

Pediatric Neurosurgery

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Outline

1. Embryology
2. Hydrocephalus
3. Spina bifida
4. Occult Tethered cord
5. Encephalocele



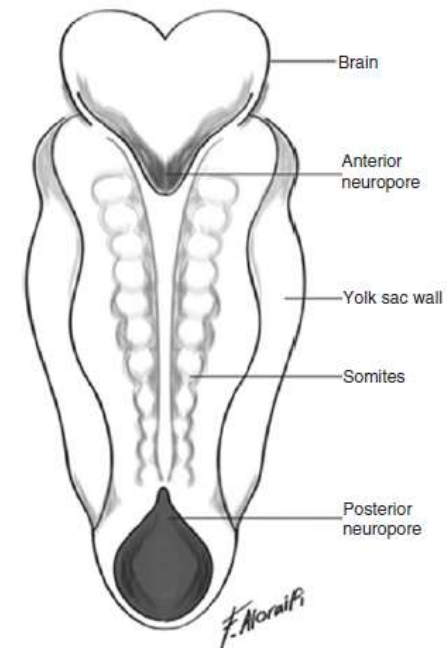
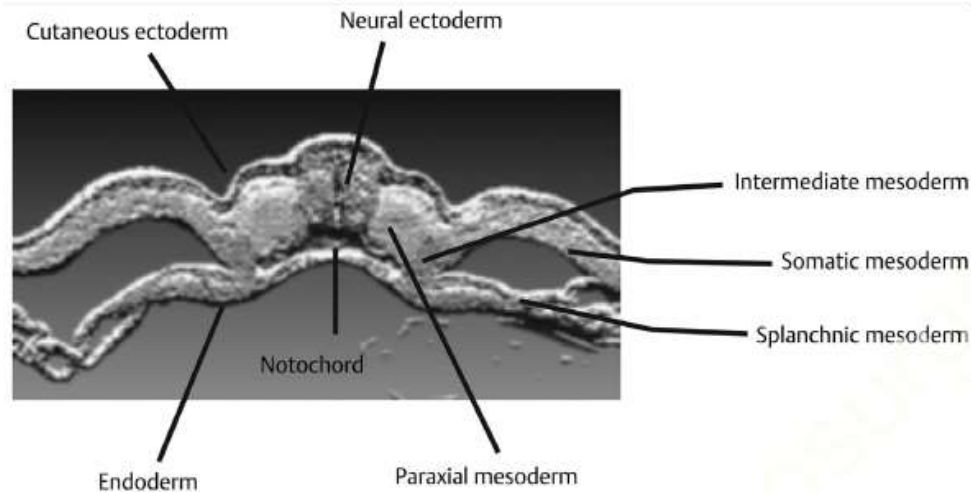
1. Embryology



Neural plate and Neural tube

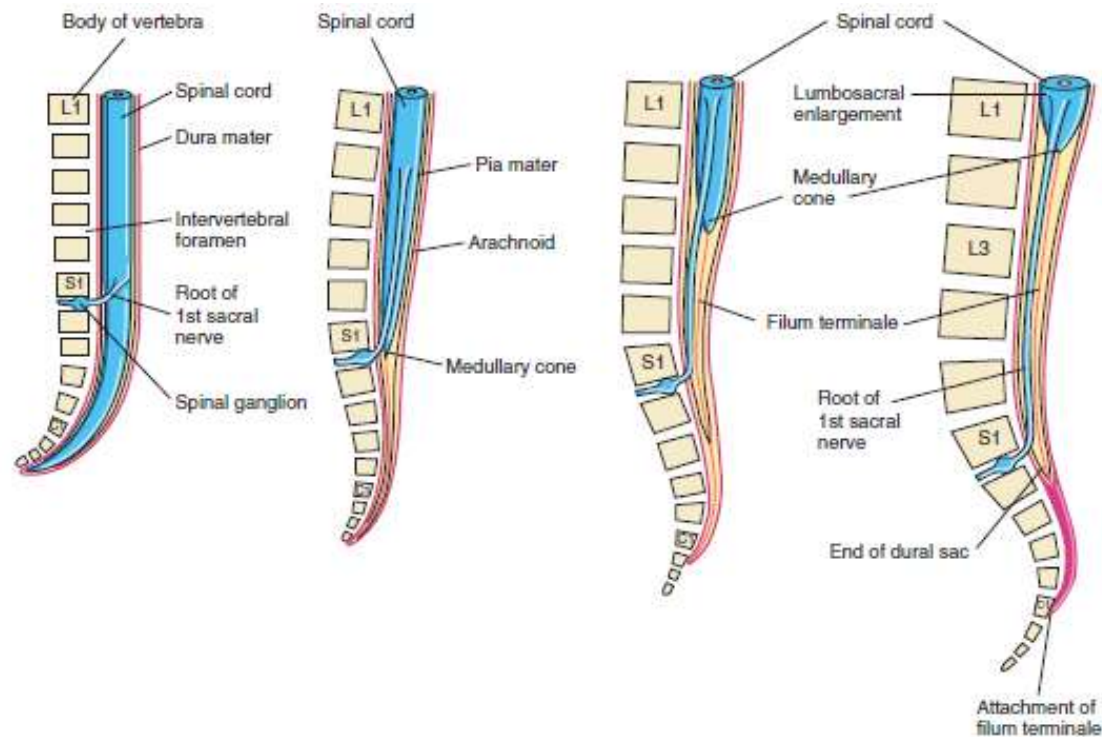
- Notochord (mesoderm) induces ectoderm to form neural plate (future brain & spinal cord)
- **Wk 4**
 - Primary neurulation - Neural tube is formed by fusion of neural folds (middle part>>cephalic/>>caudal)
 - Neural crest develop outside margin of tube (give PNS, microglia, part of meninges with mesenchyme)
- **Wk 5**
 - secondary neurulation - Caudal (sacral) part of the spine is formed by recanalization
 - CSF forms
- **~20 wk**
 - Myelination starts during continues post-natal





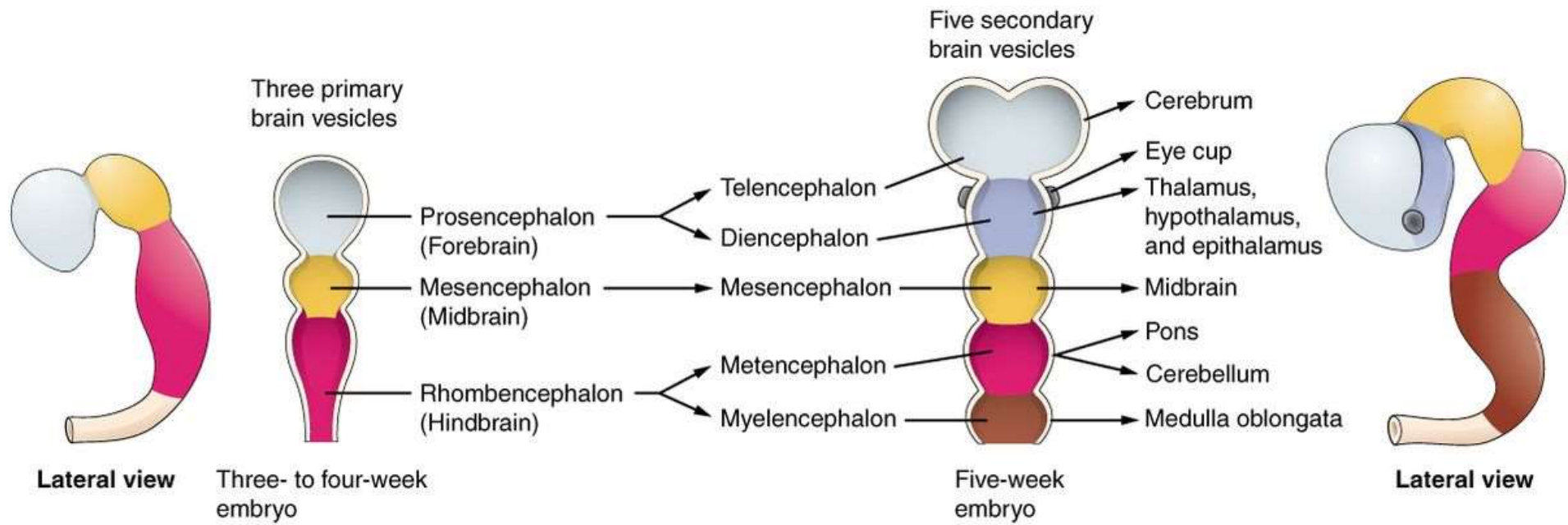
- Chick embryo showing 3 germ layers, notochord and neural plate. As neurulation proceeds the caudal part is last to close





- **Development of the spinal cord: Conus medularis** (end of cord) - position changes- 8 wk – C1, 24 wk– S1, Birth - L2-L3, Adult – L1-L2 . **Cauda equina** (nerves below conus) – initially each pass through its own foramina, later run obliquely. **Filum terminale** (pia matter)- runs downward from conus to attach to coccyx





- **Development of the brain:** Vesicles form on neural tube



2. Hydrocephalus



2.1. Epidemiology

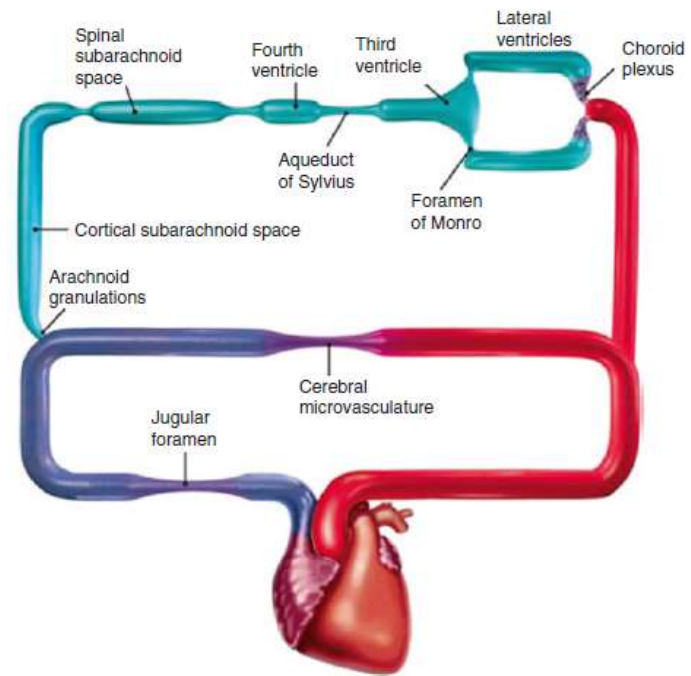
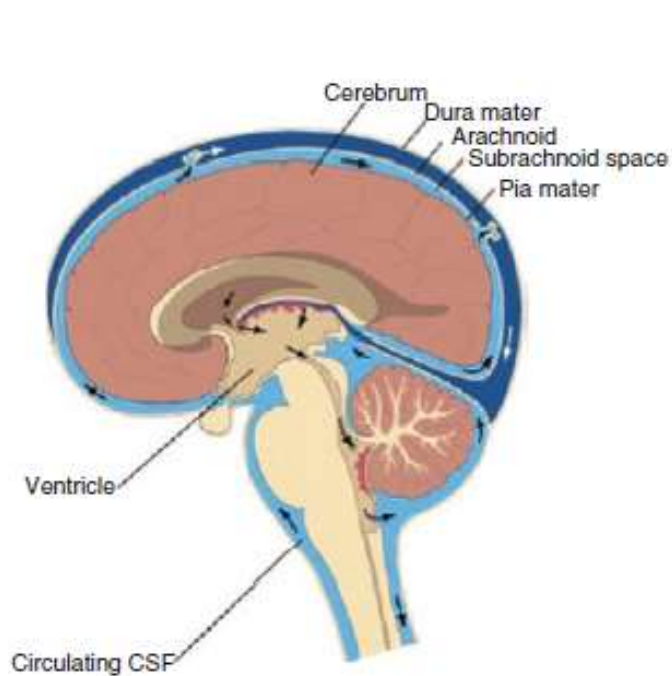
- **0.9 -3 per 1000 live birth**
- common clinical problem in the developing countries



2.2. Pathophysiology

- Excess CSF within the ventricular system and cisterns leads to increased ICP
 - CSF volume (normal = 150ml, 20ml/hr)
- disturbance of prod. (choroid plexus papilloma/carcinoma)
- obstructive (obstructed flow) – ventricle distal to obstruction remain normal
- Communicating (failure of reabsorption- obstruction at level of arachnoid granulation)- all ventricles affected





- **CSF drainage pathways:** A schematic section through the brain and spinal cord showing the drainage pathways for CSF. choroid plexuses of the lateral ventricles >>foramina of Monro >>third ventricle. >> cerebral aqueduct of Sylvius>>fourth ventricle >>basal cisterns>> foramina of Luschka and foramen of Magendie>> subarachnoid space (over the surface of the cortex)>>drains back into the blood via the arachnoid granulations into the superior sagittal sinus



2.3. Etiology

- **Congenital**

- MMC (11-16%) = ass with aqueduct stenosis, chiari malformation
- Post Intraventricular hemorrhage (PIH) = subsequent fibrosing arachnoiditis, 15-30% may require shunt
- Primary aqeductal stenosis (20%)
- Posterior fossa malformations
 - Chiari herniation of cerebellar tonsils and medulla through foarmen magnum (into spinal canal)
 - Dandy walker malformation hypoplastic cerebellar vermis

- **acquired**

- Post Infectious (PHH) (60% in developing) – intrauterine, bacterial meningitis, CNS TB
- tumor - post fossa (4th Ventricle), selar/pineal (aqueduct, 3rd ventricle), choroid plexus ca (inc prod)
- Postop



2.4. Presentation

- **Raised ICP** - Irritability, dec LOC, vomiting, poor feeding, developmental delay, FTT, seizures
- Inc head circumference and bulging fontanel, splitting sutures and of scalp veins
- papilledema, CN palsy, Drowsiness and lethargy, and ultimately coma.
- eye lid retraction & loss of up-gaze (sun setting sign)
- Opisthotonic posturing, bradycardic and apnoeic episodes



2.5. Investigations

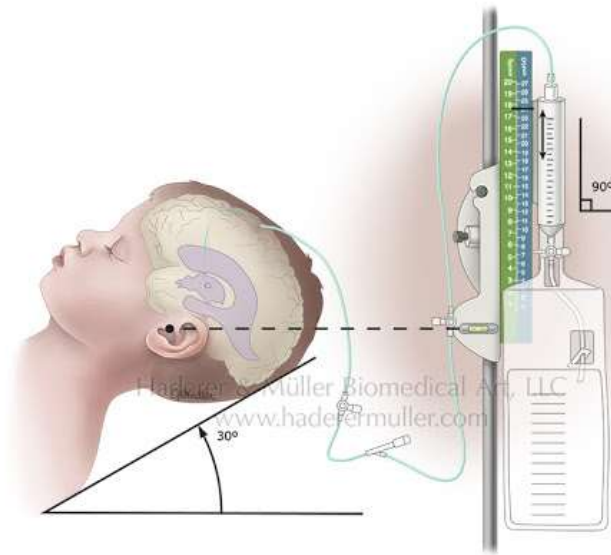
- **U/S** - Screening choice for <18 mo
- **CT** – for simple shunting
- **MRI** – mandatory for endoscopic surgery
- **CSF analysis** - infection or haemorrhage is suspected,



2.6. Treatment

- **Pharmacotherapy** – *not effective in decreasing HCP*
- **LP drainage** – *historically practiced, but now shown no benefit rather inc risk of infection*
- **EVD (External ventricular drain)** - temporary (PHH, infection, high protein)
- **Subcutaneous reservoir ventricular catheter** - temporary
- **VPS (Ventriculoperitoneal shunt)** – mainstay
- **ETV (Endoscopic Third ventriculostomy)**- especially for aqueduct stenosis





- **EVD** drained into a sterile closed circuit system which can be continuously controlled by a simple gravity based outflow valve Infection and dislodgement are potential complications.

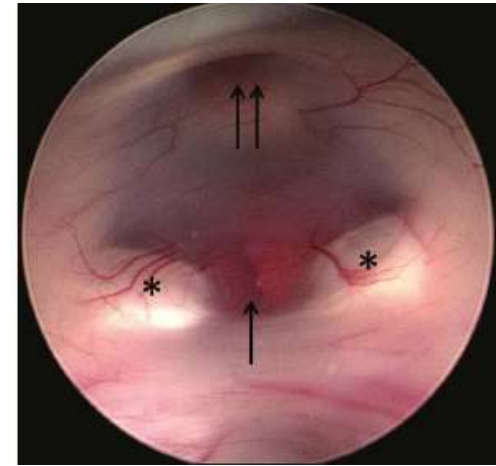
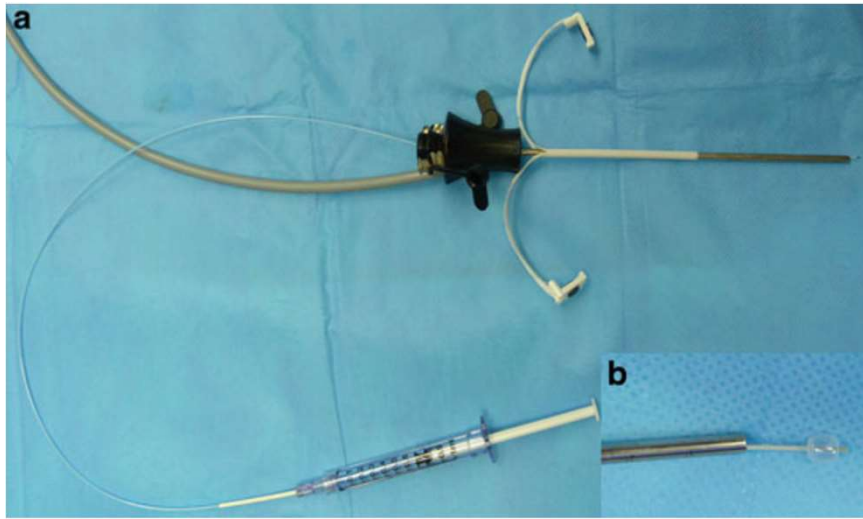




- **subcutaneous CSF reservoir** – frontal subcutaneous reservoir with catheter entering ventricle. allows repeated bedside aspiration of CSF, has lower infection rates compared to EVD. Complications and limitations include skin erosion and only intermittent intracranial pressure control.



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- **ETV** : Endoscope placed to the ventricle via a burr hole. Opening created on the floor of the third ventricle for CSF to bypass the obstruction and absorbed naturally. avoidance of foreign body implantation and a more physiological CSF circulation.



ETV vs Shunting

- **Immediate surgical risk**
 - Mortality= shunt < 0.1%, ETV up to 1% (damage to brain structures and adjacent vessels)
- **long-term follow-up,**
 - Overall complication= shunt >40% and mortality rate raises, ETV 6-20%
 - Infection = 1-5% for ETV and less severe, 1-20%for shunting
- Children younger than 12-18mo have high comp in shunt as well as ETV



ETV success score

- advantages of shunt independence makes it an attractive option but patient selection is crucial
- **(ETVSS) - patient's age, HCP etiology, previous shunt surgery**
 - High failure = Young , previous shunt, Postinfectious
 - Medium = nontectal brain tumor, MMC, and history of IVH
 - good indications for ETV = aqueductal stenosis and tectal tumors as well as other typical occlusive three-ventricular entities are good risk, with age >1yr and no previous shunt
- Role of ETV as **secondary option after multiple shunt failure** is gaining popularity
 - For PHH or PIH (majority of neonatal HCP) after primary shunt failure



2.7. Ventriculoperitoneal shunt



Consideration: Indications

- **Whenever multiple compartments need to be drained**
 - indication for endoscopic fenestration must be considered.
 - If it is unavoidable to put more than one central catheter, it is generally recommended to connect those in front of the valve, otherwise differential ICP can cause problems.
- Ongoing CNS infection is an absolute contraindication for shunting
- Inc blood cell count and/or protein content of the CSF better managed with temporary EVD



Consideration: Types of shunt

- A broad **variety of shunt devices** on the market.
 - Prevent Over-drainage - **antisiphoning or antigravity devices** (standard differential pressure valves do not properly address over drainage in upright position)
 - Prevent infection - **antibiotic-impregnated systems** (AIS) is recommended.
- **mechanism of hydrodynamics** – remains personal choice
 - Differential pressure valve (most widely used) valve will let the CSF flow out of the valve when the pressure difference drops below a predetermined pressure threshold. (low, medium, and high)
 - Flow-controlling shunts - The progressive narrowing of the outflow orifice controls the flow of CSF
 - Adjustable valves. They permit the adjustment of valve opening pressures with an external controller



Consideration: Drainage site

- VPS (peritoneal)
 - preferred unless complicated abdomen (peritonitis, adhesions)
- VAS (Atrial) (via central venous access)-
 - Sudden death from pulmonary embolism, endocarditis and nephritis
 - need to lengthen the lower end as the child grow
- Other - pleural space, bladder, biliary tract
 - limited resorption capacity (infant's pleura can't cope with absorption of effusion)
 - Last choice

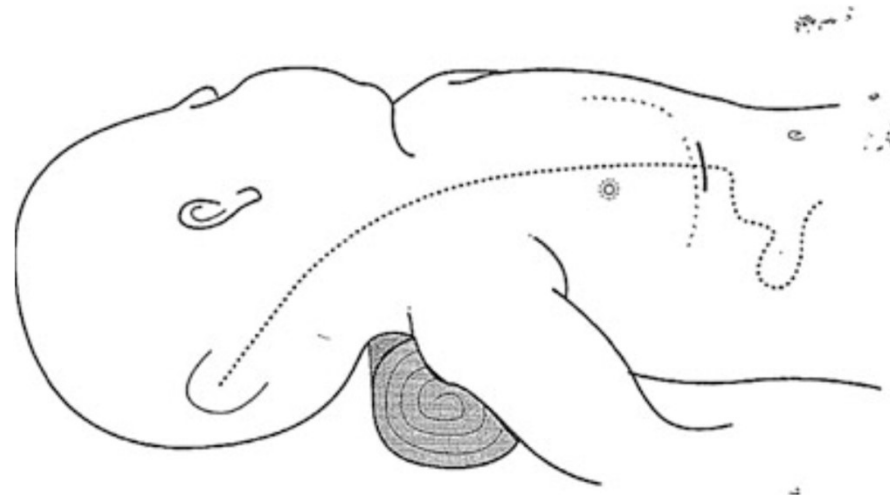


Consideration: Shunting protocol

Proven to dec infection rate

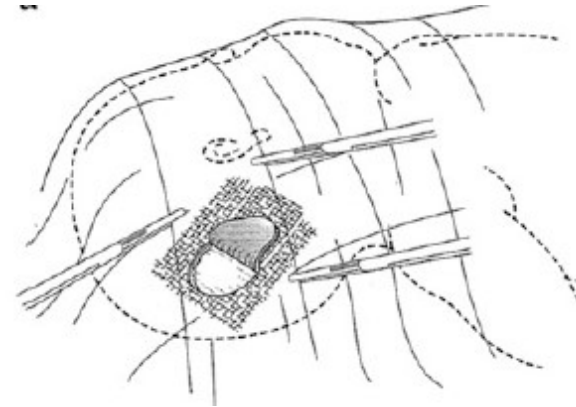
- Shunt surgery, if possible, no. 1 on schedule
- Experienced surgeon at the table
- Doors of operating room closed
- As few staff as possible
- Thorough and repeated skin disinfection by surgeon
- Antiseptic hair washing (in theater)
- Keeping hardware in sterile inner packing as long as possible
- hardware flushing and keeping in fluid (prevent air contamination)
- Don't touch hardware with sharp instruments or fingers (if avoidable)
- antimicrobial sutures





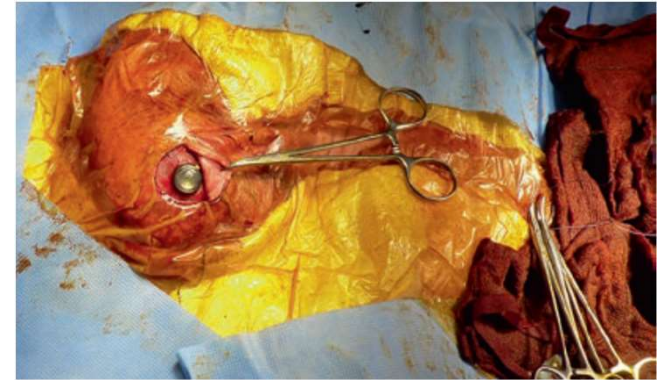
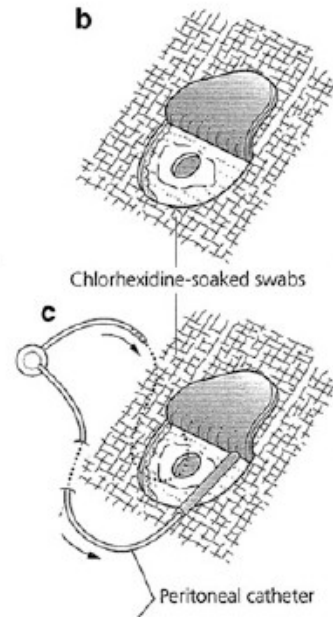
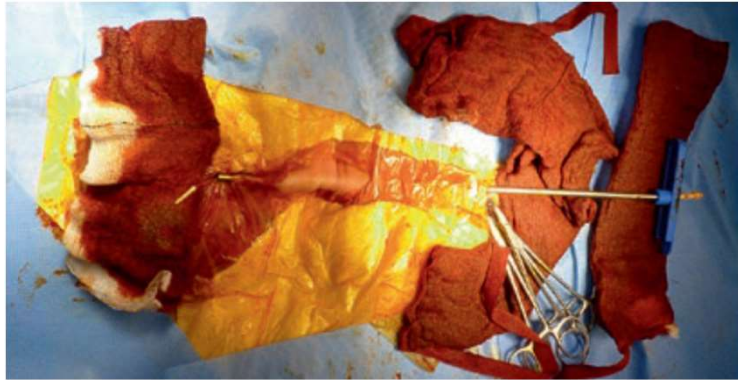
- **VPS positioning:** Head turned 45 degree to opposite side. Roll of Gamgee under the shoulders to **straighten out the neck** to allow easier passage of the tunneling cannula. Incision sites are marked. Two standard burr hole sites do exist: frontal precoronal (Kocher's point) and parieto-occipital (Frazier's point). **Keen's point** is located 3cm superior and 3 cm posterior to helix of the ear. Choice depends on the ventricular shape and surgeon's preference.





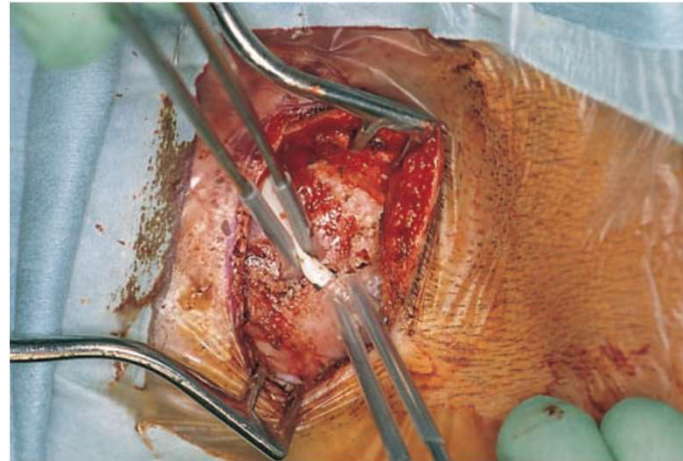
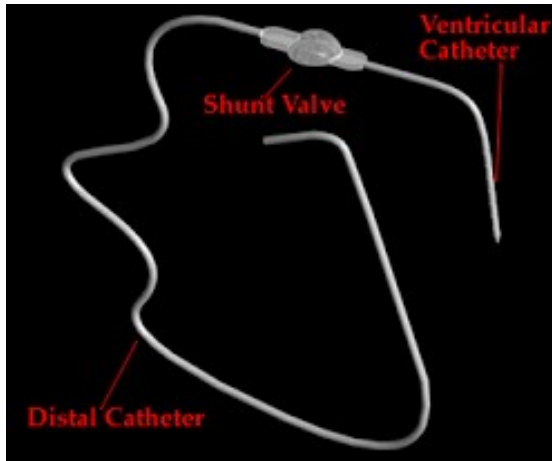
- **VPS ventricular phase: Draping** must allow palpation of important landmarks (tip of the nose, ipsilateral eye, and ear). Upper incision to the pericranium is made. The pericranium is diathermied and scraped peripherally and a **burr hole** made. In young infants, this is easily done by using a dissector or a knife as a drill. The periosteum is not detached from the skull in order to avoid ingrowth of the shunt components into the bone.





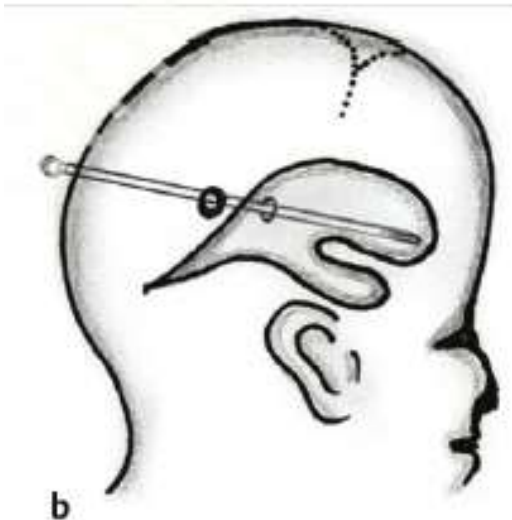
- **VPS shunt tunneling phase:** The tunneller/trochar & cannula passed percutaneously from the abdomen to the right parietal incision or from abdominal incision. The long trocar is removed, allowing passage of the peritoneal catheter along the metal cannula. Dura is opened using a knife or a small bipolar forceps, and the brain surface is coagulated. Dura is gently diathermied, and a small hole is incised in it allowing the ventricular catheter to be introduced and connected to a reservoir.





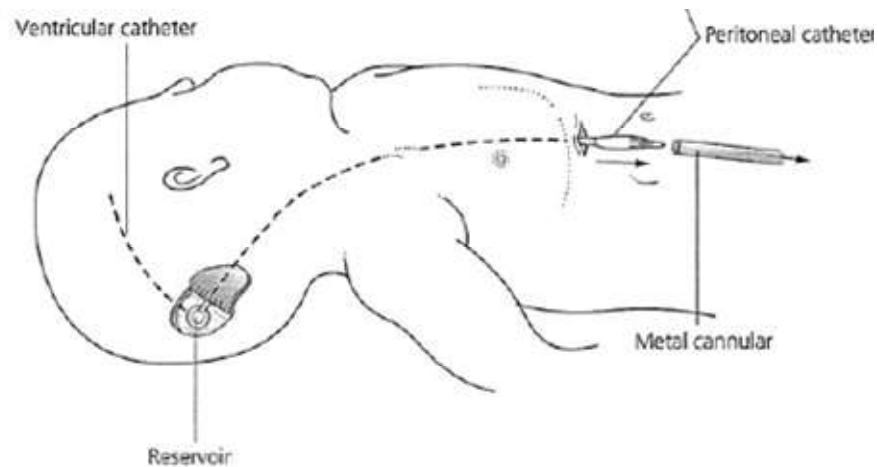
- **VPS shunt components and handling:** Most have three components: ventricular catheter, valve, and peritoneal catheter. Components are made of Sylastic, impregnated with a radiopaque material for radiographic localization. Shunt contact with the patient's skin or surgeon's gloves is avoided by the use of blunt-tipped or sleeved forceps and clean gauze





- **VPS catheter placement:** Place catheter deeply inside to avoid ingrowth of choroid plexus and early obstruction. Dura is penetrated perpendicular aiming at the inner canthus of the ipsilateral eye. Should feel entering the frontal horn 3 to 5 cm deep. The catheter is advanced 1 cm further before the stylet is withdrawn. The rest of the catheter is advanced without the inner stylet at a depth of 5 to 6 cm from the cranial vault. Fixing attachment at the outer border of the burr hole, to stitch it to pericranium and to protect the catheter from the overbending. Distal catheter is cut in a length that permits to locate the valve 2 to 3 cm distal from the burr hole (to facilitate a revision procedure through only one incision when needed)





- VPS closure:** Check for free flow from the distal (abdominal) end. If no spontaneous CSF flow occurs, the catheter end is held down in order to produce a negative pressure. As much length of catheter as possible is **placed in the peritoneal cavity** to avoid reoperation. The peritoneum is closed snugly around it. A pocket is performed between the epicranium and the periosteum for valve body and flushing chamber. All connector points will be secured with non-resorbable ligatures. The knots are placed towards the skull in order to avoid skin erosions. Generally, the shunt hardware should not be placed underneath a skin incision.



Postop care

- ECG and oxygen saturation during sleep, and when the child is without surveillance,
- regular neurological and abdominal checks and daily wound inspections.
- antibiotics for 24 hours,
- mobilization to be delayed for 6 to 12 hours,
- Discharge to home after 24 to 48 hours. A final CRP is taken before discharge.
- Wound healing should be controlled 2 weeks before exposure to water and physical stress is allowed



Postop imaging

- depends on the intraoperative course and the specific patient condition.
 - ultrasound of the brain - recommended as soon as possible.
 - Abdominal ultrasound - position of catheter and free fluid as sign of a functioning shunt.
 - Early MRI or CT scans - recommended to perform sectional imaging after 3 months
 - Earlier for complicated surgery, older children, symptomatic for shunt malfunction.



2.8 VP shunt complications



Early comp: No spontaneous CSF flow Also resistance to gentle aspiration

- **causes**

- ventricular catheter retracted out of the ventricle, or cerebral parenchyma
- blood clot, brain parenchyma, or choroid plexus. blocks the catheter
- assembly of the components have been erroneous.

- **management**

- Gently inject 2 cm of saline solution to unblock the flow.
- If resistance persists, parenchymal or cisternal location is suspected. Catheter has to be relocated



Early comp: Intracerebral or ventricular hemorrhage

- Blood flows along with CSF through the catheter.
- especially common among patients with coagulopathies
- Usually, stops spontaneously or with saline solution instillation through the catheter.
- immediate postoperative CT is mandatory
 - define the extent of the hemorrhage
 - decide whether to keep the shunt in place or change it for an external drainage.
- Rarely, an open craniotomy is required to explore and directly coagulate the injured vessel.



Early comp: Abdominal

- **Misplacement of catheter**
 - Pre-peritoneal location- resistance on placement, CSF leak through wound
 - Relocate catheter
- **Injury to viscera.**
 - peritoneal cavity might no longer be the option for CSF drainage.
 - Select to either transform for an external drainage or modify the distal location of the catheter



Early/late comp: over-drainage

- Development of **subdural hematomas**
 - in newborns with large ventricles and thin cortical mantle
 - May be avoided by high resistance valves
- **Slit ventricle syndrome** (very small ventricles) seen commonly in those shunted <2yrs



Late comp: Shunt malfunction

- 40% fail in first year
- 80% require revision after 12 years
- **Proximal obstruction**
 - Protrusion of the choroid plexus, infiltration from tumor,
 - collapse of the ventricular walls, glial tissue from astrocyte proliferation, debris, or blood
- **Distal obstruction**
 - Migration
 - Omentum obstruction



Late comp: infection

- 8-12%, 5% for antibiotic impregnated shunt
- insertion at younger age, prematurity, and CSF leak are recognized risk factors.
- **Bacteria:** Staph epidermidis, Staph aureus, Klebsiela, Pseudomona, and Candida albicans.
- **Prevention**
 - Shunt protocol
 - additional incisions where the passer stops, before traumatizing the skin when trying to force our dissector
 - avoid incisions over the burr hole to reduce the risk of CSF fistula or the exposure of the catheter entry.
- **Treatment** – usually require removal, EVD and intrathecal antibiotic



Late comp: Shunt erosion

- in malnourished and premature patients
- **Prevention**
 - Select the smallest shunt system available.
 - Draw a larger scalp flap with extensive skin dissection.
 - Avoid suture lines over any of the shunt components.



Late comp: shunt migration

- Intestine or other sites
- Identify the location and patency of the connections of the system by X-ray films.
- Remove the system. Disarticulate the components and pull out the distal catheter from exposed site
- Never try to pull it out through the cephalic incision or along the subcutaneous trajectory



Late comp: other

- **peritoneal pseudocyst**
 - Previous shunt infection or repetitive revisions are probable risk factors.
 - inflammatory cells and fibrous tissue lining on the surface of the distal catheter.
 - The removal of the whole system (related to bacterial colonization)
- **Seizure**
 - 20% to approximately 50%
 - Early shunting in younger than 2 years and history of shunt complications are significant risk factors



2.9. Prognosis

- Depends on
 - Age of onset (impact on developing brain)
 - Etiology (ass. With other brain malformations)
 - complications (Preterm infants with a history of posthemorrhagic hydrocephalus and MMC patients)
 - Obstruction
 - Infection



3. Spina bifida



3.1. Epidemiology

- 0.5–1/1000 live births
- incidence of neural tube defects is less common today and is falling
- Higher prevalence in Ethiopia (6 per 1000 live births)

ORIGINAL ARTICLE

**PATTERNS OF NEURAL TUBE DEFECTS AT TWO TEACHING HOSPITALS IN
ADDIS ABABA, ETHIOPIA A THREE YEARS RETROSPECTIVE STUDY**

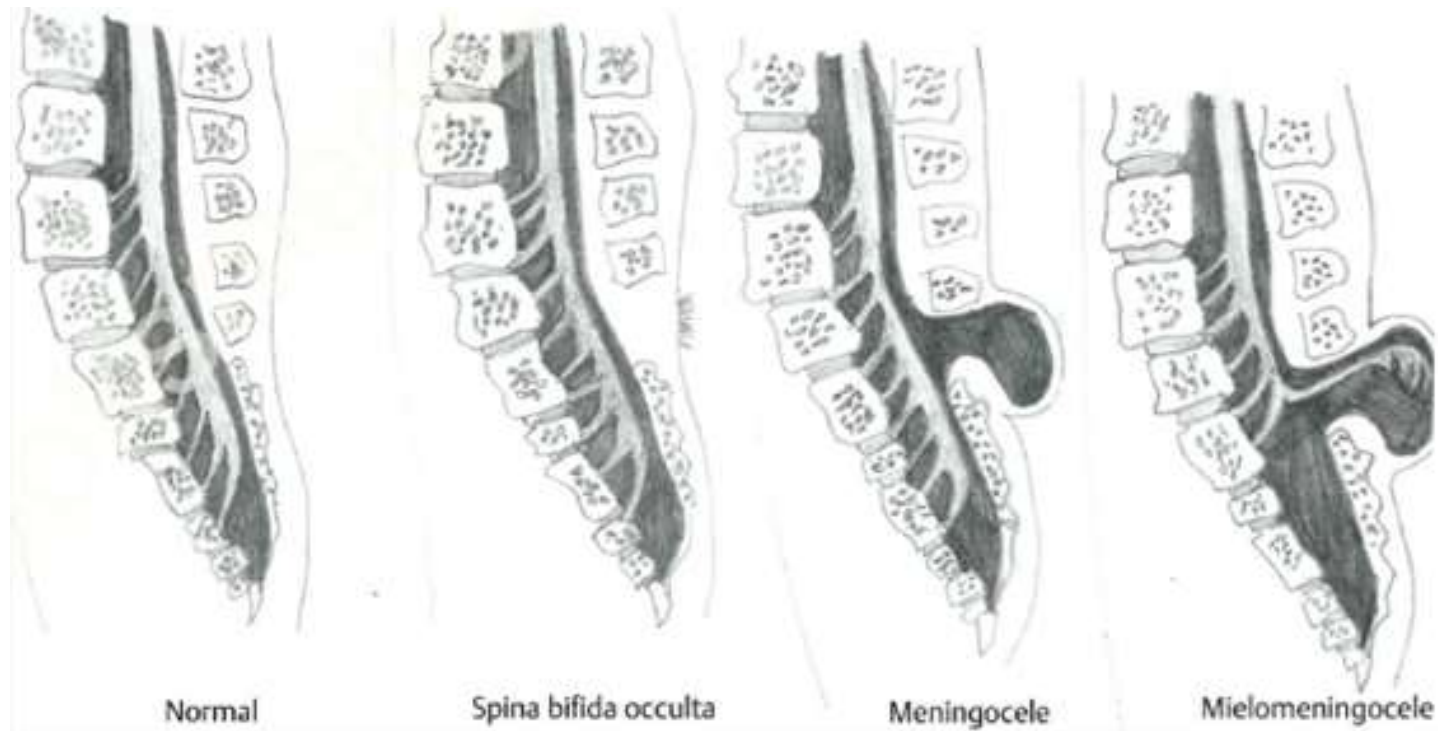
Gemechu Sorri, MD¹ Eyasu Mesfin, MD¹



3.2. Classification

- **Spina bifida occulta (5%-10%)** = secondary neurulation defect
 - Defect in posterior vertebral arch (Skin covered defect)
 - Associated lesions = (discussed later in tethered cord section)
 - clinically insignificant unless causes tethered cord
- **Spina bifida aperta (95%)** = Primary neurulation defect
 - open neural tube (exposed tissue & CSF leak)
 - **in meningocele skin usually intact





- **Spina bifida occulta** = 6 cutaneous markers (discoloration/hemangioma, tuft of hair, dimple/sinus, lipomatous mass, atretic MMC) **Spina bifida aperta** = fluctuant midline mass could be meningocele (30% tethered cord) or myelomeningocele.



3.3. Pathophysiology

- failure of closure
- persistent neural placode with concomitant **primary neurological damage**.
- exposure to the amniotic fluid leads to **secondary neurological damage**
- 73% of these infants will be **allergic to latex**.



3.4. Risk factors

- folate deficiency (higher incidence of folic acid deficiency in Ethiopia- 30%)
- family history (genetic)
 - Inc to 1 in 200 if one parent affected, 1 in 20 if one child affected, 1 in 8 if two are affected
- Anticonvulsants (valproate and carbamazepine)
- Diabetes and obesity

Prevalence of Anaemia, Deficiencies of Iron and Folic Acid and Their Determinants in Ethiopian Women

Jemal Haidar

School of Public Health, Addis Ababa University, PO Box 27285/1000, Addis Ababa,
Ethiopia and Ethiopian Health and Nutrition Research Institute, Addis Ababa



3.5. Presentation

- midline cutaneous defect, more often lumbosacral (80%)
 - *Occasionally more than one lesion can be found and there is often marked kyphosis or scoliosis present
- Neurological deficit with variable severity. (Paraplegia, urinary dysfunction)
- Associated nervous system developmental anomalies.
 - *chiari II malforamation – always associated but symptomatic in 10%
 - *Hydrocephalus – in 85%



3.6. Investigations

- **MRI** is the most reliable study
 - Not mandatory in myelomeningocele cases,
- **Plain X-ray and CT scan** with 3D reconstruction
 - precise site of the dysraphism bone defect and other orthopedic anomalies.
- **EMG**
 - evaluate the extent of neurological damage





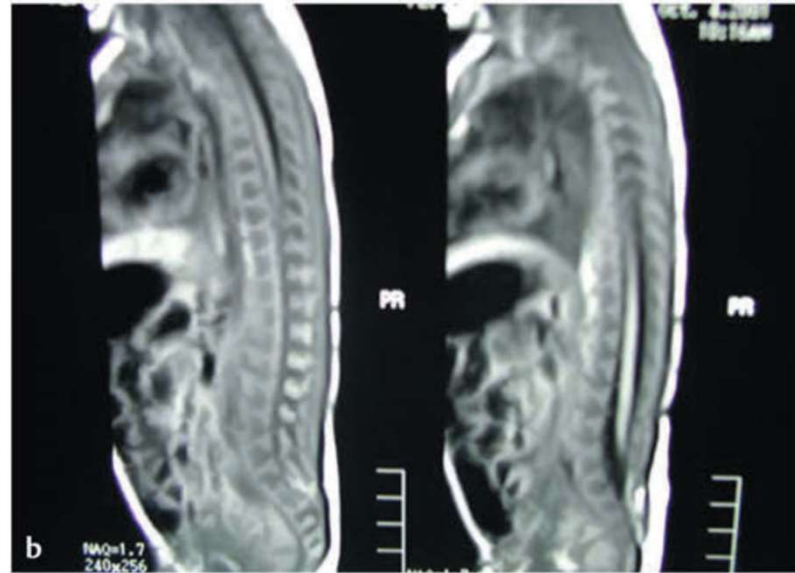
- **Myelomeningocele** – a lumbosacral lesion is demonstrated showing a central neural plaque surrounded by dystrophic epidermis





- **Thoracic Meningocele:** intact skin covering sac of meninges





- **Atretic meningocele:** characteristic blue hue. Sagittal T1-weighted MRI showing absence of communication between the spinal canal and subcutaneous tissues.





- **Anterior meningocele** = T1-weighted sagittal MRI shows point of communication of sacral MMC



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3.7. Treatment



Preop considerations

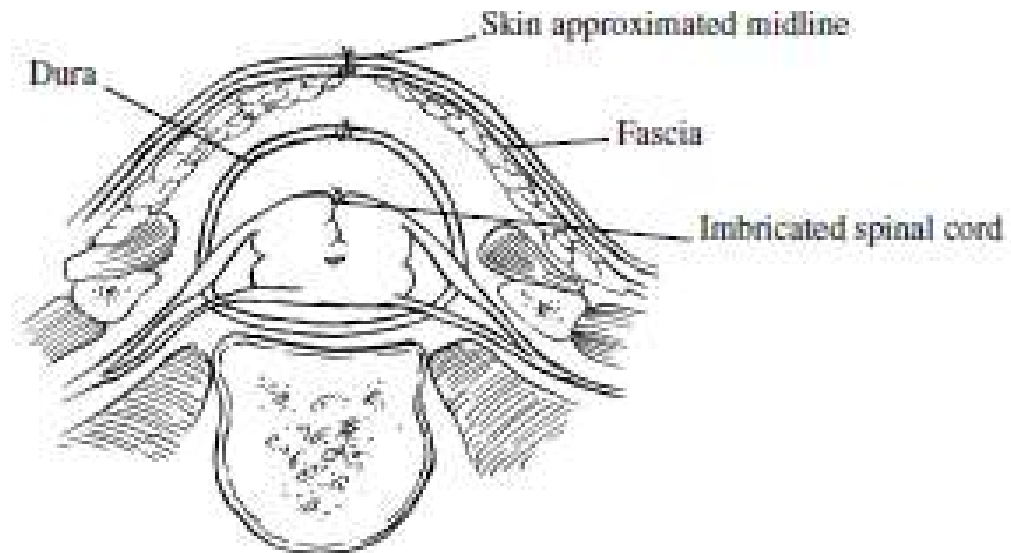
- **Repair should be done as soon as possible**
 - Repaired within the first 5 days
 - shorter hospital stay, antibiotic therapy, and a lower complication rate.
 - Repaired 72 hours after birth
 - Increased febrile UTI, VUR, hydronephrosis, and secondary tethered cord
- **Initial care-** for all lesions not covered with skin
 - **Position** lateral or prone to ensure no pressure is placed on the neural placode
 - **Covered** in sterile saline-moistened gauze and wrapped in plastic cling film to minimize dehydration and to prevent contamination.
 - **antibiotic** if ruptured



Goal of Surgery

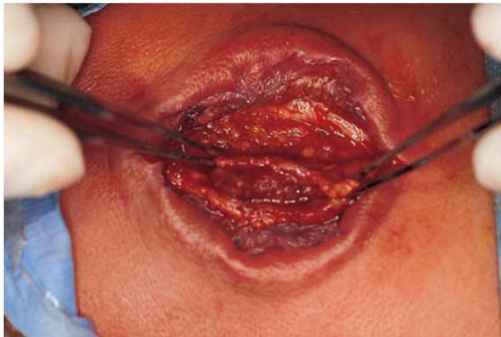
- **Preserve existing function**
- **Prevent an infection of the CNS**
- **Prevent leak** (watertight closure of dura and good skin cover)
- **Prevent secondary tethering**





- **Primary closure:** principle of surgery is to reconstruct the spinal cord by five-layer closure.





- **Primary closure: Lumbosacral MMC.** Incise and **drain CSF**. identification of the placode>>resection of cutaneous elements from edges of the placode>> **approximation of the lateral edges of placode** to transform into a neural tube>> **mobilization of underlying dura** from fascial layer>> **creating a dural sac** as spacious as possible >> lumbosacral fascial layer (optional)>>subcutaneous tissue, and **skin**. table is in the Trendelenburg position to minimize CSF drainage from the spinal defect.



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- **Primary closure:** closure of the skin is usually possible. Sometimes it may require considerable undermining and lateral releasing incisions to allow mobilization. May also need additional complex flap techniques, Associated vertebral kyphosis may occasionally require an osteotomy to facilitate closure in the primary setting



3.7. Outcome



Wound complications

- the commonest complication
- tension on the skin edges or skin necrosis, CSF leak, infection and dehiscence
- Infection is 1–1.5% and usually occurs 5–7 days
- involvement is deep to the lumbodorsal fascia, it may cause meningitis or ventriculitis.
- Treatment
 - vigorously treated with local dressings, systemic antibiotics.
 - EVD, if hydrocephalus is present and CSF leak occurs.
 - if CSF fistula occur, immediate repair is preferred rather than conservative treatment.



Bladder and bowel issues

- Multidisciplinary team follow up to look for delayed complications & hidden associated anomalies
- only 10% of MMC have normal bladder (true continence)
 - **Urodynamic testing** in newborn period are useful
 - Monitor with **ultrasound** for silent renal deterioration
 - The introduction of **CIC**, pharmacological and external devices, biofeedback, has preserved renal function and upto 75% can be socially continent
- Most have active internal anal sphincter and tend to have a degree of constipation
 - **Bowel washout programs** also significantly improve fecal continence



Life style concerns

- **only 25% capable of employment**
- **48% unable to live without supervision**
- **40-85% are ambulatory** – capacity related to intelligence, orthopedic deformity, level of lesion
 - Below L5- most are ambulators
 - L3-L5 – functional ambulators (need support, mostly in weelchair)
 - L3 wheelchair bound
- **31% mentally delayed,**
 - Patients with episodes of shunt malfunction (blockage and infection) have lower intelligence
- Mortality
 - 85% survive infancy,
 - **14% don't survive past 5 yrs,**
 - ?41% died before 16 yrs



Recurrent tethered cord

- Upto 70% have radiologic tethering
- Secondary tethered cord with deterioration of symptoms in **30% of MMC**
- **adhesions of placode** during primary closure (limited dimensions of spinal canal at site of MMC)
- Try to prevent adhesion by
 - remodeling of the placode into a neural tube (surgical neurulation)
 - trying to create a dural sac as spacious as possible.



Hydrocephalus

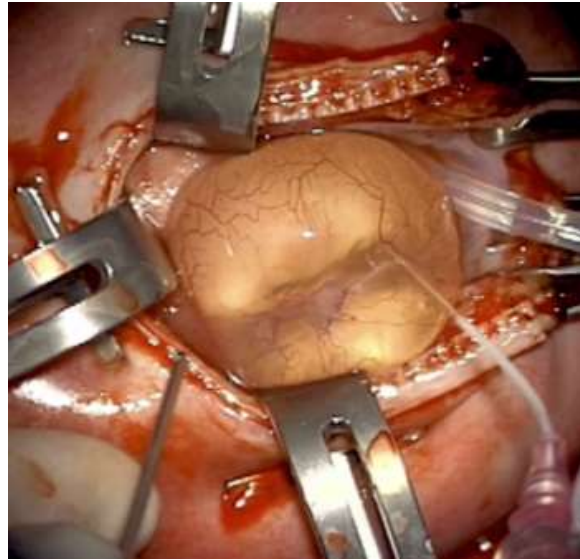
- Occurs in **85% of MMC**
- ~15% at birth, commonly noticed at 2 to 3 weeks of age (after closure)
- controversy over whether **simultaneous CSF shunt insertion and repair** of MMC increase shunt-related complications (infection, malfunction). Some recommend to **wait at least 5 days** after repair.
- Most closures are well tolerated but occasionally closure can result in **accelerated hydrocephalus**
 - generally compensated well with an open anterior fontanelle.
 - Shunting will be required when the head circumference exceeds 2 SD over the 90th centile, when there is rapid increase in HCP , ventricle >50% of skull, persistent CSF leakage from the closed back lesion.



Antenatal management

- **AFP** persistently raised at 16-18 wk
- Detail exam with **U/S or MRI**
- **Discussion to terminate** should involve obstetrician, pediatric surgeon and neurosurgeon
 - considering the level and size of the lesion & presence or absence of hydrocephalus
 - Long term issues (mobility, intellectual, continence)
- Most advocate **planned C/section**
- **Fetal surgery** is controversial





- **Fetal intervention** (repair through mini-hysterotomy): MOMS trial showed significant **reduced rates of shunt placement** at 12 mo (40% instead of 80%). Long term development of secondary tethered cord is not settled



Prevention

- **Periconceptual Folic acid Supplementation**
 - 0.1–0.4 mg/day
 - 4-5mg/d for those at high risk
- **Nutrition**
 - legume (bean, pea, lentiles), dark green Leafy vegetables
- **Fortification**
 - It is difficult to fortify foods in Ethiopia because no staple food has been identified as widely consumed in the entire country.



4. Occult tethered cord



Etiology

- anatomy of **dysraphism** creates tethered cord (strongly associated)
 - Tight filum (Thickened, fatty, short)
 - Intraspinal Lipomas (on conus(70%), filum terminale, or -subpial)
 - Split cord malformation
 - Limited dorsal myeloschisis (fibroneural stalk connecting the skin and the cord)
 - Dermoid fistula / cysts (may also have accumulation of dermis in spinal canal and infection)
- Caudal spinal cord malformations (**sacral agenesis abnormalities**) =>
 - Caudal regression synd = variable agenesis with underdeveloped limb and genitourinary & GI malformation
 - Currarino triad = partial agenesis with ARM (anal stenosis) and presacral mass (teratoma/ant MMC)



Pathophysiology

- **Stretching** of the cord, but conus is in a normal position. (**True TCS**)
 - Includes **inelastic filum terminale** and **caudal intraspinal lipoma** (lipoma attached inf. to conus)
- May have **additional focal compression (Relative TCS)**
 - Symptom might not be due to tethering but due to primary neurulation defect (MMC) or lipoma
- Repetitive strain leads to **ischemia** of the cord (Reversibility depends on traction force and duration)
- Extensive, **irreversible deficits** (?myelodysplasia- don't benefit from dethethering)



Presentation

- cutaneous manifestations of occult dysraphism (40%) – stigmata along dorsal midline
- Usually asymptomatic and become **evident during growth**
 - Urological (68 to 100%) – incontinence, reemergence of enuresis
 - Neurologic - gait weakness
 - Orthopedic - club foot
 - Pain – perineal area (more in adults)



Imaging

- **Ultrasound** can be used but needs MRI for confirmation.
 - Pulsation of cord is useful to evaluate recurrent tethering
- **MRI**
 - abnormal caudal position of the conus
 - Thickened filum terminale (wider than 2 mm) or fatty (bright attenuation on T1-weighted sequence)
 - posteriorly located filum terminate (b/c covering the shortest distance)
 - syrinx within the distal cord
 - additional anomalies along the spine. (Occult Dysraphism, Dermoid cyst/fistula, Split cord malformation)



Timing of surgery

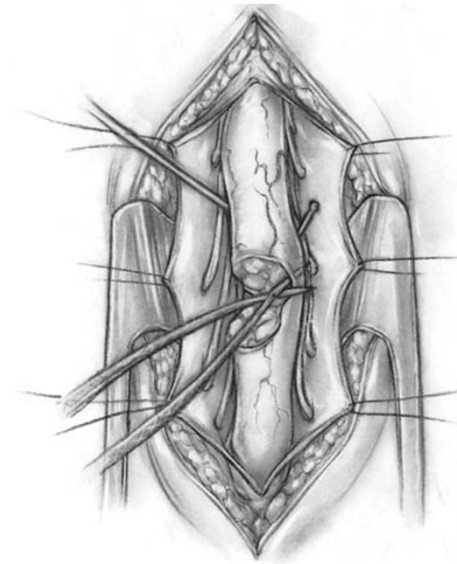
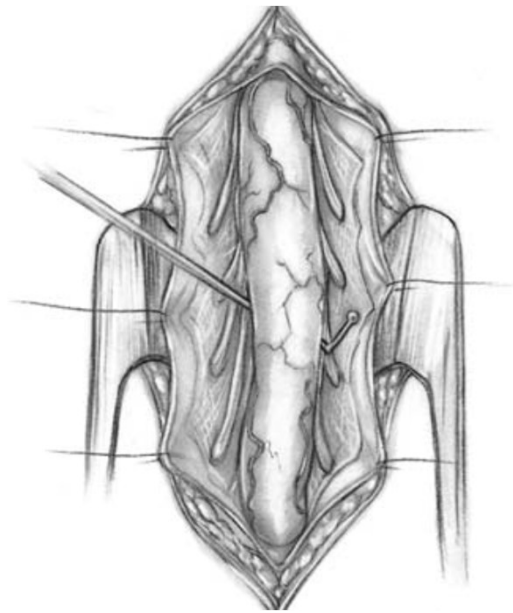
- Surgery should be done before irreversible ischemic change occurs
- **tight filum** = Indication of treatment of is **controversial**.
 - Some suggested neurological deterioration > 50% without surgery
 - Some patients managed conservatively also had improvement
- **Intraspinal lipomas** = also controversial
 - Most suggest **prophylactic total resection** for asymptomatic in young child (< 2 years)



Goal of surgery

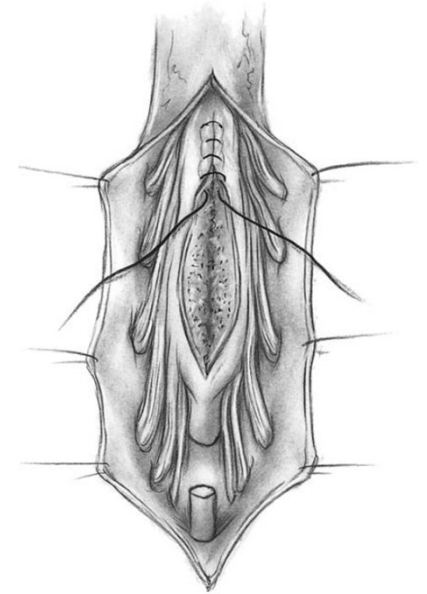
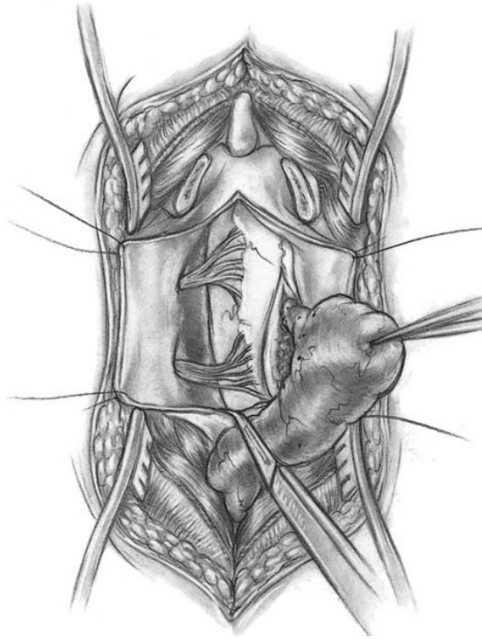
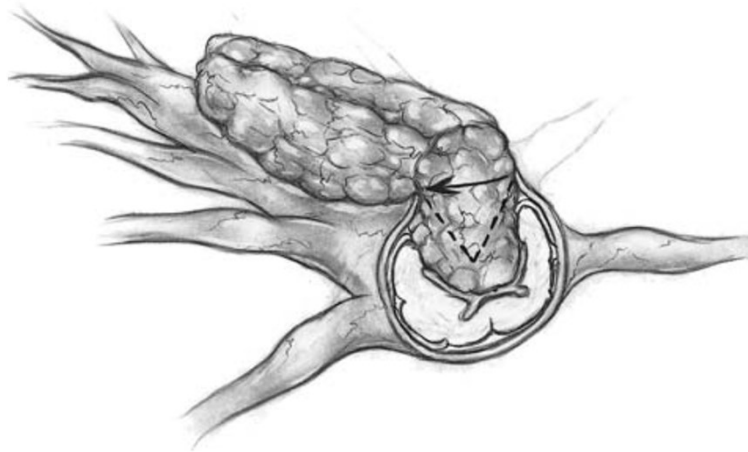
- Free cord from tethering structures
- Prevent re-tethering (create spacious dural sac)
- Preserve neurological tissue





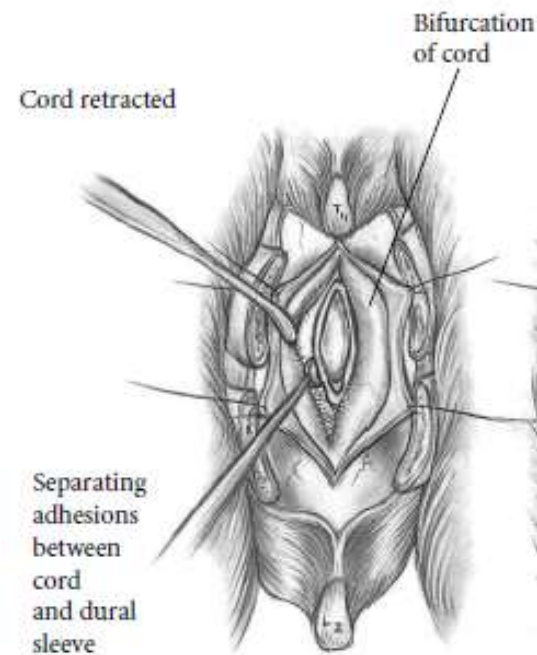
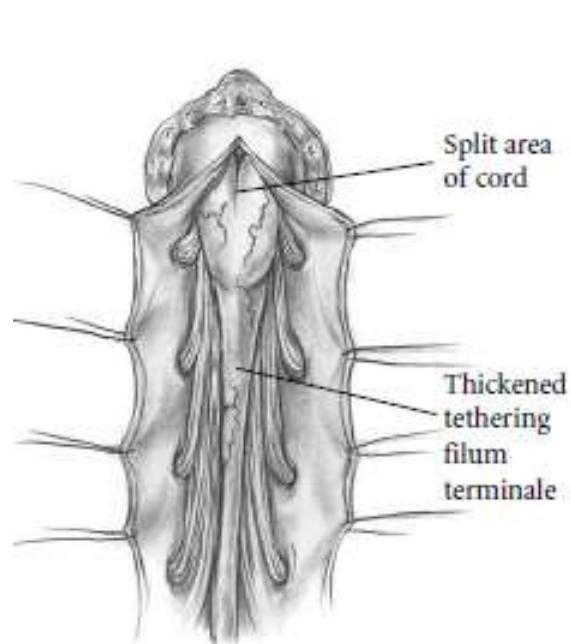
- **Surgical dethethering (tight filum)**: Exposure of tight filum and isolate from the caudal nerve roots with hook. Section distal filum after coagulation.





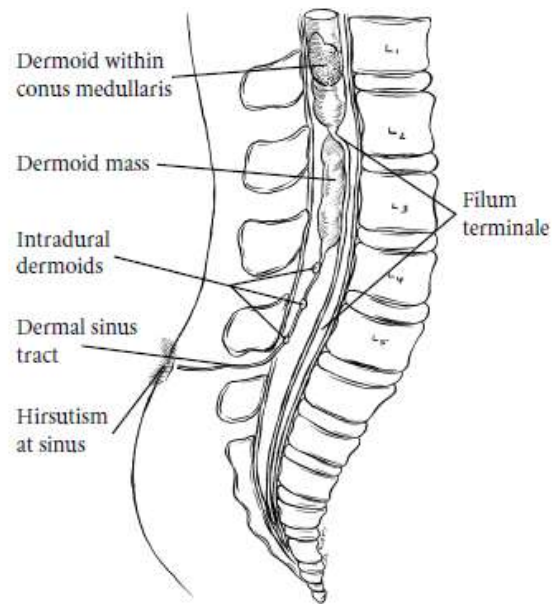
- **Surgical dethethering (intraspinial lipoma):** Superficial mass must be removed to allow visualization of the intradural structures After removal, dura opening and disrupt lipoma adhesion to neural tissue. Cut thick filum and inverted pial sutures to reapproximate dorsal tube.





- **Surgical dethethering (split cord malformation type I)**: exploration of the spinal canal may be complicated by the abnormal shape of vertebrae. Open the dura and detheter the cords by dissecting non functional medial nerve roots. Subsequent one dura sac should be created for both hemi cords





- **Surgical dethethering (dermoid fistula/cyst):** Radical dissection of intradermal and intradural extension. Total resection might require extensive multilevel intradural exposure of the fistula, and laminotomy with repositioning of the laminae.



Outcome

- **Surgical morbidity is low** (Same complications as MMC repair)
 - Intraoperative neuromonitoring decreases neurologic morbidity in spina bifida surgery
- **Best results in True TCS with early untethering** (only mild symptoms present)
- **Recurrent tethering is very low.** Occurs in
 - After closure of MMC
 - After partial resection of intraspinal lipomas



5. Encephalocele



Classification

- Cranial neural tube defect (10-20% of NTD)
- **Etiologic**
 - **Primary** - rare neurodevelopmental lesions present at birth
 - **Secondary** - acquired due to traumatic, postsurgical, inflammatory or neoplastic
- **Location**
 - **anterior** = sincipital (between frontal or facial bones), basal (inferiorly through the skull base),
 - * more common in asia
 - **Posterior** = Occipital (80-90%), temporal, parietal



Pathophysiology

- Development occur after primary neurulation, during **mesodermal differentiation**
- typically contain well developed neural and mesenchymal tissues encompassed by skin
- protrusion of cerebral tissue, leptomeninges, and CSF through calvarial and dural defects
- May occur with other anomalies as part of a syndrome



Presentation

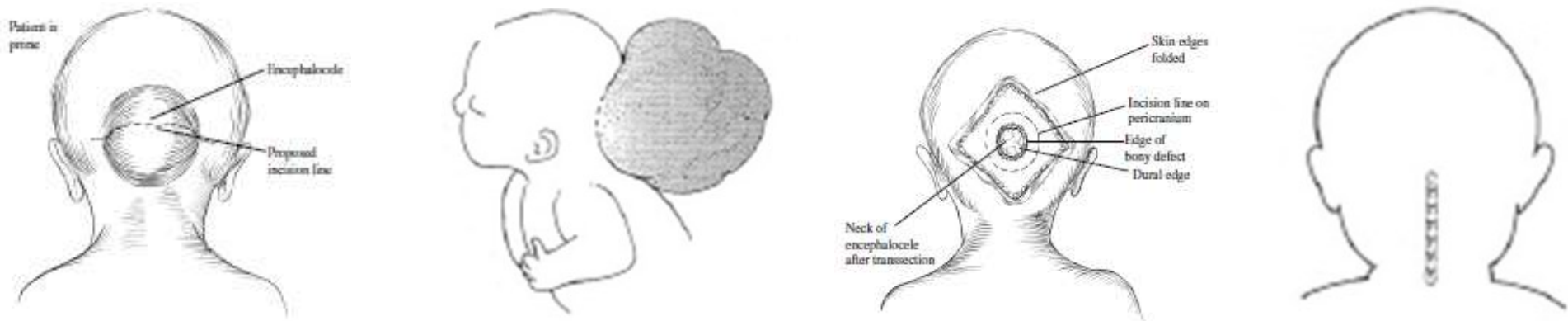
- Most are **obvious at birth** with microcephaly
- **Anterior lesions** are difficult to diagnose
 - **Pulsatile mass** that inc in size during crying is encephalocele (d/t from nasal polp...)
 - delayed diagnosis may become evident with **CSF leak & meningitis**
 - may also cause **airway obstruction**



Management

- Imaging - Craniospinal MRI should be done for all
 - Determine severity and extent
 - Look for ass. Malformations
- Conservative
 - for microcephaly and large amount of brain in encephalocele (death is inevitable)
- Surgery
 - For most patients
 - Aim is to prevent infection and preserve function





- Large lesions may be aspirated to facilitate handling and dissection. Transverse or ellipse Incision around base of encephalocele. excise extracranial non functioning brain tissue. Water tight dural closure. dural graft may be required (pericranium is ideal). Restoration of cranial contour with good skin coverage.



Outcome

- Complications
 - Nurse opposite to site to minimize pressure on wound and Observe for ICP (**HCP**)
 - **CSF leak** are managed with additional skin suture and lumbar drain (if HCP r/o)
 - **Meningitis** is common with ant. Encephalocele repair (proximity to nose)
- Prognosis depends on
 - Anatomical site
 - Volume of neural content
 - Coexisting cerebral and extracerebral malformation



References

1. Pediatric Surgery General Principles and Newborn Surgery, 2020
2. Rickham's Neonatal Surgery, 2018
3. Handbook of pediatric Neurosurgery, 2018

