

## **Isolated Frontosphenoidal Synostosis: An Underestimated Cause of Synostotic Frontal Plagiocephaly**

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### **Abstract**

**Background:** Unilateral fusion of the coronal (frontoparietal) suture is the most common cause of synostotic frontal plagiocephaly. Frontal flattening can also arise from localized fusion of the frontosphenoidal suture, a rare and often misdiagnosed form of craniosynostosis. We present our institutional experience with this rare clinical entity to highlight the unique and often elusive phenotype.

**Methods:** A retrospective chart review of our craniofacial database was performed. All patients with isolated synostosis of the frontosphenoidal suture confirmed by computed tomography (CT) were included in our series. Demographic data, clinical and radiographic findings, and clinical course were documented.

**Results:** Three patients demonstrated synostotic frontal plagiocephaly caused by isolated frontosphenoidal synostosis. All patients were female and nonsyndromic. Mean age at presentation was 4.8 months (range, 2.0-9.8). Two patients had fusion of the right side. On initial presentation, patients exhibited normal head circumference with frontal flattening and supraorbital rim recession on the involved side. Additionally, two patients had anterior displacement of the ipsilateral ear. CT imaging was required to confirm the diagnosis in all patients and was done at a mean age of 5.4 months (range, 2.1-10.8). One patient underwent CT imaging at 2.1 months of age and all sutures were patent. Based on this finding, the patient was diagnosed with deformational plagiocephaly and underwent cranial molding helmet therapy with minimal improvement. Repeat CT at 10.7 months of age demonstrated a new fusion of the frontosphenoidal suture. All three patients underwent fronto-orbital correction at mean age of 12.1 months (range, 7.8-16.1).

**Conclusions:** Isolated frontosphenoidal synostosis is a rare and oftentimes misdiagnosed cause of frontal plagiocephaly. This rare entity should be considered in any infant with frontal plagiocephaly presenting with orbital rim retrusion falling posterior to the anterior cornea, or that does not improve with growth.