432 Surgery Team

8 Common Congenital Neurosurgical Diseases

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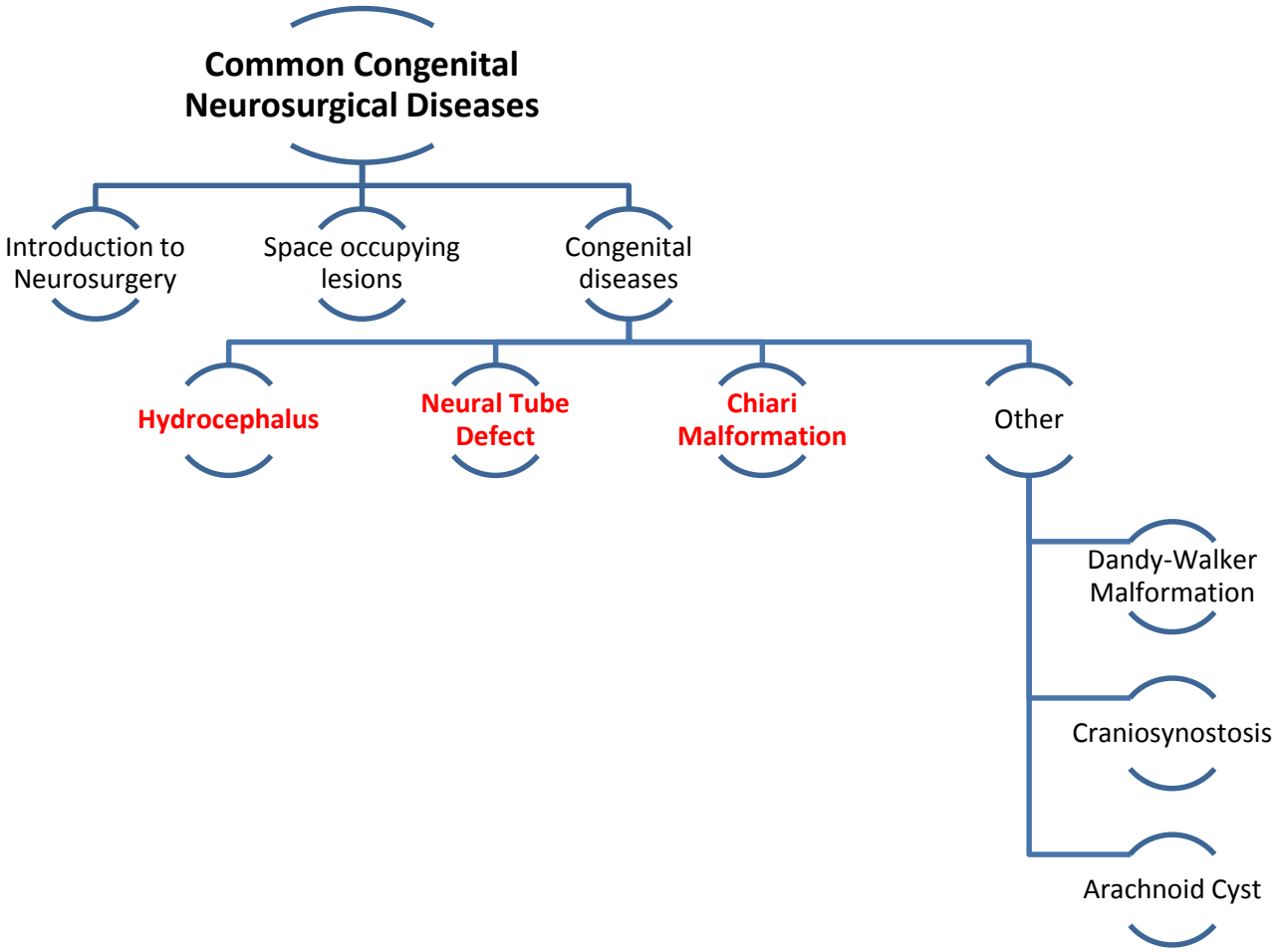
COLOR GUIDE: • Females' Notes • Males' Notes • Important • Additional

Objectives



Helpful Video

- Hydrocephalus

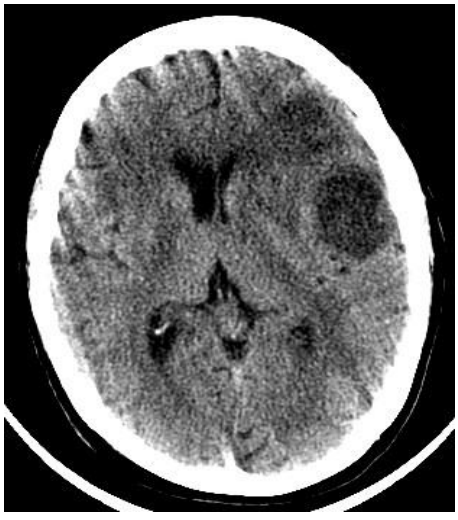
▪ Space occupying lesions:

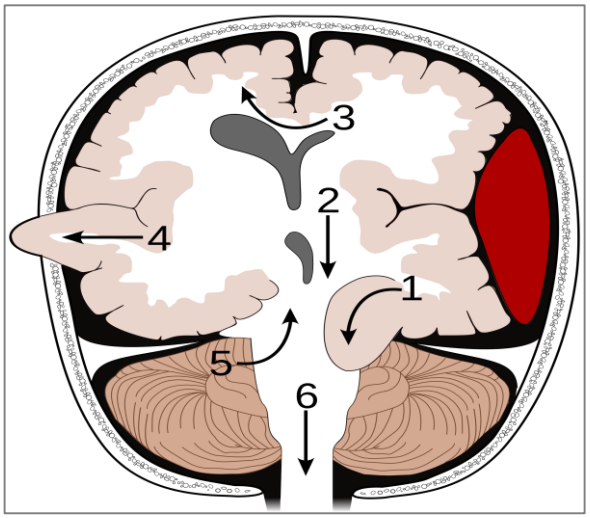
Any space occupying lesion will cause the following:

- ✓ **Local compression**
  With local symptoms.
- ✓ **Mass effect & Herniation**
  Mass will cause compression and lead to herniation of brain into small orifices the most common is **Uncal herniation** (see the pic).
  Usual presentation is contralateral weakness and ipsilateral pupil dilatation (affront in cranial nerve 2 and efferent in cranial nerve 3 which is usually compressed by the herniation).
- ✓ **High ICP**
  First 10 – 20 increasing in CSF pressure there will be compensation, but more than that → coma and death may happens.
  Most important signs and symptoms indicate high ICP are:
  1. Headache.
  2. Papilledema.
  3. Nausea.
  4. Vomiting.
  5. Decrease level of consciousness.

Differential Diagnosis of CNS space-occupying:

- Vascular
- Inflammatory/Infectious
- Neoplastic
- Degenerative/Deficiency/Drugs
- Idiopathic/Intoxication/Iatrogenic
- Congenital
- Autoimmune/Allergic/Anatomic
- Traumatic
- Endocrine/Environmental
- Metabolic





Types of brain herniation
1) Uncal transtentorial. 2) Central.
3) Cingulate. 4) Transcalvarial.
5) Upward. 6) Tonsillar.

Hydrocephalus

- Hydrocephalus is an increase in the CSF volume, associated with increased intra cranial pressure and ventricular size.
- Not the same as Ventriculomegaly

Note:

- It is an accumulation of CSF within the cerebral ventricle and is usually associated with altered ICP.
- The pressure is usually high, and sometimes normal, but rarely low (negative pressure hydrocephalus).
- When the ventricles are large but the patient is asymptomatic, that is not hydrocephalus; it's just hydrocephalus exvacuo "old name" or ventriculomegaly. So when you see large ventricles, it does not indicate hydrocephalus UNLESS there are symptoms of pressure changes of the brain.

Physiology:

- Total volume of CSF in the ventricles varies from 5-15 ml in neonates to 150 ml in adults. *(Depends on age and weight)*
- **Produced mainly by choroid plexus** and the extracellular fluid of the brain. (A tumor of the plexus can increase CSF production every day the plexus produces 500ml of CSF)
- Rate of production is 0.3-0.4 ml/minute
- Only very high ICP will reduce CSF production; usually at the point when brain perfusion is affected.

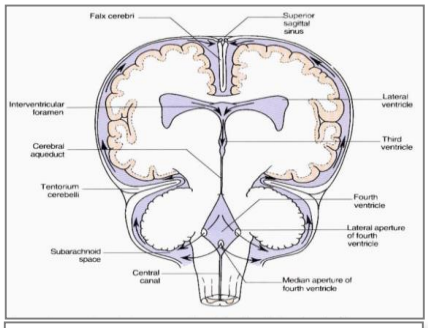


Figure 1

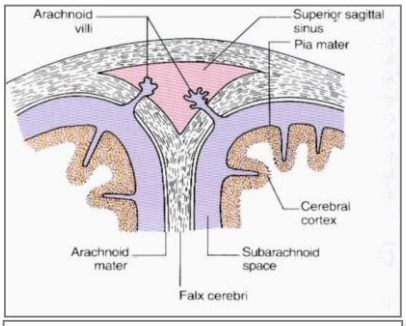
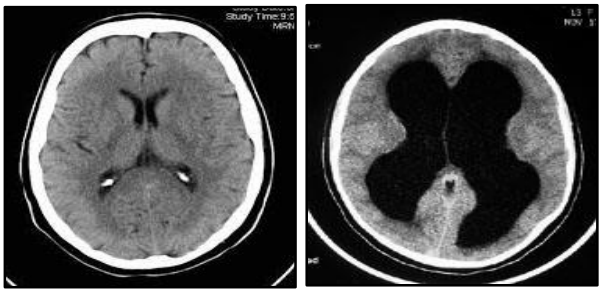


Figure 2

Notes:

Figure 1: *Coronal view of the brain*
1. Both lateral ventricles will drain CSF into 3rd ventricle through the foramen of Monro ***IMP***
2. From 3rd ventricle the CSF will pass to the 4th ventricle in the posterior fossa through the aqueduct of Sylvius ***IMP***
3. From 4th ventricle CSF circulates around the brain then passes through the three apertures (median foramen of Magendie & 2 lateral foramina of Luschka) to circulate around the brain, spinal cord and in central canal of spinal cord.
Figure 2: *This small cut section at the level of superior sagittal sinus (which is one of the dural venous sinuses of brain) shows the arachnoid granulations, which are an extension of the arachnoid. CSF drains into the arachnoid granulations (villi), then through the core of the villi into the venous circulation.*
So CSF circulates within ventricles then around the brain then it is reabsorbed in the cerebral ventricles.
It is a process of active formation and it does no stop it. It may be affected by ↑ICP BUT it does not stop it.
If there is any problem affecting absorption or the pathway of CSF→ accumulation of CSF and ↑ICP (which decreases CSF production).



Classification of Hydrocephalus:

Based on	Types	Remarks
Pathophysiology of hydrocephalus	**Overproduction of CSF.**	Choroid plexus papilloma.
	Obstruction of CSF flow.	Tumors: especially in or near the ventricles. Congenital anomalies.
	Under absorption of CSF.	Post meningitis. Post SAH (Subarachnoid hemorrhage).
Site of obstruction	**Communicating**	**All** ventricles are dilated. **Overproduction** or under absorption of CSF. **No obstruction** in the pathway of CSF within the ventricles (the ventricles can communicate with each other)
	Non-Communicating	**Partial** dilatation **Blockage** of the flow of CSF. ✓ **Congenital**, since birth. ✓ **Acquired**, develops after birth as a result of injury, tumors or meningitis. ✓ **Obstruction** within ventricles or the pathway of CSF (obstruction to the CSF flow at the foramen of Monro, the third ventricle, the aqueduct of Sylvius, the fourth ventricle, or the foramina of Magendie or Luschka.)
Etiology Not important to memories the etiology the Q's will ask: which one is a cause of hydrocephalus?	**Congenital** (primary)	• **Aqueductal anomalies** most common (see page 9) • **Dandy Walker malformation** (see page 9) • **Chiari II malformation** • Myleomeningocele • Intrauterine viral infection (CMV, mumps, rubella, varicella) • Toxoplasmosis • Congenital tumors • **Vein of Galen aneurysms** (see page 9) • Chromosomal anomalies (trisomy 13 and 18) • Congenital or primary hydrocephalus

	Acquired (Secondary, Most of the cases)	• Germinal plate hemorrhage "immature blood vessel walls": → in premature babies <1500 gm (30%-40%) (Increase in blood pressure may cause bleeding which cause obstruction → hydrocephalus developed as non-communicable, after that when the blood is absorbed by the meninges inflammation occurs and hydrocephalus become communicable).
		• Post-meningitis (non-communicable)
		  • 1% of survivors of bacterial meningitis
		  • More in neonates
		  • Especially Gram -ve organisms (i.e. E. coli).
		  • Rare but important, postnatal cysticercosis.
		• **Tumors** Most common cause
		• SAH (due to trauma)
		• Severe TBI (Traumatic Brain Injury)

Clinical features:

Infants & young children:

1. Increasing head circumference. (Scalp bones are still soft. So the head circumference increases abnormally – not according to curve of growth)
2. **Irritability, lethargy, poor feeding, and vomiting.** Mother complain.
3. Bulging anterior fontanelle. Wide, full and tense.
4. Widened cranial sutures.
5. McEwen's (cracked-pot) sign with cranial percussion **(!)**.
6. Scalp vein dilation (collateral venous drainage).
7. Sunset sign (downward deviation of the eyes) **(!)**
8. Epidsodic 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)*
   (!) The doctor said that these two signs are specific for hydrocephalus.

Notes:

Normal pressure hydrocephalus (NPH)
-Usually in elderly
-Presents with dementia-like symptoms
-Patient's family complain of patient's memory loss
-It is treatable
-Investigate with CT and MRI → you'll see ventricular dilatation more than cortical atrophy

Juvenile & adult:

1. Headaches
2. Nausea / Vomiting
3. Decreased level of consciousness
4. Focal neurological deficit (rare)
5. Papilledema

Investigation

CT (method of choice) or MRI: (CT is usually used because it is faster, show the type of hydrocephalus and can rollout hemorrhage). Classical Radiological Sign: **Separation of sutures, Pressure of the gyri on the bone (!).**

- The pattern of ventricular enlargement can help delineate the cause:
  - Lateral & 3rd ventricle dilatation
    → Normal 4th ventricle: suggests aqueduct stenosis
    → Deviated or absent 4th ventricle: suggests posterior fossa tumor
  - Generalized dilatation: suggests a communicating hydrocephalus.

Treatment

- **Surgical treatment:** Choroid plexus coagulation: We use endoscope, introduce telescope in the ventricle, and it helps in reduction of CSF formation
- **Intracranial Shunts:**
  ✓ Endoscopic Third Ventriculostomy **(First line treatment)**
  - The endoscope is passed through a burr hole to the third ventricle where the floor 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.

✓ Ventriculocisternostomy:

- Shunt between lateral ventricle and the cisterna magna.
- It has high morbidity and mortality. (Not done any more)

- **Extracranial Shunt:**
  - From the ventricular system, usually the lateral into another body cavity; the peritoneal cavity (VP Shunt), right atrium (VA Shunt) and occasionally pleural cavity.
  - Aim is to normalize the intracranial pressure
  - Specially designed shunt valve with the appropriate rate of flow and pressure.
  - Regulate the CSF flow in a unidirectional way.
  - Shunts are made of silicon which is well tolerated by the body. It causes minimal or no tissue reaction or intravascular thrombosis
  - It is done if you have communicating hydrocephalus or if you don't have the facility or capability to do endoscopic third ventriculostomy. (it is used for both communicable and non- communicable)
  - Complications of VP-Shunt:
    - Immediate operative complications
    - Shunt malfunction
    - Shunt infection

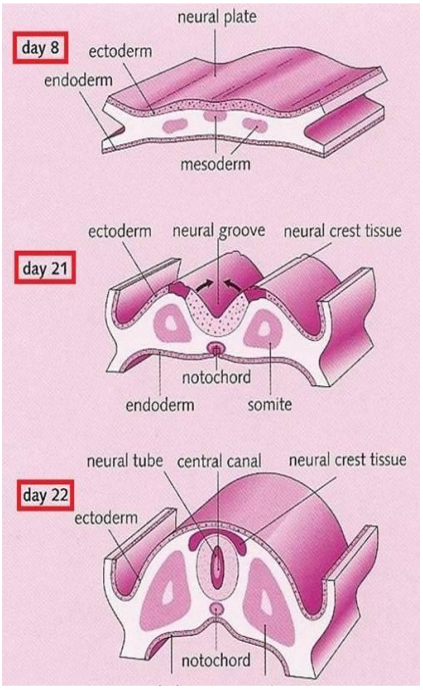
▪ Neural Tube Defect (NTD)

Spinal Dysraphism

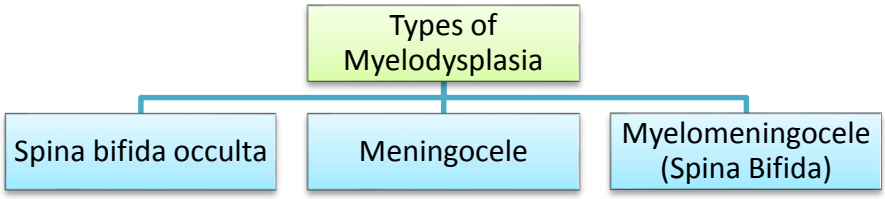
- Failure of closure of posterior neural arch
- Two major types: Open or Closed

Incidence

- 80% in lumbosacral region (Occasionally in the head)
- 2/1000 birth
- Risk increase to 5% if a sibling is affected
- Teratogens
- How to prevent?
  By giving folic acid supplement during pregnancy.

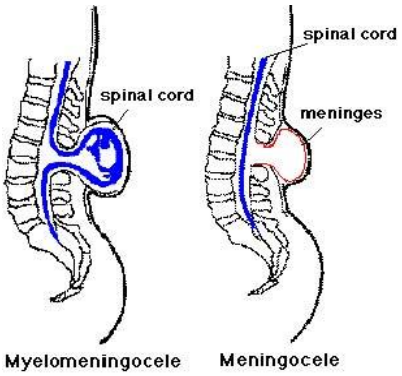


Neural fold fuse in 3ed week to 4th.
If it failure occur before 3ed week it will be open and sever. If it is late, it will be close and less sever.



Spina bifida occulta (occulta = not seen, usually benign)

- 5-10% of population. (Common)
- Not clinically significant. Found incidentally when the patient does x-ray.
- Sometime you find tuft of hair, dimple sinus or port wine stain
- high incidence of underlying defect
- no treatment required



Meningocele

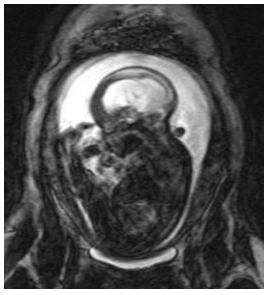
- Cystic CSF-filled cavity lined by meninges.
- No neural tissue.
- Communicates with spinal canal.
- Look for other congenital Anomalies.
- Seldom any neurological deficit.
- Dx: U/S or MRI.
- Tx: Excision; **urgent** in case of CSF leak (rupture).

Notes:

From Examination
-Locally: Size, content
-Assess skin quality (for surgery)
-If there's CSF leak :
**it is a surgical emergency*
**Risk of Infection*
-Translumination test to know the content
-Neurological examination
-Examine the sphincter to see if there's any problem with the anus (incontinence)
-Check for other associated anomalies (E.g.: Kyphosis), size of the head.
-Get the family collaborations from other specialties depending on the child's problem

Meningomyelocele

- Spinal cord and roots protrude through the bony defect.
- Lie within cystic cavity.
- Observe limb movements (degree & level of neurological damage)*.
- Note dilated bladder & patulous annual sphincter.
- DX: U/S or MRI.
- Gross hydrocephalus, multiple serious congenital anomalies.
- Many adopt through conservative treatment but if ruptured it's a surgical emergency → immediate closer and replacement of neural tissue.



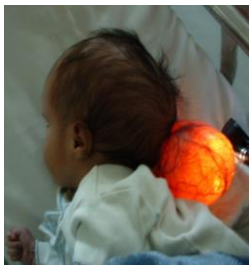
* Because the spinal cord is outside, the patient will have motor, sensory and autonomic deficit in the lower limb so we should observe it to determine the degree and level of damage (Raslan: they are born with paraplegia).

Antenatal diagnosis

- Maternal U/S,
- MRI
- Serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase
- Contrast enhancing amniography
- possibility of therapeutic abortion
- Family history for siblings with NTD

▪ Other congenital anomalies

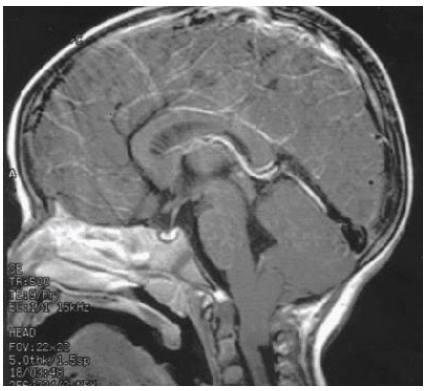
Encephalocele



- An encephalocele is a protrusion of the brain and/or meninges through a defect in the skull that is "closed" or covered with skin.
- Usually occipital
- May contain occipital lobe, or cerebellum
- Often associated with hydrocephalus
- Immediate treatment if ruptured
- Outcome depends upon contents

Chiari Malformation

When part of the cerebellum is located below the foramen magnum by > 3mm, it is called a Chiari malformation.



- **Type I**
  - Extension of the cerebellar tonsils into the foramen magnum, without involving the brain stem.
  - Manly in adult with neck pain and cough
  - Associated with: Syringomyelia and Hydrocephalus
- **Type II**
  - Extension of both cerebellar and brain stem tissue into the foramen magnum.
  - Associated with: **Myelomeningocele**, and Hydrocephalus.
- **Type III**
- **Type IV**

Notes (Raslan):

AQUEDUCTAL ANOMALIES

- *Aqueduct is the passage of CSF between the 3rd & 4th ventricles-passes in the midbrain*
- *It is still known as aqueductal stenosis, however by definition, the correct term is "aqueductal atresia" because no aqueduct is found in most of cases - but in some cases (who develop hydrocephalus late in their childhood) it allows a small amount of CSF to pass through so it is usually called stenosis*

DANDY WALKER MALFORMATION (LARGE DANDY WALKER CYST)

- *By definition it is congenital hypoplasia or even aplasia of cerebellum associated with formation of a large CSF cavity within the posterior fossa due to the obstruction of CSF flow by a large cyst (which doesn't allow CSF to pass from the 4th ventricle and circulate around the brain)*
- *There are different types of Dandy Walker cyst according to the volume of cerebellum that's involved*
- *Most of cases of Dandy Walker malformation are associated with hydrocephalus*
- *In short: large CSF cyst on posterior fossa due to agenesis of the cerebellum that communicates with the 4th ventricle, and causes hydrocephalus.*

VEIN OF GALEN ANEURYSMS

- *A large vascular malformation where there is a direct communication between the arterial system and venous system (shunting), leading to dilatation of the Vein of Galen (one of the deep venous structures in the brain) and to obstructive hydrocephalus.*
- *What is the clinical manifestation for such cases in neonates?*

o 1st and most important is heart failure (the size of the arteriovenous shunt that can steal 80% or more of the cardiac output), then symptoms of hydrocephalus (developmental delay, seizures, headaches)

- *Rx: treat the cause, which is the aneurysm (stop passage of blood from artery to vein by embolization) - and no need for shunt to treat the hydrocephalus*

o Also shunts can burst the aneurysms and cause fatal hemorrhage

ARACHNOID CYSTS

- *Developmental anomaly -> Intra-arachnoid cavity filled with CSF -> Benign*
- *Treatable -> Good prognosis*
- *Majority in the sylvian fissure*
- *Space occupying lesion symptoms -> Convulsions, raised ICP*
- *In supracellar space -> Can produce endocrine dysfunction*
- *Diagnosis*

o CT scan (Not the best) Can be confused with a tumor

o MRI is more precise

- *Treatment -> Shunting*

ANENCEPHALY *(NO BRAIN)*

- *Defective closure of the rostral neural tube*
- *Results in anencephaly or encephalocele*
- *Neonates with anencephaly have a rudimentary brainstem, or midbrain, no cortex or cranium.*
- *Rapidly fatal condition if born a live.*

DIASTEMAMYELIA

- *A bone or fibrous band divide spinal cord in two longitudinal sections* ***(!)***
- *Associated lipoma may present, which tethers cord to vertebra*
- *Signs and symptoms include weakness, numbness in feet, urinary incontinence, decreases or absent reflexes in feet*
- ***Dx: CT***
- *Rx: surgery to free cord*

SUMMARY

1. **Hydrocephalus** is increase in (CSF volume, ICP, ventricular size).
2. Caused by Overproduction of CSF, under absorption of CSF → (communicating -all the ventricles are dilated-) or obstruction of CSF flow → (non-communicating -partial dilation-).
3. Etiology of Hydrocephalus:
    - Congenital (Aqueductal anomalies - Dandy Walker malformation - Chiari II malformation)
    - Aquired (Tumors – SAH – post meningitis)
4. Clinical features:
    - In infants & young children (increase head size – poor feeding - vomiting, - McEwen's sign – Sunset sign)
    - In adults (headache – papilledema – nausea\vomiting – decrease consciousness)

5. Dx: CT sacn.
6. Treated with (surgery – intracranial shunt – extracranial shunt)

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7. **Neural Tube defect:**
    - **Spina bifida occulta**: patient have (tuft of hair – dimple sinus – port wine stain), no treatment is required.
    - **Meningocele**: cystic cavity lined by meninges and filled with CSF, Dx: MRI or U\S, treated with excision → (surgical emergency if ruptured and CSF leaked).
    - **Meningomyelocele**: spinal cord and roots protrude though the bony defect in a cyst, Dx: MRI or U\S, examine the lower limb to determine the level of damage and note if there is dilated bladder or patulous anal sphincter.

- NTD is diagnosed before delivery by (maternal U\S - Serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase).

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8. **Chiari Malformation:** When part of the cerebellum is located below the foramen magnum:
    - Type 1: extension of the cerebellar tonsils only into the foramen magnum.
    - Type 2: extension of the cerebellar tonsils and the brainstem into the foramen magnum.

Questions

1) Which statement is true:
   a. Spina bifida occulta is a neurosurgical emergency.
   b. Meningocele contains spinal cord.
   c. Spina dysraphism occurs most commonly in lumbosacral region.
   d. None of the above

2) The most common type of cerebral herniation is:
   a. Central
   b. Cingulate
   c. Transtentorial
   d. Tonsillar

3) The investigation of choice in increased ICP is:
   a. Skull x-ray
   b. MRI
   c. Lumbar puncture
   d. CT scan

4) Hydrocephalus is defined as:
   a. Accumulation of CSF with ICP
   b. Soft tissue contusion
   c. Intracranial hemorrhage
   d. Enlargement of the head

5) Obstructive hydrocephalus is best treated by:
   a. Surgery
   b. Drainage
   c. Craniotomy
   d. Endoscopic third ventriculostomy

6) In Spina Bifida, the following is correct except:
   a. It is a failure of closure of posterior neural arch
   b. Contains spinal cord in menengeocele
   c. There are two types, Open and Closed
   d. 80% in lumbosacral region

7) Which of the following can cause communicable hydrocephalus?

a. Tumor Cousin Obstruction
b. Post Meningitis
c. Germinal Plate Hemorrhage
d. Aqueductal Anomalies

8) **OSCE Q**: Child with low back skin discoloration, in examination of the back there was tuft of hair and skin pigmentation. What should be done after that?

a. Do investigation to rollout internal anomalies.
b. CNS examination
c. Argent Surgery
d. A and B

Answers:

1st Questions: C
2nd Questions: C
3rd Questions: D
4th Questions: A
5th Questions: D
6th Questions: B
7th Questions: C
8th Questions: D