

# CNS Pathology

## lecture 1

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# Lecture 1

- Malformations
- Edema and parenchymal injury

# malformations

- Incidence: 1-2%
- Complications: mental retardation, and cerebral palsy .
- More in the setting of multiple brain defects.

# etiology

- prenatal or perinatal insults ... interfere with normal CNS development or cause tissue damage.
- Insults **early** during gestation cause more severe damage.
- types of insults: **gene mutations , chemicals infections.**

- Not all developmental disorders cause specific gross or microscopic findings, but they can produce mental problems.

# Types of malformations

- Neural tube defects.. MOST COMMON
- Forebrain malformations
- Posterior fossa defects
- Spinal cord anomalies.

# Neural tube defects

- Neural tube (NT) is formed early in the development of the brain.
- NT gives rise to the ventricle, brain and spinal cord.
- Anterior part of NT..brain
- Posterior part.. Spinal cord.
- Partial failure or reversal of NT closure.. Malformation.

# NT defects

- Recurrence in subsequent pregnancies: 4-5%
- So.. Possible **genetic** component.
- **Folate deficiency** early in gestation increase the risk.
- Prenatal folic acid .. Reduce the risk by 70%.



# NT defects

- Most common form: **posterior end** of the NT..  
Where **spinal cord** forms.
- Can be mild or severe.
- Mildest form: Asymptomatic bony defects:  
**spina bifida occulta**

# Myelomeningocele

- Myelomeningocele : extension of CNS tissue through a defect in the vertebral column , mostly in lumbosacral region.
- Motor and sensory defects in lower extremities + problems with bowel and bladder control.

# myelomeningocele



# Defects of anterior part on NT

- Defect of the Anterior part of NT causes anencephaly and encephalocele.
- **Anencephaly**: absent brain and top part of the skull.
- **Encephalocele**: malformed CNS extending through a defect in the cranium.

# Forebrain malformations

- Volume of the brain can be affected in some malformations.
- Large: **megalencephaly**.
- Small: **microencephaly**.
- Microencephaly is more common than megalencephaly.
- Microencephaly associated with microcephaly.

# microcephaly



# microencephaly

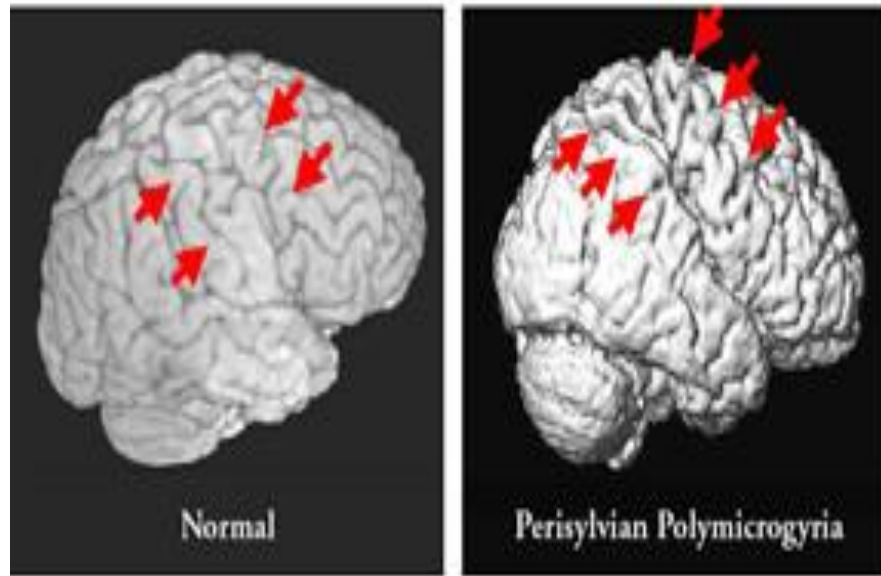
- Associated with: chromosomal anomalies , alcohol, HIV.
- Decreased generation of cortical neurons.

# Disturbed neuronal differentiation

- Disturbed neural migration and differentiation .. Problem in gyration and the six layered neocortical architecture.
- 1. Lissencephaly= **agyria** .. Smooth surface and only four layers.
- 2. **Patchy gyria**
- 3. **polymicrogyria**: cobblestone appearance.



# polymicrogyri



# Posterior fossa abnormalities

- Cerebellum affected.
- **Arnold –Chiari malformation**... small posterior fossa, mis-shaped cerebellum, downward extension of the vermis through foramen magnum.
- **Chiari tupe 1** malformation.. Milder.
- **Dandy- Walker** malformation.. Enlarged posterior fossa and absent cerebellar vermis.

# Spinal cord abnormalities

- **Hydromyelia** : expansion of the central canal of the cord.
- **Syringomyelia**: fluid filled clefts in the inner portion of the cord.

- BRAIN EDAMA AND HYDROHEPAHALUS

# Brain edema

- Cerebral edema: accumulation of excess fluid within the brain parenchyma.
- Two types: vasogenic and cytotoxic edema..  
Usually coexist

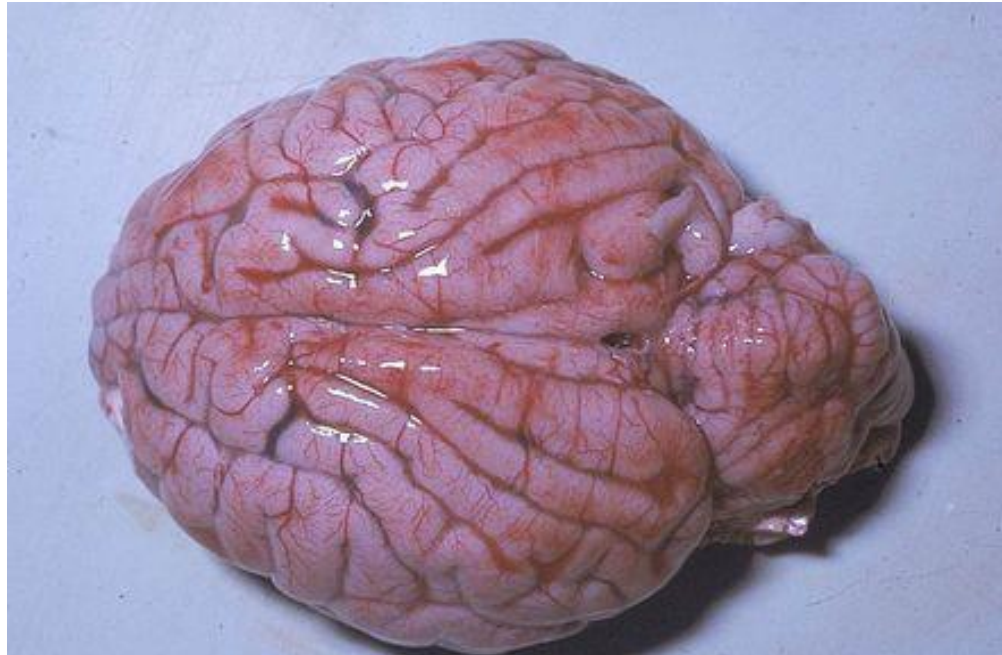
# Vasogenic edema

- Due to disruption of blood brain barrier.
- So: shift of fluids from vessels to brain tissue.
- Can be generalised or localised ( inflammation or tumors).

# Cytotoxic edema

- Due to neuronal or glial cell membrane injury.
- Causes: toxins or hypoxia.

# Brain edema





# hydrocephalus

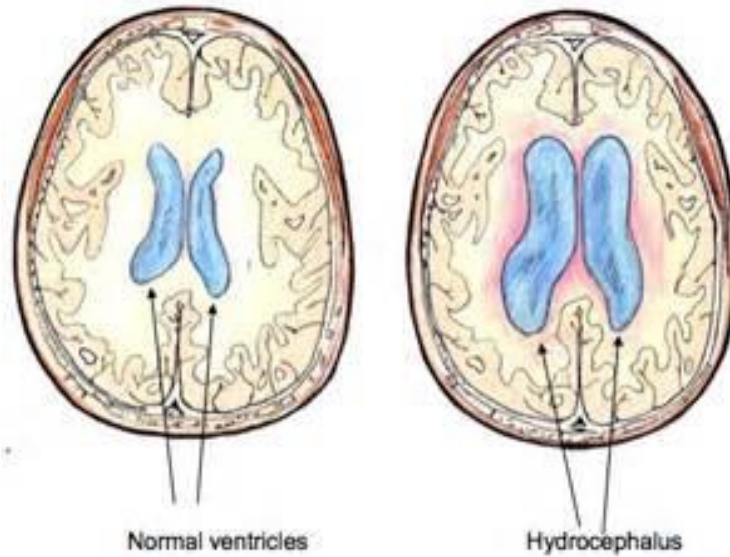
- Increased CSF within ventricles.
- Caused by overproduction or decreased resorption of CSF.
- Overproduction: rare, due to choroid plexus tumors.
- Decreased resorption.. Can be localised or generalised.

- Localised causes: tumors...
- Localised: **noncommunicating** hydrocephalus.
- Generalised: **communicating** hydrocephalus.

# hydrocephalus



# hydrocephalus



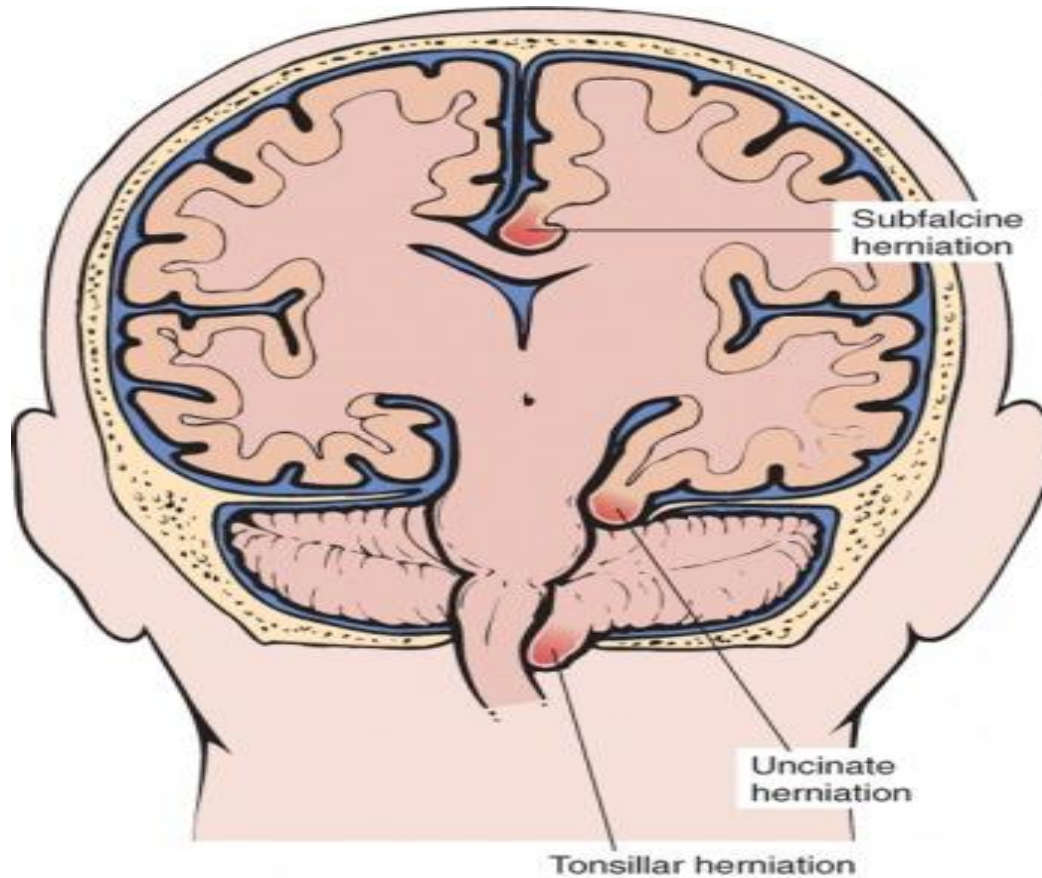
# herniation

- Increased volume of tissue inside the skull.. Increased intracranial pressure which causes focal expansion of the brain tissue .
- Because the cranial vault is subdivided by rigid dural folds (falx and tentorium).... The expanded brain tissue is displaced in relation to these folds.
- Expantion: herniation

# herniation

- Subfalcine = cingulate
- Transtentorial = uncinata
- Tonsillar.

# herniation



# Cingulate herniation

- cingulate gyrus displaced under edge of falx
- Can cause compression of anterior cerebral artery



# Transtentorial herniation

- Medial aspect of temporal lobe compressed against the free margin of the tentorium.
- **Third cranial nerve** compressed.. Dilated pupil, impaired ocular movement on the side of the lesion
- **Posterior cerebral artery** can be affected.. Ischemic injury to tissues supplied by it including visual cortex.

# Tonsillar herniation

- Displaced cerebellar tonsils through foramen magnum
- Brain stem compression... respiratory and cardiac centres in medulla compromised.
- **LIFE THREATENING**