PAEDIATRIC NEUROSURGERY: HYDROCEPHALUS AND CRANIOPATHIES. SPINA BIFINA AND OTHER DEVELOPMENTAL MALFORMATIONS

34484 Pathology of the nervous system

Neurosurgery

Topic 16

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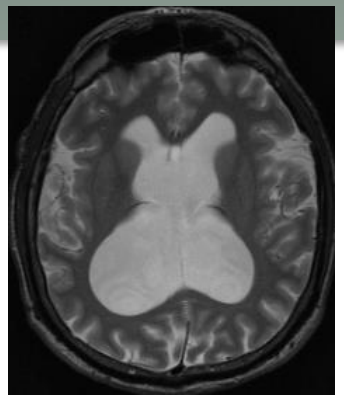
KEY POINTS

- **Hydrocephalus**
- **Craniopathies**
  - ***Craniosynostosis***
  - ***Cranioencephalic dysraphisms***
- **Spina bifida**
  - ***Spinal dysraphisms***
- **Other developmental malformations**
  - *Chiari and Arnold-Chiari malformations*
  - *Syringomyelia*

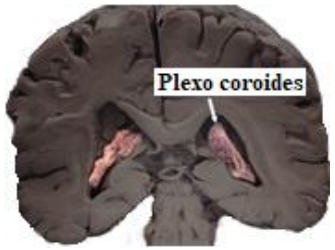




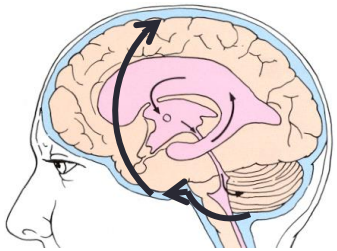

Cerebrospinal fluid (CSF)

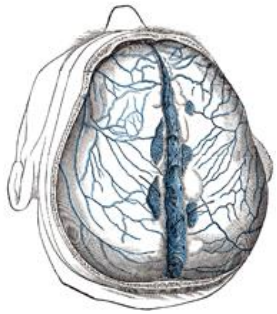
REMEMBER?

- Clear transparent fluid, 130 mL
- Function: protects CNS (brain + spinal cord)
  - Mechanical: damping (buoyancy, trauma)
  - Chemical: neuroendocrine factors regulation, elimination of metabolic debris
  - Physical: keeps ICP within normal limits (avoids ↑ ICP)
- Production – Circulation – Reabsorption



60% ~ 480 ml/d





Choroid **C**reates – ***A***rachnoid ***A***bsorbs

Cranial cavity

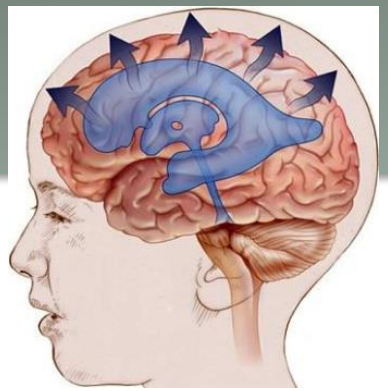
REMEMBER?

- Adult skull = bone shield
- **Infant skull**
  - Sutures and fontanelles: allow ↑ cranial cavity capacity (maintaining ICP)

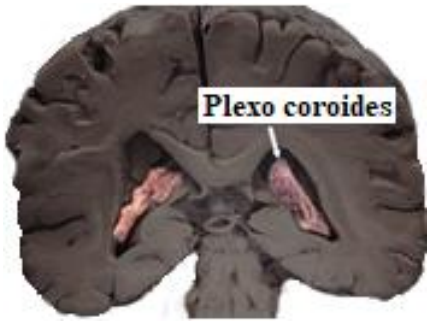
  ⇒ Larger compensation of an ↑ICP

Age group	Normal range (mmHg)
Adults and older children	< 10-15
Young children	3-7
Newborns	1.5-6

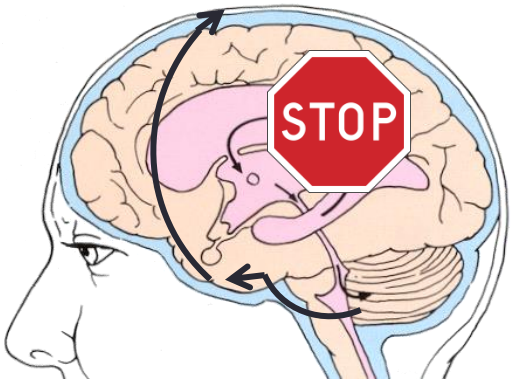
HYDROCEPHALUS



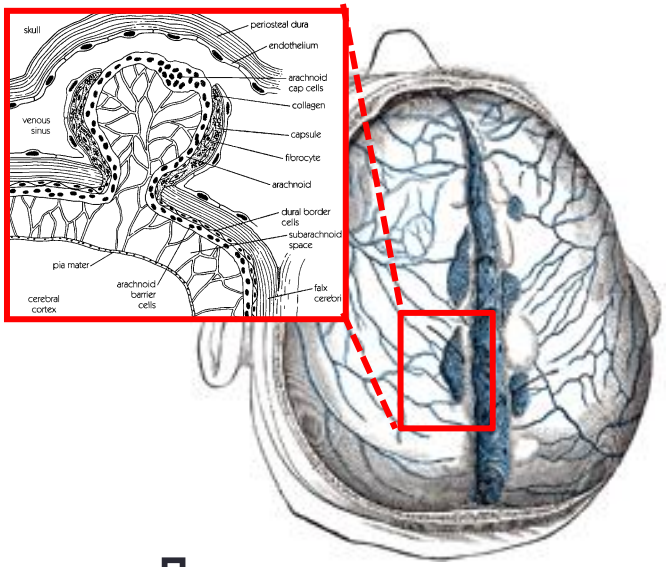
- ↑ **intracranial content of CSF**
  - One cause of ICHT (mass-blood-CSF)
- Greek *hydroképhalos* (*hydros* = water, *képhalé* = head)



⇧ **Production**
(very rare)

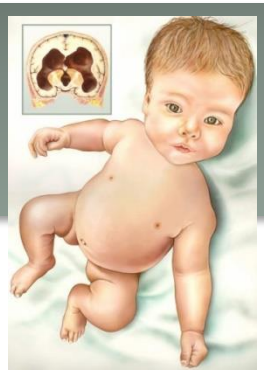


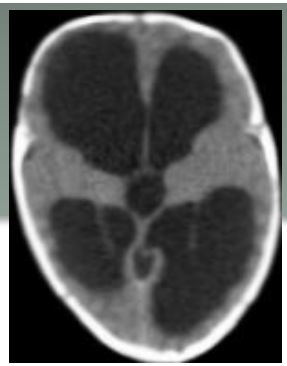
Circulation
(obstruction to flow)
Hydrocephalus non communicans



⇩ **Reabsorption**
Hydrocephalus communicans

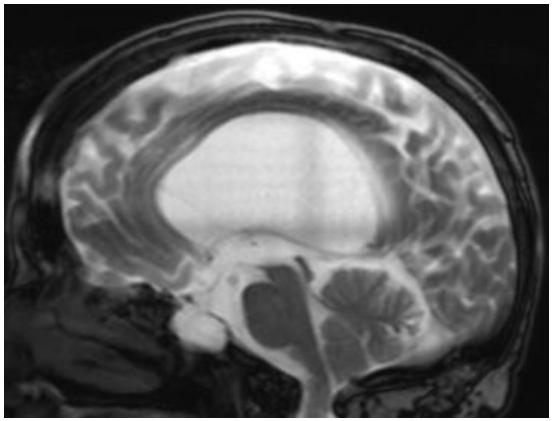
Types of hydrocephalus





Prematurity

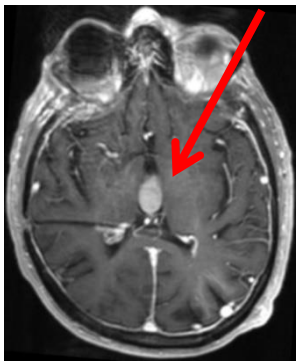
- **According to moment of presentation**
  - Congenital (at birth)
    - *Intraventricular haemorrhage, Sylvian aqueduct stenosis*
  - Acquired
    - *Tumours, cysts*
    - *Haemorrhages, infections*



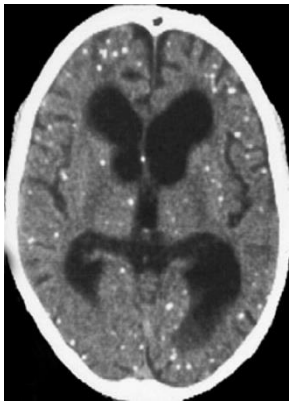
Sylvian aqueduct stenosis

- **According to CSF circulation**

Thalamic haemorrhage



Tumour III ventricle

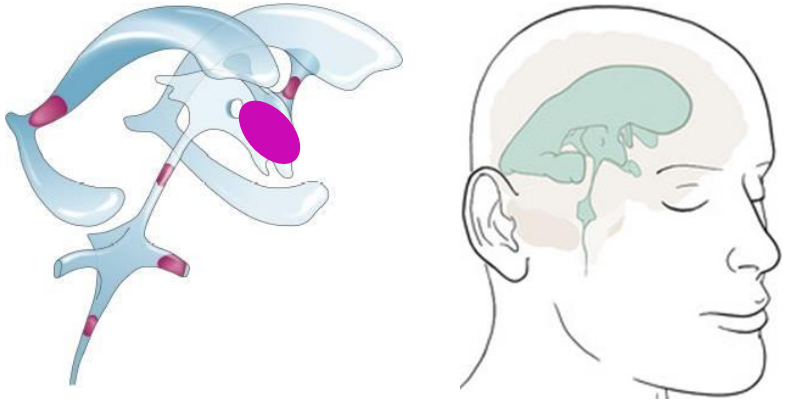


Cysticercosis

Tuberculous meningitis

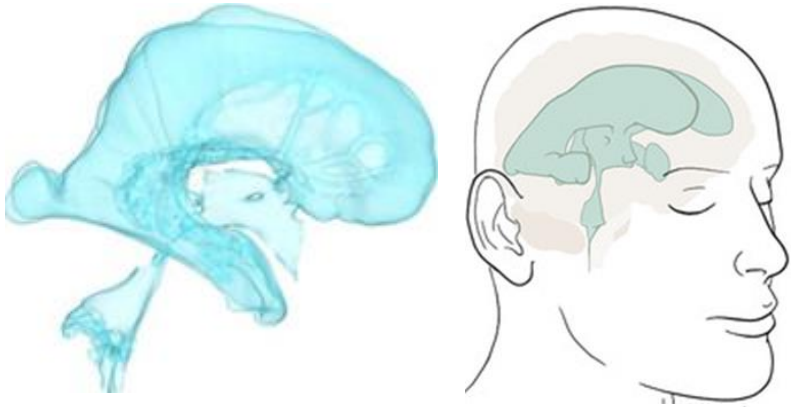
Types of hydrocephalus

- **According to moment of presentation**
- **According to CSF circulation**
  - **Non-communicating (obstructive)**
    - *CSF flow obstruction in the ventricular system (foramen of Monro, Sylvian aqueduct, fourth ventricle)*
    - *Anatomy anomaly, tumours…*
  - **Communicating**
    - *CSF circulates free in the ventricular system*
    - *Deficit of drainage or reabsorption (meninges, venous sinuses…)*



NON-communicating hydrocephalus

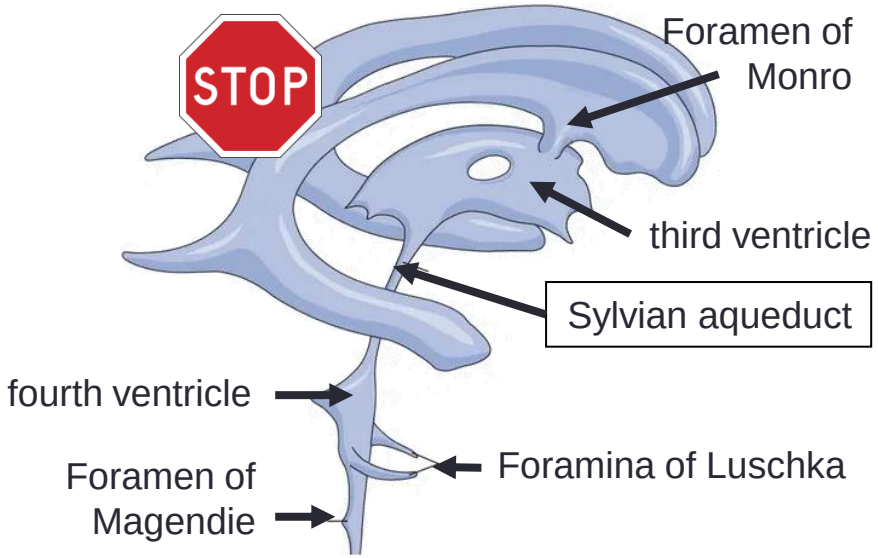
- *Points of obstruction*

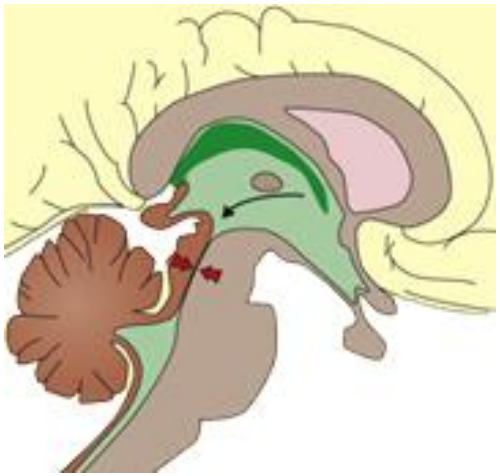


Communicating hydrocephalus

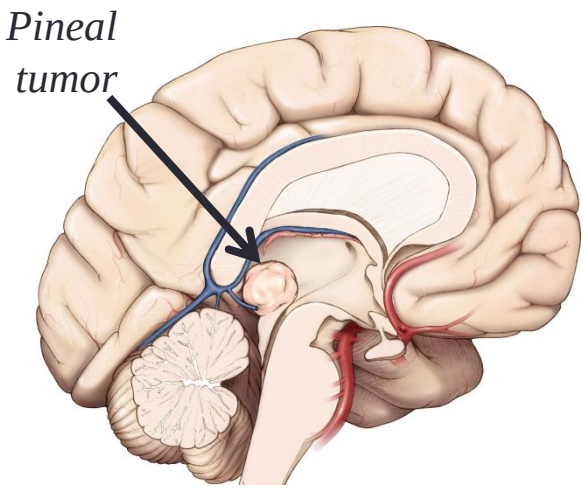
Non-communicating hydrocephalus

- Intraventricular tumours
- Pineal region tumours
  (hydrocephalus, alt vermis, dilation fourth ventricle)
- Sylvian aqueduct stenosis
- Dandy-Walker malformation

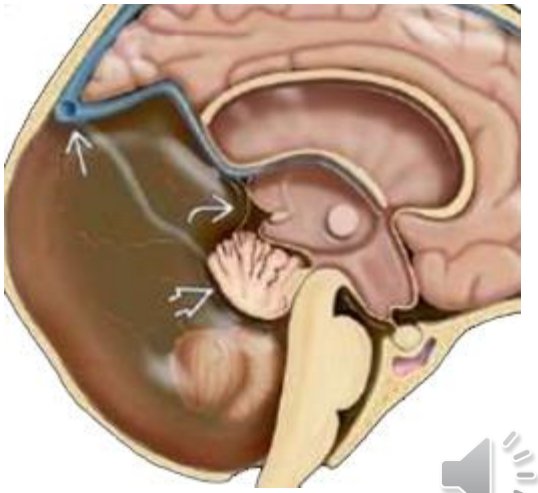




Sylvian aqueduct stenosis

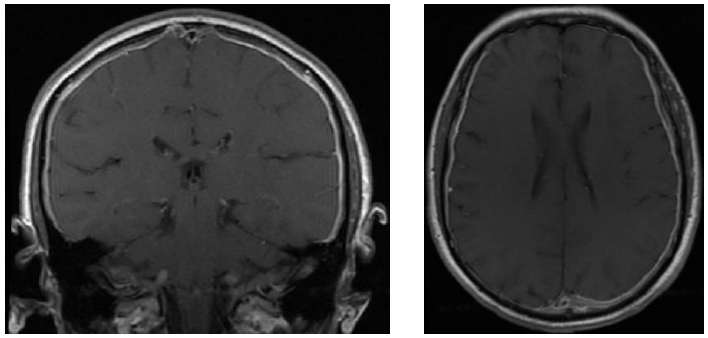


Pineal tumour



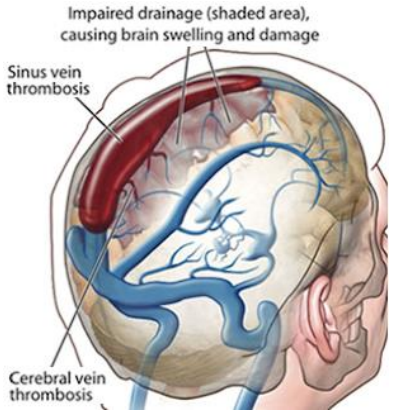
Dandy-Walker malformation

Communicating hydrocephalus



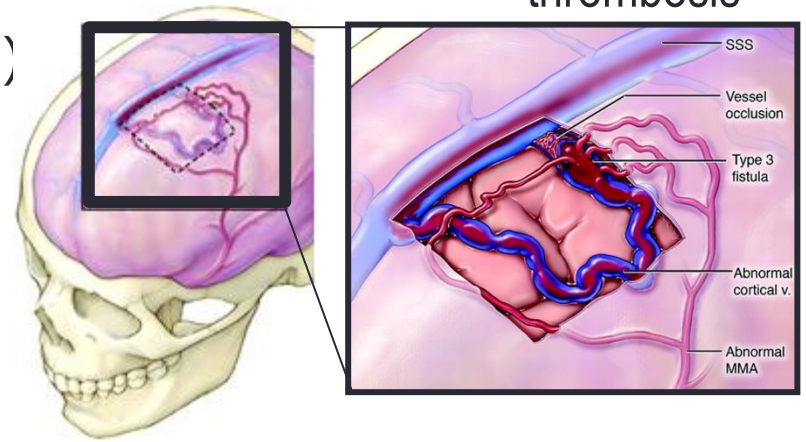
MRI: Meningeal carcinomatosis

CT: SAH



Superior longitudinal sinus thrombosis

- Meninges
  - Meningitis
  - Subarachnoid haemorrhage (SAH)
  - Meningeal carcinomatosis
  - Meningeal lymphomatosis
- Vascular
  - Venous sinus thrombosis
  - Arteriovenous fistula



Dural arterio-venous fistula

Hydrocephalus clinical features

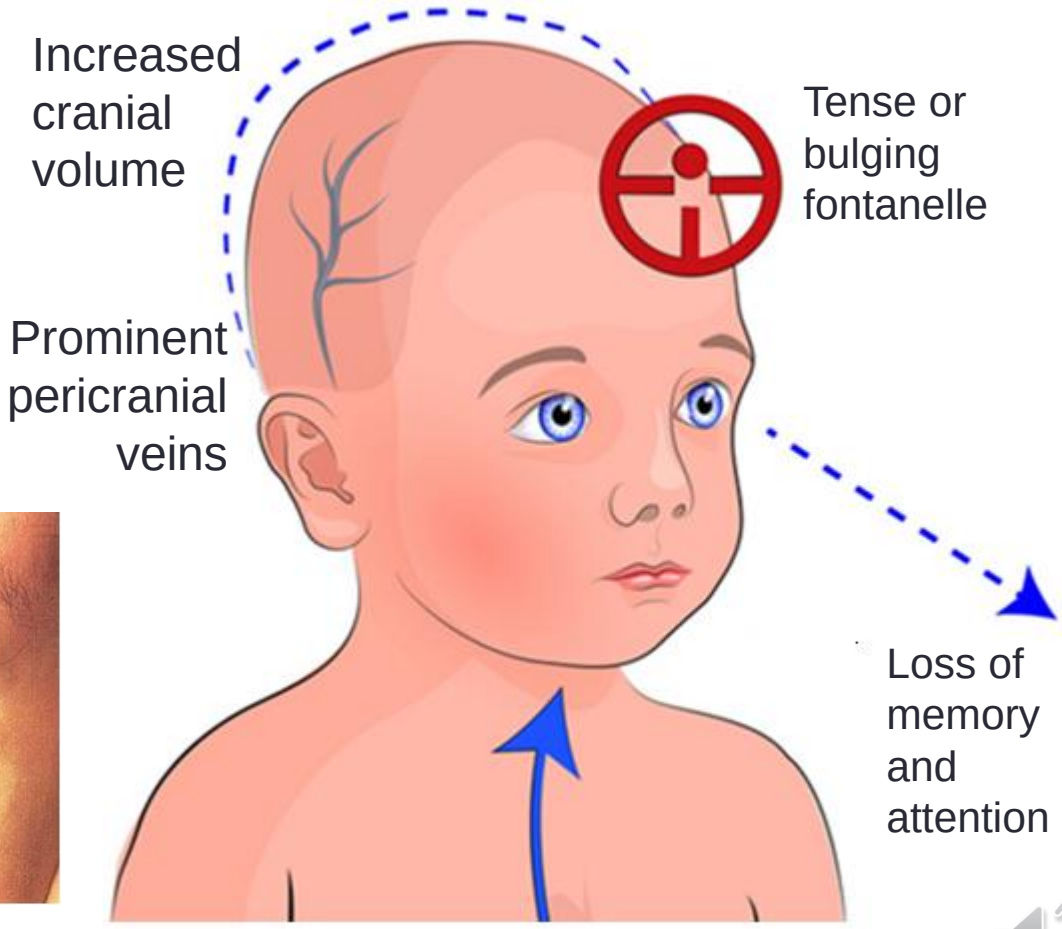
- **Infant (open sutures and fontanelles): morphology + raised intracranial pressure**
  - *1.1:1000 infants*
  - Increase in head circumference
  - Bulging fontanelles (tense)
  - Prominent pericranial veins
  - Irritability, crying
  - Poor food intake, lethargy
  - Setting sun phenomenon
  - ❖ *Chronic → psychomotor retardation*
- **Child and adult**

CT Neonatal hydrocephalus

Raised intracranial pressure in young children

REMEMBER?

Sunset eye sign
(setting sun phenomenon)



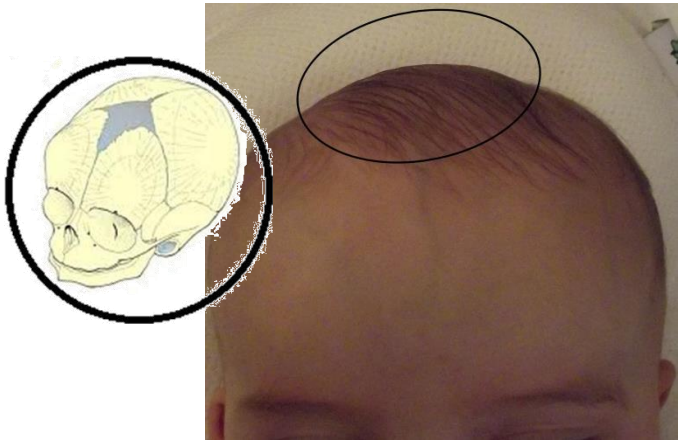
Raised intracranial pressure in children

- **Infant: symptoms**
  - Irritability, crying
  - Poor food intake, lethargy

Macrocephaly

Sunset eyes

Convergent strabismus



Tense fontanelle

Hydrocephalus clinics

- **Infant (open sutures and fontanelles): morphology and raised intracranial pressure**
- **Child and adult: raised intracranial**

Acute	Chronic
○ Headache ○ Nausea and vomiting ○ Papilledema ○ Sixth cranial nerve palsy (diplopia) ○ Gait disturbances ○ Parinaud Syndrome (upright conjugate gaze palsy)	○ Headache ○ Nausea and vomiting ○ Optic nerve atrophy – blindness ○ VI cranial nerve palsy (diplopia) ○ Spastic paraparesis ○ Parinaud syndrome (upward vertical conjugate gaze palsy) ○ Dysmetria upper limbs ○ Endocrine abnormalities ○ Cushing's triad ○ Impairment level of consciousness

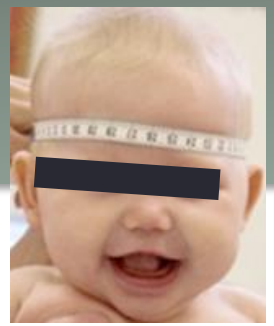
Raised intracranial pressure clinical features

REMEMBER?

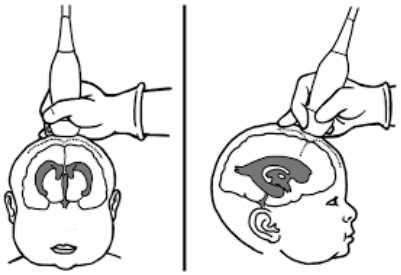
- **Headache**
  - ↑ at night, may wake up the patient
  - ↑ with Valsalva manoeuvres
- **Vomiting**
  - ↑ in the morning (on waking)
  - Projectile vomiting
- **Papilledema** (fundoscopy)
- **Decreased level of consciousness**
- **Cushing's triad**
  - Increased blood pressure, bradycardia, and irregular breathing (complete in only 30% cases)
- **Diplopia** (VI cranial nerve palsy), blurred vision
- **Focal neurological signs**

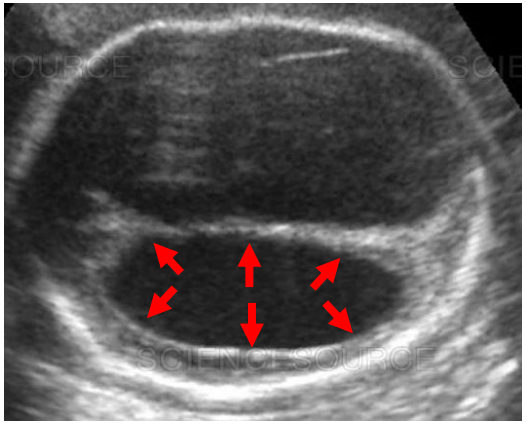
Diagnosis in infants

- Clinical suspicion:
  - ↑ head circumference
- Trans-fontanelle echography
- Imaging
  - Cranial XR (NOT as diagnostic tool)
    - *Infant: diastasis of sutures*
    - *Child/adult, chronic cases:*
      - *Copper-beaten skull*
      - *Enlarged, eroded or decalcified sella turcica*
  - **CT / MRI**
    - *Hydrocephalus*
    - *ETIOLOGY*



Head perimeter measurement



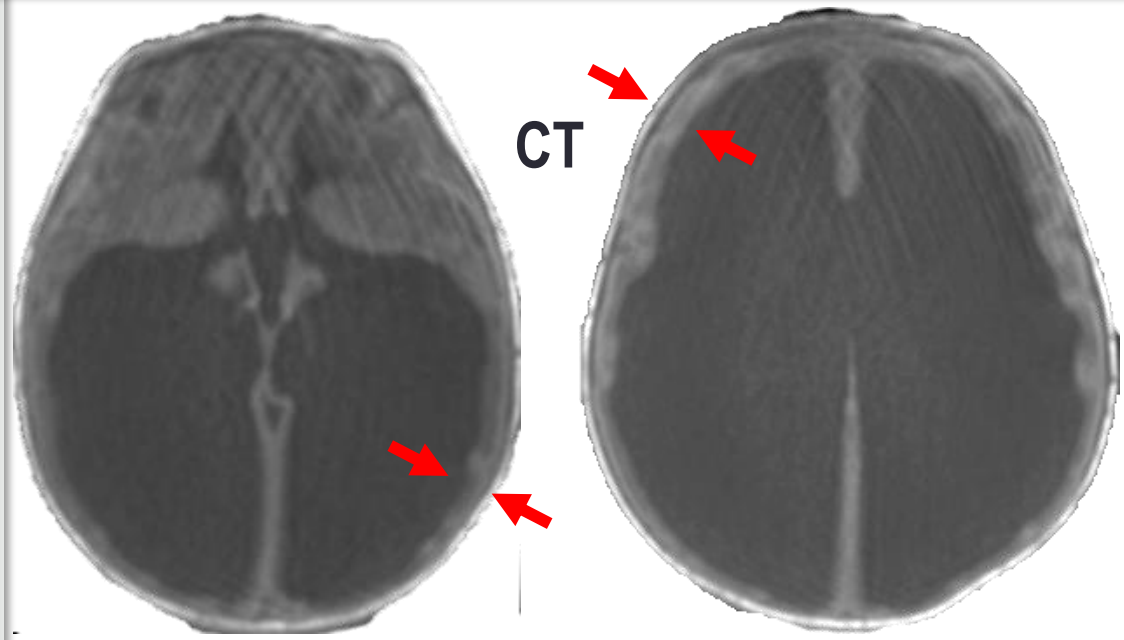


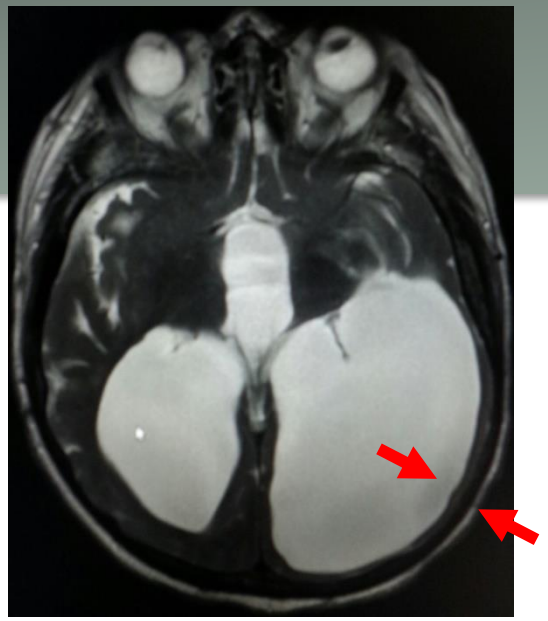
Echography

3D CT

Cranial X-ray (chronic ICHT)

CT and MRI

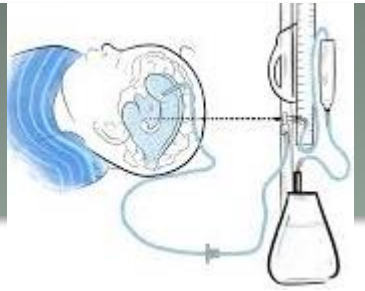




MRI

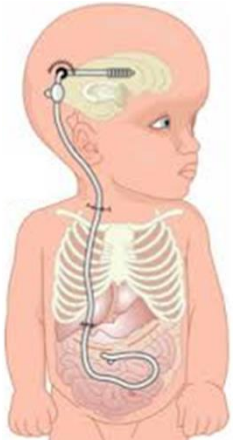
- Congenital / newborn hydrocephalus
  - Brain parenchyma atrophy
- Etiology diagnosis
  - Prematurity: intraventricular haemorrhage

Treatment: CSF drainage

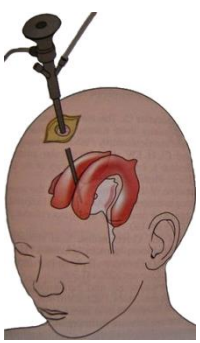


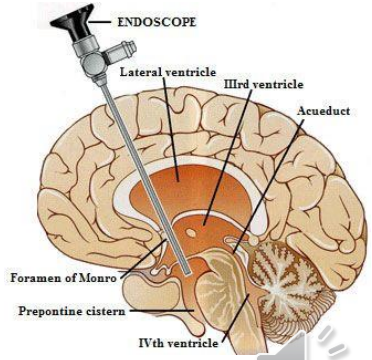
External ventricular drain

- External CSF drain (temporary measure) (EVD)
  - Intraventricular haemorrhage (prematurity / subarachnoid haemorrhage)
  - Meningitis, ventriculitis
  - Postsurgical
- CSF shunt
  - Ventricleperitoneal shunt (VP shunt)
  - Ventricleatrial shunt (VA shunt)
- Endoscopic ventriculostomy
  - Perforation floor third ventricle
    - *CSF flows directly to subarachnoid space*
    - *Only if obstruction at level of third ventricle*
    - *Sylvian aqueduct or posterior fossa*



Ventricleperitoneal shunt





Endoscopic ventriculostomy

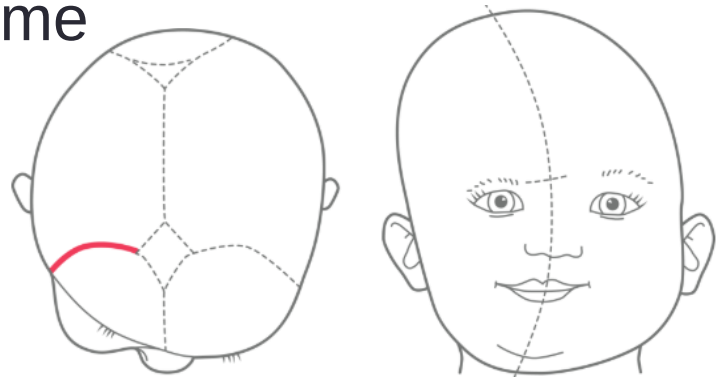
CRANIOENCEPHALIC AND SPINAL MALFORMATIONS

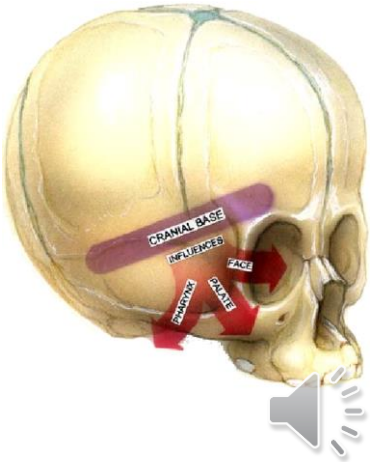
- Skin
  - Dermal sinus
  - Sinus Pericrania
- Nervous system
  - *Cranial and spinal* ***dysraphisms***
  - Arachnoid cysts
  - Dandy-Walker malformation
  - Alterations in neuronal migration
- Bony structures
  - *Cranial and spinal* ***dysraphisms***
  - **Craniosynostosis** (craniostenosis)
  - **Craniocervical junction malformations**: Chiari

1. Craniosynostosis
2. Cranial dysraphisms
3. Spinal dysraphisms
4. Craniocervical junction malformations and syringomyelia

1. CRANIOSYNOSTOSES

- Early closure of 1 or more cranial sutures
  - Abnormal development of the skull → facial asymmetry
- Primary alteration of the mesenchyme
  - Usually genetic mutation
  - Sometimes hereditary
- Incidence 3-5 / 10,000 newborns
- Clinical features:
  - Facial asymmetry
  - Cranial dysmorphia → give name to craniosynostosis
  - Compression of brain → ICHT → cranial nerves lesion
  - ➢ <u>Isolated or syndromic</u>
    - *Polydactyly, syndactyly*





Craniosynostoses

- Diagnosis
  - Clinical features: cranial morphology
    - *Palpation of sutures? Not always absent, and absence is not always diagnostic*
    - *Rule out SYNDROMES*
  - 3D high resolution multi-slice CT

Oxycephaly – Cloverleaf skull

Polydactyly

Syndactyly in a syndrome

Syndactyly

Craniosynostoses

- Diagnosis
  - Clinical features
  - 3D high resolution multi-slice CT

Simple (1 suture)

- **Scaphocephaly**
- **Trigonocephaly**

Midline

- Plagiocephaly
- Brachycephaly

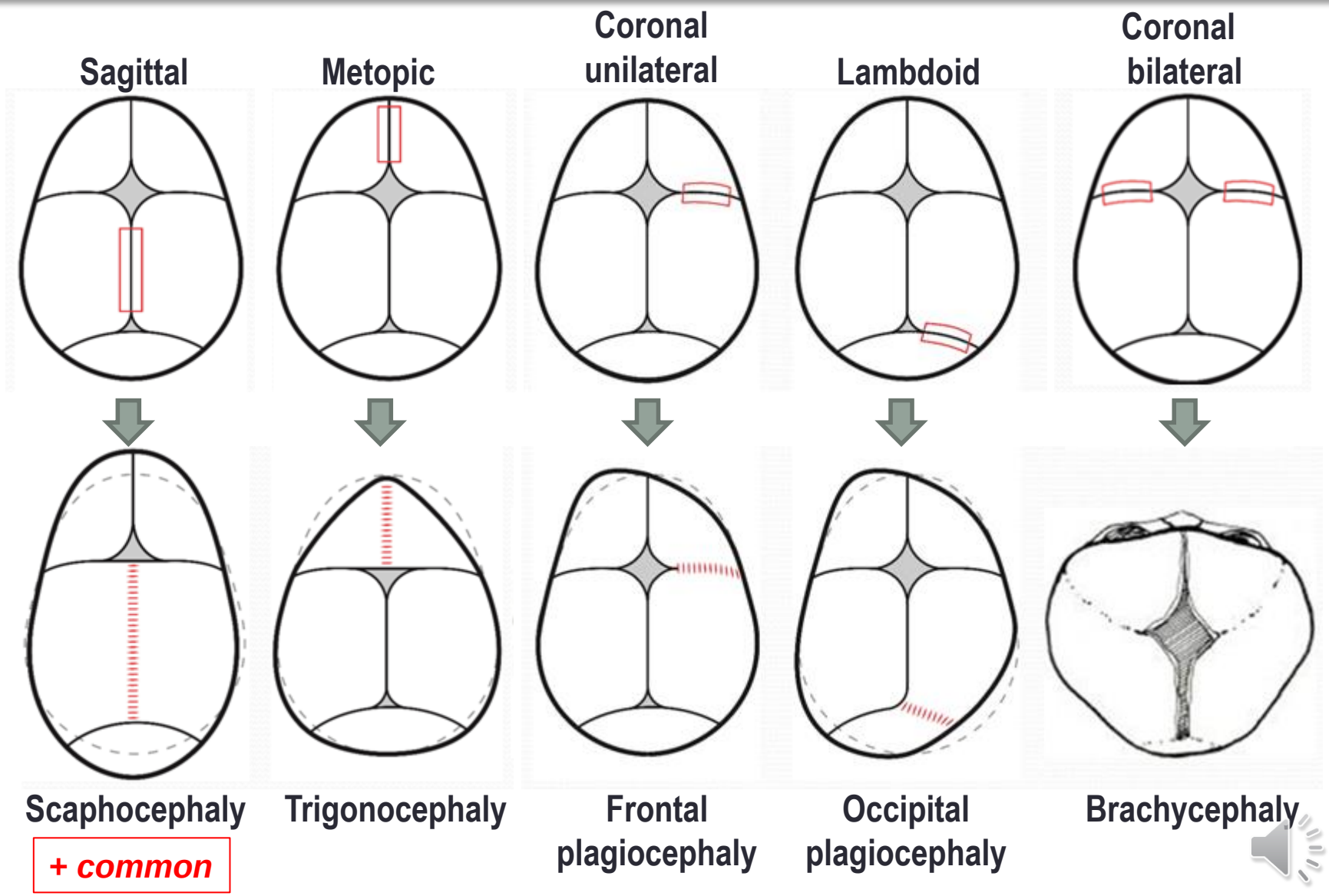
Complex (≥ 2 sutures)

- Turricephaly (acrocephaly, open sagittal suture)
- Oxycephaly. Cloverleaf skull

Syndromic (40%)

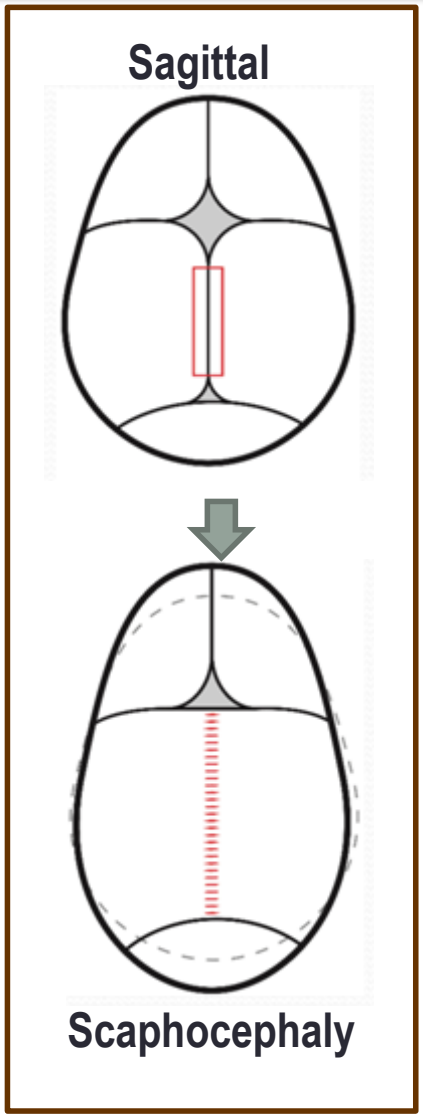
- **Crouzon syndrome**
- Acrocephalosyndactyly
  - ***Type 1 – Apert syndrome***
  - *Type 3 – Saethe-Chotzen syndrome*
  - *Type 5 – Pfeiffer syndrome*
- Acro cephalopolysyndactyly
  - *Type 2 –Carpenter Syndrome*
  - *Type 3 – Sakati-Nyhan-Tisdale syndrome*

Simple craniosynostoses



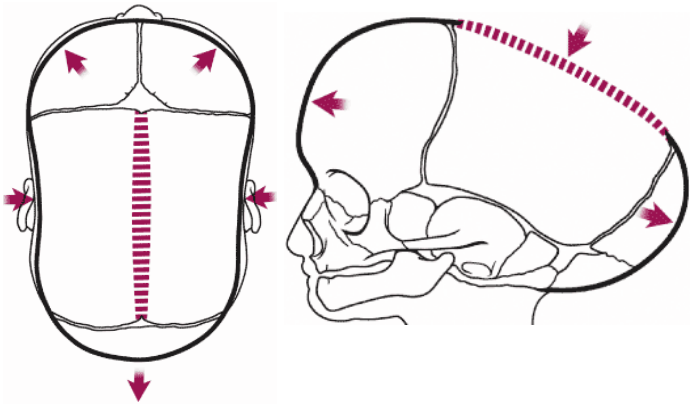
+ common

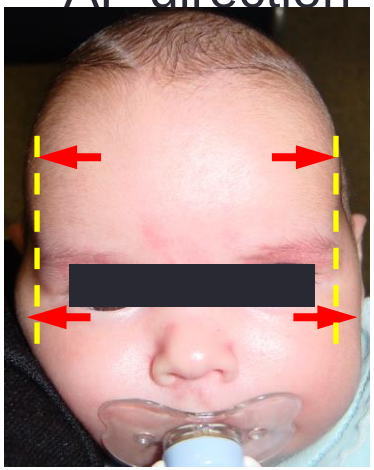
Simple craniosynostoses



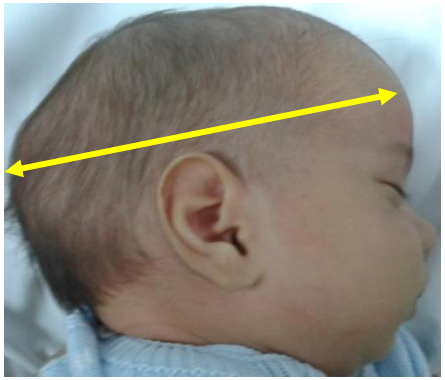
1. Scaphocephaly

- Greek *skáphē* 'boat'
  - *Dolichocephaly*
- Most common craniosynostosis (1/4.000)
- Early closure sagittal suture → skull grows in AP direction

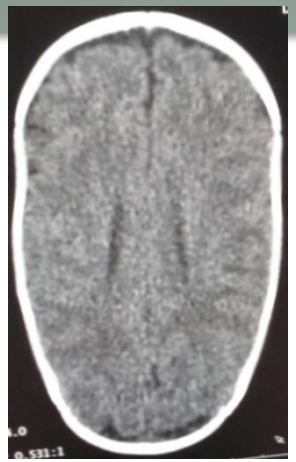




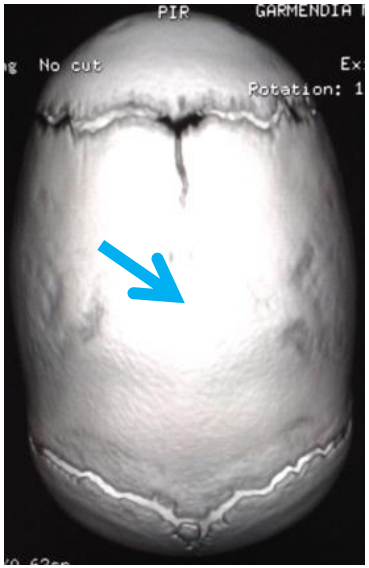
1. Scaphocephaly (dolichocephaly)

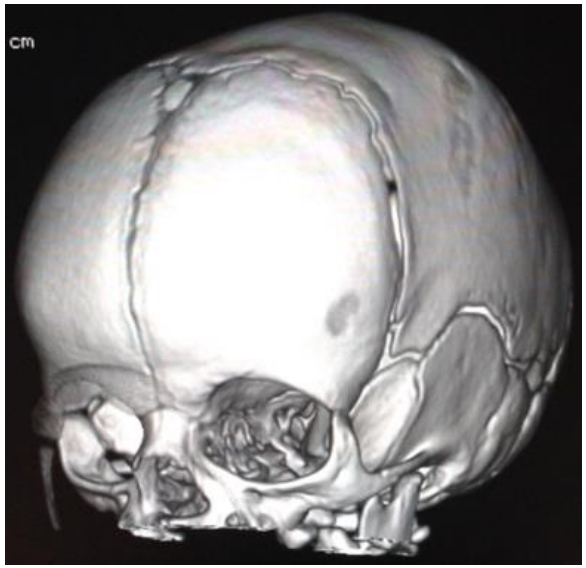


CT

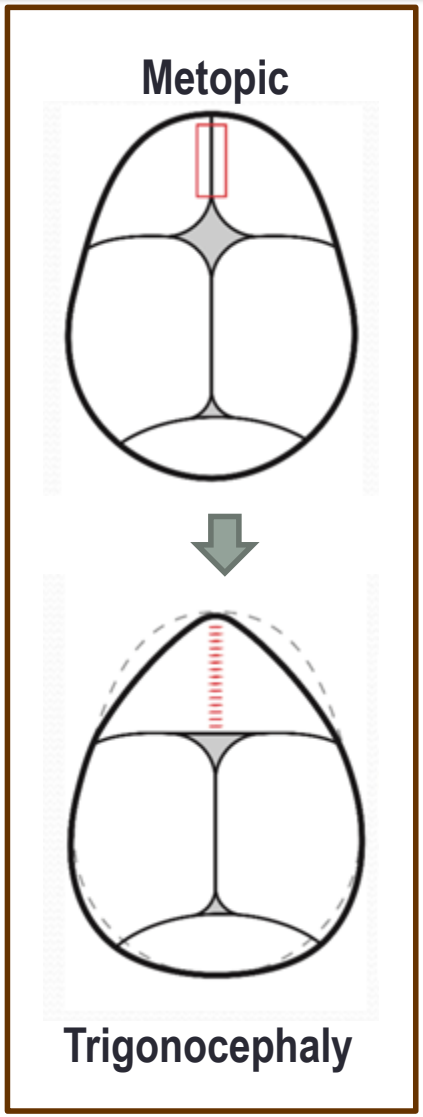


3D CT



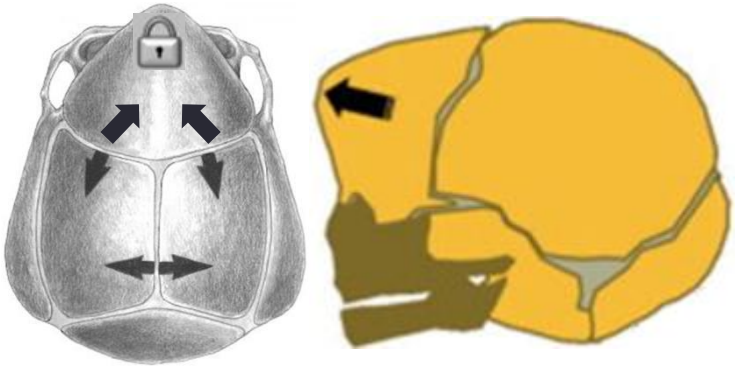


Simple craniosynostoses



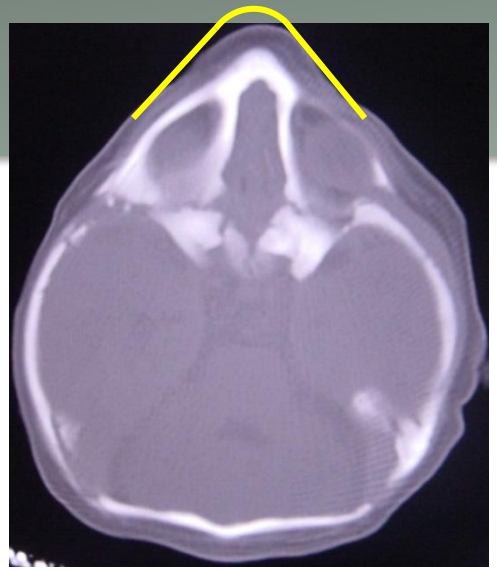
2. Trigonocephaly

- Closure metopic suture → frontal bones cannot grow
- Narrow triangular forehead and palpable border
- Hypotelorism

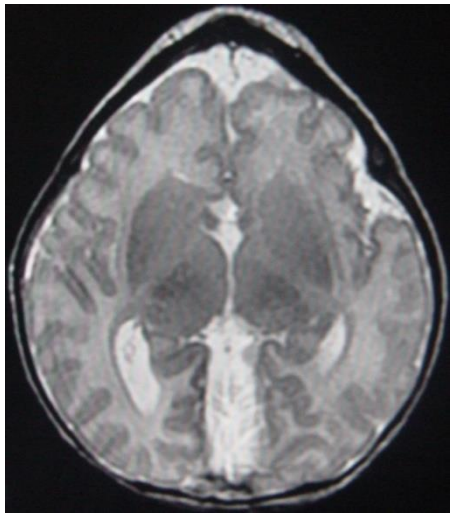


2. Trigonocephaly

CT

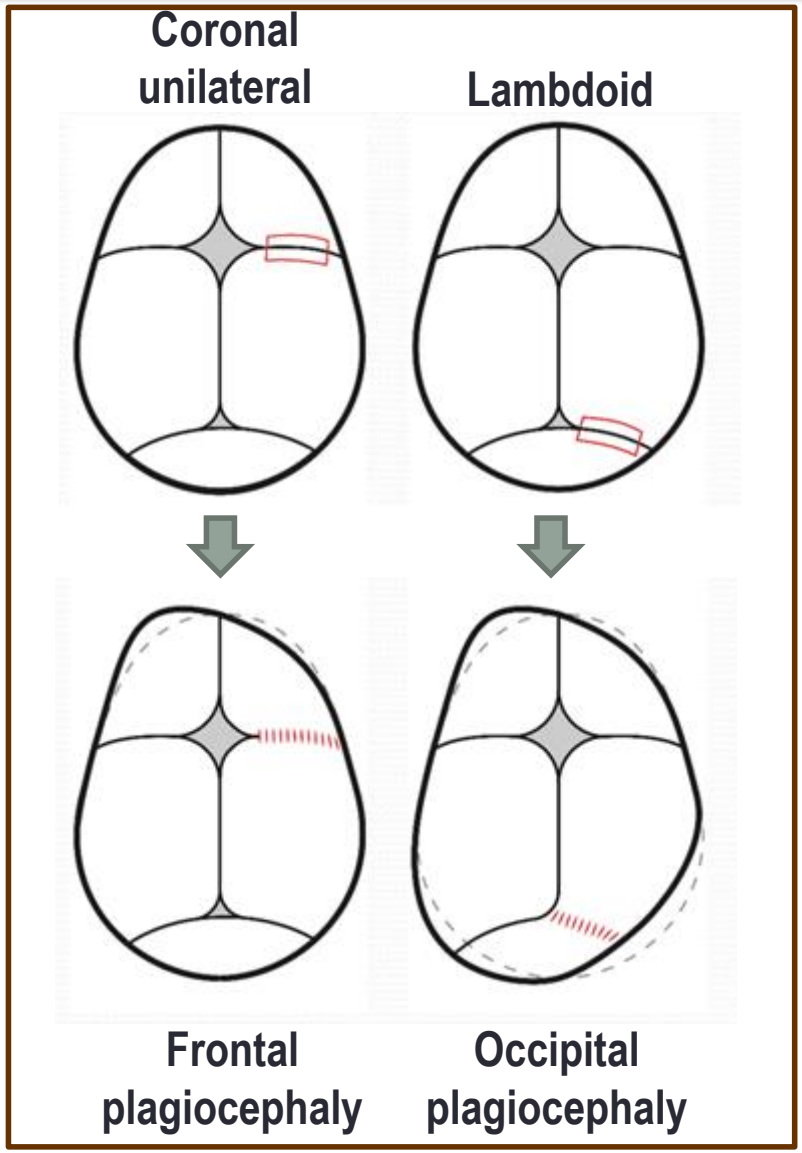


MRI



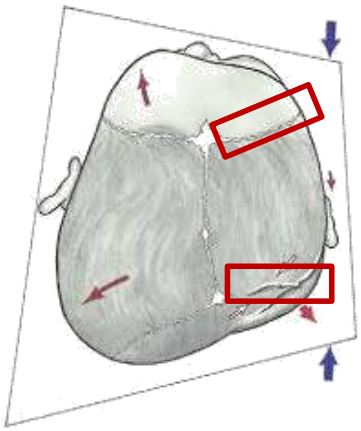
3D CT

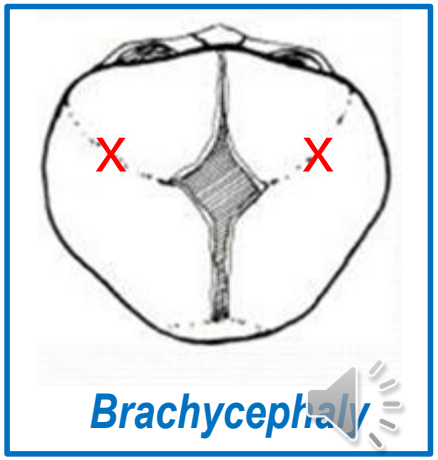
Simple craniosynostoses



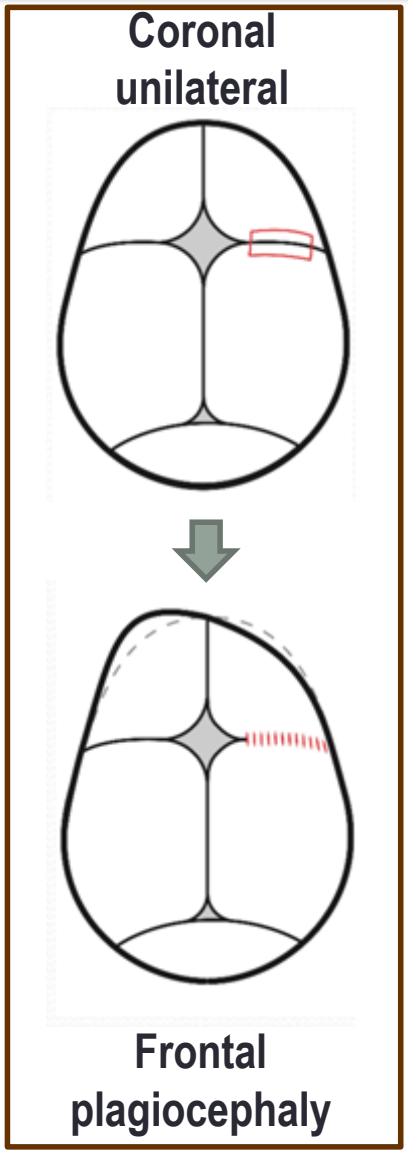
3. **Frontal plagiocephaly**
   - Closure **coronal** suture one side
   - ➢ *(Both sides = brachycephaly)*
4. **Occipital plagiocephaly**
   - Closure **lambdoidal** suture one side

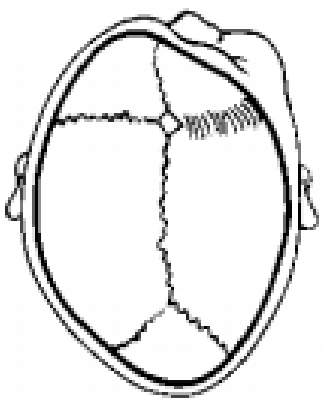
⇨ *Oblique skull, bulging of side that grows (contralateral)*

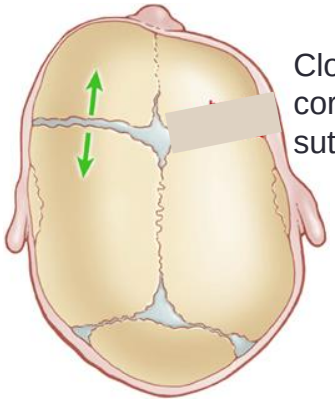




3. Frontal plagiocephaly





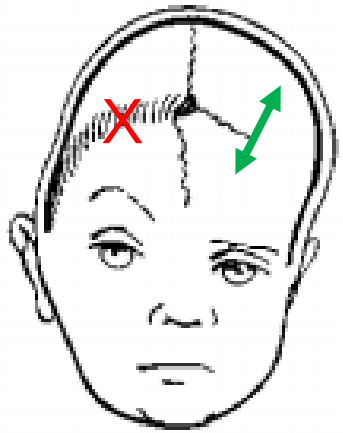


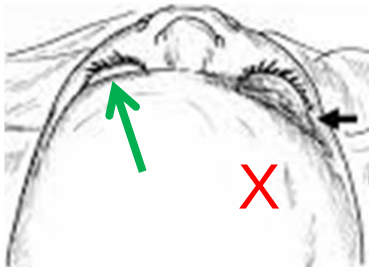
3. Frontal plagiocephaly

– Closure **coronal** suture one side

⇨ *Oblique skull, bulging of side that grows (contralateral)*

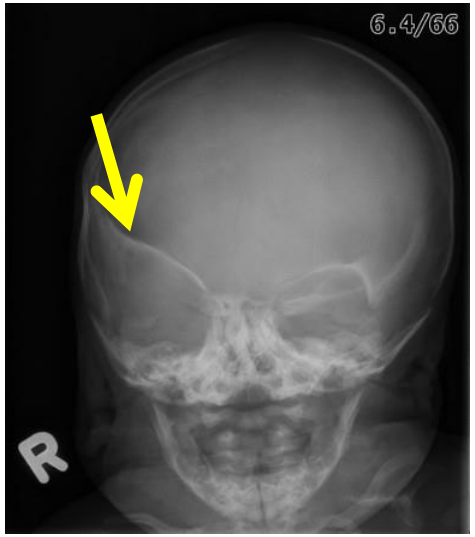
– *Bulging of frontal healthy side, facial asymmetry, harlequin eye*





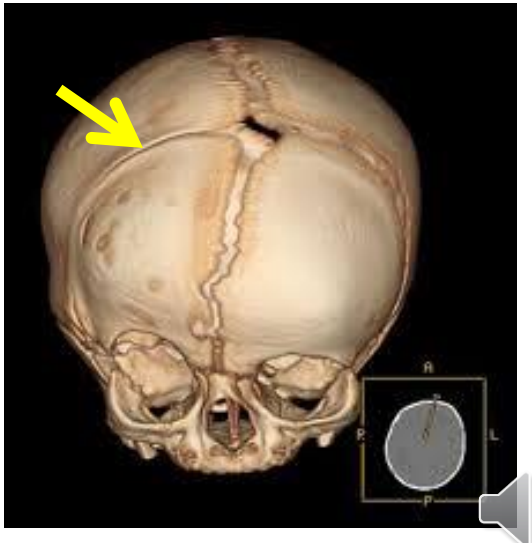
3. Frontal plagiocephaly (right side)

Simple XR

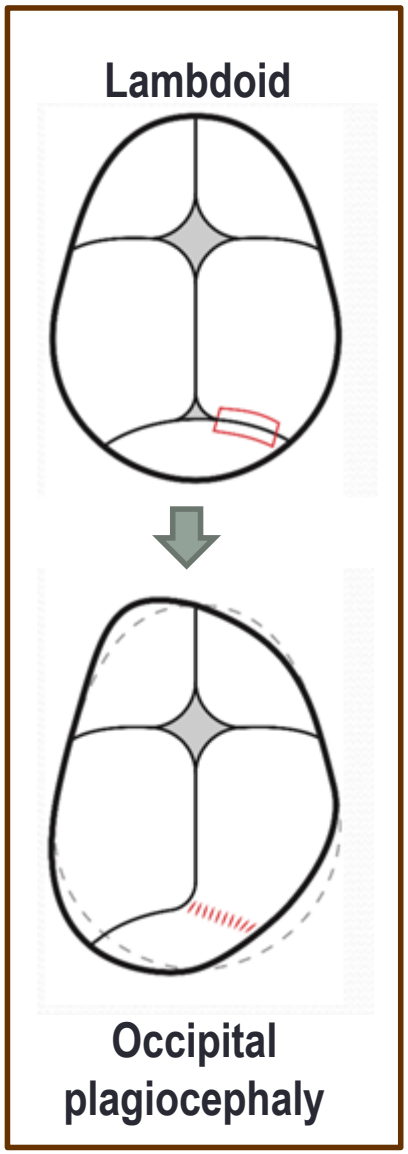


CT

3D CT



Simple craniosynostoses



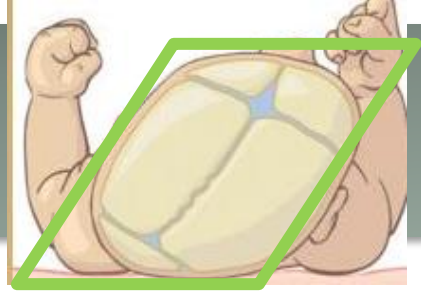
4. Occipital plagiocephaly

- Closure **lambdoid** suture one side

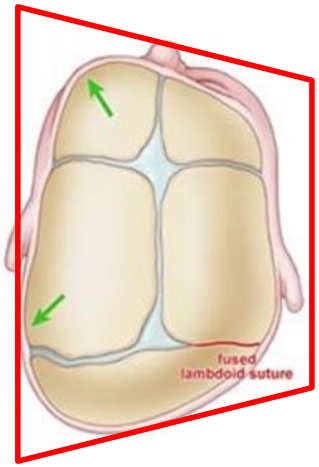
⇨*Oblique skull, bulging of side that grows (contralateral)*

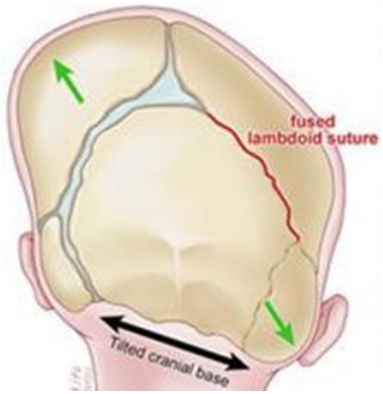
- Occipital bulging, facial asymmetry (contralateral compensatory growth)

4. Occipital plagiocephaly

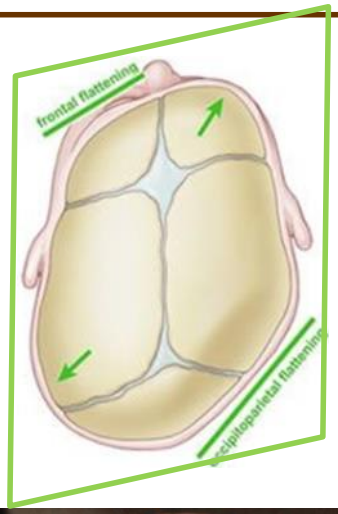


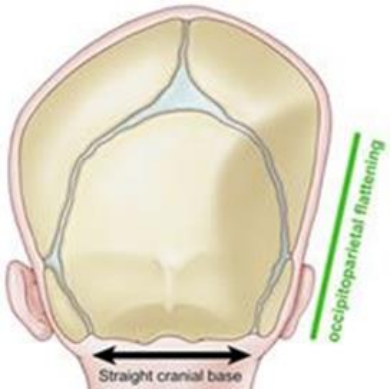
*TR*ue – *Tr*apezoid facial asymme*TR*y

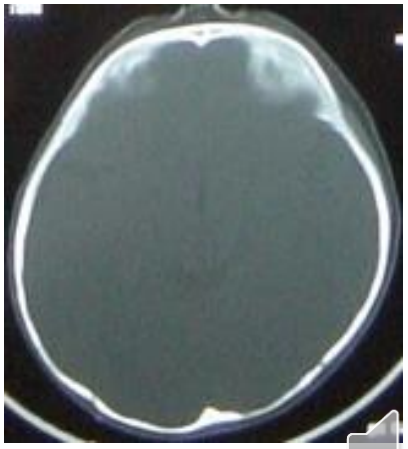




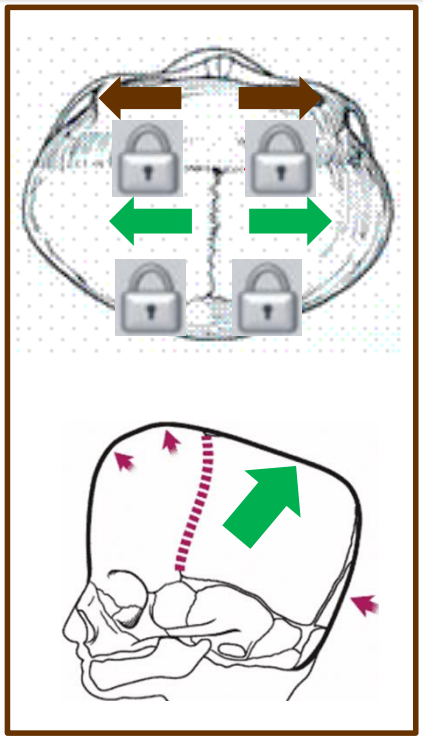
Positional (rhomboidal)



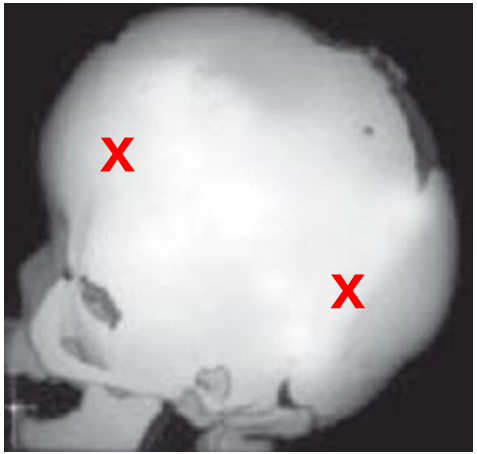




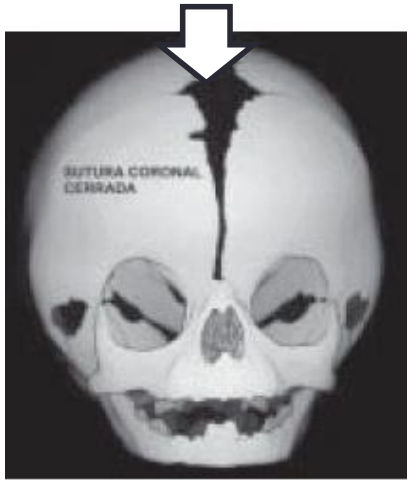
Complex craniosynostoses

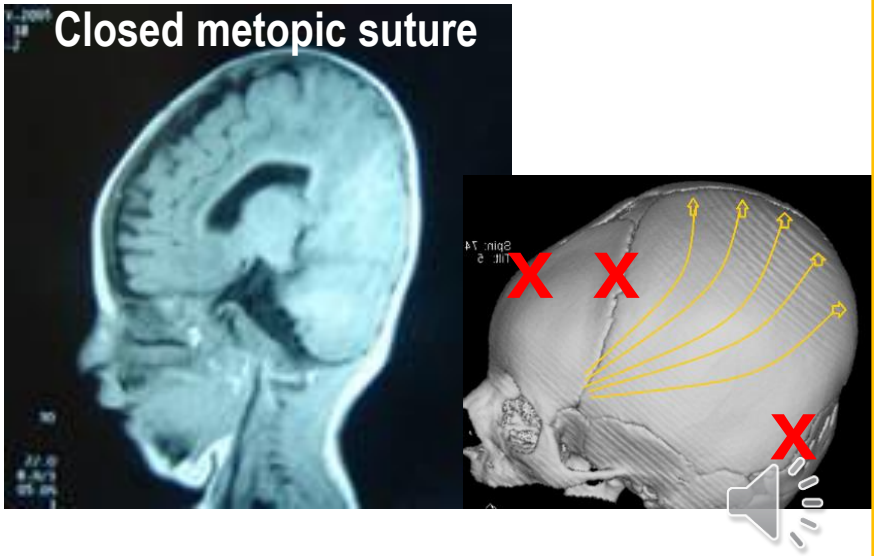


1. **Turricephaly (acrocephaly)**
   - Bilateral closure <u>coronal</u> and lambdoid (± metopic) sutures
   - Associated with syndromes
   - <u>*Open sagittal suture*</u> = skull can only grow upwards

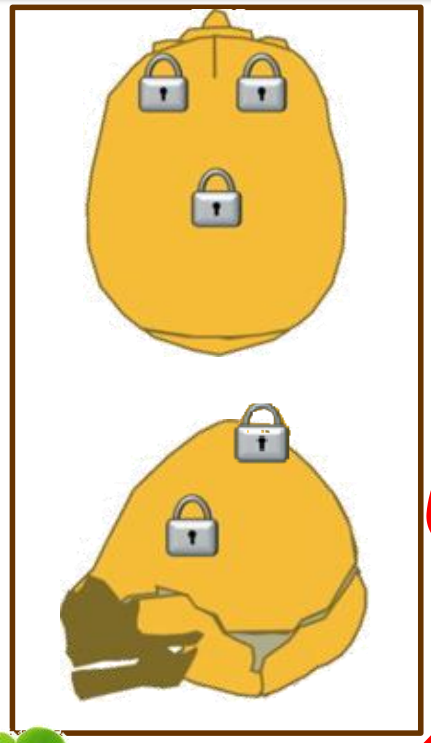


Open sagittal (and metopic) suture



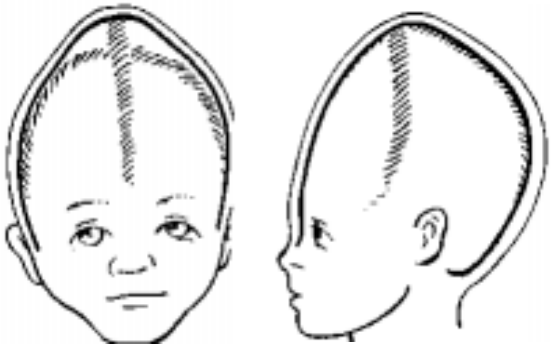


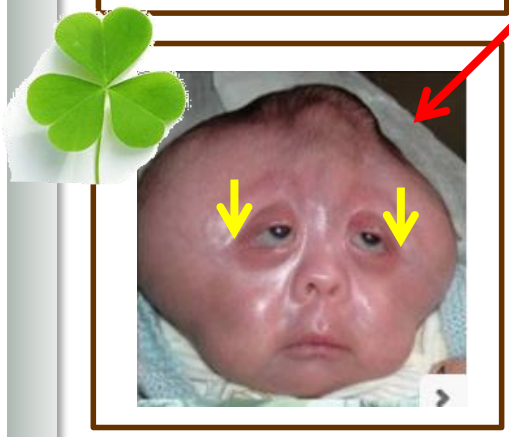
Complex craniosynostoses

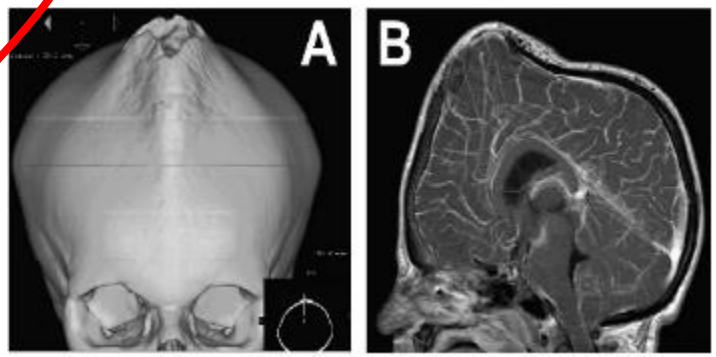


2. Oxycephaly

- Closed <u>coronal</u> and sagittal sutures
  - ➢ *"all the sutures" → cloverleaf skull*
- ↑ ICP ⇨ mental retardation + optic nerve atrophy







3D CT and MRI (4-year-old): the brain pushes towards the fontanelle

X-rays (18 months): copper beaten skull due to chronic raised intracranial pressure

Syndromic craniosynostoses

Crouzon's Syndrome

- Craniofacial dysarthrosis
- 1.6:100.000

Apert's Syndrome

- Acrocephalosyndactyly type 1
- 1.2-1.5:100.000

- Mutation gen FGFR2 (*fibroblast growth factor receptor 2*), locus 10q26 → hyperactive protein → early fusion
  - *Autosomal dominant disorder*
- Complex craniosynostosis (coronal and other) ⇨ acrocephaly
- Affects first branchial arch = medial facial area
  - *Shallow orbital socket (hypertelorism, prominent eyes, visual disturbances, strabismus)*
  - *Small pointed nose, respiratory problems (OSA in 50%)*
  - *Underdeveloped upper jaw, dental abnormalities, narrow ear canal*

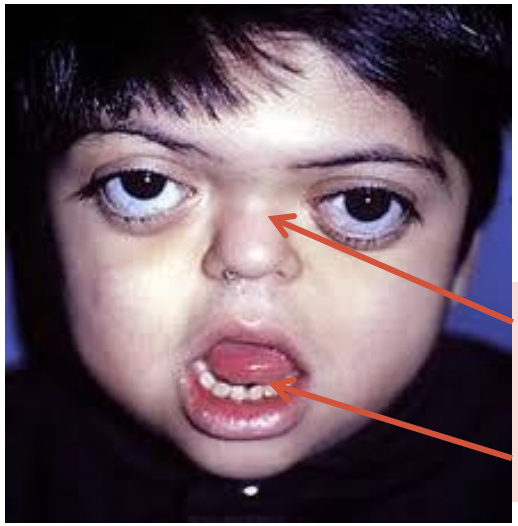
- Hydrocephaly (30 %)

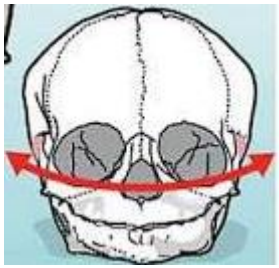
- Syndactyly 2^{nd}-3^{rd}-4^{th} fingers

Syndromic craniosynostoses

Crouzon's Syndrome

- Craniofacial dysarthrosis
- Normal intelligence
- Hydrocephalus (30 %)
  - *EARLY TREATMENT!!*

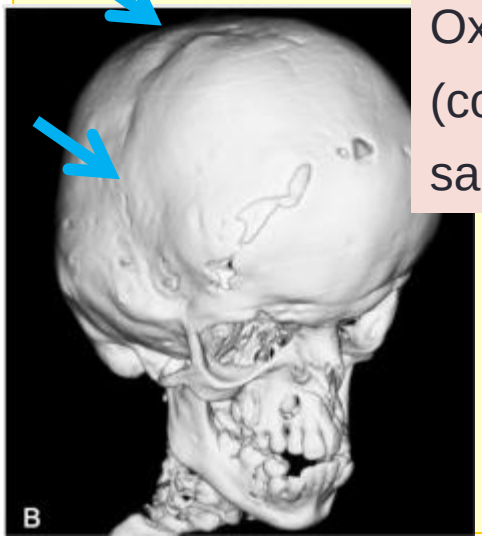




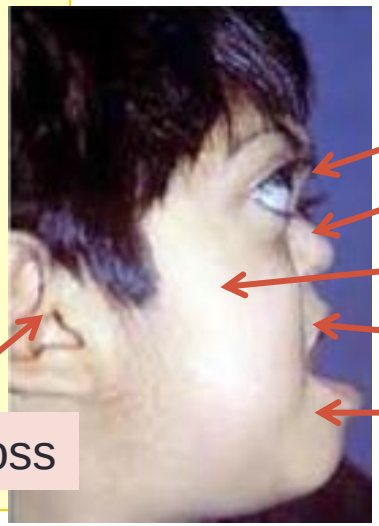
Hypertelorism

Cleft palate

Dental alt



Oxycephaly (coronal, sagittal)



Hearing loss

Exophthalmos

Pointed nose

Hypoplastic upper jaw

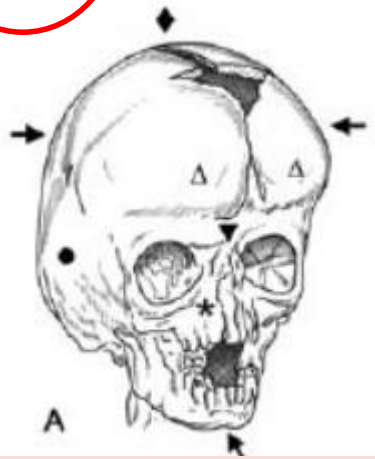
Short upper lip

Prominent inferior lip

Syndromic craniosynostoses

Apert syndrome

- Acrocephalosyndactyly type 1
- Symmetric syndactyly 2nd-3rd-4th fingers and toes
- Hyperhidrosis
- Mental retardation
- Ogival and cleft palate

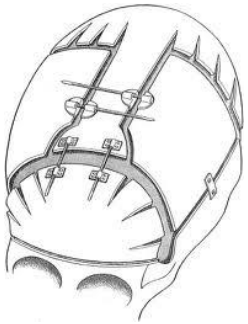


Acrocephaly (turricephaly, open sagittal suture)

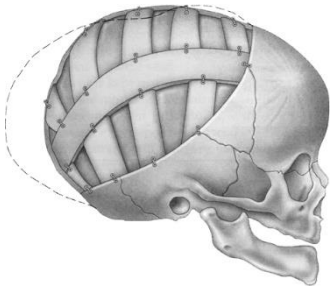
Craniosynostoses: treatment

- Conservative
  - Orthopaedic helmet (postural plagiocephaly > others)

Synostectomy



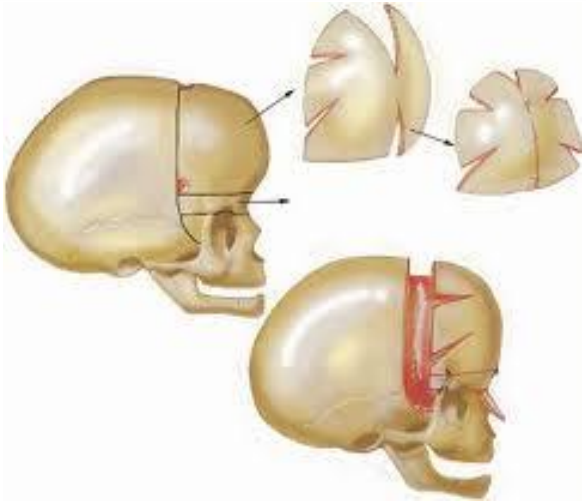
Skull expansion



Skull remodeling

- Surgical
  - Infants 3-6 months old
  - Indications
    - ICHT, optic nerve atrophy
    - Avoid psychomotor retardation (hydrocephalus, brain compensation)
    - Aesthetic
  - Options
    - Ventricle-peritoneal shunt
    - **Osteoclastic techniques**
    - Open suture = synostectomy
    - Multiple fragmentation = morcellation
    - **Remodelling techniques**

Craniosynostoses: treatment



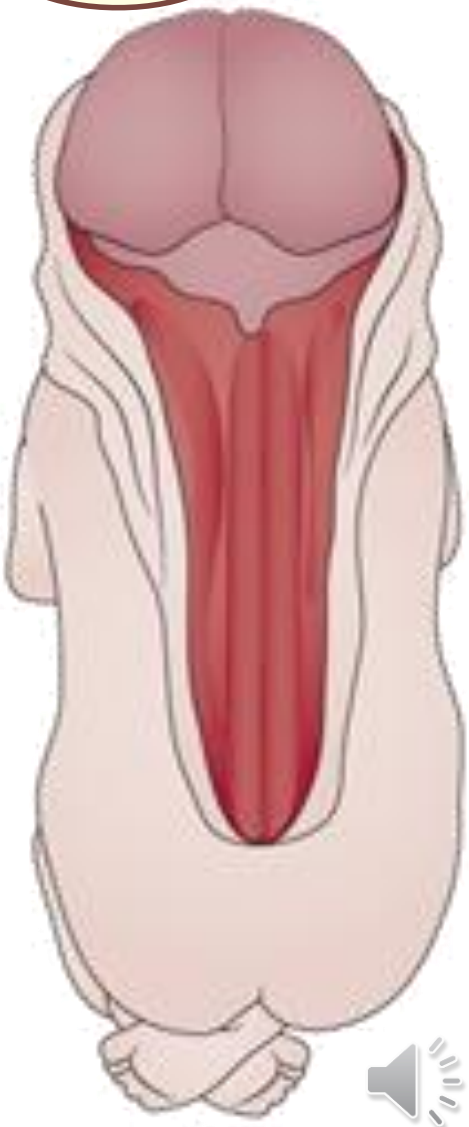
- Options
  - Ventricleperitoneal shunt
  - **Osteoclastic techniques**
  - **Remodelling (complex) techniques**
    - Craniofacial advancement (Crouzon's syndrome)

DYSRAPHISMS

REMEMBER?

- "Dysraphism" = "malformation – midline"
- Defects of fusion and formation of posterior midline (neural tube and mesoderm around it)

1:1000



➢ *It affects bone structures and nervous system*
  - Skull ± brain
  - Spine ± spinal cord

➢ *Congenital (some with genetic defect)*

7 %

Encephalocele

Anencephaly

Cranioschisis

Iliencephalocele

Craniorachischisis

Craniorachischisis

Rostral neuropore

Closure

Caudal neuropore

Occult spinal dysraphism (OSD)

50 %

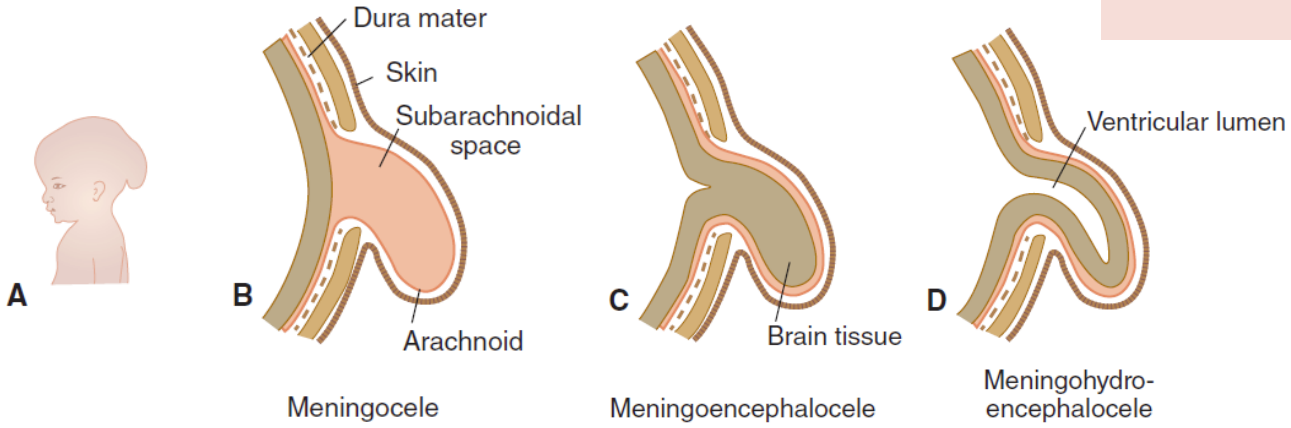
Spina bifida

Rachischisis

2. CRANIOSCHISIS

- Greek *schistós*, split
- Failure to close the rostral neuropore → Affects skull and neural tube (brain)
- Severity of **encephalocele**:
  - B. Meningocele = only meninges
  - C. Meningoencephalocele = + encephalon
  - D. Meningoencephalohydrocele = + ventricle

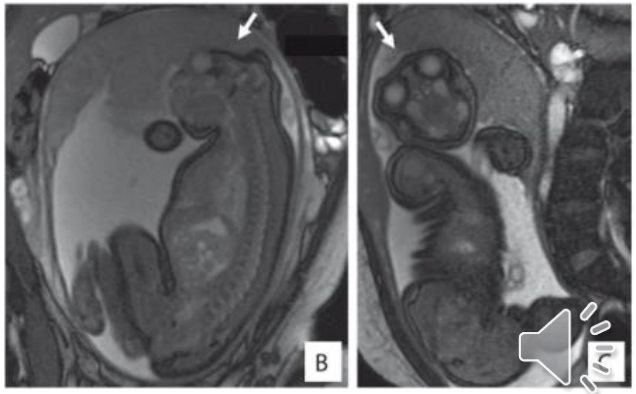
➢ ***Anencephaly*** = *meninges and "brain" open to amniotic fluid (no closure of neural tube)*



Anencephaly

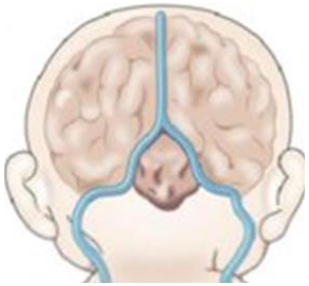
- Partial absence of encephalon
  - Brainstem is present
- Incidence “1:1000 pregnancies” (?)
- Diagnosis: intrauterus
  - **US 14 w**
    - *Absence of cranial vault > 12 sem*
  - Fetal MRI
- Prognosis: mortality 100 % < 1 yr
  - Intrauterine death (23 %) / perinatal (35 %) / < 7 dol (99 %)
  - *Survival depends on amount and viability of brain that remains*

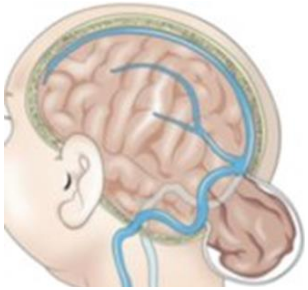
Up: 1st trimester echography (10 w)
Down: MRI 2nd trimester (diag 20 w)

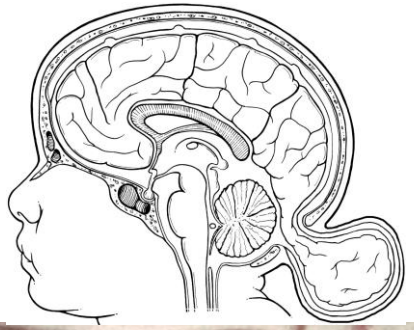


Encephalocele

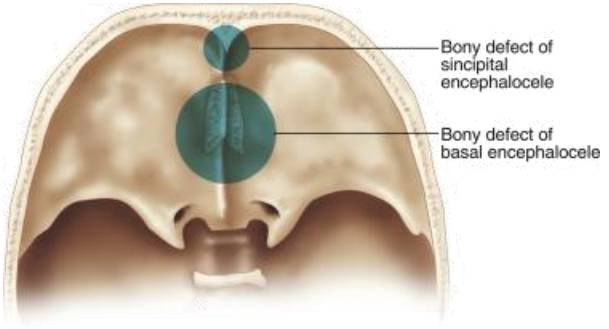
- 1 : 5-10.000 live newborns (F > M)
- Occipital (75 %) > frontobasal (15 %, anterior) > other
- Prognosis depends on size, location, involvement of nervous tissue, covering, and association with syndromes
  - Mortality ⇧

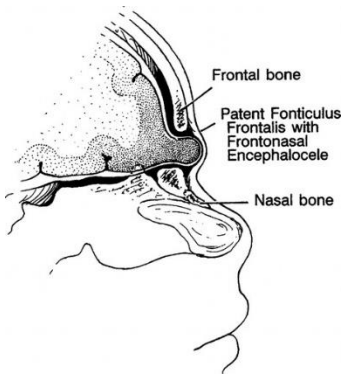






Occipital encephalocele





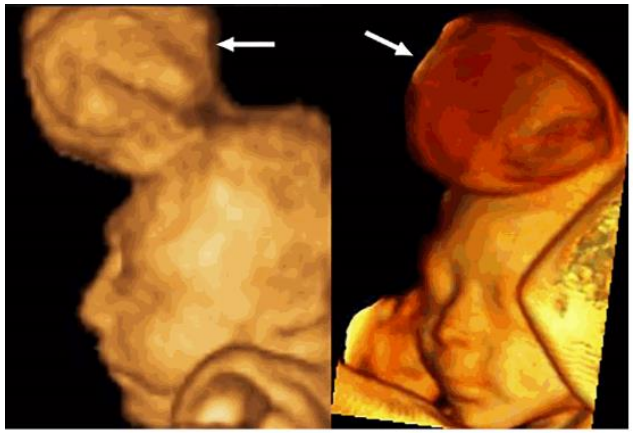
Frontobasal encephalocele

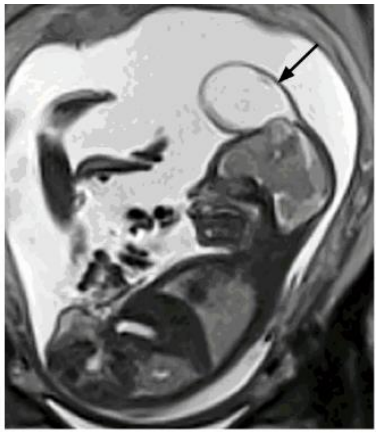
Encephalocele

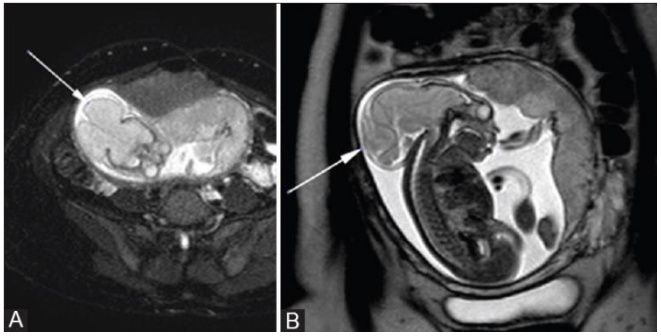
- Diagnosis
  - Fetal US ⇨ Abortion?
  - Fetal / newborn MRI

Fetal ultrasound (11 weeks)
occipital encephalocele

Differential diagnosis: Echo 3D and MRI frontal *meningocele* (26 weeks)

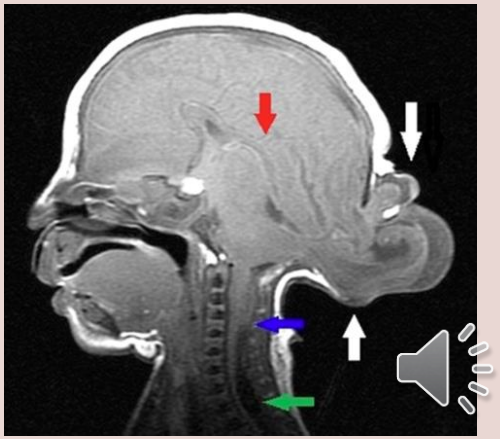






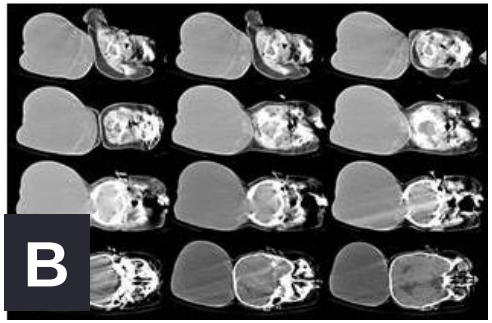
Fetal MRI
(22 w)

Newborn
MRI

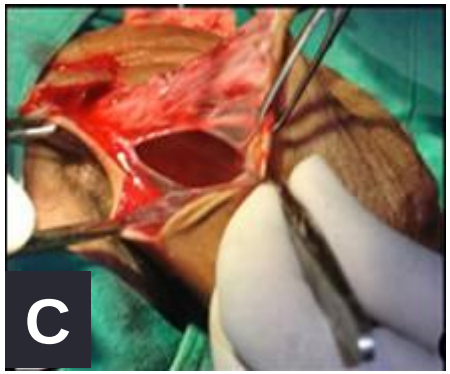


Encephalocele

- Treatment = repair and closure of the defect
  - Goal = preserve healthy brain tissue
  - Variable results but mortality ⇧
    - *Meningocele 11 %*
    - *Meningoencephalocele > 70 %*



A.3-year-old boy with frontal encephalocele.
B. Same boy, 3 months after surgery.

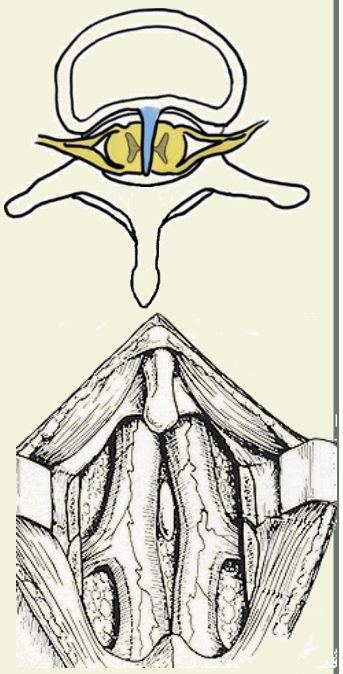


A. Girl 4 months, neurologically normal.
B. CT: Encephalocele with herniation of a thin layer of brain tissue inside the sac. Occipital bone defect.
C-D. Surgical repair

3. RACHISCHISIS

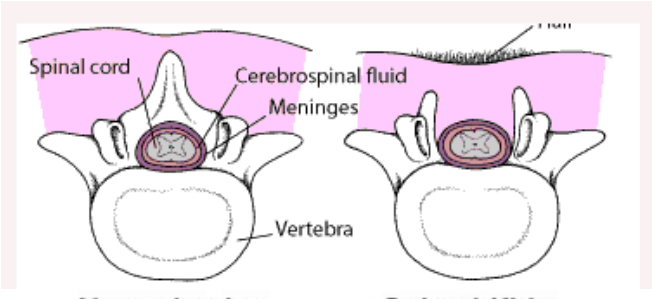
- Closure defects of the neural tube at the level of the spine
  - *Open (80 %) or closed*
- Incidence 1-2 : 1.000 live newborns
  - Most common malformation of CNS
- Types of spina bifida:
  - Spina bifida occulta = vertebral arches (associates skin alt)
  - Meningocele = Meninges
  - Myelomeningocele = + Spinal cord **Open**
  - Lipomyelomeningocele = + Adipose tissue
  - *Myeloschisis* = open and flattened spinal cord in thoraco-lumbar region **Open**
  - *Diastematomyelia* (greek *diástēma, 'interval, distance')*

Myeloschisis



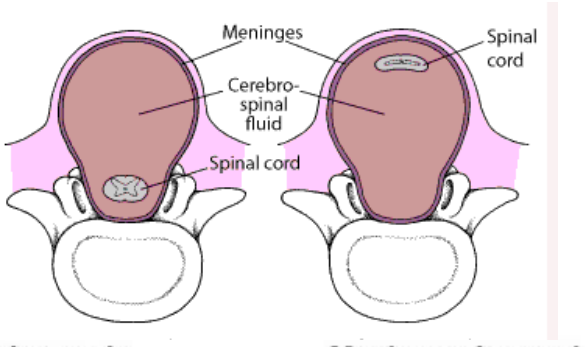
Diastematomyelia

Spina bifida



Normal spine

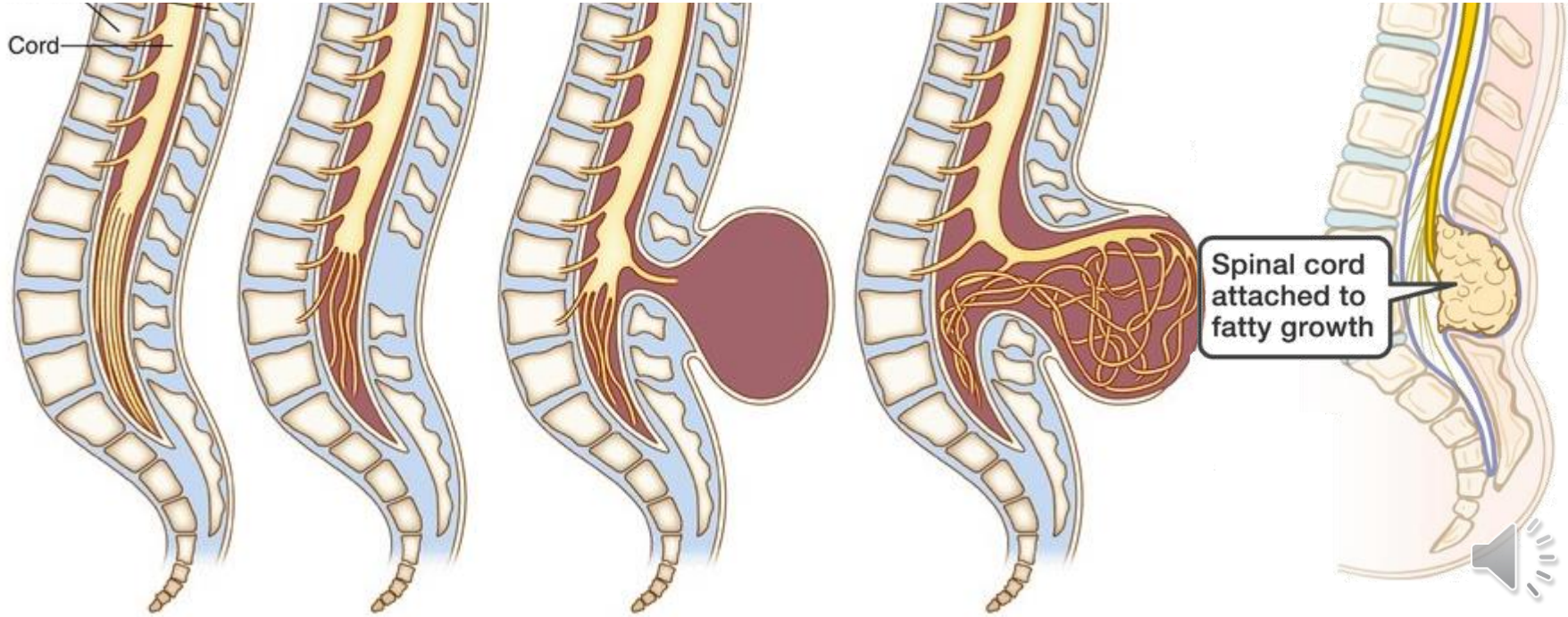
Spina bifida occulta



Meningocele

Myelomeningocele

Lipomyelomeningocele

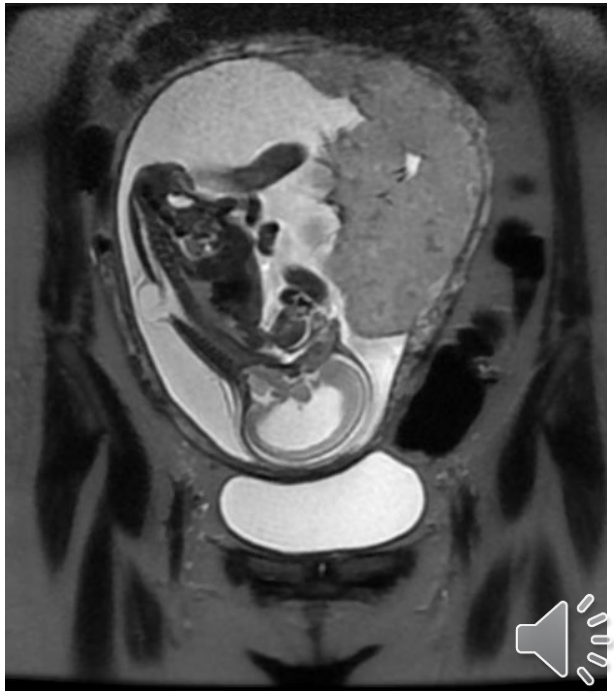


Spina bifida

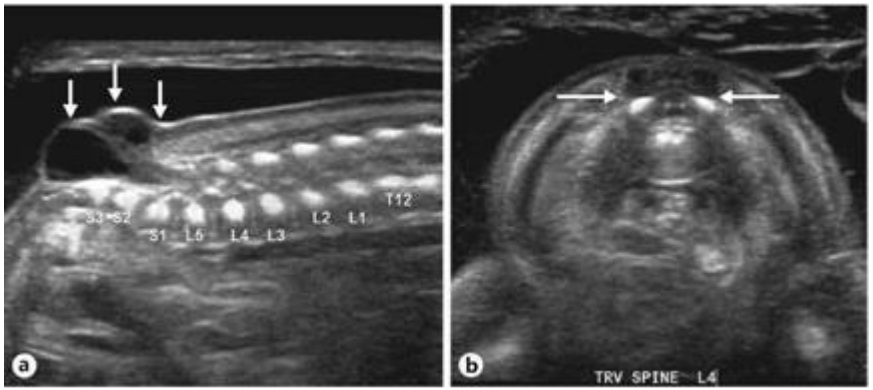
- Prenatal diagnosis:
  - Suspicion: ↑ Alfa-fetoprotein in maternal serum 2nd trimester
  - Fetal US 2nd trimester (18-22 week): precise diagnosis
  - Fetal MRI

2D and 3D fetal ultrasound, **11 w** (trisomy 18)

Fetal MRI
22 w

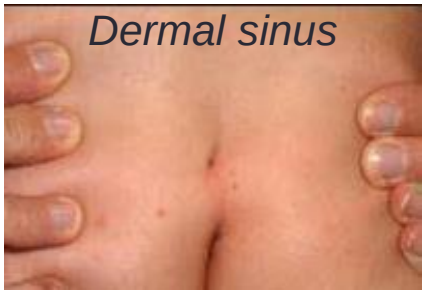


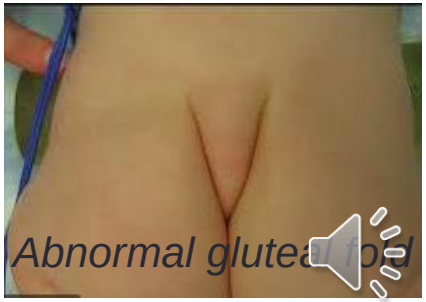
Fetal ultrasound 19 weeks: Myelomeningocele L4-S4



Spina bifida

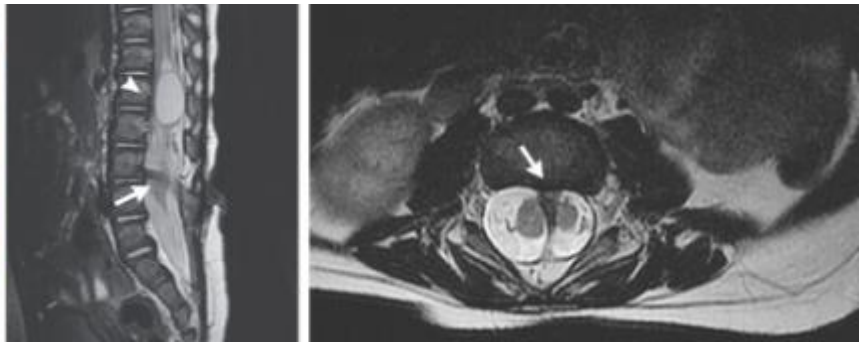
- Postnatal diagnosis:
  - Clinical features:
    - *Lump*
    - *Suspicious skin lesion: port wine stain, hypertrichosis, hemangioma, dermal sinus, abnormal gluteal fold, vestigial tail …*
    - *Progressive neurological disorder, other malformations (club feet)*
  - Image: MRI



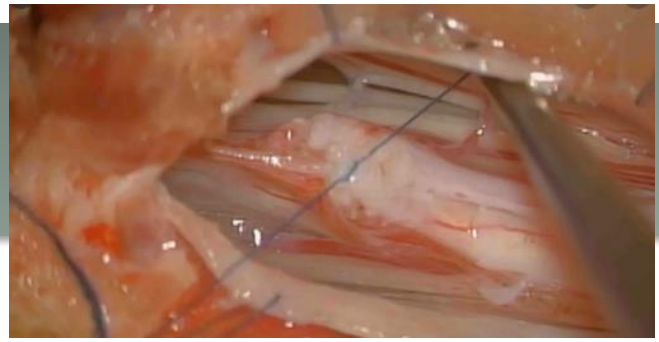


Hypertrichosis

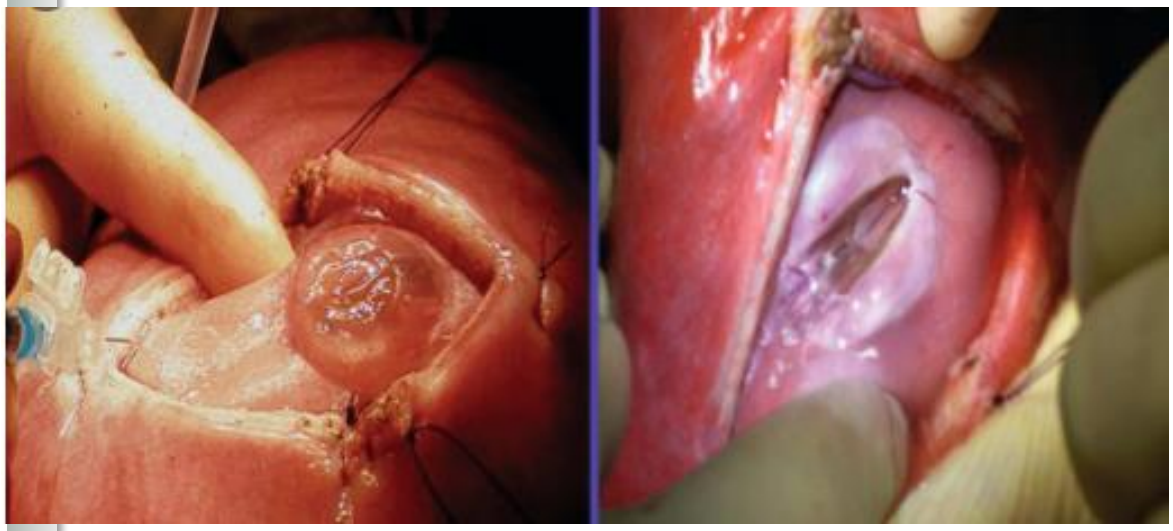
Diastematomyelia



Spina bifida

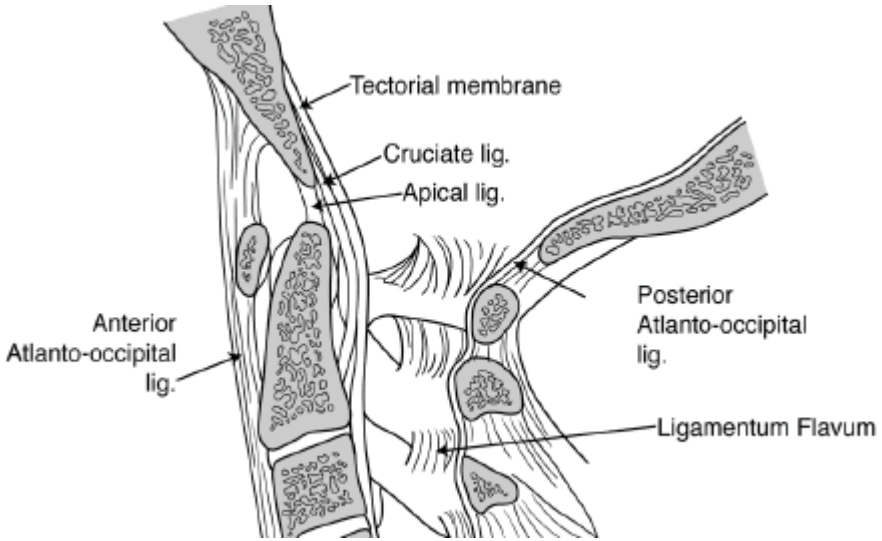


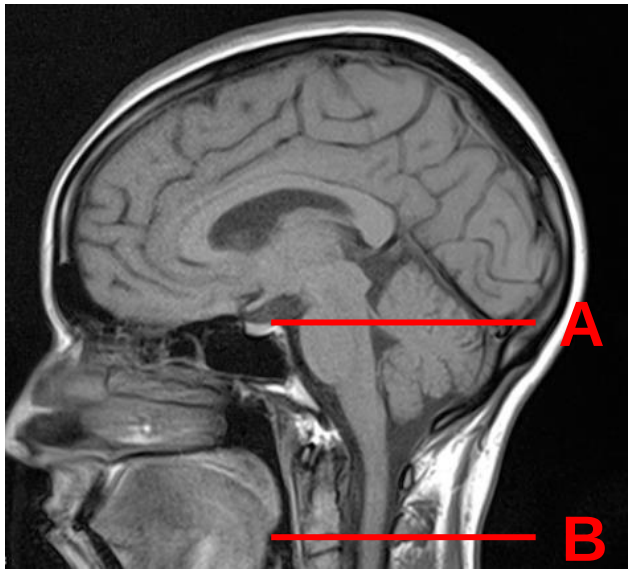
- Treatment: EARLY surgical
  - Meningocele: Dura + soft tissue repair
  - Myelomeningocele: + spinal cord
    - *Intrauterine surgery*
    - *When open, URGENT!*
  - Lipomyelomeningocele: + detach spinal cord
  - Diastematomyelia: + remove bone spike



4. CRANIO-CERVICAL JUNCTION

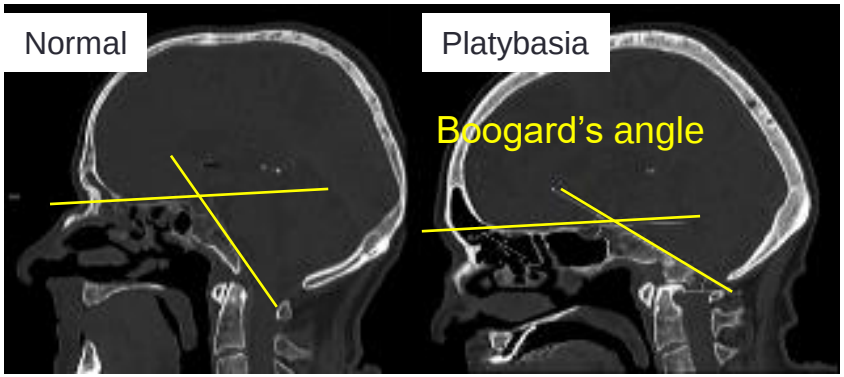
- Cranio-cervical junction (CCJ)
  - Space between the lower portion of the occipital bone around the foramen magnum (A) and the first two cervical vertebrae (B)
  - Funnel where the cerebellum rests and the brain stem joins the spinal cord

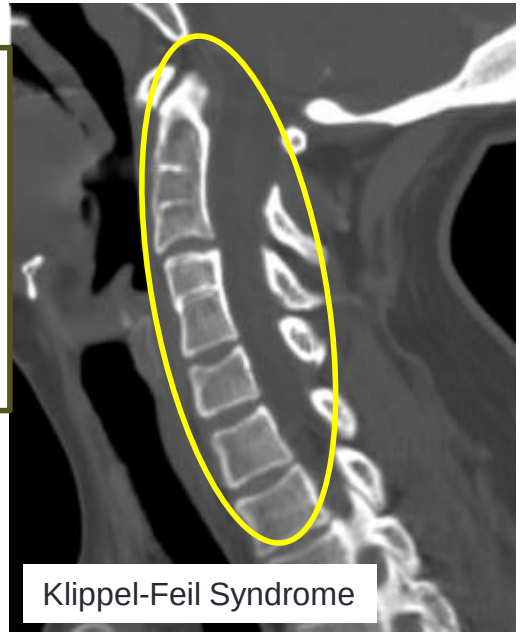


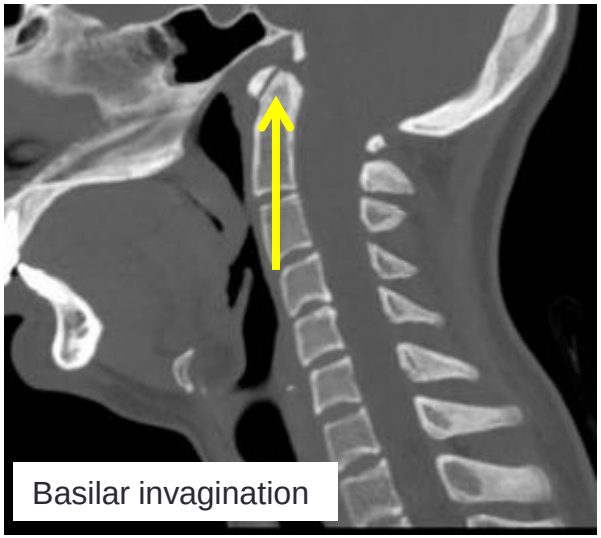


Cranio-cervical junction malformations

- Bone malformations
  - Platybasia
  - Basilar invagination
  - *Other CCJ malformations*
  - Klippel-Feil syndrome
- Neurological malformations
  - (Arnold-) Chiari malformations
  - Syringomyelia



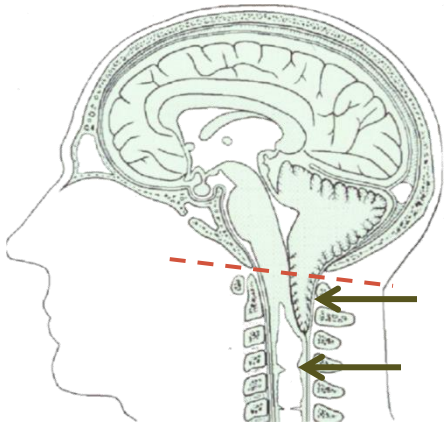




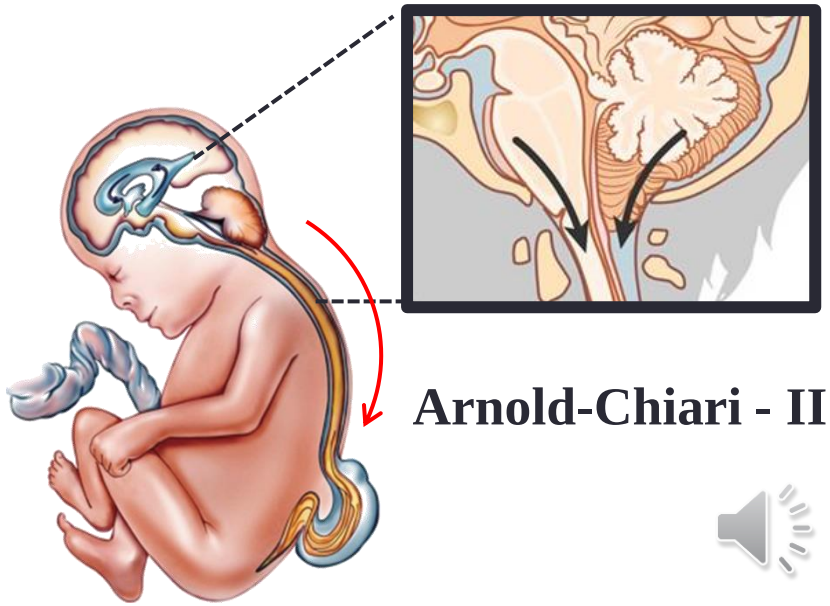
Chiari malformation

- Displacement of cerebellar tonsils > 3 mm through foramen magnum
  - **Chiari malf** – type I – Adults, incidental (5-30:1.000) – Only tonsils/cerebellum – Associated with syringomyelia + other malf skull base (short neck 25 %) or spine (kyphoscoliosis)
- **Arnold-Chiari malformation** – type II – Children (0'4:1.000) – Also involves brain stem – Associated with myelomeningocele and hydrocephalus

Normal



Chiari - I



Arnold-Chiari - II

Chiari type I malformation

- 90 % asymptomatic
  - Displacement of tonsils
    - *3 – 5 mm → asymptomatic*
    - *5 – 10 mm → symptoms in 30 % (syringomyelia)*
    - *> 12 mm → "always symptoms"*

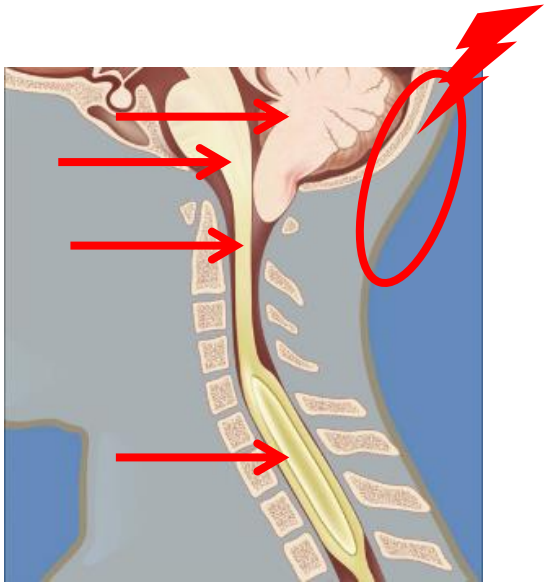
- Clinical features: woman 25-40 years old
  - Occipitocervical pain (80 %) that ↑ with Valsalva (cough)
  - Syringomyelia (40-75 %), paresthesia (60 %)

- Physical exam
  - 25 % short neck
  - Central cord syndrome (syringomyelia)
  - Compression of nerve structures

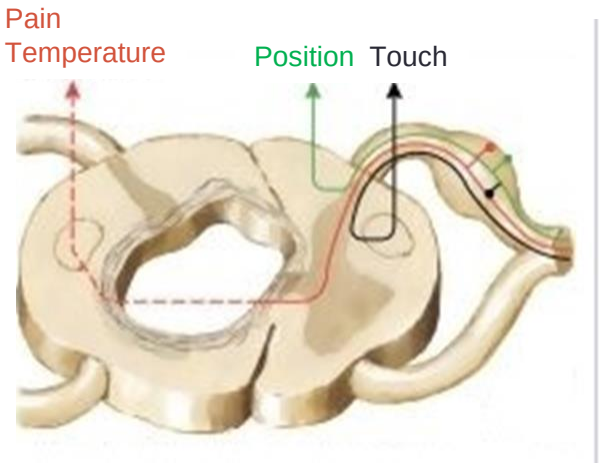
Chiari type I malformation

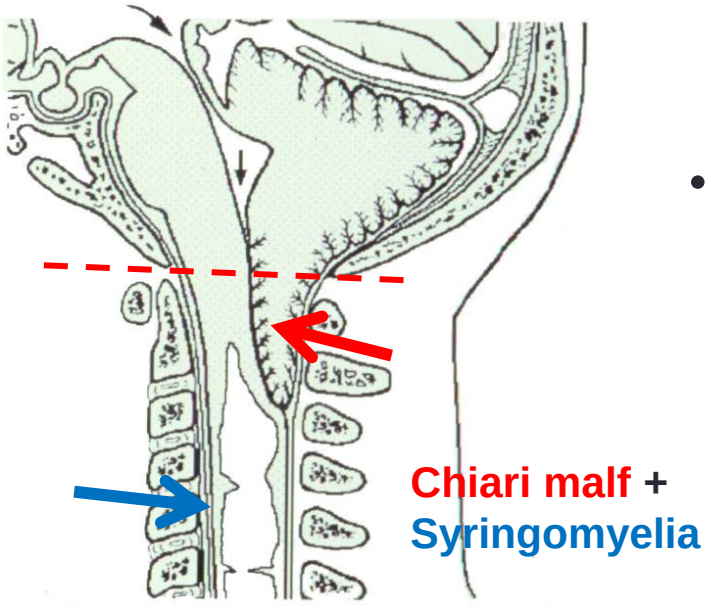
- Pathophysiology and clinical features
  - Altered CSF dynamics:
    - ***Headache or occipitocervical pain*** *(80 %) that ↑with Valsalva (cough) > vertigo, tinnitus, diplopia*
  - Brainstem compression:
    - ***Paresthesias*** *(60 %), spasticity (40 %, hyperreflexia upper arms), drop attack, apnoea*
    - *Lower cranial nerves (15-25%): absence of gag reflex > dysarthria, dysphonia*
  - Cerebellar syndrome (11-75 %):
    - *Trunk ataxia optokinetic nystagmus, dysmetria*
  - **Centro-medullary syndrome** (syringomyelia) (40-75%)

➢ *Lhermitte's sign: "electrical discharge" from neck to the arms and torso on head flexion*



Syringomyelia

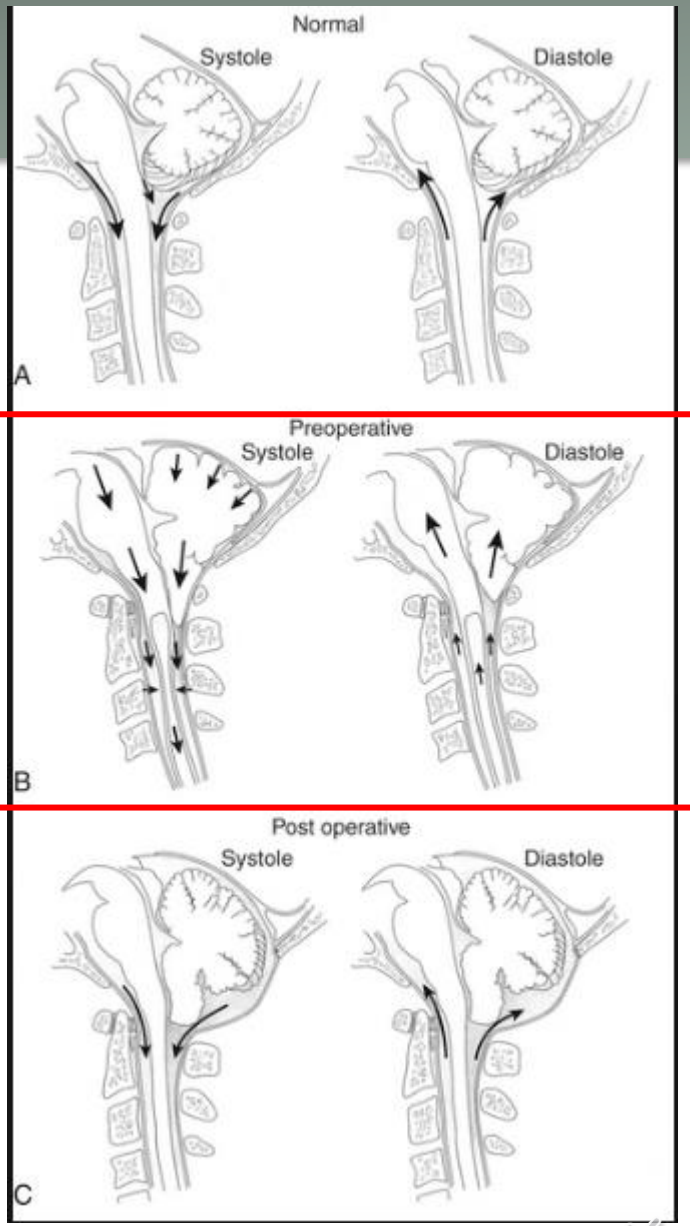


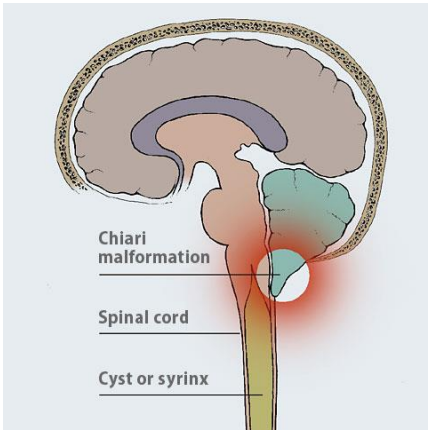


- Cysts (syringo, syrinx) along the center of spinal cord
  - 90% syringomyelia is associated to Chiari or other CCJ malformations
    - *Rest: Tumoral, traumatic, infection…*
    - *In posterior fossa or spinal cord*
  - Located cervical-dorsal
    - *Medulla oblongata: syringobulbia*
- Progressive, never reverts

Syringomyelia

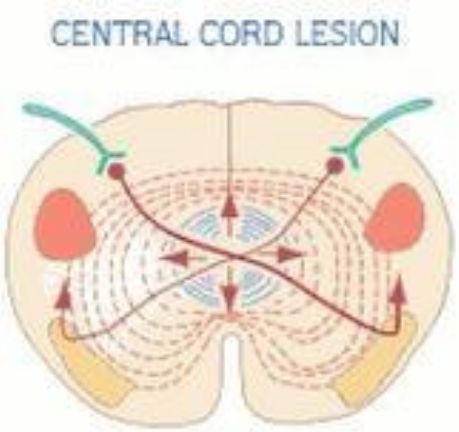
- Pathogenesis: hydrodynamic theory
  - Subarachnoid block of CSF circulation ⇨ ↓ buffering capacity of CSF pressure waves with each cardiac systole
    - *In Chiari, blockage at the foramen magnum*
  - The pulsatile increase in pressure would eventually cause formation of a cyst = syringomyelia

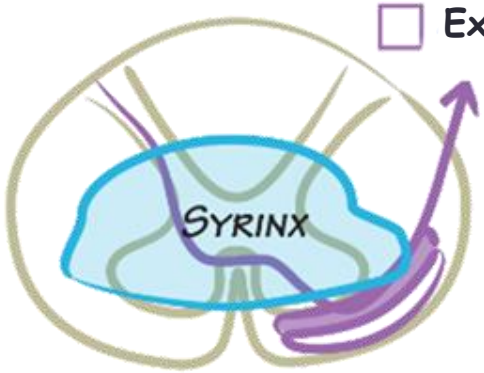


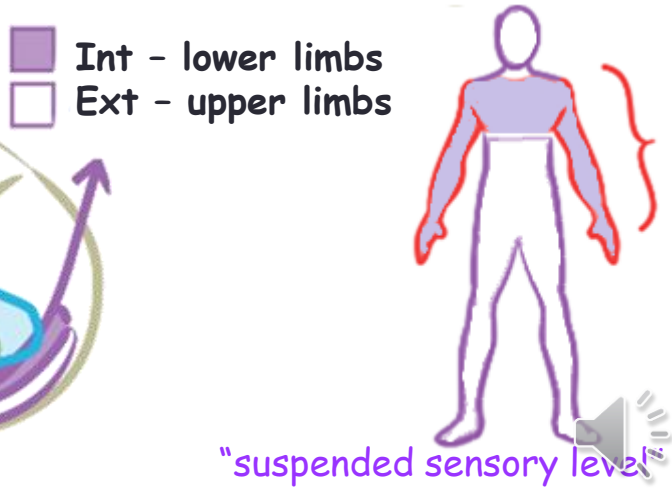


Syringomyelia

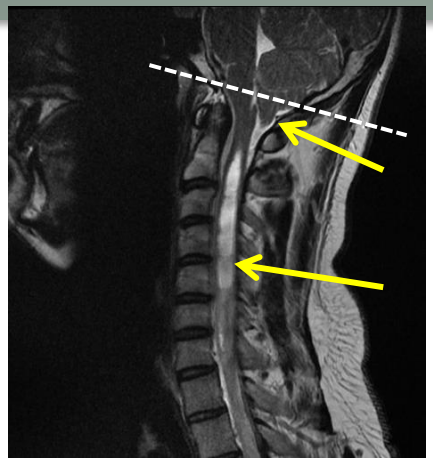
- Clinical features:
  - Presentation: woman, 25-40 years old
  - Central medullary syndrome
    - *Thermoalgesic dissociation*
    - *Inadvertent burns*
    - *Distal muscle atrophy upper limbs*
    - *Loss of reflexes and strength, muscle stiffness*
  - Syringobulbia: affects cranial nerves IX-XII



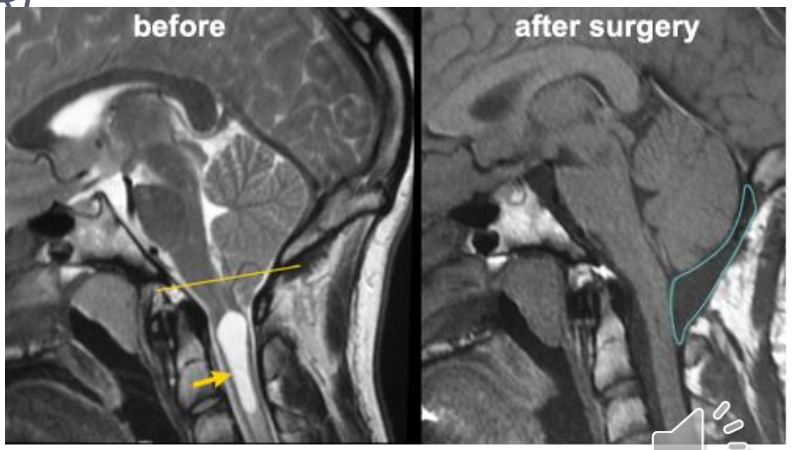




Cranio-cervical junction malformations



- Diagnosis
  - Clinical examination
    - *Associated malformations*
    - *Signs and neurological deficits*
  - MRI
    - *Associated bone malformations*
    - *CNS malformation or damage*
    - *Volumetric and CSF flow dynamics studies*
    - *Syringomyelia: complete spinal cord MRI*
- Treatment: surgery
  - CCJ decompression
  - Other pathologies
    - *Myelomeningocele*
    - *Only syringomyelia: syringopleural shunt*



SUMMARY KEY CONCEPTS TOPIC 2

- Hydrocephalus
  - Increase in intracranial CSF
  - Evolves to cerebral atrophy, psychomotor retardation, and blindness
- Craniosynostosis
  - Simple = aesthetic defect / avoid psychomotor retardation and blindness
  - Complex and syndromic = other malformations
- Cranioschisis and spina bifida
  - Need surgical repair (except occulta asymptomatic)
  - Prognosis depending on nervous tissue damage
- Cranio-cervical junction malformations
  - Chiari-I + symptomatic ⇨ CCJ decompression
  - Syringomyelia ⇨ treatment of its cause

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Recommended videos in English (1)

- **Hydrocephalus**
  - https://www.youtube.com/watch?v=JLNI2upLi7I
  - https://www.youtube.com/watch?v=Vjg99T8MALE
  - https://www.youtube.com/watch?v=bQCgIthM01I
- **Spinal dysraphysm/spina bifida**
  - https://www.youtube.com/watch?v=jlDZA2PNW2o
  - https://www.youtube.com/watch?v=vnJIzbDvxBs
  - https://www.youtube.com/watch?v=7eV3DXyBiNI
  - https://www.youtube.com/watch?v=jlDZA2PNW2o&t=30s

Recommended videos in English (2)

- **Craniosynostosis**
  - https://www.youtube.com/watch?v=RQYPgwVzzxI
  - https://www.youtube.com/watch?v=-tyE7XKodJg
- **Chiari Malformation**
  - https://www.youtube.com/watch?v=ImWtvtSQx50
  - https://www.youtube.com/watch?v=dHM5sDaHskY
  - https://www.youtube.com/watch?v=yJ2nVrhIQEo
- **Syringomyelia**
  - https://www.youtube.com/watch?v=KLH-3SzsPYM
  - https://www.youtube.com/watch?v=nAJy1JSXHCI
  - https://www.youtube.com/watch?v=RBMxQRicVCk

Recommended videos in Spanish (1)

- **Hidrocefalia**
  - https://www.youtube.com/watch?v=Id2KoXsJ9_0
  - https://www.youtube.com/watch?v=mTvabN5TpyQ
  - https://www.youtube.com/watch?v=zIX6EVhFBUc
- **Defectos del tubo neural**
  - https://www.youtube.com/watch?v=1m6iyrUmZp4
  - https://www.youtube.com/watch?v=xzgkznTDHsE
  - https://www.youtube.com/watch?v=NLReOYkk4J8
- **Espina bífida**
  - https://www.youtube.com/watch?v=F4NNjD4utks
  - https://www.youtube.com/watch?v=U39ym-vSCM0
  - https://www.youtube.com/results?search_query=espina+bifida+espa%C3%B1ol

Recommended videos in Spanish (2)

- **Craneopatías**
  - https://www.youtube.com/watch?v=HHgOIcdc_yQ
  - https://www.youtube.com/watch?v=MRZHwbK3GBA
- **Malformación de Chiari**
  - https://www.youtube.com/watch?v=wFJ7JSyt6JA
  - https://www.youtube.com/watch?v=J-m_Ikwl-h8
  - https://www.youtube.com/watch?v=LxatScJpZmE
- **Siringomielia**
  - https://www.youtube.com/watch?v=vnUA69xopFk
  - https://www.youtube.com/watch?v=XU9IHtes_3M&t=39s

Recommended videos in German

- **Hydrocephalus**
  - https://www.youtube.com/watch?v=YhKhZF0xI0Q&t=170s
  - https://www.youtube.com/watch?v=9aOdmQ2b9wg&t=2s

QUESTIONS?

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