A Surgical Technique for Management of the Metopic Suture in Syndromic Craniosynostosis

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Objectives: The objective is to describe a new surgical procedure developed in the San Jose Pediatric University Hospital for the management of syndromic synostosis of the metopic suture in a patient clinically diagnosed with Saethre–Chotzen syndrome.

Methods: The diagnosis of Saethre–Chotzen syndrome, bilateral coronal sutures, and metopic suture synostoses was made through photographic, anthropometric, exophthalmometric, and computed tomography analysis. The keel-like frontal bone deformity was corrected following resection using a fusiform osteotomy, remodelling was obtained by milling the edges, and the bony fragments were repositioned and fixed on the posterior wall of the frontal bone. Additionally, a fronto-orbital advancement with a self-stabilizing bar was performed.

Results: The 1-year postoperative results showed improvement in the position of the fronto-orbital bar, adequate cranial expansion, satisfactory correction of the upper facial third alteration, and correction of the keel-like deformity.

Conclusions: The surgical approach has not previously been described in the literature and offers an alternative management for syndromic craniosyntostosis of the metopic suture, avoiding skull irregularities.

Key Words: Craniosynostosis, fronto-orbital advancement, keel skull, plagiocephaly, Saethre–Chotzen syndrome, trigonocephaly

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Craniosynostosis is a congenital condition in which 1 or more cranial sutures are prematurely fused either during embryogenesis or in the postnatal period. In 1851, Virchow described that the sutures with sinostosys restrained perpendicular cranial growth that is then compensated for with expansion in the direction of the suture line. Nonsyndromic or isolated suture synostosis is the most common abnormality, the incidence has been reported as 0.4 to 1 per 1000 live births.[1,2] Syndromic craniosynostosis is less common even though more than 150 syndromes have been described until today.[3,4] Among those, Saethre–Chotzen syndrome is third in prevalence (1:25,000–50,000 live births). The incidence of trigonocephaly is controversial and accounts for 3% to 50% of all craniosynostosis, with male predominance in all reported series.

H Saethre first described this syndrome in 1931, followed by F Chotzen in the next year. It has an autosomal-dominant inheritance, with a complete penetrance and variable expression, although sporadic cases have been described.[1,2]

This article describes a case of bilateral synostosis of the coronal sutures and metopic suture in a patient diagnosed with Saethre–Chotzen syndrome. The objective is to present the management with a new surgical technique for metopic synostosis. Furthermore, relevant concepts in the treatment of this condition are discussed.

METHODS

The procedure performed at the San Jose University Pediatric Hospital was on a 4-month-old female patient with Saethre–Chotzen syndrome and synostosis of both coronal sutures and metopic suture. We performed a fronto-orbital advancement and an innovative frontal cranioplasty technique with a fusiform resection of the metopic ridge and the remodeling and reposition of the bone fragment to the posterior wall of the frontal bone. The main purpose of this technique is to correct the front-orbital deformity avoiding irregularities as well as to produce cranial expansion. The patient was followed up for 1 year.

CLINICAL PATIENT

A female patient was born preterm on gestational week 37 by cesarean section from an 18-year-old mother with gestational diabetes and preeclampsia. The parents were nonconsanguineous. The mother denies use of alcohol, tobacco, or other teratogens. Transfontanelar, abdominal, and cardiac sonographic studies were within the normal ranges. The mother and the maternal uncle had phenotypic characteristics of craniofacial alterations without syndromic diagnoses; however, the maternal uncle had a history of surgical intervention for cranial decompression during his first months of life.

The first examination of the patient was performed in the San Jose University Pediatric Hospital at the age of 4 months (Fig. 1), where the craniofacial characteristics were evaluated with imaging, anthropometric, photographic, and exophthalmometric studies (Fig. 2). The patient showed microcephaly, a low-set hairline, metopic ridge, retrusion of the upper orbital rings, medial epicanthal folds, hypotelorism, low nasal dorsum, caudal inclination of the lip corners, arched palate, and a short neck. Exophthalmus and exorbitism were not observed, and the middle third of the face was normal. Brachycephaly with a normal facial height was identified with anthropometric indexes (Fig. 2). The physical examination also showed clinodactyly in the fifth finger of both hands and a lack of syndactyly.

On examination, the mother of the patient showed similar syndromic characteristics of low-set hairline, retrusion of the upper orbital rings, brow asymmetry, left palpebral ptosis, medial epicanthal folds, arched palate, reduced anterior-posterior skull

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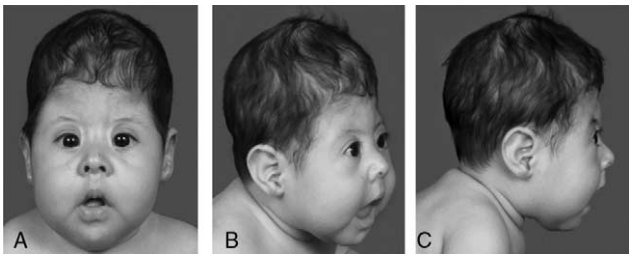

FIGURE 1. Preoperative images. (A) Frontal view. (B) Right oblique view. (C) Lateral view.

dimension, and increased vertical dimension. Furthermore, she presented brachydactylic toes (Fig. 3).

SURGICAL TECHNIQUE

The coronal approach was performed using a zigzag pattern starting in a retroauricular position, just above the pinna, bleeding was minimized with a vasoconstrictor solution at a dermic and subgaleal level as well as placing hemostatic clips to avoid the indiscriminate use of electrocautery to prevent alopecia. The dissection was performed at a subgaleal level up to the temporal line, continuing at a subperiosteal level, and a frontal craniotomy was performed by neurosurgery, carefully separating the dura of the skull. Once the osteotomies were completed, the advancement was performed with a tongue in groove self-stabilizing supraorbital bar.

The metopic ridge was removed with a fusiform osteotomy, and borders of the bone fragments obtained from it were milled on their borders and repositioned on the posterior wall of the frontal bone to correct the deformity (Fig. 4).

The cranial volume increased by performing temporoparietal barrel osteotomies (Fig. 5). The bony fragments were fixed with an absorbable material containing polyglycolic acid and polylactic acid. The patient progressed adequately during her 3-day stay in the intensive care unit and was then transferred to a regular floor without complications.

RESULTS

Follow-up was performed at 12 months, standardized photographs were taken to confirm the correction of the metopic ridge and the adequate projection of the supraorbital bar, showing the global improvement of the upper facial third deformity with a normal facial height. A computed tomography of the face and skull was performed with an ascertain correction of the brachycranic skull. Neurologically, the patient showed normal development with growth and developmental controls within the normal limits for her age (Fig. 6).

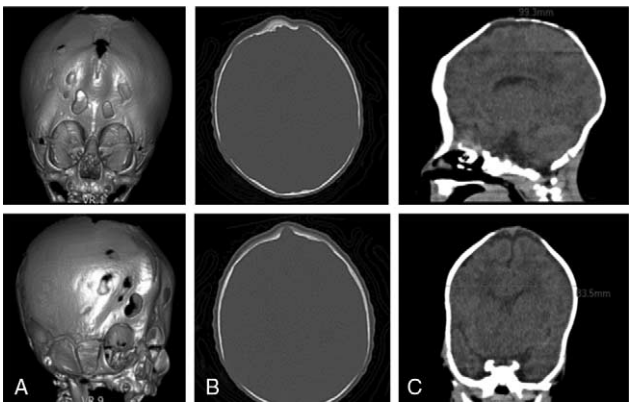

FIGURE 2. Preoperative computed tomography. (A) Three-dimensional reconstruction. (B) Axial view showing omega sign and metopic synostosis. (C) Coronal and sagittal view showing decreased intercoronal distance and small ventricles.

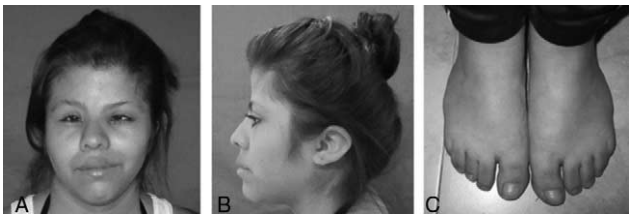

FIGURE 3. Mother's pictures. (A) Frontal view. (B) Lateral view. (C) Brachydactylic toes.

DISCUSSION

This patient is an example of the correction of the cranial deformity in a Saethre–Chotzen syndrome patient. Correction was performed by supraorbital advancement with a self-stabilizing bar, a frontal cranioplasty, and barrel osteotomies.

Saethre–Chotzen syndrome is characterized by a decreased anterior-posterior diameter or brachycephaly, upper eyelid ptosis, low-set hairline, and ears with decreased dimensions with a prominent crura and helix fold, antimongoloid palpebral fissures, and flattened nasal bridge.[5] Any cranial suture may be affected, but the bilateral involvement of the coronal suture is the most common, followed by unilateral coronal synostosis and then metopic suture involvement in terms of frequency. Presentation together with pansynostosis or multiple suture synostoses is rare.[1,6]

Furthermore, the cervical vertebra may be fused, and the limbs may present brachydactyly, syndactyly, clinodactyly, and carpal and tarsal fusion, this is why it is considered to be among the acrocephalosyndactyly group. Syndactyly is usually simple and incomplete in the second and third fingers and in the third and fourth toes.[1]

Patients with deafness associated with nerve compression and disorders in tear production with normal intelligence have been reported, although some have mental retardation.[1,2,4,7–9] Other skeletal findings are retrusion of the upper maxilla, class III underbite or distoclusion, and palatal cleft or arched palate, and exophthalmos at a lower frequency than is found in other symptomatic craniosynostoses, for which Paul Tessier named this entity "Apert Syndrome of the upper facial third."[10,11] The phenotypical characteristics are variable and depend upon the location of the mutation in the TWIST1 gene on chromosome 7p21. Furthermore, the TWIST1 gene is associated with the domain of the TWIST box, responsible for inhibition of the cranial synostosis by blocking Runx2, a protein required for the induction of osteoblast differentiation and endochondral ossification.[1]

Muenke syndrome and Pfeiffer syndrome are included among differential diagnoses showing similar phenotypical characteristics, the only difference being a mutation located on chromosome 5. The

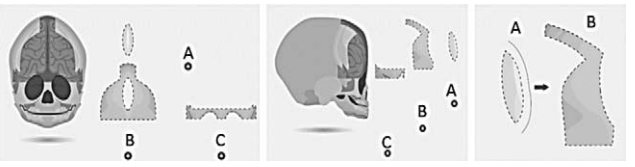

FIGURE 4. Osteotomies design. (A) Fusiform metopic ridge resection and edge milling. (B) Frontal bone graft. (C) Supraorbital bar bone graft.

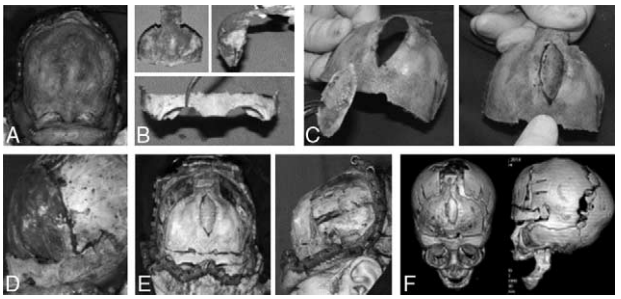

FIGURE 5. Surgical technique. (A) Metopic suture synostosis. (B) Frontal bone and supraorbital bar grafts segments. (C) Frontal cranioplasty: metopic ridge fusiform bone fragment after osteotomy an edge milling and its placement on the posterior wall of the frontal bone. (D) Tongue in groove self-stabilizing supraorbital advancement. (E) Frontal and lateral views after bone fragments were fixed with an absorbable material. (F) Postoperative computed tomography.

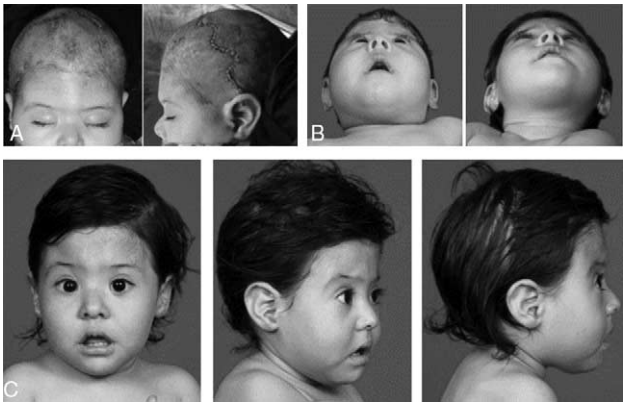

FIGURE 6. (A) Immediate postoperative result: frontal and lateral views. (B) The difference from basal view before and 1 year after the surgery. (C) One-year follow-up result: frontal, oblique, and lateral views.

mutation in Muenke syndrome is in the gene encoding the receptor for Fibroblastic Growth Factor 3 and shows a higher incidence of mental retardation and psychomotor development. The mutation in Pfeiffer syndrome is located in the gene encoding Fibroblastic Growth Receptors 1 and 2.[12]

In our hospital, the surgical management is conducted between the 4th and 6th months of life, although some patients might require intervention at an earlier phase when endocranial hypertension is documented.

The supraorbital bar techniques may vary according to the abnormality observed, in this patient, projection was the main improvement aim, requiring advancement with a self-stabilizing bar technique with a tongue in groove design for self-support.[9]

Several techniques for the management of the metopic ridge have been described, most suggesting frontal cranioplasty designs that include multiple osteotomies, such as the open-wing,[13] cathedral dome,[14] endocranial placement of the osteosynthesis material,[15] and endoscopic techniques.[16]

The advantage of this technique is the lower number of bone fragments that require repositioning. Its execution is thus simpler, and the bony gap is minimized when repositioning the spindle-shaped segment to the posterior wall of the frontal bone, thus avoiding future cranial defects and irregularities. However, as occurs in all cranial expansion techniques, the creation of dead space between the dura and the skull may increase the risk of complications, such as the formation of an epidural abscess or a hematoma.[13]

The need for a reintervention in the patient is subject to an increase in intracranial pressure. About 35% to 42% of the patients with Saethre Shotzen suffer from recurrent endocranial hypertension that requires further surgical expansion by craniotomy and cranial remodeling. A second reintervention is estimated to occur 4.3 years after the first surgery due to recurrence of the retrusion of the upper third or a beliebe of intracranial hypertension in patients who had the first remodeling procedure before 15 months of age.[12]

Whitaker et al described an assessment classification for patients with syndromic craniosynostosis according to the type of reintervention performed after cranial remodeling. They classified the patients into 4 categories as follows: those not requiring a reintervention, those requiring a minor intervention for bone or soft tissue outlining, those requiring a new major surgery with osteotomy or bone graft; and finally, those requiring a new major surgery that is similar to or more extensive than the initial cranioplasty.

Several patient series have described that up to 65% of the patients require a secondary major intervention,[1] characteristic shared across all syndromic craniosynostosis due to the possible involvement of intrinsic genetic defects in the bone, although these recurrences are more common in Apert syndrome. For this reason, overcorrection of 2 to 3 mm advancement over the planned treatment is important.[9,11]

Midface deformity at age 5 to 7 years is managed with a monobloc or Lefort III procedure or even a facial bipartition. Finally, girls between 13 and 15 years of age and boys between 15 and 17 years of age undergo orthognathic surgery for a definitive occlusal correction.[17]

CONCLUSIONS

The metopic synostosis fusiform osteotomy and frontal cranioplasty technique is useful in the management of the metopic keel-like deformity and should be considered for patients with syndromic craniosynostosis of the metopic ridge. Additional studies with a greater number of patients are required to standardize the technique.

In our hospital, surgical management is carried out between the 4th and 6th months of life, although some patients might require intervention at an earlier phase when endocranial hypertension is documented. Reinterventions vary according to the severity of the deformity and the presence of endocranial hypertension; therefore, monitoring the neurological development of these patients is essential.

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