

Surgical Treatment of Metopic Synostosis

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Abstract: Metopic synostosis is characterized by keel-shaped forehead (trigonocephaly), prominent midline ridge of the forehead, bitemporal narrowing, bilateral retrusion of supraorbits, egg-shaped orbits, low nasal dorsum, and reduced volume of the anterior cranial fossa. The mainstay treatment is early surgical intervention before the age of 12 months, which usually consists of bifrontal craniotomy with bilateral recontouring, lateral advancement, and lateral displacement of the superior orbital rims. Here, we have developed a new simplified technique for surgical treatment of trigonocephaly.

A total of 60 cases of trigonocephaly were operated on between January 1995 and January 2010 by the first author. Surgical outcomes were evaluated 6 months after surgery using postoperative photographs and clinical examination notes, and scaling was made using the Whitaker classification. The evaluation showed that 85% of them were in class I, 11.6% were in class II, and 3.3% were in class III. No case was in class IV. Only the last 10 cases received the new surgical technique, and all were in class I.

Complication rate was 38.3% for all cases and was only 20% for the last 10 cases, that is, the new technique group. Revision rate for trigonocephaly surgery was 13.3%, and the most common reason was hardware removal. None of the patients from the new technique group underwent revision surgery.

We believe that our new technique is fast and easy, can provide sufficient bone graft, and is more useful for older patients (>1 y). Early postoperative results have been promising.

Key Words: Trigonocephaly, metopic, synostosis, cranial suture

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Craniosynostosis has been recognized since the time of Hippocrates.¹ Single-suture synostosis is the most common form of craniosynostosis.² The rate of isolated nonsyndromic craniosynostosis in newborn population has been reported to be approximately 1 in 2000 to 3000 live births.³ True incidence of trigonocephaly (premature closure of metopic suture) is somehow controversial. Although many authors believe that trigonocephaly is not a common

form of craniosynostosis, with an estimated incidence of 0.3 to 7 per 1000 to 15,000 live births (ie, 3%–10% of all craniosynostosis require surgical treatment),^{1,4–7} others have reported a higher incidence (14%–30% or even as high as 50%).^{8–12} In all reported series, a male predominance was evident. The male-to-female ratio has been reported from 2:1 up to 6.5:1.^{1,9,11} Most cases are sporadic, but an autosomal-dominant mode of inheritance has also been described.^{1,4}

It is appropriate to describe metopic synostosis as “premature fusion” because the metopic is the only calvarial suture that normally fuses in humans.^{4,13} Although some reports indicate that this fusion may be secondary to the influence of regional dura growth factor-mediated signals, the exact cause of this deformity still remains unknown.³

Metopic synostosis results in keel-shaped forehead (trigonocephaly), prominent midline ridge of the forehead, bitemporal narrowing and compensatory increased biparietal growth, bilateral retrusion of supraorbits, hypotelorism with or without epicanthal folds, egg-shaped orbits, low nasal dorsum, and reduced volume of anterior cranial fossa.^{14–16} Also, 17% to 25% of patients have associated anomalies.¹ Although there are many reports that indicate that trigonocephaly may be associated with mental and behavioral problems,^{8,17} the correlation between severity of skeletal disfigurements and mental development is somehow controversial.^{18,19}

Apart from very mild cases, early surgical interventions for trigonocephaly have been strongly recommended.²⁰ The optimal time is considered before the age of 12 months,¹⁰ usually between 3 and 6 months,^{7,14,21} because of the high probability of passive postoperative endocranial remodeling, the likelihood of reossification of calvariectomy defects, the malleability of the calvarial bone, and the favorable effect on minimizing facial dysmorphism.²² Although there is a concept that surgery may also improve cognitive function,²⁰ generally it is believed that intracranial anomalies are not secondary to synostosis and surgery is done mainly for the aesthetic purpose.⁷

The standard approach to metopic synostosis in infants has been bifrontal craniotomy with bilateral recontouring, lateral advancement, and lateral displacement of superior orbital rims.⁸ In this article, we set out to describe a new simplified technique for surgical treatment of trigonocephaly and review our postoperative results for this surgery, especially those treated by the new technique.

MATERIALS AND METHODS

All craniosynostosis surgeries done by the first author between January 1995 and January 2010 were reviewed retrospectively. Records of the isolated metopic synostosis patients were selected and reviewed carefully. Also, all demographic data, including sex and age (at first visit and at the time of surgery), plus surgical technique, postoperative results and complications, follow-up schedule, repeated operations, and its causes were recorded. All patients' photographs and videotapes (before and after surgery) were reviewed. The new technique was used since January 2009 on the last 10 patients, that is, the new technique (NT) group. Hence, they were reviewed separately.

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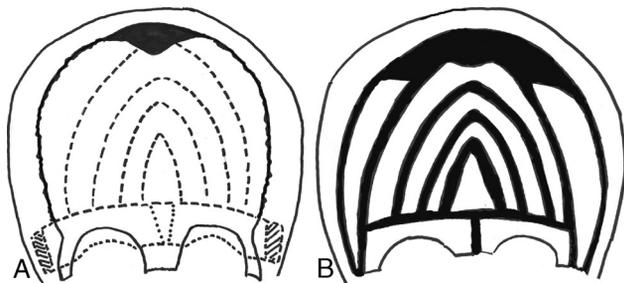


FIGURE 1. A, Schematic drawing for horseshoe-like osteotomy. Usually, the outer borders of osteotomy lines correlate with coronal sutures, but in some cases, it may be necessary to include some parts of the parietal bones for better configuration. B, After rearrangement of bone segments.

The surgical outcomes were evaluated 6 months after surgery using postoperative photographs and clinical examination notes. The Whitaker classification²⁰ was used for scaling of results, which consists of 4 classes: class I, excellent result, no revision necessary; class II, satisfactory result, soft tissue revision indicated; class III, marginal result, bony irregularities present, requiring contouring with bone graft or alloplast/osteobiologics; and class IV, unacceptable result, repeat craniotomy and fronto-orbital reshaping necessary.

Surgical Technique

In this section, the operation that was undertaken for the NT group is described. First, the patient is anesthetized by a pediatric anesthesiologist, using endotracheal intubation, 2 large-bore intravenous lines and 1 arterial line. Second, Foley catheter is inserted, and urine output is recorded. After local infiltration with saline solution containing 1:200,000 epinephrine, a lazy-w bicoronal incision is made. Anteriorly based frontal flap is raised in a subgaleal plane. Afterward, this plane is changed to a subperiosteal plane 1.5 to 2 cm above the superior orbital rims. Temporalis muscles are also elevated in association with the perisoteal flap.

Marking is done and the frontal bony segment (including frontal and some parietal bones) is removed in 1 piece by the neurosurgeon. Supraorbital bar is then removed, which includes minimal part of the temporal bone. Immediately lateral to supraorbital bar, a quadrangular-shaped osteotomy is made, and the bone piece is removed.

Using a wedge osteotomy in the midline of bandeau, the supraorbital bar is split in 2 pieces. Then these segments are rotated anterosuperiorly and reattached to each other by a 5-hole bioabsorb-

able plate (Onion, Finland) without using any bone graft. Afterward, the bandeau is refixed in an advanced position using 20-hole bioabsorbable plates bilaterally. No fixation at the midline is used. It should be mentioned that, in some cases, the superior orbital rims are too prominent and must be trimmed for a better configuration.

Thereafter, the frontal bone is then split by making multiple concentric horseshoe-like osteotomies (Fig. 1). These osteotomy lines must be at least 10 to 15 mm apart from each other. Usually, this results in 5 to 6 bone pieces. These segments are then rearranged and refixed in position with bioabsorbable plates (Onion; Fig. 2). Sometimes removing one or moving some pieces is necessary for having a more natural convex forehead profile. Fixation of these segments is easily done with bioabsorbable plates (or even Vicryl suture in some points).

After copious irrigation and careful hemostasis check, perisoteal flap and temporalis muscles are resuspended by Vicryl sutures. No drain is used. The galeal layer is approximated by Vicryl sutures, and the scalp skin is closed by staples (Fig. 3).

A special bulky cotton dressing is used for the scalp, which makes the patient unable to rotate his/her head. Blood loss is estimated and compensated during surgery by transfusion. The patient is extubated in the operating room and transferred to neonatal intensive care unit for the next 24 hours. Also, intravenous third-generation cephalosporin is used for infection prophylaxis. Finally, the patient is discharged from the hospital 3 days after surgery.

RESULTS

From January 1995 to January 2010, a total of 152 craniosynostosis patients were operated on by the first author. From these patients, 60 cases were trigonocephaly, 52 cases were anterior plagiocephaly, 28 cases were brachycephaly, and 12 cases were lambdoid synostosis and multiple-suture synostosis.

Patients with trigonocephaly consisted of 45 boys and 15 girls. The mean ages of the patients at the first visit and at the time of surgery were 8.4 months (range, 3–46 mo) and 10.1 months (range, 4–48 mo), respectively. Follow-up visits were done at 1, 3, 6, and 12 months after surgery and then once a year. Photographs were taken after 3, 6, and 12 months and also 5 years after surgery. The first computed tomography was performed 1 year after surgery for all patients. Early postoperative results were determined 6 months after surgery. On the basis of the Whitaker classification,²⁰ the population of this study was classified as follows: 85% in class I, 11.6% in class II, and 3.3% in class III. No one was in class IV.

Surgical complications for all trigonocephaly surgeries consisted of 10 cases of palpable plates or screws, 1 case of infection, 1 case of bleeding, 5 cases of hematoma, 2 cases of seroma, 2 cases of cerebrospinal fluid leakages, and 2 cases of recurrent bitemporal

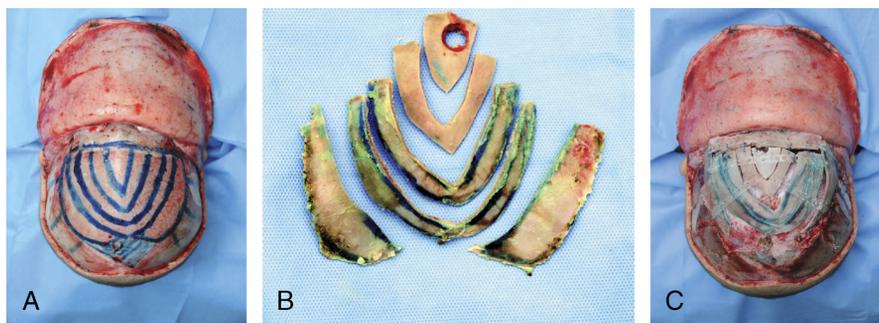


FIGURE 2. A, Marking for osteotomy. B, Bone segments after completion of osteotomy. C, After rearrangement and fixation of bone segments.

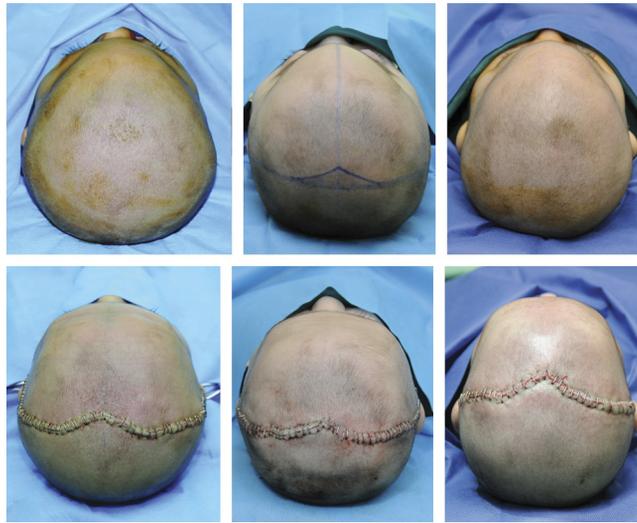


FIGURE 3. Preoperative (top) and immediate postoperative (bottom) photographs of 3 patients operated on with the new technique.

narrowing. Repeated operations were done for 8 cases, including 4 cases of hardware removal, 1 case of bleeding control, 1 case of infection drainage, 1 case of hematoma drainage, and 1 case of soft tissue revision.

From this population, 50 patients were operated on by common techniques and 10 patients were chosen to receive the new developed technique, that is, the NT group. Surgical complications of the NT group were limited to 1 case of cerebrospinal fluid leakage and 1 case of seroma, both treated nonsurgically. All cases from the NT group were classified as class I in the Whitaker classification 6 months after surgery.

DISCUSSION

Our 15 years of experience with craniosynostosis included 152 cases, which consisted of 60 cases of trigonocephaly (39.4%). Hence, in our series, trigonocephaly was the most common form of craniosynostosis, which is not entirely comparable with the previous literature. Regarding that, we did not have any mild cases (who are not referred usually for surgical treatment); its true incidence may be higher. Also, a pan-European study exists, in which an increased incidence of metopic synostosis has been demonstrated.²³ The male-to-female ratio in our study was 3:1, which is comparable with the reported male dominance in this malformation around the globe.

The treatment of trigonocephaly is challenging: it is a bilateral disfigurement, it involves the midline of the forehead and both orbits (which makes camouflaging difficult), it will worsen if remains untreated,²⁰ and it associates with mental and behavioral problems.^{8,17} Surprisingly, surgery cannot improve the cognitive state⁸ or normalize growth.²⁴

There are many different surgical approaches and techniques to treat trigonocephaly. The much older techniques of strip craniectomy (first reported by Matson)⁴ and lateral canthal advancement (first reported by Hoffman¹⁴) have evolved to the open fronto-orbital remodeling (first reported by Marchac)⁴ and more recently into minimally invasive endoscopy and distraction osteogenesis.^{3,25,26} The mainstay of all open surgical techniques is to remodel and to reposition the frontal bone and supraorbital bar.⁸ However, the fact that associated hypotelorism should be surgically corrected or not is a matter of debate.^{4,6,7,10}

Fifteen years of craniosynostosis surgery and treating many cases of trigonocephaly led us to revolutionize the older time-consuming surgical techniques and develop a newer, simplified one. Regarding the extension of the supraorbital bar (bandeau), in this new technique, we terminate the lateral extension of the supraorbital bar osteotomy line just beyond the zygomaticofrontal suture. Instead of advancing a part of the squamous portion of the temporal bone¹⁶ (“tongue-in-groove” technique), we excised a rectangular bone piece just lateral to the supraorbital bar (where the bone is depressed; Fig. 1). When the supraorbital bar is advanced, this gap will become larger.

Reshaping of the supraorbital bar is done by splitting and rotating it at the midline, but different techniques are used for reattaching these segments. Some authors use interpositional bone graft between 2 segments.¹⁶ Selber et al²⁰ have reported that they used another segment of the bone graft at the posterior side of the interpositional segment for additional buttressing. They also used inlay bone strut to orbital roof. Other authors believe that bone graft does not improve the hypotelorism state, and hence, they do not use it.¹⁴ We prefer the latter approach and do not use any bone graft here and just reattach the 2 segments by a 5-hole bioabsorbable miniplate.

Recontouring of the frontal bone does not require a unique technique. The bone may be reshaped by multiple peripheral cuts (or wedge osteotomies) and outward bending (greenstick fracture) of the segment.²⁰ In older patients, when the bone is thicker and harder, this technique may not be applicable, and the frontal bone must be split into multiple, irregularly shaped pieces. Rearranging and reattaching these puzzle-like segments is sometimes difficult and time consuming. Some authors have used individualized templates as a guide for remodeling²⁷ and some use barrel-staving osteotomy technique for a better configuration.⁸

Our new technique consists of performing multiple concentric horseshoe-like osteotomies at the frontal bone segment. These segments (usually 5–6) are then rearranged in position. Sometimes, it is necessary to remove 1 segment for better configuration and convexity of the frontal bone. This will provide an extra source for bone graft. In our last 2 patients, the outer bone segment was split into 2 segments, and the latter were advanced anteriorly, filling the gap between the supraorbital bar and the squamous bone (Fig. 1). Because of the long arms of the bone segments, fixing them is



FIGURE 4. Scaling is done by comparing preoperative and postoperative photographs. Left, Preoperative photographs of a patient (same patient in Fig. 2). Right, Postoperative photographs 6 months after surgery.

easy and can be quickly done with 3 to 4 long (20-hole) absorbable plates. There are few reports about using spring in addition to frontal remodeling in the hope that it will also correct the associated hypotelorism.^{4,28} However, we believe that the hypotelorism state will be corrected spontaneously.

There are many ways for evaluating postoperative surgery results: clinical appearance, standardized photography,¹⁴ three-dimensional computed tomographic scans and volumetric studies,^{2,16} standardized cephalometry, and anthropometric studies.²⁴ We prefer to evaluate our results by clinical examination and standardized photography only (Fig. 4), avoiding unnecessary x-ray exposure to our patients. Because of simplicity and reliability, we reported our postoperative results based on the Whitaker classification. In this respect, from 60 cases of trigonocephaly, 85% were in class I, 11.6% were in class II, 3.3% were in class III, and no one is in class IV 6 months after surgery. Also, considering the results of our new osteotomy technique, all NT group patients were in the Whitaker class I. Although this can show that our new technique is promising, long-term follow-up of patients is needed for it to be judged more accurately.

On the whole, 23 complications occurred (38.3%) in the postoperative period, which included 10 cases of palpable hardware. Since the time of using bioabsorbable plates instead of metallic ones, this complication never occurred again. However, the complication rate for the NT group was only 20%.

The overall revision rate for trigonocephaly surgery was reported to be 15% to 20% in the literature, and the most common reason for secondary surgery has been temporal reconstruction.¹⁴ Recurrence of bitemporal depression may not be considered as a failure of surgery because it is a consequence of restricted growth of the frontal bone in a lateral direction secondary to the absence of a functional metopic suture.¹⁰ Because cranioplasty cannot create a normal functional metopic suture, it is not surprising that bitemporal depression can reoccur even after appropriate surgical treatment. There is also a report about coronal suture transplantation to overcome this problem.²⁹ In our series, the revision rate for this surgery has been 13.3%, and the most common reason has been hardware removal. Up to now, we have not reoperated on any of the NT group patients.

CONCLUSIONS

We believe that our new technique for the osteotomy of the frontal bone has several new advantages. First, it is fast and easy to learn and easy to do. Second, it provides sufficient bone in case more bone graft is needed. Third, because it has long segments of bones, it is much easier to be fixed by plates and screws. Fourth, for older patients (>1 y) whose greenstick fracture of bone segment is much more difficult, this can be a more reasonable substitute. Last, its initial postoperative results have shown to be promising.

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