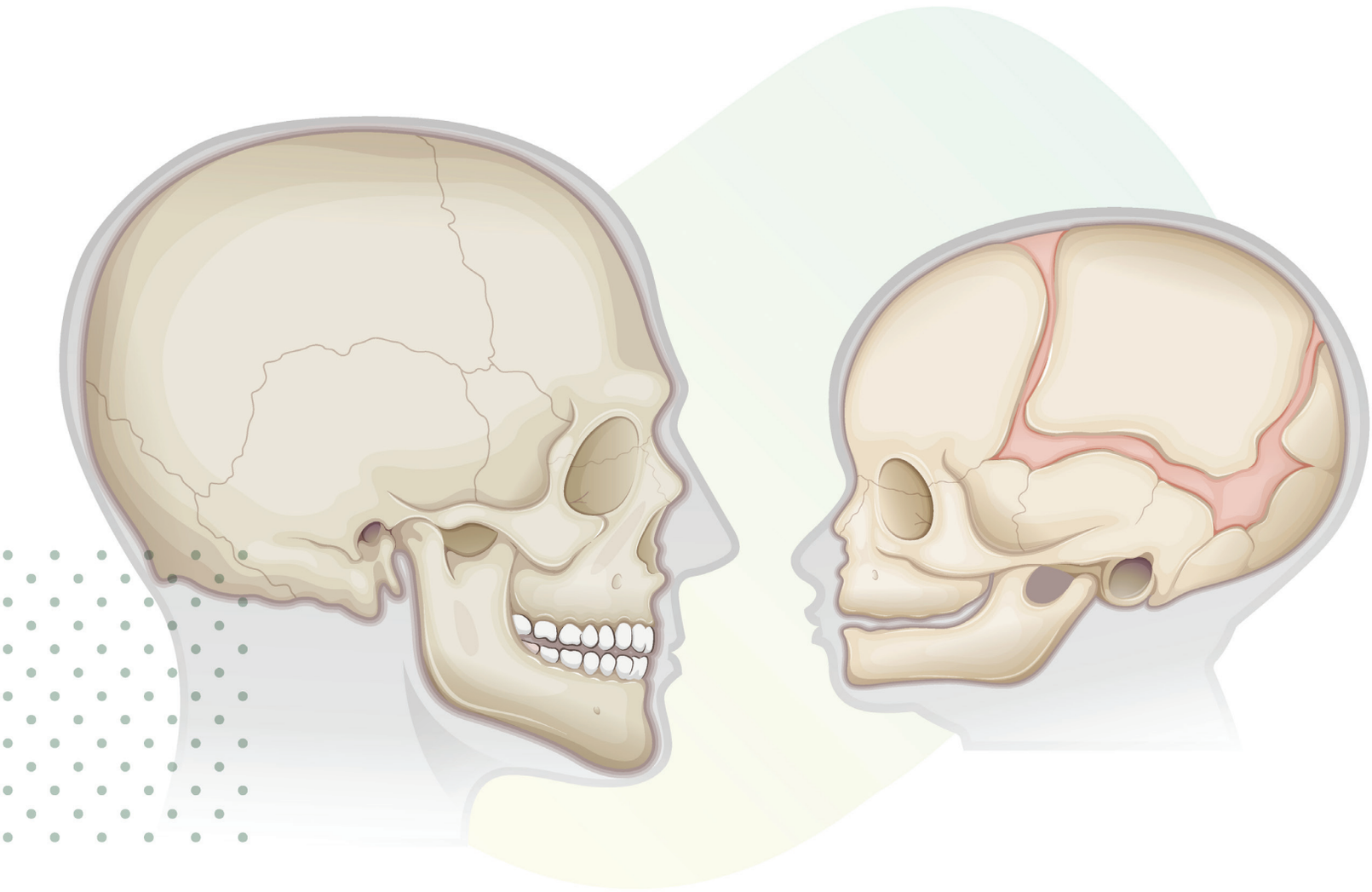

Craniosynostosis

Part 1 of 3

Kevin B. Frey, CST

Craniosynostosis is a complex congenital condition that affects the normal growth and shape of an infant's skull due to the premature fusion of one or more cranial sutures. This abnormal fusion can lead to increased intracranial pressure, developmental delays, and distinct craniofacial deformities if left untreated. Understanding the foundational aspects of this condition is essential for healthcare professionals involved in pediatric care, surgery, and diagnostics.

In part one of this continuing education series, the relevant anatomy, pathology, epidemiology, and types of craniosynostosis will be explored in depth. The discussion will also highlight the various risk factors that contribute to the development of craniosynostosis and outline the clinical and radiological methods used to confirm a diagnosis.

ANATOMY OF THE NEWBORN SKULL

The skull of vertebrates consists of two divisions – neurocranium that surrounds and covers the brain and viscerocranium that forms the face. The focus of this discussion is on the neurocranium that is divided into two sections.

- Membranous neurocranium: Forms through the process of intramembranous ossification and in process creates the frontal bone, the squamous portion of the temporal bone, the intraparietal portion of the occipital bone, and the parietal bone.[1]
- Cartilaginous neurocranium: Forms through the process of endochondral ossification and creates the ethmoid and sphenoid bones, the petrous and mastoid portions of the temporal bone, and the occipital bone.[1]

LEARNING OBJECTIVES

- Detail the anatomy of the newborn skull
- Discuss the pathology and epidemiology of craniosynostosis
- List the risk factors for craniosynostosis
- Assess the various types of craniosynostosis
- Explain the various types of scaphocephaly craniosynostosi.
- Identify methods used for confirming craniosynostosis diagnosis

KEYWORDS

craniosynostosis, helmet therapy, neurocranium, nonsyndromic craniosynostosis, Omega sign, scaphocephaly (sagittal craniosynostosis), sutures, syndromic craniosynostosis

DEFINITIONS

Autosomal dominant inheritance: A genetic condition or trait that can be inherited by a child from the parent. The condition occurs when a variant is present on one allele of a given gene. One copy of a mutated gene from one parent can cause the genetic condition. A child whose parent has the mutated gene has a 50% chance of inheriting that genetic condition.

Bossing: Protuberance of the skull that can be normal but may be associated with certain pathologies such as types of craniosynostoses. It can occur with any bone of the skull.

Bregma: Anatomical landmark where the coronal and sagittal sutures intersect, located at the junction of the frontal bone and two parietal bones.

Fibroblast growth factor receptor pathway (FGFR): Receptors that bind to members of the protein group called fibroblast growth factors (FGF). FGFs are cell signaling proteins produced by macrophages that are needed for the normal development of human cells of which 23 members of the FGF proteins have been identified in humans. Abnormalities in their function will cause developmental defects in infants. Therefore, the FGFR receptors are also involved in pathological conditions. For example, FGFR-2 is associated with Crouzon's syndrome.

Perisutural sclerosis: Increased thickening of the bone adjacent to a cranial suture that can be an indication of premature fusion.

Hypoechoic gap: Normal appearance of a cranial suture in an ultrasound image. The image will show the gap as a dark line, indicating the suture is normal and functioning as the ossification center. When the suture is prematurely fused, the gap will not show up on the ultrasound image.

Optical coherence tomography: A non-invasive test in which light waves are used to take cross-section images of the retina. The ophthalmologist can view each layer of the retina and optic nerve, allowing them to measure their thickness and monitor changes over time.

Otitis media with effusion: Thick or sticky fluid that collects behind the eardrum in the middle ear.

The cranial sutures form at the junctions of the cranial bones. They are only located between the cranial bones and are joined together by a thin layer of connective tissue called a suture ligament. During fetal development the bone tissue is placed along the suture lines, therefore, the cranial sutures are the ossification centers of the skull.[1] The sutures allow the brain to enlarge and grow through this process of progressive bone accumulation that contributes to skull expansion and a symmetrically shaped head. The sutures are immovable and therefore are classified as a synarthrotic joint. However, in the newborn, the sutures allow the bones to slightly move during the birthing process.[2]

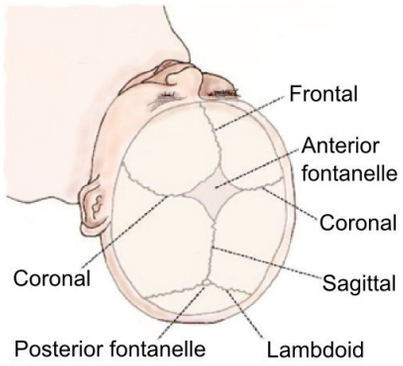

Figure 1. Sutures of the bony cranium

The bones that compose the skull are two frontal and parietal bones and one occipital bone. The sutures are as follows **(Figure 1)**.

- Metopic: Also known as the frontal, interfrontal, and median frontal suture that is a vertical suture that connects the two halves of the frontal bone. It may fuse as early as the third month of age and is usually fused in all infants by nine months. By age seven the suture has usually disappeared, but in rare cases it can still persist. This is referred to as metopism. Its persistence is clinically insignificant except clinicians and neurosurgeons should confirm its presence because it can be mistaken for a frontal bone fracture.[3]
- Coronal: Connects the frontal bones with the parietal bones. The suture extends from ear to ear.
- Sagittal: Connects the two parietal bones in the midline of the skull.
- Lambdoid: Connects the parietal bones to the occipital bone extending across the back of the skull.
- Squamosal: Connects the squamous portion of the temporal bone with the parietal bones **(Figure 2)**.

CRANIOSYNOSTOSIS: PATHOPHYSIOLOGY, CAUSES, AND EPIDEMIOLOGY

Craniosynostosis occurs when one or more cranial sutures ossify too early and do not function as the skull's ossification center(s). The growth of the skull is impeded causing various deformations depending on the cranial suture(s) that have ossified. Virchow's (Rudolf Virchow, 1821-1902, "Father of Modern Pathology") classic law of craniosynostosis states that premature fusion of a cranial suture results in limited development of the skull perpendicular to the fused suture and to compensate the skull grows in the direction parallel to the fused suture, providing room for the brain tissue.[4]

The two types of craniosynostosis are nonsyndromic and syndromic. Nonsyndromic is the most common. Its cause is not known, but research has focused on being a mix of genetic and environmental factors. Syndromic is caused by alterations in specific genes that then cause various types of syndromes including Apert, Crouzon, and Pfeiffer. The syndromes affect skull development and therefore are risk factors for craniosynostosis.

Approximately 20% of all syndromic craniosynostosis cases are genetically based, through autosomal dominant inheritance.[5] About 86% of those cases involve a single-gene mutation and 15% are related to chromosomal abnormality.[5] The genes affected most often are those in the fibroblast growth factor receptor pathway.

Depending on the research report, estimates of the incidence of craniosynostosis ranges from 1 in 2,000 to 2,500 live births with a male-to-female ratio of 4 to 1.[1,5] Nonsyndromic craniosynostoses account for 75% to 95% of cases, while syndromic account for 5% to 15%.[1,5] Statistics vary in published research articles, but sagittal craniosynostosis occurs most often in 40% to 60% of cases.[1,5] Depending on the research article, unilateral coronal craniosynostosis is next with 20% to 25% of cases but also reported to be 15% to 20% with metopic craniosynostosis reported 15% to 30% of cases.[1,5] Lambdoid craniosynostosis occurs fourth in 1% to 5% of cases. Ossification of two or more sutures is very rare and usually occurs in syndromic cases.

Multiple risk factors have been associated with development of craniosynostosis including the following.

- Environmental: Advanced parenteral age, use of tobacco products, and in vitro fertilization have been associated with craniosynostosis development.
- Ethnicity: Studies have shown that the Caucasian race has higher rates of craniosynostosis as compared to other ethnic groups.[6] Additionally, a higher incidence of metopic craniosynostosis (see below for description of craniosynostosis types) occurs in Caucasians.[5]
- Gender: As previously stated, males have a higher

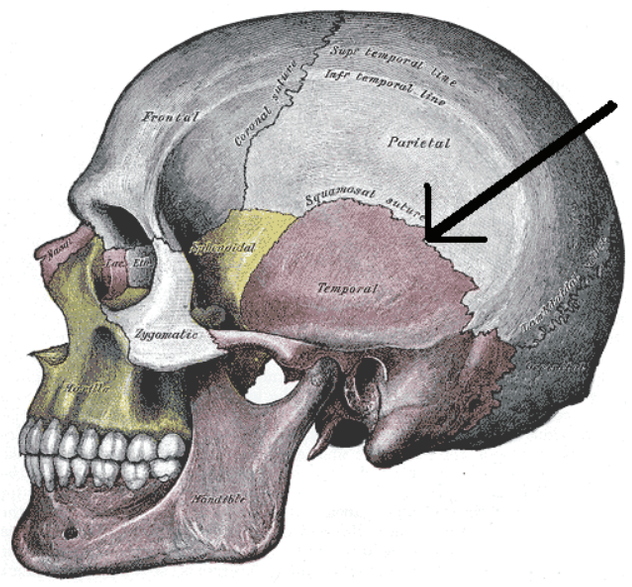

Figure 2. Squamosal suture

"The gold standard for imaging is computed tomography (CT) scan that provides the ability to assess all sutures because of the three-dimensional image of the skull."

occurrence of craniosynostosis than females. Males are more likely to have sagittal and metopic craniosynostosis, whereas females are more predisposed to unilateral coronal craniosynostosis.[5]

- Mechanical forces: Multiple births, bicornuate uterus, and pathologies such as hypoxic-ischemic encephalopathy are associated with single-suture craniosynostosis.[5]
- Thyroid disease: Increases the risk for the development of single-suture synostosis, particularly sagittal craniosynostosis. However, diagnosed thyroid disease can be managed to avoid fetal complications.
- Vitamins: Vitamins B6, C, and E lower the risk for certain types of craniosynostosis, whereas vitamins such as B12 may increase the risk of metopic craniosynostosis.[5]

Evaluating the head shape is an important clinical diagnostic method as it provides the physician with a starting point for deciding the course of treatment **(Figure 5A)**. Applying Virchow's law explains the characteristics associated with the head shapes of the various types of craniosynostosis. Only after birth can deformity be clearly confirmed, including the ability to palpate the bony ridge associated with scaphocephaly.

- Scaphocephaly (sagittal synostosis): Premature fusion of the sagittal suture **(Figures 3 and 5B)**. The shape of the head is long and narrow with a larger anteroposterior diameter. A ridge, called the Omega sign, that can be felt on palpation is present along the sagittal suture **(Figure 4)**.[1] Early ossification of the suture is associated with a more prominent scaphocephalic deformity as compared to late ossification that is associated with less of a deformity.[1]

Because Parts II and III articles will focus on surgical procedures for treating scaphocephaly, the following information regarding specific subtypes is provided:

- Dolichocephaly: Entire sagittal suture is ossified, characterized by an elongated, narrow head.
- Leptocephaly: Anterior third of the suture is ossified, characterized by a narrowing of the frontal and parietal bones.
- Cynocephaly: Middle third of the suture is ossified, characterized by a depression in the cranial bone posterior to the coronal sutures.
- Batrocephaly: Middle and anterior third of the suture are ossified, characterized by occipital bossing.[1]

> "Premature fusion of a cranial suture results in limited development of the skull perpendicular to the fused suture and to compensate the skull grows in the direction parallel to the fused suture."

- Sphenocephaly: Middle and poster third of sutures are ossified and is the most common type. Characterized by bossing of the bregma and frontal bone.[1]
- Clinocephaly: Referred to as saddle head. It can occur later during sagittal craniosynostosis, marked by concavity along the middle of the midline.[7]
- Anterior plagiocephaly: Premature fusion of a single coronal suture **(Figure 5C)**. The forehead looks flattened on the affected side with the chin deviating towards the unaffected side and on the affected side the eye appears larger, the eyebrow is raised, and the ear is anteriorly shifted.[5]
- Posterior plagiocephaly: Premature fusion of a single lambdoid suture **(Figure 5D)**. Characterized by flattening of the parietal bone and occiput on the affected side as well as an enlarged mastoid process and the ear is positioned inferior and posterior as compared to the normal side.[8]
- Pachycephaly: Premature fusion of bilateral lambdoid sutures causing a total flatness of the back of the skull.
- Trigonocephaly: Premature fusion of the metopic suture **(Figure 5E)**. The forehead is narrow and pointed and when viewing the head from above it presents a triangular shape.[8]
- Brachycephaly: Premature fusion of both coronal sutures **(Figure 5F)**. The condition causes shortened anteroposterior and widened transverse diameters of the skull.[8]
- Oxycephaly: Also called acrocephaly, high-head syndrome, or turricephaly. This occurs because of the fusion of more than one suture resulting in a towering appearance of the skull.[5]
- Kleeblattschadel: Also called cloverleaf skull, is a rare and devastating form of craniosynostosis caused by the

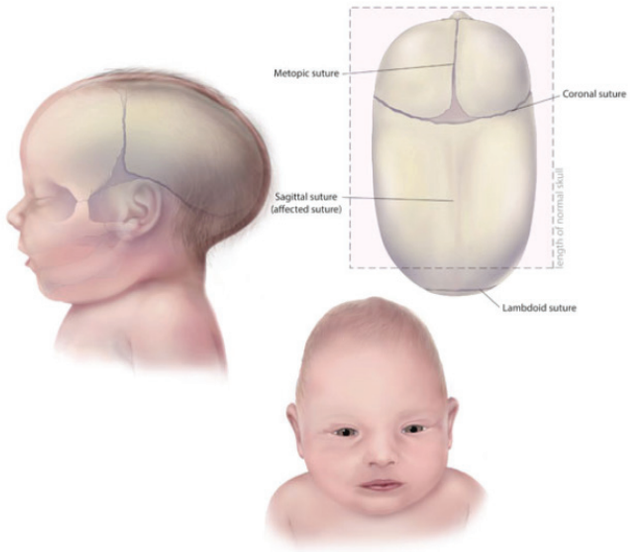

Figure 3. Sagittal Craniosynostosis

premature fusion of multiple cranial sutures, usually the coronal and lambdoid.[5] Characterized by several conditions including facial deformity, hydrocephalus, increased intracranial pressure, neurological deficits, small limb development, and temporal bulging.[9]

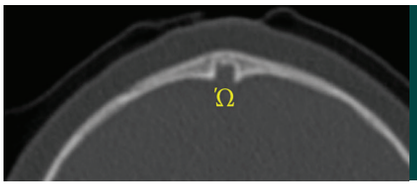

Figure 4. Omega sign – sagittal craniosynostosis

DIAGNOSING CRANIOSYNOSTOSIS

The diagnosis is initially clinical. Depending on the type of craniosynostosis, the physician is able to visualize the cranial deformations as well as palpate the skull. For example, the physician will be able to palpate the ridge (Omega sign) associated with sagittal craniosynostosis as well as palpate the absence of the displacement of the two bones adjacent to the suture. However, a final diagnosis is confirmed through radiological imaging. Plain x-rays are helpful in showing the bony ridge, perisutural sclerosis, and inability to clearly see the suture. Cranial ultrasound is used specifically to confirm ossification of the suture. A normal ultrasound will show a hypoechoic gap, whereas the gap will not be seen in prematurely fused sutures.[1,5]

The gold standard for imaging is computed tomography (CT) scan that provides the ability to assess all

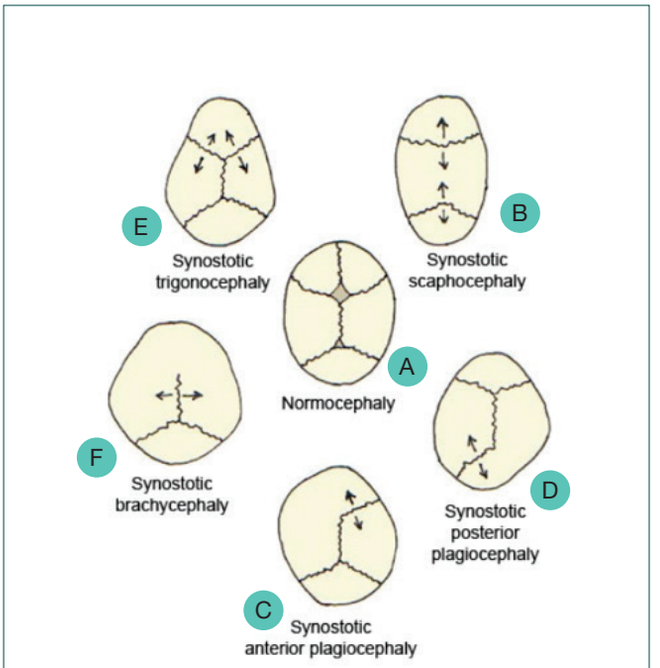

Figure 5. Types of single suture craniosynostosis: A) Normocephaly B) Scaphocephaly C) Anterior plagiocephaly, D) posterior plagiocephaly, E) trigonocephaly, F) brachycephaly

sutures because of the three-dimensional image of the skull. CT scans can also help assess the four ventricles in patients who exhibit hydrocephalus. Surgeons often use the CT scans for preoperative planning purposes. Magnetic resonance imaging (MRI) is not as effective as CT scan in diagnosing craniosynostosis, but provides additional evaluation of hydrocephalus or other intracranial abnormalities.[1,5]

For cases in which syndromic craniosynostosis is suspected, genetic testing is essential, particularly for FGFR gene mutations. Additionally, intracranial pressure (ICP) assessment and monitoring should be completed.[10,11] Increased ICP is a result of hydrocephalus, abnormal osseous changes to the skull base that affects venous outflow, and midface hypoplasia that causes sleep apnea.[5] The initial ophthalmological examination is performed to evaluate for papilledema. An optical coherence tomography may be completed because of its effectiveness in identifying elevated ICP.[11-13] However, if these tests are inconclusive, intracranial pressure monitoring should be completed.[14]

Patient with syndromic craniosynostosis who are suspected for sleep apnea may undergo a polysomnogra-

phy.[15,16] Hearing evaluations may also be performed, as the patient can suffer from hearing loss in both nonsyndromic and syndromic craniosynostosis due to otitis media with effusion.[5]

Part II in the January edition of *The Surgical Technologist* will discuss the endoscopic-assisted repair for craniosynostosis.

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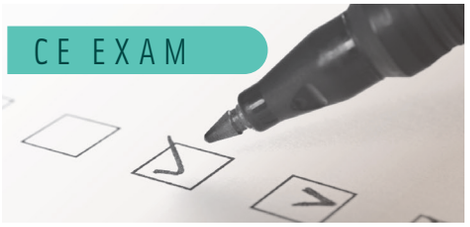

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Craniosynostosis, Part 1

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1. **Females have a higher incidence of craniosynostosis as compared to males.**
a. True
b. False

2. **Which of the following is considered the "gold-standard" imaging technique for diagnosing craniosynostosis?**
a. MRI
b. X-ray
c. CT scan
d. Ultrasound

3. **Which of following is the term applied to the bony ridge present in scaphocephaly?**
a. Omicron
b. Lambda
c. Omega
d. Alpha

4. **Which of the following terms refers to the normal appearance of a cranial suture in an ultrasound?**
a. Hypoechoic gap
b. Bregma
c. Perisutural sclerosis
d. Bossing

5. **Which of the following cranial sutures is clinically significant in that it can be mistaken for a frontal bone fracture?**
a. Coronal
b. Lambdoid
c. Squamosal
d. Metopic

6. **Which of the following is the most common type of craniosynostosis?**
a. Metopic
b. Sagittal
c. Coronal
d. Lambdoid

7. **Which of the following neurocranial divisions forms the ethmoid and sphenoid bones?**
a. Cartilaginous
b. Viscerocranium
c. Membranous
d. Synarthrotic

8. **Which of the following causes syndromic craniosynostosis?**
a. Lack of vitamin B6
b. Thyroid disease
c. Gene changes
d. Encephalopathy

9. **Which of the following subtypes of scaphocephaly is most common?**
a. Leptocephaly
b. Batrocephaly
c. Dolichocephaly
d. Sphenocephaly

10. **Which of the following does not contribute to increased intracranial pressure in syndromic craniosynostosis?**
a. Midface hypoplasia
b. Otitis media with effusion
c. Hydrocephalus
d. Osseous changes to skull base

CRANIOSYNOSTOSIS, PART 1
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