Non-syndromic craniosynostosis treated by frontal orbital advancement: A case report

Khoo Kay Wai, MD¹, Arman Zaharil Mat Saad, MSurg (Plastic)², Mohd Ali Mat Zain, MSurg (Plastic)²

¹Reconstructive sciences unit, Health Campus, USM, Kubang Kerian, Kota Bharu, Kelantan Malaysia, ²Department of Plastic and Reconstructive Surgery, General Hospital Kuala Lumpur, Kuala Lumpur, Malaysia

SUMMARY
Craniosynostosis is a premature pathologic fusion of one or more cranial vault sutures leading to abnormally-shaped skull. It can occur in isolated event (non-syndromic), or it can occur in conjunction with other anomalies in well-defined patterns (syndromic). The diagnosis rests on clinical examination and confirmation is generally on the computed tomography scan. The need for surgery is both for cosmetic and functional reasons. Here we describe a case of non-syndromic craniosynostosis that was treated with frontal orbital advancement (FOA). The potential benefits of FOA need to be carefully weighed against the potential complications when deciding for any surgical intervention.

INTRODUCTION
Craniosynostosis, defined as premature fusion of cranial sutures, Virchow created a classification system for the types of skull deformity observed in craniosynostosis and made the important observation that premature suture fusion resulted in compensatory growth in other areas of the skull.¹ Craniosynostosis is classified as single suture versus multiple sutures and as either syndromic or non-syndromic. Non-syndromic craniosynostosis typically involve a single suture, the most common types being sagittal, unicoronal, bicoronal, metopic, and lambdoid. Bilateral coronal fusion produces brachycephaly (flat head syndrome) which is skull shortening in the anterior to posterior dimension.

Frontal orbital advancement (FOA) originated from the changing concepts of treatment of craniofacial dysostosis introduced by Tessier in 1967.¹ FOA is still considered to be the gold standard in the treatment of coronal suture synostosis due to its universality of indications, no matter the age of the patient or severity of deformities. Local data are limited on the techniques and repair of craniosynostosis. Meanwhile, internationally a lot of techniques and evolutions of the technique described which signify that so far no one single procedure is considered perfect in managing this complex case. The variability on the surgical access (open versus endoscopic), bony incision, resection and remodelling pattern, and fixation methods are among topic being discussed. In this case we used a non-absorbable suture which gave less rigid fixation but serve as good modality for holding osteotomized bony components without major complications and managed to achieve satisfactory outcome.²

CASE REPORT
AZ, a girl aged 1-year-old was referred to plastic surgery team for abnormal skull shape. She was born full term via normal vaginal delivery with uneventful pregnancy. The child does not have any medical or surgical problems. She has two normal elder brothers who are eight years old and five years old.

According to parents, the child had normal developmental milestones. Clinically, the child was noted to have flat forehead, increased bitemporal diameter and mild proptosis. A clinical diagnosis of brachycephaly was made. Computer tomography was carried out to confirm the diagnosis and also for surgical planning. Computer tomography showed isolated bicoronal craniosynostosis.

The child underwent FOA in a joint effort with the neurosurgical team. A bicoronal incision was performed behind the coronal suture and directed laterally above the ears. Clips were placed on the incised edges to reduce blood loss. The dissection was performed in the subperiosteal plane and extend laterally detaching the temporal muscles of the temporal bone and down to the level of supraorbital rims. The supraorbital and supratrochlear nerves were preserved.

A bifrontal craniotomy was carried out anteriorly leaving 10mm of orbital rim and posteriorly as far behind the coronal suture. The orbital osteotomies were performed with an oscillating saw. The frontal bone and the frontal orbital bar were removed as separate pieces. 1cm of strip craniectomy was performed over bilateral coronal sutures. The frontal bone and frontal orbital bar were reshaped by thinning and burring down the thickened areas of the bone. The reshaped frontal bone was attached to the frontal orbital bar with non-absorbable sutures. Two bone grafts from the strip craniectomy were added to the advanced lateral orbit and secured with non-absorbable sutures. Drains were left in place and the skin closed in layers.

The child’s recovery was uneventful and was discharged on the post-operative day-10. Patient is currently on regular follow up.

DISCUSSION
Since craniosynostosis first began to be surgically treated in 1890, much controversy has evolved regarding the procedure
for correction of bicoronal craniosynostosis. Surgical procedures have expanded from simple suturectomies to frontal calvarial vault remodeling consisting of bifrontal craniotomies and FOA.1

In 1892, Lannelongue described bilateral strip craniectomies, but this method was associated with high morbidity and mortality due to major blood loss.3 Simple suturectomy is nowadays considered to be insufficient to correct the complex three-dimensional growth restrictions in bicoronal craniosynostosis.

On the other hand, the FOA procedure manages to restore the volume of the anterior vault and corrects the morphological changes. This method restores both volume and shape of the skull. Thus, FOA has been considered to be the best treatment of bicoronal craniosynostosis.

Complications arising from FOA are common. Mortality rates varied between 0 and 4.5%. There has been a general decline in reported mortality rates, and the most recent series report mortalities of less than 1%. Commonest cause of death was from intraoperative haemorrhage followed by intracranial problems (cerebral oedema, bleeding, tonsillar herniation). Cardiac arrhythmias and pulmonary embolus were also reported.3 Another complication is related to the osteotomy across the anterior skull base which creates a communication between the nasal cavities and the anterior cranial fossa may lead to infection or CSF rhinorrhea.4 CSF leaks may occur but most of these settled spontaneously, and nearly all responded to lumbar drainage (if required).

Frontal bone infection due to devascularization during surgery can be problematic. The incidence of frontal bone flap necrosis requiring debridement and a subsequent cranioplasty varied between 3-20%.4

The incidence of postoperative seizures is not widely commented on in the literature. As the appearance of seizures may be a late event, McCarthy et al. in their 20-year review of syndromic craniosynostosis outline the problem and found postoperative seizures principally a problem in Apert syndrome.

CONCLUSION
The functional and aesthetic benefits of FOA are well documented, but these advantages are associated with a significant complication rate. The potential benefits of FOA need to be carefully weighed against the potential complications when recommending surgery.

REFERENCES