Surgical Treatment of Trigonocephaly
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Summary
Metopic synostosis is a relatively rare form of nonsyndromic synostosis. Premature closure of the metopic suture results in deformation of the anterior calvarium, in the phenotypic features of trigonocephaly. Trigonocephalic deformities are recognized because of a pathognomonic 'keel-shaped' deformity of the forehead with a prominent midline ridge, bilateral frontotemporal constriction with compensatory biparietal expansion, supraorbital and lateral orbital retrusion and hypotelorism.

This article presents a surgical technique used over 5 years to treat trigonocephalic children. Retrospective analyses were performed on 9 patients with metopic synostosis treated during 2005-2010. The study included reviews of preoperative and postoperative computed tomography scans, operative techniques, clinical outcomes and complications. Operative techniques included fronto-orbital bandeau tilting after expansion with a midline interpositional -bone graft, detrangulation of foreheads with an inward bending at lateral orbital wall, multiple zigzag osteotomy of frontal bone, barrel-stave osteotomy of parietal bone.

The average age of the patients at time of surgery was 11 months. Interorbital distances widened from 18.8 mm to 20.9 mm, biocular distance widened from 67.56 mm to 74.39 mm, interorbital distance widened from 13.44 mm to 15.65 mm, intertemporal distance widened from 58.22 mm to 64.16 mm. Mean follow-up was 34 months, and no neurological sequelae or other significant complications were encountered.

Trigonocephaly requires surgical correction involving anterior two-thirds calvarial remodeling with fronto-orbital advancement and frontal-bone remodeling. The described operative approach minimizes bone defects by adopting multiple zigzag osteotomy of the frontal bone. This modality results in significant improvements in skull form and high patient/parent satisfaction.

Key words: Trigonocephaly, metopic synostosis, surgery

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INTRODUCTION

Metopic synostosis is a relatively rare form of nonsyndromic synostosis (25,35). In large serials, metopic synostosis accounts for 7-23% of the synostosis operations performed (1,5,15,19,29). The incidence of metopic synostosis was reported to be 0.3 per thousand 1,000 live births (10). Under normal conditions, metopic suture remains patent until 2-3 years of age and so frontal bone development continues (8). However, premature closure of the metopic suture results in trigonocephaly. In trigonocephaly, a ‘keel-shaped’ deformity of the forehead is present and this is characterized by a prominent midline ridge, bilateral frontotemporal constriction with compensatory biparietal expansion, supraorbital and lateral orbital retrusion and hypotelorism (1,13,27,33) Furthermore, the patient is protected from psychiatric effects of craniofacial anomaly in the long-term (1,15). In the studies performed, increase in self-esteem, decrease in hyperactive behavior and decrease in anxiety was observed in the children undergoing craniosynostosis surgery (7).

In this study, fronto-orbital remodeling surgical technique that we used in the treatment of trigonocephalic cases and the results we obtained are presented.

MATERIAL AND METHODS

Nine patients who were diagnosed as trigonocephaly and operated by Reconstructive Surgery and Neurosurgeon teams during 2005 and 2010 were analyzed retrospectively. The ages, genders of the patients, sutures held, surgical interventions performed, hospitalization periods, complications and follow-up periods, preoperative and postoperative computed tomography scans were investigated. Preoperative and postoperative intercanthal, biocular, interorbital and intertemporal distances were measured.

SURGICAL TECHNIQUE

Coronal incision was planned as to be zigzag osteotomy. Scalp was dissected until temporal region and supraorbital region at subgaleal plane. Dissection was
performed until frontozygomatic and frontonasal sutures and 2 cm inward from orbital ceilings by passing to subperiostal plan at 1-2 cm superior part of supraorbital rim. Temporal muscle was removed until squamous suture subperiostally. Supraorbital nerves were released from supraorbital rim (Figure 1). Craniotomy line was passing through just anterior part of coronal suture and 1 cm superior part of supraorbital rim and supraorbital segment in 2 cm width which was extending 5 cm to temporal region by passing 2-3 mm top of frontonasal and frontozygomatic sutures was marked. Bifrontal craniotomy was performed, duramater was dissected from the floor of anterior cranial fossa, supraorbital horizontal bar was removed approximately 2 cm in width together with its temporal elongation. Supraorbital bar was remodeled by a midline interpositional-bone graft and/or absorbable plaque-screw and so hypotelorism and frontal ridge had been corrected (Figure 2). Additionally, bitemporal narrowing was corrected by forming green-stick fracture at lateral orbital walls. Remodeled supraorbital bar was fixed with absorbable plaque screws, wire and silk suture.

Forehead and skull-cap reconstruction was performed by frontal bone flap after osteotomies performed to ensure anterior cranial widening. Widening was assured by multiple green-stick fractures and whole fractures performed at the peripheral parts of the bone, but the bone integrity was not deteriorated. Therefore, an increase was provided in anterior 2/3 cranial volume. However, bone defect area was minimized by osteotomies performed. Scalp was closed by adapting pericranial flap lifted in its place (Figure 3).

![Figure 1a: Dissection of scalp at subgaleal plane and appearance of subperiosteal dissection at approximately 2 cm superior part of supraorbital region. Figure 1b: Exploration and preservation of supraorbital nerves.](image)

![Figure 2: Appearance of bone segments removed after osteotomies. Supraorbital bar, remodelling by a midline interpositional-bone graft and/or absorbable plaque-screw.](image)
RESULT

Nine patients, 6 males and females were operated due to metopic synostosis between 2005 and 2010. Personal and familial histories of the patients were nonspecific. These patients were evaluated genetically and no genetic problem was detected. The patients operated due to trigonocephaly were nonsyndromic and their average ages were 11 months. The assessments of the patients were performed clinically and by quantitative CT measurements. Preoperative and postoperative intercanthal, biocular, interorbital and intertemporal distances were evaluated by measurement of the images at the tomography (Figure 4). While preoperative average intercanthal distance, biocular distance, interorbital distance and intertemporal distances were 21.78 mm, 67.56 mm, 13.44 mm and 58.22 mm, respectively, postoperatively these values were determined to be 23.67 mm, 74.39 mm, 15.65 mm and 64.16 mm, respectively (Table 1). Duration of the operation was average 150 minutes. Blood loss was approximately 250 cc. Postoperative average hospitalization duration was 7.4 days and no complication was observed during postoperative period. Mean follow-up of the patients was 34 months (6 months and 5 years) and reoperation was not required during this period. The patients were evaluated according to Sloan classification of surgical results regarding cosmetic improvement (Table 2) and photographic documentations were prepared during three months follow-ups. In Sloan classification, 7 patients were assessed to be Sloan 2 (Figures 5, 6) and 2 patients to be Sloan 3.
Table 1: The results of preoperative and postoperative intercanthal, biocular, interorbital and intertemporal distance measurements.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Intercanthal distance (mm)</th>
<th>Biocular distance (mm)</th>
<th>Interorbital (CT) distance (mm)</th>
<th>Intertemporal (BT) distance (mm)</th>
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Table 2: Sloan classification after craniosynostosis correction operation

Class 1  good–excellent result, no visual irregularity and no irregularity felt by palpation

Class 2  Good–excellent result, no visual irregularity, irregularity felt by palpation is present but reoperation is not required (e.g., palpable but invisible wire, plaque-screw or bony prominences)

Class 3  Good–excellent result together with visual irregularities, but reoperation is not required (e.g., visible plaque-screw, wire, bony prominences or bone defects not affecting surgical correction)

Class 4  Good–excellent result together with visual or palpable irregularities requiring surgical correction (e.g., removal of plaque-screw)

Class 5  Presence of problem affecting surgical correction but not requiring reoperation (e.g., presence of mild asymmetry of forehead)

Class 6  Asymmetry disturbing correction in general and severe sufficient enough to require reoperation

Class 7  Presence of problems affecting surgical outcome and reoperation offered by the surgeon but unwillingness of the family against reoperation

Figure 4a: Appearances of intercanthal and biocular distances, Figure 4b: Images of interorbital and intertemporal distances in tomography
Figure 5a-b: Preoperative appearances of 6 months trigonocephaly patient

Figure 5c: Appearance of preoperative 3 dimensional CT of the patient

Figure 5d-e-f-g: Postoperative appearances of the patient just after operation, after 1 month, 6 months and 18 months, Sloan Class 2.

Figure 6a-b: Preoperative appearances of 8 months trigonocephaly patient, CT scan imagining

Figure 6c-d-e: Postoperative appearances of the patient just after operation and after 8 months, Sloan Class 2.
DISCUSSION
Trigonocephaly results from premature closure of the metopic suture. Metopic synostosis can be encountered in clinic with a spectrum ranging from mild to severe deformity. In the cases with mild deformity, a mild ridge occurs in metopic suture localization and a mild distortion in supraorbital, orbital and cranial anatomy. If this protrusive condition does not improve until 3 years of age in these cases, excellent outcomes can be obtained with conventional methods. Baumgartner has suggested microscopic minimal invasive suturectomy technique in the patients younger than 3 months. Marked frontal protruding structure, hypotelorism, supraorbital rim and lateral forehead retrusion was present in the cases with more severe deformity and orbital structure changed as a result of these.

The deformities occurred due to premature closure of the metopic suture is wanted to be corrected by the surgery. Various techniques were described within years for the surgical treatment of craniosynostosis. These are ranging from simple suturectomy to calvarial bone remodeling. In recent years, endoscopic procedures and distraction osteogenesis are tested.

The most important goal of anterior calvarial remodeling for the correction of trigonocephaly is the reshaping of the frontal bandeau. Required planning should be made for frontal bandeau advancement and remodeling and leading to symmetrical and satisfying supraorbital rim contours. Supraorbital bar should be removed and expanded with an interpositional wedge-bone graft and pathological deformity in supraorbital bar should be corrected by application of bone graft as inverse wedge. So, significant correction occurs in hypotelorism and bifrontal narrowing. Green-stick fractures are formed at lateral orbital walls to correct bitemporal constriction and to normalize contour. Furthermore multiple osteotomies and green-stick fractures of the frontal and parietal bones are performed to increase the anterior cranial volume. The goal of the osteotomies performed is to minimize the surface area of the bone defect as much as possible, therefore multiple zigzag osteotomies were recommended. Minor irregularities can be removed by application of calcium phosphate cement. Calcium phosphate cement is biocompatible and it increases osteoconduction. Besides that it is easy to use and it increases the accuracy of reconstruction. Remodeling capacity of reconstruction is quite high in immature bones. In all cases, relapse risk in the long term is tried to be minimized by performing overcorrection during operation.

Treatment of metopic synostosis is multidisciplinary, plastic and reconstructive surgeon, neurosurgeon, pediatrician and ophthalmologist are necessary. Vast majority of these patients have developmental delay. Therefore, these patients should be evaluated in this respect and necessary support should be provided. Timing of surgery is controversial in the cases with severe deformity. Aim is best long-term cosmetic result and minimal risk. Patients in the serials of Kelleher et al. were operated while they were 12 months but the cases in other serials were operated during 3-9 months period. Although the patients in Kelleher's study were more elderly than the other studies, long-term cosmetic result obtained was satisfying and blood loss of the patients was too less. While 400 ml of blood loss occurred from the patients operated at average 7.5 months, 200 ml of blood loss occurred from 12 months infants. This caused decrease in operation morbidity.

Symptomatic palpability of the plaque-screw used during the operation is a general problem. This problem is removed.
by using absorbable plaque-screw. Sufficient rigidity is provided with absorbable plaque-screws until completion of bone healing.

Head circumference was used to evaluate the outcome during postoperative period. However, no correlation was found between change in head circumference and satisfaction level. Although CT scans are used in cephalometric measurements and anthropometric measurements, there are so many technical problems for standardization of radiographic data in the patients with marked asymmetry (4,6,20,31). In addition, the patient receives extra radiation from these imagining techniques and since the average age of the patients operated due craniosynostosis is low, it is required to procedural sedation to the patients during these imagining. These measurements take a lot of time. The most important criterion is self-perception of the patient. But objective self assessment of the patient can be lacking. Thus, assessment disharmony between surgeons and patients is too meaningful (32). Satisfaction of the patients and their relatives are high for both scar and skull form following trigonocephaly surgery. Many scales indicating this satisfaction were constituted (12,14). In their serial including 250 patients, Sloan et al., examined the cases regarding residual deformity, irregularity, complication, mortality and need for additional surgery. They evaluated the results with their own scales (29). Also we evaluated our cases by using this scale. Plaque-screw and/or wire were visible subcutaneously only in 2 cases, plaque-screw and/or wire could be noticed in 7 cases by palpation. None of these patients was operated due to these complaints. Follow-ups of the patients and their parents with quite high satisfaction are continuing.

Number of the patients is quite limited due to cultural reasons. More patient is required to evaluate the long-term results and treatment parameters. Surgery outcomes are generally satisfying for patient parents and the patient due to cosmetic improvements in skull form.

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