Common congenital neurosurgical diseases

Objectives:

- Introduction to Neurosurgery
- Approaching neurological symptoms
- Congenital diseases: Hydrocephalus, Neural Tube Defect, Chiari Malformation

  Dandy-Walker Malformation

  Craniosynostosis

  Arachnoid Cyst

Resources:

- Davidson
- Current diagnosis & treatment
- Raslan
- Doctor's note

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[Color index | Important | Notes | Extra]

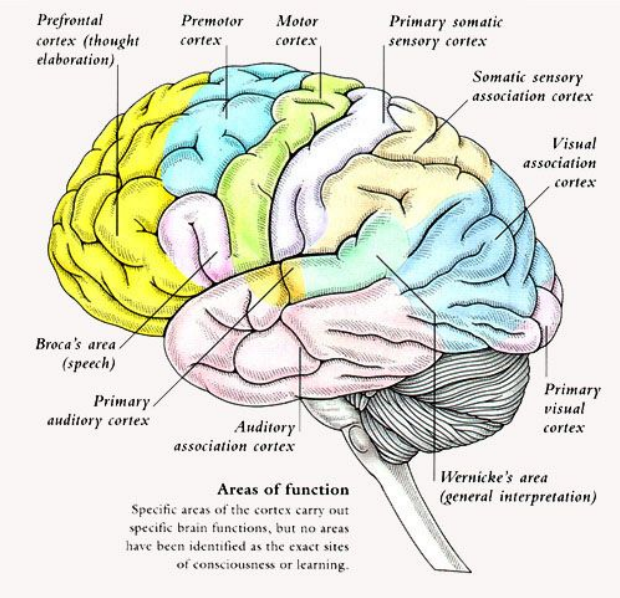
[Editing file]

Basic review:

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, is refined during the general and neurological examinations, and is reassessed after any relevant diagnostic studies are completed. The most important thing to learn from this lecture is how to localize the lesion. e.g. frontal, parietal etc

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

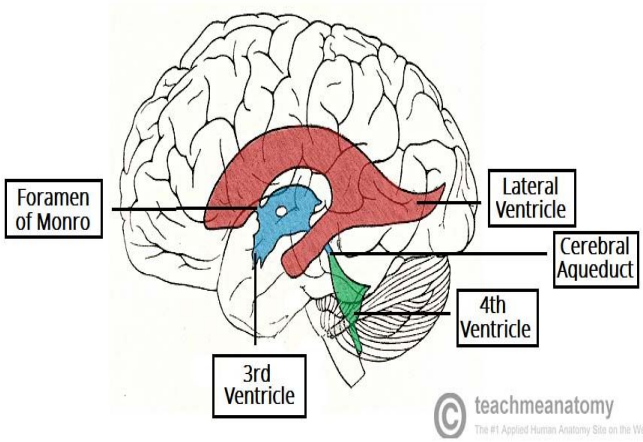
1. What is the necessary minimal amount of neuroanatomy that must be damaged in order to produce the patient's symptoms/signs?
    a. A patient with monocular blindness must have a lesion on the same side somewhere between the cornea and optic chiasm.
    b. A patient with an absent tendon reflex must 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?

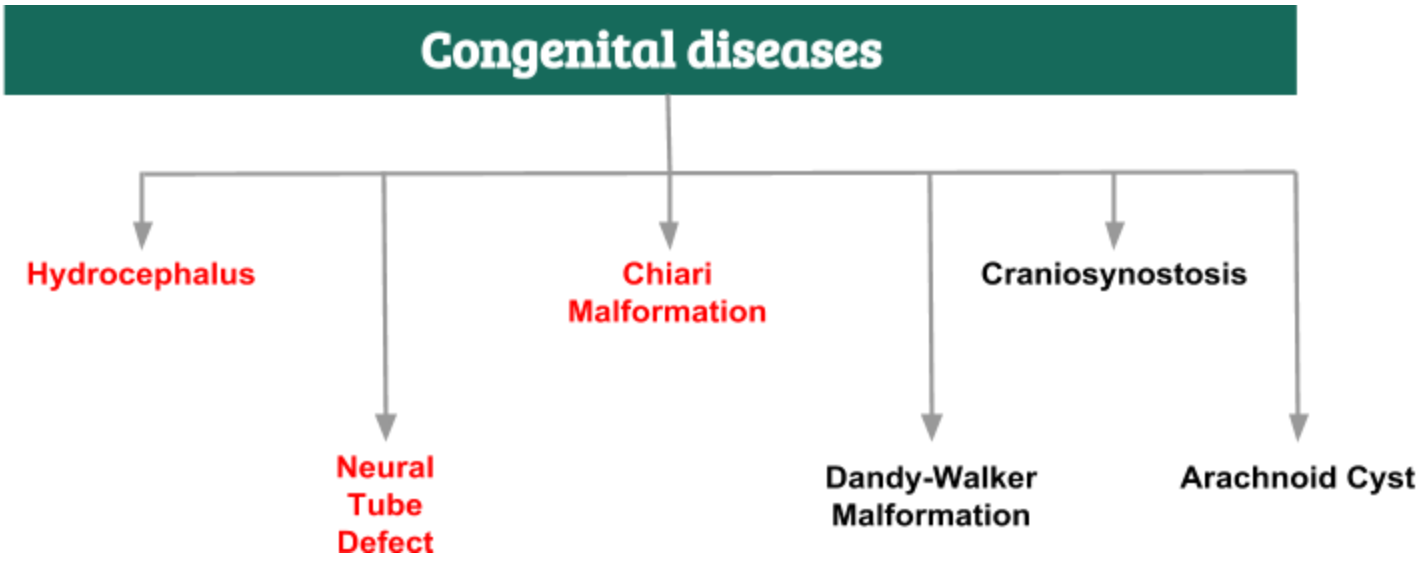


If you want to know more about how to localize a lesion, you can check this file from 431. Highly recommend it

★ 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.





اللي بالأحمر مهم، واللي بالأسود بس أبيكم تعرفون ال Definition and the pathology!

Approaching Neurological Symptoms

Headache or facial pain:

In taking history, not every pain in the head considered headache, it could be facial or dental problems. So make sure is it headache or something else!

MCQ: Compare between the two types of headache.

Primary headache (not serious)	**Secondary headache** (serious)

• Are benign, recurrent headaches not caused by underlying disease **nor mass** or structural problems
• Examples: Migraines, **tension headaches** (**MCQ**:most common type of headache! وهو زي الصداع الي يجيكم آخر السنة وتتعبون من التفكير بوش سويتوا بحياتكم), cluster headaches.
• Very common, You don't need to do any further investigations.
• More common in females.
• It's **classical**:
  ○ Family history, half of the face, episodic, more common at night, with stress, photophobia, nausea, intermittent.
	• Are caused by an underlying disease : **masses**, tumor, hematoma, vascular disease, حاجة بتكبر بالدماغ بتسبب هذا الصداع
	• International Headache Society (IHS) classification
	• Certain "**Red flags**" indicate a secondary headache **MCQ**: Which of the following criterias is low or high risk for headache? (**SSNOOP**) is a mnemonic to remember the red flags for identifying a secondary headache
	  ○ Systemic symptoms (fever or weight loss)
	  ○ Systemic disease (HIV infection, malignancy) In HIV, it's because of the increased risk of lymphomas and infections, in CNS, lymphomas are the ones we're afraid of.
	  ○ Neurologic symptoms or signs
	  ○ Onset sudden (thunderclap headache)
	  ○ Onset after age 40 years
	  ○ Previous 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 ، مانتكلم عن الأطفال لأن المفروض والطبيعي إنه مايجيهم صداع نتكلم عن الشباب
- Features typical of primary headache
- History of similar headache
- No abnormal findings on neurologic exam
- No concerning change in normal headache pattern
- No high-risk comorbid conditions (for example, HIV) Why ? B.c High risk of neoplasms and CNS infection.
- No new concerning history or physical examination findings

ممكن تجيك بنت شابه ٢٥ سنة أول مره يجيها صداع فايش تسوي ؟ تخليها تراجعك بعد فترة بدون ما تطلب لها CT لان تتوقع انه عندها الشقيقة بس ذي أول مره تجيها عشان كذا سويت لها فولو اب to make sure she is fine

Differential Diagnosis of headache: (Please do not be specific by giving a differential diagnosis, ALWAYS be systematic! امشوا على اللسته هذي بأي إختبار بحياتكم ولما تفكرون)

- Vascular : aneurysm, AVM , hemorrhage
- Inflammatory/Infectious : MS / meningitis, fungal or viral infection, abscess.
- Autoimmune/Allergic/Anatomic
- Neoplastic two types: primary and secondary tumors(metastasis).
    - Most common primary tumors: مابغاكم تحفظون الأسماء بس اعرفوهم لأنهم الأكثر انتشاراً عندنا
        1. Meningioma (which is a **good** tumor, Arise from the meninges, very benign)
        2. Glioblastoma (very **bad** it's the WORST, survival 1-2 year)
        3. Pituitary adenoma (common, Cushing diseases, acromegaly)
        4. Schwannomas which happens with 8th cranial nerve (present with hearing loss, vertigo)
- 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	

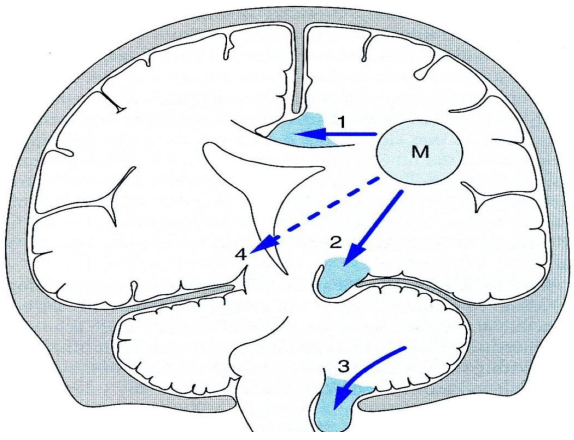
Differential Diagnosis of CNS space-occupying:

Neoplastic → Vascular → Congenital → Inflammatory → Infectious.

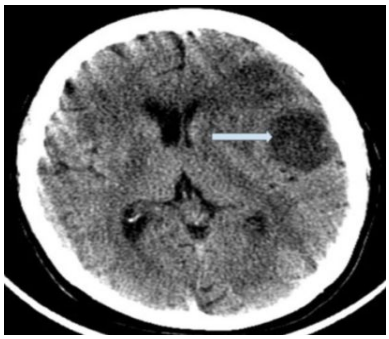
لما يجيكم المريض، دائماً حطوا ببالكم هالثلاث نقاط:

1- **Localization** of the pathology
2-What causes it? Most likely **diagnosis (pathology)**
3-Severity, how bad it is

In picture:
1- Local compression
2- Mass effect & herniation
3- 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. **MCQ**: 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.



CT Scan: Left brain lesion

هنا مثلاً هذي ال lesion إيش ممكن تكون وإيش ممكن تسبب؟

نمشي زي ماعلمتكم فوق بالترتيب: it could be vascular, inflammatory etc..

إيش تسبب: بما إنها في

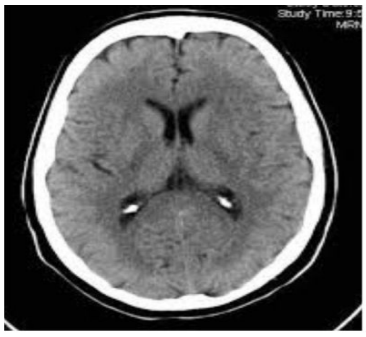
frontal lobe specifically in front of the central sulcus, so contralateral weakness and if it's the dominant hemisphere maybe it'll cause aphasia too.

This picture might come in the OSCE station, prepare yourselves ;p

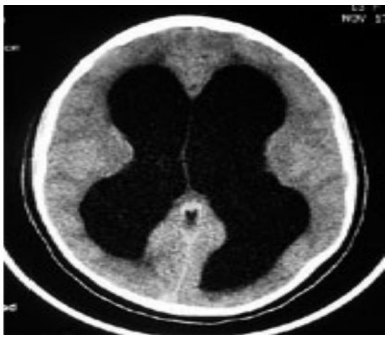
Common Congenital Neurosurgical diseases

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 elderly can have ventriculomegaly but it's due to brain shrinking(atrophy)! So, every active hydrocephalus is ventriculomegaly, but not the opposite.

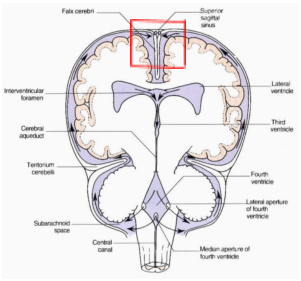
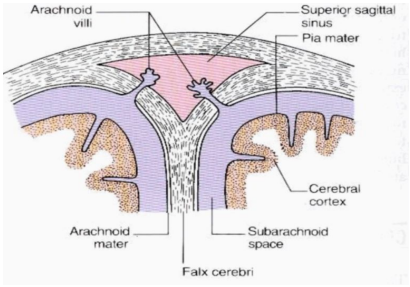
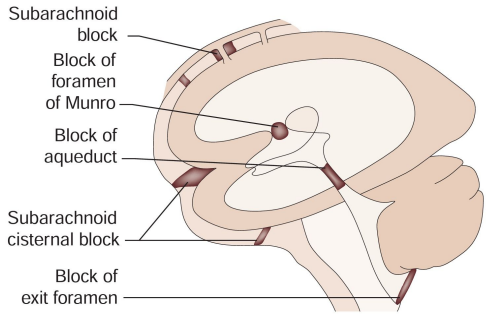


Normal



Hydrocephalus (in the lateral ventricles)

Hydrocephalus can be classified by pathology or by location

Physiology of CSF	Causes of Hydrocephalus (pathophysiology)IMP
• 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.   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 (MCQ).	• Obstruction of CSF flow (non-communicating), can be caused by tumors, malformation. • Under absorption of CSF. CSF production = CSF reabsorption = ~500 mL/d in normal adults Normal CSF volume ~150 mL (50% spinal, 50% intracranial 25 mL intraventricular, 50 mL subarachnoid) • Overproduction of CSF while we have increasing in size  Fig. 24.17 Hydrocephalus: sites of cerebrospinal fluid (CSF) blockage.

Types of Hydrocephalus: (obstructive and non obstructive) it's better for classification.

MCQ: which one of the following causes obstructive or nonobstructive hydrocephalus?

Communicating (Non obstructive) Usually **chronic** presentation, like post meningitis	Unimpaired connection of CSF pathway from lateral ventricle to subarachnoid space **Causes:** • 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)
Usually **acute** presentation
more common
You need to know at what level is the obstruction and what ducts are going to be dilated! Comes in the OSCE as a picture
	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

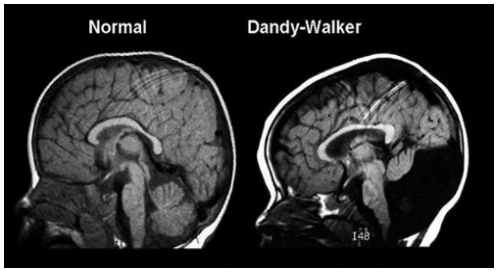
Etiology:

MCQ:

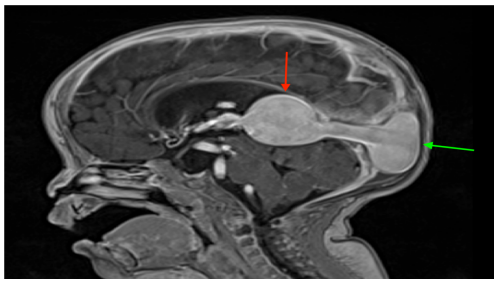
- Which one of the following isn't a congenital cause of hydrocephalus? So you better Memorize them.
- What's the most common cause of congenital hydrocephalus? Aqueductal anomalies\stenosis (obstructive)

Congenital	• Aqueductal anomalies(stenosis): (**most common cause**): Infants will come after 2 weeks of birth with vomiting, sleepy, crying, increased head circumference very rapidly and CT will show obstruction (stenosis) at the level of the aqueduct → non communicating. ○ 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) MCQ • Dandy Walker malformation (more details below) • Chiari II malformation • Myelomeningocele. • Vein of Galen aneurysms (more details below) • 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 **MCQ** • Post-meningitis **non-obstructive**. very common, hydrocephalus is the most common complication. Happens due to scarring after inflammation. If there is abscess it is mass obstruction but usually it isn't. • Tumors (usually obstructive) • 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 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**: Incoordination, ataxia, nystagmus

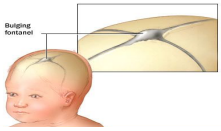
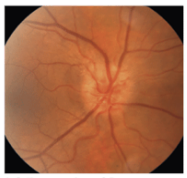
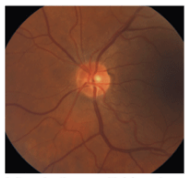


★ Vein of Galen aneurysms: very rare(obstructive hydrocephalus)

لما يشوفونها بالبداية بيحسبونها ورم لكن لما يمشون بباقي التشخيصات المحتملة على نفس الترتيب اللي سويناه بيعرفون انها ممكن تكون هذا المرض ويتأكدون من الشكل انها سموث مو نفس الورم

- 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.

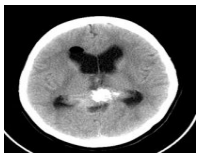
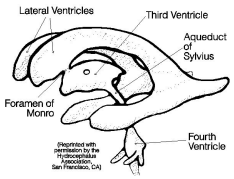
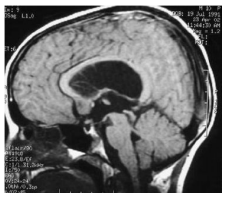
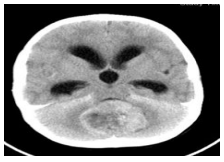
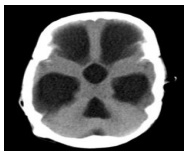
Clinical features:

Infants & young children MCQ	Juvenile & adult: (↑ ICP)
MCQ: the following are clinical features in infants except? 1. Increasing head circumference. 2. Irritability, lethargy, poor feeding, and vomiting. 3. Bulging anterior fontanelle. >> 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. Episodic bradycardia and apnea. very high ICP leads to pressure on brainstem. 	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 question asks what's the next step in this case (picture), LP or brain imaging? choose brain imaging  

Investigations:

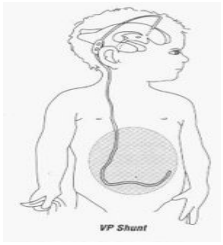
CT or MRI:

The pattern of ventricular enlargement can help delineate the cause:

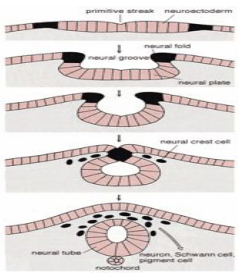
Lateral ventricles dilatation	Obstructive hydrocephalus caused by 3rd ventricle tumor. (absent 3rd and 4th ventricles)	
Lateral & 3rd ventricle dilatation	→ **Normal 4th ventricle:** suggests aqueduct stenosis	 
	→ **Deviated or absent 4th ventricle:** suggests posterior fossa tumor	
Generalized dilatation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

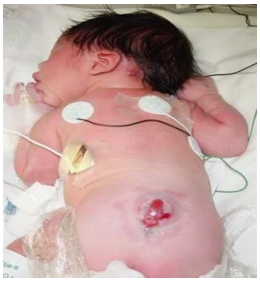
Treatment:

1- Communicating : Medical or surgical.
2- Obstructive: : SURGICAL TREATMENT. **VP shunt**



2- Neural Tube Defect (NTD):



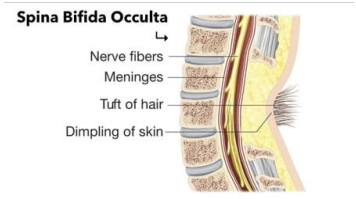
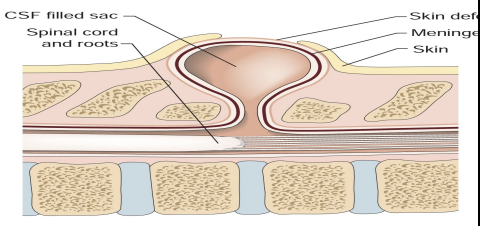
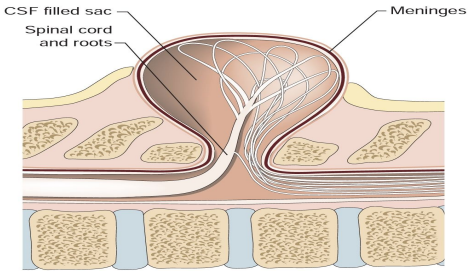


- Spinal Dysraphism is **Failure of closure of <u>posterior</u> neural arch**.

- Might happen between 3rd to 8th week (**MCQ**). The earlier it happens the more severe the symptoms.

Two major types: Open or Closed

Types of myelodysplasia: MCQ+OSCE الصور الي بالجدول ممكن تجي بالأوسكي ونسألكم عن الأعراض وطرق العلاج

Spina bifida occulta (closed) **MCQ**	**Meningocele** (closed) protrusion of meninges	**Meningomyelocele** (open) protrusion of meninges and spinal cord
• 5-10% of population , the commonest • not clinically significant • Asymptomatic usually, may be tuft of hair, dimple sinus or port wine stain • high incidence of underlying defect • no treatment required Reassure patient  بسيطة جداً ويمكن خمسه منكم الحين عندهم هذا الشيء ولا يدرون عنه من اسمها occulta خفيفة بسيطة ما تبان	• Cystic CSF-filled cavity **lined by meninges** prone to infection. • **no neural tissue** • communicates with spinal canal • look for other cong. Anomalies • Urgent excision if CSF leak (If ruptures), otherwise deferred • seldom any neurological deficit. (LL weakness-urinary retention) • Diagnosed by U/S or MRI.+look for signs of infection. • sac containing CSF only compared to 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 • 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. لما يكلمونا الطوارئ يقولون وحده ولدت ماكانت تتابع معنا والبيبي عنده spina bifida ايش أول سؤال نسألهم ؟ هل هي مفتوحة ولا مقفله فيه فلويد يطلع او لا ؟ لان لو مفتوحة الان الان لازم نروح نقفلها لأنه عرضة للانفكشن Treatment: close it! 

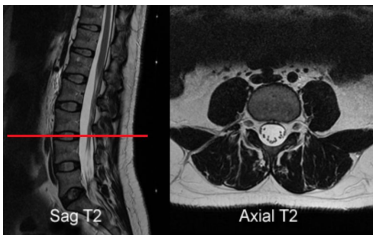
Questions in history taking in Spina Bifida :
Ask about the motor function (nerves are protruding), signs of infection (meningitis), urinary retention, pregnancy and antenatal care.

Incidence:

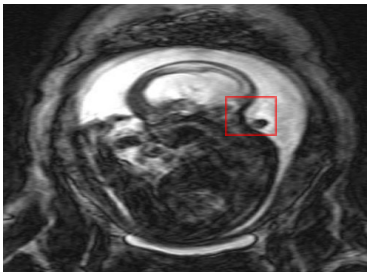
- In low-socioeconomic
- 80% in lumbosacral region (Occasionally in the head)
- 2/1000 birth
    - Risk increase to 5% if a sibling is affected. More common in SA
    - Teratogens ex: antiepileptics الصيدلي، بالغالب كمان هذا من الأسباب اللي ترفع النسبة عندنا غير انو مافي تخطيط للحمل عندنا يقدر يصرف دواء 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), 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

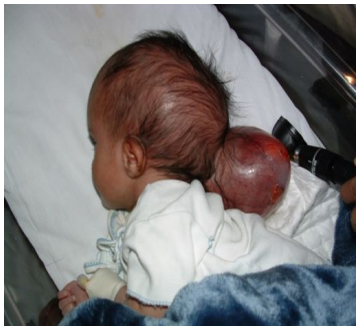


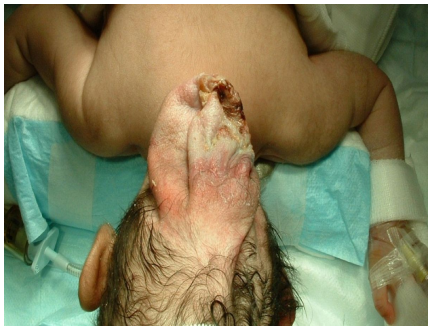
Antenatal diagnosis: Red square in the picture: is not cute! It's BAD, will grow after birth (encephalocele)



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

3- Other congenital: Anomalies:





★ **Encephalocele .** Not imp

- Usually occipital
- may contain occipital lobe, or cerebellum
- often associated with hydrocephalus
- Immediate treatment if ruptured
- outcome depends upon contents. Usually they don't survive

★ **Chiari Malformation:** مهم نعرف الأنواع

It is the herniation of posterior fossa's content below the foramen magnum.

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 - choking (due to compression to brainstem) - ataxia - nystagmus - hydrocephalus.
Type II children type A Triad! MCQ	More serious than type 1, Extension of both cerebellar and brainstem tissue into the foramen magnum, Associated with: Myelomeningocele, Hydrocephalus, Herniation
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.

*Both of below are Not in dr's slides but he said they're IMP to know and may come in MCQs

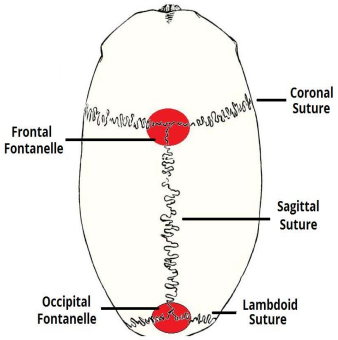
★ Arachnoid cyst:

- 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: know the cause + Types

Premature closure or absence of a cranial suture. The effect depends on the site and number of sutures involved. The brain has reached 85% of its adult size by the age of 2 years but continues to grow slowly after this time.

- 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. Many cases of plagiocephaly are due to head moulding, when the baby lies on its back to sleep. This usually resolves when the child starts to sit and walk.



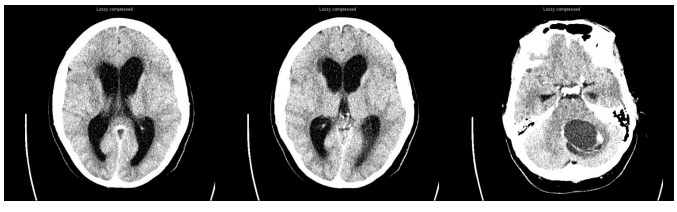
Sometimes, more than one suture can be affected. This can be syndromal (e.g. Crouzon's or Apert's syndrome).These syndromes are associated with characteristic craniofacial deformities. Craniosynostosis may also lead to a reduction in cranial volume, causing raised ICP. Surgery can be undertaken to remodel the skull into a more acceptable shape or to increase the cranial volume.

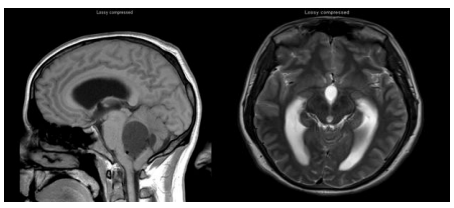
(Pic) : In neonates, the incompletely fused suture joints give rise to membranous gaps between the bones, known as fontanelles. The two major fontanelles are the frontal fontanelle (located at the junction of the **coronal and sagittal sutures**) and the occipital fontanelle (located at the junction of the sagittal and lambdoid sutures)

Case from Dr.:

4 y/o, No past medical history, Worsening headache, occipital area, for 7 weeks,His headache worsened last night. Associated with: Dizziness,loss of balance,N/V twice over the last 3 weeks.

- **red flag**: New headache,localized, worsened last night is a sign of high ICP specifically hydrocephalus N/V also sign of high ICP.
- **localization** in brain (cerebellum) because of symptoms , **Pathology** in CNS (not vascular because it is worsening that means something is getting bigger in size either tumor or abscess but it is not abscess because there is no fever) , **severity** (there are alot of Red flags).
- **Ddx**: Hydrocephalus caused by: neoplastic. Vascular (bleeding unlikely), congenital, meningitis is unlikely why? no fever, duration 7 weeks!
- **CT** scan: Mass in posterior fossa causing obstruction at level of fourth ventricle (lateral ventricle and third are dilated but not the fourth)
- **MRI**: more details





- Since the mass is in the posterior fossa Signs may be caused by focal compression of the cerebellum which are: VERY IMP. **OSCE**

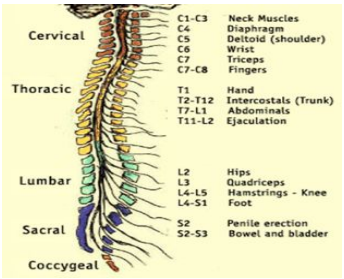
• Gait ataxia • Truncal ataxia • Limb ataxia :Finger-nose and heel-knee-shin, intention tremor, dysmetria (past pointing), dysrhythmia.	• Cerebellar dysarthria • Hypotonia • Rapid alternating movements (dysdiadochokinesia) • Tremor • Nystagmus gaze-evoked, horizontal drift followed by a fast correction

At your level I expect you to know:
1- How to Examine & Localize lesions
2- Clinical presentation associated with the lesion
3- The Most common Pathology.

In the exam, I might give you symptoms and examination findings and ask where is the disease ?
Or the opposite:
For example, There is a mass compressing frontal lobe what are you going to ask and examine?

So make sure you study the function of these areas very well:

- Frontal/motor cortex (Dominant vs nondominant hemispheres)
- Parietal/sensory cortex.
- Cerebellum.
- Spinal cord (Cervical/Thoracic/lumbar).
- Cervical roots.
- Lumbar roots.



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?

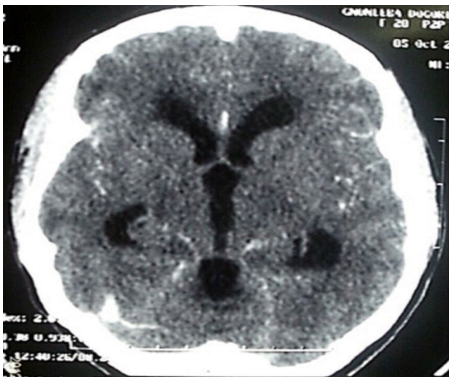
< 95% survival for the first 2 years, compared with 25% in patients not undergoing surgical procedures

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

Folic acid

MCQS

1) A 20 years old female patient with no medical history was referred to our department for 1 month progressive onset of intense headache. On clinical examination, the patient was complaining of worsening headache associated with vomiting. However, no neurologic deficit was noticed. The CT scan revealed an important dilatation of lateral, third and fourth ventricle. What's the first management option you should do?



A. Ventriculoperitoneal shunt
B. Reassure patient
C. Order an MRI scan
D. Observe

2) Manifestations of uncal herniation include which of the following?

A. Sensory deficits
B. Dilated pupil
C. Quadrantanopia
D. Monocular blindness

3) The investigation of choice in increased ICP is:

A. Skull x-ray
B. MRI
C. Lumbar puncture
D. CT scan

Answers:

1- A
2- B
3- D