

Central Nervous System Module -I

2nd Year MBBS(LGIS) **Development of Cranium**



Presenter: Prof. Dr. Ifra Saeed

Date: 00-00-25



First Ten Minutes

Prof. Umar's Model of Teaching Strategy Self Directed Learning Assessment Program

Objectives : To cultivate critical thinking, analytical reasoning, and problemsolving competencies.

To instill a culture of self-directed learning, fostering lifelong learning habits and autonomy.

How to Assess?

- Ten randomly selected students will be evaluated within the first 10 minutes of the lecture through 10 multiple-choice questions (MCQs) based on the PowerPoint presentation shared on Students Official WhatsApp group, one day before the teaching session.
- The number of MCQs from the components of the lecture will follow the guidelines outlined in the **Prof. Umar model of Integrated Lecture**.

Component	Core	Horizontal	Vertical	Spiral
of LGIS	Knowledge	Integration	Integration	Integration
No of MCQs	6-7	1-2	1	1

Professor Umar Model of Integrated Lecture



Motto

Vision; ThDream/Tomorrow



- To impart evidence based research oriented medical education
- To provide best possible patient care
- To inculcate the values of mutual respect and ethical practice of medicine

Learning Objectives

At the end of the session, student will be able to

- Describe development of Cranium
- Discuss development of Visceral and Neurocranium Cranium
- Discuss congenital abnormalities associated with Cranium
- Integrate physiological and biochemical aspects with development of Midbrain
- Approach towards patient of Microcephaly
- Correlate and build core knowledge on the basis of latest research
- Bioethics related to microcephalic patient

Cranium The Viscerocranium vs. Neurocranium

2 main parts that fuse together

The Skull = 22 bones

viscerocranium

- Facial skeleton and bones of the jaw
- ~14 bones

e.g. nose, mouth, cavities



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Neurocranium

- Protects the brain
- 8 bones

Development of the Cranium

The cranium (skull) develops from mesenchyme around the developing brain. The cranium consists of:

- The neurocranium, a protective case for the brain
- The viscerocranium, the ske

Core Knowledge

Bones of Skull



Development of Neurocranium Cranium

Cartilaginous NeurocraniumMembranous Neurocranium

Initially, the cartilaginous neurocranium or chondrocranium consists of the cartilaginous base of the developing cranium, which forms by fusion of several cartilages





 Later, endochondral ossification of the chondrocranium forms the bones in the base of the cranium. The ossification pattern of these bones has a definite sequence, beginning with the occipital bone, body of sphenoid, and ethmoid bone.



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The parachordal cartilage, or basal plate, forms around the cranial end of the Notochord and fuses with the cartilages derived from the sclerotome regions of the occipital somites. This cartilaginous mass contributes to the base of the occipital bone; later, extensions grow around the cranial end of the spinal cord and form the boundaries of the foramen magnum



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- The hypophysial cartilage forms around the developing pituitary gland (Latin, hypophysis cerebri) and fuses to form the body of the sphenoid bone.
- The trabeculae cranii fuse to form the body of the ethmoid bone, and the ala orbitalis forms the lesser wing of the sphenoid bone.



Otic capsules develop around the otic vesicles, the primordia of the internal ears and form the petrous and mastoid parts of the temporal bone. Nasal capsules develop around the nasal sacs and contribute to the formation of the ethmoid bone.



Membranous Neurocranium

Membranous Neurocranium consists of neural crest cells and paraxial mesoderm which form the main portion of the roof and lateral sides of the neurocranium.

Intramembranous ossification occurs in the mesenchyme at the sides and top of the brain, forming the calvaria (cranial vault).



During fetal life, the flat bones of the calvaria are separated by dense connective tissue membranes that form fibrous joints, the sutures



Molding

Six large fibrous areas-fontanelles-are present where several sutures meet. The softness of the bones and their loose connections at the sutures enable the calvaria to undergo changes of shape during birth, called molding.



Molding of Fetal Cranium

During molding of the fetal cranium (adaptation of fetal head to the pelvic cavity during birth), the frontal bones become flat, the occipital bone is drawn out, and one parietal bone slightly overrides the other one. Within a few days after birth, the shape of the calvaria returns to normal



Cartilaginous Vicserocranium

Most mesenchyme in the head region is derived from the neural crest.

Neural crest cells migrate into the pharyngeal arches and form the bones and connective tissue of craniofacial structures. Homeobox (Hox) genes regulate the migration and subsequent differentiation of the neural crest cells, which are crucial for the complex patterning of the head and face. The parts of the fetal cranium are derived from the cartilaginous skeleton of the first two pairs of pharyngeal arches

- The dorsal end of the first pharyngeal arch cartilage forms two middle ear bones, the malleus and incus.
- The dorsal end of the second pharyngeal arch cartilage forms the stapes of the middle ear and the styloid process of the temporal bone. Its ventral end ossifies to form the lesser horn (Latin, cornu) and superior part of the body of the hyoid bone.
- The third arch cartilages give rise to the greater horns and the inferior part of the body of the hyoid bone.
- The fourth pharyngeal arch cartilages fuse to form the laryngeal cartilages, except for the epiglottis

Membranous Viscerocranium

- Intramembranous ossification occurs in the maxillary prominence of the first pharyngeal arch and subsequently forms the squamous temporal, maxillary, and zygomatic bones.
- The squamous temporal bones become part of the neurocranium. The mesenchyme in the mandibular prominence of the first pharyngeal arch condenses around its cartilage and undergoes intramembranous ossification to form the mandible. Some endochondral ossification occurs in the median plane of the chin and in the mandibular condyle.

Newborn Cranium

- After recovering from molding, the newborn's cranium is rather round and its bones are thin. Like the fetal cranium, it is large in proportion to the rest of the skeleton, and the face is relatively small compared with the calvaria.
- The small facial region of the cranium results from the small size of the jaws, virtual absence of paranasal (air) sinuses, and underdevelopment of the facial bones.



Postnatal Growth of the Cranium

The fibrous sutures of the newborn's calvaria permit the brain to enlarge during infancy and childhood. The increase in the size of the calvaria is greatest during the first 2 years, the period of most rapid postnatal growth of the brain.

The calvaria normally increases in capacity until approximately 16 years of age. After this, it usually increases slightly in size for 3 to 4 years because of thickening of its bones..

- There is also rapid growth of the face and jaws, coinciding with eruption of the primary (deciduous) teeth. These facial changes are more marked after the secondary (permanent) teeth erupt.
- There is concurrent enlargement of the frontal and facial regions, associated with the increase in the size of the paranasal sinuses (e.g., frontal and ethmoid sinuses). Most paranasal sinuses are rudimentary or absent at birth. Growth of these sinuses is important in altering the shape of the face and in adding resonance to the voice

Kipple Feil Syndrome

The main features of this syndrome are shortness of the neck, low hairline, and restricted neck movements. In most cases, the number of cervical vertebral bodies is fewer than normal.

The number of cervical nerve roots may be normal but they are small, as are the intervertebral foramina. Individuals with this syndrome are often otherwise normal, but the association of this anomaly with other congenital anomalies is not uncommon.



Vertical Integration

ACRANIA

In this condition, the calvaria is absent and extensive defects of the vertebral column are often present

- Acrania associated with meroencephaly or anencephaly (partial absence of the brain) occurs approximately once in 1000 births and is incompatible with life.
- Meroencephaly results from failure of the cranial end of the neural tube to close during the fourth week. This anomaly causes subsequent failure of the calvaria to form



Craniosynostosis

- Prenatal closure of the cranial sutures results in the most severe abnormalities.
- The cause of craniosynostosis is unclear.
- Homeobox gene Msx2 and Alx4 mutations have been implicated in cases of craniosynostosis and other cranial defects.
- A recent epidemiologic study of maternal drug use found a strong association between anticonvulsant use during early pregnancy and infant craniosynostosis. These abnormalities are much more common in males than in females and are often associated with other skeletal anomalies.

Scaphocephaly

- The type of deformed cranium produced depends on which sutures close prematurely.
- If the sagittal suture closes early, the cranium becomes long, narrow, and wedge shaped-scaphocephaly
- This type of cranial deformity constitutes about half the cases of craniosynostosis.



Brachycephaly, Plagiocephaly, Trigonocephaly

- Another 30% of cases involve premature closure of the coronal suture, which results in a high, tower-like cranium-**brachycephaly**. If the coronal suture closes prematurely on one side only, the cranium is twisted and asymmetrical-**plagiocephaly**.
- Premature closure of the frontal (metopic) suture results in a deformity of the frontal bone and other anomalies-trigonocephaly



Fetal anomaly diagnosis and termination of pregnancy



Spiral Integration https://doi.org/10.1111/dmcn.15528Citation

Bioethics

Microcephaly measurement in adults and its association with clinical variables

Head circumference was related to cerebral volume. Due to its low cost and easy use, head circumference can be used as a screening test for microcephaly, adjusting it for gender and height. Microcephaly was associated with dementia at old age.

Adult; Microcephaly, classification; Cephalometry; Dementia; Data Mining

Reference: Rev. Saúde Pública 56 27 May 20222022 • https://doi.org/10.11606/s1518-8787.2022056004175

Spiral Integration

Family Medicine

Updates on Clinical and Genetic Heterogeneity of ASPM in 12 Autosomal Recessive Primary Microcephaly Families in Pakistani Population

 Microcephaly (MCPH) is a genetically heterogeneous disorder characterized by non-progressive intellectual disability, small head circumference, and small brain size compared with the age- and sex-matched population. MCPH manifests as an isolated condition or part of another clinical syndrome; so far, 25 genes have been linked with MCPH.

Reference: 2021 Jul 6. doi: 10.3389/fped.2021.695133