







Objectives:

- Introduction to Neurosurgery.
- Approaching neurological

neurosurgical diseases

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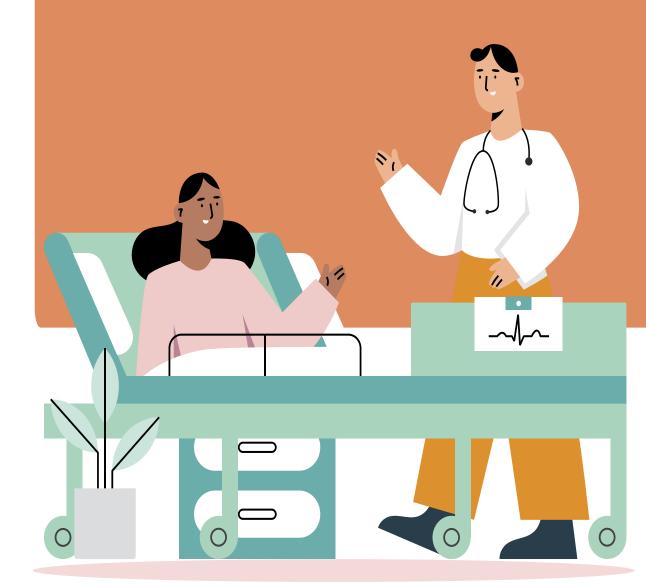
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Color Index:



- symptoms.
- Congenital diseases, including:
 - Hydrocephalus.
 - Neural tube defect.
 - Chiari Malformation.
 - Dandy-WalkerMalformation.
 - Craniosynostosis.
 - Arachnoid Cyst.











Basic review: From 435, for your knowledge.

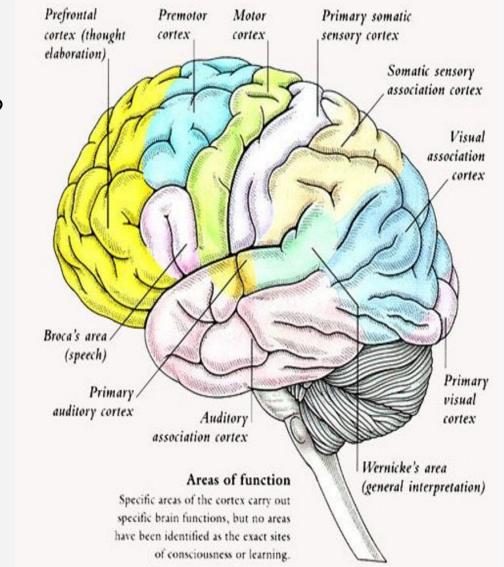
The Localization of a lesion requires an understanding of the anatomy and physiology of the nervous system, its blood supply, and the disease processes that affect it. The process of localization begins during history taking, and is refined during the general and neurological examinations, and reassessed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





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 <u>check this file</u> from 431. Highly recommend it

\star 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**.

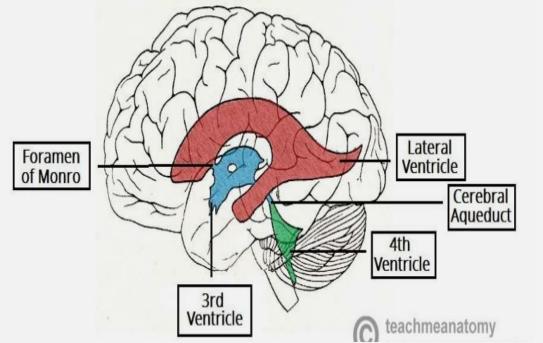
■ The third ventricle is situated in between the right and the left thalamus.

■ 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.



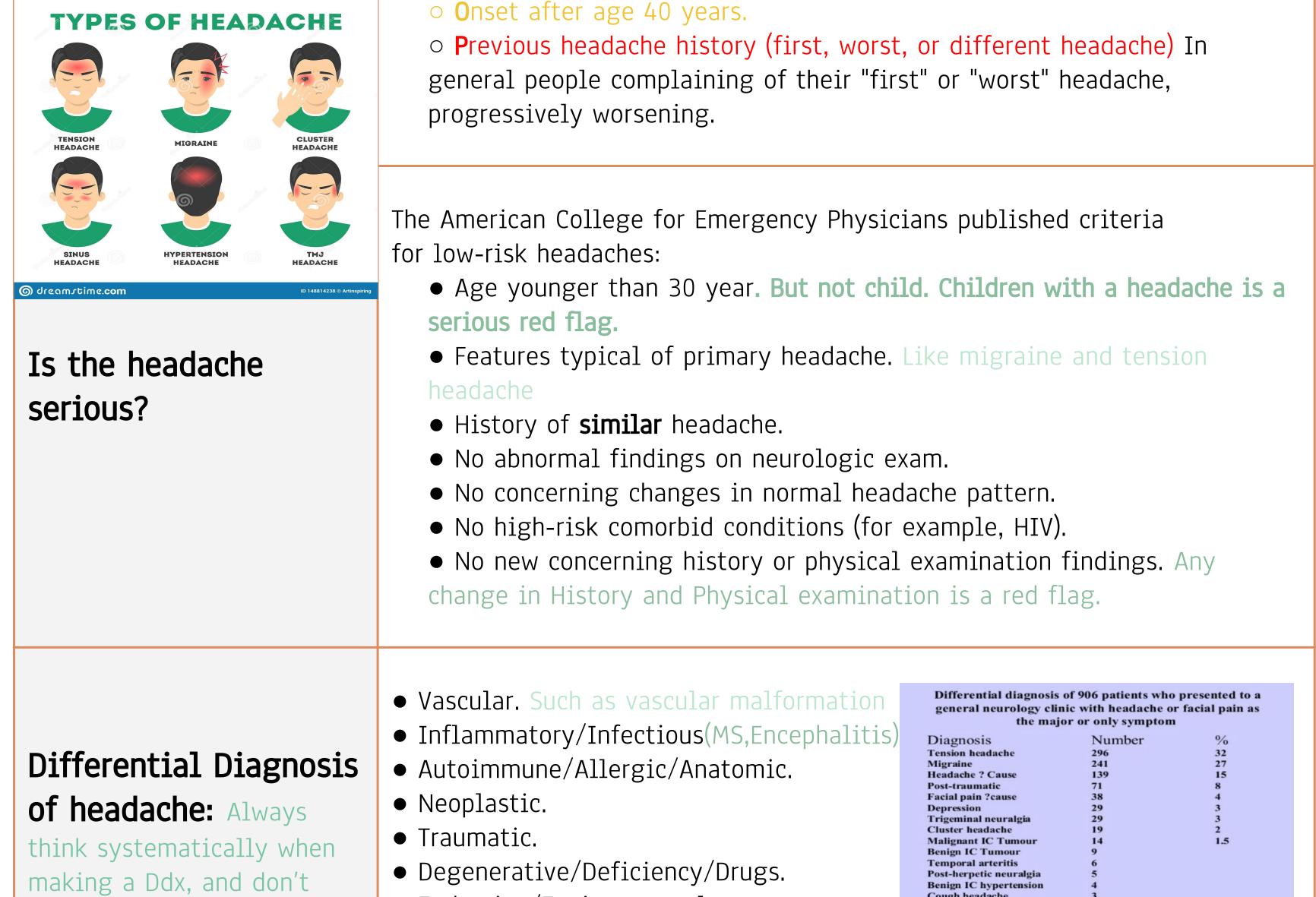


Approaching Neurological Symptoms

Headache or facial pain (Not every pain in the head is headache, it could be

facial or dental problem).

Primary headache (not serious) don't need imaging	 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.
Secondary headache (serious) need imaging	 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 Systemic symptoms (fever or weight loss) Systemic disease (HIV infection, malignancy). Neurologic symptoms or signs. Onset: sudden (thunderclap headache).



jump to conclusions to diagnose immediately!

- Endocrine/Environmental.
- Idiopathic/Intoxication/Iatrogenic.
- Congenital.
- Metabolic.

 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:

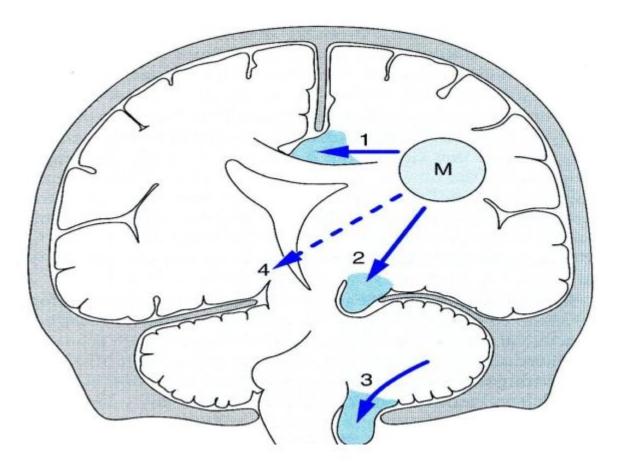
Neoplastic \rightarrow Vascular \rightarrow Congenital \rightarrow Inflammatory \rightarrow Infectious.

حطوا ببالكم هالثلاث نقاط الدائما ، لما يجيكم المريض :

- 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!



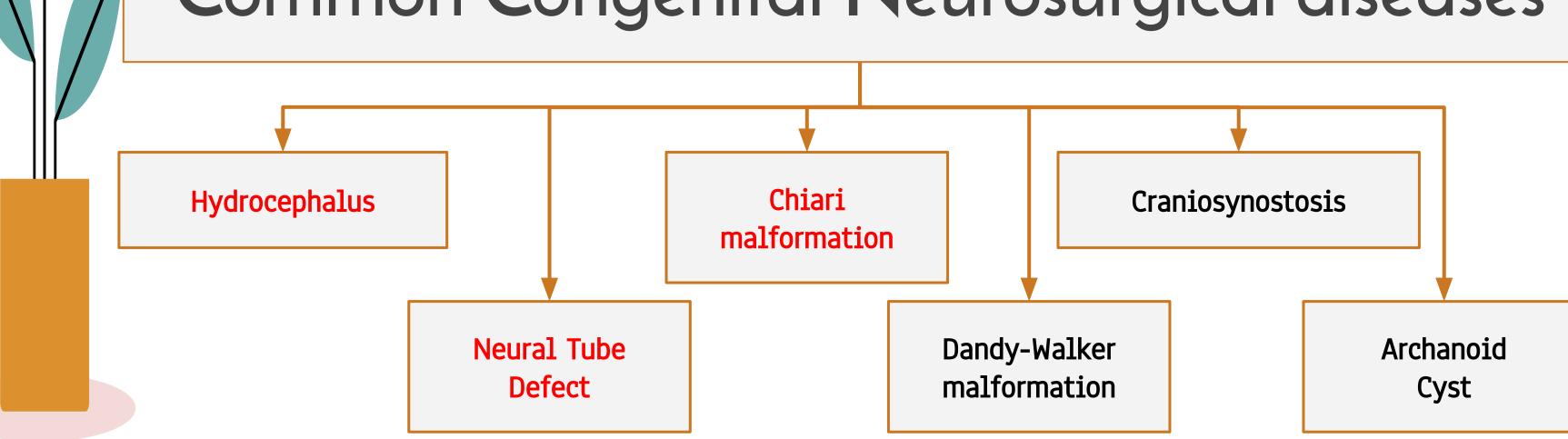
Local compression	Mass effect & Herniation	High ICP
Functional areas: -Motor cortex → weaknessSensory → numbness or seizureCerebellar → tremor, dysarthria, ataxia or even truncal ataxiaPituitary adenoma → visual loss.	 When it starts to increase in size, the brain tissue will shift to the other side. 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? Contralateral weakness (right) hemiparesis and hemiplegia. Compression of midbrain cerebral peduncles. 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! ! يما طول أجي المستشفى dilated pupil dilated pupil للمستشفى واحنا طفشانين بالأوسكي نحب نسألهم ، واحنا طفشانين بالأوسكي نحب نسألهم ، واحنا طفشانين بالأوسكي نحب interesting and the syndrome (notch). 	 Headache. Nausea. Vomiting. High BP. Impaired level of consciousness. Papilledema. You need to remember them by heart.

which shows a left sided brain mass

منا مثلا هذي ال lesion إيش ممكن تكون؟

نمشي زي ماعلمتكم فوق بالترتيب إيش تسبب: بما إنها في : it could be vascular, inflammatory etc. 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

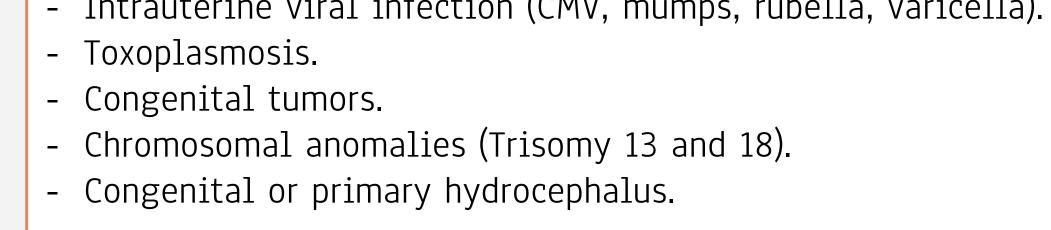


1- Hydrocephalus could be congenital or acquired

- Hydrocephalus: is an increase in the CSF volume, associated with increased ventricular size.
- Not the same as Ventriculomegaly (ex vacuo dilatation) which happens as result of Brain atrophy (Ex:Dementia) . Not all hydrocephalus can cause ICP

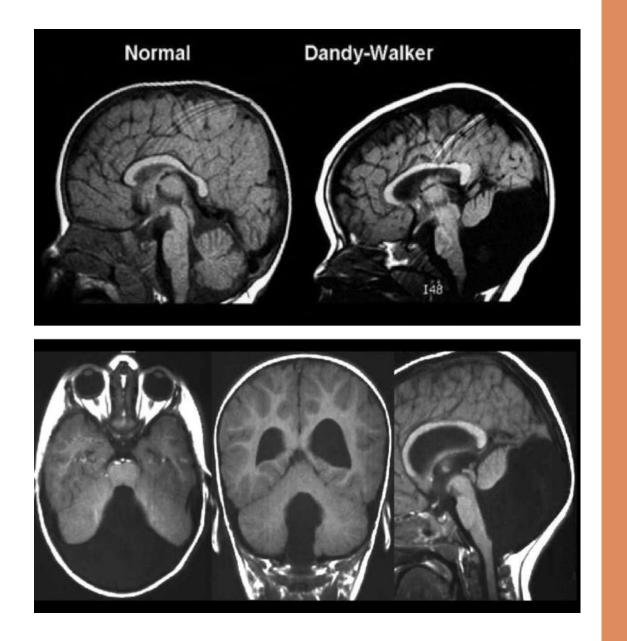
_	Total volume of CSF in the ventricles varies from 5-15 ml in neonates to 150 ml in adults. Produced mainly by choroid plexus.	
- Physiology of CSF -	Rate of production is 0.3-0.4 ml/minute. Absorption occurs via arachnoid villi obstruction at this level or = superior sagittal sinus(the area where arachnoid villi located) because of fibrosis or previous inflammation it will cause under absorption hydrocephalus imp (MCQ)	
Pathophysiology	 Obstruction of CSF flow. Most common and urgent one Under absorption of CSF. CSF production = CSF reabsorption Overproduction of CSF (rarely, due to CSF overproduction, e.g., because of a choroid plexus papilloma) 	
Types of Hydrocephalus:		
Communicating	 Overproduction or under absorption of CSF All ventricles are dilated No obstruction in the nathway of CSE within the ventricles (the ventricles can 	

(NOT ODSTRUCTIVE)	communicate with each other)	
<section-header></section-header>	 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. You should read about csf pathway production could be mcq qustion 	
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. Chiari II malformation. Wein of Galen aneurysms. Intrautering viral infection (CMV, mumps, rubella, varicella) 	

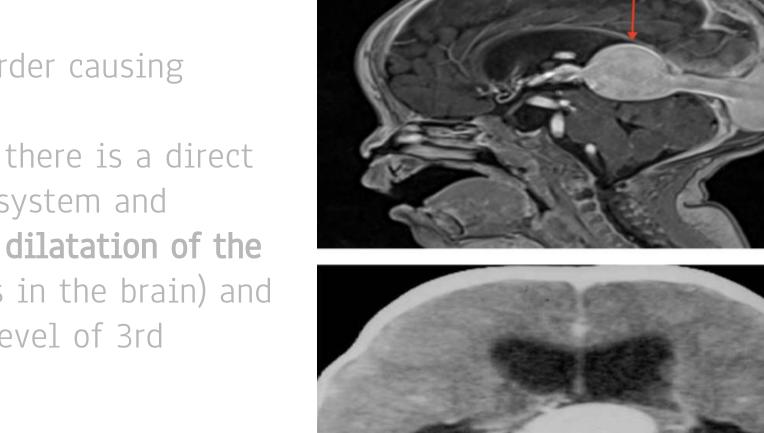


Acquired	 Germinal plate hemorrhage: in premature babies <1500 gm (30%-40%) (<u>immature</u> blood vessel wall) can be acute or chronic: if acute it is obstructive there is a mass, if chronic (delay) due to problem in absorption it can be obstructive or communicating. Post-meningitis non-obstructive. 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(Cerebellar and hydrocephalus symptoms):
 Incoordination, ataxia, nystagmus.



- Vein of Galen aneurysms: (very rare disorder causing obstructive hydrocephalus)
 - 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 of Hydrocephalus:

	-As the hydrocephalus worsens, the eyes
<section-header><section-header></section-header></section-header>	 may become downcast (sunsetting). The child may be floppy and develop apnoeic spells and episodes of bradycardia. After closure of the fontanelles children have symptoms of raised ICP (head-ache, vomiting and drowsiness). The eyes may develop a squint secondary to VI cranial nerve palsy. Irritability, lethargy, poor feeding, and vomiting. Bulging anterior fontanelle. (picture) Widened cranial sutures. MacEwen's (cracked-pot) sign with cranial percussion.(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. Scalp vein dilation (collateral venous drainage). Sunset sign (downward deviation of the eyes). Episodic bradycardia and apnea(Due to the compression on BrainStem because of increase IP which led to herniation)
Juvenile & adult: (high ICP)	 Headaches. Nausea. Vomiting. Decreased level of consciousness. Focal neurological deficit (rare). Papilledema. Scenario: patient has meningitis, headache, v/n. You

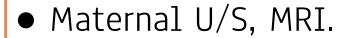
	need to do lumbar puncture, but before that you have to rule out high ICP. How? By fundoscopy (papilledema) or brain imaging (CT) if the question asks what's the next step in this case (picture), LP or brain imaging? choose brain imaging		
CT or MRI: ⊤	Investigations: CT or MRI: The pattern of ventricular enlargement can help delineate the cause:		
Lateral ventricles dilatation	Obstructive hydrocephalus caused by 3rd ventricle tumor. (absent 3rd and 4th ventricles)		
Lateral & 3rd	→ Normal 4th ventricle: Suggests aqueduct stenosis.		
ventricle dilatation	\rightarrow Deviated or absent 4th ventricle: Suggests posterior fossa tumor.		
Generalized dilatation	Suggests a communicating hydrocephalus.		



2-Neural Tube Defect (NTD)

- Spinal Dysraphism is Failure of closure of posterior neural arch (Happens between the 3rd to 8th week of gestation).
- Two major types: **Open** or **Close**.

Types of myelodysplasia		
Spina bifida occulta (closed)	 5-10% of population. Common incidental finding not clinically significant tuft of hair, dimple sinus or port wine stain High incidence of underlying defect. No treatment required. Do you need to do MRI in spina bifida occulta? Ans: No If there are risk factor or sign in clinical exam you should do MRI to rule out any associated spinal neural abnormalities Common scenario: young patient in the gym complaining from back pain we do x-ray we only see sign of bony deformity but otherwise the patient is intact they have neural defect and they go to spina bifida occulta no treatment is needed If you are 60 or 70 student in class 3 of you you have spina bifida occulta and you don't no about it 	
Meningocele (closed) protrusion of meninges	 Cystic CSF-filled cavity lined by meninges prone to infection. No neural tissue unlike meningomyelocele. Communicates with spinal canal. Look for other congenital anomalies. Seldom any neurological deficit.t. (LL weakness urinary retention) Diagnosed by U/S or MRI.+look for signs of infection. Usually it is mild because neural tissue not involved -Urgent excision if there's CSF leak(If it ruptures) otherwise no need to remove it 	Wyelomeningocu
Meningomyelocele (open) protrusion of meninges and spinal cord	 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 anal sphincter Gross hydrocephalus, multiple serious congenital anomalies. Diagnosed by U/S or MRI(becaues its usually associated with chiari -malformation. Is it open or closed ? if open risk of infection is high bc CSF come out and treatment is urgent you need to close it surgical If closed treatment not urgent but still you need to fix it Prognosis depends on the site of the lesion,The higher its the more worse prognosis the same concept is applied on meningocele 	
Incidence		
 80% in lumbosacral region (Occasionally in the head). 2/1000 birth: the reason they have NTD low folate or folate deficiency and antiepileptic medication Risk increase to 5% if a sibling is affected. Teratogens. How to prevent? By giving folic acid supplements before 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		



• Serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase.

• Contrast enhancing amniography.

• possibility of therapeutic abortion.

3- Other congenital anomalies:

Encephalocele

 \circ Usually occipital.

- \circ May contain occipital lobe, or cerebellum.
- \circ Often associated with hydrocephalus.
- \circ Immediate treatment if ruptured .
- \circ Outcome depends upon contents.



Chiari Malformation

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

In adults	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 Commonly In children	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

 \circ 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.
- \circ 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:

Premature closure of the Sagittal suture.

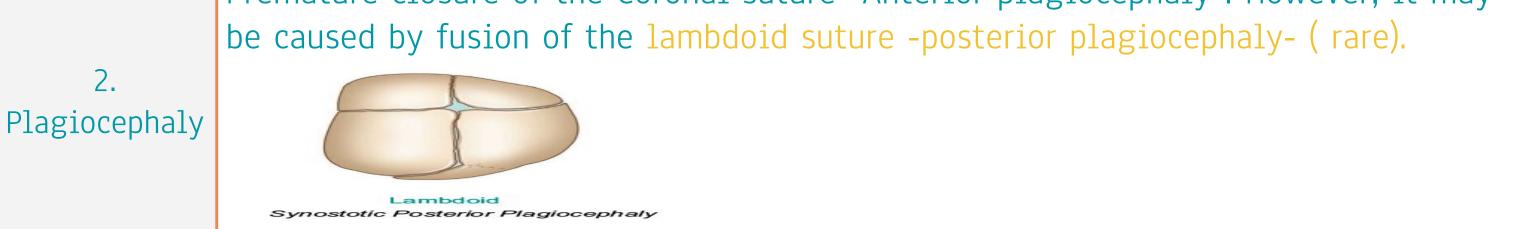
Growth normally occurs perpendicular to the suture line. In this case, the skull cannot widen, as there is no growth at the fused suture, which may be palpable as a ridge in the midline.

1. Scaphocephaly

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

No growth at fused suture



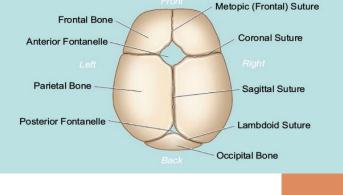
• Sometimes, more than one suture can be affected. This can be syndromal (e.g. Crouzon's or Apert's syndrome) which are associated with characteristic craniofacial deformities.



Crouzon's syndrome

Anterior Carlos Contractor a la contractor a





Surgical 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

0

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

Cont.Surgical Recall:

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?

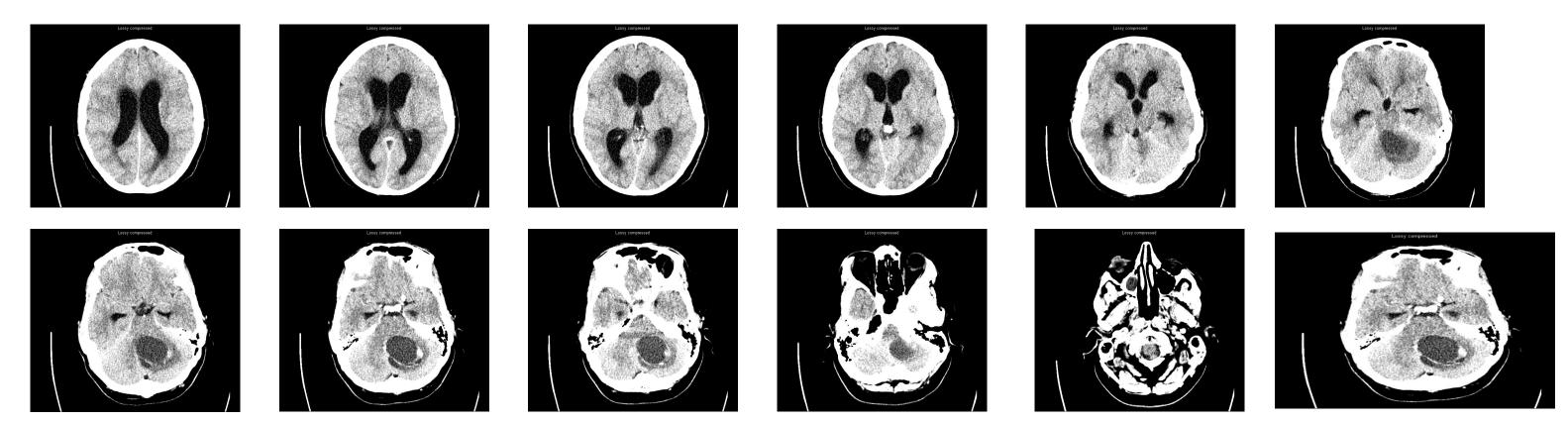
< 95% survival for the first 2 years, compared with 25% in patients not undergoing surgical procedures Which vitamin is thought to lower the rate of neural tube defects in utero? Folic acid



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).m



- 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.
- 2. 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.
- 3. 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.

4. 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.





Rapid alternating movements (dysdiadochokinesia).

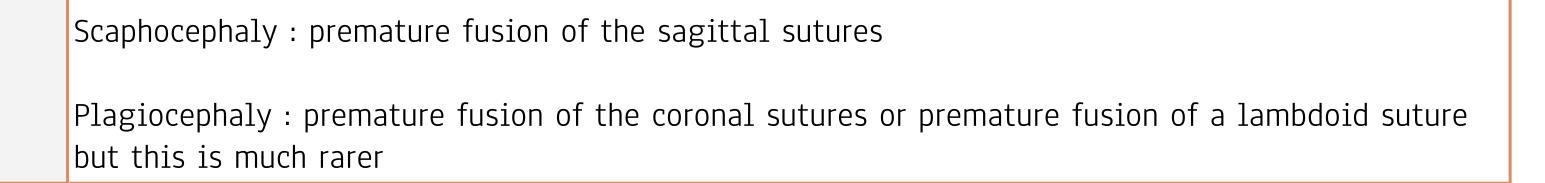


• Nystagmus gaze-evoked, horizontal drift followed by a fast correction.

Summary

Congenital diseases	Important notes
	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
1.Hydrocephalus	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
	1.Spina bifida occulta: .patient have (tuft of hair – dimple sinus – port wine stain), no treatment is required

	partene nave (tare of han aimpie sinds 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
2.Neural Tube defect	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
	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:
3.other congenital	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 \cdot
	3.Craniosynostosis:
	Premature closure or absence of a cranial suture



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