

# **MICROCEPHALY and HYDROCEPHALUS**

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# Objectives:

## The student should be able to:

1. Define and identify causes of micro and macrocephaly.
2. Discuss the physiology of CSF production.
3. Classify hydrocephalus and enumerate different causes of it.
4. Describe the clinical presentation, diagnosis and treatment of hydrocephalus.

# Microcephaly and Macrocephaly

## Definition:

- **Microcephaly** represents a head circumference below the 3rd percentile.
- **Macrocephaly** represents a head circumference above the 97th percentile.

# MICROCEPHALY

## Causes:

- Premature closure of skull sutures (**craniosynostosis**), rare.
- Usually due to **small brain due to insult** (infectious, toxic, metabolic, vascular) e.g. rubella, CMV, Fetal alcohol syndrome
- **Genetic disorder:** microcephaly vera (AR), trisomies 21, 18, 13, ....

# MACROCEPHALY

## Causes:

- **Macrocrania** (increased skull thickness) due to (bone metabolism or hypertrophy of bone marrow secondary to hemolytic anemia).
- **Hydrocephalus** (enlarged ventricles).
- **Megalencephaly** (enlarged brain) due to:
  - Disorders causing proliferation of brain tissue (e.g. neurofibromatosis, tuberous sclerosis)
  - An accumulation of abnormal metabolic substances (Tay Sach's, mucopolysaccharidoses).

# Hydrocephalus

## Definition:

- An excessive amount of cerebrospinal fluid (CSF) and is dilated because of increased pressure

## PHYSIOLOGY:

- CSF is produced - choroid plexus.
- Circulates through the ventricular system
- Absorbed into the systemic circulation.

# Hydrocephalus

## CSF production:

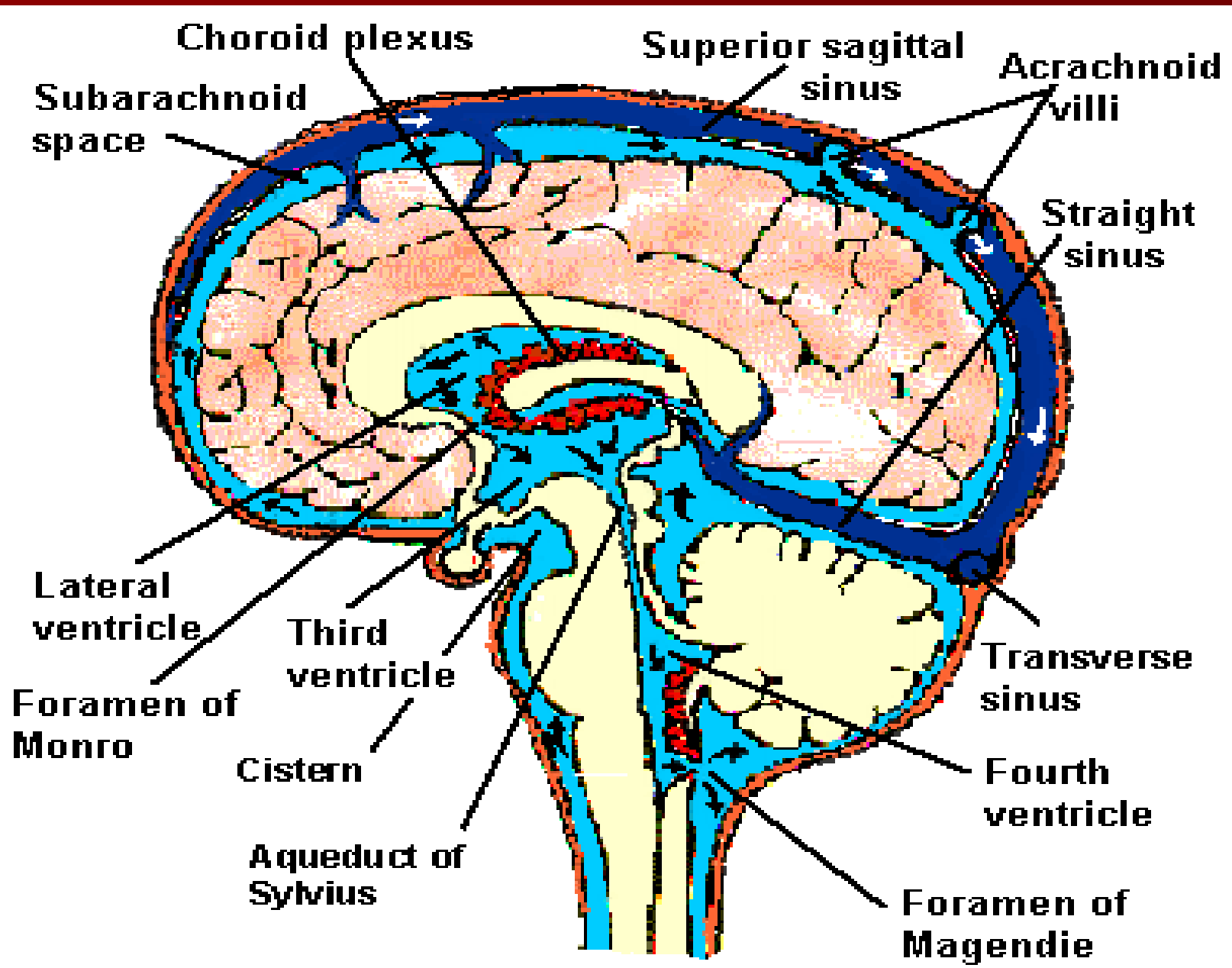
- Choroid plexus - located in the cerebral ventricles (also in 3<sup>rd</sup> and 4<sup>th</sup>).
- The volume: infants= 50 mL & 150 mL adults.
- CSF formation continues in raised intracranial pressure unless extremely high

# Hydrocephalus

## Ventricular system:

- Lateral ventricles via foramen of Monro to third.
- Third ventricle via aqueduct of Sylvius to fourth
- The fourth ventricle via two lateral foramina of Luschka and one midline foramen of Magendie, to subarachnoid spaces known as cisterns.
- CSF is absorbed via arachnoid villi into the sagittal sinus.





# Hydrocephalus

## PATHOGENESIS:

- Imbalance between production and absorption of CSF.
- **Three mechanisms:**
  1. Obstruction of CSF pathways (obstructive or non-communicating): most common.
  2. Impaired venous absorption.
  3. Over secretion of CSF.

# Choroid Plexus Papilloma



# Hydrocephalus

## Congenital causes:

- **Neural tube defects:** Arnold-Chiari malformation type 2.
- **Isolated aqueduct stenosis** (X-linked hydrocephalus).
- **Vein of Galen malformation** (compression of the aqueduct of Sylvius).
- **Intrauterine infection** (aqueduct stenosis).

# Congenital causes cont.

## ■ **The Dandy-Walker malformation:**

Classic triad:

- Complete or partial agenesis of the vermis.
- Cystic dilation of the fourth ventricle.
- Enlarged posterior fossa.

= Leads to secondary obstruction of the foramina of Luschka and Magendie.

# Acquired hydrocephalus:

- **CNS infections** (Pneumococcal meningitis/tuberculosis).
- **Hemorrhage** into the subarachnoid space.

# Hydrocephalus

## CLINICAL FEATURES:

(Depends on the age, the lesion, the duration and rate of ICP).

- **Headache** is a prominent symptom in older.
- **Changes** in personality and behavior.
- **Increased ICP** (in the posterior fossa often leads to nausea, vomiting and decreased appetite).

# CLINICAL FEATURES cont.

## In infants:

- A large head or rapid increase in the head size.
- A bulging anterior fontanel.
- Sun-setting of the eyes (Pressure on the midbrain may result in impairment of upward gaze).
- Developmental delay.
- Poor feeding, irritability, reduced activity and vomiting.



# CLINICAL FEATURES cont.

## In older children and adults:

- Blurred or double vision.
- Sun-setting of the eyes.
- Problems with balance, coordination or gait
- Developmental delay or loss of development.
- Change in personality

# Hydrocephalus

## Physical examination ( ICP):

- **Head size.**
- The **anterior fontanel** full or distended.
- The **scalp veins** dilated and prominent.
- **Setting-sun sign.**
- **A cracked pot sound or MacEwen's sign** (indicating separation of the sutures).
- **Papilledema.**
- Stretching of motor cortex may results in spasticity of the extremities, especially the legs).





# Hydrocephalus

## Complications:

- Intellectual impairment
- Neurological damage.
- Problems with the surgical shunt.
- Infection at the site of the shunt

# HYDROCEPHALUS

## Diagnostic Investigations:

- **Ultrasound of brain** (anterior fontanel):
  - Shows ventricular enlargement
- **CT/MRI of head:**
  - Shows ventricular enlargement, periventricular lucency, narrow/absent sulci ± 4<sup>th</sup> ventricular enlargement.

# HYDROCEPHALUS

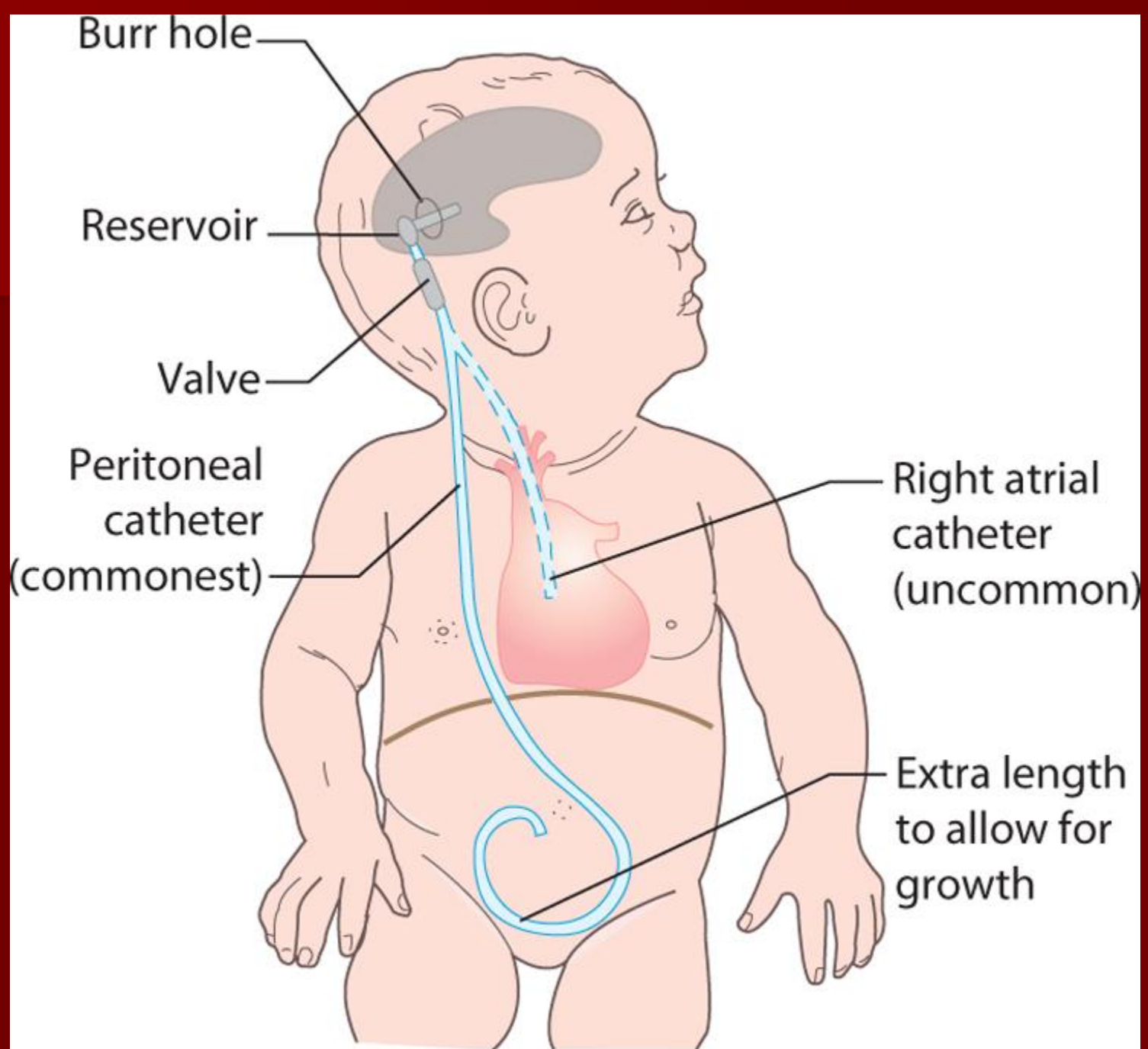
## Treatment:

- Serial Spinal taps.
- Surgery- remove obstruction if possible.
- Endoscopic third ventriculostomy (ETV)
- Shunts.
- Acetazolamide (decreases blood flow to choroidal arteries , therefore decreasing CSF production).

# Surgical shunts:

- Ventriculo Peritoneal Shunt
- Ventriculo Pleural Shunt
- Ventriculo Atrial Shunt.
- Lumboperitoneal shunt.





# Complications of a Shunt

- Shunt blockage.
- Shunt infection.
- Over drainage/Siphon effect.
- Hemorrhage along the tract.
- Shunt migration/disconnection.
- Seizures.