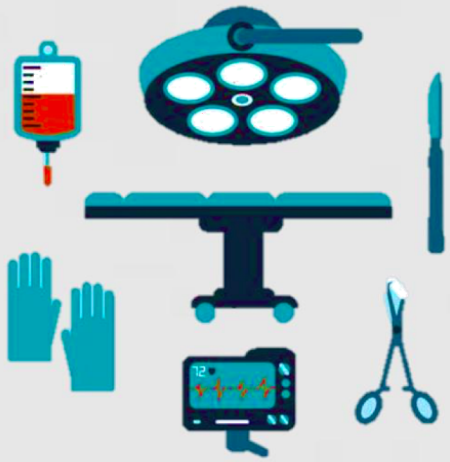

Common congenital neurosurgical diseases

Objectives:

- Introduction to Neurosurgery.
- Approaching neurological symptoms.
- Congenital diseases, including:
  - Hydrocephalus.
  - Neural tube defect.
  - Chiari Malformation.
  - Dandy-Walker Malformation.
  - Craniosynostosis.
  - Arachnoid Cyst.

Resources:

- Davidson's.
- Current diagnosis & treatment.
- Raslan.
- 436 Doctor's slides.
- 435' team work.

Done by: Nehal Beyari, Abdulrahman Zekry, Razan Alotaibi, Khalid Aleedan.
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Revised by: Yara Aldigi

Color Index:

Notes , Important , Extra , Davidson's

Editing File

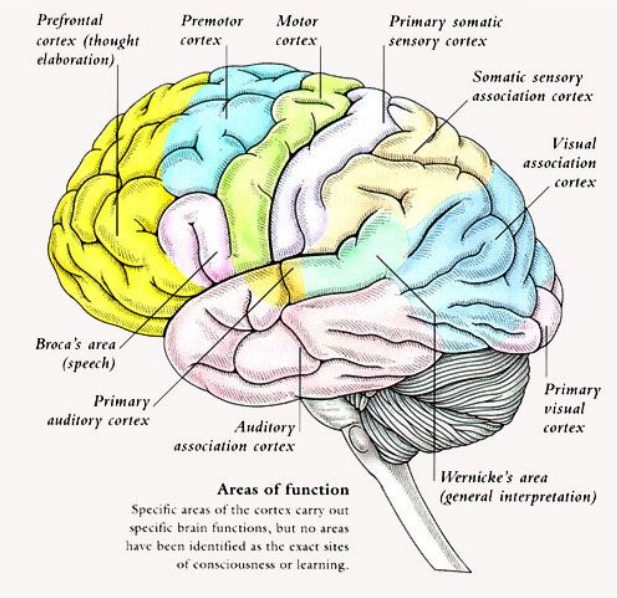
Feedback

Basic review: From 435, for your knowledge.

The Localization of a lesion requires an understanding of the anatomy and physiology of the nervous system, its blood supply, and the disease processes that affect it. The process of localization begins during history taking, and is refined during the general and neurological examinations, and reassessed after any relevant diagnostic studies are completed.

★ **During the process of localization, ask yourself three questions:**

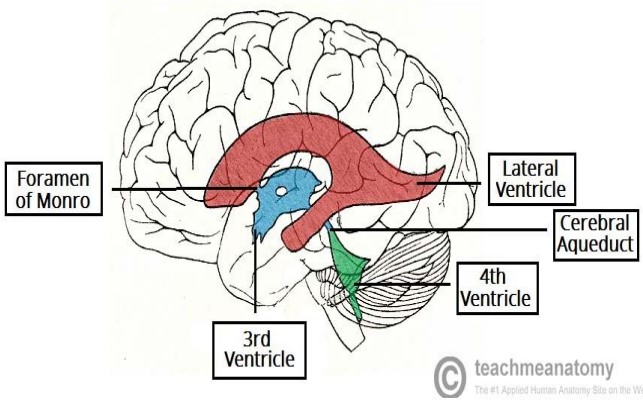
1. What is the necessary minimal amount of neuroanatomy that must be damaged in order to produce the patient's symptoms/signs?
    a. A patient with monocular blindness must have a lesion on the same side somewhere between the cornea and optic chiasm.
    b. A patient with an absent tendon reflex must have a lesion within the afferent-efferent arc serving that reflex.
    c. A patient with aphasia must have a lesion of the dominant (95% left) cerebral hemisphere.
2. After localizing the lesion, does it explain all the findings? If the answer is no, you should carefully re-examine the data and your conclusions. Either the localization is wrong or the disease process is multi-focal or diffuse.
3. After localizing the lesion, what else should be present? That's another way of saying, does the patient have the expected neighborhood signs to go along with your proposed localization?



If you want to know more about how to localize a lesion, you can check this file from 431. Highly recommend it

★ Ventricular system :

- Is a set of communicating cavities within the brain. These structures are responsible for the production, transport and removal of cerebrospinal fluid, which bathes the central nervous system.
- They are lined by ependymal cells, which form a structure called the choroid plexus. It is within the **choroid plexus that CSF is produced.**
- In total, there are four ventricles; right and left lateral ventricles, third ventricle and fourth ventricle.
    - The lateral ventricles are connected to the third ventricle by the foramen of Monro.
    - The third ventricle is situated in between the right and the left thalamus.
    - The fourth ventricle is the last in the system. It lies within the brainstem, at the junction between the pons and medulla oblongata. Drains fluid into two places :
        - Central spinal canal – Baths the spinal cord
        - Subarachnoid cisterns – Baths the brain, between arachnoid mater and pia mater. Here the CSF is reabsorbed back into the circulation.



Approaching Neurological Symptoms

Headache or facial pain: Not every pain in the head is headache, it could be facial or dental problem.

Primary headache (not serious)	Secondary headache (serious)
• Are benign, recurrent headaches not caused by underlying disease or structural problems. • Examples: Migraines, tension-type headaches, cluster headaches. • Tension-type headache is one of the most common causes of primary headache, it comes with stress. (important) • Primary headaches usually have very typical presenting features.	Are caused by an underlying disease: 1. International Headache Society (IHS) classification 2. Certain "**Red flags**" indicate a secondary headache may be dangerous (**SSNOOP**) is a mnemonic to remember the red flags for identifying a secondary headache ○ **S**ystemic symptoms (fever or weight loss). ○ **S**ystemic disease (HIV infection, malignancy). ○ **N**eurologic symptoms or signs. ○ **O**nset: sudden (thunderclap headache). ○ **O**nset after age 40 years. ○ **P**revious headache history (first, worst, or different headache) In general people complaining of their "first" or "worst" headache, progressively worsening.

Is the headache serious?

The American College for Emergency Physicians published criteria for **low-risk headaches**:

- Age younger than 30 year. But not child. Children with a headache is a serious red flag.
- Features typical of primary headache.
- History of similar headache.
- No abnormal findings on neurologic exam.
- No concerning changes in normal headache pattern.
- No high-risk comorbid conditions (for example, HIV).
- No new concerning history or physical examination findings. Any change in History and Physical examination is a red flag.

Differential Diagnosis of headache: Always think systematically when making a Ddx, and don't jump to conclusions to diagnose immediately!

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

Differential diagnosis of 906 patients who presented to a general neurology clinic with headache or facial pain as the major or only symptom

Diagnosis	Number	%
Tension headache	296	32
Migraine	241	27
Headache ? Cause	139	15
Post-traumatic	71	8
Facial pain ?cause	38	4
Depression	29	3
Trigeminal neuralgia	29	3
Cluster headache	19	2
Malignant IC Tumour	14	1.5
Benign IC Tumour	9	
Temporal arteritis	6	
Post-herpetic neuralgia	5	
Benign IC hypertension	4	
Cough headache	3	
Subdural haematoma	2	
Sinus infection	1	

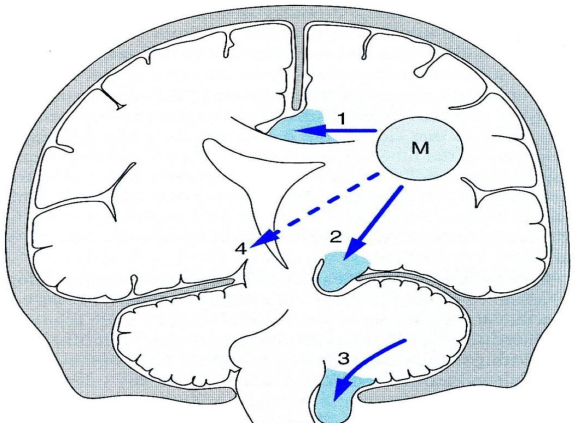
These are the most common causes of headache, there might be a question saying: which of the following is the most common cause of headache? Listing 3 from the ones below and only one from above (which is the answer).

Differential Diagnosis of CNS space-occupying:

Neoplastic → Vascular → Congenital → Inflammatory → Infectious.

لما يجيكم المريض، دائماً حطوا ببالكم هالثلاث نقاط:

1. **What is causing the pathology (ddx)?**
2. **Where is the pathology (location)?**
3. **Severity.**



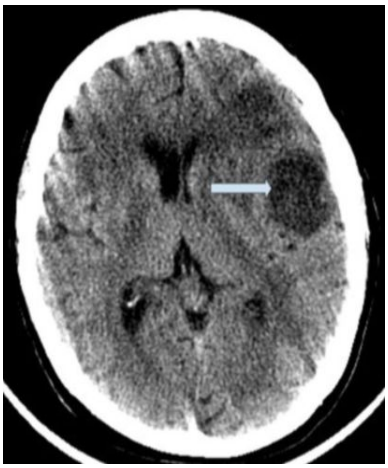
*What can cause these symptoms? (look at the picture):

1- Local compression

2- Mass effect & herniation

3- High ICP. Be careful when using terminology, all of us have ICP, what's abnormal is **high** ICP!

Local compression	Mass effect & Herniation	High ICP
Functional areas: -Motor cortex→ weakness. -Sensory→ numbness or seizure. -Cerebellar→ tremor, dysarthria, ataxia or even truncal ataxia. -Pituitary adenoma→ visual loss.	When it starts to increase in size, the brain tissue will shift to the other side. Most common and serious. - If there is a **left** side epidural hematoma increasing in size causing **Left uncal herniation**, what other 2 major symptoms you are going to see in the exam? 1. Contralateral weakness (right) hemiparesis and hemiplegia. Compression of midbrain cerebral peduncles. 2. Ipsilateral fixed dilated pupil (afferent in **cranial nerve 2** and efferent in cranial nerve 3 which is usually compressed by the herniation) Uncal herniation is an URGENT case! لما يكلموني ويقولوا عندك مريض فيه dilated pupil على طول أجي للمستشفى! Uncal herniation associated with Kernohan syndrome (notch). هذي السندروم هي حقت الدوافير،واحنا طفشانين بالأوسكي نحب نسألهم إياها	S/S: • Headache. • Nausea. • Vomiting. • High BP. • Impaired level of consciousness. • Papilledema. You need to remember them by heart.



This is a CT Scan of a patient with HIV presenting with headache, nausea and vomiting which shows a left sided brain mass

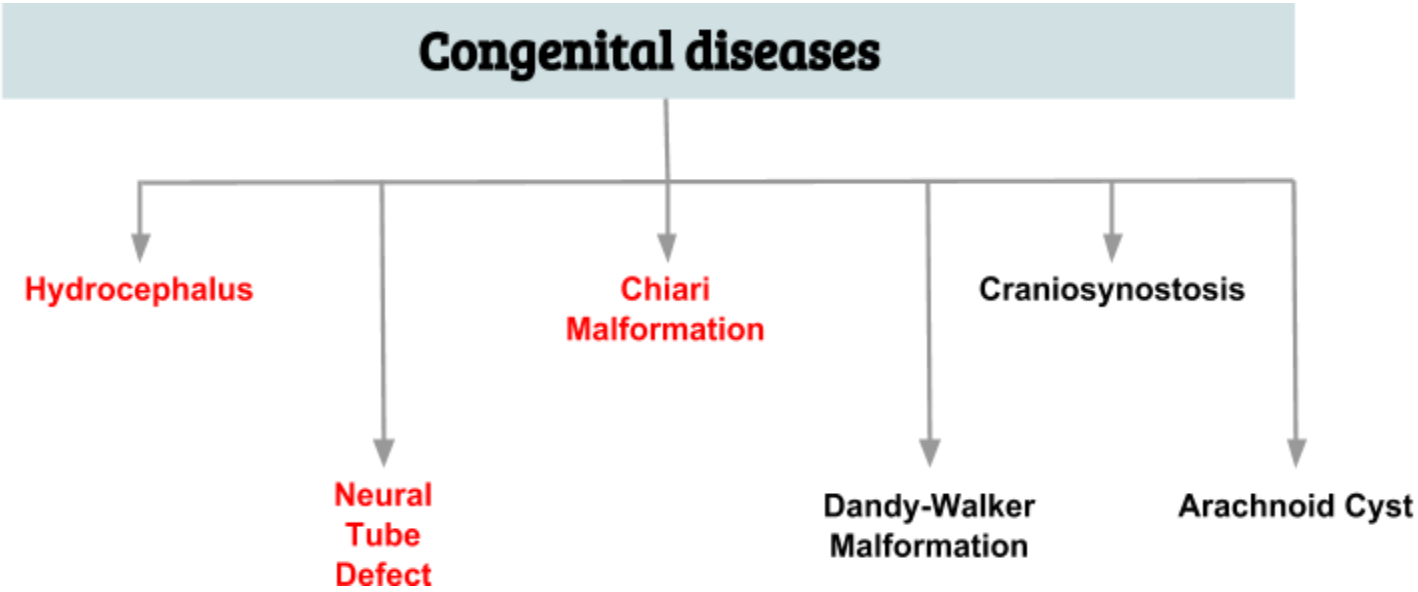
هنا مثلاً هذي ال lesion إيش ممكن تكون؟

نمشي زي ماعلمتكم فوق بالترتيب: vascular, inflammatory etc..

إيش تسبب: بما إنها في

frontal lobe specifically in front of the central sulcus, so contralateral weakness and if it's the dominant hemisphere maybe it'll cause aphasia too.

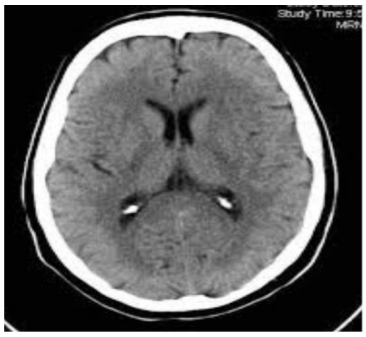
Common Congenital Neurosurgical diseases



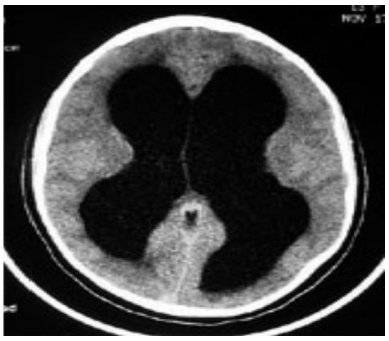
1. Hydrocephalus:

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

Not every increase in size means there is **active hydrocephalus** or dense pressure. For example, an 80 y/o patient with dementia can have ventriculomegaly but it's due to brain shrinking (atrophy)! So, every active hydrocephalus is ventriculomegaly, but not the opposite.



Normal

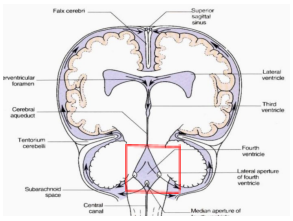


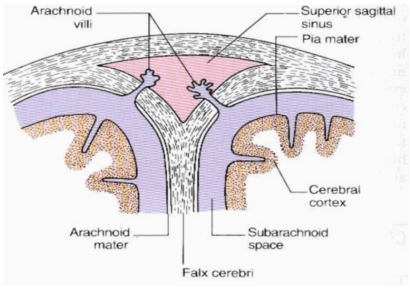
Hydrocephalus

Physiology of CSF

Helpful video

- Total volume of CSF in the ventricles varies from 5-15 ml in neonates to 150 ml in adults.
- Produced mainly by choroid plexus.
- Rate of production is 0.3-0.4 ml/minute.
- Reabsorbed in the superior sagittal sinus.





هذه الصورة تكبير للمربع اللي بالاحمر:

Arachnoid villi (site of obstruction in hydrocephalus due to meningitis, site of fibrosis in subarachnoid hemorrhage)
Where does the production & absorption of CSF happen?
Produced from the lateral ventricle (choroid plexus) and absorbed into the Superior sagittal sinus via Arachnoid villi.

Causes of Hydrocephalus (pathophysiology)

- Obstruction of CSF flow. The most common.
- Under absorption of CSF. CSF production = CSF reabsorption, it happens when a person is (for example) producing 700 ml and only absorbing 300 ml.
- Overproduction of CSF

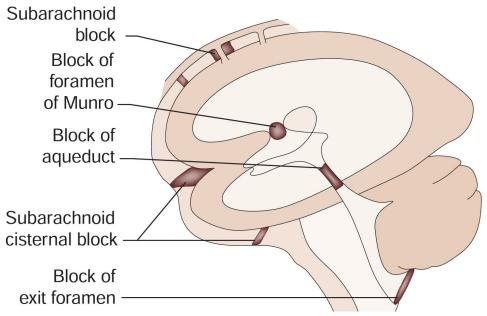


Fig. 24.17 Hydrocephalus: sites of cerebrospinal fluid (CSF) blockage.

Types of Hydrocephalus:

Communicating (Non obstructive) Usually **chronic** presentation, like post-meningitis	• Overproduction or under absorption of CSF • All ventricles are dilated • No obstruction in the pathway of CSF within the ventricles (the ventricles can communicate with each other)
Non-Communicating (obstructive)	Complete or incomplete obstruction of CSF within or at the exit of the ventricular system.

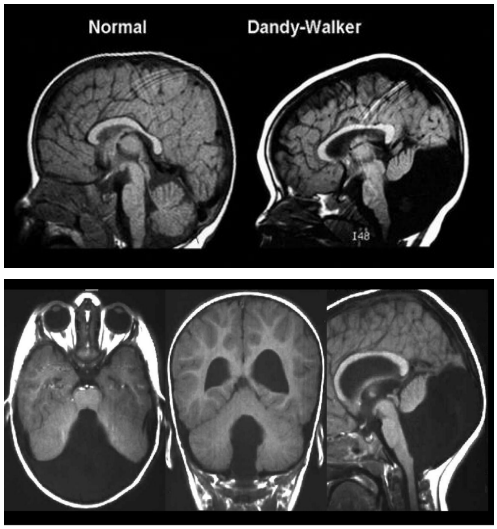
	Causes: ● Blockage of the flow of CSF (obstruction) within ventricles or the pathway of CSF (obstruction to the CSF flow at the foramen of Monro, the third ventricle, the aqueduct of Sylvius, the fourth ventricle, or the foramina of Magendie or Luschka): ○ Congenital, since birth. ○ Acquired, develops after birth. ● Partial dilatation.

- **Etiology:**

Congenital	● Aqueductal anomalies (stenosis): (**most common cause**): Infants will come after 2 weeks of birth with vomiting, sleepiness, crying, increased head circumference very rapidly and CT will show obstruction (stenosis) at the level of the aqueduct → non communicating hydrocephalus. ○ In MRI and CT scans we'll see the lateral ventricle and the 3rd are dilated but not the 4th. (abnormal communication between the 3rd and 4th) ● Dandy Walker malformation. Common ● Chiari II malformation. Common ● Myelomeningocele. ● Vein of Galen aneurysms. Rare. ● Intrauterine viral infection (CMV, mumps, rubella, varicella). ● Toxoplasmosis. ● Congenital tumors. ● Chromosomal anomalies (Trisomy 13 and 18). ● Congenital or primary hydrocephalus.
Acquired	● Germinal plate hemorrhage: in **premature** babies <1500 gm (30%-40%) (immature blood vessel wall) can be acute or chronic: if acute it is obstructive there is a mass, if chronic (delay) due to problem in absorption it can be obstructive or communicating. ● Post-meningitis **non-obstructive**. Very common, hydrocephalus is the most common complication. Happens due to scarring after inflammation which decreases the absorption of CSF. If there is abscess it is mass obstruction but usually it isn't. ● Tumors. ● SAH (subarachnoid hemorrhage) bleeding in the space causes inflammation> fibrosis of arachnoid villi > decrease absorption of CSF(communicating hydrocephalus), if acute obstructing, if chronic communicating. ● Severe TBI (traumatic brain injury) if acute obstructing, if chronic communicating. ● Dural venous sinus thrombosis or Superior sagittal sinus thrombosis.

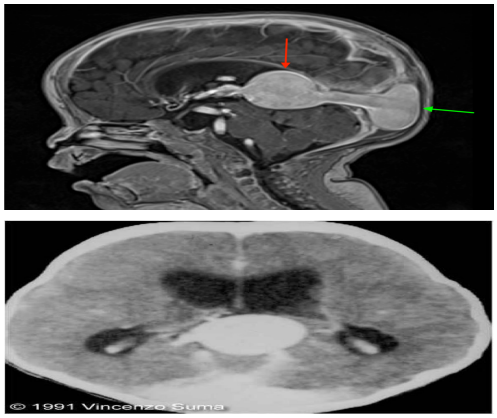
★ **Dandy Walker malformation:** know the **definition** and **location** of the lesion.

- 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) Most of cases of Dandy Walker malformation are associated with hydrocephalus.
- **Presentation** (both cerebellar and hydrocephalus symptoms): Incoordination, ataxia, nystagmus.



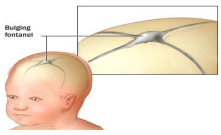
★ **Vein of Galen aneurysms:** (very rare disorder causing obstructive hydrocephalus) لما يشوفونها بالبداية بيحسبونها ورم، ولكن لازم تنتبهون لأن لو سويتو للمريض عملية بينزف ويموت

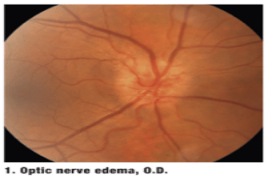
- A large vascular malformation where there is a direct communication between the arterial system and venous system (shunting), leading to dilatation of the Vein of Galen (deep venous structures in the brain) and to obstructive hydrocephalus at the level of 3rd ventricle.

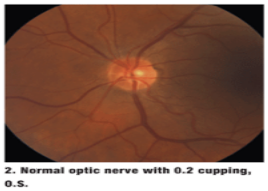


Clinical features:

Infants & young children	Juvenile & adult: (↑ ICP)
1. Increasing head circumference. 2. Irritability, lethargy, poor feeding, and vomiting. 3. Bulging anterior fontanelle. (picture). 4. Widened cranial sutures. 5. MacEwen's[1] (cracked-pot) sign with cranial percussion. 6. Scalp vein dilation (collateral venous drainage). 7. Sunset sign (downward deviation of the eyes). 8. Episodic bradycardia and apnea. Very high ICP leads to pressure on brainstem causing herniation and those are very bad signs.	1. Headaches. 2. Nausea. 3. Vomiting. 4. Decreased level of consciousness. 5. Focal neurological deficit (rare). 6. Papilledema. Scenario: patient has meningitis, headache, v/n. You need to do lumbar puncture, but before that you have to rule out high ICP. How? By fundoscopy (papilledema) or brain imaging (CT) if the question asks what's the next step in this case (picture), LP or brain imaging? choose brain imaging



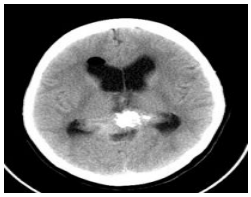
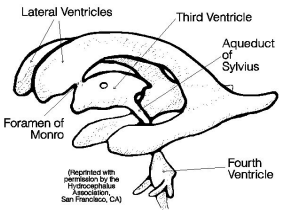
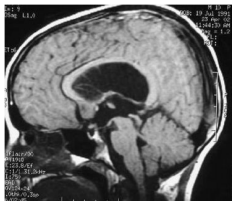
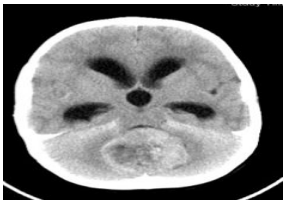
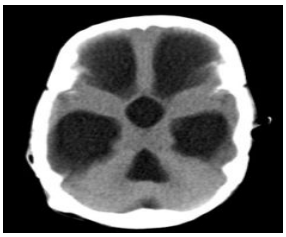

1. Optic nerve edema, O.D.


2. Normal optic nerve with 0.2 cupping, O.S.

[1] A sign to detect hydrocephalus and brain abscess. Percussion (tapping) on the skull at a particular spot (near the junction of the frontal, temporal and parietal bones) yields an unusually resonant sound in the presence of hydrocephalus or a brain abscess.

Investigations:

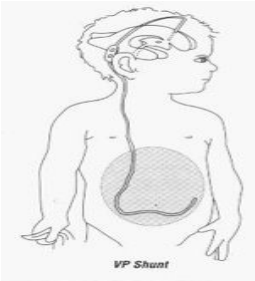
CT or MRI: The pattern of ventricular enlargement can help delineate the cause:

Lateral ventricles dilatation	Obstructive hydrocephalus caused by 3rd ventricle tumor. (absent 3rd and 4th ventricles)	
Lateral & 3rd ventricle dilatation	→ **Normal 4th ventricle:** Suggests aqueduct stenosis.	 
	→ **Deviated or absent 4th ventricle:** suggests posterior fossa tumor compressing on the 4th ventricle.	
Generalized dilatation	Suggests a communicating hydrocephalus.	

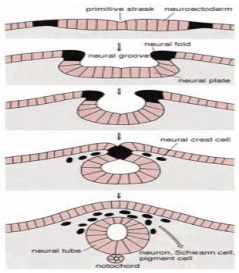
Treatment:

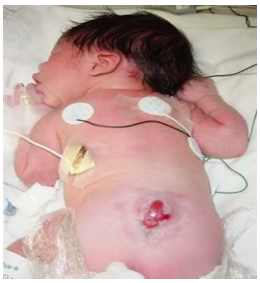
1- Communicating : Medical or surgical: communicating hydrocephalus is usually treated medically.

2- Obstructive: : SURGICAL TREATMENT: VP shunt. If it's an obstructive agent, we either do a VP shunt or surgically remove the obstructing agent.



2- Neural Tube Defect (NTD):



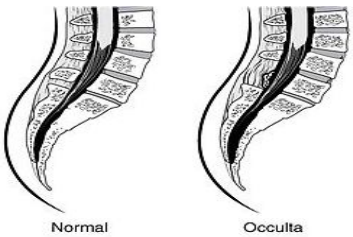
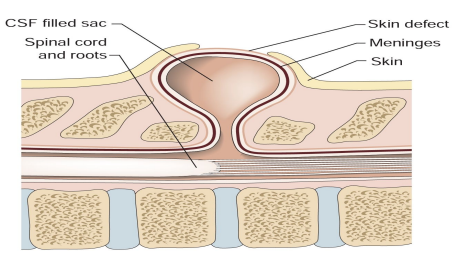
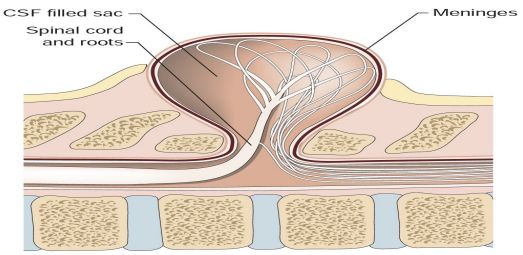


- Spinal Dysraphism is **Failure of closure of <u>posterior</u> neural arch**.

- Might happen between 3rd to 8th week of gestation.The earlier it happens the more severe the symptoms.

- Two major types: Open or Closed.

Types of myelodysplasia:

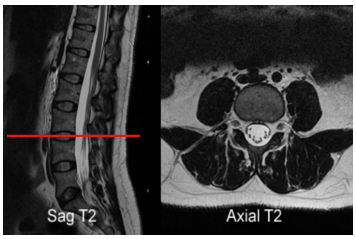
Spina bifida occulta (closed)	**Meningocele** (closed) protrusion of meninges	**Meningomyelocele** (open) protrusion of meninges and spinal cord
• 5-10% of population • not clinically significant • Asymptomatic usually, may be tuft of hair, dimple sinus or port wine stain. • High incidence of underlying defect. • No treatment required. • Common presentation: a male in his twenties presenting with back pain during weight lifting. MRI shows that everything is normal except for a little opening in the lamina.  	• Cystic CSF-filled cavity **lined by meninges** prone to infection. • **No neural tissue** • Communicates with spinal canal. • Look for other congenital anomalies. • Urgent excision if there's CSF leak (if it ruptures), otherwise no surgery needed. • Seldom any neurological deficit. (LL weakness-urinary retention) • Diagnosed by U/S or MRI.+look for signs of infection. • A sac containing CSF only compared to meningomyelocele. • Prognosis usually related to the location, the higher it is in the spinal cord the worse the prognosis.  	• 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. • Gross hydrocephalus, **multiple** serious congenital anomalies. • Diagnosed by U/S or MRI Why would we do MRI of head? It's associated with chiari malformation. • Prognosis is determined based on the site of the lesion, the higher it is the more worse the prognosis is. لما يكلمونا الطوارئ يقولون وحده ولدت ماكانت تتابع معنا والبيبي عنده spina bifida ايش أول سؤال نسألهم ؟ هل فيه فلويد يطلع او لا ؟ لان لو مفتوحة الان الان لازم نروح العملية ونقفلها لأنه عرضة للانفكشن. Treatment: close it! 

Questions in history taking in Spina Bifida:
Ask about the motor function (nerves are protruding), signs of infection (meningitis), urinary retention, pregnancy and antenatal care.

Incidence:

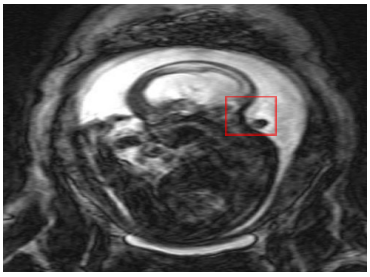
- 80% in lumbosacral region (Occasionally in the head).
- 2/1000 birth:
    - Risk increase to 5% if a sibling is affected. More common in SA.
    - Teratogens ex: antiepileptics بالغالب كمان هذا من الأسباب اللي ترفع النسبة عندنا غير انو مافي تخطيط للحمل، الصيدلي عندنا يقدر يصرف دواء وما يسأل اذا فيه حمل او لا فيؤثر على الجنين

How to prevent? By giving folic acid supplement before pregnancy in high dose, and plan pregnancy.



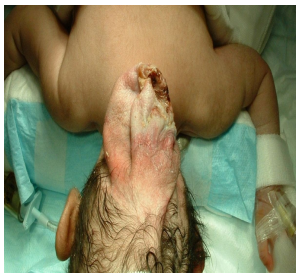
Lesion will happen at this level of the spine (see picture - red line), so all the structures in this area can be affected, and the severity varies from pt to another. Example: one might come with only skin manifestations, others come with canal involvement etc.

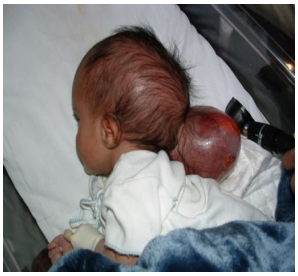
Antenatal diagnosis: Red square in the picture: its not cute! It's BAD, it will grow after birth (encephalocele). Not important.



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

3- Other congenital anomalies:





★ Encephalocele:

- Usually occipital
- May contain occipital lobe, or cerebellum
- Often associated with hydrocephalus
- Immediate treatment if ruptured
- Outcome depends upon contents.
- Very bad and they usually don't survive especially if it ruptures.

★ Chiari Malformation:

It is the herniation of posterior fossa's content below the foramen magnum. You only need to know the first 2 types.

Type I Adults type Mostly doesn't need surgical intervention	Extension of the cerebellar tonsils into the foramen magnum, without involving the brain stem. The only type that can be acquired. Clinical features: headache - n/v - electrical shock - ataxia - nystagmus - hydrocephalus (papilledema).

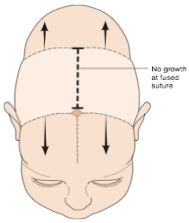
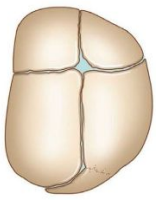
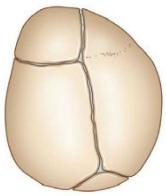
Type II children type A triad!	More serious than type 1, Extension of both cerebellar and brainstem tissue into the foramen magnum, **associated with: Myelomeningocele, Hydrocephalus.**
Type III	Rare, herniation of the cerebellum with or without the brainstem through a posterior encephalocele.
Type IV	Rare, Cerebellar hypoplasia or aplasia with normal posterior fossa and no hindbrain herniation.

★ Arachnoid cyst:

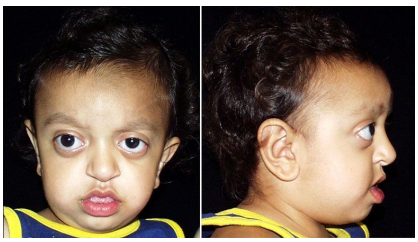
- Cystic collections of CSF of developmental origin.
- Arachnoid cysts may gradually increase in size, either due to CSF being driven in through a valve-like opening or by active secretion of fluid from the cyst wall.
- Occasionally patients present with mass effects > Convulsions, raised ICP
- More often they are discovered **incidentally** on CT or MRI.
- Treatment: shunting.

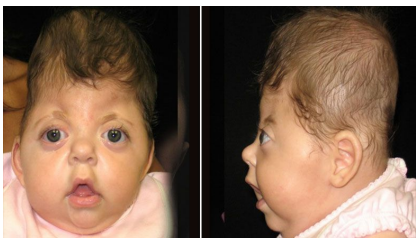
★ Craniosynostosis:

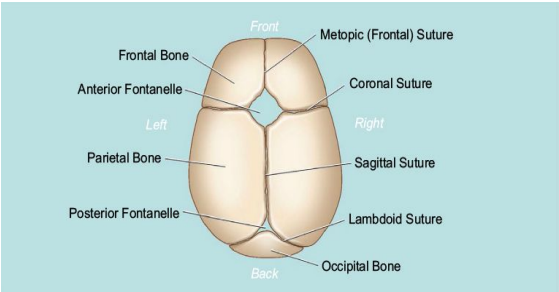
- Definition: it is the premature closure or absence of a cranial suture.
- We have three types of cranial sutures: Sagittal, Coronal and Lambdoid.
- Each suture fuses at a different age, but premature fusion leads to asymmetrical skull growth.
- Fusion of a single suture is associated with certain typical head shapes, which are **either:**

1. Scaphocephaly	2. Plagiocephaly
Premature closure of the Sagittal suture. Growth normally occurs perpendicular to the suture line. In this case, the skull cannot widen, as there is no growth at the fused suture, which may be palpable as a ridge in the midline. There is compensatory growth at the coronal and lambdoid sutures leading to an elongated head shape (enlarged antero-posterior diameter). 	Premature closure of the Coronal suture -Anterior plagiocephaly-. However, it may be caused by fusion of the lambdoid suture -posterior plagiocephaly- (rare).   Lambdoid *Synostotic Posterior Plagiocephaly* Unicoronal *Synostotic Anterior Plagiocephaly*

- Sometimes, more than one suture can be affected. This can be syndromal (e.g. Crouzon's or Apert's syndrome) which are associated with characteristic craniofacial deformities.


Crouzon's syndrome


Apert's syndrome



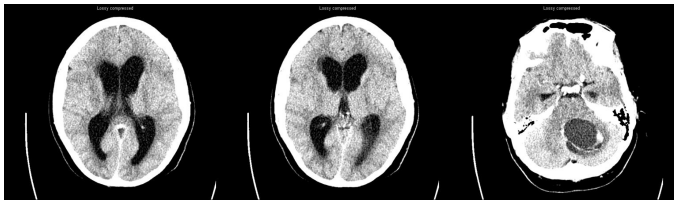
A case from the Doctor's slides

A 4 y/o patient presented with a 7 week period of worsening headache in the occipital area, and worsened last night:

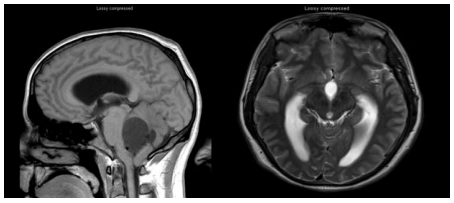
- He has no past medical history.
- The headache is associated with dizziness, loss of balance, nausea and vomiting (twice over the last 3 weeks).

4. **Red flag**: patient is very young, worsening headache (a sign of a space occupying lesion which is increasing in size), localized, worsened last night (a sign of high ICP -specifically hydrocephalus-), N/V (also a sign of high ICP), dizziness and loss of balance.
5. **Again, you need to think about three things in every presentation:**
    a. **What is causing the pathology (ddx)?**
        i. Could be vascular (less common in pediatrics).
        ii. Could be neoplastic (or benign lesion).
        iii. Abscess (but we can exclude it because there's no fever).
    b. **Where is the pathology (location)?** Posterior fossa (most probably a lesion compressing on the cerebellum).
    c. **Severity:** very severe and it's progressing.
6. Examination:
    a. Vital signs (if it's a severe case): high blood pressure, bradycardia, high ICP, breathing problems.
    b. Cranial nerves examination:
        i. 2: affected due to papilledema.
        ii. Cranial nerves that are responsible for balance: 8, 9, 10, 11, 12: nystagmus, ataxia.
7. Investigations:

CT scan: Mass in the posterior fossa causing obstruction at level of the fourth ventricle (lateral ventricle and third are dilated but not the fourth)



MRI: more details



- Since the mass is in the posterior fossa, so findings on examination may be caused by **focal compression of the cerebellum** which are:

• Gait ataxia. • Truncal ataxia. • Limb ataxia: Finger-nose and heel-knee-shin, intention tremor, dysmetria (past pointing), dysrhythmia.	• Cerebellar dysarthria. • Hypotonia. • Rapid alternating movements (dysdiadochokinesia). • Tremor. • Nystagmus gaze-evoked, horizontal drift followed by a fast correction.

Recall :

What is Hydrocephalus?
Abnormal condition consisting of an increased volume of CSF along with distension of CSF spaces.

What are the three general causes?
1. Increased production of CSF
2. Decreased absorption of CSF
3. Obstruction of normal flow of CSF (90%of cases)

What is the normal daily CSF production?
< 500 mL

What is the normal volume of CSF?
< 150 mL in the average adult

Define"communicating" versus "non communicating" hydrocephalus.
Communicating—unimpaired connection of CSF pathway from lateral ventricle to subarachnoid space.
Noncommunicating—complete or incomplete obstruction of CSF flow within or at the exit of the ventricular system.

What are the specific causes of hydrocephalus?
Congenital malformation
Aqueductal stenosis
Myelomeningocele
Tumors obstructing CSF flow
Inflammation causing impaired absorption of fluid
Subarachnoid hemorrhage Meningitis
Choroid plexus papilloma causing production of CSF.

What are the signs/ symptoms?
Signs of increased ICP: HA, nausea, vomiting, ataxia, increasing head circumference exceeding norms or age

How is the diagnosis made?
CT scan, MRI, measurement of head circumference

What is the treatment?
1. Remove obvious offenders
2. Perform bypass obstruction with ventriculoperitoneal shunt or ventriculoatrial shunt

What is the prognosis if untreated?
50% mortality; survivors show decreased IQ (mean 69); neurologic sequelae: ataxia, paraparesis, visual deficits

What are the possible complications of treatment?
1. Blockage/shunt malfunction 2. Infection

What is hydrocephalus ex vacuo?
Increased volume of CSF spaces from brain atrophy, not from any pathology in the amount of CSF absorbed or produced

What is a "shunt series"?
Series of x-rays covering the entire shunt length—looking for shunt disruption/ kinking to explain malfunction of shunt

Spinal Dysraphism/ Neural Tube Defects

What is the incidence?
< 1/1000 live births in the United States

What are the race/gender demographics?
More common in white patients and female patients

Define spina bifida occulta.
Defect in the development of the posterior portion of the vertebrae

What are the signs/ symptoms?
Usually asymptomatic, though it may be associated with other spinal abnormalities; usually found incidentally on x-rays

What is the most common clinically significant defect?
Myelomeningocele: herniation of nerve roots and spinal cord through a defect in the posterior elements of the vertebrae; the sac surrounding the neural tissue may be intact, but more commonly is ruptured and therefore exposes the CNS to the external environment

What are the three most common anatomic sites?

1. Lumbar region
2. Lower thoracic region
3. Upper sacral region

What are the signs/ symptoms?

Variable from mild skeletal deformities to a complete motor/sensory loss; bowel/ bladder function is difficult to evaluate, but often is affected and can adversely affect survival

What is the treatment?

With open myelomeningoceles, patients are operated on immediately to prevent infection

What is the prognosis?

< 95% survival for the first 2 years, compared with 25% in patients not undergoing surgical procedures

Which vitamin is thought to lower the rate of neural tube defects in utero?

Folic acid

Summary:

Congenital diseases	Important notes
1.Hydrocephalus	**Types:** **1.Communicating:** Unimpaired connection of CSF pathway from lateral ventricle to subarachnoid **2.Non-Communicating:** Complete or incomplete obstruction of CSF within or at the exit of the ventricular system **2.Etiology:** Congenital: (Aqueductal anomalies - Dandy Walker malformation - Chiari II malformation · Acquired :Tumors – SAH – post meningitis **3.Clinical features:** In infants & young children (increase head size – poor feeding - vomiting, - McEwen's sign sunset sign)i · In adults (headache – papilledema – nausea\vomiting – decrease consciousness ·) **4.DX: CT , MRI** **5.treatment :** **1.Communicating** : Medical or surgical **2- Obstructive** : SURGICAL TREATMENT
2.Neural Tube defect	**1.Spina bifida occulta:** .patient have (tuft of hair – dimple sinus – port wine stain), no treatment is required **2.Meningocele:** Cystic CSF-filled cavity lined by meninges prone to infection-seldom any neurological deficit.- Diagnosed by U/S .or MRI **3.Meningomyelocele:** Spinal cord and roots protrude through the bony defect,lie within cystic cavity observe limb movements note dilated bladder & patulous annual sphincter gross hydrocephalus, multiple serious congenital anomalies **4. Diagnosed:** by U/S or MRI
3.other congenital	**1.Encephalocele:** Usually occipital often associated with hydrocephalus Immediate treatment if ruptured **2.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 · **3.Craniosynostosis:** Premature closure or absence of a cranial suture Scaphocephaly : premature fusion of the sagittal sutures Plagiocephaly : premature fusion of the coronal sutures or premature fusion of a lambdoid suture but this is much rarer

Questions

1-Which one of the following types of headaches is the most common one?
A-Cluster Headache
B-Migraine
C-Tension Headache
D-Drugs induce Headaches

2-What is the most common cause of Hydrocephalus?
A-Obstruction of CSF flow
B-Under absorption of CSF
C-Overproduction of CSF
D-None of the above

3-What is the most common cause of Congenital Hydrocephalus?
A-Dandy Walker malformation
B-Aqueductal stenosis
C-Vein of Galen aneurysm
D-Trisomy 13 and 18

4-Which one of the following describes this type of Chiari malformation (Extension of both Cerebellar and Brainstem into Foramen magnum)?
A-Type 1
B-Type 2
C-Type 3
D-Type 4

Answers
1- C, 2- A, 3- B, 4 -B

BEST OF LUCK!❤