Development of secondary unilateral coronal suture synostosis with a sagittal suture synostosis in a nonsyndromic patient

Case report

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Multiple-suture synostosis is typically associated with syndromic craniosynostosis but has been occasionally reported in large series of nonsyndromic children. The diagnosis of multiple fused sutures usually occurs at the same time, but rarely has the chronological development of a secondary suture synostosis been noted. The development of secondary bicoronal suture synostosis requiring surgical intervention has only been reported, to date, after surgical intervention and is hypothesized to arise from a disruption of inhibitory factors from the dura. The disinhibition of these factors permits the sutures to then fuse at an early stage. The authors report on a patient who developed secondary unilateral coronal synostosis after the diagnosis of an isolated sagittal synostosis. The secondary synostosis was identified at the time of the initial surgical intervention and ultimately required a second procedure of a frontoorbital advancement.

The clinical appearance of this phenomenon may be subtle, and surgeons should monitor for the presence of secondary synostosis during surgery as it may require intervention. Failure to identify the secondary synostosis may necessitate another surgery or result in a poor cosmetic outcome. The authors recommend close clinical follow-up for the short term in patients with isolated sagittal synostosis.

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Key Words • craniosynostosis • sagittal suture • coronal suture • secondary synostosis • congenital

Craniosynostosis occurs in approximately 3.5–4.5 per 10,000 live births and can involve any of the cranial sutures. Sagittal synostosis (scaphocephaly or dolichocephaly) is the most commonly encountered isolated single-sutured synostosis, accounting for more than 50% of cases.1,5,7 Unilateral coronal synostosis (anterior plagiocephaly) is the second most common, whereas metopic and lambdoid synostosis are less commonly encountered.7 Multiple-suture synostosis is also not uncommon but has been reported almost exclusively in syndromic craniosynostosis.3,6 Few cases of nonsyndromic multiple-suture craniosynostosis, or newly appearing secondary synostosis, have been reported, but most fused sutures are diagnosed concurrently, and plans for reconstruction include all deformities.3 We report on a patient in whom sagittal suture synostosis was diagnosed and confirmed by CT scanning and in whom a unilateral coronal suture synostosis subsequently developed.

Case Report

Examination. This 2-month-old girl was born at 35 weeks’ gestation. She had a concomitant history of multiple congenital malformations including a tracheoesophageal fistula, duodenal atresia, horseshoe kidney, patent ductus arteriosus, and atrial septal defect. On examination, the patient had obvious scaphocephaly, with a soft and flat anterior fontanelle and a frontooccipital circumference at the 50th percentile corrected for her prematurity. A CT scan confirmed the presence of a fused sagittal synostosis with the remaining sutures appearing normal (Fig. 1). Reconstruction surgery was recommended at age 2–3 months.

Operation. At 3 months of age, the patient underwent a classic suturectomy in which the previously described clamshell technique was used.9 Intraoperatively, the right coronal suture was clearly visualized; however, the left coronal suture was not. The authors report on a patient in whom sagittal suture synostosis was diagnosed and confirmed by CT scanning and in whom a unilateral coronal suture synostosis subsequently developed.

Follow-Up and Second Surgery. At follow-up, the patient had developed cosmetic facial asymmetry with char-
Secondary unilateral coronal suture synostosis

![Fig. 1. A: Preoperative 3D CT reconstruction showing sagittal synostosis with both coronal sutures open. B: Lateral view 3D reconstruction displaying an elongated head from fusion of the sagittal suture. C: Axial CT with bone windows displaying the scaphocephalic head shape.](image)

A characteristic recession of the left side of the forehead and supraorbital rim along with nasal deviation to the right (Fig. 2). The patient’s left anterior plagiocephaly continued to progress and at the age of 11 months (7 months after the initial surgery), the patient underwent a second surgery: a frontoorbital advancement and reconstruction with cranial vault remodeling.

**Postoperative Course.** The patient’s postoperative course was uneventful, and she was placed in a helmet for 4 months postoperatively. At the 6-month follow-up, the patient showed normal head contour. The patient did not undergo a complete series of genetic tests to search for abnormal chromosomal deletions.

**Discussion**

Sagittal synostosis can produce anterior and posterior calvarial deformities resulting from the compensatory growth that begins once the sagittal synostosis has occurred in utero. Other associated deformities include retrocoronal or prelambdoid constriction and bossing of the forehead. The diagnosis of sagittal synostosis can typically be made by physical examination.

However, diagnosis of certain types of multiple-suture craniosynostosis can be more challenging, and some forms can be very subtle, leading to a delay in the diagnosis. In a small subgroup of patients there may be fused sutures that are perpendicular to one another (bilateral coronal-sagittal) resulting in a nearly normal appearing head shape. This expression of combined synostosis has been termed a “balanced dysmorphism” because the sagittal fusion offset or counterbalanced the bilateral coronal or lambdoid synostosis. The cranial appearance of these patients can be quite normal and they may not be suspected of having craniosynostosis until later in childhood. Some centers obtain a CT scan on any child who has a sustained decline in the percentile growth head circumference, even if the head shape appears “normal.”

Even in patients with balanced dysmorphism, early diagnosis of multiple-suture synostosis is important as up to 77% of patients with multiple-suture synostosis will have clinical or radiographic signs suggestive of elevated intracranial pressure. Multiple-suture synostosis also leads to complex skull base deformities, orbitocranial deformities, misalignment, and psychological problems that may require early intervention.

**Fig. 2. A: Follow-up 3D CT reconstruction obtained 7 months after the initial suturectomy using the clamshell technique, showing interval development of facial asymmetry. B: Lateral view 3D CT reconstruction. C: Axial CT scan with bone windows highlighting the asymmetry of the frontoorbital calvaria.**
example, those with Crouzon, Apert, Pfeiffer, and Muenke syndromes), and rare variants of multiple-suture craniosynostosis in nonsyndromic children is significantly less common. Chumas et al. found that 3% of their 1474 patients with craniosynostosis had “unclassified” multiple-suture synostosis; the most common patterns included unilateral coronal-sagittal, bilateral lambdoid-sagittal, and sagittal metopic. Greene et al. reported unusual combinations of craniosynostosis in 7.5% of their series, and the most commonly encountered combined synostosis involved fusion of bilateral coronal-sagittal sutures. Sloan et al. reported a 12% incidence of atypical multiple suture synostosis from a series of 250 patients; the most common combinations included coronal-unilateral lambdoid and metopic-sagittal suture synostosis. In all of these previous series, the children presented with multiple-suture synostosis, and none were reported to develop a delayed-onset secondary suture synostosis. We report a case in which a coronal suture fused after the initial diagnosis of an isolated sagittal synostosis. Whether our case reflects an earlier presentation of an evolving process leading to multiple-suture synostosis or an entirely different pathological process is unclear. This one patient represents the only case of a delayed or secondary synostosis encountered at our institution from 66 patients with sagittal synostosis who required surgical intervention between January 2007 and March 2011.

Arnaud et al. reported a 10% incidence of secondary coronal synostosis after scaphocephaly correction using the standard “H” craniectomy, with 1% requiring another surgical intervention. A subgroup of this series with milder forms of scaphocephaly that were not surgically treated rarely developed secondary coronal synostosis (1.2%), and none required surgical intervention. Arnaud et al. postulated that surgery might have interrupted the signaling mechanism of the infantile dura to maintain a patent suture, thus leading to a secondary synostosis and suggesting that surgery seems to play a role in the appearance of a secondary coronal stenosis. All of their patients also developed bilateral coronal synostosis, which differed from our case. Again, perhaps the timing of diagnosis influenced the presentation, but it seems unlikely that our case would have progressed to bilateral coronal synostosis given more time. Although, the etiology of the secondary synostosis remains unclear, it is possible that mechanical forces from sagittal synostosis contributed to the development of coronal synostosis.

Although the incidence of secondary coronal synostosis is low, surgeons should remain cognizant of its occurrence and monitor for it intraoperatively especially if there is a significant interval delay prior to surgery. Failure to treat the secondary synostosis may necessitate another surgical intervention that may have been avoided. A detailed genetic screening of all nonsyndromic multiple-suture synostosis patients should be considered, although it was not performed in our patient.

Conclusions

We report on a patient in whom secondary unilateral coronal synostosis developed after being diagnosed with sagittal synostosis. The clinical appearance of this phenomenon may be subtle and surgeons should monitor for its presence during surgery, as it may require intervention. Failure to treat the secondary synostosis may necessitate another surgery. We recommend close clinical and radiological follow-up in patients with isolated sagittal single-suture synostosis.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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