Hydrocephalus & Common Neurosurgical Congenital CNS Malformations

With all courtesy to our colleagues, Raslan and his team, a lot of our work is based on their Manual to Surgery Booklet.

- **Important**
- Mentioned by doctors but not in slides
- Additional notes from Surgical Recall 6th edition or Raslan's booklet
- Not mentioned by the doctor

431 SURGERY TEAM

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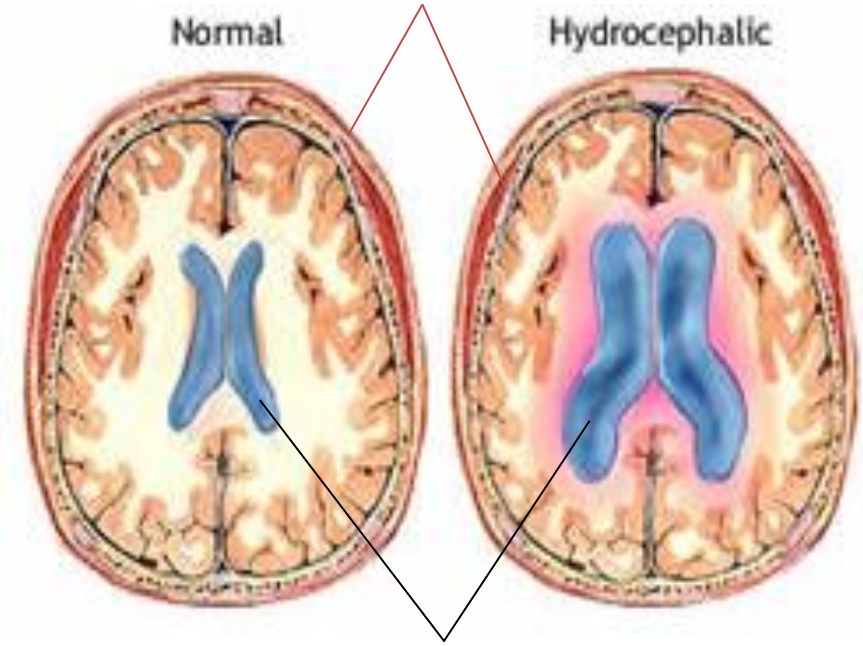
Cephalus; Head



Hydrocephalus =

(Hydro = Water)

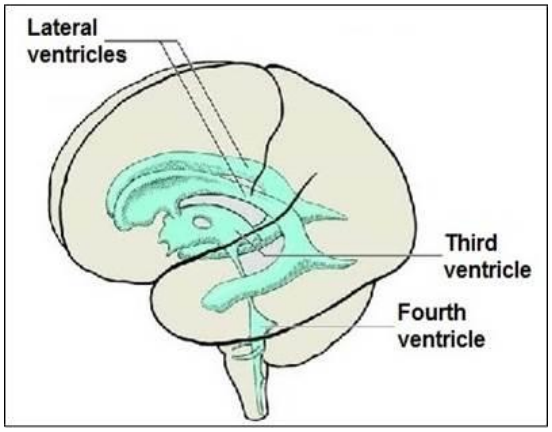
+

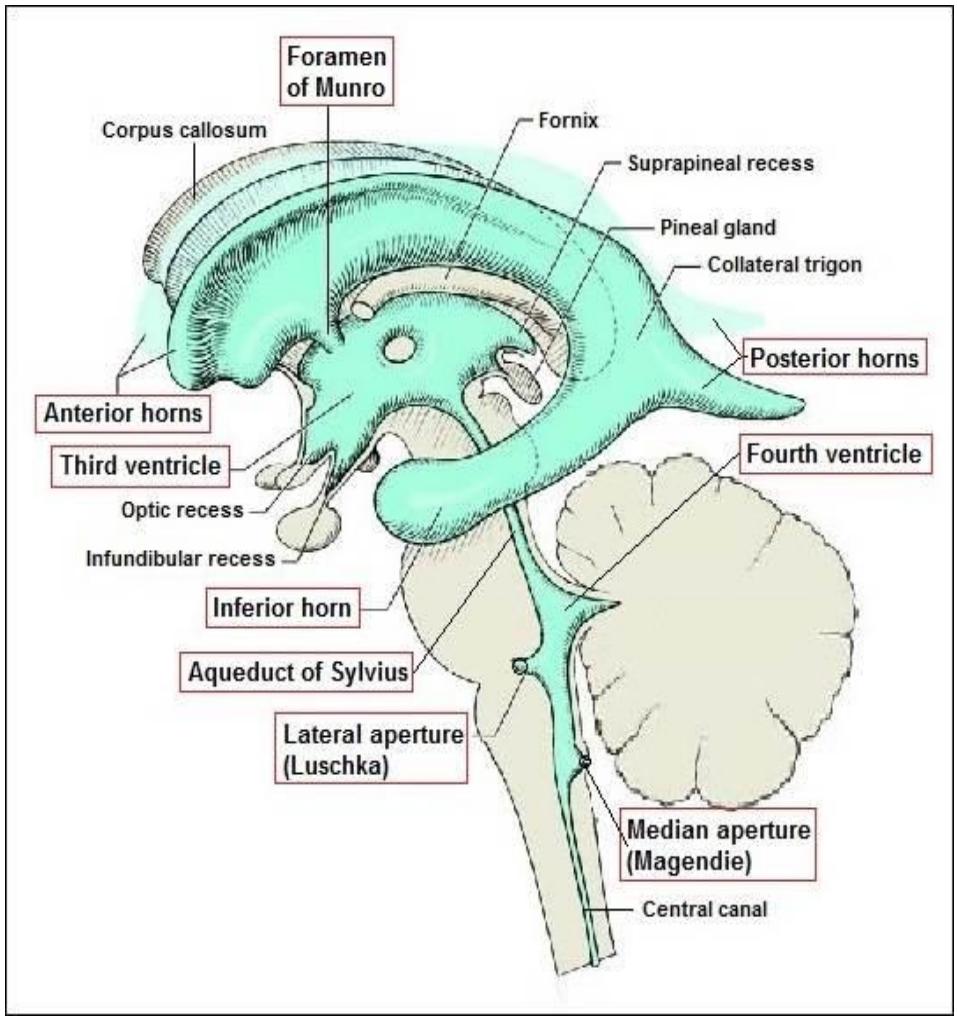
(Cephalus = Head)

- Hydro; Water; ↑ CSF in the head causes Ventriculomegaly & ↑ ICP

Anatomy

of The Ventricular System





Physiology of CSF Production:

- Total volume of CSF in the ventricles ranges from ~10 ml in neonates to ~150 ml in adults.
- Produced mainly by the choroid plexus & to a lesser extent by extracellular fluid of the brain.
- Rate of production is 0.3-0.4 ml/minute (~500 ml/day).
- Only very high ICP will reduce CSF production; usually when brain perfusion is decreased.

Physiology of CSF Flow & Absorption:

- CSF Flow:
  - ☐ Through anatomical CSF spaces
- CSF Absorption:
  - ☐ Mainly at the arachnoid granulations

Pathogenesis of Hydrocephalus

✓ Excessive CSF production:

Choroid plexus papilloma

✓ CSF flow obstruction:

Tumors: especially in or near the ventricules

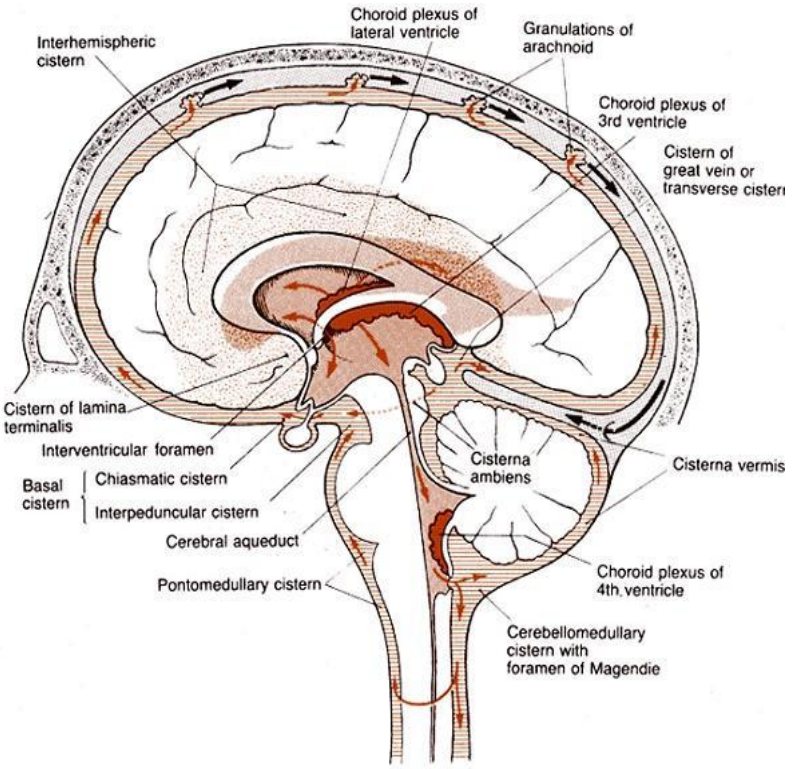
Congenital anomalies

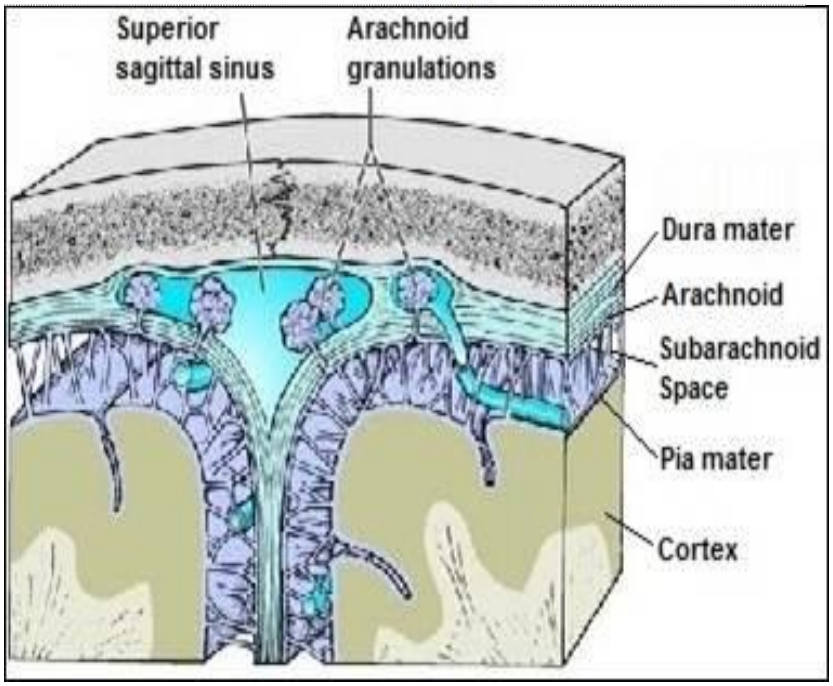
✓ Decreased CSF absorption:

Post meningitic

Post SAH

✓ Decreased brain volume: Brain atrophy: Hydrocephalus ex vacuo





Causes of Hydrocephalus

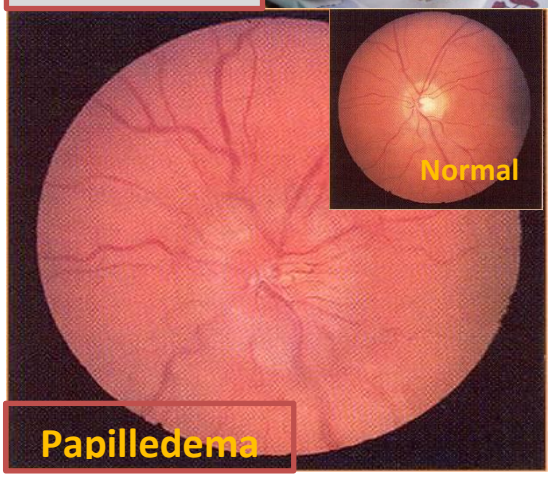
- ❖ Developmental anomalies:
  - Aqueductal anomalies
  - Dandy Walker malformation
  - Chiari II malformation
  - Myelomeningocele
- ❖ Infections: Meningitis
- ❖ Tumors
- ❖ Intracranial hemorrhages
- ❖ Traumatic brain injury
- ❖ Chromosomal anomalies (trisomy 13 & 18)

Classification of Hydrocephalus

Based on	Types	Remarks
Etiology	Congenital H.	▪ Present at birth or diagnosed in utero
	Acquired H.	▪ Develops after birth
Site of obstruction	Communicating H.	▪ Obstruction distal to ventricular system ▪ All ventricles dilated
	Non-communicating (Obstructive) H.	▪ CSF flow obstruction within ventricular system ▪ Only parts of ventricular system are enlarged
Intracranial pressure	Hydrocephalus with high pressure	▪ Most symptomatic hydrocephali ▪ Symptoms & signs of raised ICP
	Hydrocephalus with normal pressure	▪ May be asymtomatic ▪ NPH: Triads:Dementia, gait disturbance, incontinence


Macrocephalus


Sunset Sign



Clinical Presentation:

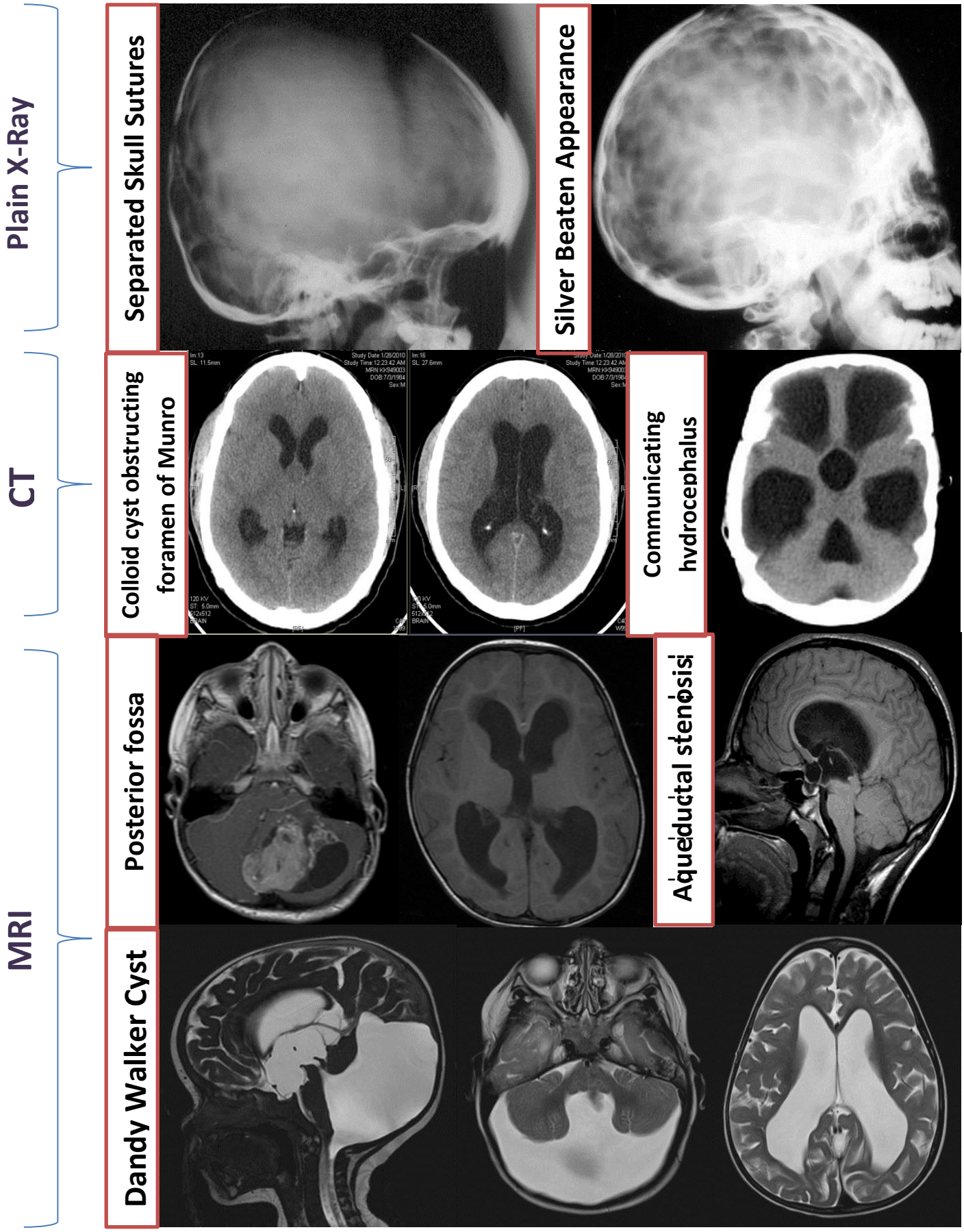
I- Infants & Young Children

- Increasing head circumference
- Irritability, lethargy, poor feeding, and vomiting
- Bulging anterior fontanelle
- Widened cranial sutures
- McEwen's *cracked pot sign with cranial* percussion
- Scalp vein dilation (increased collateral venous drainage)
- Sunset sign (forced downward deviation of the eyes, a neurologic sign almost unique with hydrocephalus)
- Epidsodic bradycardia and apnea

II- Juvenile & Adult

- Symptoms & signs of raised ICP:
  - Headaches
  - Vomiting
  - Visual disturbance
  - Papilledema
- Decreased level of consciousness.
- Seizures
- Focal neurological deficit
- Collection of CSF around previous shunt site

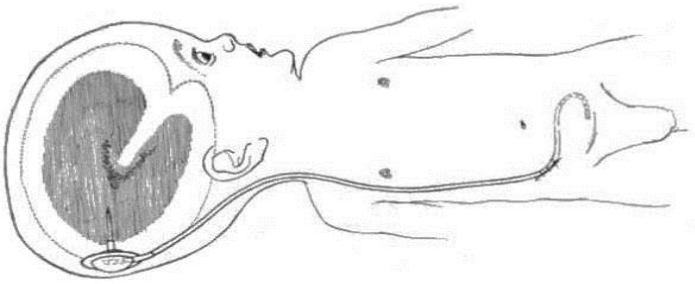
Radiological Investigations of Hydrocephalus of different causatives:



☐ **CT scan:**
- Allows direct visualization of the ventricular system.
- Shows acute and chronic ventricular enlargement.
- Often shows site and cause of ventricular obstruction.
- Method of choice for emergency.

☐ **MRI:**
- Shows more anatomical details in multiple planes
- Allows better visualization of:
  - Obstructive lesion
  - Associated brain anomalies
- Shortcomings:
  - Long imaging time.
  - Often anesthesia needed for children.
  - Not routinely available in emergency.

☐ **Treatment:**
- Removal of obstructive lesion
- CSF shunting procedures:
  - Ventriculo-peritoneal (VP shunt)
  - Ventriculo-atrial (VA shunt)
- Endoscopic 3rd ventriculostomy



☐ **Ventriculo-peritoneal Shunt:**
- Diversion of excessive ventricular CSF to the peritoneal cavity.
- Aim is to normalize the intracranial pressure.
- Shunt system consists of 3 components:
  - A Ventricular catheter.
  - A valve that allows unidirectional CSF outflow at a certain pressure range or flow rate.
  - A long peritoneal catheter.
- Shunts are made of biocompatible silicon & cause no or minimal tissue reaction or intravascular thrombosis.

☐ **Complications of VP-Shunt:**
- Immediate operative complications
- Shunt malfunction
- Shunt infection

☐ **Operative Complications of VP-Shunt:**
- Misplacement of:
  - ventricular catheter
  - peritoneal catheter
- Intracerebral / intraventricular hemorrhage
- Injury to abdominal viscera
- Pneumothorax
- Convulsions

☐ **VP-Shunt malfunction:**

- Most common shunt complication
- Incidence:
  - In the first few months after surgery: 25 to 40% of cases
  - Later 4 to 5 % per year.
- Causes:
  - Obstruction by cell debris, choroid plexus, or blood clot
  - Accounts for >50% of all shunt malfunctions
  - Migration, disconnection, or rupture shunt catheter(s)
  - Shortening of peritoneal catheter as child grows
  - CSF encystation around the peritoneal catheter
- Over drainage:
  - Subdural fluid collection
  - Slit ventricle syndrome

☐ **VP-Shunt Infections:**

- Occurance: About 5%
- Organisms:
  - *Staphylococcus epidermidis : ~40%*
  - *Staph aureus: ~20%*
  - *Streptococci & gram -ve organisms: less frequent.*
- Clinical features:
  - Onset ~8-10 weeks after shunt insertion.
  - Fever, malaise, headache & irritability ± neck stiffness.
  - Peritonitis is less common.
- Diagnosis: By CSF & blood examination & culture
- Treatment: Shunt removal, external ventricular drain, antibiotics until CSF is clean followed by new shunt insertion
- May result in additional neurological/intellectual impairment

☐ **Endoscopic 3rd Ventriculostomy:**

- Suitable for cases with patent external CSF spaces
- The endoscope is passed through a burr hole to the third ventricle
- The floor of 3rd ventricle is fenestrated just anterior to mamillary bodies to allow for CSF to exit the ventricle

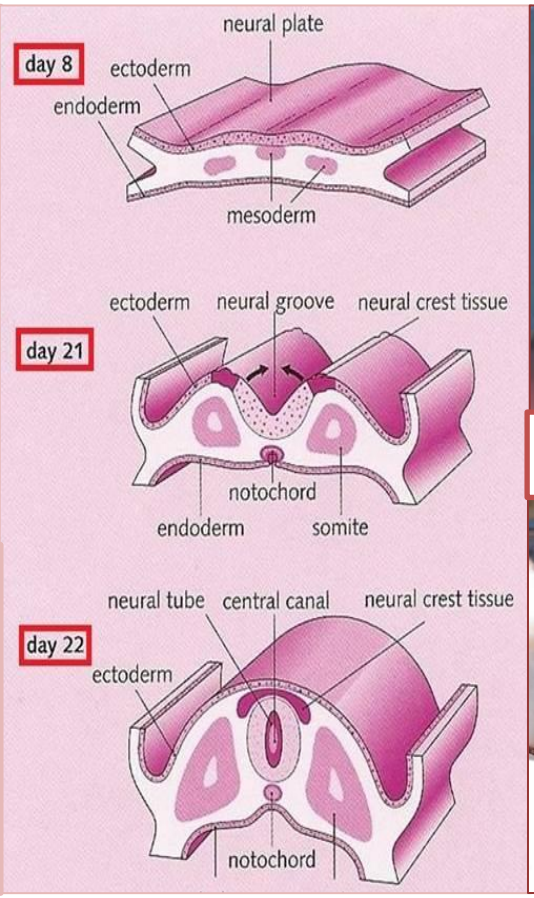
☐ **Neural Tube Defects (NTD):**

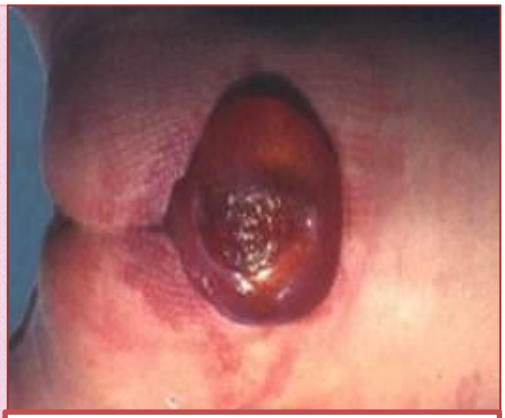
- Anomalies arising from: Incomplete or faulty closure of the dorsal midline embryonal structures
- **Two groups:**
  - Spinal dysraphism: by far larger
  - Cranial dysraphism

❖ **Spinal dysraphism:**

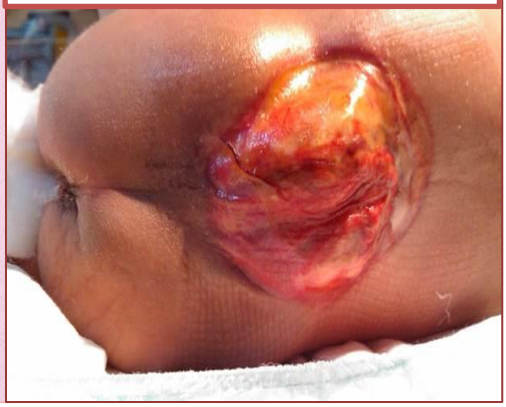
Various Forms:

❶ Myelomeningocele
❷ Meningocele
❸ Spina bifida occulta





Myelomeningocele



❶ Myelomeningocele

- Most important dysraphic disorder
- Incidence: 0.2-2/1000 live births
- Risk increases to 5% if a sibling is affected
- Slight female preference
- More common in whites

○ **Etiology:**

- Uknown
- Genetic factors
- Teretogens, e.g. sodium valproate

- 80% are in lumbosacral region
- Spinal cord and roots protrude through the bony defect

○ **They can be:**

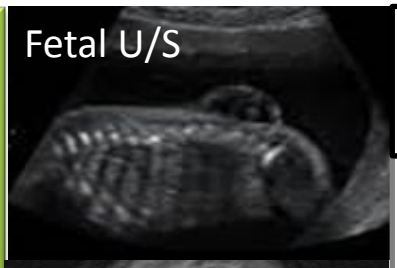
- Closed: neural tissue lies within a cystic cavity, lined with meninges and/oder skin
- Open: dysplastic neural tissue is exposed to air

○ **Associated anomalies:**

- Hydrocephalus
- Chiari type II
- Aqueduct forking
- Many others: Microgyria, ectopic grey matter, platybasia, etc.

Antenatal diagnosis

- Fetal U/S
- **In high risk patients:**
  - MRI
  - Screening maternal serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase
  - Contrast enhanced amniography
  - Possibility of therapeutic abortion



Myelomeningocele Assessment & Management

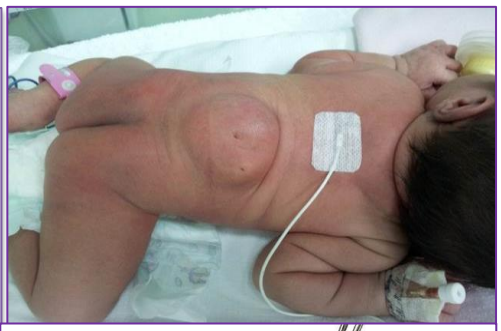
- Careful **examination of the lesion** for:
  - Presence of neural elements
  - Quality of the skin of the sac
  - Any CSF leakage
- Transillumination
- Observe limb movements: spontaneous & to pain (degree & level of neurological damage)
- Note dilated bladder & patulous annual sphincter
- Look for associated anomalies: e.g. hydrocephalus, scoliosis, foot deformities
- Investigations: U/S or MRI
- **Treatment:**
  - Immediate closure & replacement of neural tissues into spinal canal to prevent infection
  - Hydrocephalus need to be managed early to prevent CSF leakage from the wound
  - In patients with multiple serious congenital anomalies; many adopt thoughtful conservative treatment

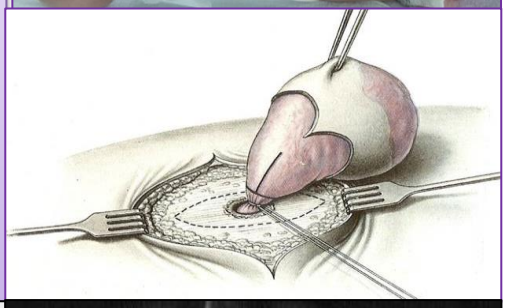
Myelomeningocele Long Term Care

- Regular follow up in multidisciplinary **spina bifida clinic** consisting of specialists in urology, orthopedics, pediatrics, neurosurgery, physical therapy, psychology & social services
- Goal: Early detection of problems and prompt treatment
- Urological: urinary incontinence, vesicoureteric reflux, recurrent UTI, renal impairment
- Pediatrics: hypertension and stunted growth
- Orthopedic: feet deformity, and tendon transfer, pelvic and spine deformities
- Neurosurgery: tethered cord, chiari II malformation, shunted hydrocephalus
- Physiotherapy: limb weakness, deformities, contractures
- Psychology: emotional stresses on patient & family
- Social services: supply of equipment, individual help

❷ Meningocele

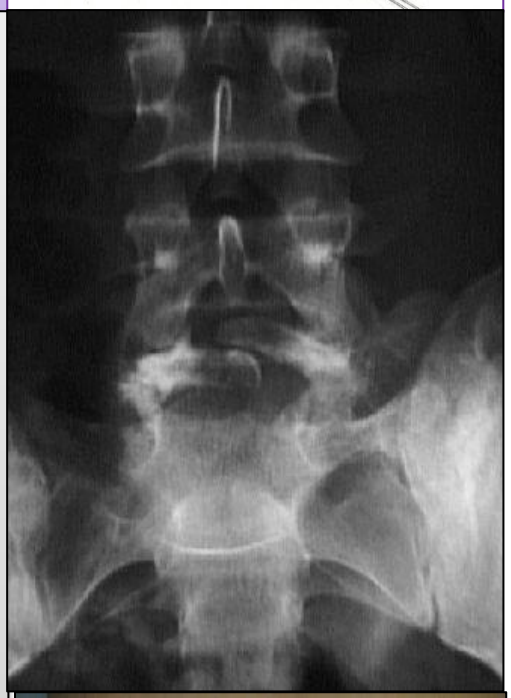
- Cystic CSF-filled cavity lined by meninges & skin
- Communicates with spinal canal
- No neural tissue
- Seldom any neurological deficit
- Rarely associated with other congenital anomalies
- U/S or MRI
- Tx: Excision; urgent in case of CSF leak





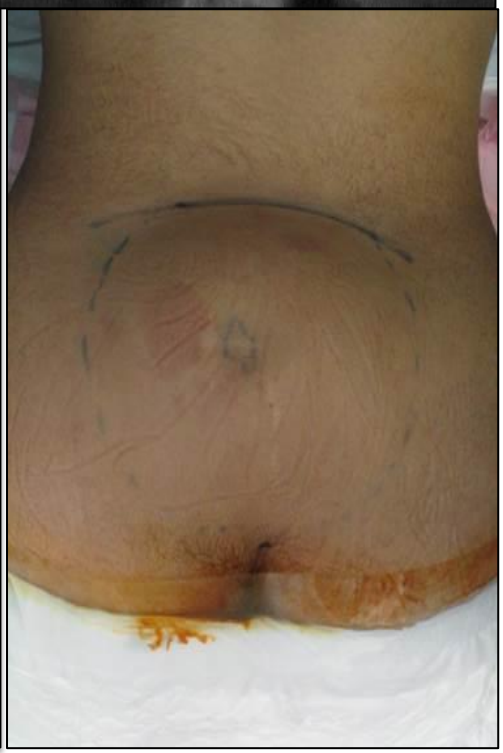
❸ Spina Bifida Occulta

- A bony defect of the lamina
- Usually in the lumbosacral region
- Affects 5-10% of population
- Clinically not significant
- No treatment required
- **But**: Rule out associated cutaneous abnormalities



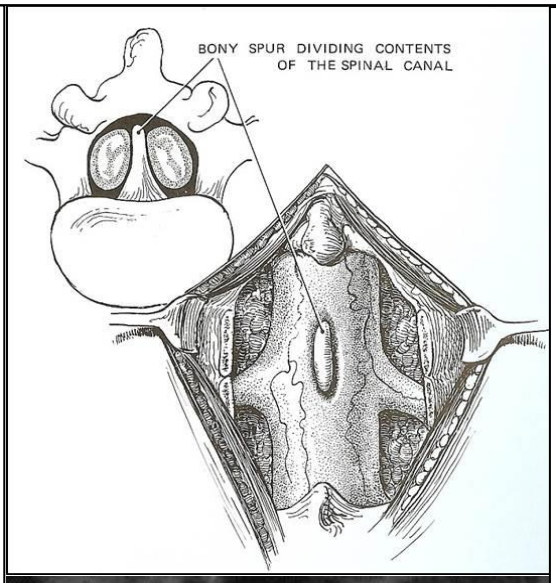
Spina Bifida Occulta: Caution!

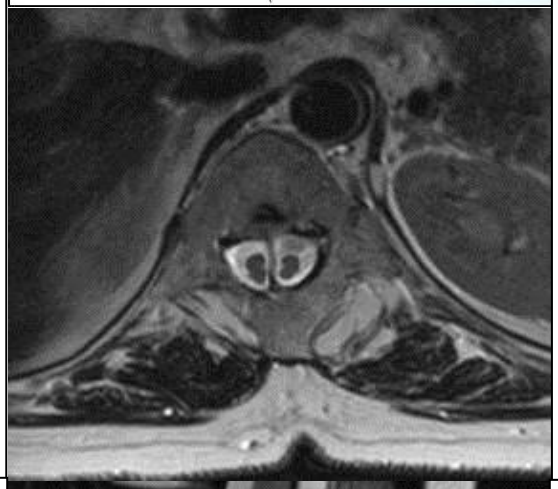
- In individuals with additional lumbosacral cutaneous abnormalities:
  - Tuft of hair
  - Dimple
  - Sinus
  - Port wine stain
  - Subcutaneous lipoma
- High incidence of other occult spinal anomalies:
  - Diastematomyelia
  - Intraspinal lipoma
  - Dermoid tumor
  - Tethered cord due to thickened filum terminale.



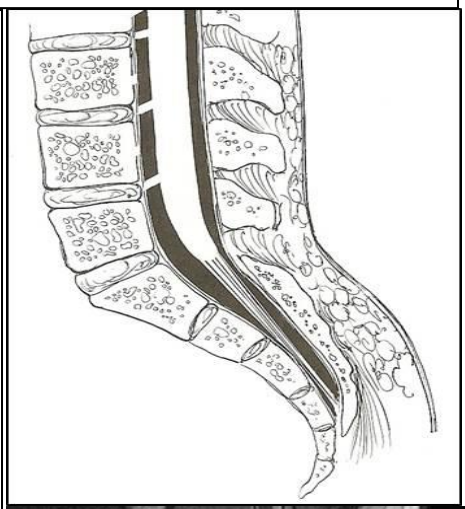
Other Spinal Dysraphic Anomalies

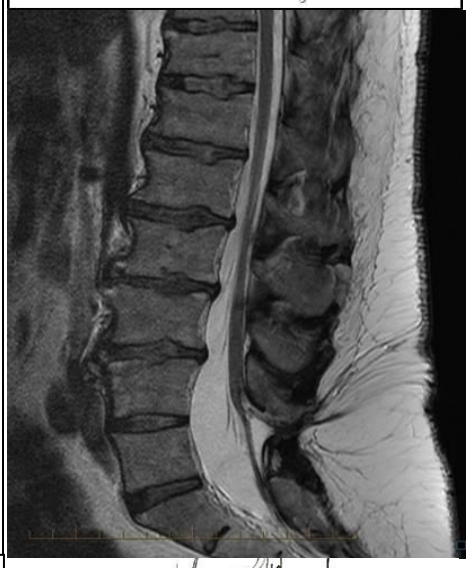
Diastematomyelia



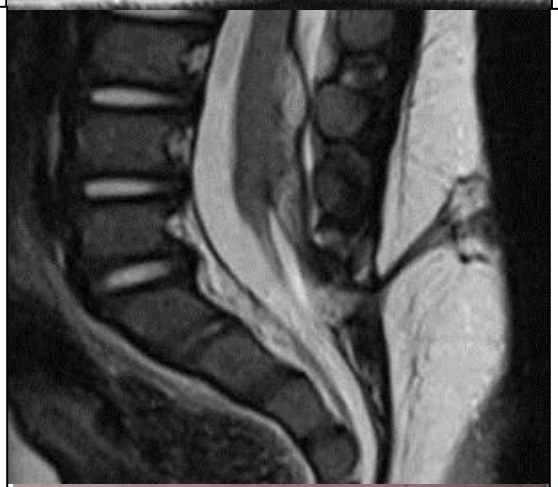


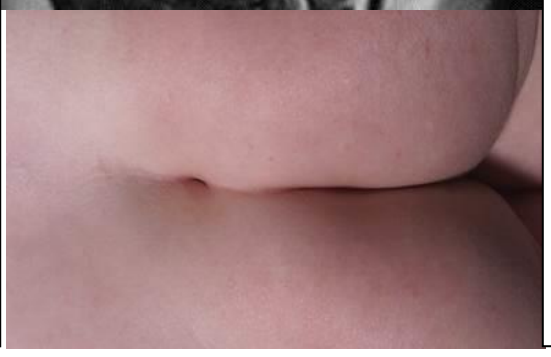
Tethered Spinal Cord



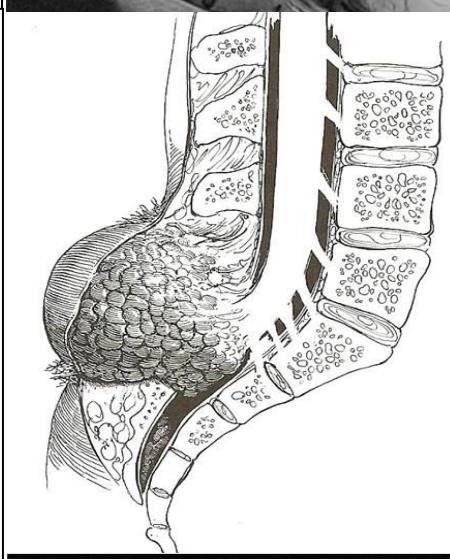


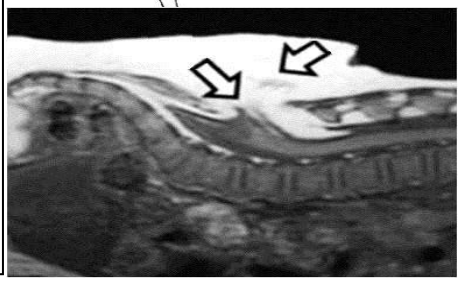
Dermal Sinus





Lipomyelomeningocele

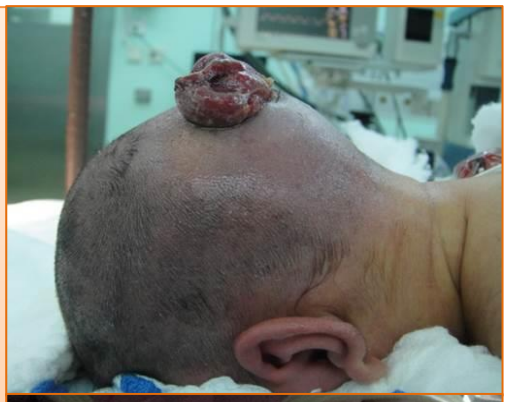




Cranial Dysraphysm

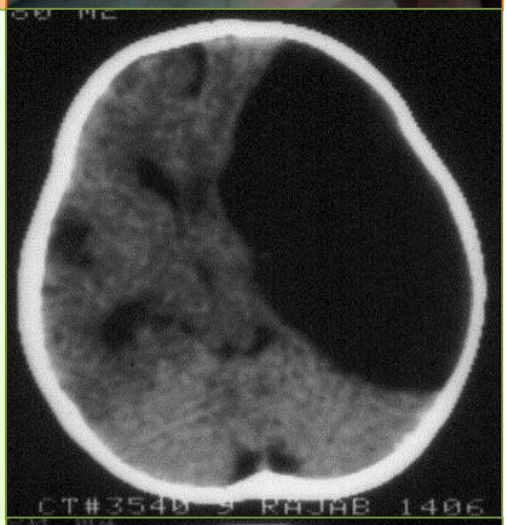
Encephalocele

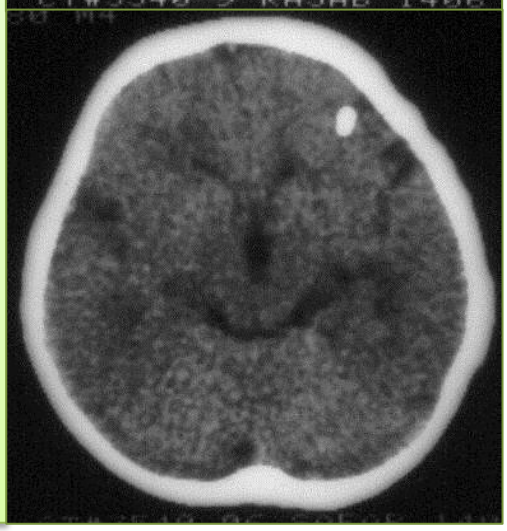
- Usually occipital, less often ethmoidal
- May contain occipital lobe or cerebellum
- Often associated with hydrocephalus
- Immediate treatment if ruptured
- Outcome depends on contents



Arachnoid Cysts

- Benign developmental CSF containing cysts
- Predomenantly (~50%) located in the sylvian fissure
- Can cause:
- Increase ICP
- Convulsions
- Neurological defecit
- Endocrine dysfunction
- Imaging: CT (Pre & post cystoperitoneal) & MRI
- Tx: Cystoperitoneal shunt or endoscopic fenestration; rarely excision.

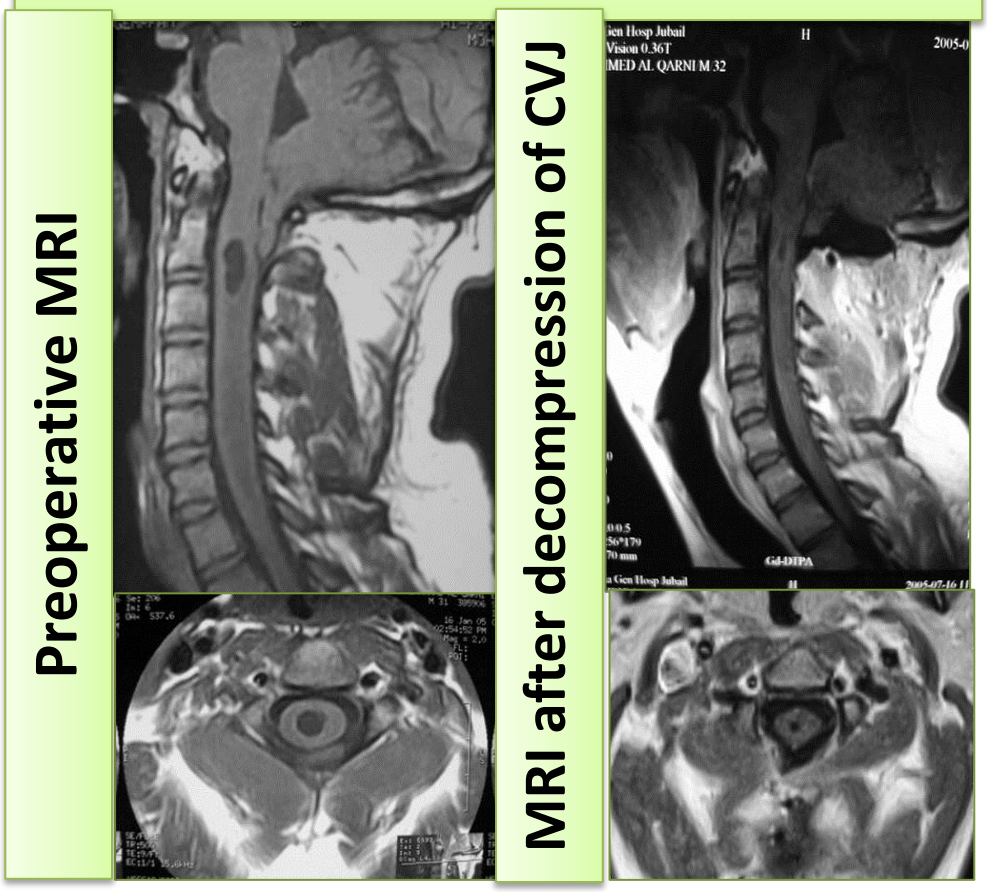




Chiari Malformation (CM) & Syringomyelia

- CM is a complex developmental malformation characterized by caudal displacement to variable degrees of parts of the cerebellum, medulla oblongata and 4th ventricle into the cervical canal
- Syringomyelia is cavitation within the spinal cord
- Hydromyelia refers to dilatation of the central canal that occurs in associations with CM

Chiari Malformation with hydromyelia treated by decompression of CVJ



Presntation & Management

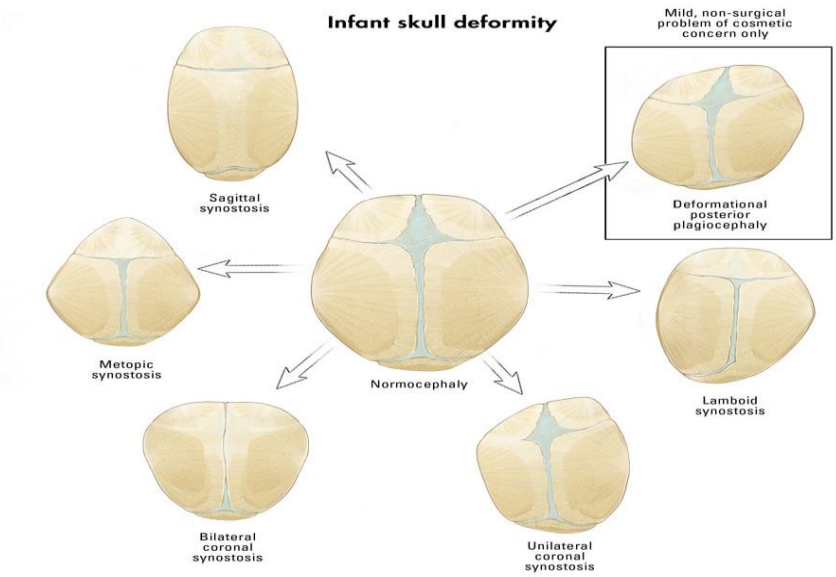
Chiari Malformation:

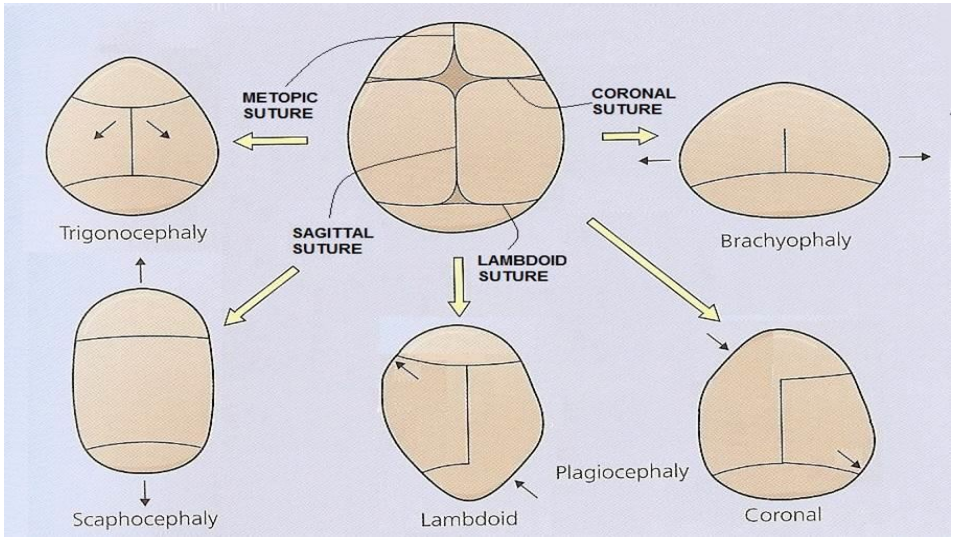
- Occipital headache
- Nystagmus
- Spastic paresis of upper/lower limbs
- Ataxia
- Lower cranial nerve defecits
- Dx: MRI
- Tx: Decompression of craniovertebral junction

Syringo/hydromyelia:

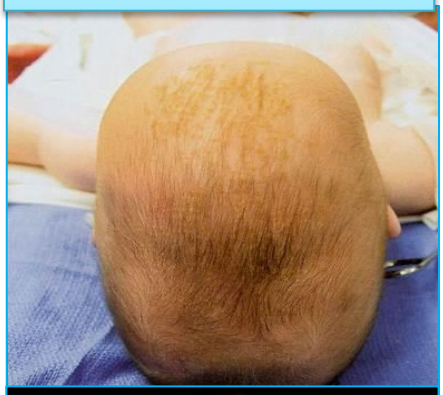
- Dissociated sensory loss in “cape-like” distribution
- Wasting of small hand muscles
- Dx: MRI
- Tx: Syringostomy, syringo-subdural shunt

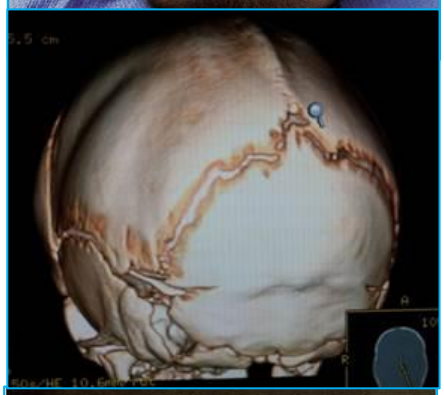
Cranial Synostoses

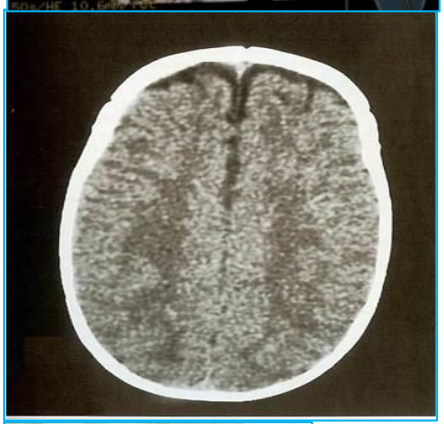


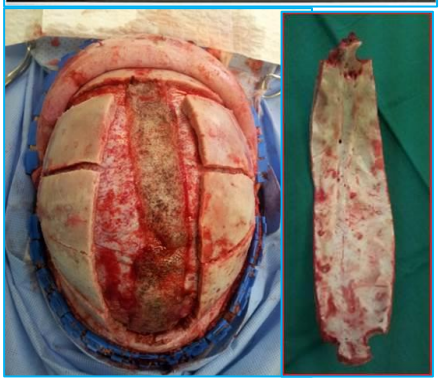


Sagittal Suture Synostosis: Scaphocepaly

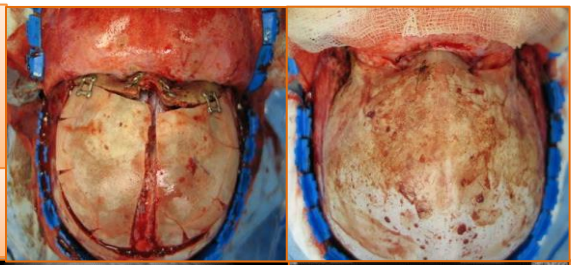


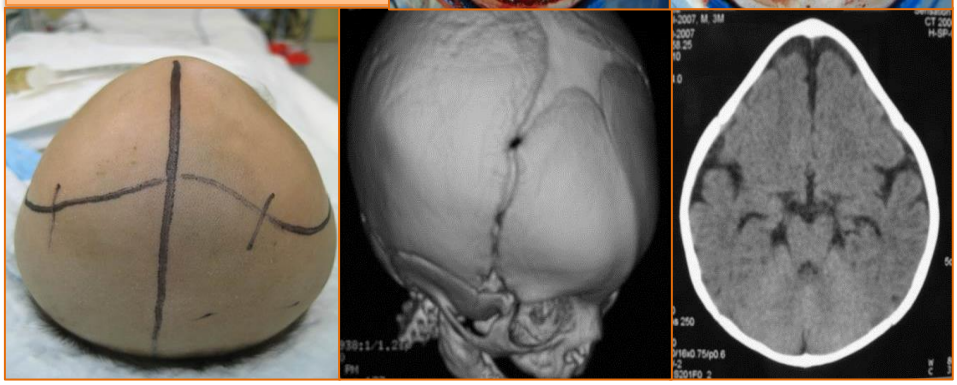






Metopic Suture Synostosis: Trigonocephaly





Disease	Definition	Classification	Radiological investigation	Treatment
Hydrocephalus	Increased amount of CSF in the head due to either: -Excessive CSF production. -CSF flow obstruction. -Decreased CSF absorption.	Based on: ***Etiology** -Congenital. -Acquired. ***Site of obstruction:** -Communicating -Non-communicating (Obstructive) ***Intracranial pressure:** -Hydrocephalus with high pressure. -Hydrocephalus with normal pressure	-Plain X-ray -CT–scan -MRI	-Removal of obstructive lesion. -CSF shunting procedures. -Endoscopic 3rd ventriculostomy.
Neural Tube Defects	Anomalies arising from incomplete or faulty closure of the dorsal midline embryonal structures	-Spinal dysraphism: *Myelomeningocele (closed/ Open) *Meningocele *Spina bifida occulta Others: *Diastematomyelia *Tethered spinal cord. *Dermal sinus *Lipomyelomeningocele -Cranial dysraphism *Encephalocele. *Arachnoid Cysts.	*Myelomeningocele: -Fetal U/S -In high risk patients: -MRI. -Screening maternal serum/amniotic fluid. - Contrast enhanced amniography. *Meningocele U/S or MRI *Arachnoid Cysts: CT & MRI	*Myelomeningocele: -Immediate closure & replacement of neural tissues into spinal canal to prevent infection. -Hydrocephalus needs to be managed early to prevent CSF leakage from the wound. -In patients with multiple serious congenital anomalies; many adopt thoughtful conservative treatment. *Meningocele: Excision; urgent in case of CSF leak

				*Spina bifida occulta: No treatment required *Arachnoid Cysts: Cystoperitoneal shunt or endoscopic fenestration; rarely excision
Chiari malformation (CM)	a complex developmental malformation characterized by caudal displacement to variable degrees of parts of the cerebellum, medulla oblongata and 4th ventricle into the cervical canal		MRI	Decompression of craniovertebral junction
Syringo/hydromyelia	S: cavitation within the spinal cord. H: dilatation of the central canal that occurs in associations with CM		MRI	Syringostomy, syringo-subdural shunt
Cranial Synostoses	Premature closure of one or more of the sutures between the skull plates	-Sagittal suture synostosis: Scaphocephaly -Metopic suture synostosis: Trigonocephaly	Plain x-rays	

From surgical recall:

1. **What are the signs/ symptoms of hydrocephalus?**

Signs of increased ICP: HA, nausea, vomiting, ataxia, increasing head circumference exceeding norms for age

2.**What is hydrocephalus ex vacuo?**

Increased volume of CSF spaces from brain atrophy, not from any pathology in the amount of CSF absorbed or produced

3. **Define spina bifida occulta?**

Defect in the development of the posterior portion of the vertebrae

4. **What is the most common clinically significant defect?**

Myelomeningocele: herniation of nerve roots and spinal cord through a defect in the posterior elements of the vertebra(e); the sac surrounding the neural tissue may be intact, but more commonly is ruptured and therefore exposes the CNS to the external environment

5. **What are the signs/ symptoms?**

Variable from mild skeletal deformities to a complete motor/sensory loss; bowel/ bladder function is difficult to evaluate, but often is affected and can adversely affect survival.

6. **What is syringomyelia?**

Central pathologic cavitations of the spinal cord.

7. **What is the definition of craniosynostosis?**

Premature closure of one or more of the sutures between the skull plates.

8. What are the types?

Named for the suture that is fused (e.g., sagittal, coronal, lambdoid); sagittal craniosynostosis accounts for >50% of all cases; more than one suture can be fused, and all or part of a suture may be affected.

9. **How is the diagnosis made?**

Physical examination can reveal ridges along fused sutures and lessened suture mobility; plain x-rays can show a lack of lucency along the fused suture, but are rarely required.

10. **What are the indications for surgery?**

Most often the reasons are cosmetic, as the cranial vault will continue to deform with growth; occasionally, a child will present with increased ICP secondary to restricted brain growth.

11. **What is the timing of surgery?**

Usually 3 to 4 months of age; earlier surgery increases the risk of anesthesia; later surgeries are more difficult because of the worsening deformities and decreasing malleability of the skull.

Questions:

1. Myelomeningoceles are congenital malformations of the spinal cord. Which of the following findings are not commonly associated?

A. Hydrocephalus.

B. Chiari II malformation.

C. A midline dorsal spinal mass easily noted at birth.

D. Skin, bone, and dural defects superficial to the neural placode.

E. Mandatory urinary incontinence.

Answer: E

DISCUSSION: Myelomeningoceles are usually associated with hydrocephalus and the Chiari II malformation. The myelomeningocele sac is a midline dorsal spinal mass associated with defects in the skin, bone, and dura overlying the neural placode, and the sac is readily apparent at birth. Although the innervation of the bladder is dysmorphic, the majority of patients can achieve social urinary continence through the use of clean intermittent bladder catherization.

2. Hydrocephalus is defined as:

Answer: Accumulation of CSF with ICP.

3. obstructive hydrocephalus is best treated by:

Answer: Endoscopic third ventriculostomy

4. A mother brought her 3 month/old baby to the clinic concerning about his large head in addition to poor feeding that followed by vomiting. On examination, scalp veins were dilated, and the eyes were forcibly deviated downward. The cause(s) of this condition is (are):

A. CSF overproduction.
B. Obstruction of CSF flow.
C. Under absorption of CSF.
D. All of the above.

Answer: D

5. Which of the following lesions is not one of the cutaneous stigmata of occult spinal dysraphism?

A. Midline lumbar capillary hemangioma.

B. Focal hairy patch over the thoracolumbar spine.

C. Dermal sinus located above the midsacrum.

D. Midline subcutaneous lipoma.

E. Café-au-lait spot over the thoracolumbar spine.

Answer: E

DISCUSSION: Café-au-lait spots are not a feature of spina bifida occulta. The other four skin features all may be associated with significant intradural pathology and warrant further investigation, most commonly with magnetic resonance imaging (MRI). A dermal sinus tract that overlies the coccyx is a pilonidal sinus and is not likely to be associated with intradural pathology.

6. Which statement is true?

A. Spina bifida occulta is a neurosurgical emergency
B. Meningocele contains spinal cord
C. Spinal dysraphism occurs most commonly in the lumbosacral region
D. Non of the above.

Answer: C

7. VP shunt was inserted to the patient. The commonest complication could occur is:

A. Tube fracture.
B. Shunt infection.
C. Shunt migration.
D. Shunt obstruction.

Answer: D