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MEDICAL CENTER

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DEPARTMENT OF IMAGING SCIENCES

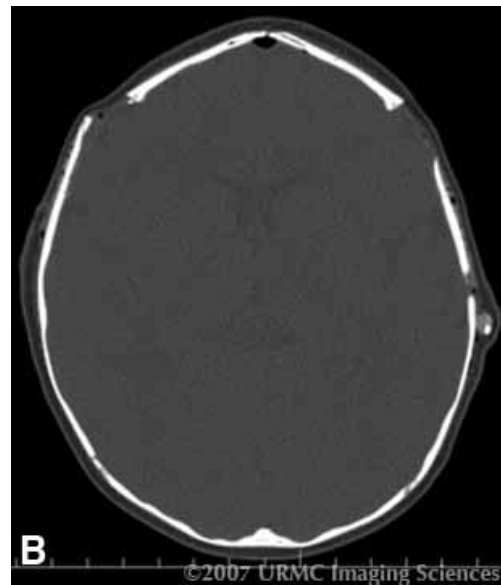
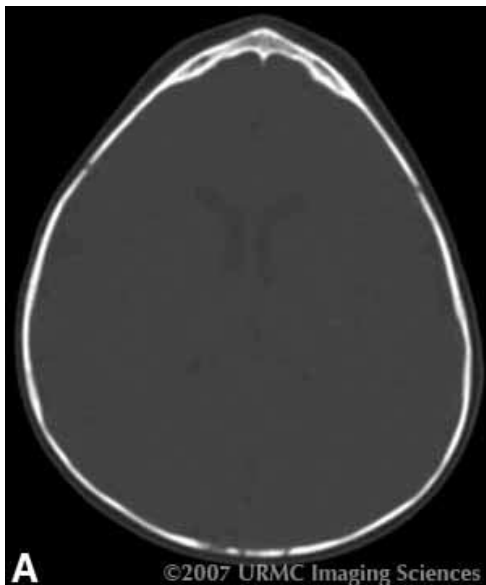
**Imaging Sciences Interesting Cases**

**CASE 25**

**Igor Mikityansky, MD, MPH**

**CLINICAL PRESENTATION:** The patient is a 5 month-old with skull deformity.

**IMAGING FINDINGS:** CT scan of the head with 3D reconstruction demonstrates fusion and bridging of the metopic suture.



**Post repair**



## **DIAGNOSIS: Trigenocephaly**

**DISCUSSION:** Trigenocephaly is a triangular, forward pointing skull due to premature closure of the metopic fissure. The metopic suture normally closes between 1 and 2 years of age and is completely fused by 3 years. In trigenocephaly, the premature fusion is completed prior to birth.

Trigenocephaly is seen in 10-20 % of all craniosynostosis patients. The incidence is 1 in 2,500 with 3 to 1 male predominance. The highest incidence of premature metopic suture closure has been described in Japanese.

In 1862, Hermann Welcker, prosector, professor, and later director of the Anatomical Institute at the University of Halle an der Salle, was first to use the term "trigenocephaly" to describe the shape of the skull in a patient with metopic suture synostosis. The deformity is most commonly an isolated finding without a known cause. Familial cases are seen in 2 to 5%. In 10 to 20% it is associated with Opitz C, Say-Meyer, or Frydman syndromes. In addition, other syndromes, such as Carpenter, Baller-Gerold, and Seathe-Chatzen may cause premature metopic suture closure. Multiple chromosomal defects without named syndromes have association with this deformity as well.

The facial features of trigenocephaly include keel-shaped forehead, absence of frontal eminences, retruded lateral orbital rims, epicanthus, and hypotelorism. Decreased interpterional width and reduced frontal height cause anterior displacement of coronal suture, which leads to reduced capacity of the anterior fossa. The increased interparietal width is thought to be compensatory. In some cases cerebral midline defects and cognitive deficiency are seen.

The general consensus is that a minor form of trigenocephaly consisting of prominent metopic ridge without other abnormalities does not need an operation. Management of other cases is controversial. Intracranial pressure is reported to be elevated in 8% cases. In addition, some researchers speculate that small anterior fossa restricts brain growth leading to mental retardation. However, this relationship is not clearly established. Most authors agree that the main reason for surgical repair is aesthetics. Although the surgery can be performed at any age before 24 months, some studies suggest that repair within the first 6 months (3-9 months range) may prevent frequently seen development of astigmatism and strabismus. Recontouring of the fronto-orbital bar is most commonly performed. Overall, trigenocephaly has the best postoperative results compared to other craniosynostosis.

## **REFERENCES:**

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2. Zumpano MP, Carson BS, Marsh JL, Vanderkolk CA, Richtsmeier JT. Three-dimensional morphological analysis of isolated metopic synostosis. Anat Rec. 1999 Oct 1;256(2):177-88. [PubMed]