Calvarial Remodeling For The Treatment of Scaphocephaly in A 4-Moth-Old Boy: Case Report

4 Aylık Hastada Skafosefalinin Tedavisi İçin Yeniden Şekillendirme: Olgu Sunumu

ABSTRACT

Scaphocephaly is the most common type of craniosynostosis. Simple sagittal suturectomy is still the operation of choice for infants under the age of 6 months. We describe a 4-month-old boy who underwent calvarial remodeling for the treatment of scaphocephaly. The patient was operated on in the prone position using a transverse incision halfway between the anterior fontanelle and the occipital prominence. Sagittal strip craniectomy was coupled with bilateral parietal wedge craniectomies and occipital craniotomy. Bone edges of the wedge craniectomies were drawn to each other and the occipital craniotomy flap repositioned following remodeling. The described technique provided an immediate restoration of the cranial shape and the cosmetic outcome, 6 months after the surgery, was pleasing. Although an extensive procedure, calvarial remodeling can be the procedure of choice for a better cosmetic outcome even before 6 months of age.

KEY WORDS: Scaphocephaly, craniosynostosis, remodeling

ÖZ

Skafosefali, en sık görülen kranyosinostoz tipidir. Basit sagital sütürektomi altı aydan küçük hastalarda halen tercih edilen bir tedavi yöntemidir. Bu makalede, skafosefali nedeniyle kalvaryal yeniden şekillendirme yapılmış 4 aylık bir erkek çocuk sunulmuştur. Hasta yüzüstü yatar pozisyonda, ön fontanel ile oksipital çıkıntıyı ortalayan yatay bir kesi ile opere edildi. Hastaya sagital sütürektomi, iki taraflı paryetal üçgen şekilli kranyektomi ve oksipital kranyotomi yapıldı. Daha sonra, üçgen kranyektomi alanlarının kemik kenarları birbirine doğru çekildi ve oksipital kemik flebi küçültülerek yerine konuldu. Uygulanan teknik, kafa şeklinin ameliyat esnasında düzeltilmesini ve 6 ay sonraki kontrolünde tatmin edici bir kozmetik sonuca ulaşılmasını sağlamıştır. Sonuç olarak skafosefalinin tedavisinde yeniden şekillendirme, daha büyük çaplı bir girişim olmakla beraber, daha tatminkâr bir kozmetik sonuç için 6 aydan küçük hastalarda da uygulanabilinir.

ANAHTAR SÖZCÜKLER: Skafosefali, kranyosinostoz, yeniden şekillendirme

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INTRODUCTION

Scaphocephaly is the most common type of craniosynostosis (6). Despite the introduction of numerous sophisticated techniques for calvarial remodeling, simple sagittal suturectomy still seems to be the procedure of choice in infants younger than 6 months of age (12). In this technical note, a 4month-old boy who underwent calvarial remodeling for the treatment of scaphocephaly is described.

CASE

A 4-month-old boy presented with the typical findings of scaphocephaly: frontal bossing, biparietal narrowing and occipital bossing (Figure 1A, 1B). Neurological examination did not reveal any pathology. Pre-operative x-rays and three dimensional computerized tomography scans confirmed a fused sagittal suture. An operative decision was made for the correction of scaphocephaly.

The patient was placed in the prone position with



the chest elevated by chest rolls and the head resting face down on a gel-padded pediatric horseshoe head holder, paying attention to avoid any pressure on the eyeballs (Figure 2A). A transverse incision, positioned halfway between the anterior fontanelle and the bottom of the occipital prominence was used. Prior to the incision, 40 mg lidocaine containing 0.025 mg epinephrine in 10 cc normal saline was injected intradermally. The epidermis was incised with a knife and the scalp opened and dissected away from the underlying periosteum using needle-tip unipolar cautery (Figure 2B).

After the flaps were mobilized, sagittal strip craniectomy with a width of 3 cm, occipital craniotomy with a diameter of 4 cm and encompassing the occipital prominence, and bilateral parietal wedgeshaped craniectomies at the sites of maximal parietal narrowing were performed using a high-speed drill (Figure 2C). The bone edges of the wedge-shaped craniectomies were drawn to each other by using sutures to widen the biparietal diameter. The occipital dura was plicated by coagulation using bipolar cautery. The occipital craniotomy flap was then repositioned, following removal of a 1 cm strip around its periphery (Figure 2D).

The subcutaneous tissue was closed using interrupted sutures, and the skin was closed with a stapler. No drain was placed (Figure 2E). The total amount of blood loss was 80 cc, and the total time of surgery was 105 minutes. No blood transfusion was needed. There were no intra- or post-operative complications. The patient was discharged on the 5th post-operative day, and the staples were removed on the 10th post-operative day as an outpatient procedure.



Figure 1A, 1B: Pre-operative photographs of the patient. (with the permission of the family)



Figure 2A: The patient was placed prone



Figure 2B: Transerse incision was used, and the skin flaps were mobilized



Figure 2C: Sagittal strip craniectomy, bilateral parietal wedge craniectomies, and occipital craniotomy were performed.



Figure 2D: Bone edges of the wedge-shaped craniectomies are drawn to each other, and occipital craniotomy repositioned following remodeling.



Figure 2E: The skin was closed with a stapler and no drain was used.

The patient was called for a follow-up examination 6 months after surgery. The cosmetic result was pleasing, and the family was satisfied with the outcome (Figure 3A, 3B).

DISCUSSION

Premature closure of the sagittal suture is the most common type of craniosynostosis, accounting for up to 60% of cases (6). Fused parietal bones act as a single growth center with reduced growth potential and compensatory growth occurs at neighboring sutures (12). Affected children present with biparietal narrowing, frontal bossing, ridging of the fused



Figure 3A, 3B: Post-operative photographs of the patient. (with the permission of the family)

sagittal suture and occipital bossing, giving rise to the so-called "scaphocephaly" appearance (14). Although it seems to be an aesthetic consideration, scaphocephaly is also a functional disorder. Children with uncorrected scaphocephaly are socially isolated and stigmatized (2). As adults, they are more likely to suffer from personality disorders (4).

Removal of the stenosed sagittal suture was first described by Lane in 1892 (7). Since then, numerous procedures have been developed for the treatment of scaphocephaly including strip craniectomies (13), extended craniectomies (3) and calvarial vault remodeling (10). In this report, the patient was operated on by using a technique first described by Albright (1).

Timing of the surgery is an important issue in the treatment of scaphocephaly. The optimal time for operating seems to be 3-4 months as the blood volume at this age is greater than that of the newborn period, the 8 to 10 week period of

physiologic anemia has passed, the skull has greater pliability than at 6-12 months, bone defects fill with new bone formation more readily, and the subsequent brain growth further improves the postoperative skull shape (1). Furthermore, despite the correction of the skull shape, it has been observed that the neurocognitive performance of scaphocephalic children does not reach the same level as their matched controls at school age if the surgery is performed after 6 months of age; early correction of the deformity seems to decrease this developmental delay (15).

Despite the introduction of numerous sophisticated techniques, sagittal suturectomy still seems to be the procedure of choice in infants younger than 6 months of age because of its simplicity and the rarity of complications (12). The mainstay of sagittal suturectomy is unrestricted brain growth which has been thought to reshape the calvarium; however prospective data have shown that only one-fourth of the children who have undergone sagittal suturectomy before the age of 6 months have retain a normal cephalic index (5). On the other hand, calvarial remodeling provides an immediate restoration of the skull shape, giving rise to an aesthetically pleasing cranial shape in the very early post-operative period (8).

When calvarial remodeling and simple suturectomy were compared, remodeling led to a greater intra-operative blood-loss but the total time of hospitalization was similar, and there were no significant complications in either group (9). Prospective data have shown that the cephalic index has been normalized in nearly all patients who have undergone calvarial remodeling before the age of 6 months (5). Furthermore, Panchal et al (11) have performed a photographic assessment of head shape following calvarial remodeling. The photographs of the operated children and their matched controls were evaluated by both lay and professional observers. The results indicated that children who had undergone calvarial remodeling for the treatment of scaphocephaly before the age of 6 months were indistinguishable from their peers on the basis of a fully haired head.

In conclusion, calvarial remodeling should be the procedure of choice, even before 6 months of age, for an aesthetically pleasing cranial shape although it is a more extensive procedure than sagittal suturectomy.

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