Congenital Neurosurgical Diseases

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Modified from Dr. Essam Elgamal

Learning Objectives

- **Introduction to Neurosurgery**
- **Approaching neurological symptoms**
- **Congenital diseases**
  - Hydrocephalus
  - Neural Tube Defect
  - Chiari Malformation
  - Dandy-Walker Malformation
  - Craniosynostosis
  - Arachnoid Cyst

Neurosciences

-Basic science

-Clinical

→Neurosurgery

→Neurology

→Psychiatry

Vascular
Oncology
Skull-Base
Spine
Pediatrics
Functional
Epilepsy
Peripheral Nerves
Critical Care
Trauma

? →

Headache or facial pain

Headache

- **Primary vs. secondary headaches**

Primary headaches

Are benign, recurrent headaches not caused by underlying disease or structural problems

Examples: Migraines, tension-type headaches, cluster headaches

Secondary headaches

Are caused by an underlying disease

→International Headache Society (IHS) classification

→Certain **"red flags"** indicate a secondary headache may be dangerous.

Headache

Vascular
Inflammatory/**I**nfectious
Neoplastic
Degenerative/**D**eficiency/**D**rugs
Idiopathic/**I**ntoxication/**I**atrogenic
Congenital
Autoimmune/**A**llergic/**A**natomic
Traumatic
Endocrine/**E**nvironmental
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	

http://www.slideshare.net/oxfordshireloc/headache-dr-paul-davies

Headache

Is the headache serious?

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

-Age younger than 30 years

-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)

-No new concerning history or physical examination findings

Headache

Red Flags:

-In general

People complaining of their "first" or "worst" headache

Progressively worsening

- The American Headache Society recommends using "SSNOOP", 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)

Onset after age 40 years

Previous headache history (first, worst, or different headache)

Differential Diagnosis of CNS *space-occupying*

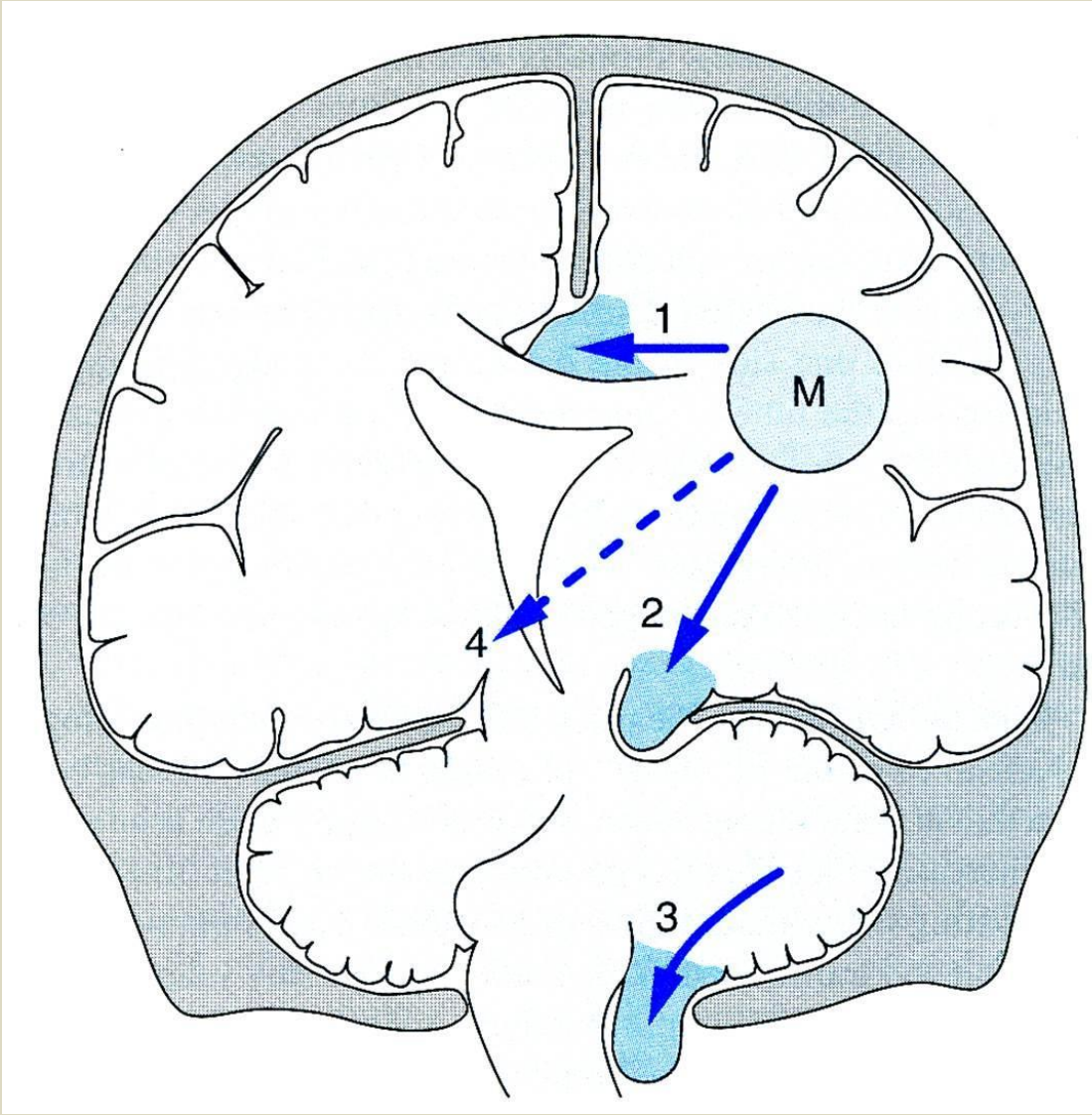
- **V**ascular
- **I**nflammatory/**I**nfectious
- **N**eoplastic
- **D**egenerative/**D**eficiency/**D**rugs
- **I**diopathic/**I**ntoxication/**I**atrogenic
- **C**ongenital
- **A**utoimmune/**A**llergic/**A**natomic
- **T**raumatic
- **E**ndocrine/**E**nvironmental
- **M**etabolic

Neoplastic
Vascular
Congenital
Inflammatory
Infectious

1- Local compression

2- Mass effect & Herniation

3- High ICP

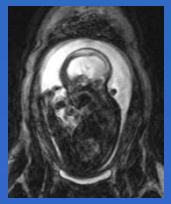


Hydrocephalus

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

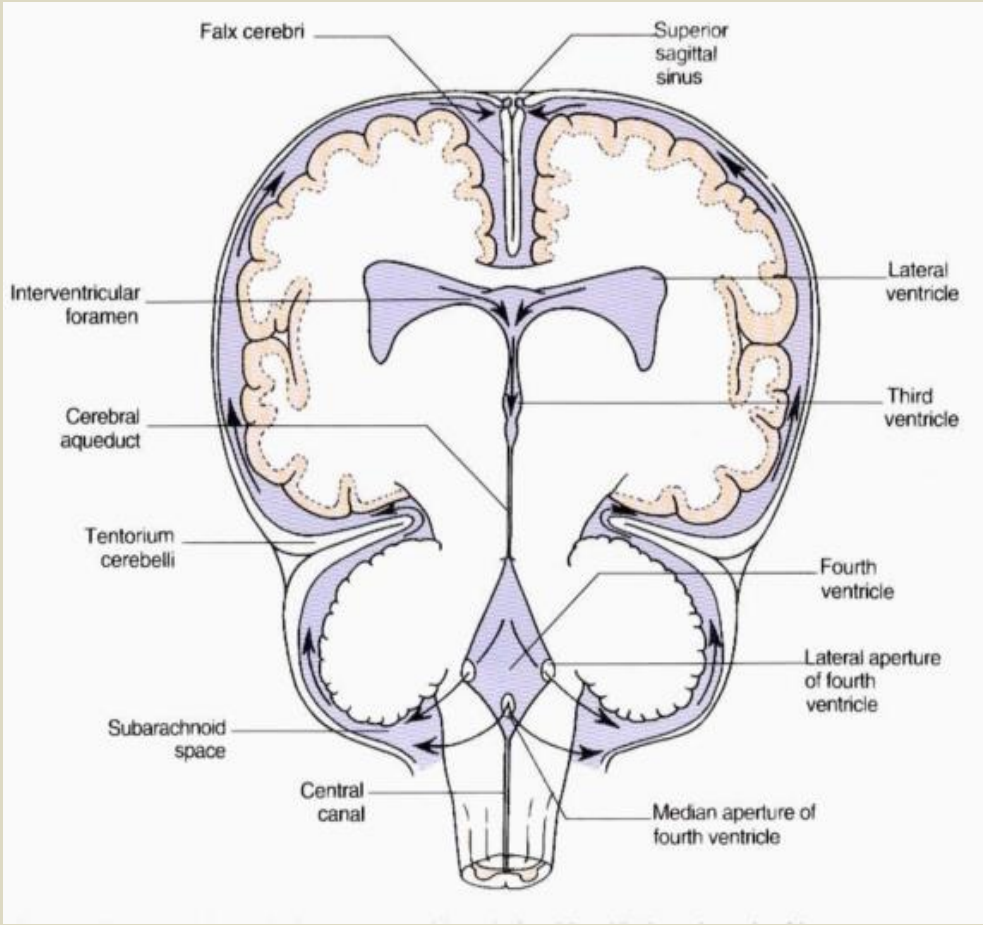
Causes of hydrocephalus

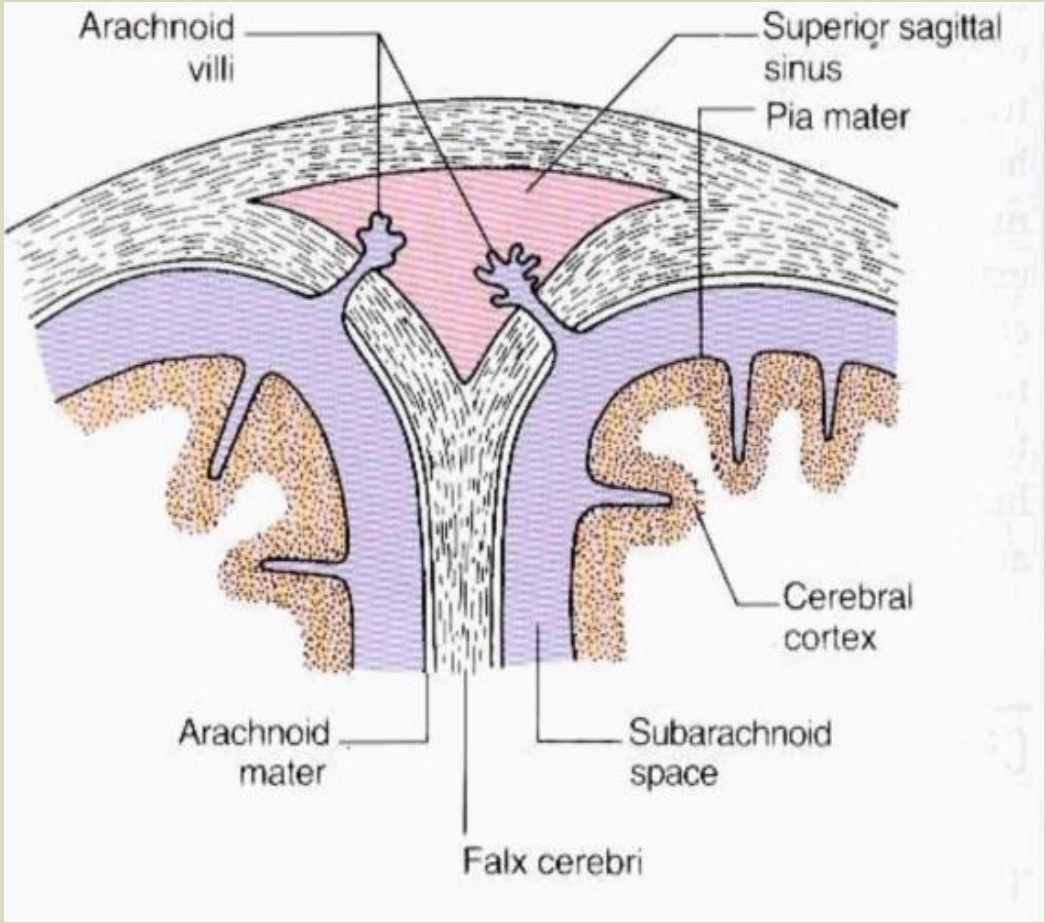
- Overproduction of CSF
- Obstruction of CSF flow
- Under absorption of CSF

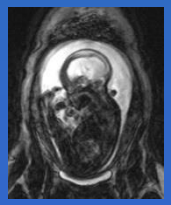


Physiology

- 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.




Arachnoid villi
Superior sagittal sinus
Pia mater
Cerebral cortex
Arachnoid mater
Subarachnoid space
Falx cerebri



Types of hydrocephalus

- **Communicating:**

  Overproduction or under absorption of CSF

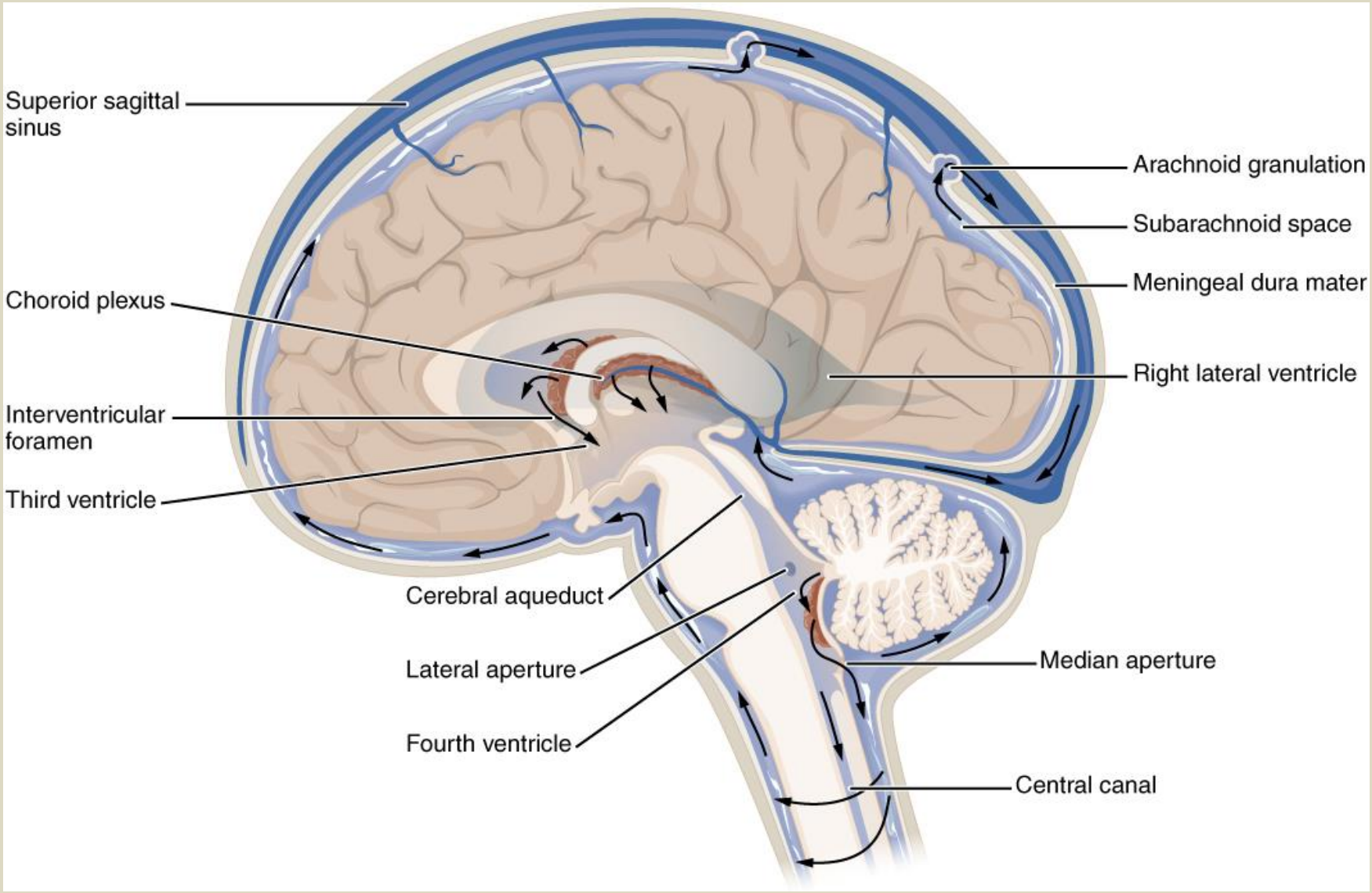
- **Non-Communicating:**

  blockage of the flow of CSF

→ Congenital, since birth

→ Acquired, develops after birth

Types of hydrocephalus



Etiology

- **Congenital**

  Aqueductal anomalies

  Dandy Walker malformation

  Chiari II malformation

  Myleomeningocele

  Intrauterine viral infection (CMV, mumps, rubella, varicella)

  Toxoplasmosis

  Congenital tumors

  Vein of Galen aneurysms

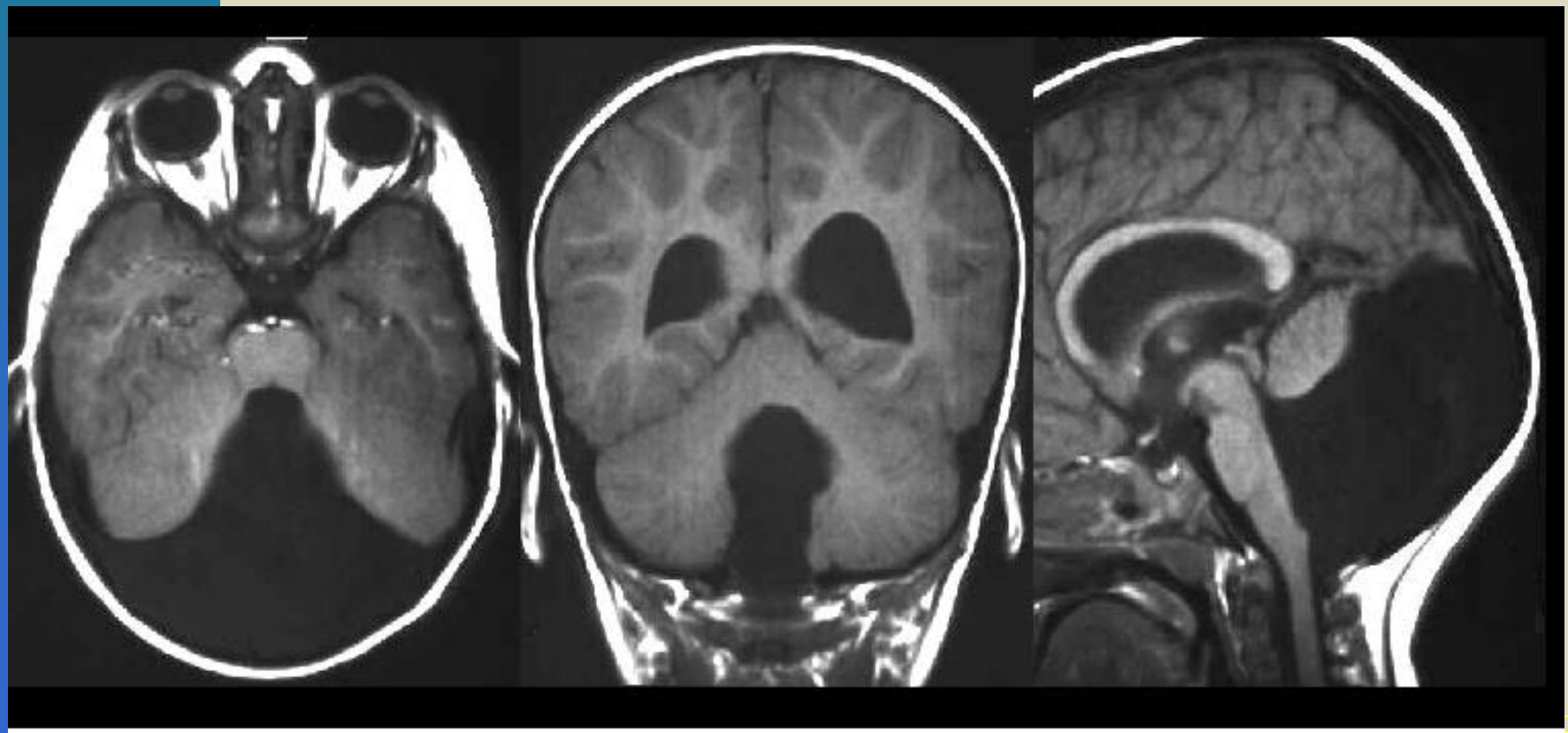
  Chromosomal anomalies (trisomy 13 and 18)

  Congenital or primary hydrocephalus.

Etiology

- **Congenital**

  Dandy Walker malformation



Etiology

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Aqueductal anomalies
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Congenital tumors
Vein of Galen aneurysms
Chromosomal anomalies (trisomy 13 and 18)
Congenital or primary hydrocephalus.

Etiology

- **Acquired**

Germinal plate hemorrhage:

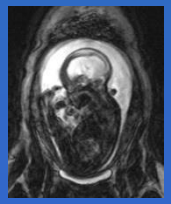
in premature babies <1500 gm (30%-40%)

Post-meningitis

Tumors

SAH

Severe TBI



Clinical features

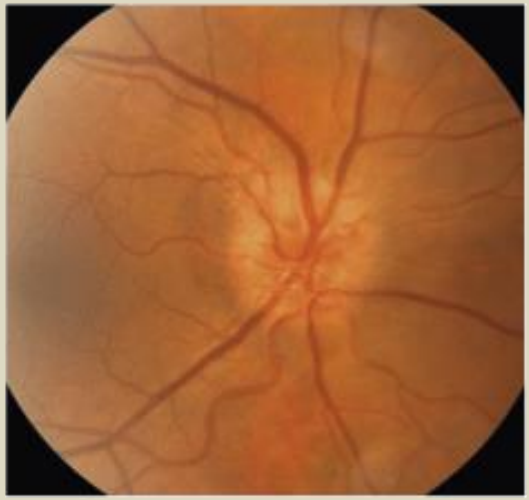
- *Infants & young children:*

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. Epidsodic bradycardia and apnea.

Clinical features

- **<u>Juvenile & adult:</u>**

  Headaches
  Nausea
  Vomiting
  Decreased level of consciousness
  Focal neurological deficit (rare)
  Papilledema



1. Optic nerve edema, O.D.

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

Investigations

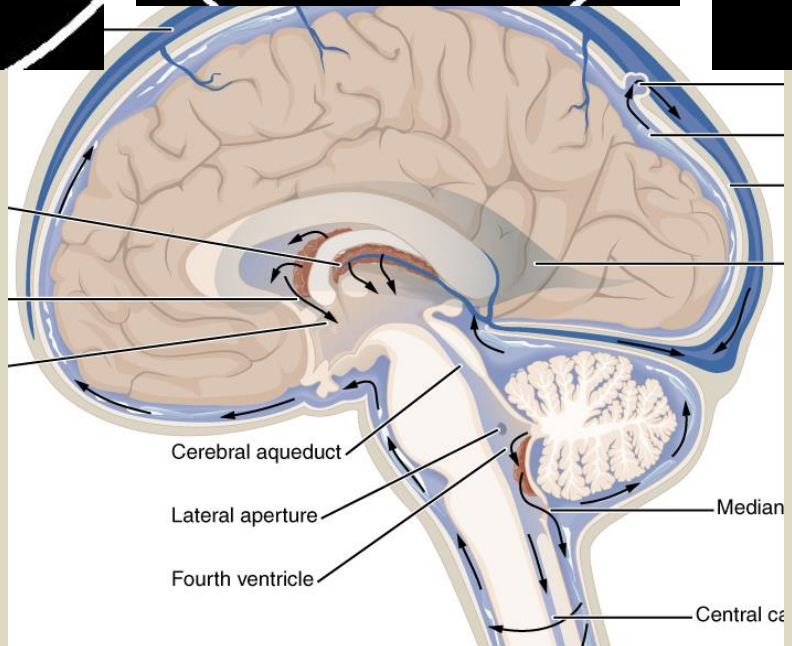
- *CT or MRI*:

- The pattern of ventricular enlargement can help delineate the cause:
- Lateral & 3rd ventricle dilatation

  → normal 4th ventricle: suggests aqueduct stenosis

  → deviated or absent 4th ventricle: suggests posterior fossa tumor
- Generalized dilatation: suggests a communicating hydrocephalus.

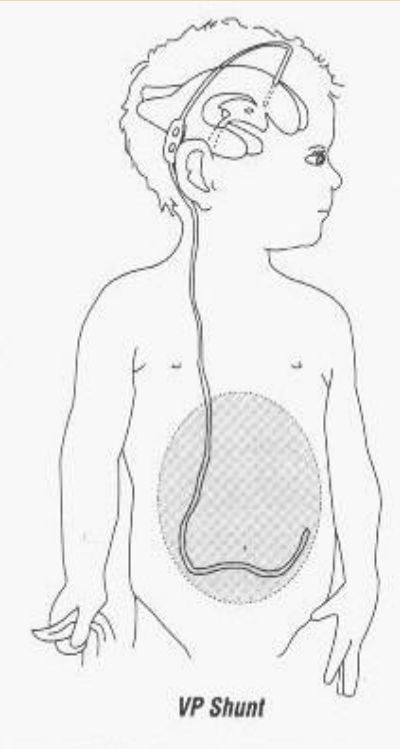
Investigations

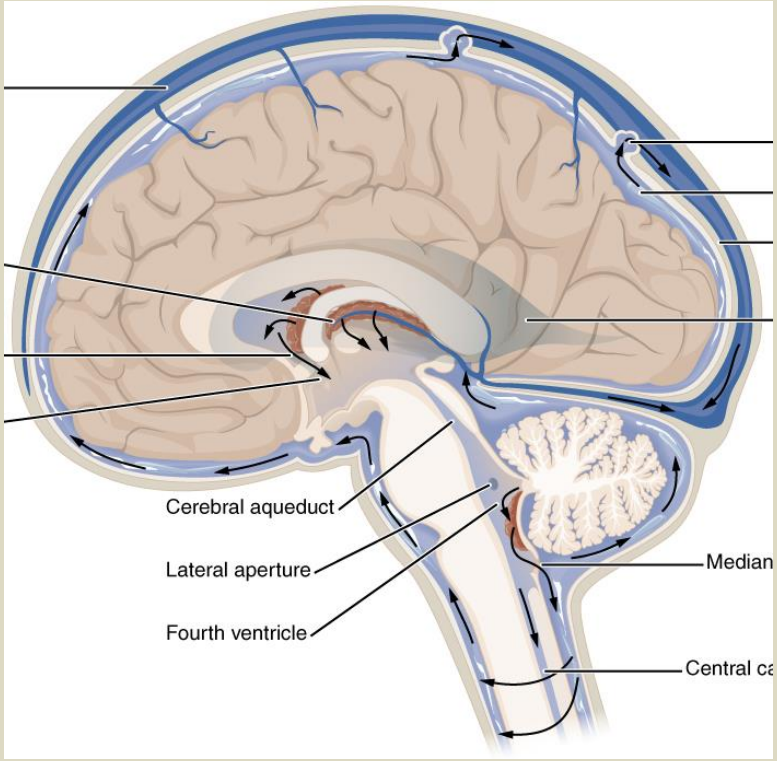


Treatment

Communicating: Medical or surgical

Obstructive: **SURGICAL TREATMENT**



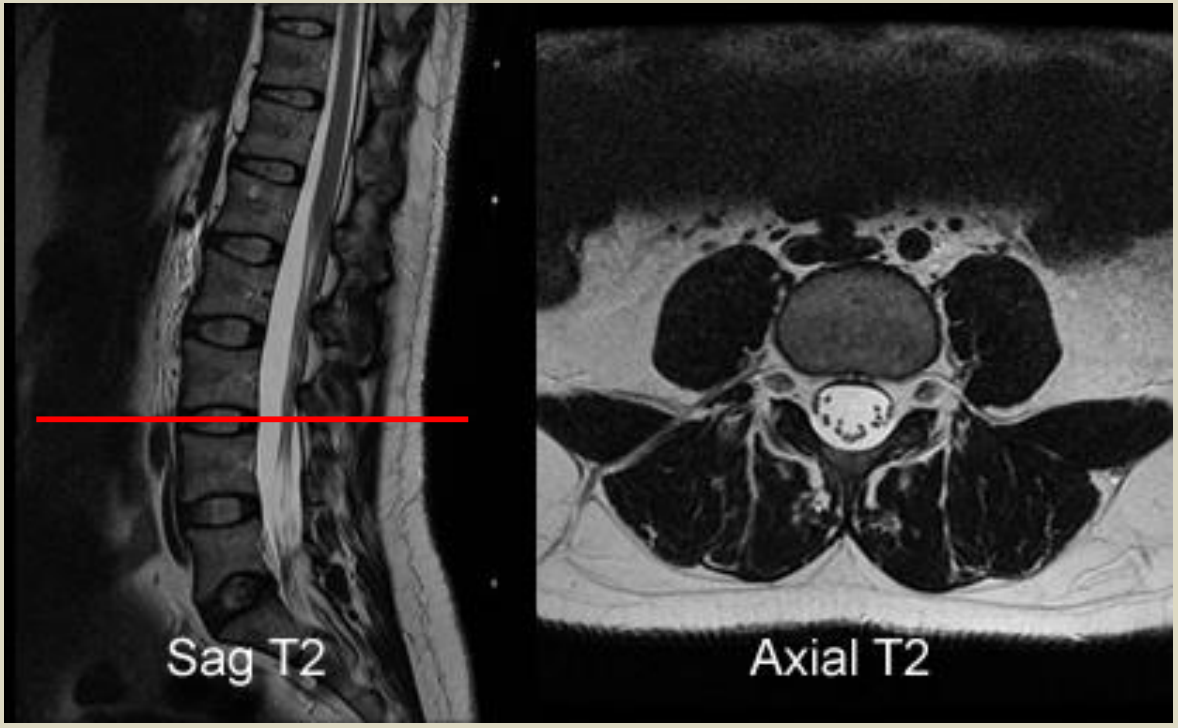


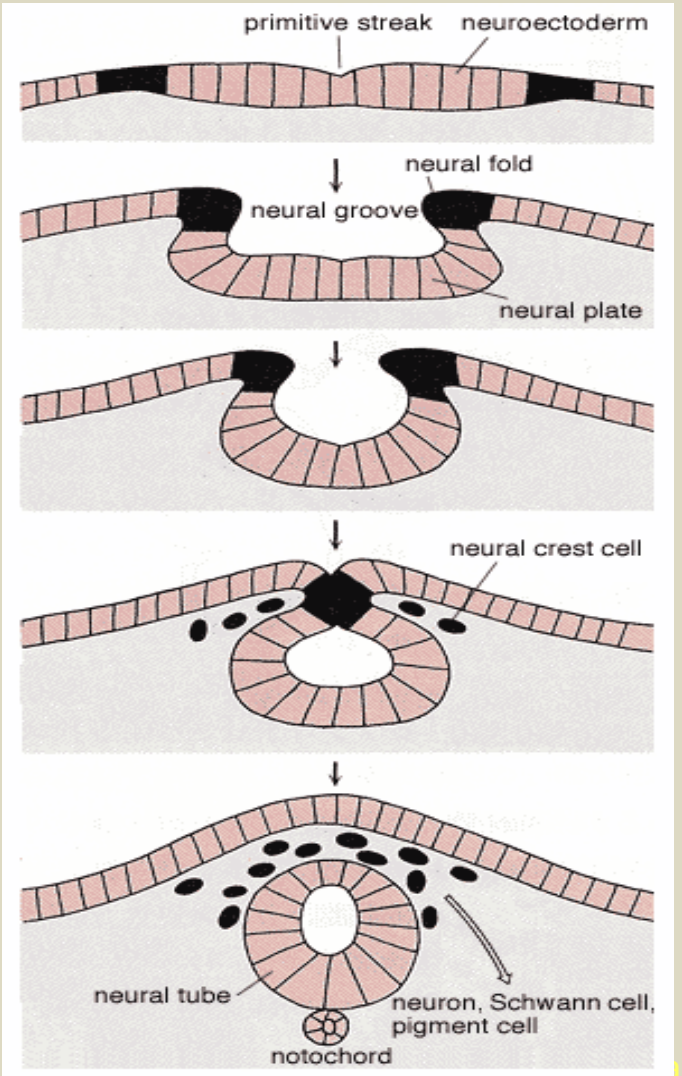
Neural Tube Defect

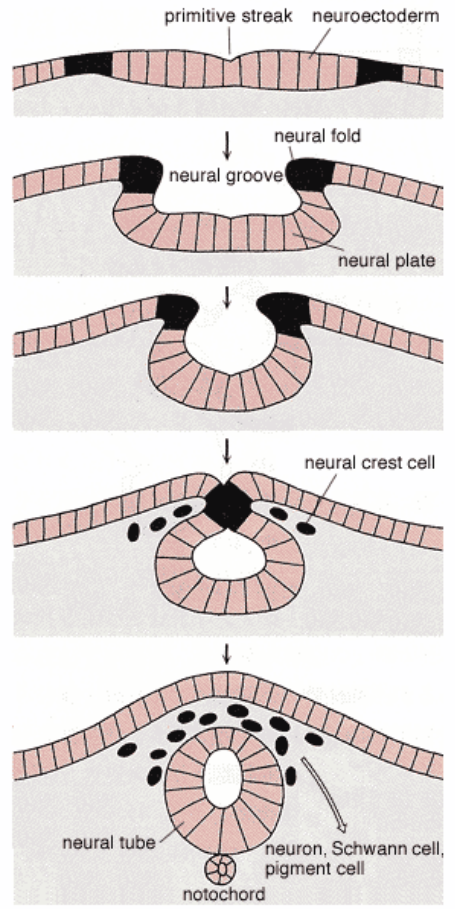
Spinal Dysraphism

Failure of closure of posterior neural arch

Two major types: Open or Closed






primitive streak
neuroectoderm
neural fold
neural groove
neural plate
neural crest cell
neural tube
neuron, Schwann cell,
pigment cell
notochord

Types of Myelodysplasia

- Spina bifida occulta
- Meningocele
- Myelomeningocele

NTD

- <u>Spina bifida occulta</u>

  5-10% of population

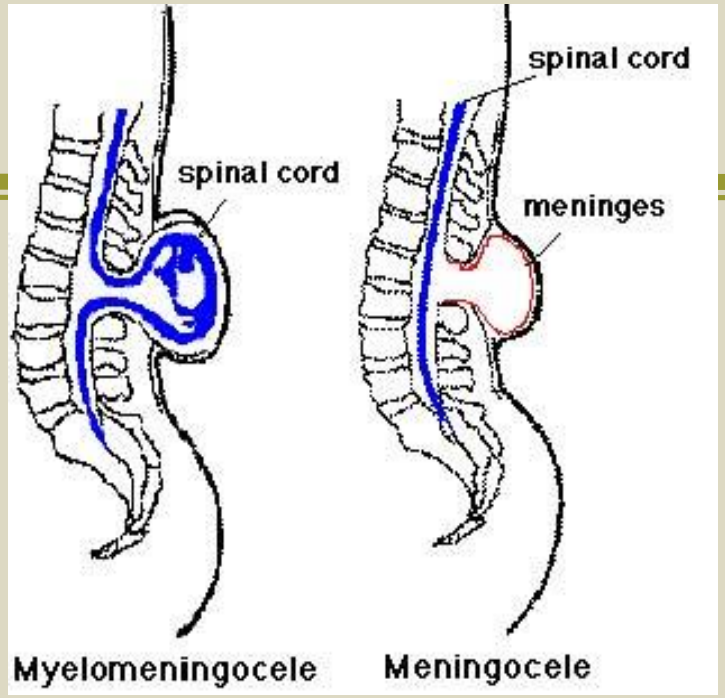
  not clinically significant

  tuft of hair, dimple sinus or port wine stain

  high incidence of underlying defect

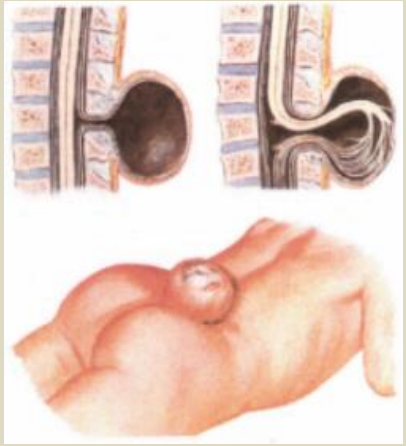
  no treatment required

NTD



- Meningocele

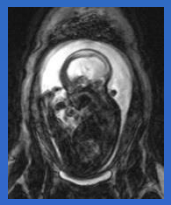
-Cystic CSF-filled cavity lined by meninges
-no neural tissue
-communicates with spinal canal
-look for other cong. Anomalies
-seldom any neurological deficit
-U/S or MRI



NTD

- 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
U/S or MRI
gross hydrocephalus, multiple serious cong. anomalies



NTD

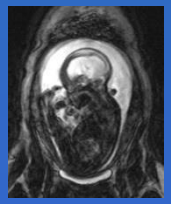
- Incidence

  2/1000 birth

  Risk increase to 5% if a sibling is affected

  Teratogens

  How to prevent?



NTD

- <u>Antenatal diagnosis</u>

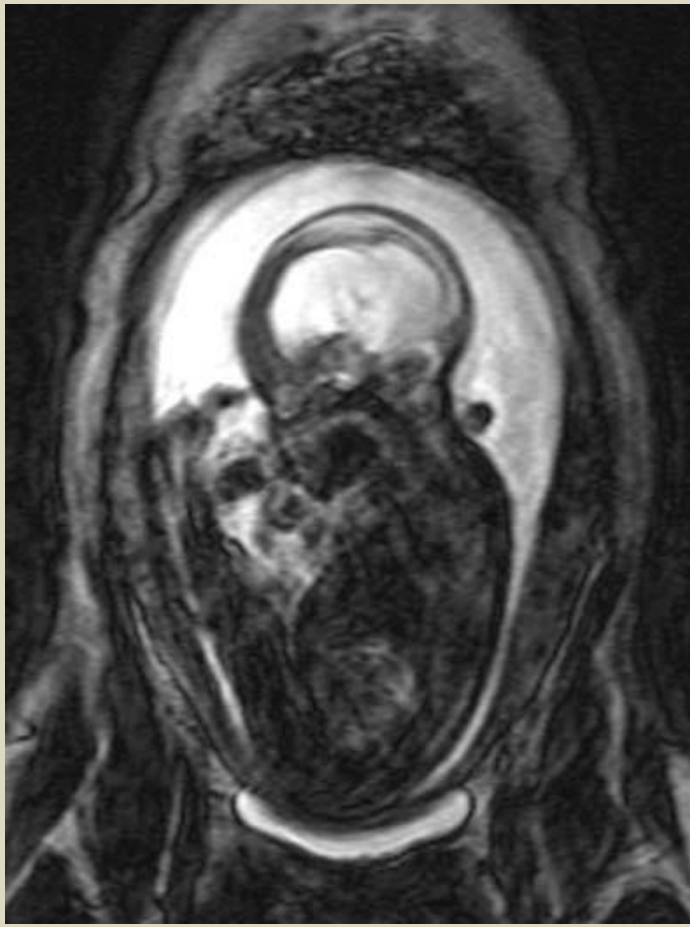
  Maternal U/S,

  MRI

  Serum/amniotic fluid for alpha-fetoprotein & acetylcholinesterase

  Contrast enhancing amniography

  possibility of therapeutic abortion



Other congenital anomalies

- Encephalocele

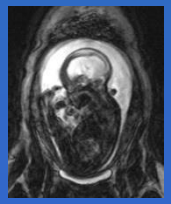
Usually occipital

may contain occipital lobe, or cerebellum

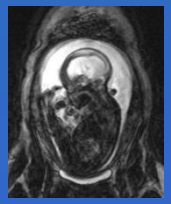
often associated with hydrocephalus

Immediate treatment if ruptured

outcome depends upon contents



Other congenital anomalies



Chiari Malformation

When part of the cerebellum is located below the foramen magnum, it is called a Chiari malformation. http://www.ninds.nih.gov/disorders/chiari/detail_chiari.htm

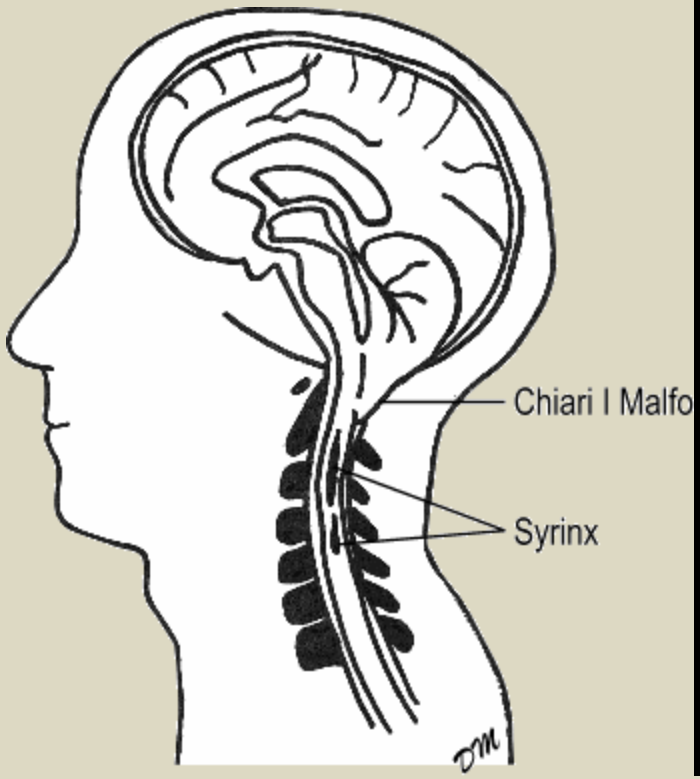


Figure 2. Chiari I Malformation

Chiari Malformation (Types)

- ***Type I***

  -Extension of the cerebellar tonsils into the foramen magnum, without involving the brain stem

- ***Type II***

  -Extension of both cerebellar and brain stem tissue into the foramen magnum

  -Myelomeningocele

  -Hydrocephalus

- ***Type III***
- ***Type IV***

Learning Objectives

- **Introduction to Neurosurgery**
- **Approaching neurological symptoms**
- **Congenital diseases**
  - Hydrocephalus
  - Neural Tube Defect
  - Chiari Malformation
  - Dandy-Walker Malformation
  - Craniosynostosis
  - Arachnoid Cyst

-4 y/o

-No past medical history

-Worsening headache, occipital area x 7 weeks

-His headache worsened last night

-Dizziness

-loss of balance

-N/V twice over the last 3 weeks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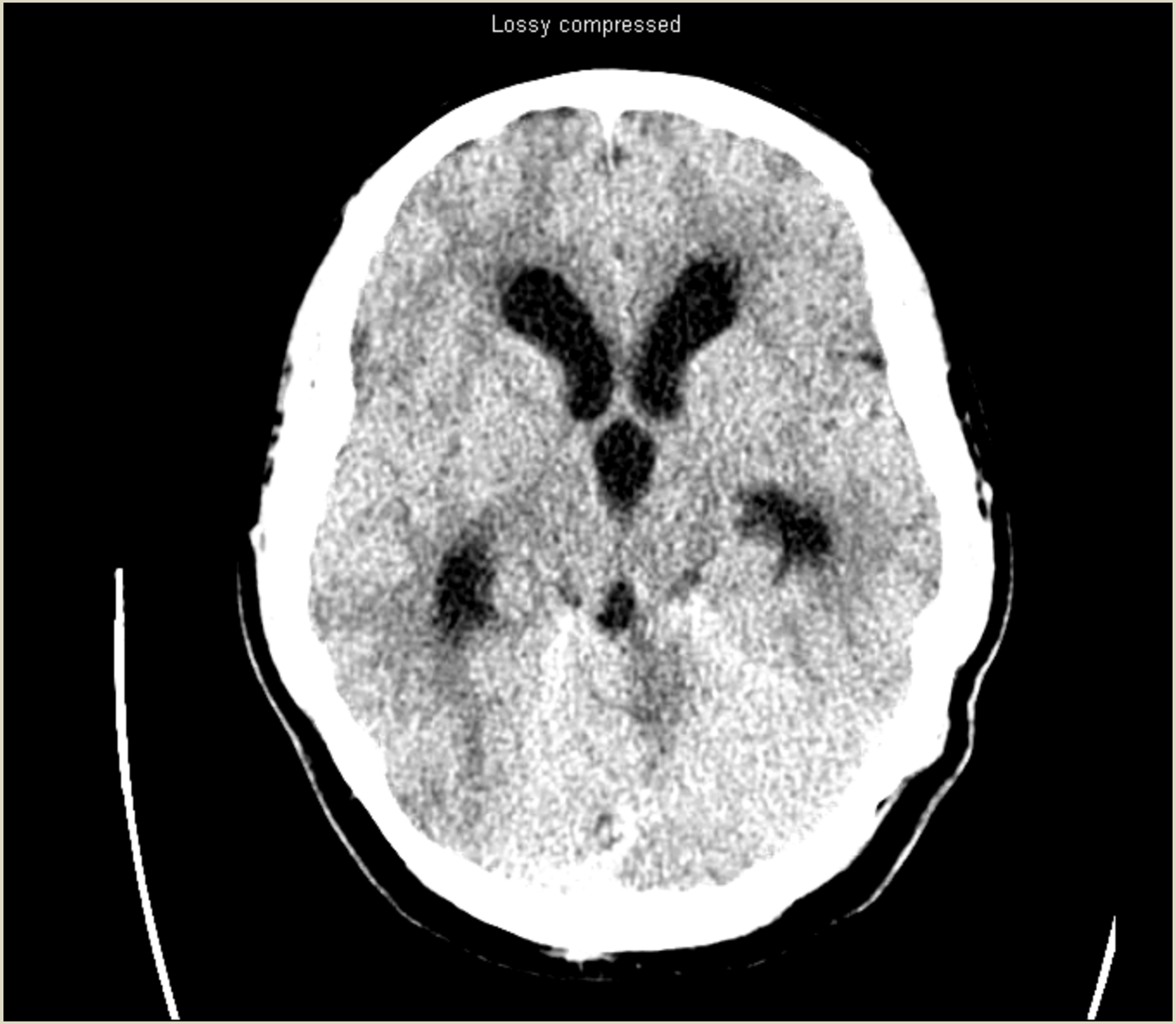
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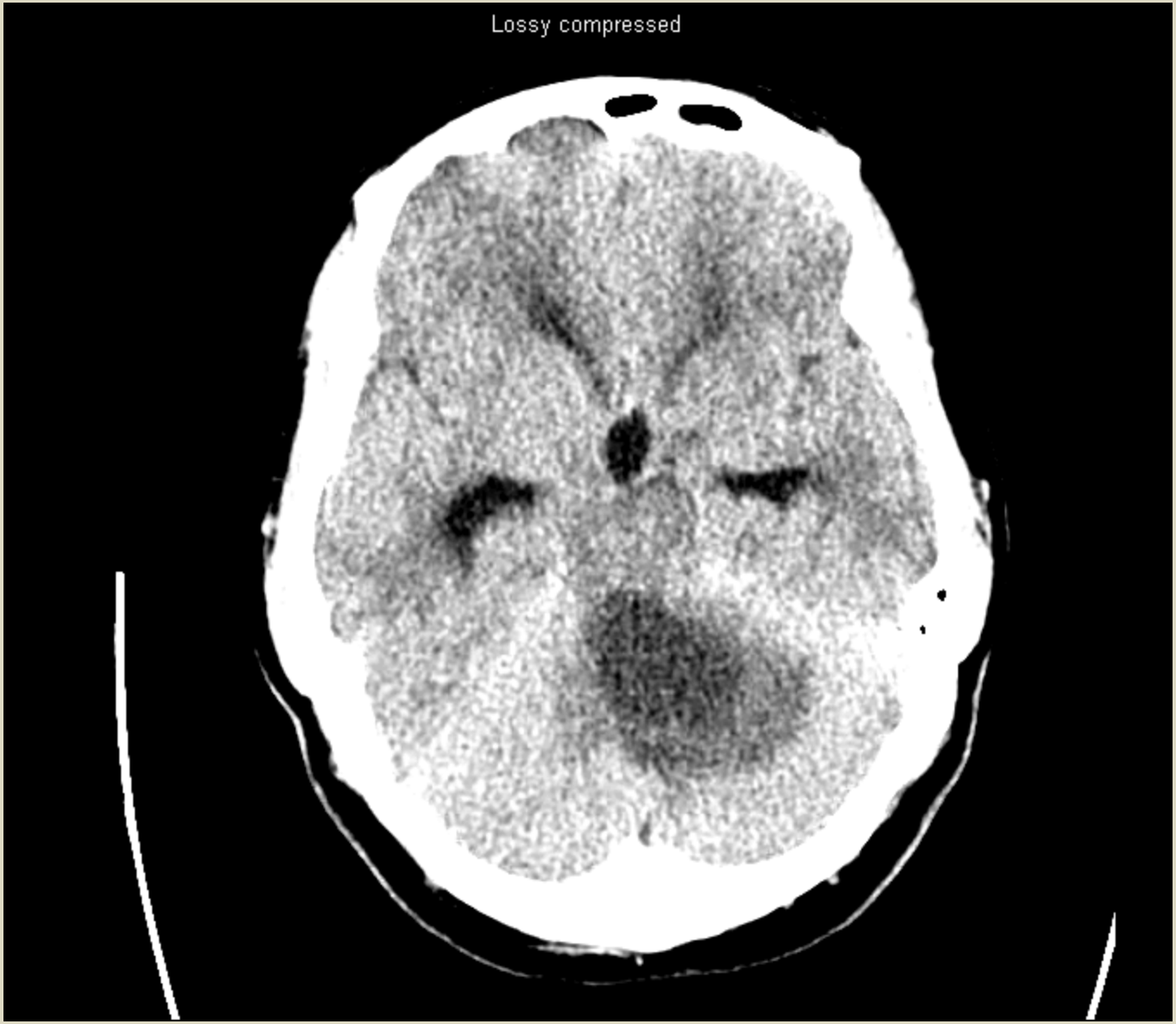
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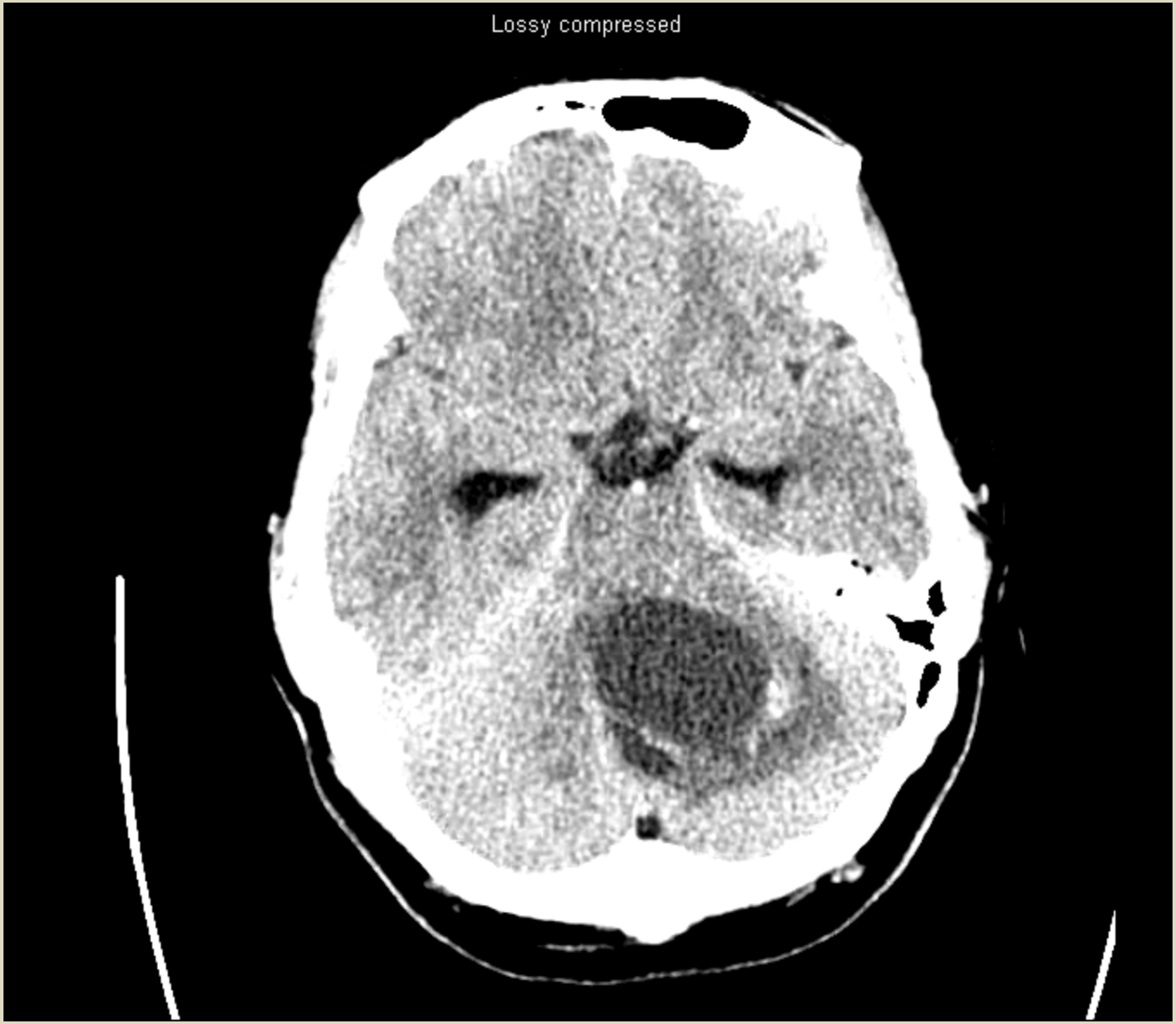
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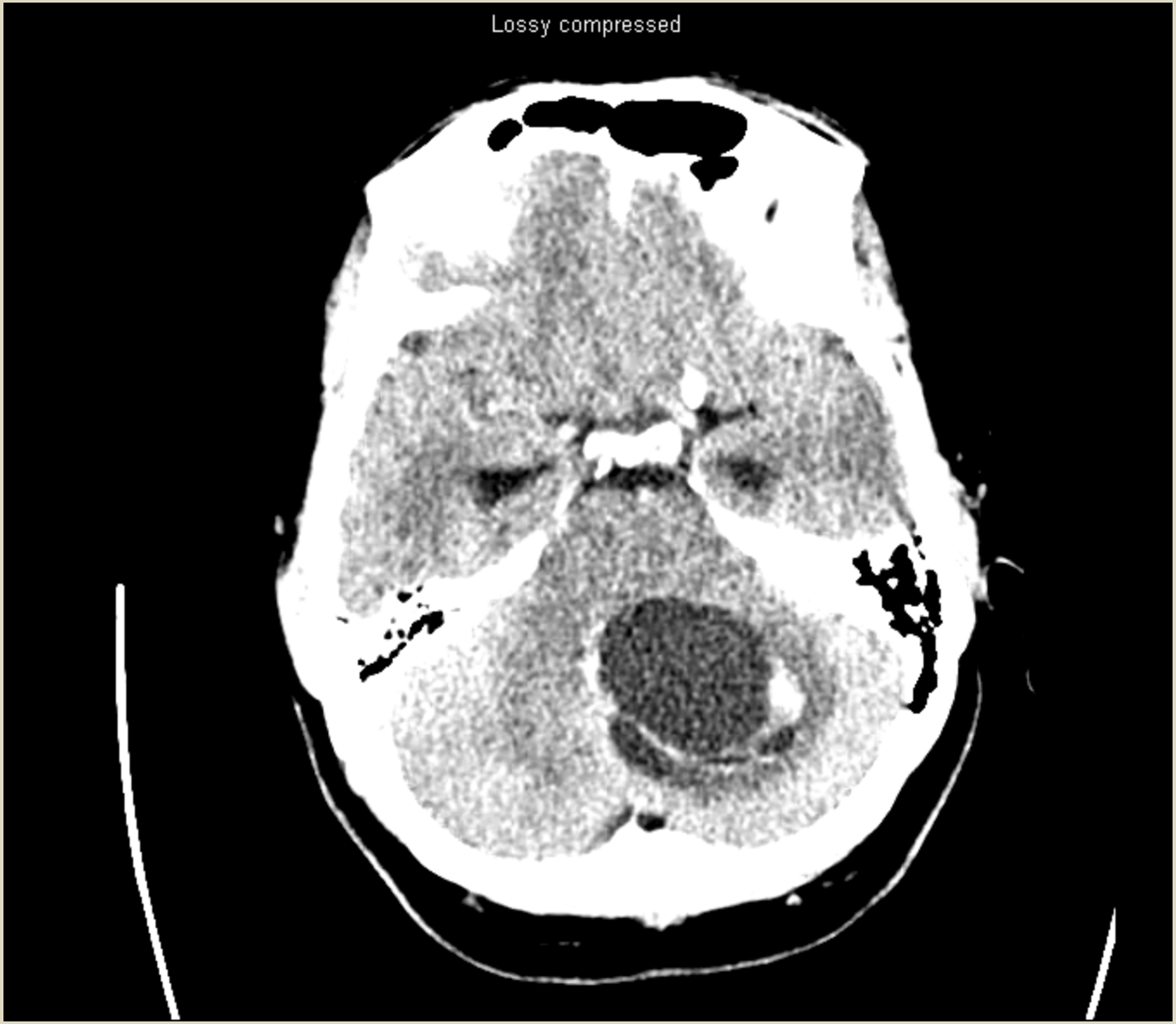
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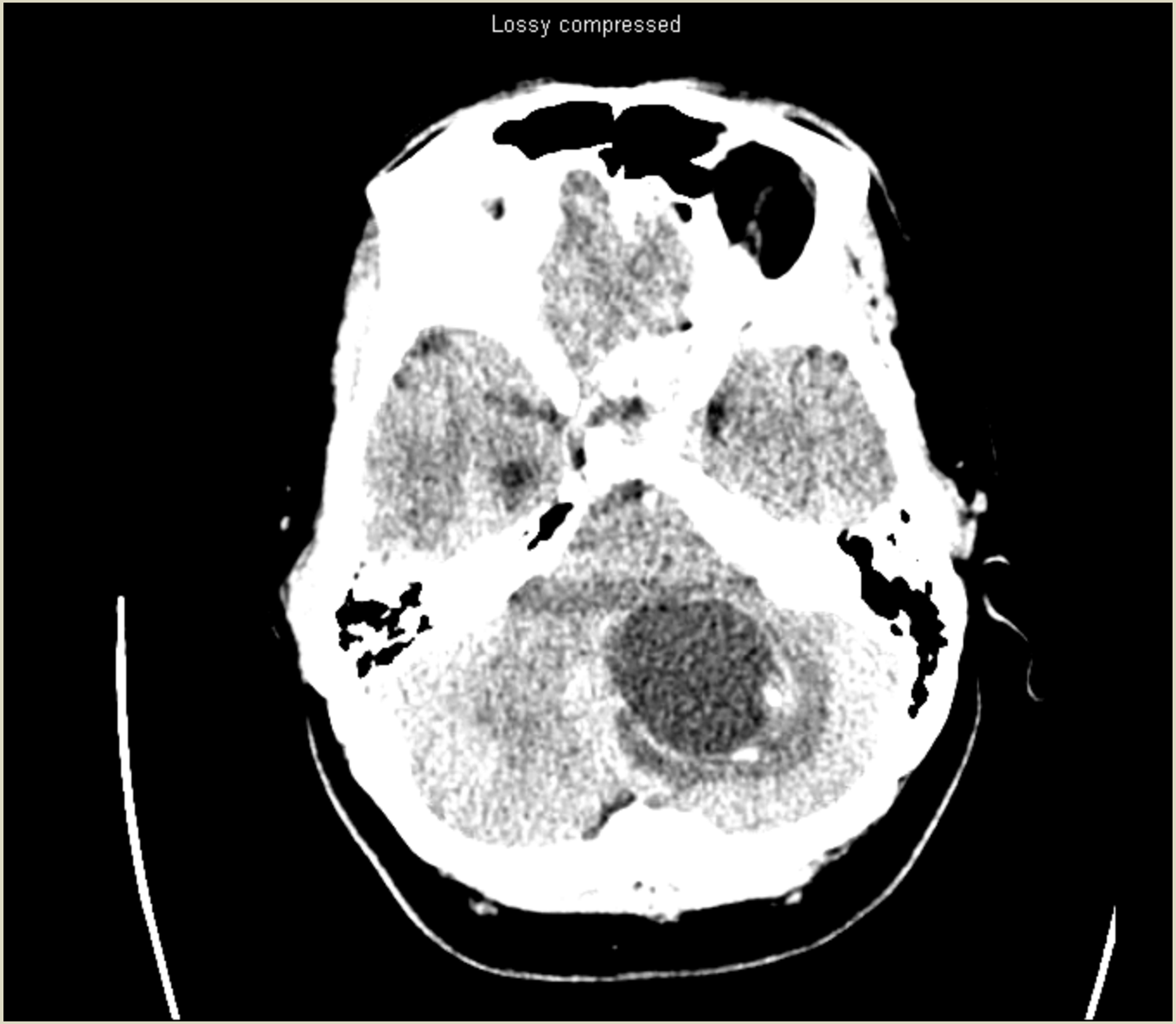
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- Other exam findings?
- D/D?
- What do u want to do?


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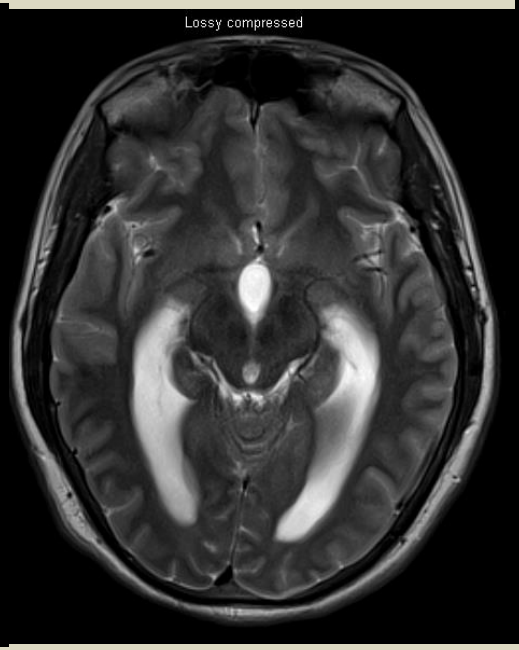
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What do you want to do?

Cerebellar sign

-Gait ataxia

-Truncal ataxia

-Limb ataxia Finger-nose and heel-knee-shin

→ intention tremor , dysmetria (past pointing) &dysrhythmia

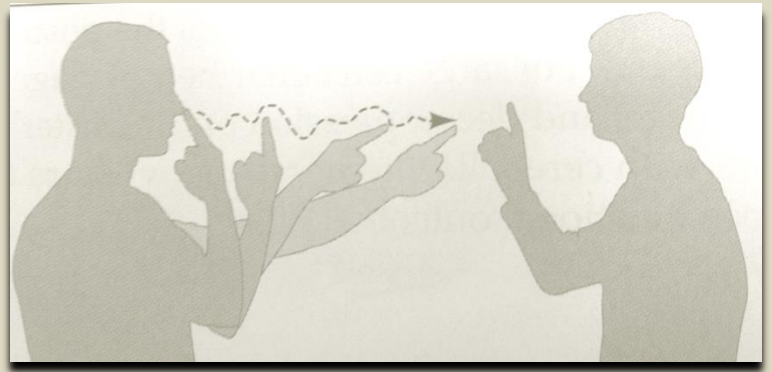
-Cerebellar dysarthria

-Hypotonia

-Rapid alternating movements (dysdiadochokinesia)

-Tremor

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



Thank You

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