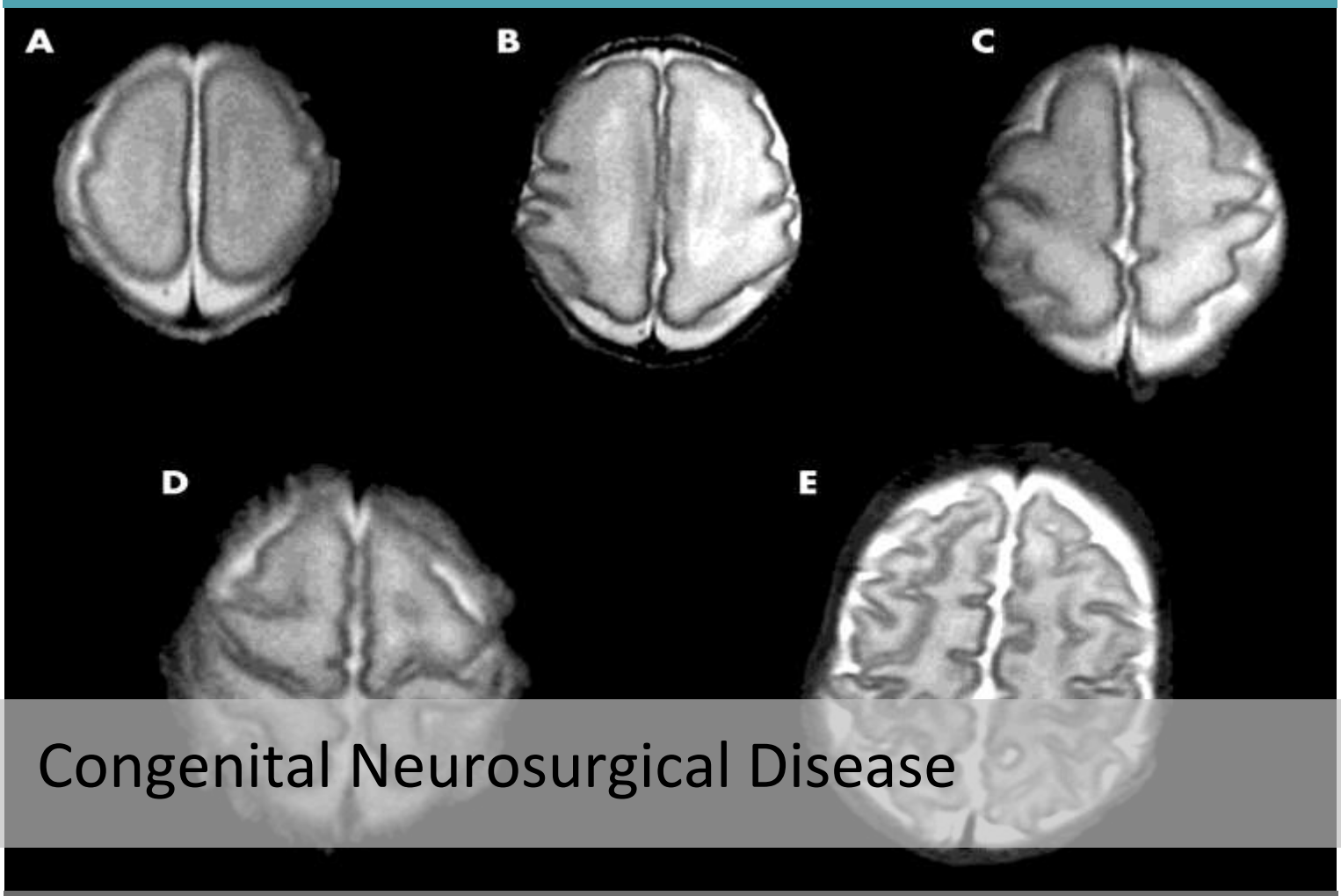

Congenital Neurosurgical Disease

429 Surgery Team

Source: Dr. Essam Elgamal's lecture

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Special thanks to: Abdullah Alaogayil

Team notes are in blue & in boxes

Questions: http://ask.fm/TeamNotes429

HYDROCEPHALUS

- Hydro: water, and cephalus: head
- Hydrocephalus is an increase in the CSF, associated with increased ICP IMP
- Ventriculomegaly secondary to cortical atrophy; aka hydrocephalus ex vacuo

- 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 ex vacuo "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.

- In short: hydrocephalus is the presence of increased CSF and increased ICP

CAUSES (PATHOPHYSIOLOGY)

- Overproduction of CSF
- Obstruction of CSF flow
- Under absorption of CSF into the blood stream (leads to accumulation of CSF)

- CSF is produced by the choroid plexus of the ventricles
  - A tumor of the plexus can increase CSF production
- Everyday the plexus produces 500ml of CSF

PHYSIOLOGY

- Total volume of CSF in the ventricles varies from 5-15 ml in neonates to 150 ml (cc) in adults. (Depends on age and weight)
- Produced by active secretion of choroid plexus and the extracellular fluid of the brain **IMP**
- Only very high ICP will reduce CSF production, usually at the point when brain perfusion is affected

- Rate of production is 0.3-0.4 ml/minute → 500 ml CSF/day – 150 ml in the CNS → 350 CSF absorbed everyday.
- It is a process of active formation and it does not 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)

Figure 1: Coronal view of the brain

1. Both lateral ventricles will drain CSF into 3rd ventricle through foramen of Monro **IMP**
2. From 3rd ventricle the CSF will pass to 4th ventricle in 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.

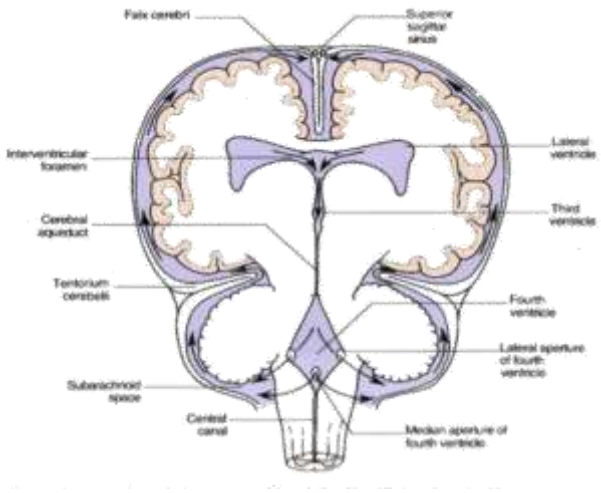


Figure 1

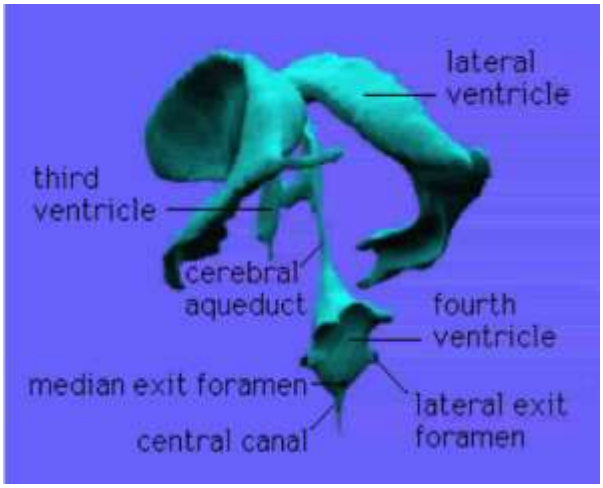


Figure 2

Figure 2 shows the communication between the ventricles

2 lateral ventricles inside cerebral hemisphere - they have a frontal horn, occipital horn & temporal horn

Both communicate through foramen of Monro> 3rd ventricle> cerebral aqueduct> 4th ventricle

Figure 3: *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.

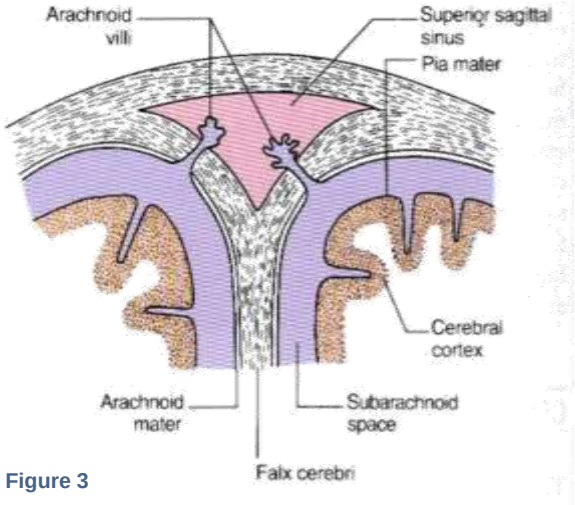


Figure 3

EPIDEMIOLOGY

The overall incidence of infantile hydrocephalus is 1/1000 live birth and 2/1000 in some countries like Africa and India

* It is not a common disease

TYPES

COMMUNICATING

- Overproduction or under absorption of CSF
- No obstruction in the pathway of CSF within the ventricles (the ventricles can communicate with each other)

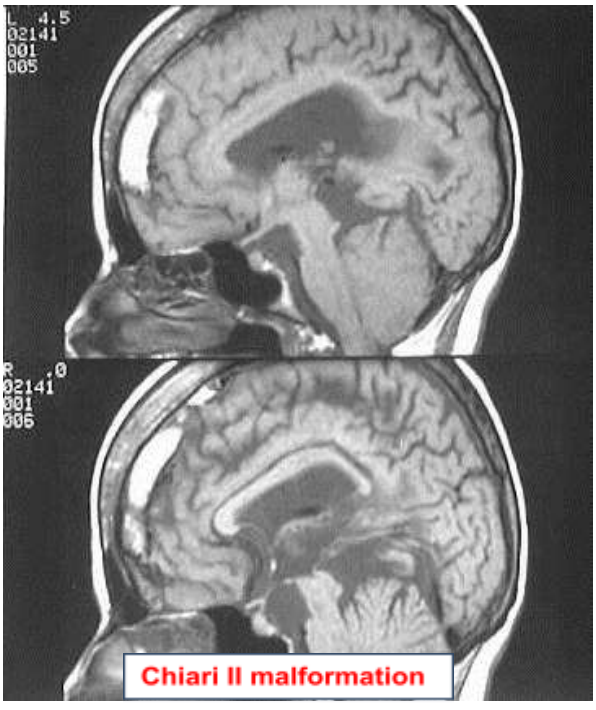
NON-COMMUNICATING

- Blockage of the flow of CSF
- 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.)
  - Congenital, since birth
  - Acquired, after birth due to injury (trauma, hemorrhage), tumors or meningitis

ETIOLOGY

CONGENITAL

- Aqueductal anomalies
  - Aqueductal stenosis **MOST COMMON CAUSE OF CONGENITAL HYDROCEPHALUS**
- Dandy Walker malformation
- Chiari II malformation *associated with meningomyelocele*
- Myleomeningocele
- Intrauterine viral infection (CMV, mumps, rubella, varicella)
- Toxoplasmosis
- Congenital tumors
- Vein of Galen aneurysms
- Chromosomal anomalies (trisomy 13 and 18)
- Congenital or primary hydrocephalus (idiopathic; no known reason)



AQUEDUCTAL ANOMALIES

Aqueduct is the passage of CSF between the 3rd & 4th ventricles-passes in the midbrain

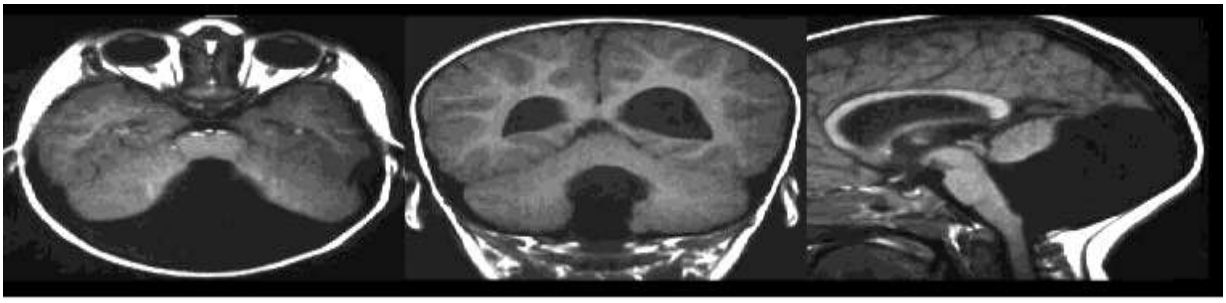
It is still know as aqueductal stenosis, however by definition, the correct term is "aqueductal atresia" because no aqueduct is found in most of cases - but in some cases (who develop hydrocephalus late in their childhood) it allows a small amount of CSF to pass through so it is usually called stenosis

DANDY WALKER MALFORMATION (LARGE DANDY WALKER CYST)

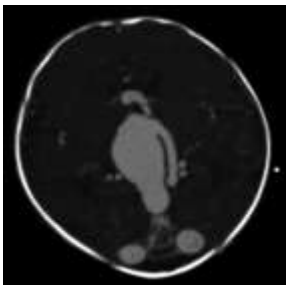
You can see the volume of the cerebellum is reduced.

- 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.

Dandy-Walker malformation is characterized by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlargement of the posterior fossa
Presentation: In-coordination, ataxia, nystagmus



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?
    - 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)

- R_x: 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
    - Also shunts can burst the aneurysms and cause fatal hemorrhage

ACQUIRED

- Germinal plate hemorrhage: especially in preterm infants "immature blood vessel walls"
    - Leads to intracranial hemorrhage in premature babies <1500 gm (30%-40%)
- 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.

CLINICAL FEATURES

INFANTS AND 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. (Leads to delayed development)
3. Bulging anterior fontanelle. (Wide, full and tense)
4. Widened cranial sutures
5. McEwen's cracked pot sign with cranial percussion IMP *(sounds like you're tapping on a cracked pot)*
6. Scalp vein dilation (collateral venous drainage).
7. Sunset sign (downward deviation and failure of accommodation of the eyes) IMP *not only infants but also in children and sometimes adults*
    - It 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"

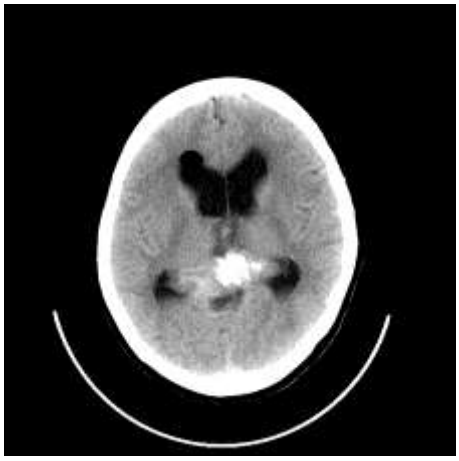
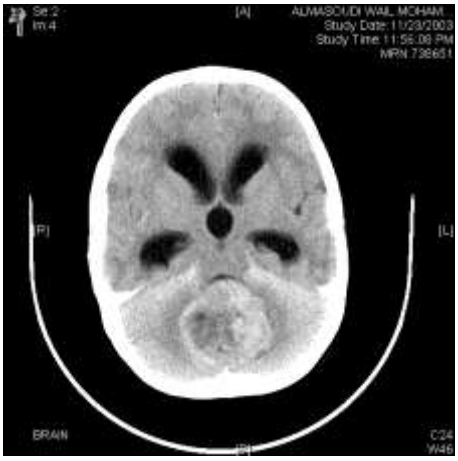
8. Episodic bradycardia and apnea (If hydrocephalus is left untreated - the increased intracranial pressure will press on the brain stem –where the respiratory centers are located- which will lead to this)

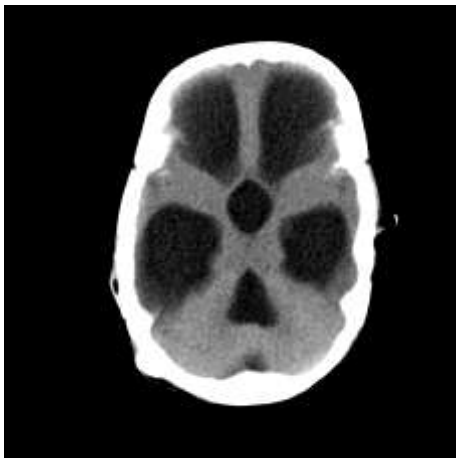
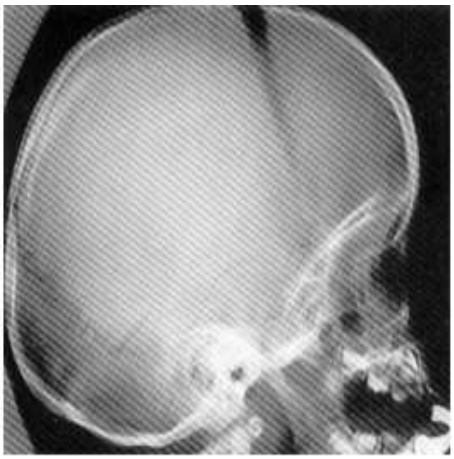
JUVENILE AND ADULTS

- Headaches. (Usually in the early morning. Why? Because during sleep, the patient will hypo ventilate, which will lead to ↑ CO_2 → ↑ vasodilation → blood stasis & ↑ intracranial pressure)
- 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)
- Seizures. (Acute manifestation)
- Decreased level of consciousness
- Focal neurological deficit (Depending on the cause of the hydrocephalus)
- Collection of CSF around previous shunt site

Night (sleep) → hypoventilation→ increased Co2 retention → vasodilatation → symptoms are worse
Vomiting (on waking up) → hyperventilation→ washing out of Co2 → vasoconstriction and less dilations → symptoms improve

INVESTIGATIONS

	
Lateral ventricles dilated but 3rd ventricle not very dilated > lesion in 3rd ventricle that prevents CSF from entering the 3rd ventricle at level of foramen of **Monro**	Posterior fossa tumor

	
Communicating hydrocephalus	X-ray is not a diagnostic procedure for hydrocephalus now You will see widening of the suture of skull secondary to increased intracranial pressure

- CT scan *easy, available and you can do it in minutes*
- The pattern of ventricular enlargement can help delineate the cause:
- 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'
- Generalized dilatation: suggests a communicating hydrocephalus.

TREATMENT

There's no medical treatment, 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

SURGERY

1. Endoscopic choroid plexectomy was tried but found unsuccessful. Not done now b/c all children die during.
2. Replaced now by Choroid plexus coagulation
   We use endoscope, introduce telescope in the ventricle, and it helps in reduction of CSF formation

INTRACRANIAL SHUNTS

In obstructive hydrocephalus where the subarachnoid spaces are still patent

a. Endoscopic third ventriculostomy: **1st line treatment** now IMP!
   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.

b. Ventriculocisternostomy “Torkildsen operation”:
Shunt between lateral ventricle and the cisterna magna.
It has high morbidity and mortality. (Not done anymore)

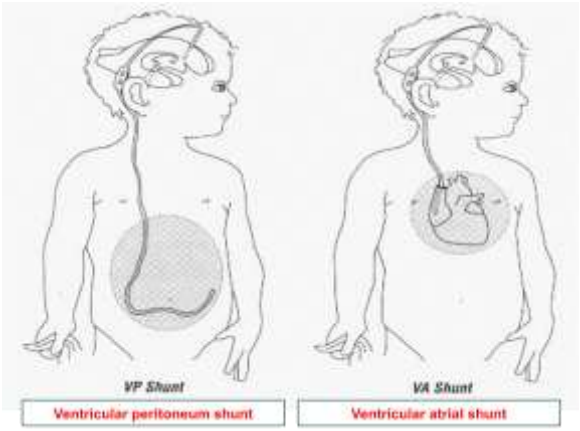
EXTRACRANIAL SHUNTS

Extra-cranial Shunt: if you have communicating hydrocephalus or if you don’t have facility or capability to do endoscopic third ventriculostomy

- *We insert a catheter into the ventricles and connect it to a valve that regulates the volume of CSF, and then distal catheter in distal cavity that can absorb the CSF, usually the peritoneal cavity, plural cavity and the blood stream in the right atrium*
- *Ventricular peritoneal shunt is standard for every patient who has hydrocephalus, especially communicating hydrocephalus*
- *Ventricular plural shunt is only for babies 3 years or older - b/c younger babies have a small chest cavity and then suffer of hydrothorax*
- *Ventricular venous or atrial shunt is suitable for all ages, but usually we do it for patients who can’t do ventricular plural or peritoneum shunt or endoscopic third ventriculostomy.*
- *Shunt stays forever*

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



COMPLICATIONS OF VP SHUNT

COMMONEST COMPLICATION IS OBSTRUCTION FOLLOWING BY INFECTION

1. Mechanical failure
    a. Under drainage or over drainage
    b. Blockage (Obstruction)
    c. Improper placement or
    d. Migration of the shunt system.

    25 to 40% in the first few months after surgery, later on 4 to 5 %

SHUNT BLOCKAGE

If you press valve and there is no fluid passing through shunt > distal obstruction
If you press shunt & valve collapse and doesn't refill again > proximal obstruction

50% of mechanical failure, highest in the immediate post-operative period

- Proximal occlusion in brain: By brain debris or choroid plexus
  - Ideally the catheter should lie away from the choroid plexus
- Shunt valve blockage: By brain debris or blood clots or due to failure of the valve system
- Distal obstruction in abdomen
  - Less frequently than the proximal obstruction
  - Accumulation of particles
  - Catheter encysted or isolated in one area of the peritoneal cavity
    Following infection or adhesion
- Catheter migration, disconnection, or fracture
  Sometimes catheter pass through stool or coming out through the anus

SHUNT INFECTION

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.
- **Clinical features:** *infection leads to increase CSF volume so patient suffer from hydrocephalus symptoms again*
  - Early, within 8-10 weeks.
  - Fever, malaise, headache & irritability, neck stiffness.
  - Peritonitis is less common.
  - Patients with Staph epidermis may look remarkably well with only intermittent fever or irritability.
  - Diagnosed by blood culture, routine blood examination and CSF examination.
- Treatment of shunt infection
  - Removal of shunt and the external ventricular drainage plus antibiotics.
    - Take the shunt out → put temporally external drainage → treat the infection by antibiotics till the CSF cleared of infection → then replace the external drainage by internal shunt

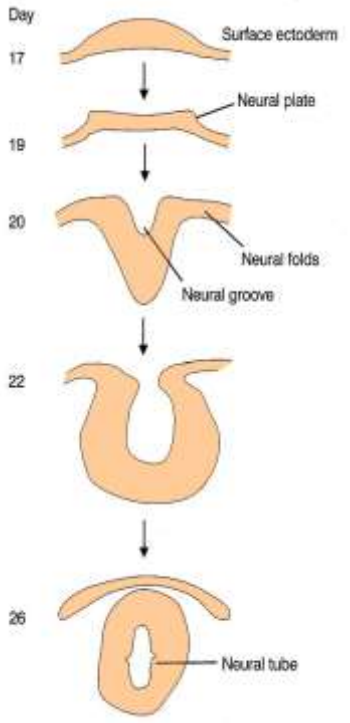
        - External drainage very useful b/c we can drain the CSF, take sample of CSF for culture and to inject antibiotics intra-ventricular to treat infection or speed up the clearance of infection
    - Treatment with antibiotics alone. For patient who have very low grade of infection & Clear CSF
        - Antibiotic prophylaxis is controversial however it was found that intra-operative antibiotics or antibiotics for the first 24 hours give the best results

LESS COMMON COMPLICATIONS

- Subdural collection from over drainage
- Slit ventricle syndrome, (over drainage) > cause collapse of ventricles
- Disconnection or fracture of shunt tubing
- Seizures

DEVELOPMENT OF THE NERVOUS SYSTEM

DEVELOPMENTAL SEQUENCE



- Neural plate invaginates as neural folds push up
- Neural folds eventually form neural groove
- Cells of neural fold eventually meet
- Form the neural tube
- Developmental Sequence
- Neural tube runs anterior – posterior along embryo.
- Surrounding ectoderm eventually encloses neural tube.
- When neural tube closes off brain and spinal cord are formed and this process finishes in day 28 from when the pregnancy begins.
- Spinal cord covered posteriorly by spinal presses and Laminae
- Developmental Sequence
- Neural tube runs anterior – posterior along embryo
- Surrounding ectoderm eventually encloses neural tube
- When neural tube closes off brain and spinal cord are formed and this process is completed on day 28 of pregnancy begin > so if the spina bifida formed at 28 day it's formed!
- Spinal cord covered posteriorly by spinal process and laminae

REMEMBER

- Spinal cord covered posteriorly by spinal process and laminae. Any failure in the fusion of posterior components of spinal cord will lead to spinal bifida (it is also called open neural defect, or neural tube defect)
- If the S.C out said the spinal canal it will not function and it will affect the baby lower limb

NEURAL TUBE DEFECT (SPINAL DYSRAPHISM OR BIFIDA)

- Failure of closure of posterior neural arch

Types: Open or Closed

Open	Closed
• occurs when the brain and/or spinal cord are exposed at birth through a defect in the skull or vertebrae .	• occurs when the spinal defect is covered by skin.

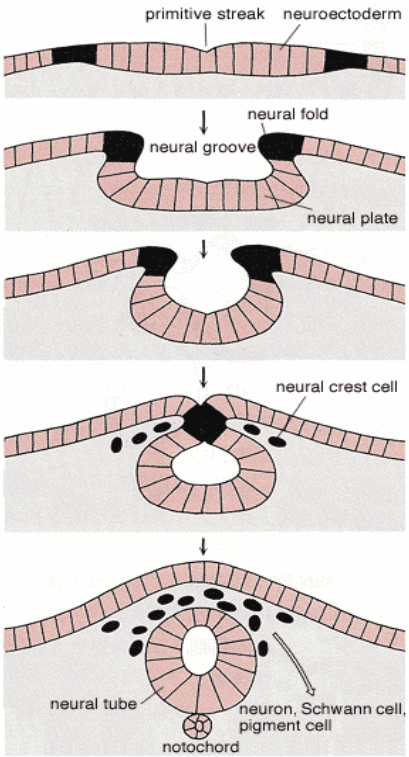
- 80% in lumbosacral region

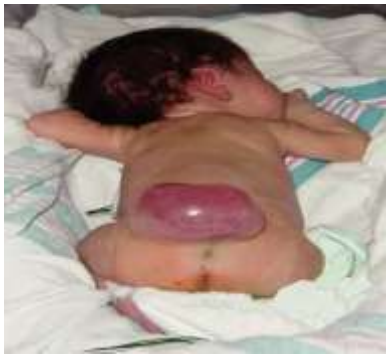
CNS: neuroectoderm in origin > neural plate > neural groove > neural fold > more folding > ends try to proximate and touch each other > forming neural tube > migrate rostral forming brain & caudally > spinal cord

Then it'll be covered by mesoderm > forms the bones "bodies vertebra" & lamina & spinal process (posteriorly) > failure of that called spina bifida

- Myelomeningocele: herniation of S.C > if S.C outside the canal & will effect the lower limb of baby b/c it's not functioning
- Meningocele: only contains CSF covered by meninges

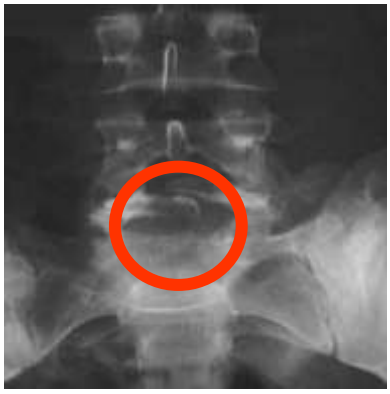
Commonly in lumber area





TYPES OF SPINAL BIFIDA OR MYELODYSPLASIA

- Spina bifida occulta
- Lipomeningocele "lump of fat in spinal canal"
- Meningocele
- Myelomeningocele (Spina bifida)



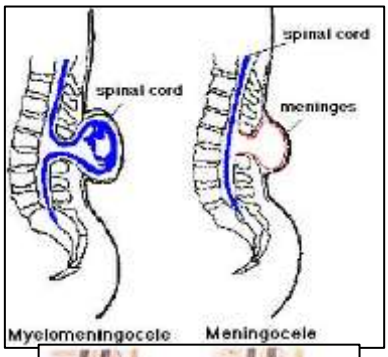
Spina Bifida Occulta: The two laminae try to reach each other but are not touching each other and do not form

1. SPINA BIFIDA OCCULTA

a. Found incidentally when the patient does x-ray for any reasons.
b. 5-10% of population so it's common.
c. not clinically significant "asymptomatic"
d. tuft of hair, dimple sinus or port wine stain
e. high incidence of underlying defect
f. no treatment required, U/S or MR

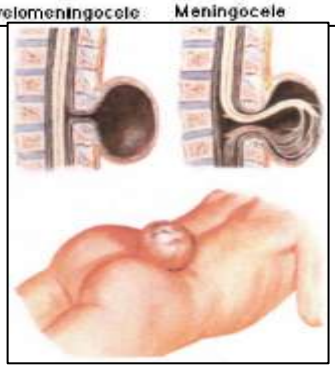
2. MENINGOCELE

(Sac filled with CSF but no spinal cord)



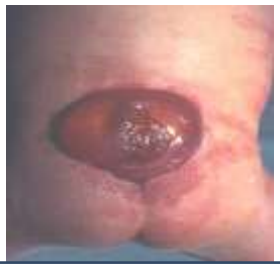
a. Cystic CSF-filled cavity lined by meninges no neural tissue → so no neural abnormalities "motor or sensory deficit in lower limbs" but some have some autonomic deficits like sphincters; Incontinent or nocturnal enuresis.
b. communicates with spinal canal
c. look for other cong. Anomalies; seldom any neurological deficit
d. DX: U/S or MRI
e. **Urgent** excision if CSF leak "if it rupture occurs", otherwise deferred perhaps indefinitely if small.

3. MENINGOMYELOCELE



a. Spinal cord and roots protrude through the bony defect, lie within cystic cavity, if ruptured, CSF will leak trans-illumination (emergency case).
b. Because the S.C outside >> the pt will have autonomic, sensory and motor deficit in the lower limbs and they are born with paraplegia.
c. Observe limb movements (degree & level of neurological damage) and note dilated bladder & patulous annual sphincter.
d. Dx :U/S or MRI

e. If ruptured; immediate closure & replacement of neural tissues into spinal canal .
f. Look for other cong. Anomalies : gross hydrocephalus , Chiari malformation and other multiple serious cong. Anomalies
g. Many adopt thoughtful conservative treatment.
h. Ruptured meningiocele/ meningiomyocele is a surgical ER

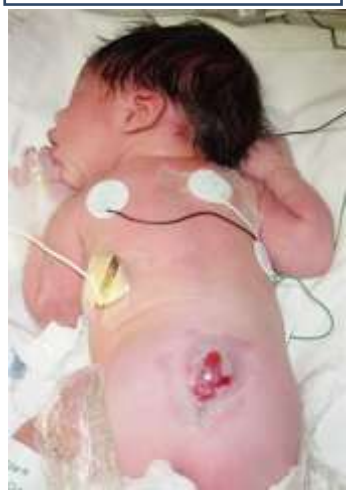


Meningomyelocele baby: thin layer of membrane > if it ruptures > leak of CSF > infection "meningitis"

4. ANENCEPHALY

(No brain)

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



CLINICAL MANIFESTATIONS

According to the level of spinal bifida (Meningomyelocele)

- Lower Lumbar (L5,S1) → Distal weakness in the feet and sphincters incontinent
- Upper lumbar (L1) → Complete paraplegia of the lower limbs.

Myeloschisis or rachischisis: where there is defect in the bone and the spinal cord exposed outside no skin covering > type of Meningomyelocele Sever neurological deficit in the lower limbs.

INCIDENCE

- 2/1000 births, .2-.3/1000 in Scandinavia b/c they put folic acid in flour and they do therapeutic abortion once baby diagnosed with spina bifida.

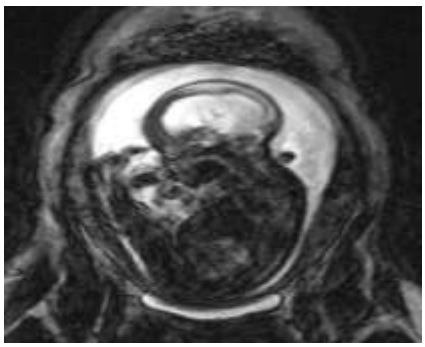
Risk increase to 5% if a sibling is affected

Anencephaly

CAUSES

1. The main cause is Folic acid deficiency; since the development of the nervous system end in day of 28 of pregnancy. The woman should take folic acid when they plan to get pregnant.
2. Teratogens; Sodium Valporate (antiepileptic drug , if you start giving this medication, once she becomes pregnant she may develop baby with spina bifida > so you need to change it before pregnancy)

ANTENATAL DIAGNOSIS



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

* Associated with Hydrocephalus, Chiari II and aqueduct forking

PREVENTION

Give folic acid supply during pregnancy

TREATMENT

- Short term treatment:
  - If the defect has normal skin covering>>> Treatment in elective basis (no needs to emergency surgery)
  - Thin or no skin covering at all, ruptured sac >>>early surgery as possible to prevent meningitis.

But even we do the surgery the problems did not solve, neurological deficit or other congenital CNS anomalies. They need Long term treatment and that includes:

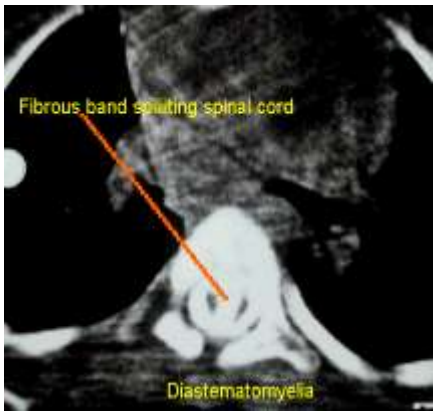
- Long-term treatment
  - Early treatment of hydrocephalus
  - Regular follow up in spina bifida clinic
  - Urological, orthopedic, paediatrics, and physical therapy
  - Urological; urinary incontinence, vesicoureteric reflux, repeated UTI, renal impairment, hypertension and stunted growth
  - Orthopedic; feet deformity, and tendon transfer, pelvic and spine deformities
  - Neurosurgical; tethered cord (needs detether) , chiari II malformation, shunted hydrocephalus
  - At least once or twice a year you have to see them

OTHER CONGENITAL ANOMALIES

- Tethered spinal cord
- Diastematomyelia
- Lipomeningocele: Definition: fat in the meningocele.
- Congenital dermal sinus: The baby is born with back dermal sinus with tuft of hair, check for spina bifida in this baby.

1-DIASTEMAMYELIA

- A bone or fibrous band divides spinal cord in two longitudinal sections **IMP!**
- Associated lipoma may be present, which tethers cord to vertebra
- Sings &Symptoms include weakness, numbness in feet, urinary incontinence, decreased or absent reflexes in feet
- Dx: CT
- Rx - surgery to free cord



2- CHIARI I MALFORMATION

Cerebellum herniation through foramen magnum >> crowding of the area >> affect the CSF circulation >> dilation of the CSF within the S.C; this is called syringomyelia

- Two types:
  - Type A → no spinal bifida (Meningomyelocele)
  - Type B → with spinal bifida

** Chiari II malformation, in addition to cerebellar herniation, there's also brain stem herniation.*

CLINICAL MANIFESTATION OF CHIARI MALFORMATION

The babies have clinical manifestation called foramen magnum syndrome, which include symptoms like:

- Pressure of the S.C in that area.
- Stretching of lower cranial nerves, associated syringomyelia
- Leads to neck pain, Headache, upper limbs symptoms, weakness of hands , loss of fine touch ,loss in temperature sensation in the tip of the fingers ,upper motor neurons manifestations of the lower limb.

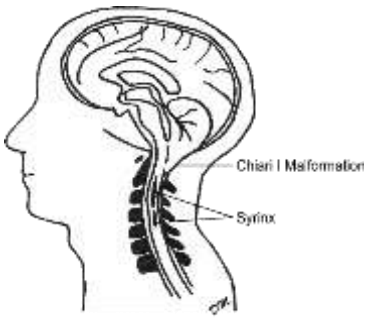


Figure 2. Chiari I Malformation

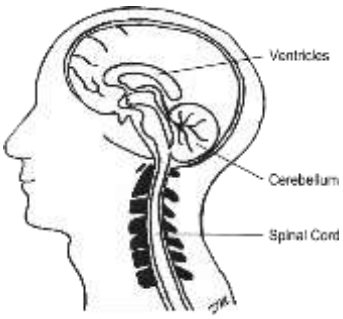


Figure 1. Normal

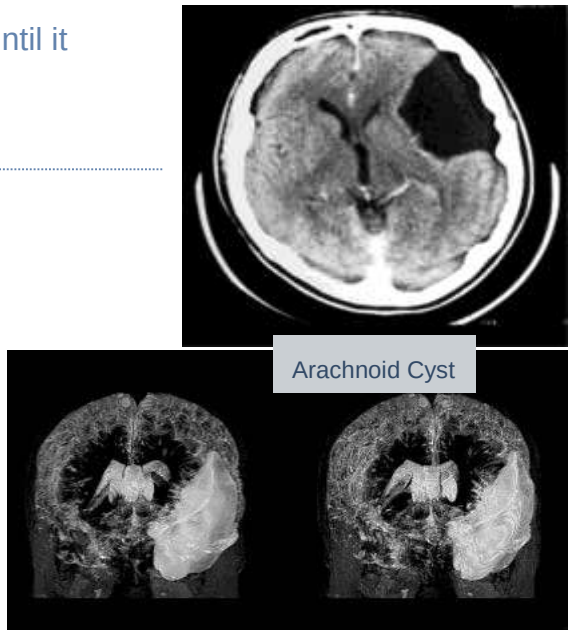
Downward herniation of cerebellar tonsil through foramen magnum into cervical spinal canal

OTHER CONGENITAL ANOMALIES

ARACHNOID CYST:

Large cyst around the brain, it is asymptomatic until it becomes large then the symptoms appear

→ Treat if large



Arachnoid Cyst

ENCEPHALOCELE

Large cyst in the skull contains CSF or brain

- Usually occipital
- May contain occipital lobe, or cerebellum often associated with hydrocephalus Immediate treatment if ruptured outcome depends upon contents

Encephalocele

Emergency if it ruptures

MCQS

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- In arachnoid cyst, what is false?

A) Occurs commonly in the temporal area.
B) All cases should be treated urgently to avoid complications
C) May present with seizures
D) May be asymptomatic

3- The most common type of cerebral herniation is:

a) Central
b) Cingulate
c) Transtentorial
d) Tonsillar

4- The investigation of choice in increased ICP is:

a) Skull x-ray
b) MRI
c) Lumbar puncture
d) CT scan

(Extra Notes)

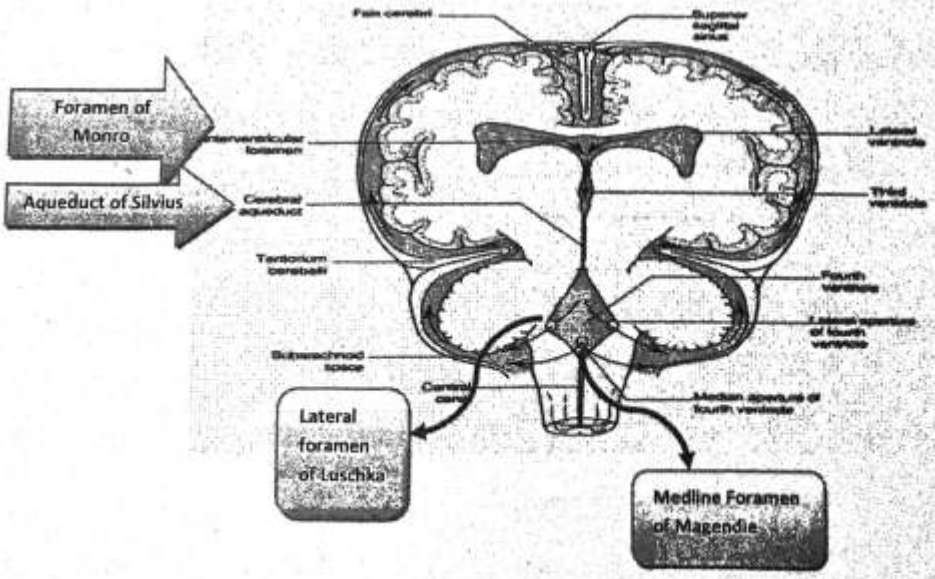
Hi everybody >>>>>>>>>

I know that u all have troubles with the *common congenital neural tube defects* so to be helpful I thought to publish these notes said by the Dr but not in 426 notes..... Hopefully they`ll be beneficial 4 u all.....besides THE DR DIDN'T MENTION ONE SINGLE THING ABT *NEUROMIGRATION DISORDERS & DISORDERS OF CORTICAL DEVELEPMENT* OR *NEUROCUTANEOUS SYNDROMES* ! I DON'T KNOW... IT MIGHT BE IN THE COMING LECTURE HE IS SUPPOSED TO GIVE BUT SUER NOT IN THIS ONE ..... THOUGH ITS UP TO U GUYZ IF U WANT TO STUDY IT.....

Hydrocephalus is the commonest congenital anomaly in the CNS.

Large ventricles in the scan doesn`t mean hydrocephalus. Symptoms of altered ICP is by definition Hydrocephalus.

Normal circulation of the CSF in the brain:



❖ Congenital

1. **Aqueductal anomalies**

aqueductal stenosis (non-communicating type) is the commonest cause of congenital hydrocephalus.

2. **Dandy Walker malformation**

Large cyst in the post. Cerbellar fossa in which the space filled with CSF the cerebellar hemispheres are too small and no vermis.

3. **Chiari II malformation**

Herniation of the cerebellum (tonsils) downward through the foramen magnum into cervical Spinal canal.

There are 4 types of chiari but type II is usually associated with coccal neural nibe defect (myleomeningocele) which is seen in children just after birth.

4. **Myleomeningocele**

Associated with hydrocephalus not necessary an obstructive it could be a communicating type (due to venous drainage i.e. a problem of absorption)

5. **Intrauterine viral infection** (CMV, mumps, rubella, varicella)

Infection in general cause **fibrosis** in which decrease the flow rate but it is a communicating type : there is no obstruction in the ventricles or their foramen.

6. **Toxoplasmosis**
7. **Congenital tumors** children who've been born with tumor in the brain usually obstructive
8. **Vein of Galen aneurysms**

The only aneurysm that we'll hear about
Large aneurysm in the ventricular cavity.
Usually hydrocephalus is treated by SHUNTS but in this case shunt is contraindicated because it may lead to rupture of the aneurysm and causes fatal intraventricular hemorrhage.

If u saw the phrase Severe TBI = sever TB infection..... don't confuse! Nothing new.....

McEwen's *cracked pot* sign with cranial percussion i.e. as you are taping a pot of water.

Sunset sign (forced downward deviation of the eyes, a neurologic sign almost unique with hydrocephalus, u c a rim of the sclera around the pupil).

N.B: It is not safe to do a lumber puncture in a child with obstructive hydrocephalus such in compressing tumors because when the CSF is out the brain may herniated through foramen magnum (coning) which leads to death.

❖ Investigations

- ultrasound
- *CT scan*

- Lateral & 3rd ventricle dilatation
- → Normal 4th ventricle: suggests aqueductal stenosis (obstructive HC)
- → deviated or absent 4th ventricle: suggests posterior fossa tumor (look like Mickey mouse)

Intracranial shunts: "in which we drain the accumulative fluid out of the body using a cathater".

⊕ Neural Tube Defect

We call it:

- *Spinal Dysraphism*
- *spina bifida*
- *Failure of closure of posterior neural arch* Open or Closed
- *80% in lumbosacral region but we can see it anywhere in the spine*

SPO = spina bifida occulta

Spina bifida occulta very common anomaly but without symptoms.
5-10% of population
Not clinically significant discovered accidently when a pt. have a scan for any reason

It's a Tuft of hair, dimple sinus or port wine stain
High incidence of underlying defect
No treatment required, U/S or MRI

- Meningocele
- Cystic CSF-filled cavity lined by meninges
- no neural tissue the lower limbs will be normal
- communicates with spinal canal
- seldom any neurological deficit
- U/S or MRI
- urgent excision if CSF leak, otherwise deferred
- perhaps indefinitely if small

It is caused by folic acid deficiency at 28 days the problem is mothers knows they are pregnant after they m too late to correct the damage if happened.
Solution is to take folic acid supplement before deciding to conceive.

- Spinal cord and roots protrude through the bony defect,
- lie within cystic cavity, if ruptured, CSF will leak
- trans-illumination
- observe limb movements (degree & level of neurological damage)

It cannot be corrected because the neural tissue grew in the wrong place so even if we tried to put it in the right place it won't be functional effect.

- note dilated bladder & patulous annual sphincter
- U/S or MRI
- if ruptured; immediate closure & replacement of neural tissues into spinal canal
- gross hydrocephalus, multiple serious cong. anomalies; many adopt thoughtful conservative treatment

- Antenatal diagnosis it is not for ttt but for delivery safety and eases the child nursing
- Maternal U/S,
- MRI
- Serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase
- Contrast enhancing amniography

- possibility of therapeutic abortion

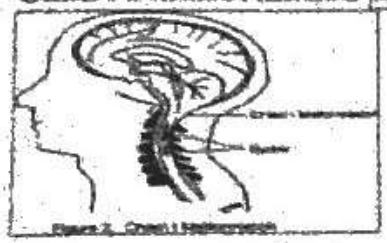
Tethered spinal cord in which the
appear with growth as S.C compres

In general those children are wheal chaired but very smart and strong, good coordination of upp limbs.

Diastematomyelia not very common

Congenital dermal sinus to prevent infec

Chiari I malformation small foramen magnum cause severe headache



Transtentorial herniation means Passing across or through either the tentorial notch or tentorium cerebell which occure in chiari II malformation which is the most common while tonsilar occure with the less frequent chiari I malformation.

GOOD LUCK DOCTORS☺
WISH ME LUCK TOO ☺

Always yours:
Nour A. AlMalki