430 SURGERY TEAM

Hydrocephalus & Common Neurosurgical Congenital CNS Malformation

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Green: Doctor's notes & explanation during the lecture.
Blue: Further explanation & team's notes.
Red: important notes.

HYDROCEPHALUS

1. Introduction :

- **Hydro** = water + **Cephalus** = head
- **Hydrocephalus** is an increase in the amount of CSF in the head.
- **Increasing of CSF in the head usually results in:**
  1. Ventriculomegaly
  2. Increased intracranial pressure

There is no water in the head except CSF . Hydrocephalus is increase of CSF within the cerebral ventricle and is usually associated with increase ICP .

2. Anatomy & Physiology :

Anatomy of cerebrospinal fluid spaces:

2 lateral ventricles inside cerebral hemisphere they are separated by corpus callosum and septum pellucidum. They are communicated through **foramen of Monro** (funnel shape foramen) to the **3rd ventricle**. 3rd ventricle has fine canal which is cerebral aqueduct or **aqueduct of Sylvius** that connected to the **4th ventricle.**

Why is 4th ventricle very important? Because it contains three outlet of ventricles

1. **One** in medial aperture : **Foramen of Magendie**
2. **Two** in the sides (lateral apertures) : **Foramina of lushki**

The importance of these foramina that CSF **gets out the ventricles** to reach **basal cisterns** and from basal cisterns it gets distributed to the rest of the **intracranial cavity.**

Physiology of CSF Production:

- Total volume of CSF in the ventricles ranges from **5-15 ml in neonates** to **150 ml (cc) in adults.** (1)
- Produced **mainly** by the choroid plexus of the ventricles (2) and to a **lesser extent** by the extracellular fluid of the brain. (3)
- **Rate of production** is 0.3-0.4 ml/minute → (~500 ml (cc) /day)
- **Only very high ICP will reduce CSF production; usually when brain perfusion (4) is decreased [Note: the slight raising or initial phase of increasing of ICP won't affect the production].**

(1) Every day the plexus produces 500ml of CSF (–) 150 ml where in the CNS = 350 CSF absorbed every day. The 350 cc of CSF means that **each day our brain is washed three times by fresh CSF.**

(2) Most of CSF is produced in choroid plexus in the **lateral ventricles.**

(3) CSF production is an **active process**. That may be affected by ↑ ICP but it **does not stop it**

(4) **Perfusion pressure =** mean systemic arterial pressure (MAP) – mean intracranial pressure

Physiology of CSF Flow & Absorption:

CSF Flow: Through anatomical CSF spaces. (Explained earlier in the anatomy)

CSF Absorption: Mainly at the arachnoid granulation

- Most of the CSF absorption occurs Para sagittal around sagittal sinus that has arachnoids granulations. If these granulations are obstructed the absorption isn't adequate.
- Most of the obstructions are not absolute / complete obstruction. **They are partial obstruction so the symptoms develop slowly.**

3. Pathogenesis of Hydrocephalus :

1. **Excessive CSF production :**
   - Choroid plexus papilloma

> [It's very special condition: tumor of choroid the plexus. results in: **1)** choroid the plexus enlarge. **2)** Increase in CSF production more than 500 cc **[multifold]. 3)** the absorptive capacity isn't enlarge that results **accumulation of CSF.]**

2. **CSF flow obstruction : [The most common cause of hydrocephalus]**
   - Tumors especially near or in the ventricles.
   - Congenital anomalies**.**
3. **Decrease CSF absorption :**
   - Post-Meningitic
   - Post Sub Arachnoid Hemorrhagic (SAH)
4. **Decrease brain volume (Brain atrophy) :**
   - **Hydrocephalus Ex Vacuo: Advancing age, after trauma or severe meningitis all will lead to brain atrophy but the head won't shrink so the empty space will be filled with CSF.**

4. Types of Hydrocephalus :

There are various classifications based on:

- **a) Site of the obstruction**
- **b) Etiology**
- **c) level of Intracranial Pressure**

a) Site of the obstruction :

I. Communicating Hydrocephalus :

- There is no obstruction within the ventricular system or in the pathway of CSF (the four ventricles are communicated with each other)
- **The entire ventricular system is enlarge**
- **Causes :**
  - CSF overproduction
  - Inadequate absorption
  - Flow obstruction **distal outside** the ventricular system (lies after the outlets of 4^{th} ventricles) **Eg :**
    - Post meningitis
    - Post subarachnoid hemorrhage
    - Chiari malformation
    - Anomalies of the posterior fossa? Around formain magnum where CSF doesn't circulate well.

II. Non-Communicating Hydrocephalus :

- There is a local Blockage in the flow of CSF
- **Only parts of the ventricular system are enlarged [**depend on the site of the obstruction}
- **Causes :** CSF Flow obstruction **within** the ventricular system
  - **Eg by :** tumors **,** Congenital anomalies **[aqueductal stenosis]**

b) Etiology :

- **Congenital (**Primary) **hydrocephalus**
- **Acquired (**Secondary)**hydrocephalus** (most cases of hydrocephalus during life)

I. Congenital Hydrocephalus :

Present since birth or diagnosed in utero.

Causes:

- **Developmental anomalies :**
  1. Aqueduct anomalies **Aqueductal stenosis MOST COMMON CAUSE OF CONGENITAL HYDROCEPHALUS**
  2. Dandy Walker malformation
  3. Chiari II malformation
  4. Myleomeningocele
- **Intrauterine infection**
  1. Viral Infection (CMV, mumps, rubella, varicella)
  2. Toxoplasmosis
- **Congenital tumors (very rare)**
- **vascular malformation : Vein of Galen aneurysms**
- **Chromosomal anomalies (trisomy 13 and 18)**
- **Congenital or primary hydrocephalus (idiopathic; no known reason)**

II. Acquired Hydrocephalus :

Develop after birth

Causes

- **Germinal plate hemorrhage:** in preterm infants <1500 gm (30%- 40%) 'immature & fragile blood vessel walls around the ventricles'
- **Post-meningitis :**

1% of survivors of bacterial meningitis
More in neonates
Especially G-ve organisms (i.e. E. coli).

- **Rare but important, postnatal cysticercosis**
- **Tumors**
- **SAH: Subarachnoid hemorrhage**
- **Severe TBI (Traumatic brain injury)**

Important 3 causes of communicating hydrocephalus:
- Post-hemorrhagic
- Post-meningitic
- Post-traumatic
3 posts for acquired causes! *And remember the cause of acquired hydrocephalus is usually outside the ventricles.*

The most common cause is tumor but **the earliest type** of **the acquired** hydrocephalus in post natal period which is bleeding in the ventricle (**Germinal plate hemorrhage).**

c) Level Of Intracranial Pressure:

I. Hydrocephalus with raised ICP:

Most of symptomatic hydrocephalus

II. Hydrocephalus with normal ICP:

1. Normal Pressure Hydrocephalus (NPH): elderly pt presents with **dementia like symptoms its treatable causes of dementia.** If pt came with dementia we have to exclude NPH

- **Communicating type of hydrocephalus**

- **Clinical presentation : classic triad of :**

1) **Gait abnormality / ataxia** [rapid Parkinson gait , shaky , unsteady , small steps 'as if the pt don't know how to walk']
2) **Cognitive deficit / dementia**
3) **Urinary incontinence**

- **Investigations : CT – MRI [ventricular dilation more than brain atrophy]**

2. Arrested hydrocephalus: mild hydrocephalus which produce symptoms for sometime **not severe symptoms** like slow enlargement of head of the baby then the head stops enlarging. Then is back to normal **{pt with Ventriculomegaly who became again asymptomatic)**

5. Clinical Features :

A. Infants And Young Children

1. **Increasing head circumference.** [Scalp bones are still soft & have connective tissues between them**. the head circumference increases abnormally not according to curve of growth.** There is **enlargement of the head which calls (macrocephalus).** Remember macrocephalus isn't hydrocephalus. Hydrocephalus is one of the causes of macrocephalus.

2. **Irritability, lethargy, poor feeding, and vomiting.** (Leads to delayed development)
3. **Bulging anterior fontanel**. [fontanel normally is 1-2 cm in diameter usually it's **soft** . if the baby is upright the fontanel is **pulsating and sunken** . But in hydrocephalus **it's tense , large and full]**

4. **Widened cranial sutures** [sutures start to separate . gaps between them]

5. **McEwen's cracked pot sign with cranial percussion** (sounds like you're tapping on a cracked pot)

6. **Scalp vein dilation (increase collateral venous drainage)** [because there is shunting of the venous blood and Skin is very thin]

7. **Sunset sign (forced downward deviation of the eyes , neurological sign almost unique with hydrocephalus)** We can see the upper limbus of the iris isn't covered by eye led .Not only infants but also in children and sometimes adults

- **Sunset sign** is secondary to dilation of posterior 3rd ventricle that will then compress the part of midbrain called **superior colliculus (contains nucleus of 3rd and 4th cranial nerves) leading to palsy of upward gaze 'failure to look upward'** (when you ask child to look upward, he will move his eyelid but won't be able to move his eye-ball > you see a white rim of sclera below the eyelid)

- And **Parinaud's Syndrome** has the same clinical manifestation, secondary to lesion in same area (area of penile gland) compressing the superior colliculus of brain stem and leading to Parinaud's sign 'sunset appearance of eyes'

8. **Episodic bradycardia and apnea** (If hydrocephalus is left untreated – the increased intracranial pressure will press on the brain stem -where the respiratory centers are located- which will lead to this)

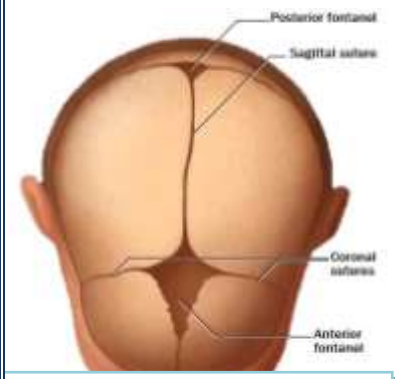


Figure 1 : Anterio frontanel

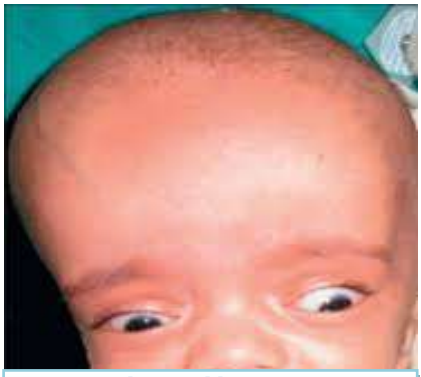


Figure 2: scalp vein dilatation & sunset sign

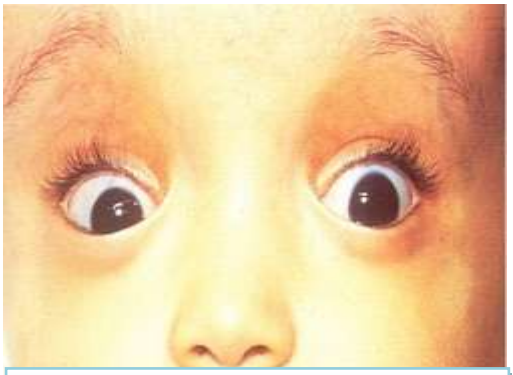


Figure 3: sunset sign

B. JUVENILE AND ADULTS

1. **symptoms and Signs of raised intracranial pressure:**

a) **Headaches**. (Usually in the **early morning or late night worse when pt lying flat or bending forward better if the pt is upright.** if left untreated the pt will suddenly vomit in a **projectile fashion .)**Why? Because during sleep, the patient will hypo ventilate, which will lead to ↑ CO_2 → ↑ vasodilation → blood stasis & ↑ intracranial pressure)

b) **Vomiting**. (The symptoms are usually relived after vomiting. Why? Because vomiting leads to hyperventilation which will reverse the CO_2 retention and cause vasoconstriction of the blood vessels → ↓ Intracranial pressure)

c) **Visual disturbances** (blurred , double vision due to **abducent nerve involvement**).

d) **Papilledema Most common clinical sign.** it is present if hydrocephalus for few days or less than 24 – 48 h **(acute obstruction)**

- **Papilledema** always presents as a bilateral . its swelling of the optic nerve
- **You have to defrentiate between Papilledema and normal fundoscopy**
- **Decription of Papilledema :** venous engorgement , blurring of optic margins , hemorrhages over optic disc and elevation of optic disc .

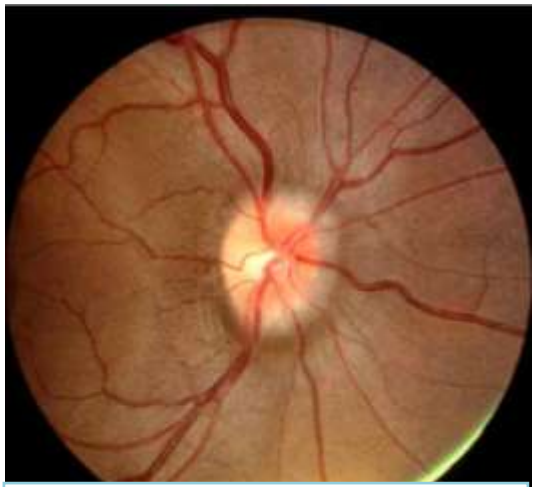


Figure 4: Papilledema

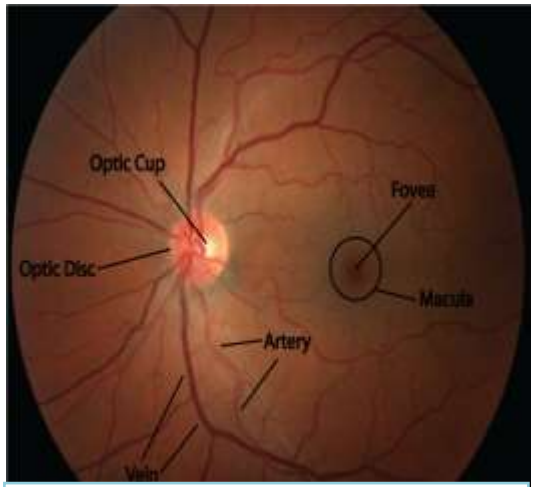


Figure 4: normal fundoscopy

2. **Decreased level of consciousness**
3. **Seizures**. (Acute manifestation) } (Depending on the underlying
4. **Focal neurological deficit** } lesion of hydrocephalus)
5. **Collection of CSF around previous shunt site** **. Iatrogenic or artificial hydrocephalus** If pt went through surgery there is bulge over that defect .

6. INVESTIGATIONS

- **Plain X-ray : shows longstanding indirect signs of increased ICP .** Xray has limited role in hydrocephalus , it Shows only **secondary changes** . very little diagnostic value .
- **Ultrasound :** to diagnose **Ventriculomegaly** in **Utero** (as early as 16 week of gestation) mostly in the west
- **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 of Emergency**
- **MRI :**
  - Shows more anatomical details in multiple planes .
  - Allows better visualization of :
    - **Obstruction lesions** specially tiny lesions such as **aquedauctal stenosis**
    - **Associated brain anomalies**
  - **Shortcoming :**
    - Long image time
    - Often anesthesia needed for children
    - Not routinely available in emergency

The pattern of ventricular enlargement can help delineate the cause:

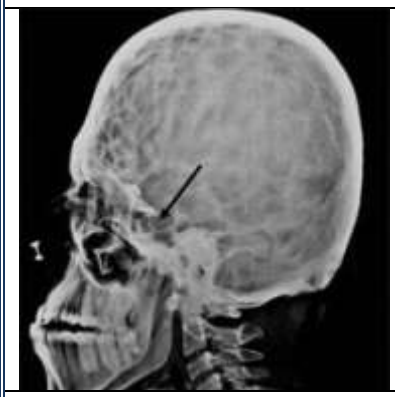
a. **Lateral & 3rd ventricle dilatation**

→ normal 4th ventricle: suggests **aqueduct stenosis** (b/w 3rd & 4th ventricles)

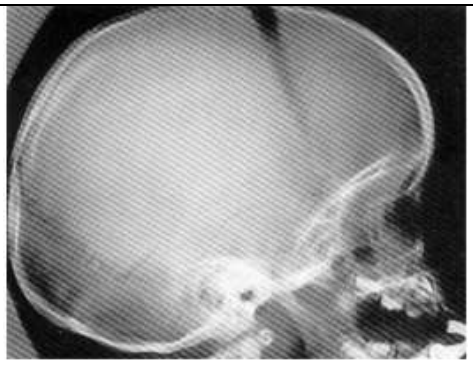
→ deviated or absent 4th ventricle: suggests **posterior fossa tumor** 'obstructive hydrocephalus

b. **Generalized dilatation:**

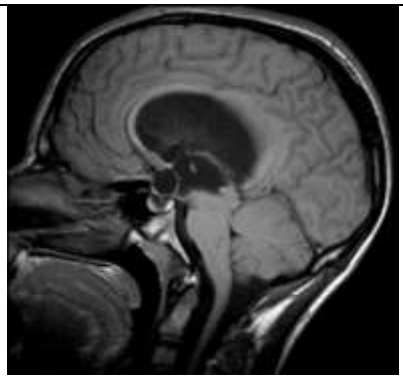
suggests a **communicating hydrocephalus.**



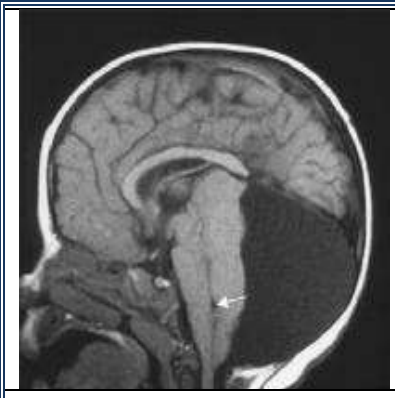
Picture 1 : silver beaten appearance : *cloudy looking appearance*



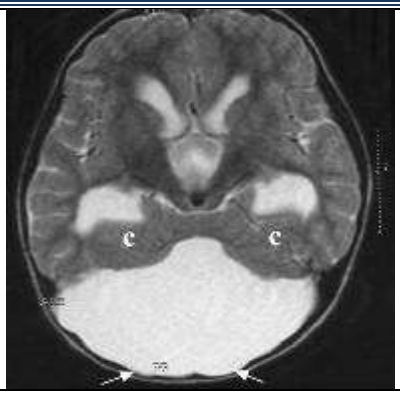
Picture 2 : Separated skull suture *secondary to increased intracranial pressure*



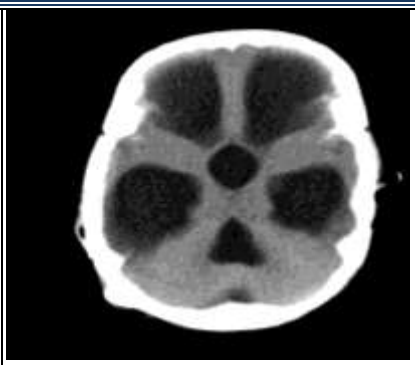
Picture 3 : Aqueductal stenosis



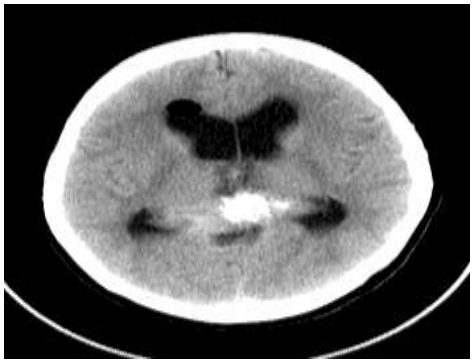
Picture 4 : Dandy-Walker malformation. Posterior fossa arachnoid cyst .
MRI shows a large posterior fossa cyst that is compressing the cerebellar hemispheres, fourth ventricle (arrow), and brainstem.



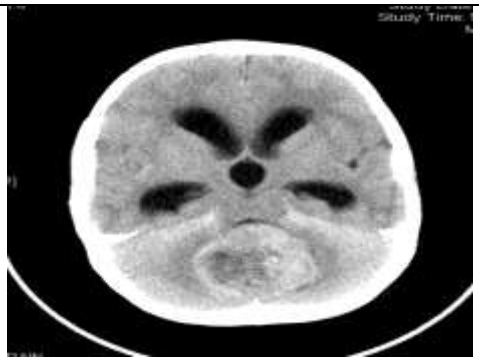
Picture 5 : Dandy-Walker malformation.
MRI shows hydrocephalus, a large cerebrospinal fluid cyst in the posterior fossa, thinned occipital bone (arrows), and hypoplastic cerebellar hemispheres with a winged appearance (c).



Picture 6 : Communicating hydrocephalus all ventricles are dilated



Picture 7 : Hydrocephalus caused by colloid cyst obstructing foramen of Monro
acquired not communicating (obstructive hydrocephalus) acute hydrocephalus (colloid cyst since birth) developmental lesion cause sudden death . major surgery with major risk .
Lateral ventricles enlarged but 3rd ventricle not dilated (normal) > lesion in 3rd ventricle that prevents CSF from entering the 3rd ventricle at level of foramen of Monro



Picture 8 : Hydrocephalus caused by Posterior fossa tumor gliomas
Lateral ventricles and third ventricles are enlarged but the fourth ventricles is compressed by the tumor

6. TREATMENT

There's no medical treatment, it's only conservative .it is mainly surgical except arrested hydrocephalus.
you can use some medication to reduce the volume of CSF and reduce ICP, but CSF is actively produced by the choroid plexus so it's difficult to get rid of the problem

Most common procedures:

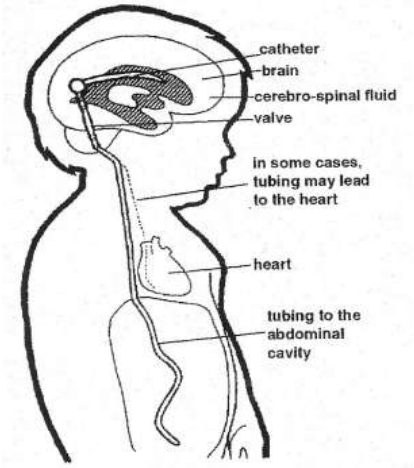
- **Ventriculo-peritoneal shunt (VP Shunt): most common procedure** From the ventricular system, usually the lateral into the peritoneal cavity not stomach
- **Endoscopic third ventriculostomy:** very popular not used to every type of hydrocephalus there is specific indications **[obstructive hydrocephalus]**

Infrequently used procedures:

- **Ventriculo-Atrial shunt (VA Shunt)** right atrium
- **Ventriculo-Pleural shunt**
- **Ventriculo- cisternal shunt**
- **Choroid plexus coagulation** used very rarely only if all other methods are failed

Ventriculo-peritoneal shunt (VP Shunt):

- Diversion of excessive ventricular CSF to the peritoneal cavity with **the Aim is to normalize the intracranial pressure**



- **Shunt systems consists with three Component**
  1. **ventricular catheter**
  2. specially designed **valve that allows unidirectional CSF outflow at a certain** pressure range or flow rate
  3. **A long peritoneal catheter**
- Shunts are made of silicon which is well tolerated by the body. **It causes minimal or no tissue reaction or intravascular thrombosis** But at the end, it is an artificial device that causes some complications.

7. COMPLICATIONS OF VP SHUNT

- **a. Operative complications**
- **b. Shunt malfunction**
- **c. Shunt infection**

Operative complications

- Misplacement of : **most common Operative complications**
  1. **ventricular catheter** [goes somewhere else]
  2. **Peritoneal catheter [** or if pt is obese and when operating with small incision and surgeon thinks he is in peritoneal cavity while he is still in subcutaneous tissue .]
- intracerebral / intraventicular hemorrhage puncturing the ventricles is a risk of bleeding
- 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 on 4 to 5 % per year
  - **[most of the pts need shunt changing at some point of their life]**
- **Obstruction :**

- Blockage of any shunt component by cell debris, choroid plexus or blood clots [because CSF isn't very clear fluid it contains cells also presence of catheter inside the ventricles cause some reactions]
- **Account of > 50% of all Shunt malfunction**
- Migration , disconnection or rupture shunt catheter(s)Sometimes catheter pass through stool or coming out through the anus
- Shortening of peritoneal catheter as child grows [because shunt is lifelong device]
- CSF encystations [make localized cavities like cysts] around peritoneal catheter

- **over drainage :**
  - subdural fluid collection
  - slit ventricle syndrome [cause collapse of ventricles]

[if the pressure of the shunt is very low it will over drain . symptoms : headache , nausea , vomiting , and dizziness . in this case shunt must be removed]

VP- SHUNT INFECTION :

- **(most common life threatening)**
- About 5% and may result in further risk of intellectual impairment
- **Organisms:**
  - **Staphylococcus epidermis about 40%.** Commonest cause. [It's normally found in the skin and it easily reaches the shunt, causing CSF infection 'meaning meningitis leading to disability']
  - **Staph Aureus about 20%**
  - **Streptococci and gram negative organisms** are less frequent. **[Most fearful infection because they produce pseudomembrane and if there is infection with gram negative organisms instead of having one communicating cavity pt will have compartmental cavities , each cavity requires shunt for its own as a total pt require 4-5 shunts but we can minimize the number of the shunts by endoscopy but we can't eliminate it]**
- **Clinical features:** like any other infection
  - Onset often within 8-10 weeks after shunt insertion
  - Fever, malaise, headache & irritability, neck stiffness.
  - Peritonitis is less common. **[peritoneum has many defense mechanism as macrophages etc . infection rate is low]**
  - Patients with Staph epidermis may look remarkably well with only intermittent fever or irritability.
- **Diagnosed:** CSF and blood examination , blood culture **[take sample of CSF through the shunt]**
- **Treatment of shunt infection:**
  - **Shunt Removal**
  - **external ventricular drainage [the pt is still having hydrocephalus** infection leads to increase CSF volume so patient suffer from hydrocephalus again**]**
  - **antibiotics until csf is clear followed by new shunt insertion** [long term treatment: takes for about 6- 7 weeks to clear the csf . when do surgeons insert new shunt again ? if three cultures were negatives & normal cell counts] Antibiotic prophylaxis is controversial however it was found that intra-operative antibiotics or antibiotics for the first 24 hours give the best results

Endoscopic third ventriculostomy:

- Used in occasion cases In obstructive hydrocephalus not communicated hydrocephalus
- devised by dandy (very high morbidity & mortality)
- The endoscope is passed through a burr hole to the third ventricle **[very simple but has to be very specific]**
- the floor of the third ventricle is fenestrated just anterior to mamillary bodies (So we bypass the obstruction in the aqueduct or posterior fossa). The hole is enlarged by introducing the endoscope or an inflatable balloon.

Neural Tube Defect (NTD):

Anomalies arising from incomplete or faulty closure of the dorsal midline embryonal structures

The neural tube closes like a zipper during the first 4 weeks of fetal development starting at the cervical area and moving down toward the sacral area. If the neural tube fails to close at any point, all areas below of the point at which the 'zipper' gets caught are affected. The sooner neural tube development stops, the more severe the birth defect.

The most common areas directly affected in spina bifida are the lumbar and sacral area of the spine

Sacral region : inner legs and maybe their feet will be involved in the disability .They may be able to walk on their own or they may need a little help, although parts of their legs may experience numbness. They may or may not have bowel and bladder control

Lower lumbar region (L4-L5): They may be able to walk on their own or they may need a little help, although parts of their legs may experience numbness. They may or may not have bowel and bladder control

Upper lumbar region (L1-L3) : numbness and/or paralysis from the waist down and will probably not be able to walk. They will be wheelchair bound and will not have control of their bowel or bladder functions

Development of the Nervous system :

- Neural plate invaginates as neural folds push up
- Neural folds eventually form nneural groove
- Cells of neural fold eventually meet
- Form the neural tube
- Developmental sequence
- Neural tube runs anterior- posterior along the embryo
- Surrounding ectoderm eventually encloses neural tube
- When neural tube closes off brain and spinal cord are fromed and this process finishes in day 28 from when the pregnancy begins

Spinal cord covered posterioraly by spinal prosseses and laminae

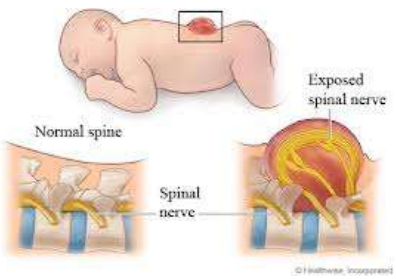
Two groups :

- Spinal dysraphism: by far larger → Myelomeningocele, Meningocele, Spina bifida occulta
- Cranial dysraphism

Raphi=midline fusion dysraphism= faulty closure or faulty union of the neural crest

Myelomeningocele:

- Most important and common dysraphic disorder
- Incidence 0.2-2/1000 live births (varies from place to another in our society it's higher a little)
- Risk increases to 5% if a sibling is affected (the family here is a risk factor , it have to be monitored in following pregnancies)
- Slight female preference



- More common in whites
- **Etiology :**
    - Unknown
    - Genetic Factors
    - Teratogens e.g. Sodium valporate , anti convulsive drugs ,,, (pregnant women should avoid these drugs even before pregnancy)

- 80% are in the lumbosacral region
- Spinal cord and roots protrude through a bony defect
- They can be :
    - Closed: neural tissue lies within a cystic cavity , lined by meningies and other skin
    - Open : dysplastic neural tissue is exposed to air (surgical challenge because you have to generate artificial skin or graft to cover the defect)

      Many of these have neurological deficit such as paraparesis, sphincter and bladder problems

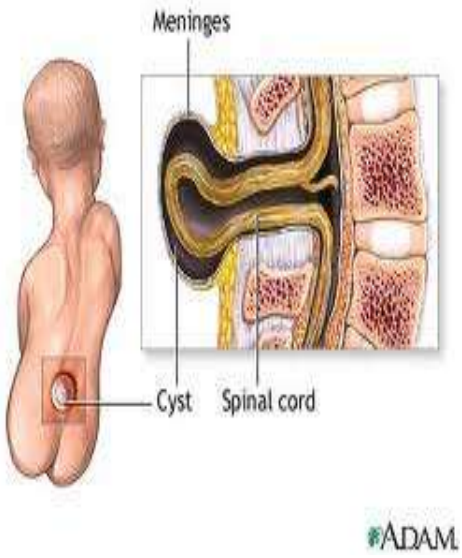
- **Associated anomalies :**
    - Hydrocephalus
    - Chiara type 2
    - Aqueduct forking
    - Many others : microgyria, ectopic gray matter , platybasia ,etc
- **Antenatal diagnosis :**
    - Fetal U/S
    - In high risk patients :
        - MRI more accurate but is kept for high risk patients
        - Screening maternal serum/amniotic fluid for alpha fetoprotein and acetylcholinestrase
        - Contrast enhanced amniography
        - Possibility of therapeutic abortion
- Assessment and management
    - Careful examination of the lesion for (local examination)
        - Presence of neural elements
        - Quality of the skin of the sac
        - Any CSF leakage → surgical emergency should be done within 24-maximum 48 hours because the risk of infections)
    - Transillumination (useful in lesions that are close or covered by skin to see if there is CSF or other solid material)
    - Observe limb movements :spontaneous and to pain (degree and level of neurological damage)
    - Note dilated Bladder and patulous (no muscle tone) annual sphincter (when the child cry he will loose stools spontaneously because there is no sphincter)
    - Look for associated anomalies e.g. hydrocephalous, scoliosis, foot deformities.
    - Investigations
        - U/S
        - MRI

- Treatment
  - Immediate closure and replacement of neural tissue 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.

- Long term care : because it will leave permanent disability even after the surgery
  - Regular follow up to detect problems early and prompt treatment
  - Urological : urinary incontinence ,vesicourteric reflux , repeated UTI,renal impairment , hypertension and stunted growth
  - Orthopedic : feet deformity , tendon transfer , pelvic and spine deformities
  - Neurosurgical : tethered cord, chiari 2 malformation and shunted hydrocephalus

Meningocele :

- Cystic CSF filled cavity lined by meningeis and skin
- Communicates with spinal canal
- No neural tissue
- Seldom any neurological deficit
- Much better prognosis
- Rarely associated with other congenital anomalies
- U/S or MRI
- Tx: excision , urgent in case of CSF leakage



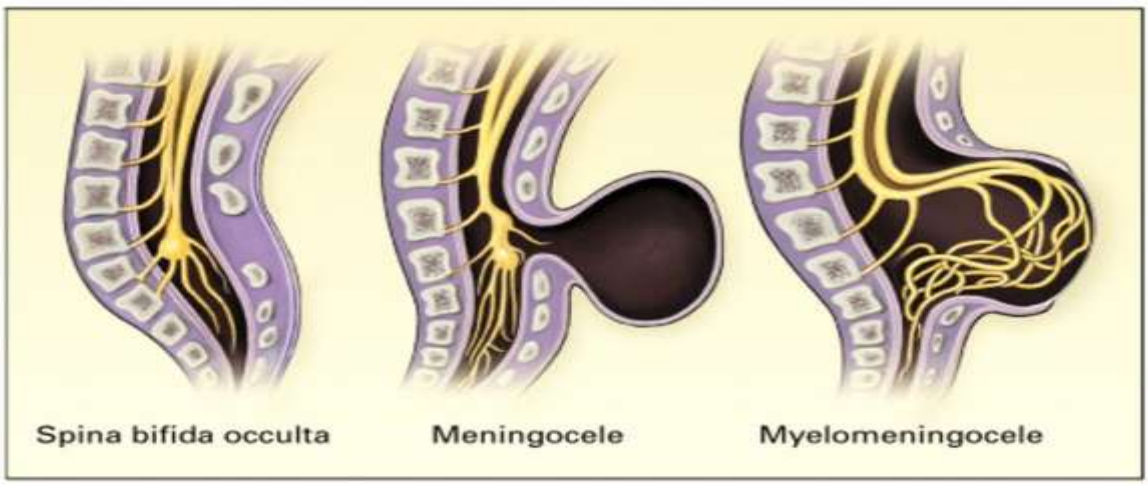
Spina Bifida occulta :

- A bony defect of the lamina
- Usually in the lumbosacral region
- Affects 5-10% of the population (common)
- Clinically not significant
- Usually asymptomatic
- Found incidentally when a patient do x-ray for another reason e.g. accident
- No treatment required BUT rule out associated cutaneous abnormalities

CAUTION!

- In individuals who have lumbosacral cutaneous abnormalities :
  - Tuft of hair in
  - Dimple (small retraction in the skin)
  - Sinus (is a dimple which goes deeper)
  - Port wine stain
  - Subcutaneous lipoma

- High incidence of other occult spinal anomalies
  - Diastomatomelya
  - Intraspinal lipoma
  - Dermoid tumor
  - Tethered cord due to thickened filum terminale



Other spinal dysraphic anomalies:

Diastematomeyelia:

- Localized splitting
- Asymptomatic at least in their early life
- Later, some start developing loss in sphincter control or weakness in the lower limbs.

Congenital anomaly, often associated with spina bifida, in which the spinal cord is split into halves by a bony spicule or fibrous band, each half being surrounded by a dural sac.

Tethered spinal cord

Involve the pulling of the spinal cord at the base of the spinal canal, literally a tethered cord. Whereas the spinal cord normally hangs loose in the canal, free to move up and down with growth and bending and stretching, a tethered cord is held taut at the end, or some point in the spinal canal. In children, a tethered cord can force the spinal cord to stretch as they grow. In adults the spinal cord stretches in the course of normal activity, usually leading to progressive spinal cord damage if untreated. It is often associated with the closure of a spina bifida—though not always, depending on the form it takes. For example it can be congenital such as in tight filum terminale, or the result of injury later in life. And this is one of the causes of recurrent meningitis in a child

Lipomyelomeningyocele:

big fat mass going through subcutaneous tissue through a big defect in the bone.

A Rare congenital condition, where a fatty mass is attached to the spinal cord and protrudes through a defect in the spinal cord.

It forms a mass under the skin and damage to this mass or compression of adjacent spinal cord can have neurological consequences.

Compression effects are more likely to occur if the patient gains or loses weight rapidly - especially during growth spurts.

It can develop anywhere along the spine but is less common in the neck and upper regions of the spine. The condition is often associated with other congenital abnormalities such as cloacal malformations or imperforate anus.

The severity of the condition is variable depending on whether neurological symptoms develop due to the attachment to the spinal cord

Dermal sinus

Cranial Dysraphism:

most are spinal but occasionally we get it in the head

Encephalocele:

a neural tube defect characterized by sac-like protrusions of the brain and the membranes that cover it through openings in the skull

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

In the west, they don't see these cases because they terminate pregnancies with these anomalies

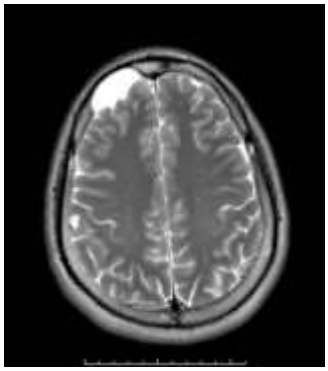
Arachnoid cyst:

Arachnoid cysts are relatively common benign lesions occurring in association with the central nervous system, both within the intracranial compartment (most common) as well as within the spinal canal. They are usually located within the subarachnoid space and contain CSF. prognosis is good

The majority of arachnoid cysts are small and asymptomatic. When symptoms occur they are usually the result of gradual enlargement resulting in mass effect. This results in either direct neurological dysfunction, or distortion of normal CSF pathways resulting in obstructive hydrocephalus [3].

Arachnoid cysts are thought to arise due to congenital splitting of the arachnoid layer with accumulation of CSF within this potential space. The cyst wall is comprised of flattened arachnoid cells forming a thin translucent membrane. There is no solid component and no epithelial lining

- Benign developmental CSF containing cyst
- Predominantly located in the sylvian fissure between frontal and temporal lobe
- Can cause:
  - Increased ICP
  - Convulsions - headache
  - Neurological deficit
  - Endocrine dysfunction
- Imaging:
  - CT (sometimes we can't differentiate between it and between epidermoid tumor)
  - MRI (more precise)



- Treatment:

  - Cystoperotineal shunt (Do CT scan pre and post) → the best
  - endoscopic fenestration
  - excision → rarely

Chiari malformation(CM) and syrengomyelia :

	CM	Syrengomyelia
Definition	CM is a complex developmental malformation characterized by caudal (downward) displacement to variable degrees of parts of the cerebellum, medulla oblongata and 4th ventricle into the cervical canal (it has various type , type 1(mildest),2,3) It may be associated with either syrengomyelia or hydromyelia*	Syringomyelia is the development of a fluid-filled cavity or syrinx within the spinal cord (is cavitations within the spinal cord) Syrengomyelia can occur alone or in association with CM
Presentation	Occipital headache Nystagmus Spastic paresis of upper/lower limbs Ataxia Lower cranial nerves defect Swallowing difficulty	dissociated sensory loss in (cape-echo) distribution wasting of small hand muscles
Diagnosis	MRI	MRI
Treatment	- decompression o f craniovertebtal junction	- syrengostomy - syrengio-subdural shunt

Hydromyelia:

Refers to dilatation of the central canal that occurs in association with CM (dilatation of the central canal of the cord caused by an increase of the)

Hydromyelia refers to an abnormal widening of the central canal of the spinal cord that creates a cavity in which cerebrospinal fluid (commonly known as spinal fluid) can accumulate. As spinal fluid builds up, it may put abnormal pressure on the spinal cord and damage nerve cells and their connections. Hydromyelia is sometimes used interchangeably with syringomyelia, the name for a condition that also involves cavitation in the spinal cord. In hydromyelia, the cavity that forms is connected to the fourth ventricle in the brain, and is almost always associated in infants and children with hydrocephalus or birth defects such as Chiari Malformation II and Dandy-Walker syndrome. Syringomyelia, however, features a closed cavity and occurs primarily in adults, the majority of whom have Chiari Malformation type 1 or have experienced spinal cord trauma

- Sometimes radiology can't distinguish 100% between them.

Central cord syndrome:

- very characteristic syndrome for hydro and syrengomyelia .

- One specific pathway passes through the center of the core , fibers cross the center of the core in front of the central canal , these fibers are mediating the sense of pain and temperature(four qualities of sensation, pain , temperature , pressure and touch) . Fibers mediating touch and pressure remain epsilateral while the fibers mediating temperature and pain cross in front of the central canal to the opposite side . Therefore , any lesion in the center will not affect touch and pressure and will affect pain and temperature →these patient will have dissociated loss of sensation . Dissociated =part of the sensation Is intact (touch and pressure) while the other part is affected (temperature and pain)

-Typically the distribution is in the shoulder, arms and upper trunk we call it (tape like) distribution so if a patient presents with tape like distribution this means that he has most likely intra medullary lesion and the common lesions are hydro or syrengiomyelia

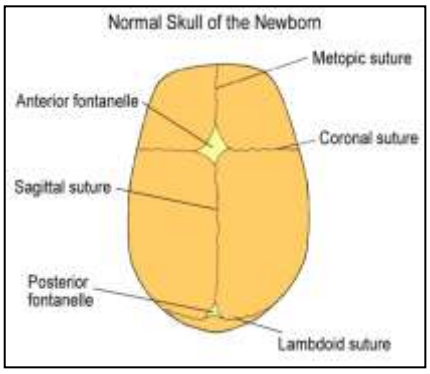
Cranial synostosis :

Syn =together ostosis=early fusion of the bone

-We are born with three patent sutures in our skull

-The coronal, the sagittal , the lamboid and the fourth is important but closes in a mature newborn which is the metopic suture.

-The sagittal suture causes the head to enlarge in lateral direction



Sagittal suture synostosis (Scaphocepaly): scaphoid = السندان

An early closure or fusion of the sagittal suture. This suture runs front to back, down the middle of the top of the head. This fusion causes a long, narrow skull. The skull is long from front to back and narrow from ear to ear.

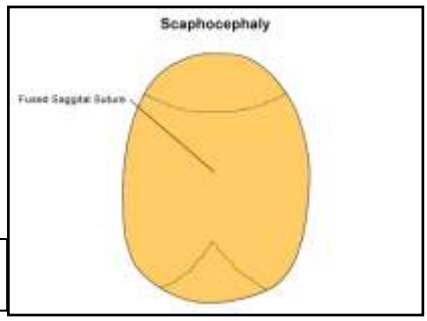


Figure : Scaphocepaly

Metopic suture synostosis (Trigonocephaly):

Fusion of the metopic (forehead) suture. This suture runs from the top of the head down the middle of the forehead, toward the nose. Early closure of this suture may result in a prominent ridge running down the forehead. Sometimes, the forehead looks quite pointed, like a triangle, with closely placed eyes (hypotelorism).

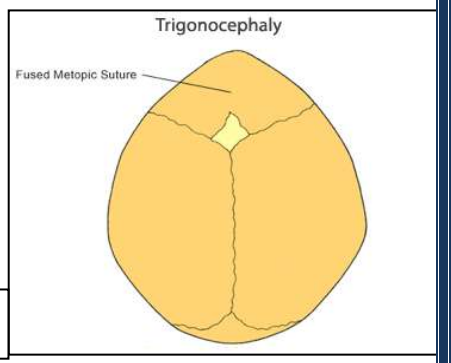


Figure : Trigonocephaly

Coronal suture synostosis (plagiocephaly):

The coronal suture runs across the top of the skull, extending almost from ear to ear .Unilateral coronal synostosis prevents the forehead and brow bone from developing normally.

Global synostosis:

All sutures are involved this is a clear Indication for surgery because it doesn't allow the brain to expand and there for the child may have brain death

What to do?

Try to mimic the natural condition by surgical removal of the suture and also removing bone around it

*Always fix the synostosis even if it was a cosmetic condition because their appearance will have a bad impact on their lives in the future .Moreover, the earlier you correct the condition the easier it would be

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429 surgery team notes

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