أهلا وسهلا

الأستاذ الدكتور يوسف حسين

كلية الطب - جامعة مؤتة – الأردن

دكتوراة من جامعة كولونيا المانيا

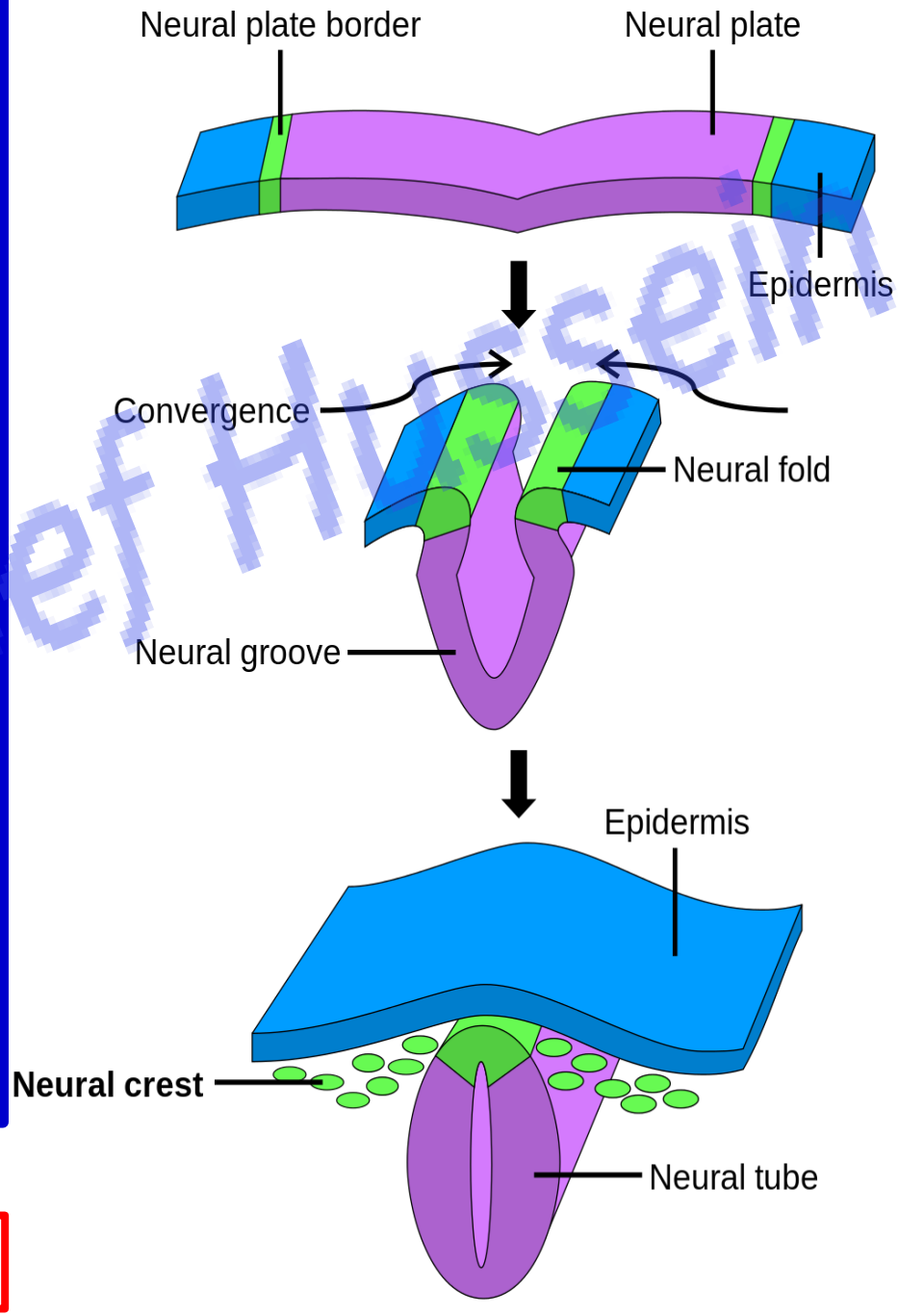
الواتس (أي استفسار)
00201224904207

Development of the neural tube:

- The neural tube develops as **neural plate** from the **ectoderm** dorsal to the notochord.
- A **neural pit** that forming the **neural groove**.
- The edges of the neural groove fused forming the **neural tube** that later on **separated** from the ectoderm.
- Neural tube lies opposite developed somites:

a- The part **cranial to the 4th somite** is dilated and forms the **brain**.

b- The part **caudal to the 4th somite** remains narrow and forms **the spinal cord.**



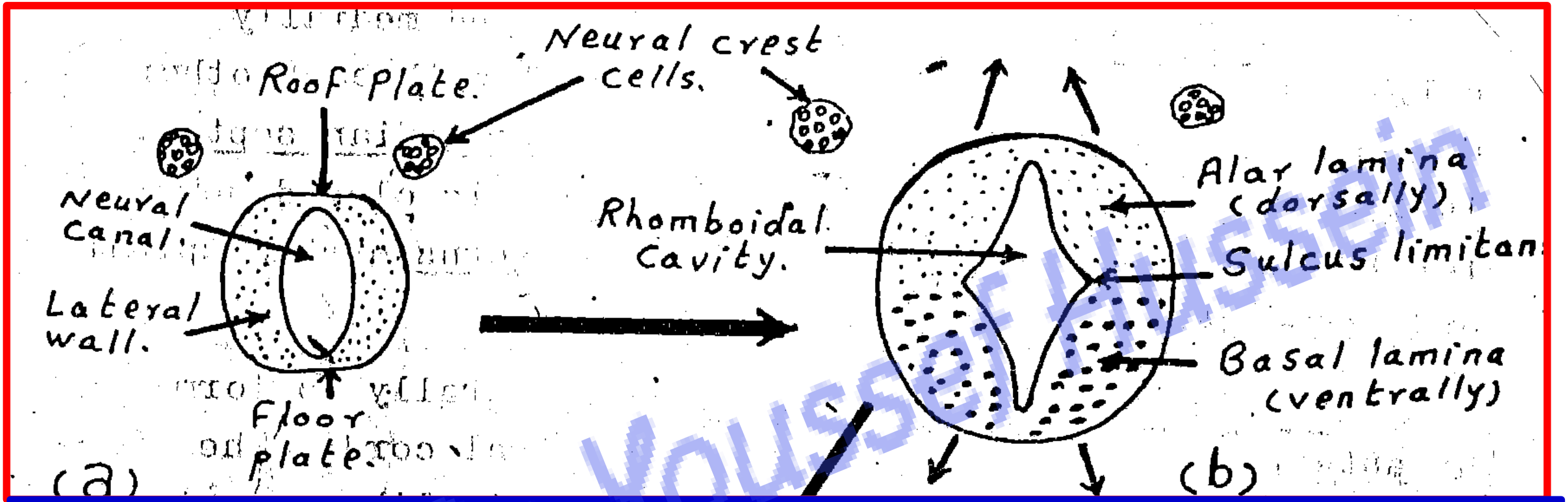
- **Central and peripheral nervous systems origins**

- **Neuroepithelia in neural tube—CNS neurons, CNS glial cells (astrocytes, oligodendrocytes, ependymal cells).**

- **Neural crest EMO PASSES**

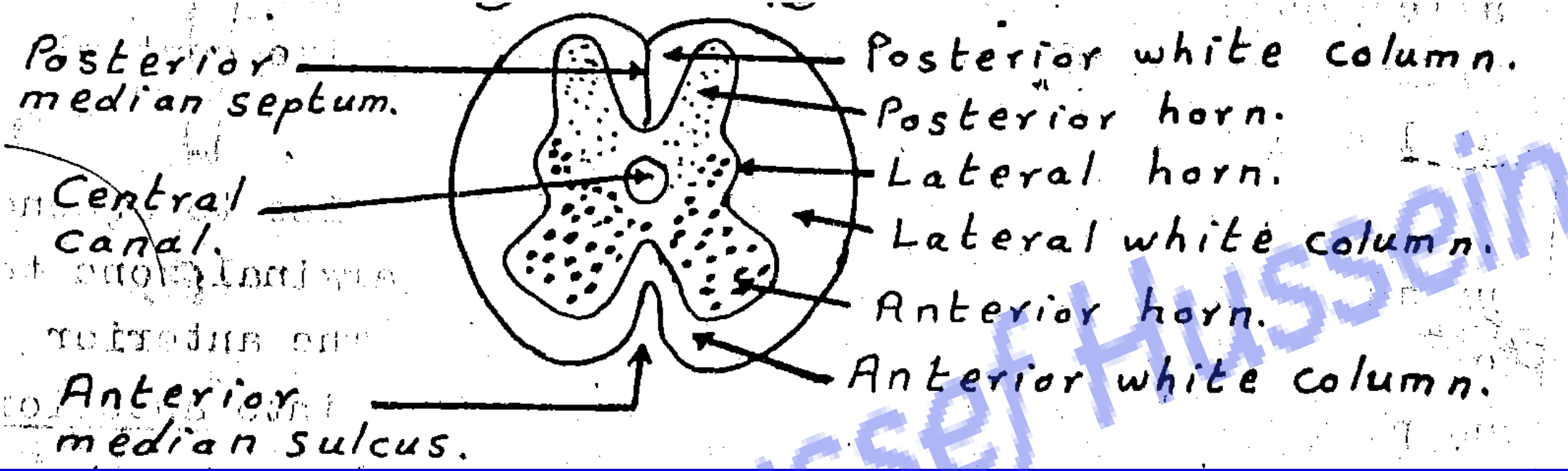
1. **Enterochromaffin cells** in the epithelial layer of the entire gastrointestinal tract
2. **Melanocytes (Pigment cells in the skin, iris and retina).**
3. **Odontoblasts (**outer layer of the dental pulp)
4. **PNS ganglia (cranial, dorsal root, autonomic)**
5. **Adrenal medulla (adrenal cortex is mesoderm)**
6. **Schwann cells**
7. **Spiral membrane (aorticopulmonary septum)**
8. **Endocardial cushions (also derived partially from mesoderm)**
9. **Skull bones** (frontal, ethmoid, and sphenoid bones derives from ectodermal neural crest and others from mesoderm)**.**
10. **Pia and arachnoid matters of the meninges (dura matter is mesoderm).**

Development of Spinal cord



** **At first**, the **neural tube** has thick lateral wall, thin roof plate and floor plate, and a narrow slit-like lumen.

- **The ventral and dorsal parts** of the lateral wall become thick by proliferation of the cells in the mantle zone. As a result, it is divided by lateral sulcus called sulcus limitans into ventral part (**basal lamina**) and dorsal part (**alar lamina**) and the cavity becomes rhomboidal in shape:



A- Ventral parts: contain motor cells, form the anterior and the lateral horns of the spinal cord.

- The **anterior horns** are found in all segments.
- The **lateral horns** are found in;

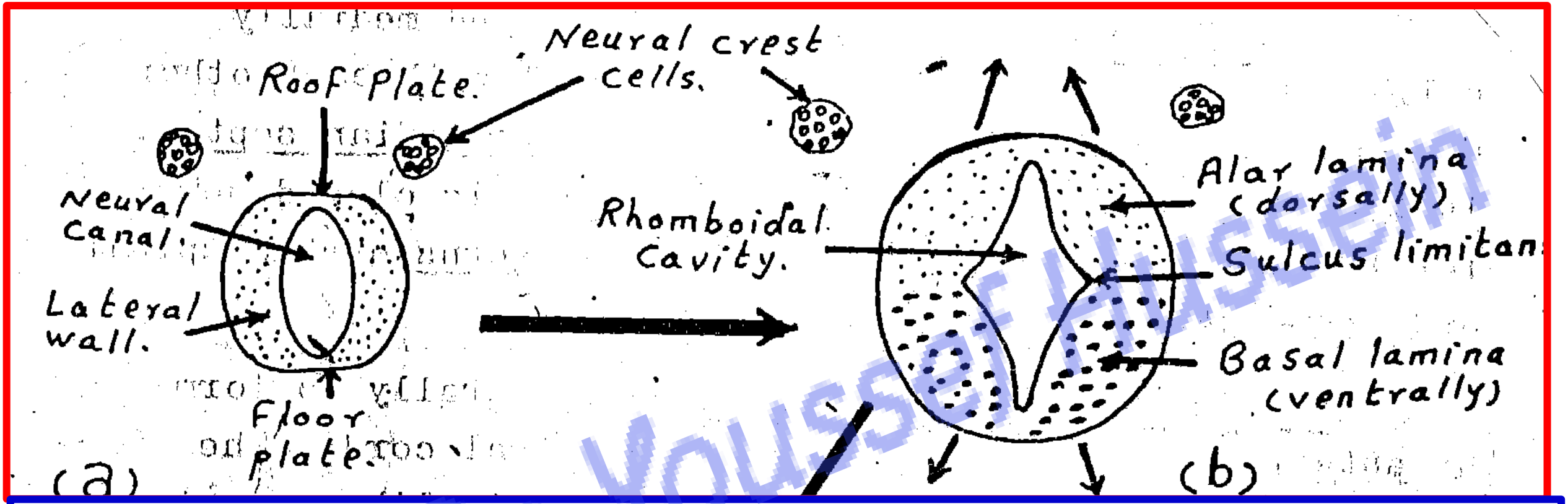
  1) All the thoracic and upper 2-3 lumbar segments (sympathetic).

  2) In the 2nd, 3rd and 4th sacral segments (parasympathetic).

B- Dorsal parts: contain the sensory cells, form the posterior horns of the spinal cord

C- The cavity becomes reduced in size (narrow) to form the central canal.

D- Marginal layer: the outer layer containing ascending and descending tracts (**white matter** of the spinal cord, anterior, lateral and posterior columns).



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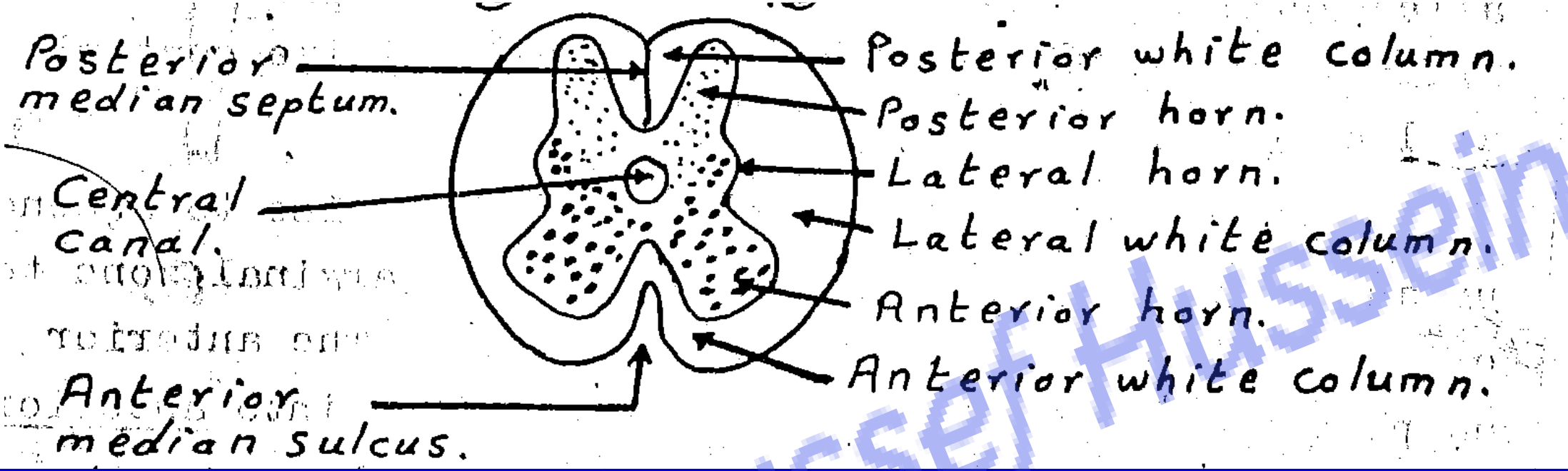

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• Congenital anomalies of the spinal cord

Spina bifida: failure of fusion of neural arch of vertebra around the spinal cord.

**** Types of spina bifida:**

a- Spina bifida occulta: bifid spines of the vertebra but no herniation. Dura is intact. Usually seen at lower vertebral levels. Associated with tuft of hair or skin dimple at level of bony defect. No increase AFP.

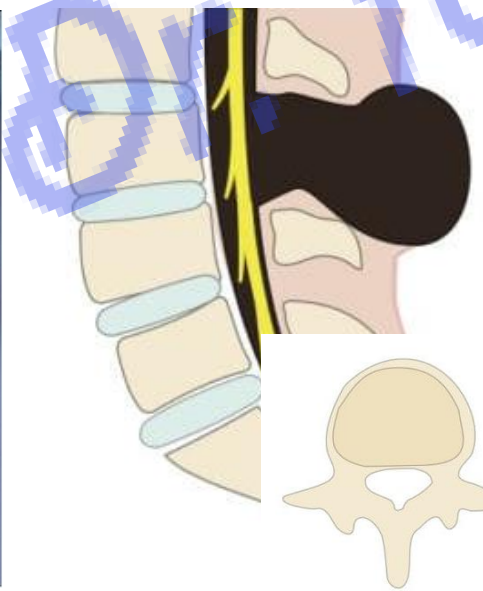
b- Meningocele; bulge of the meninges through the spina bifida (increase AFP).

c- Meningomyelocele; bulge of the meninges and spinal cord through the spina bifida. (increase AFP).

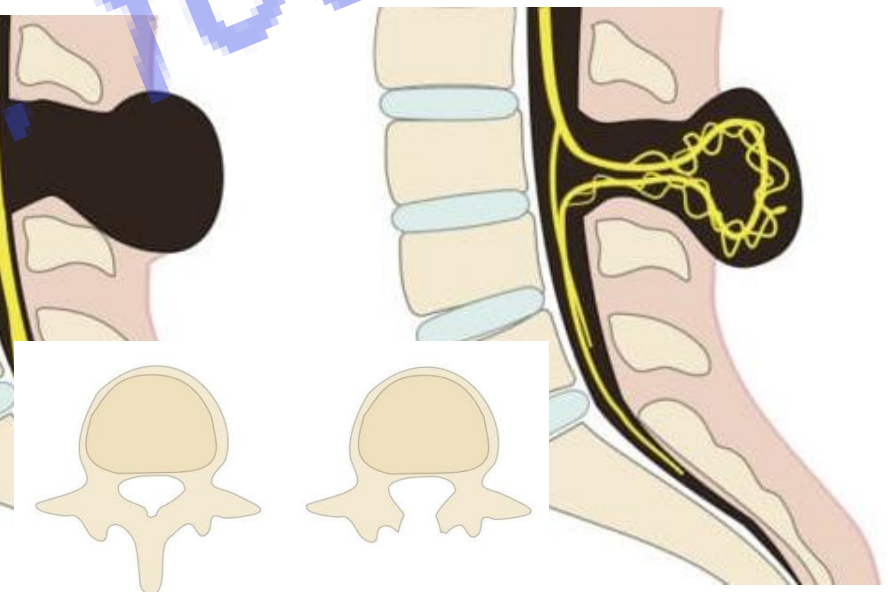
d- Myelocele; the spinal cord is exposed directly to the spina bifida. (increase AFP).

Spina bifida occulta

Meningocele

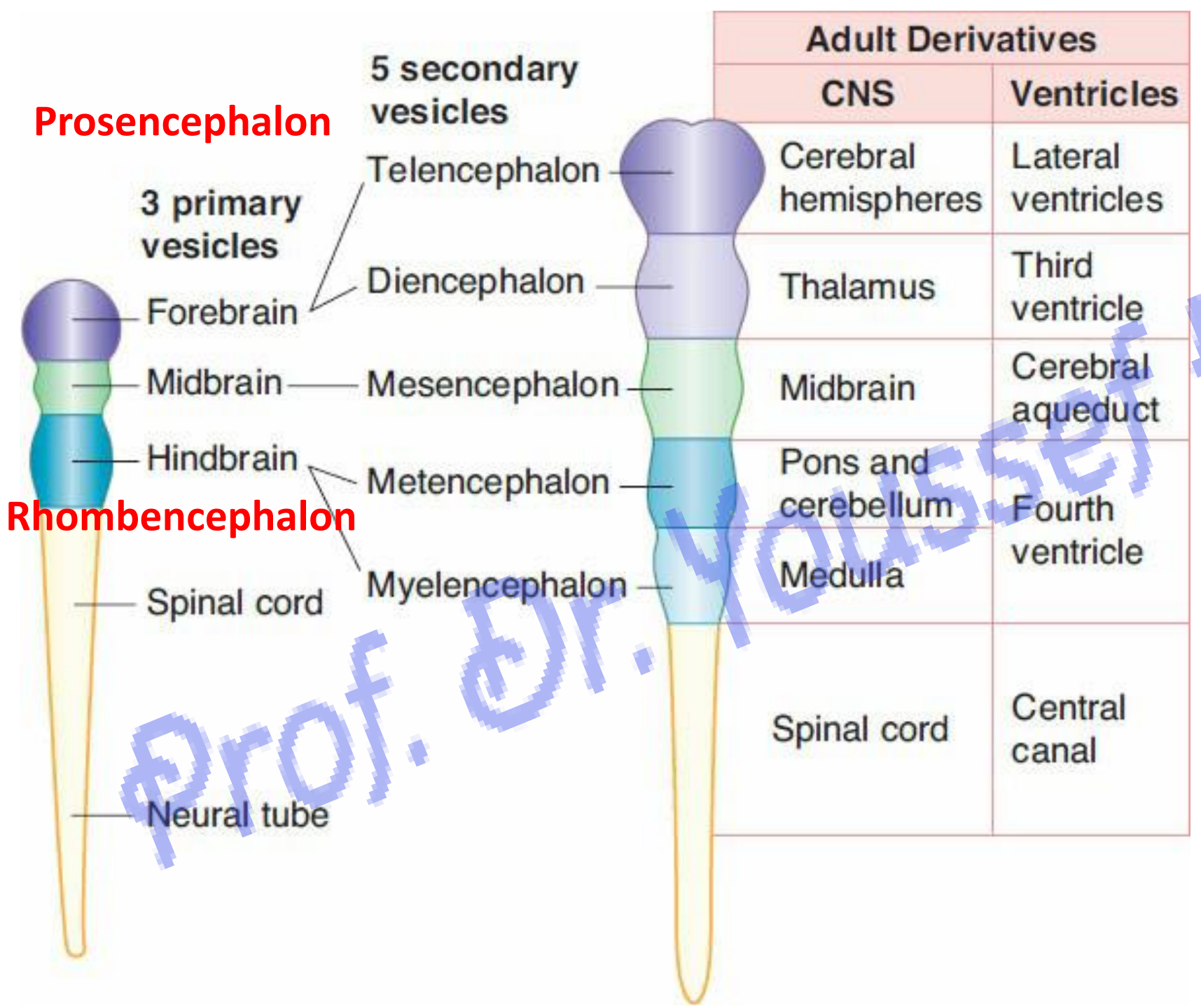


Meningomyelocele



Myelocele

Development of Brain



Adult Derivatives	
CNS	Ventricles
Cerebral hemispheres	Lateral ventricles
Thalamus	Third ventricle
Midbrain	Cerebral aqueduct
Pons and cerebellum	Fourth ventricle
Medulla	
Spinal cord	Central canal

Cerebral hemispheres, most of basal ganglia

Thalamus, hypothalamus, subthalamus, epithalamus (pineal gland), retina and optic nerve

Meningocele

herniation of a part of the meninges

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- **Meningoencephalocele** herniation of a part of the brain and its covering meninges.

- **Meningohydroencephalocele**: herniation of the meninges and part of the brain and its ventricle containing CSF

Meningoencephalocele

Anencephaly

- Failure of development of greater part of the brain and vault of the skull due to failure of cephalic part of the neural tube to close
- Incompatible with life
- Increase AFP during pregnancy
- **Often presents with polyhydramnios (decrease fetal swallowing due to lack of neural control).**

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- **Hydrocephalus**
  excessive accumulation of C.S.F in the ventricular system due to closure in the CSF circulation

- **Microcephaly**

  small skull and cerebral hemisphere

- **Cyclopia** one cerebral hemisphere, one ventricle and one median eye

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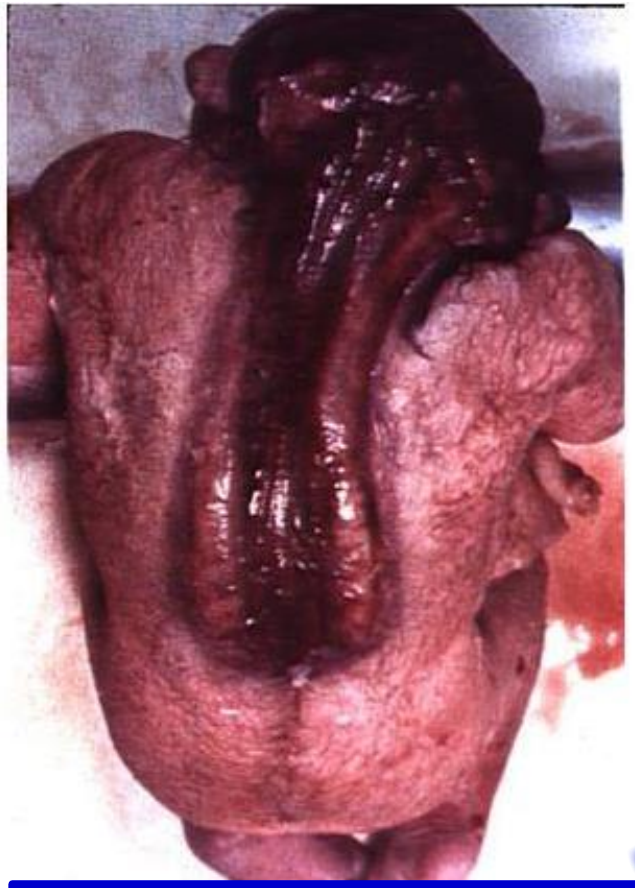


Rachischisis: spinal cord
Cranioschisis: brain

Craniorachischisis: brain & cord

- **Neural tube defects (NTD)**
- **Failure of neural tube to close completely by week 4 of development. Associated with maternal folate deficiency during pregnancy. Diagnosis: ultrasound, maternal serum AFP (increase during pregnancy).**

Craniorachischisis: Failure of closure of the neural tube, Exposed, unfused neural tissue without skin/meningeal covering, leading to longitudinal cleft in the back of the head and vertebral column

- **Brain malformations**

- **Often incompatible with postnatal life.**
- **Survivors may be profoundly disabled.**

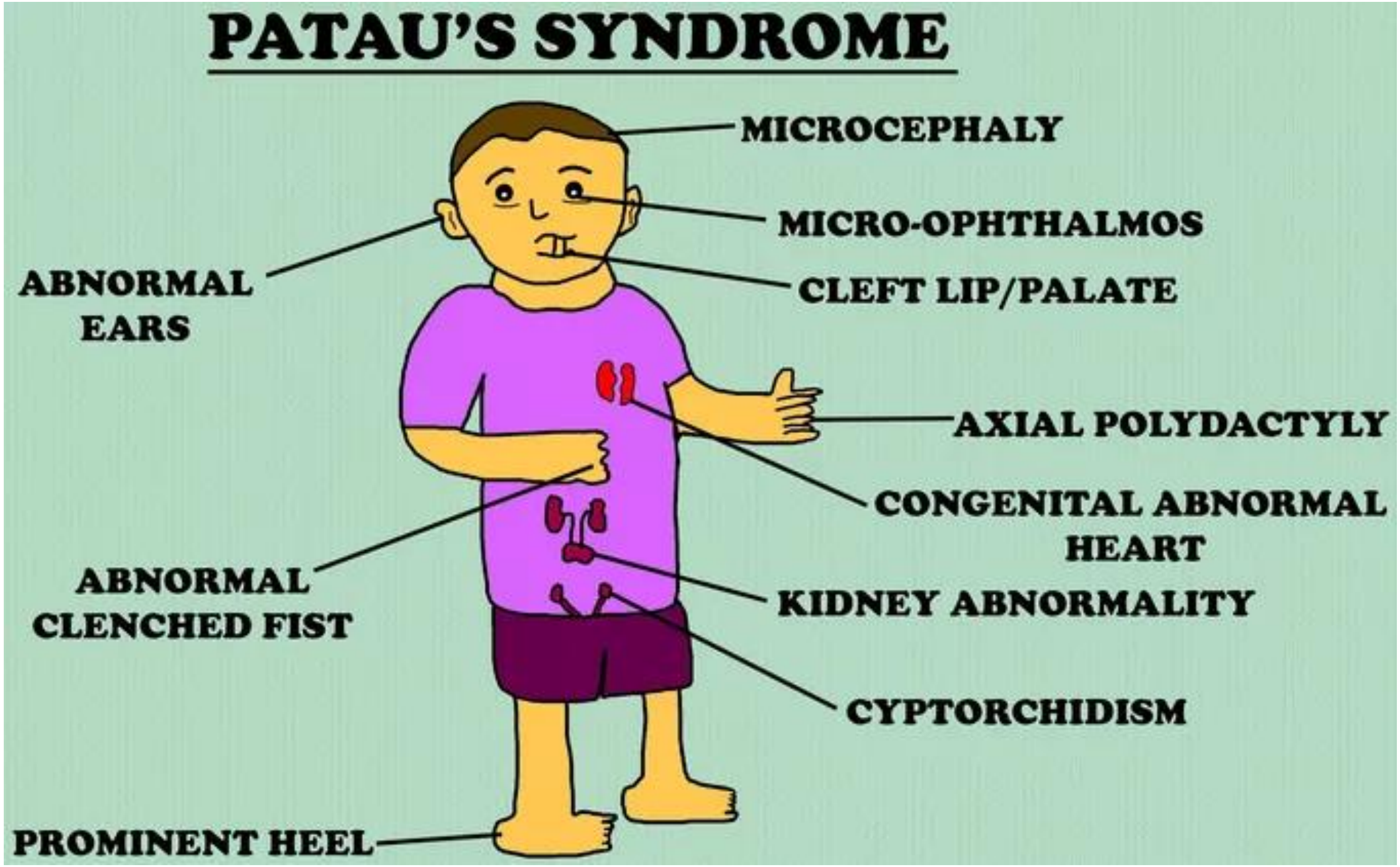
- **Holoprosencephaly**

  - **Failure of forebrain to divide into 2 cerebral hemispheres;**
  - **developmental field defect usually occurring at weeks 3–4 of development.**
  - **Associated with *SHH* mutations.**
  - **May be seen in Patau syndrome (trisomy 13), fetal alcohol syndrome.**
  - **Presents with midline defects: monoventricle, fused basal ganglia, cleft lip/palate, hypotelorism, cyclopia, proboscis (long, bendable nose).** nose-like appendage
  - **Increase risk for pituitary dysfunction (eg, diabetes insipidus).**

hej¸hôg

Sonic HedgeHog gene

PATAU'S SYNDROME

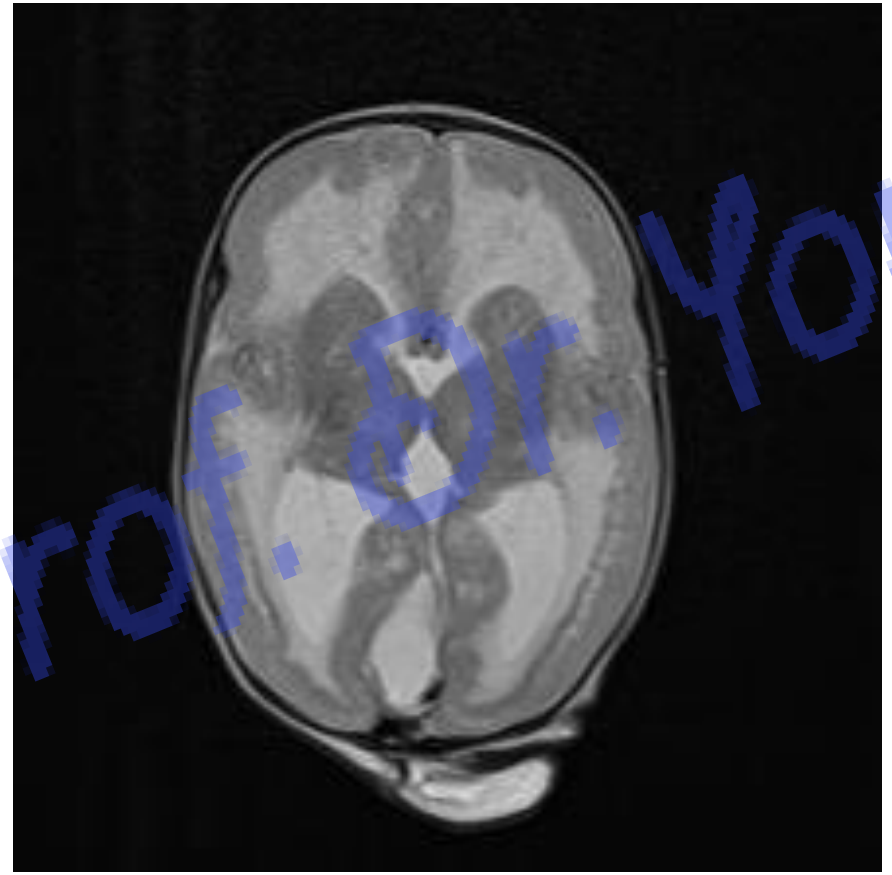


• Fetal alcohol syndrome

- **One of the leading preventable causes of intellectual disability in the US. 2° to maternal alcohol use during pregnancy.**
- **Newborns may present with developmental delay, microcephaly, facial abnormalities (eg, smooth philtrum, thin vermillion border, small palpebral fissures, flat nasal bridge), limb dislocation, heart defects.**
- **Holoprosencephaly may occur in more severe presentations.**
- **One mechanism is due to impaired migration of neuronal and glial cells.**

Lissencephaly

- **Failure of neuronal migration--smooth brain surface that lacks sulci and gyri**
- **Presents with dysphagia, seizures, microcephaly, facial anomalies.**



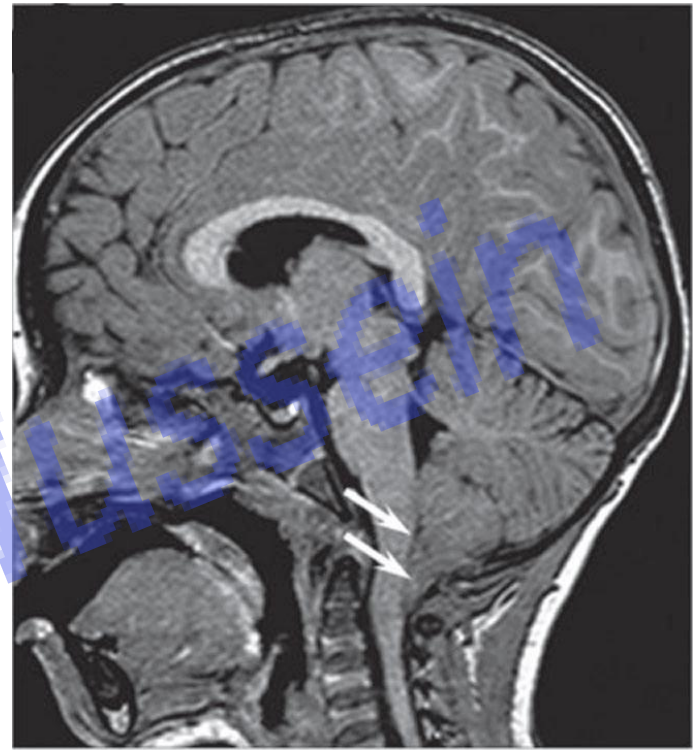
• Posterior fossa malformations

• Chiari I malformation

- Downward displacement of cerebellar tonsils through foramen magnum (1 structure).
- Usually asymptomatic in childhood, manifests in adulthood with headaches and cerebellar symptoms.
- Associated with spinal cord cavitations (eg, syringomyelia).

• Chiari II malformation

- Downward displacement of cerebellum (vermis and tonsils) and medulla (2 structures) through foramen magnum -- noncommunicating hydrocephalus.
- More severe than Chiari I, usually presents early in life with dysphagia, stridor, apnea, limb weakness.
- Associated with Meningomyelocele (usually lumbosacral).



Tonsil in the Foramen magnum

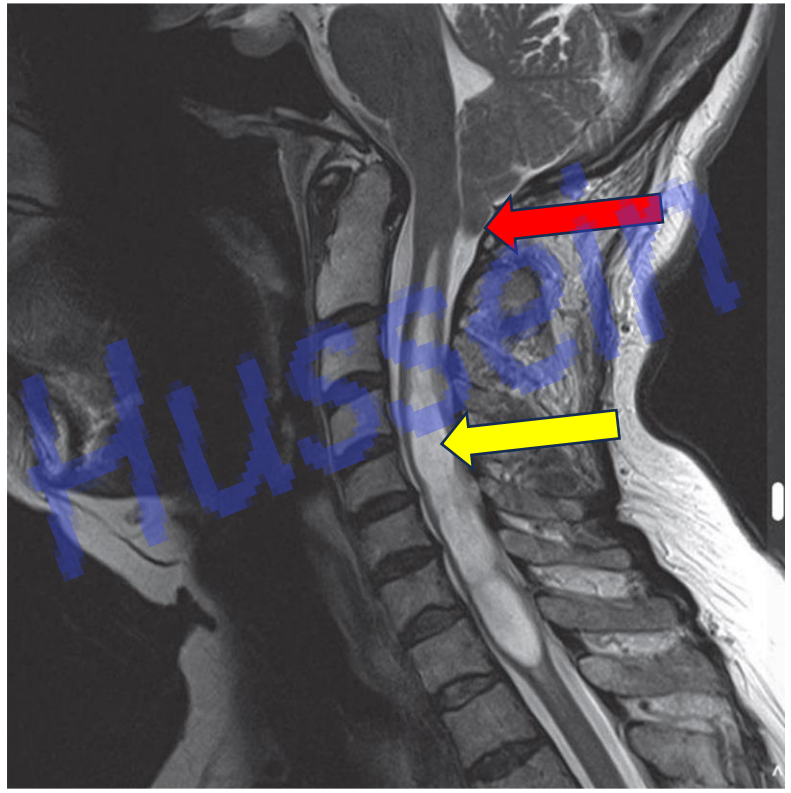
- **Posterior fossa malformations**
  - **Dandy-Walker malformation**

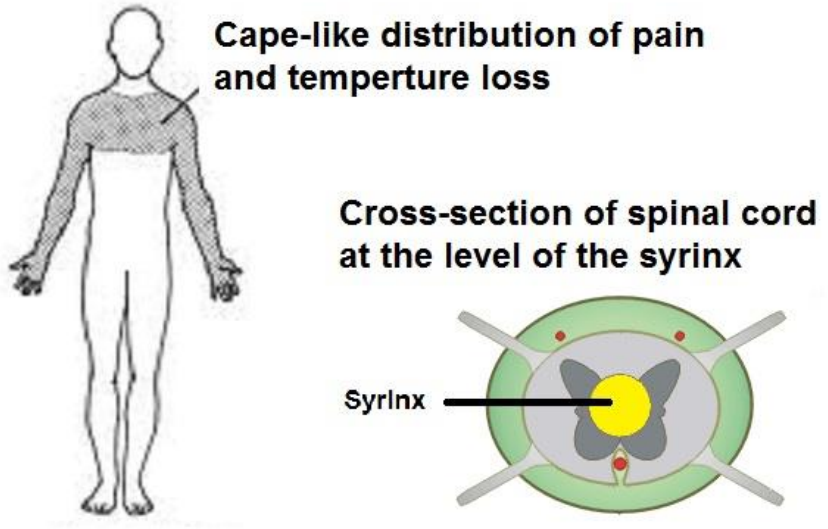
- **Agenesis of cerebellar vermis -- cystic enlargement of 4th ventricle that fills the enlarged posterior fossa.**
- **Associated with noncommunicating hydrocephalus, spina bifida.**

Enlargement of the 4th ventricle

Syringomyelia

- *Syrinx* (Greek) = tube, as in "syringe."
- Fluid-filled, gliosis-lined cavity within spinal cord (yellow arrow)
- Fibers crossing in anterior white commissure (spinothalamic tract) are typically damaged first Ž"cape like" loss of pain and temperature sensation in bilateral upper extremities.
- As lesion expands it may damage anterior horns Ž LMN deficits.
- Most lesions occur between C2 and T9.
- Usually associated with Chiari I malformation (red arrow)
- Less commonly, associated with other malformations, infections, tumors, trauma.





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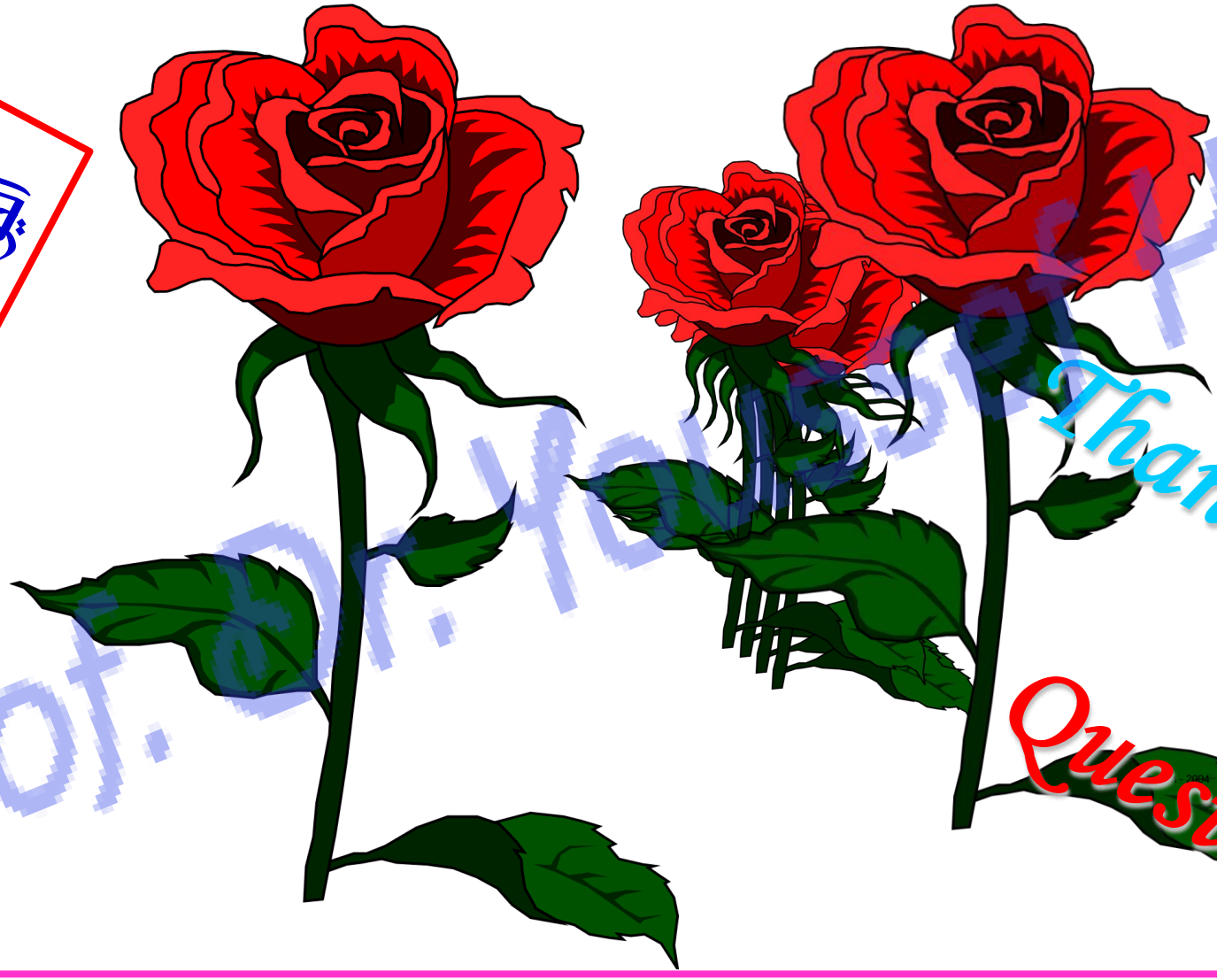

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