2024

Level 4 - Semester 7

Surgery

Congenital Anomalies

Dr. Ahmed Hassan

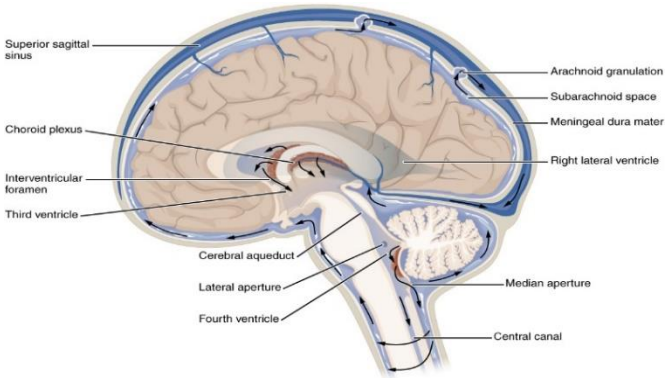
HYDROCEPHALUS

Definition

Dilatation of the ventricular system due to increase in the amount of CSF.

CSF circulation

Lateral ventricles → foramen of Monro → third ventricle → aqueduct of Sylvius → fourth ventricle → foramina of Magendie and Luschka → subarachnoid space over brain and spinal cord → reabsorption into venous sinus blood via arachnoid granulations.



Etiology

I Obstructive:

Mechanism	**Abnormal ↑ of CSF volume within cerebral ventricles** due to Impairment of circulation from one ventricle to another.
Causes	1) **Aqueductal stenosis**: **Cong. XR, acquired** as neonatal meningitis or subarachnoid hemorrhage. 2) **Dandy-Walker syndrome**: atresia of foramina of Luschka & Magendi → ballooning of 4th ventricle 3) **Arnold Chiari syndrome**: Elongation of 4th ventricle → displacement of inferior cerebellar vermis, pons, and medulla into cervical canal. 4) **Space occupying lesion**: as aneurysm of vein of Galen, brain tumor, abscess, cyst.

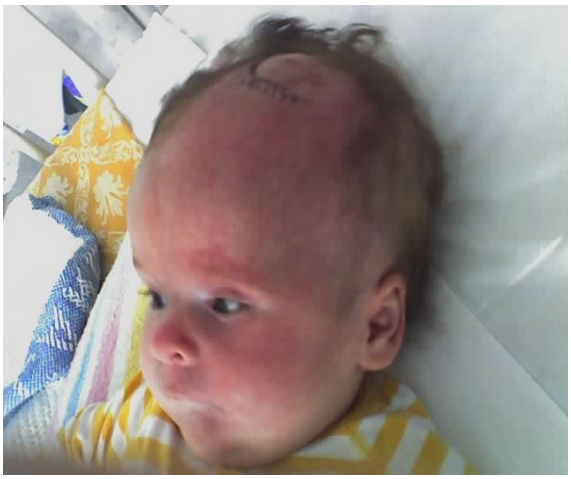
II Communicating:

Mechanism & Causes:

Excess formation	**Choroid plexus congestion** (meningitis), **Choroid plexus tumor** (papilloma).
Impaired absorption	**Hemorrhage, infection** **Leukemia infiltrates, hypervitaminosis D.**

Clinical picture

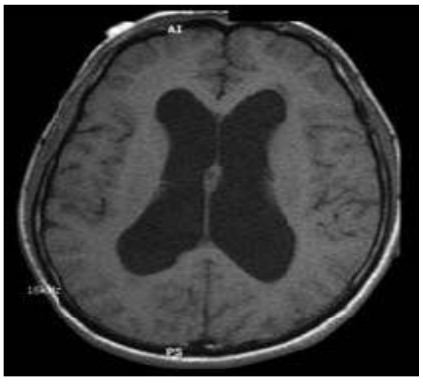
	Infancy & early childhood	During late childhood
Head	**Stretched Skin with Dilated Veins of forehead.** **Ant. Fontanelle** (Wide, Bulge, Tense) & Diastasis of skull sutures.	**Less significant head enlargement.**
Neurological Symptoms	Spasticity. Clonus. **Brisk tendon reflexes.** **+ve Babinski sign.** Papilledema. Optic atrophy. Convulsions. Squint.	Headache. Irritability. **Projectile vomiting.** Blurring of vision. **Abducent nerve palsy.** Lethargy. Poor appetite. **Pyramidal tract signs**



Investigations

1) CSF analysis.
2) Plain X-ray skull.
3) CT, MRI brain.
4) Transfontanellar US.

صورة الاشعة دي مكن تيجي في الاند ويسألك ده اي



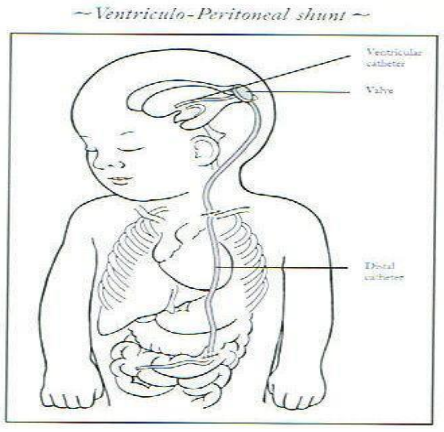
Differential diagnosis

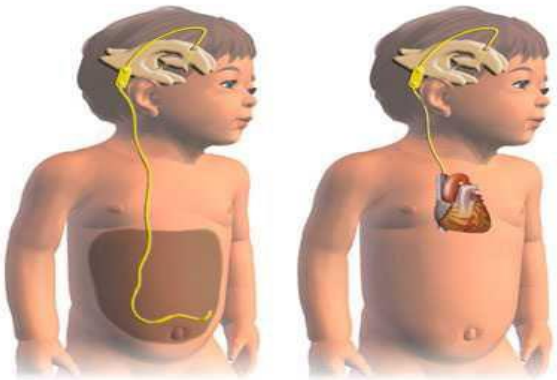
In the skull	1) Rickets. 3) Secondaries of neuroblastoma.	2) Osteogenesis imperfecta. 4) Achondroplasia.
In the meninges	1) Subdural hematoma. 2) Subdural effusion.	
In the brain	1) Hydranencephaly	2) Megalencephaly.

Treatment

W

Medical	1) Salt & H2O restriction. 2) Furosemide. 3) Acetazolamide. 4) TTT of the cause.
Surgical	1) Choroid plexectomy. 2) Removal of brain tumors. 3) Diathermy coagulation. 4) Shunt operation.
Endoscopic	--

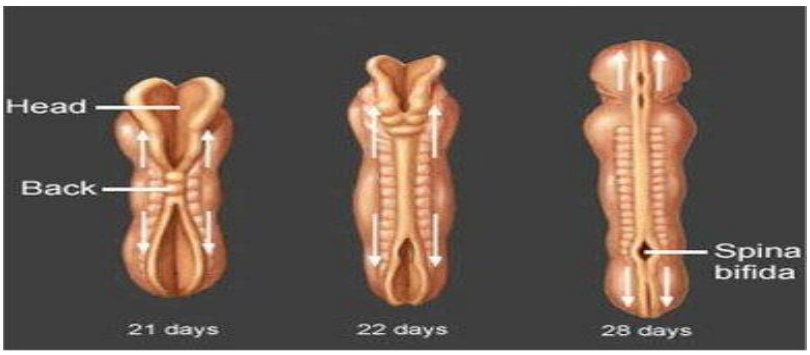


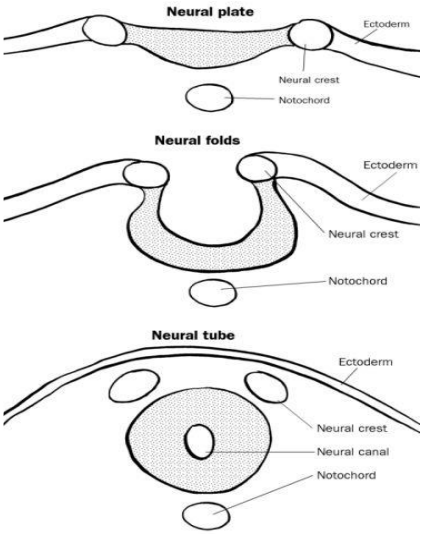


NEURAL TUBE DEFECT (NTD)

Normal embryological development

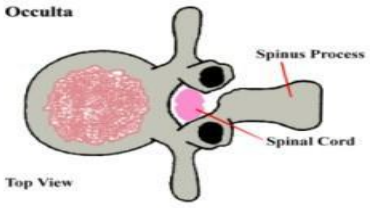
- Neural plate development → 16th day.
- Cranial closure → 24th day (upper spine).
- Caudal closure → 28th day (lower spine).

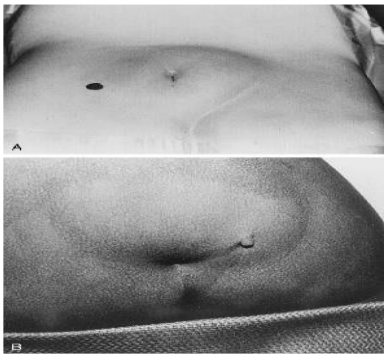




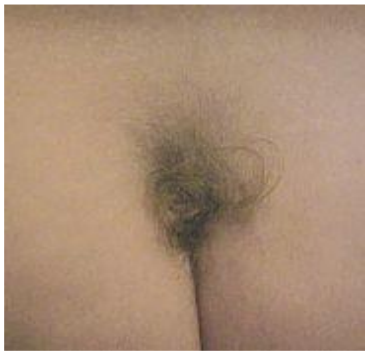
SPINA BIFIDA OCCULTA

Incidence	**Very mild & common form.** Very rarely causes disability.
Investigation	Can only detected by **x-ray** or investigating a back injury.
Clinical Picture / Association	1) **May be associated with tethered cord.** 2) **Usually associated with skin visible signs on the back:** A) Dimple. B) Small hair growth. C) Nevus flaminous (red spot) or port wine. D) Pad of subcutaneous fat.

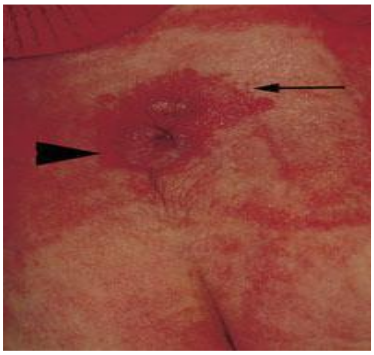




Dimple



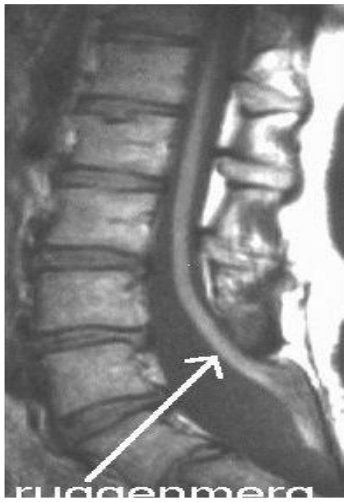
Tuft of hair



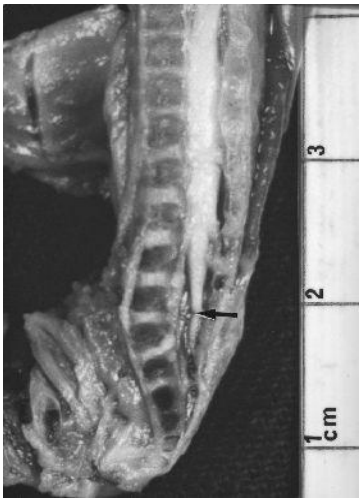
Dimple with Nevus port wine

Tethered cord

Pathology	**Normal cord ends at lower end of L1.** The spinal cord could be caught against the vertebra.
Complications	1) Motor weakness of lower limbs. 2) Sphincteric problems such as inefficient bladder control.



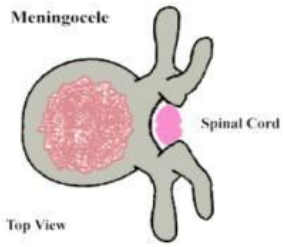
Tethered cord MRI



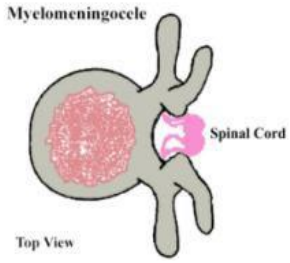
Autopsy of Infant with tethered cord

MENINGOCELE & MYELOMENINGOCELE

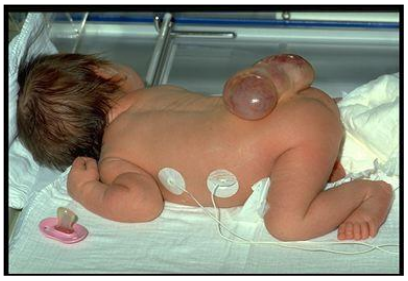
	Meningocele	Myelomeningocele
Incidence	**Least common form**	**Most serious and common**
Pathology	Sac contains meninges and cerebro-spinal fluid. CSF protects the brain and spinal cord.	The cyst not only contains meninges and CSF but also the nerves and spinal cord.
Complications	1) The nerves are not badly damaged and are able to function normally. 2) Limited disability is present.	1) The spinal cord is damaged or not properly developed **resulting in motor and sensory deficit.** 2) **The majority have bowel and bladder problems.**



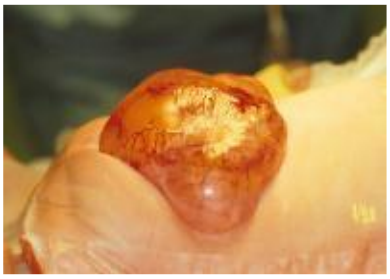
Meningocele



Myelomeningocele



Intact Myelomeningocele



Intact Myelomeningocele

Covered by thin membrane surrounded by hyper pigmentation)

The High Cost of NTDs

Financial Costs	Average estimated lifetime cost of $532,000 for each infant born with spina bifida (CDC 1999) adds an estimated 19 million dollars every year to Missouri resident lifetime costs associated with spina bifida.
Physical Costs	Possible paralysis (**the leading cause of childhood paralysis**) bowel and bladder control problems, learning disabilities, hydrocephalus, surgical procedures, latex allergies, ↑ health problems with age.
Emotional Costs	Miscarriage, stillbirth, infant mortality (death before 1st birthday), disability, feeling “different”.

Diagnosis (Prenatal detection of NTD)

Serum alpha-fetoprotein (AFP):

Normal	1) **Normal fetal glycoprotein** (MW= 70,000). 2) Present normally in **amniotic fluid and mother serum start 12 weeks increase steadily till 32 weeks.**
Diagnosis	1) **91% sensitivity** in spina bifida 2) **High maternal serum AFP > 2 multiples** of median for appropriate week of gestation **are diagnostic.**

Ultrasound:

Value	1) **Detect 90-95 % of cases of spina bifida**. 2) **100% cases of anencephaly**.
NB	In cases of elevated AFP, differentiate NTD from non-neurological causes of elevated AFP e.g., omphalocele.

Amniocentesis:

Indication	1) Pregnancies subsequent to NTD. 2) Elevated AFP with normal US.
Finding	Show **elevated AFP between 12-15 week earlier than serum AFP.**
Complication	Carries 6% risk of abortion and fetal loss.

Prophylaxis

Factors Associated with Increased Risk of NTD:

1) Family history of NTD
2) A previous pregnancy affected with NTD.
3) Maternal obesity
4) Maternal insulin-dependent diabetes.
5) **Anti-epileptic drugs** (Valproic Acid, Carbamazepine)
6) Lower socioeconomic/educational level, **dietary deficiency especially folic acid.**

The only most significant risk factor associated with NTDs is folic acid deficiency.

Folic Acid for Women:

1) As NTD occurs before diagnosis of pregnancy.
2) All women of childbearing age should receive **400 micro g (0.4 mg) folic acid daily**.
3) Women who have had a **previous child with NTD** should **receive 4000 micrograms (4 mg) of folic acid daily**.

Management

I Nursing Care:

1) Like any other neonate with congenital anomalies, efforts should be made towards careful examination and investigations to rule out other anomalies.
2) **Nursed in Trendelenburg position** → ↓ pressure & keep it away from cystic lesion.
3) **Care not to disturb intact membrane** (high incidence of infection and urgent surgery).
4) **Cover lesion with Gauze ring soaked with normal saline or Ringer solution** to prevent dryness.
5) **Avoid antiseptics** e.g., betadine → Neurotoxic affecting functioning roots in placode.
6) **Avoid mechanical trauma to placode**.
7) **No need for ultra-frequent dressing**.

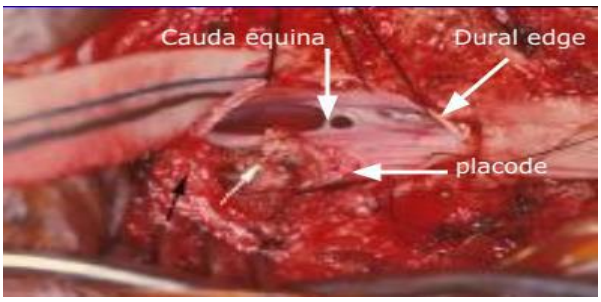
II General assessment:

Assess lesion	**Whether it is ruptured or unruptured:** 1) Ruptured lesions start prophylactic antibiotics. 2) Urgent surgery.
Measure defect size & site	For proper planning for **closure.**
Evaluation by neonatologist	**Other anomalies** (average **2-2.5% additional anomalies**). **Condition opposes with surgery** e.g., lung immaturity.
Bladder	Start with regular urinary catheterization. Urological consultation.
Orthopedic consultation	**For sever kyphotic or scoliotic deformities** and hip, knee, and foot deformities.

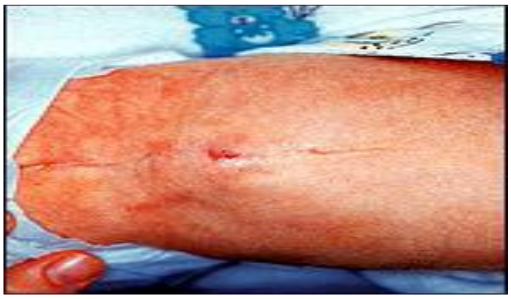
III Neurological preoperative Assessment:

1) Watch for spontaneous movement of lower limbs which are associated with better outcome.
2) Assess the lowest level of neurological function.
   A) Response to painful stimuli
   B) Differentiate between voluntary movement from reflex movement which is stereotyped and not persist after stimulus.
3) Evaluate other neurological associations:

Hydrocephalus	Anterior fontanel. Head circumference.
Chiari II	Check for inspiratory stridor and apneic episodes.



Intra-operative



Post-operative