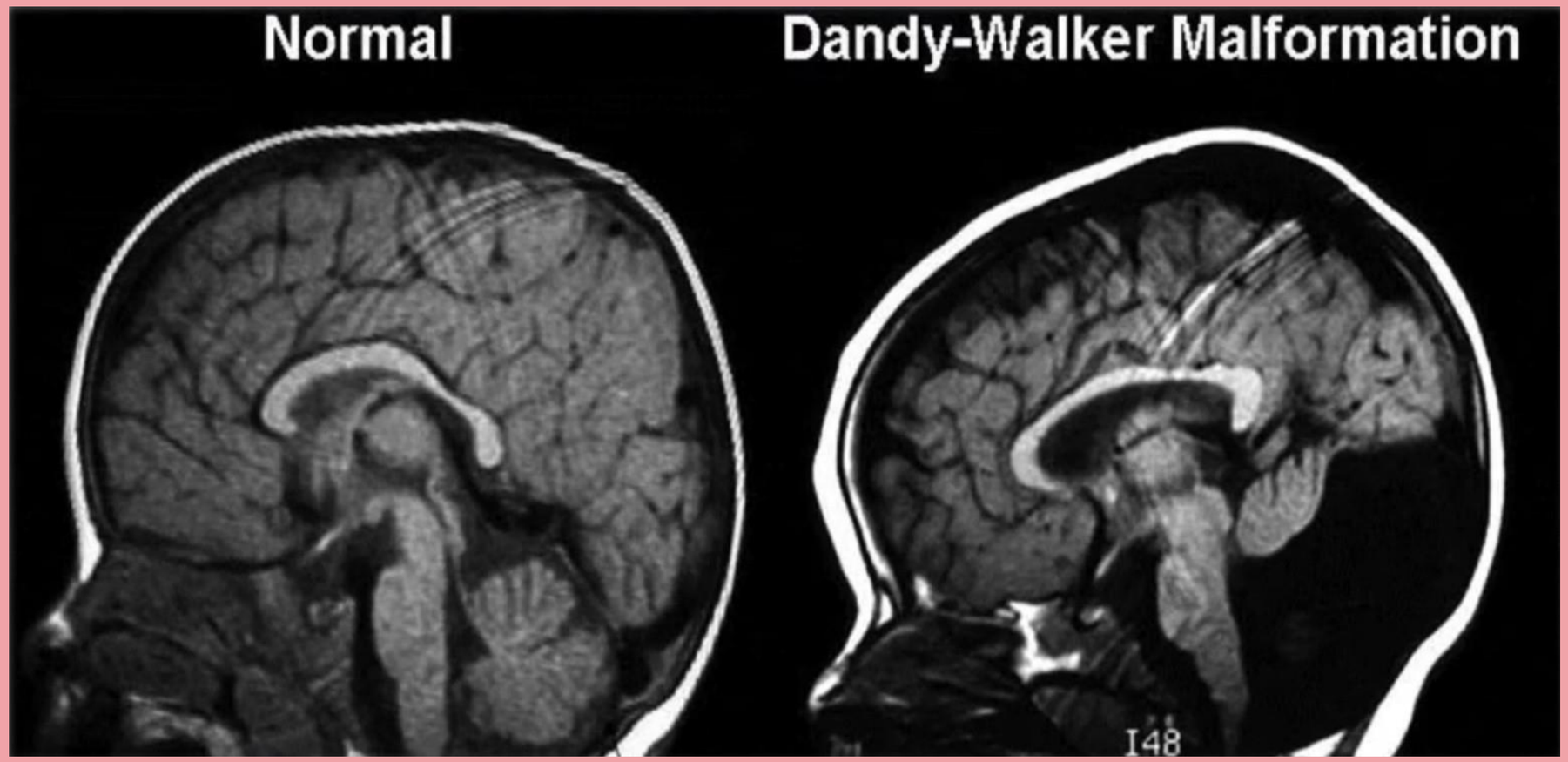
rare, congenital brain anomaly

Characterised by :

1 complete or partial agenesis of cerebellar vermis (midline part of cerebellum).

2 Cystic dilatation of the 4th ventricle (failure of foraminal outlets to open)

3 An enlarged posterior fossa with upward displacement of tentorium , lateral sinuses


Normal
Dandy-Walker Malformation
I48

symptoms:

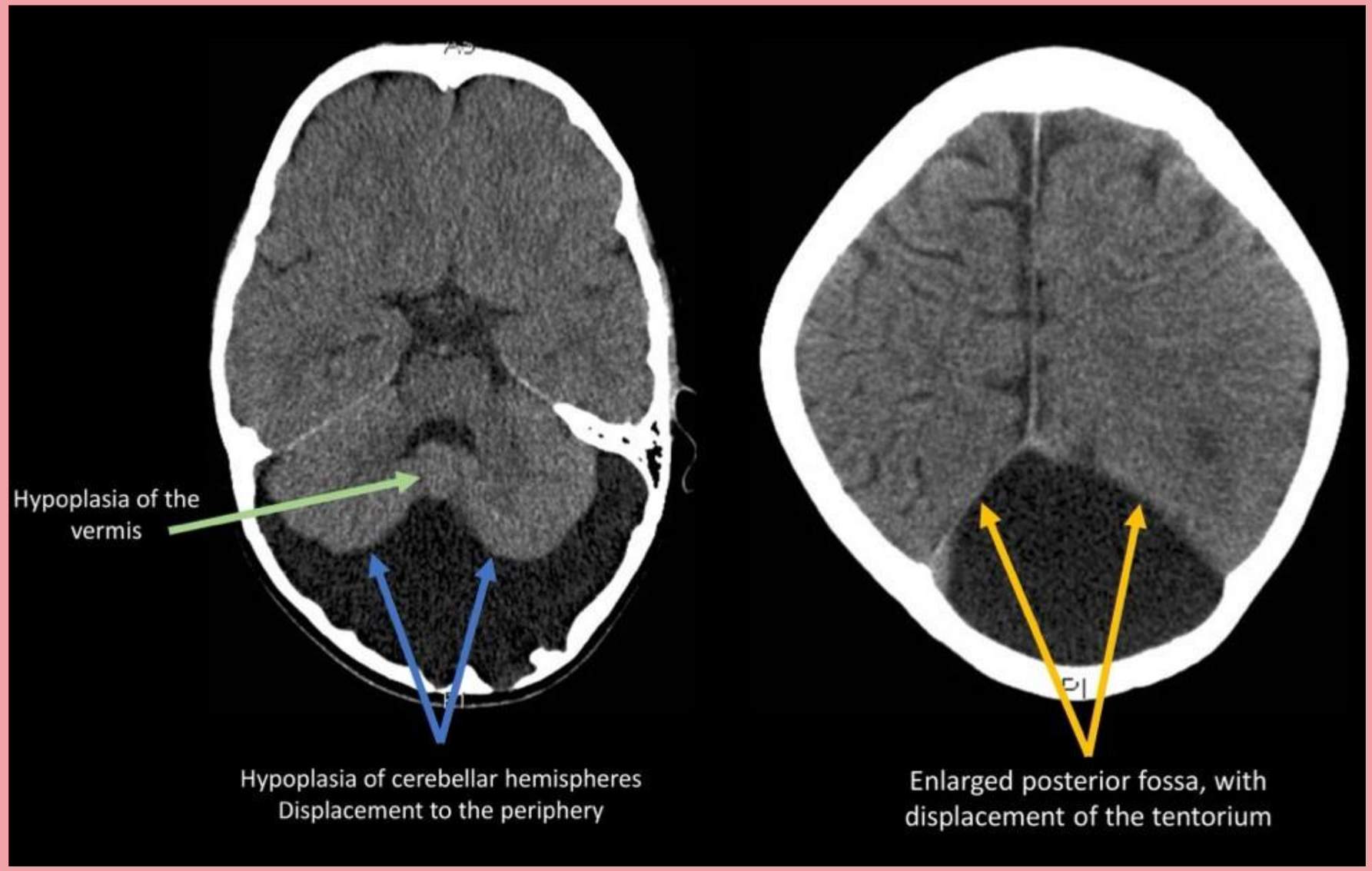
1 90%of patients have hydrocephalus (present as increasing head size, vomiting ,excessive sleepiness)

2 If not diagnosed postnatally : in childhood the major presenting features are ataxia, and delayed motor development

3 Associated anomalies : agenesis of the corpus callosum, occipital encephalocele,spina bifida, syringomyelia , cleft palate , cardiac and renal anomalies.

Male : Female = 1 : 3

May be associated with Phaces Syndrome

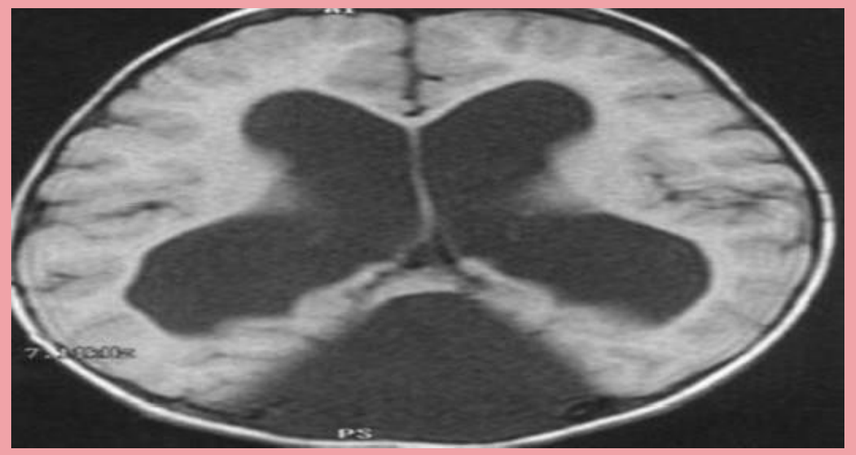

Hypoplasia of the vermis
Hypoplasia of cerebellar hemispheres
Displacement to the periphery
Enlarged posterior fossa, with displacement of the tentorium

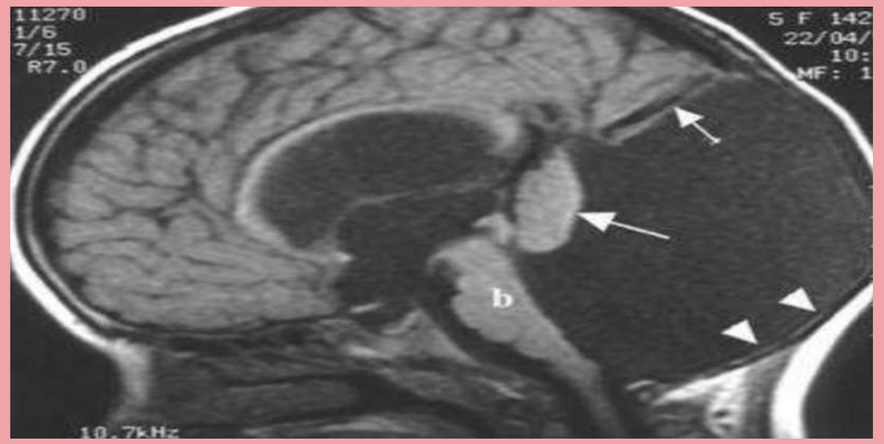
Management :

1: cyst - peritoneal shunt / VP shunt

2 : therapy : speech therapy can help with language and speech development

3 : meditation : to control seizures





Thanks