



December

Question View: All (15) ▾

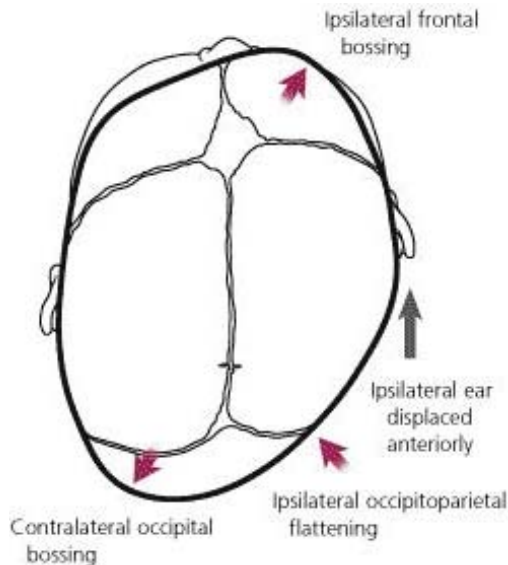
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ASSESSMENT PROGRESS: Total Questions: **15** Questions Answered: **10** Correct Answers: **9**

Question: 10

You are seeing a 4-month-old infant in the neurodevelopmental follow-up clinic. He was born at 32 weeks' gestation and his neonatal course was complicated by mild respiratory distress syndrome and necrotizing enterocolitis. His mother is concerned about his head shape. On physical examination, marked flattening of the right occiput is noted. Looking down at his head from above, you note right frontal bossing and anterior deviation of the right ear, such that his head is shaped like a parallelogram (**Figure 1**). The remainder of the physical examination demonstrates mild generalized hypotonia.

Figure 1: Vertex view. Adapted and reprinted with permission from Kabbani H, Raghuvver TS. Craniosynostosis. *Am Fam Physician*. 2004;69:2863- 2870.



Of the following, the skull deformity in this infant is MOST likely the result of:

<input type="radio"/>	A.	coronal synostosis
<input type="radio"/>	B.	deformational plagiocephaly
<input type="radio"/>	C.	lambdoid synostosis
<input type="radio"/>	D.	metopic synostosis



E. sagittal synostosis

Incorrect

Correct Answer: B

Craniosynostosis is the premature fusion of one or more of the calvarial sutures. In full-term infants, well-formed skull bones are separated by strips of connective tissue, sutures, and fontanelles (**Figure 2**). The calvarial sutures permit head malleability during passage through the birth canal, and serve as growth sites where new bone is deposited as the neurocranium expands.



Craniosynostosis occurs in 1 in 2,500 births, and 20% of cases are syndromic. The cause of craniosynostosis is unclear. Among isolated synostoses, 2% to 6% of sagittal and 8% to 14% of coronal synostoses are familial and transmitted in an autosomal dominant fashion. Fetal osteogenic growth is regulated in part by fibroblast growth factor receptor (FGFR), and mutations in the genes coding for *FGFR1* and *FGFR2* are implicated in syndromic craniosynostoses, specifically Pfeiffer syndrome (*FGFR1*) and Apert and Crouzon syndromes (*FGFR2*).

Premature fusion of calvarial sutures is a prenatal event, but initial physical examination findings may be subtle and delay diagnosis for several months. A persistent palpable ridge at the suture line in association with an abnormally shaped head suggests craniosynostosis. Plain radiography demonstrates bony bridging across the suture, sclerosis, and loss of suture clarity. Computed tomography assesses fusion of the suture, evaluates for structural abnormalities, and can exclude other causes of asymmetric cranial vault growth.

In addition to skull deformity, untreated craniosynostosis may result in inhibition of brain growth, increase in intracranial and intra-orbital pressure, asymmetry of the face, and malocclusion. Developmental delays and reduced IQs are associated with untreated craniosynostosis. Treatment options include strip craniectomy and cranioplasty, with intervention optimally occurring by 9 months of age.

Recognizable patterns of skull deformity or calvarial shape characterize each type of sutural synostosis (**Figure 3**). Growth restriction occurs in a plane perpendicular to the plane of the fused suture. Compensatory changes occur frequently and in a plane parallel to the fused suture. Clinical diagnosis can be made by viewing the infant's head from the top (vertex view) and assessing head shape. Likewise, the shape of the head can distinguish craniosynostosis, particularly lambdoid synostosis, from plagiocephaly without synostosis.

Deformational plagiocephaly, occipital plagiocephaly or plagiocephaly without synostosis, is a benign cause of skull deformity associated with occipital flattening. Typically, the head is round at birth with progressive deformation noted over weeks to months. The cause is related to positioning of the head in the same manner over a prolonged period. Risk factors for deformational plagiocephaly include abnormal fetal positioning, torticollis, hypotonia, and back-to-sleep positioning. Deformational plagiocephaly is characterized by asymmetric occipital flattening and ipsilateral forehead bossing as deforming forces are exerted in a ventral direction (**Figure 4**). The following features distinguish deformational plagiocephaly from lambdoid synostosis:

- Absence of posterior bossing (contralateral in lambdoid synostosis)

- Prominent ipsilateral frontal bossing (absent or contralateral in lambdoid synostosis)
- Anterior displacement of the ipsilateral ear (posteriorly displaced toward the fused suture in lambdoid synostosis)
- Parallelogram-shaped head (trapezoid-shaped in lambdoid synostosis)

With positional plagiocephaly, examination of the face may show flattening of the malar eminence and the mandible contralateral to the occipital flattening, with the nasal radix remaining midline. The infant in the vignette has abnormalities of his head shape consistent with deformational plagiocephaly. Treatment is nonsurgical. Conservative measures such as intentional change in positioning can improve head shape, and the use of a customized molding helmet before age 1 year can be successful in severe cases.

Coronal synostosis comprises 20% to 30% of cases of craniosynostosis. Unilateral coronal synostosis results in flattening of the forehead and frontoparietal region ipsilateral to the fused suture, with compensatory bulging of the contralateral frontoparietal region. Additional characteristic features include anterior displacement of the ipsilateral ear and deviation of the tip of the nose to the contralateral side. Bilateral coronal synostosis results in anteroposterior shortening of the skull, flattening of the occiput and temporal convexity (brachycephaly, Figure 3), and elevation of the height of the skull (turribrachycephaly).

Metopic synostosis comprises fewer than 10% of cases of craniosynostosis and results in restriction of transverse growth of the frontal bones and trigonocephaly or a triangular-shaped head (Figure 3). Narrowing of the temporal regions reduces the intermedial canthal distance, but true hypotelorism does not exist. Mild metopic synostosis may cause elevation of the suture, but no trigonocephaly.

Lambdoid synostosis is uncommon. Unilateral fusion is characterized by ipsilateral parieto-occipital flattening, contralateral parietal occipital compensatory bulging, and posterior displacement of the ipsilateral ear as the petrous portion of the temporal bone is pulled toward the closed suture. Forehead asymmetries may be associated, resulting in a trapezoid-shaped head (Figure 3). The presence of ipsilateral forehead flattening and posterior displacement of the ear help to distinguish unilateral lambdoid synostosis from deformational plagiocephaly (Figure 4). Bilateral lambdoid synostosis results in occipital flattening and increased biparietal diameter (brachycephaly).

Sagittal synostosis is the most frequently observed form of craniosynostosis, occurring in 40% to 60% of cases. Affected boys outnumber girls 4:1. Fusion of the sagittal suture is characterized by restriction in transverse growth of the skull, biparietal and temporal narrowing, and compensatory growth in the frontal and/or occipital region. The resultant head shape is scaphocephaly (dolichocephaly) (Figure 3).

More than 150 syndromes include craniosynostosis as a feature, with Apert and Crouzon syndromes accounting for the majority of cases. Typically, multiple sutures are involved. Apert syndrome is characterized by bilateral coronal synostosis with turribrachycephaly, midface hypoplasia, and complex syndactyly of the hands and feet. Crouzon syndrome is characterized by bilateral coronal synostosis, hypertelorism, midface hypoplasia with associated exorbitism, but no abnormalities of the hands or feet. These syndromes are inherited in an autosomal dominant fashion, though 50% of cases result from spontaneous mutation.

Suggested Readings

Kabbani H, Raghuvver TS. Craniosynostosis. *Am Fam Physician*. 2004;69:2863-2870. Accessed April 1, 2011 at: <http://www.aafp.org/afp/2004/0615/p2863.html>

Panchal J, Uttchin V. Management of craniosynostosis. *Plastic Reconstr Surg.* 2003;111:2032-2048. Abstract available at:
<http://www.ncbi.nlm.nih.gov/pubmed/12711969>

Persing JA. MOC-PSSM CME article: management considerations in the treatment of craniosynostosis. *Plastic Reconstr Surg.* 2008;121:1-11. Abstract available at:
<http://www.ncbi.nlm.nih.gov/pubmed/18379381>

American Board of Pediatrics Content Specification(s)

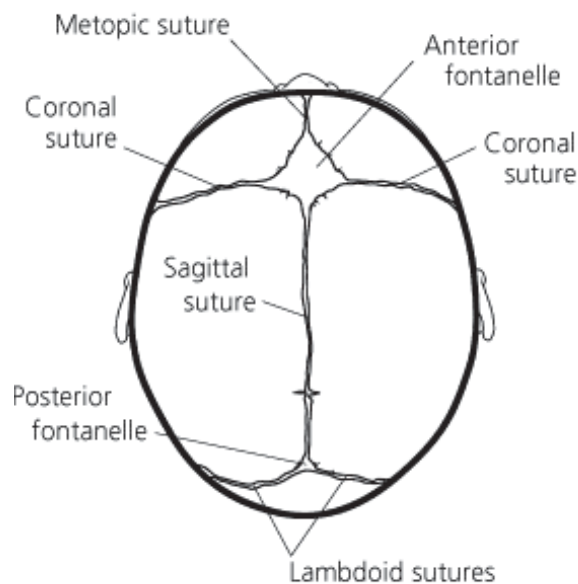
Genetics/Dysmorphism: Recognize the clinical features and know how to diagnose and manage craniofacial anomalies

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Figure 2: Calvarial sutures and fontanelles. Adapted and reprinted with permission from Kabbani H, Raghuveer TS. Craniosynostosis. Am Fam Physician. 2004;69:2863-2870.



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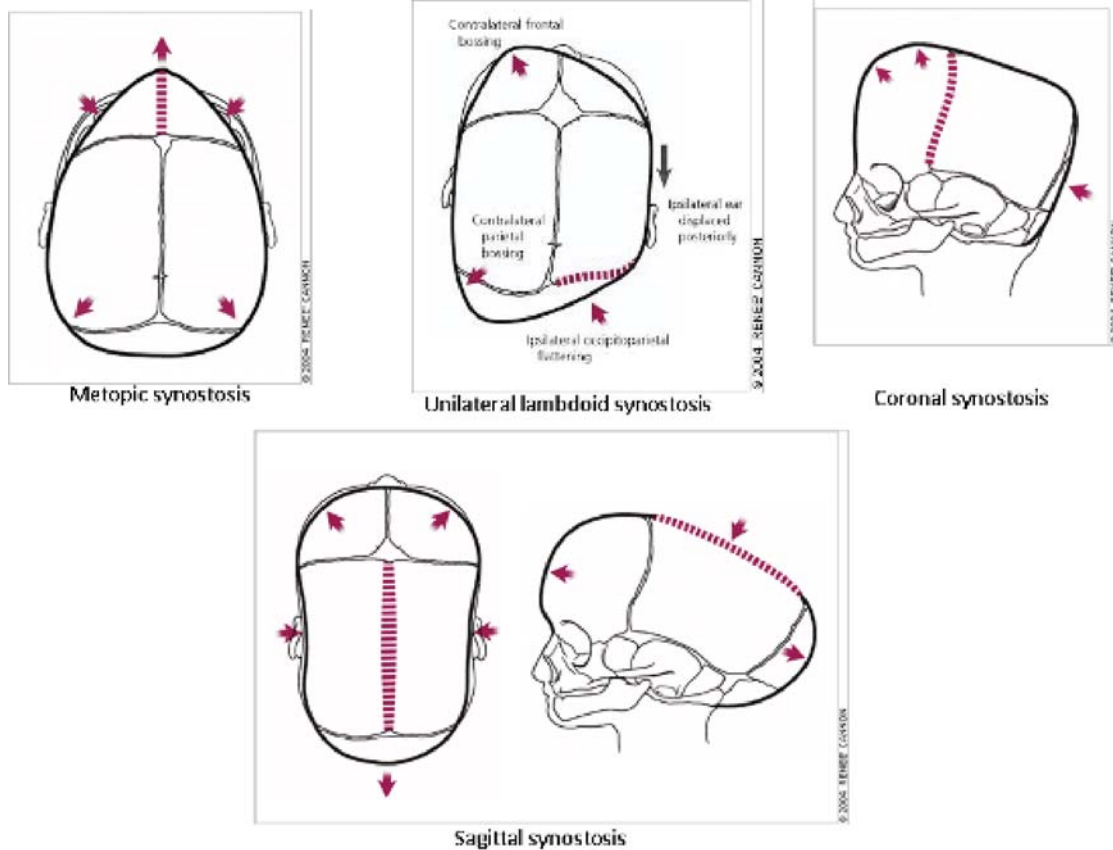
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Figure 3: Characteristic calvarial shape secondary to sutural synostoses. Adapted and reprinted with permission from Kabbani H, Raghuveer TS. Craniosynostosis. *Am Fam Physician*. 2004;69:2863- 2870.



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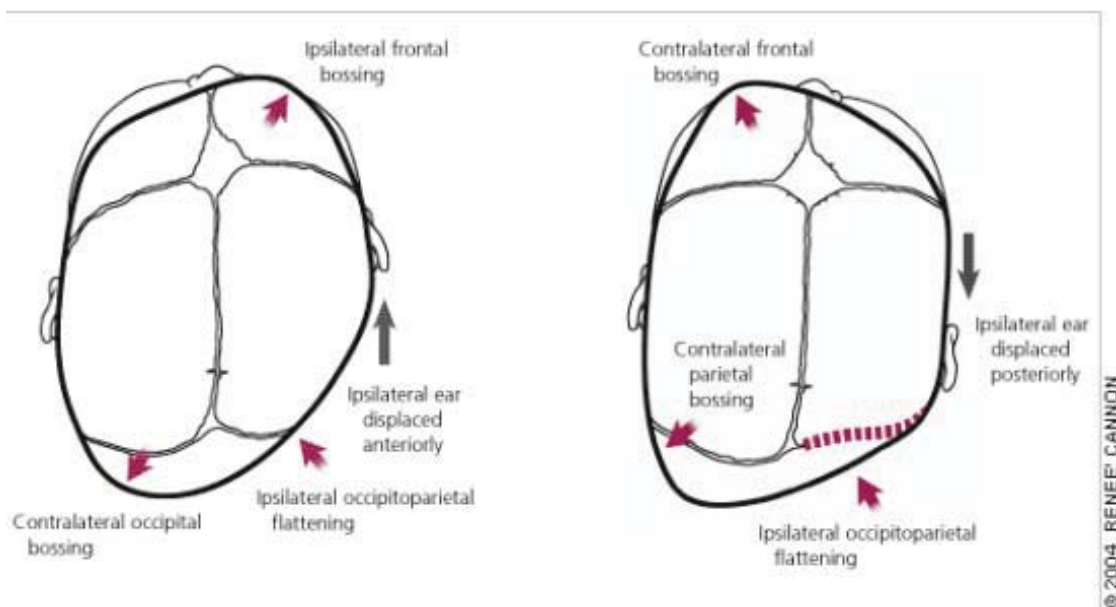
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Figure 4: Parallelogram-shaped cranium secondary to deformational plagiocephaly (left). Trapezoid-shaped cranium secondary to lambdoid synostosis (right). Adapted and reprinted with permission from Kabbani H, Raghuvver TS. Craniosynostosis. Am Fam Physician. 2004;69:2863- 2870.



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