Learning Objectives

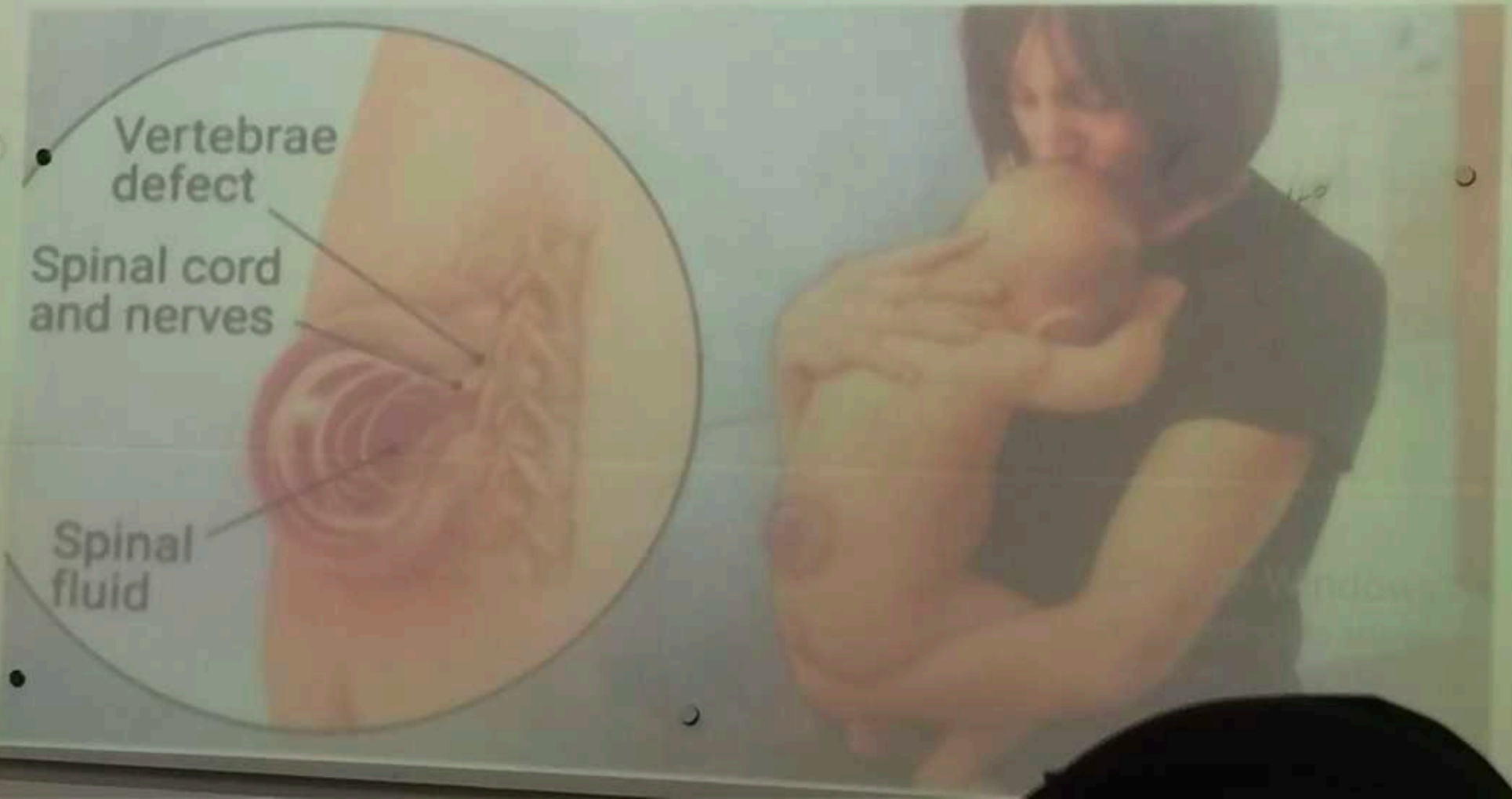
Definition of Spina Bifida and Hydrocephalus

Etiology of Spina Bifida and Hydrocephalus

Types of Spina Bifida and Hydrocephalus

Management of Spina Bifida and Hydrocephalus

Spina bifida



Embryology

- Neural tube formed by ectodermal neural crest
- Occurs by 28 days of gestation
- Neural crest deepens

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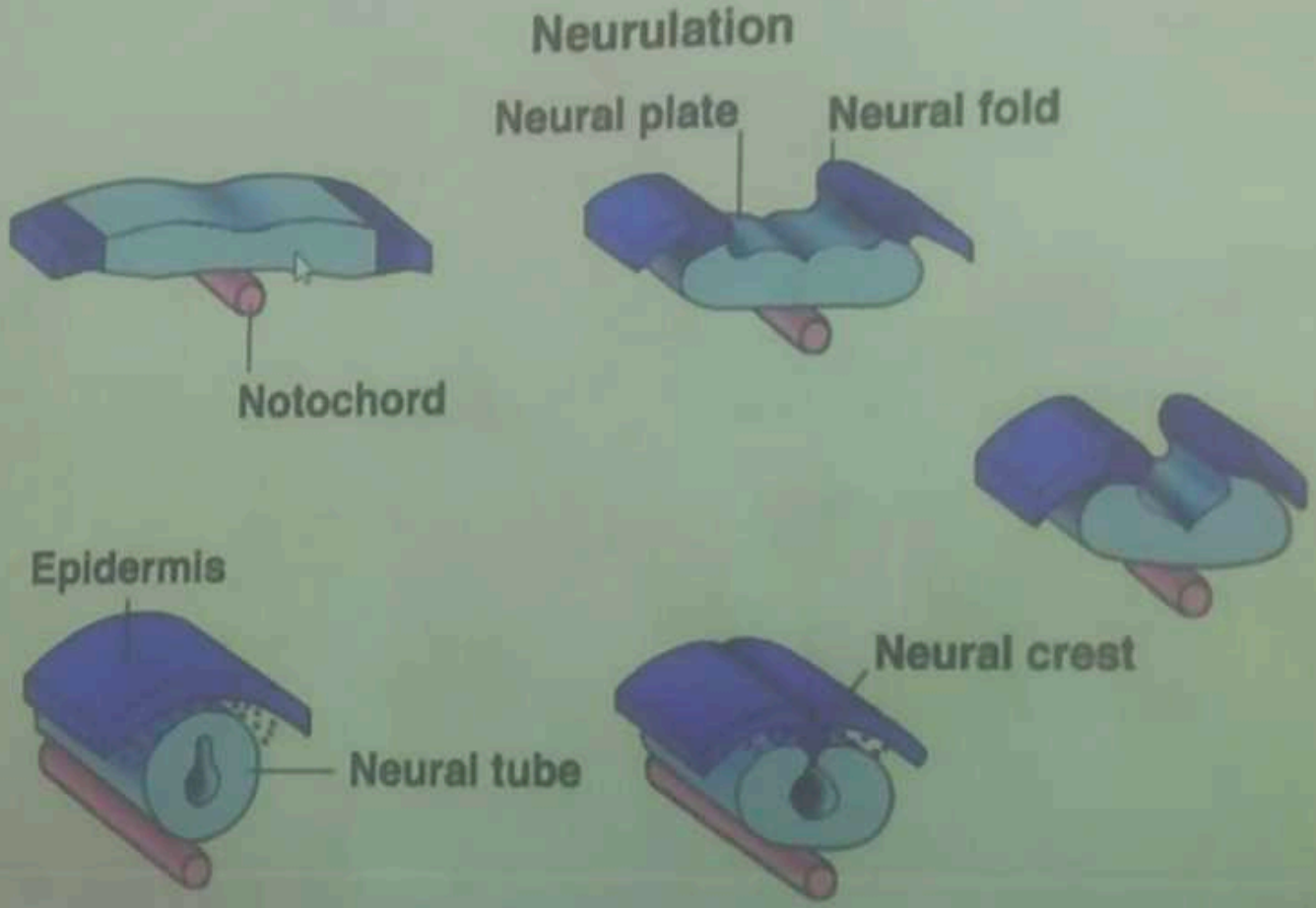
Neural groove

↓

Neural tube

} Neurulation


Neurulation
Neural plate
Neural fold
Notochord
Epidermis
Neural tube
Neural crest
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Myelodysplasia- Congenital failure of the neural tube to close

- Anencephaly- defect in anterior closure of the neural groove with brain and skull poorly developed
- Encephalocele- brain herniation through cephalad defect
- Spina bifida cystica- fusion fails in the midline or caudal neural groove with

  Meningocele- herniation of meninges

  Meningomyelocele- herniation of meninges plus neural tissue

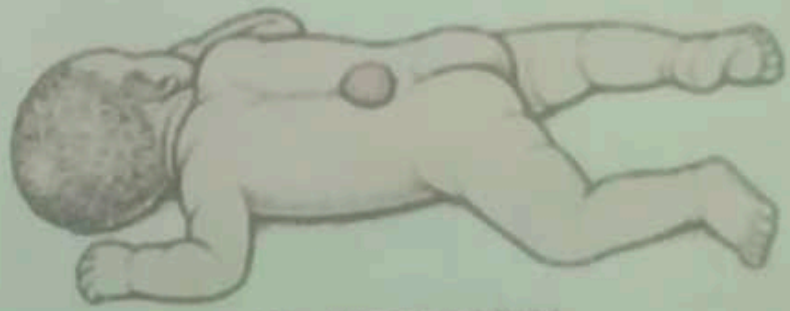
Etiology

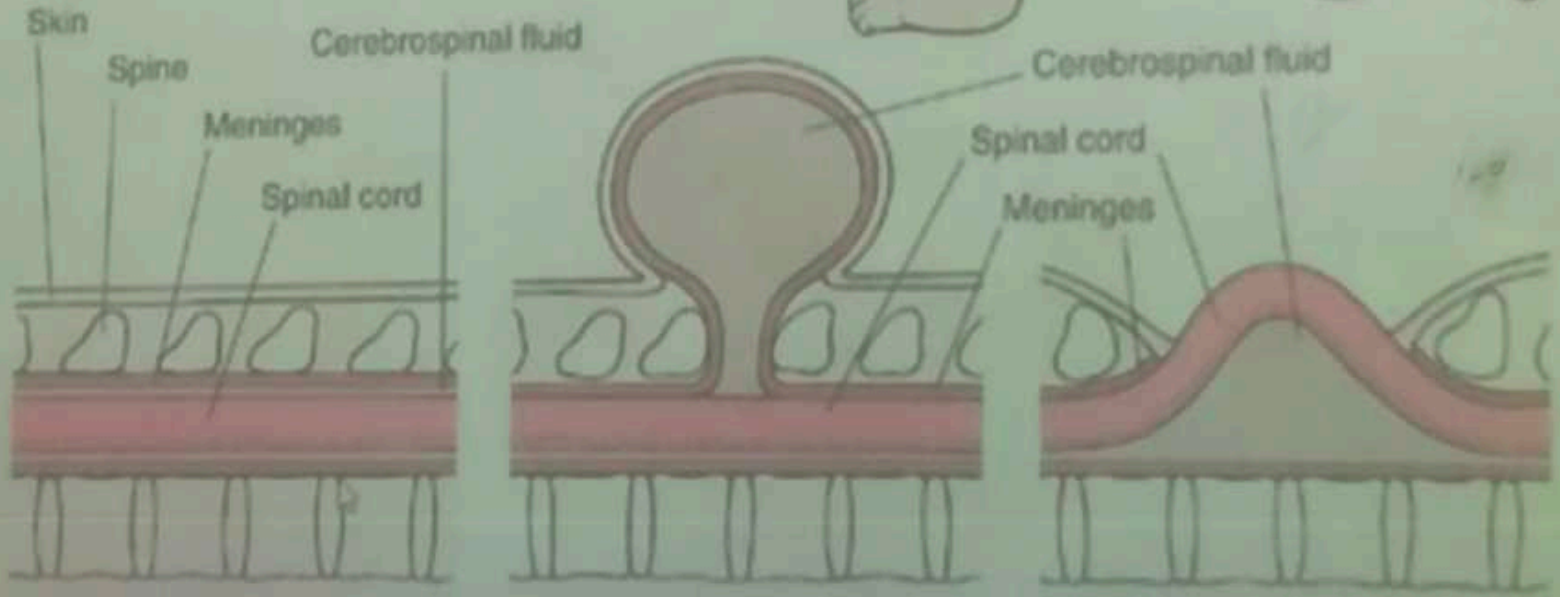
- Genetic
  - Females are more affected
  - More common in Caucasians and blacks
  - Parenteral consanguinity increases incidence
- Environmental- folic acid deficiency
- Teratogenic drugs- valpraote, carbamazapine
- Higher prevalence in lower socio economic groups

Pathophysiology

- Meningocele (20%) - sac containing meninges bulges through the defect. Overlying skin intact. Neurological manifestations usually absent
- Meningomyelocele (80%) – contains neural elements along with meninges. Majority of defects occur in lumbosacral area. Neurological deficits distal to defect are most severe.
- Spina bifida occulta – absence of bony spinous process. No clinical significance

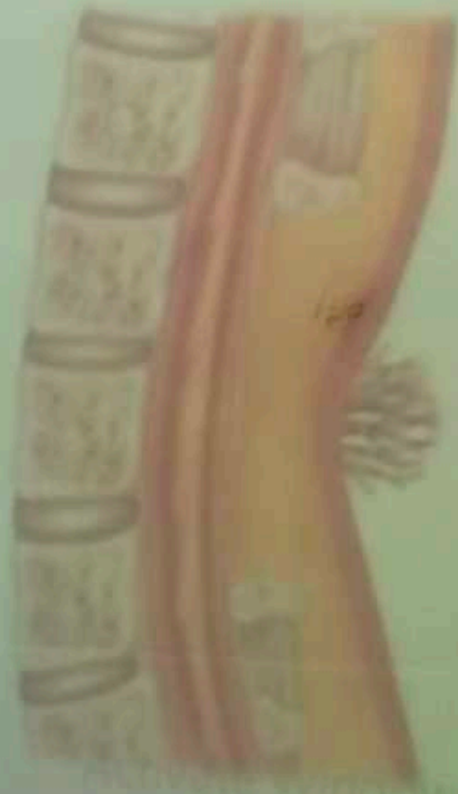
Types

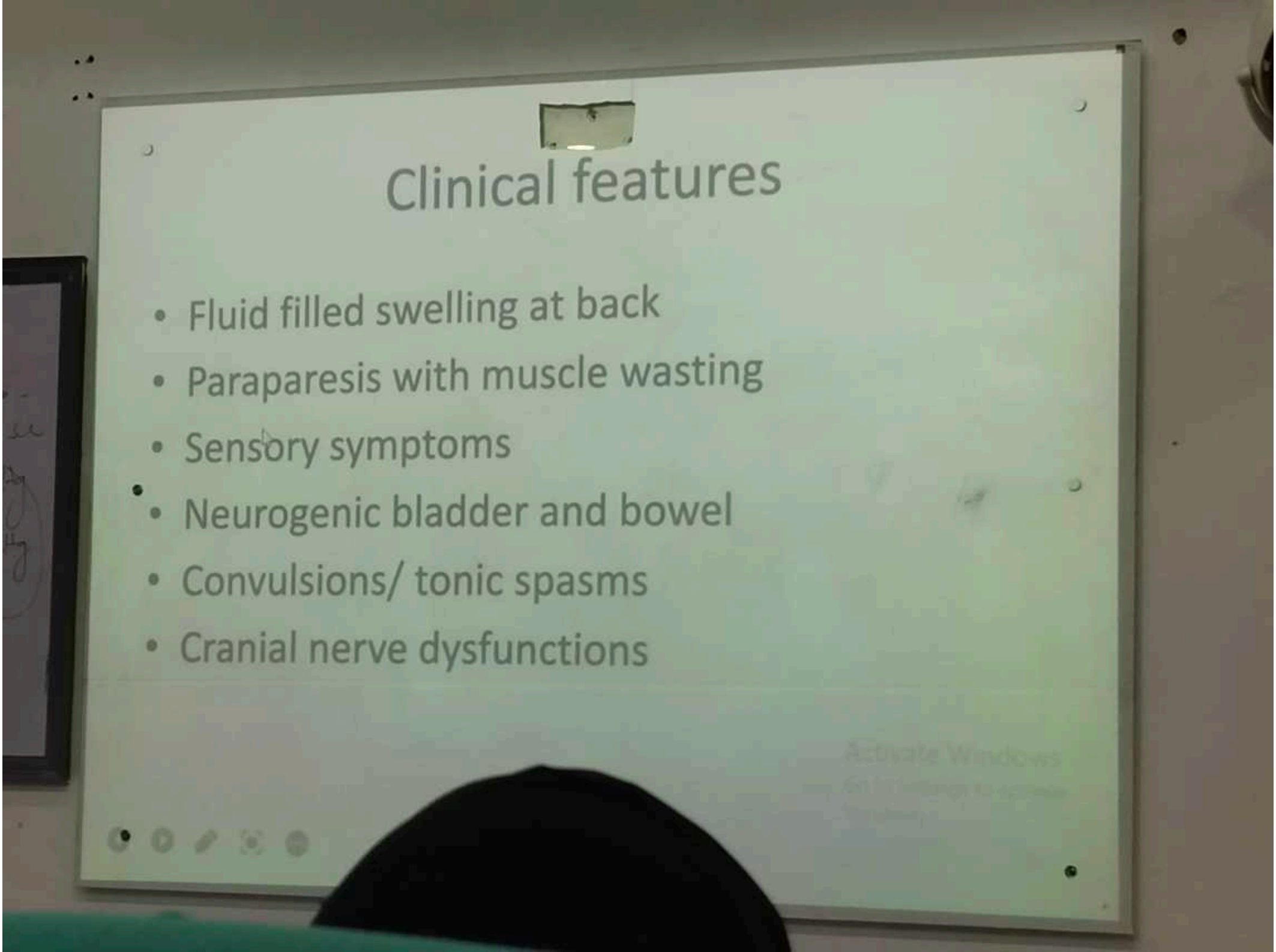




Spina bifida occulta

- *Occulta* is Latin for **"hidden"**.
- This is the mildest form of spina bifida.
- In occulta, the outer part of some of the **vertebrae** is not **completely closed**.
- The splits in the vertebrae are so small that the spinal cord does not protrude.




Clinical features
• Fluid filled swelling at back
• Paraparesis with muscle wasting
• Sensory symptoms
• Neurogenic bladder and bowel
• Convulsions/ tonic spasms
• Cranial nerve dysfunctions

Associated anomalies

- Club feet- commonest
- Arnold Chiari malformation (80-90%)
- Hydrocephalus (80%)
- Neurogenic bladder(90%)
- Musculo skeletal defects
- Urogenital anomalies
- Facial clefts
- Umbilical hernia
- Congenital heart diseases - rare

Complications

- Rupture of sac during birth process
- CSF leak
- Raised ICP- vomiting, convulsions, altered sensorium, irritability
- Infection – fever, unconsciousness, altered sensorium (meningitis)

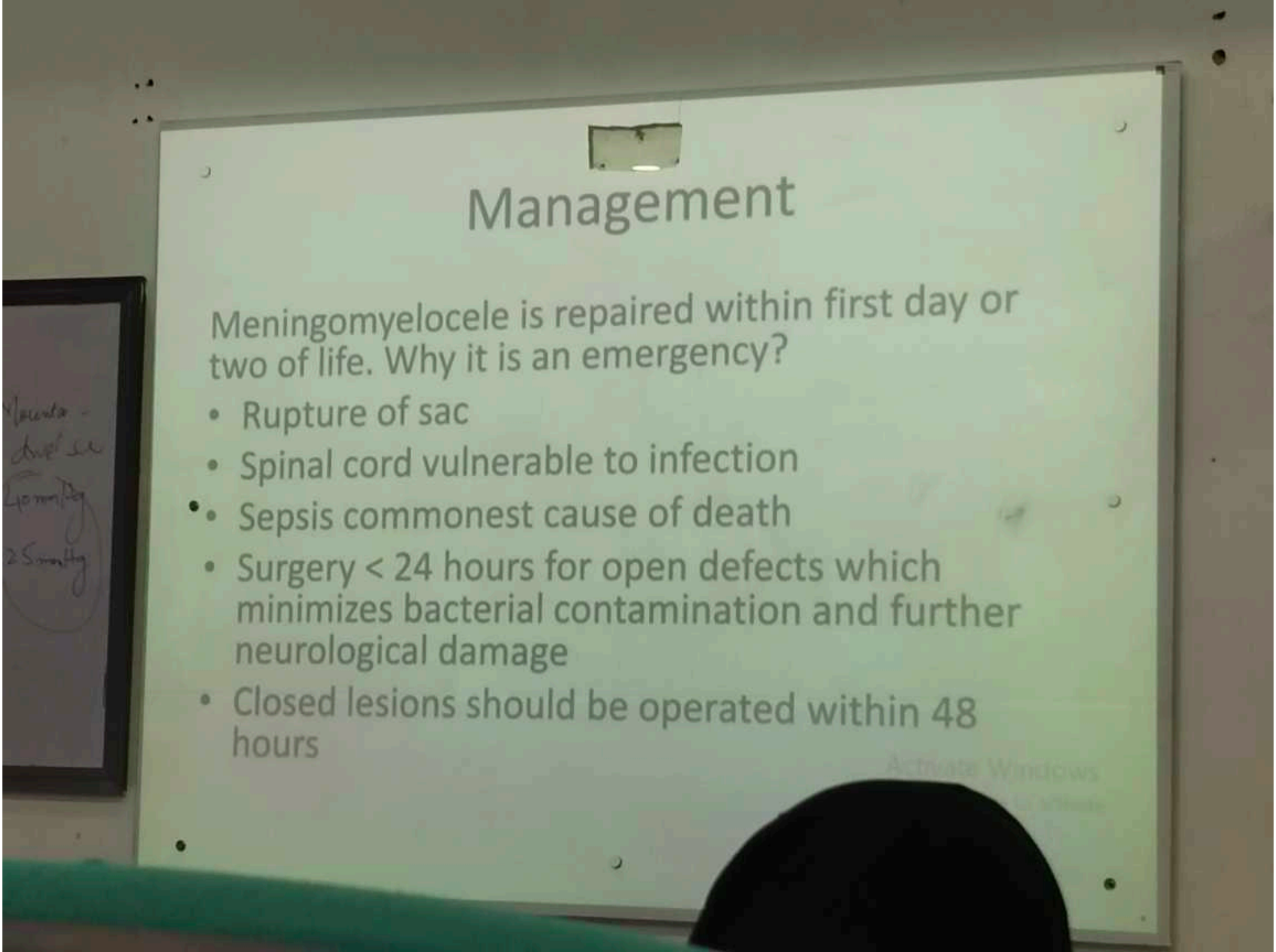
Diagnosis

Prenatal

- ↑ Acetyl cholinesterase and alpha fetoprotein in maternal serum and amniotic fluid (in 2nd
- trimester)
- Prenatal ultrasound picks up the defect in 100% of the cases

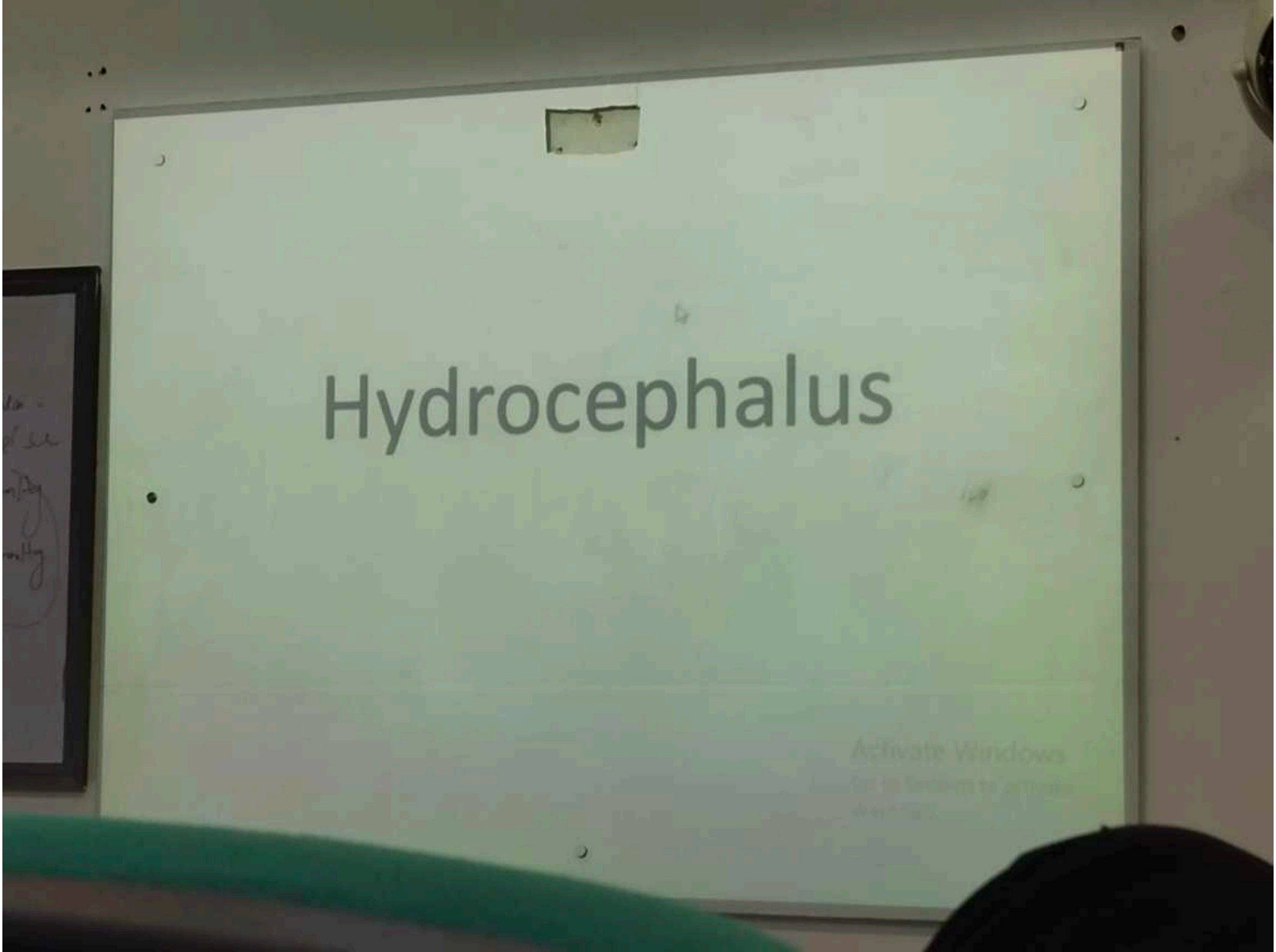
Management

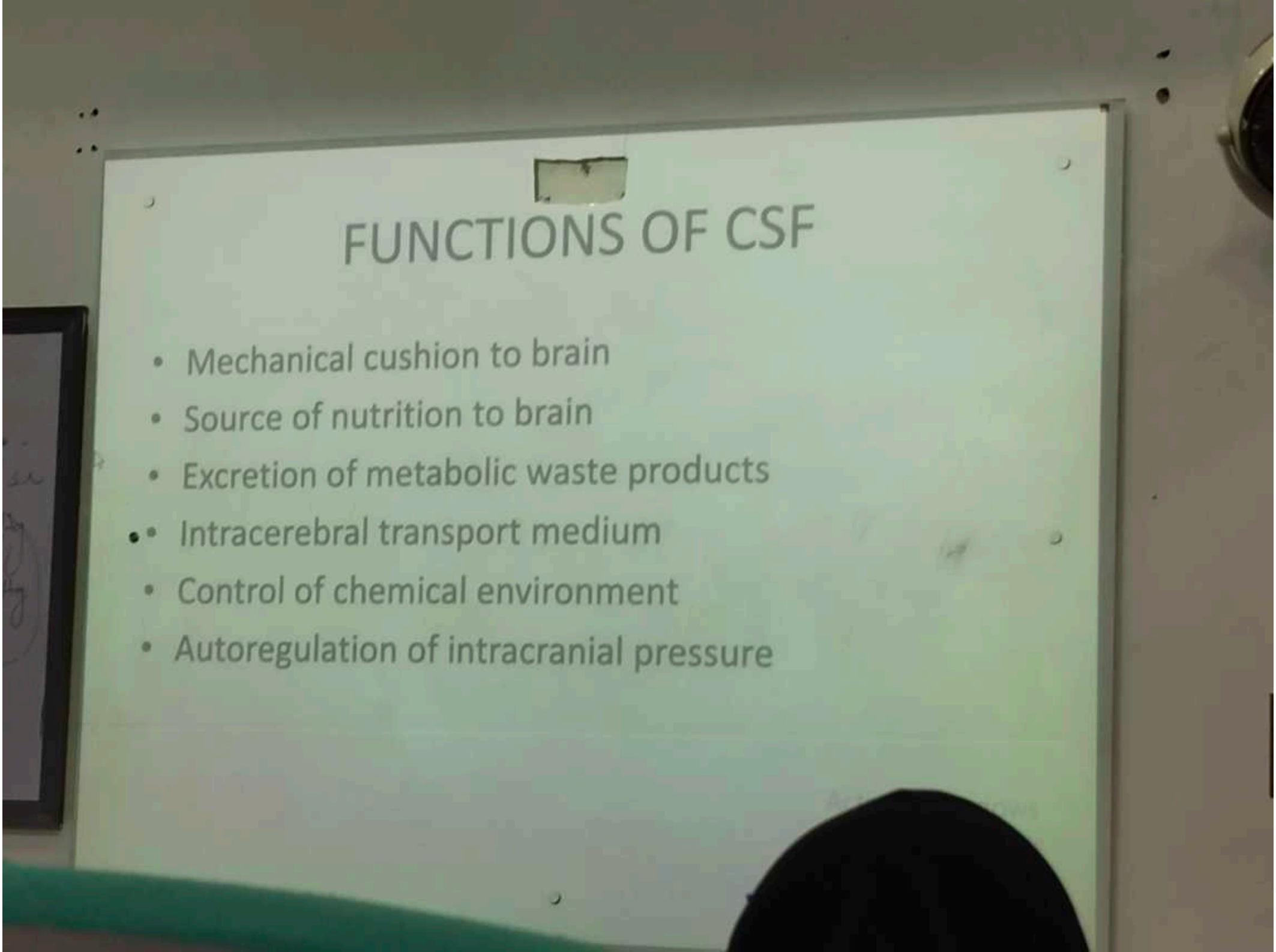
- Management of neural tube defects requires team approach with cooperation of pediatrician, neurologist, neurosurgeon and anesthesiologist
- Surgery is mainstay of treatment and includes repair of the defect and a VP shunt (if associated with hydrocephalus)


Management
Meningomyelocele is repaired within first day or two of life. Why it is an emergency?
• Rupture of sac
• Spinal cord vulnerable to infection
• Sepsis commonest cause of death
• Surgery < 24 hours for open defects which minimizes bacterial contamination and further neurological damage
• Closed lesions should be operated within 48 hours

Fetal surgery

- Presently, open midgestation surgery is largely reserved for closure of MMC
- Defect is exposed while the rest of the fetus remains bathed in amniotic fluid in the uterus
- The rationale for in utero closure of MMC appears that prolonged bathing of neurologic elements in the amniotic fluid worsens the neurologic outcome


Hydrocephalus


FUNCTIONS OF CSF
• Mechanical cushion to brain
• Source of nutrition to brain
• Excretion of metabolic waste products
• Intracerebral transport medium
• Control of chemical environment
• Autoregulation of intracranial pressure

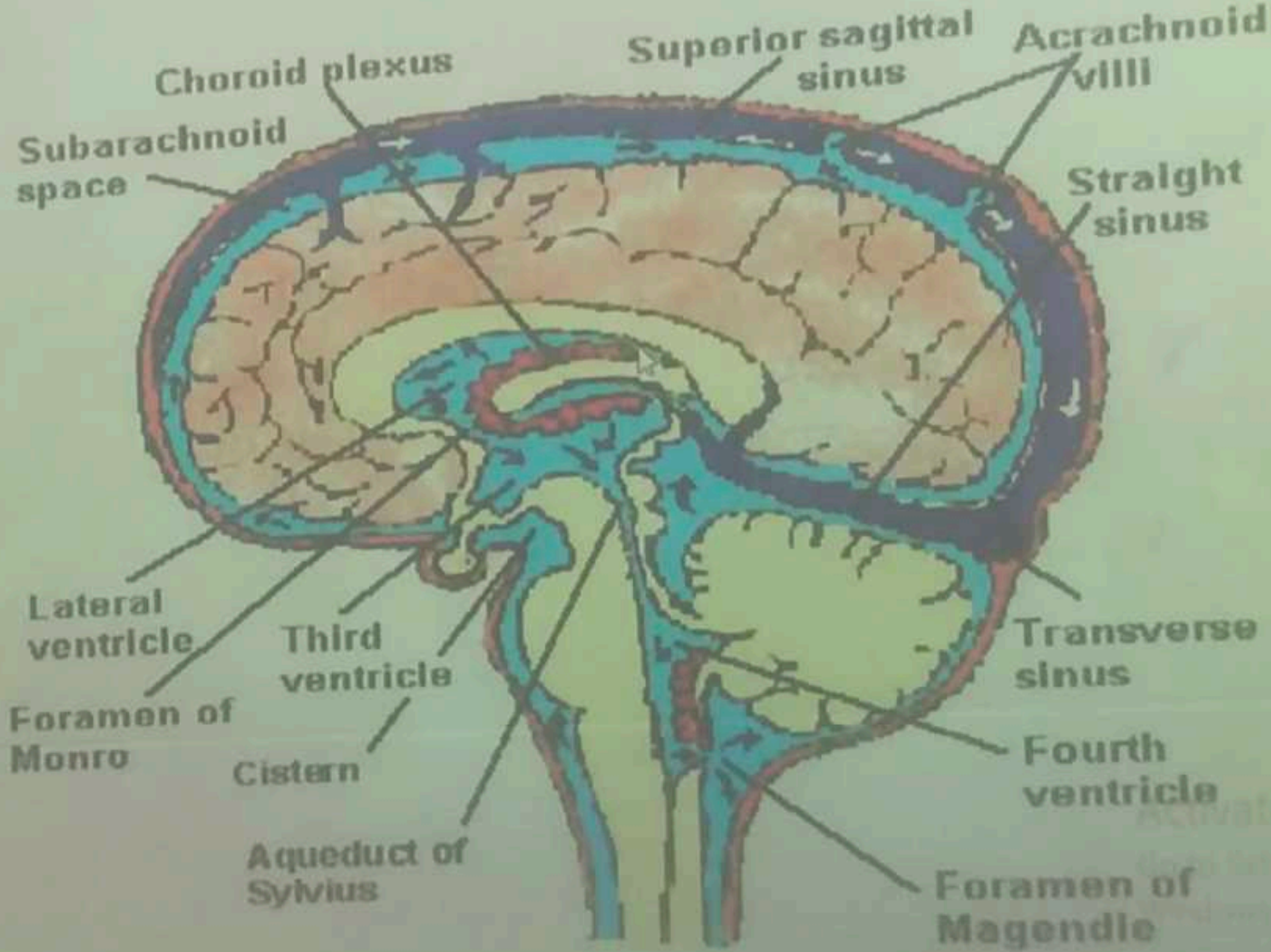
Production of CSF

- Choroidal – choroid plexus of the venticles
- Extrachoroidal
- – Ependyma
  – Neighbouring brain substance

Ventricular system

- Lateral ventricles > foramen of Monro to the midline third ventricle
- Third ventricle to fourth ventricle via aqueduct of Sylvius
- 3 exits from the 4th ventricle to cisterns
  - midline foramen of Magendie
  - paired lateral foramina of Luschka
- Cisterns connect to the subarachnoid spaces
- Absorbed at the arachnoid villi

CSF pathways



Hydrocephalus

Definition:

- It is the condition of excess cerebrospinal fluid accumulation in the head as a result of disturbance of formation, flow or absorption
- Term arises from greek, *hydro* (water) and *cephalus* (head)

Epidemiology:

- Infantile HCP: 3-4 per 1000 live births
- As a single congenital disorder: 0.9-1.5 per 1000 live birt

TYPES OF HYDROCEPHALUS

- OBSTRUCTIVE OR NON-COMMUNICATING (obstruction within the ventricular system)

- NON OBSTRUCTIVE OR COMMUNICATING (malfunction of arachnoid villi)

Etiology

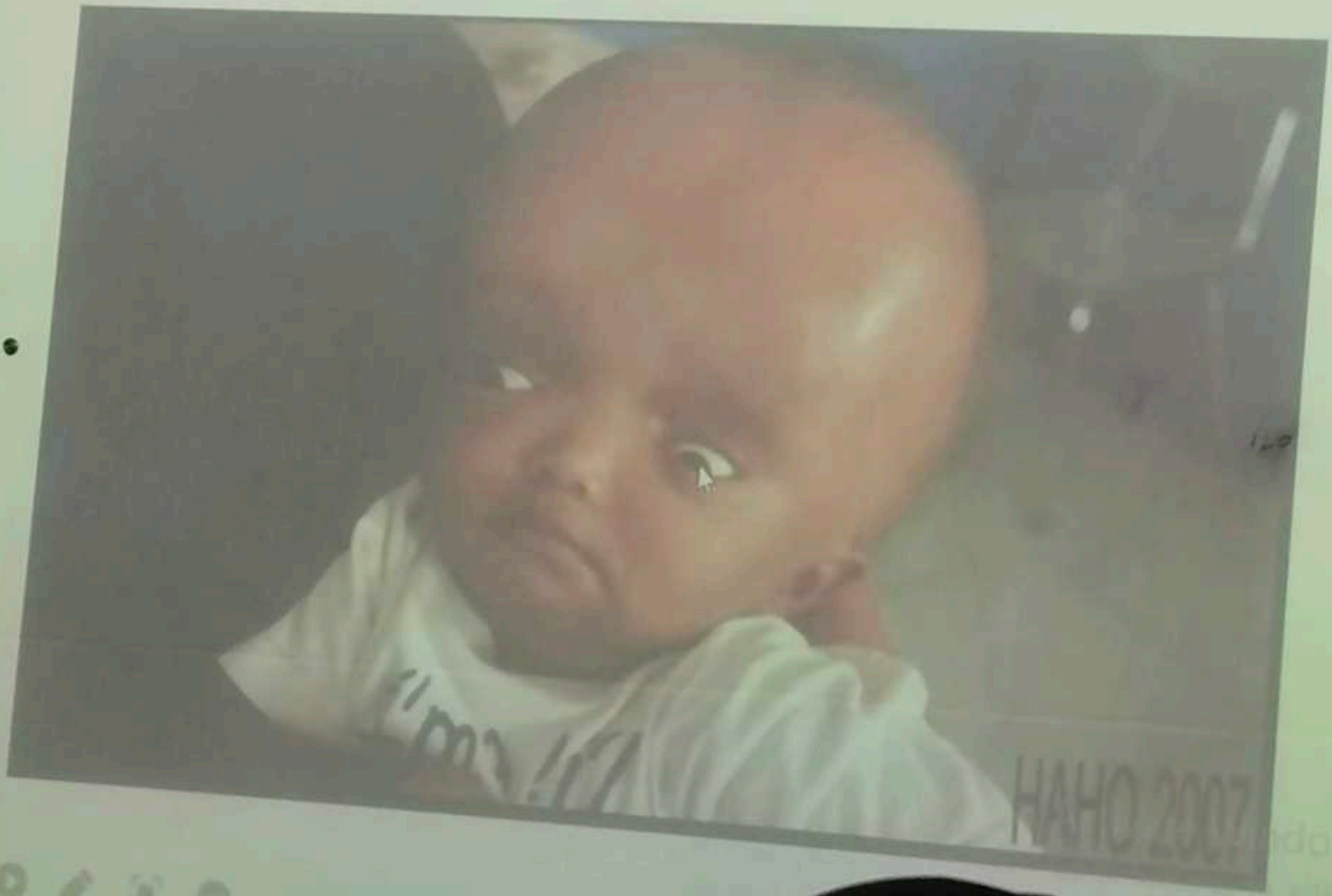
- Obstruction to flow – most common
  - Congenital
    - Neural tube defects
    - Chiari II malformation
    - Dandy- walker syndrome
  - Acquired
    - Post haemorrhagic – intraventricular
    - Post meningitic
      - Tubercular
      - Pyogenic

Clinical features

In neonatal period and children < 2 yrs

- Disproportionately increased head circumference
- Tense fontanelle, widely separated cranial sutures
- Failure to thrive, irritability and poor feeding
- ICP may be normal
- Sunset sign positive
- Crackpot sign positive
- Transillumination of head positive

HYDROCEPHALUS



Clinical features

in a child > 2 yrs

- No disproportionate enlargement of head
- Signs of raised ICP
  - Projectile vomiting
  - Headache
  - Diplopia, papilledema
  - Altered consciousness
  - Cranial nerve palsies
  - Pupillary asymmetry
  - Cushing's triad

Investigations

- Serial head circumference measurement
- Skull radiographs
- If ant fontanelle open – transcranial sonography
- If ant fontanelle closed – CT scan
- MRI

Management

- Medical (limited role in long term) - methods to reduce CSF production

  -Mannitol 1mg/kg

- -Acetazolamide 25-100mg/kg/day

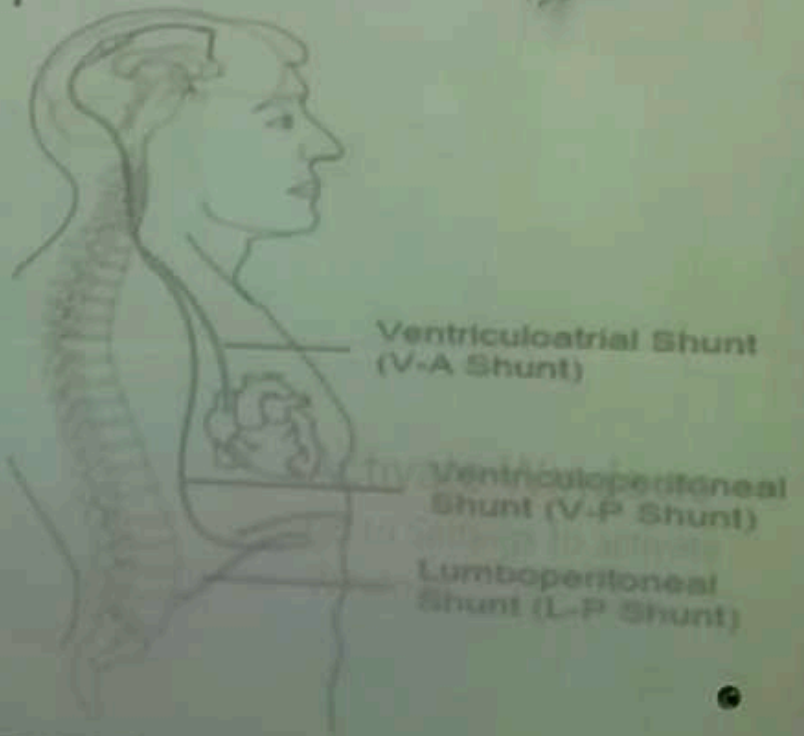
  -Oral glycerol 0.5-1g/kg(2.5% solution)

  -Furosemide 0.7 mg/kg

  -Dexamethasone 4-8 mg 4-6 hrly

Management

- Surgical
  - Reduce CSF production - choroid plexus ablation
  - Drainage of CSF – ventriculostomy
  - Drainage of CSF into an absorptive viscus
    - Ventriculoperitoneal shunt
    - Ventriculoatrial shunt
    - Lumboperitoneal shunt



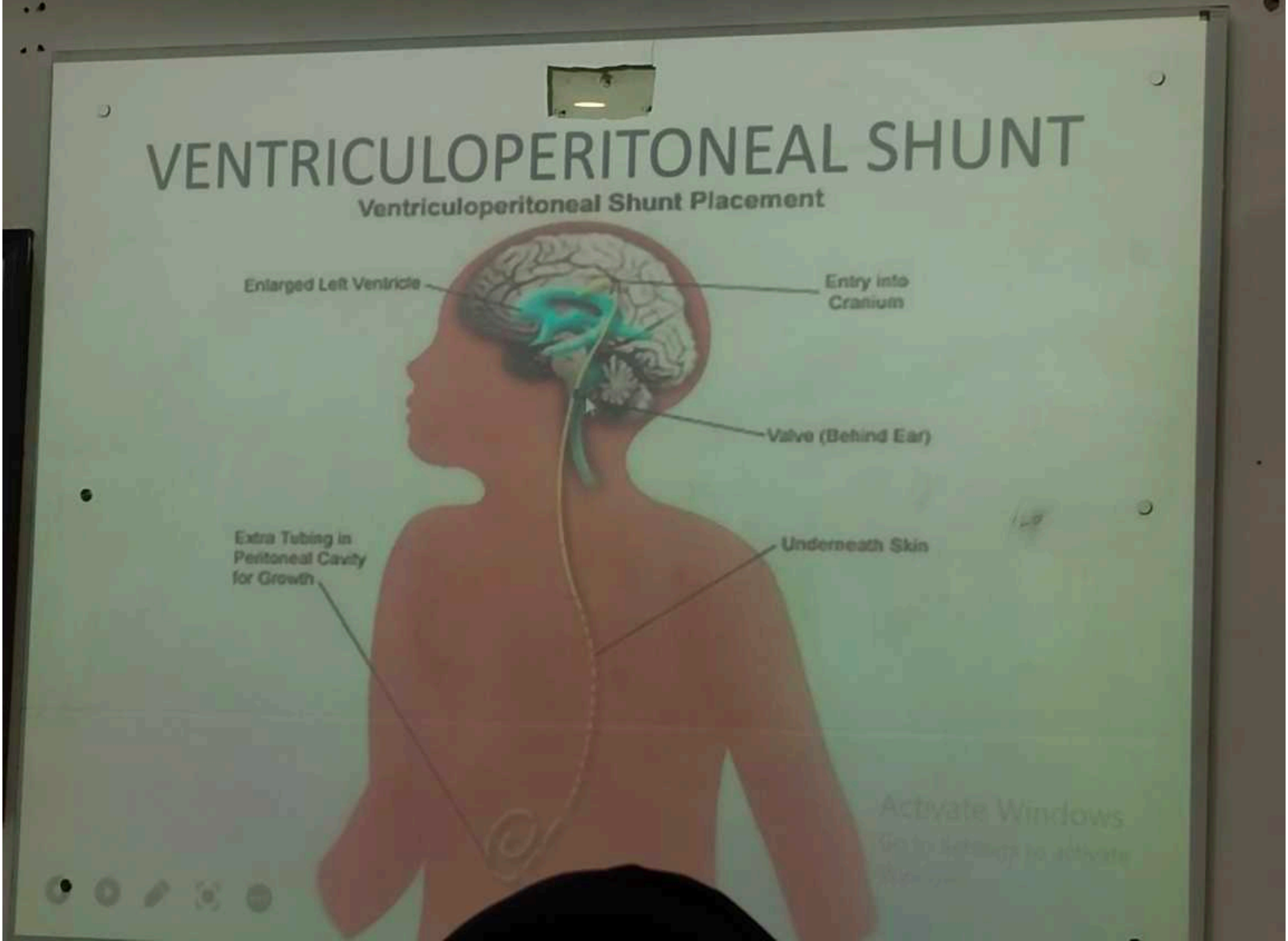
Venticulostomy

- Endoscopic procedure
- Involves fenestration of floor of the 3rd ventricle so that it communicates with basal cisterns allowing reabsorption of CSF
- Contraindications – abnormal ventricular anatomy, IVH, meningitis
- Complications – injury to basilar artery, CVS instability due to mid brain manipulation, ↑ ICP due to irrigation fluid leading to hypertension and tachycardia

Shunt

- Thin, flexible tube that directs the flow of CSF from the ventricles to another place in the body where it can be absorbed
- All parts of the shunt are under the skin
- Relieves the pressure & prevents the condition from getting worse
- It does not cure hydrocephalus

VENTRICULOPERITONEAL SHUNT



TAKE HOME MESSAGE

Spina bifida should always be classified according to their location and extent

Always look for associated congenital anomalies

- Closely monitor for signs and symptoms of raised Intracranial Pressure

Surgery is the mainstay of treatment for both spina bifida and hydrocephalus

Folic acid supplementation to mothers is essential