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Case Report

Trigonocephaly – A case series

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ABSTRACT

Purpose: To esthetically evaluate the surgical outcome in children with non-syndromic craniosynostosis.
Patients and methods: Three consecutive patients with non-syndromic trigonocephaly were included in the study.
Result: A total of three children with isolated non-syndromic metopic craniosynostosis were analyzed. The children were treated with supra-orbital bar advancement and cranial vault remodeling. The mean age at the time of initial operation was 9 months. All children achieved desirable esthetic results and there was no operative mortality.
Conclusion: The endocranial base in non-syndromic craniosynostosis has a suture-specific dysmorphology that normalizes, to variable degrees, after cranio-orbital surgery in infancy. Hence, timely restorations of facial form by surgery will not only improve morphology but also prevent functional disturbances and will result in less psychological trauma from peers and society in general.

1. Introduction

Children with metopic suture craniosynostosis account for approximately 4–10% of all patients with single-suture craniosynostosis [1]. Although rare, the hallmark cranial and facial features of metopic craniosynostosis make it a striking anomaly. The metopic suture that lies in the midline between the two frontal bones usually fuses by 2 years of age. If it fuses before birth, spectrums of craniofacial anomalies can result [2]. In the case of a premature fusion where sutural growth does not occur, the continued growth of the brain 'must find fresh directions in which to expand'. This results in compensatory growth of the skull in other areas away from the involved suture, leading to the classical recognizable shape for each involved suture according to the principles first proposed by Virchow in 1851. The restriction of ability to 'expand' in the area of the synostosed suture does not always result in a rise in overall intracranial pressure but does occur in a percentage of cases [3]. In its mildest form, metopic suture craniosynostosis can present as a bony ridge in the midline of the forehead. In its most conspicuous form, the affected child will manifest trigonocephaly and hypotelorism; the hypotelorism is accompanied by a change in the orbital shape (the so-called egg-shaped orbits) [4], and deficient lateral orbital projection [2].

1.1. Etiology

Potential risk factors include white maternal race [7], advanced maternal age [7], male infant sex [7], maternal smoking [8], maternal residence at high altitude [9], use of nitrosatable drugs (e.g., nitrofurantoin, chlordiazepoxide, chlorpheniramine) [10], certain paternal occupations (e.g., agriculture and forestry, mechanics, repairmen) [11], and fertility treatments [12]. Fibroblast growth factor and fibroblast growth factor receptor (FGFR) regulate fetal osteogenic growth and are expressed in cranial sutures in early fetal life. These factors possibly influence fetal suture patency [13].

1.2. Diagnosis

The diagnosis relies on physical examination and confirmed by radiographic studies. Clinical history should include complications of pregnancy duration of gestation, and birth weight [14]. Physical findings typically consist of bitemporal narrowing, a palpable midline forehead ridge at the site of the fused suture, and the characteristic deformity of a triangular anterior cranial vault to which the term *trigonocephaly* is applied. Compensatory parieto-occipital expansion is also seen, and hypotelorism may be observed in severe

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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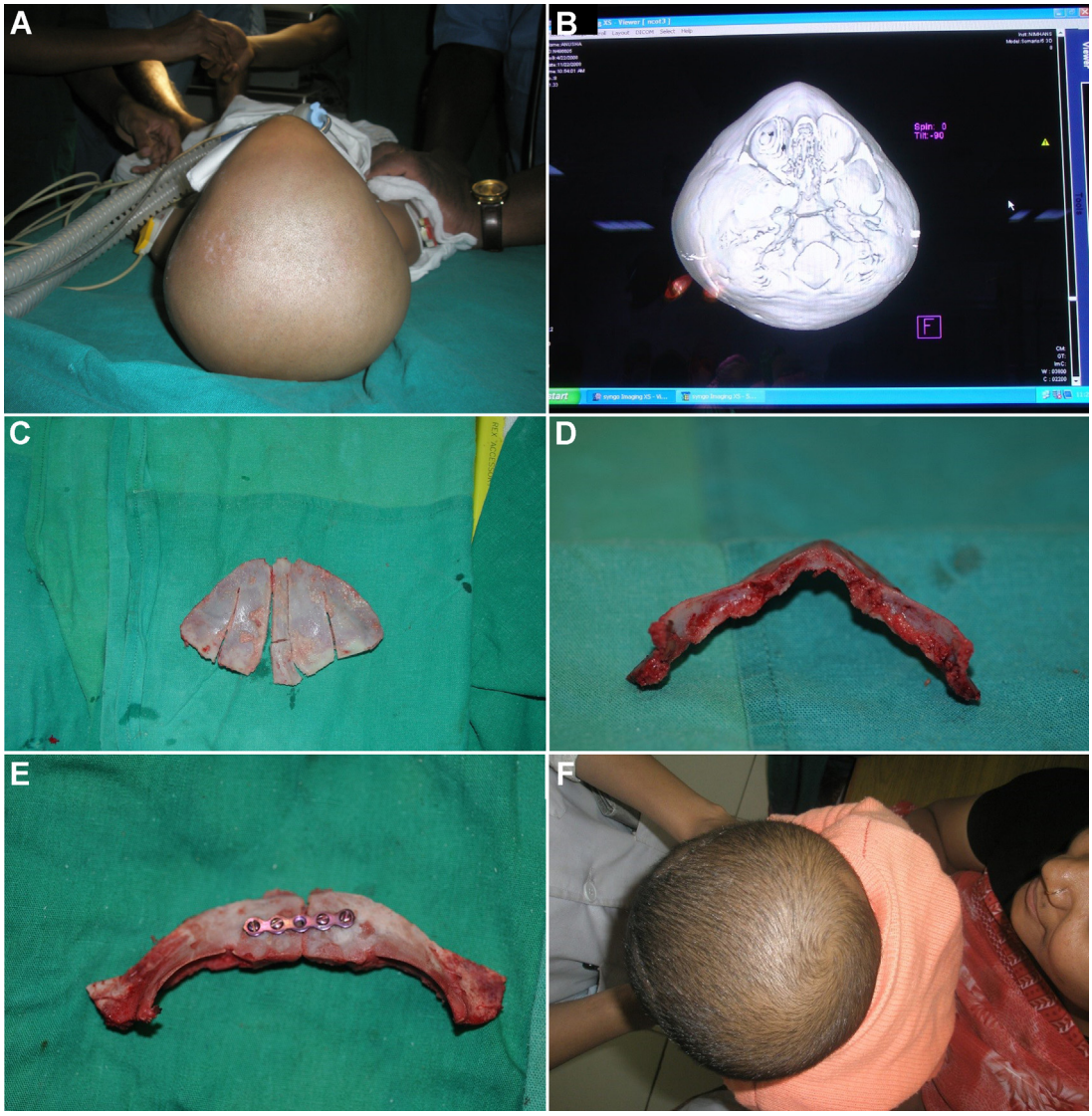

Fig. 1. (A) Pre-operative clinical picture showing shape of skull, (B) CT showing triangular deformity, (C) radial cuts made on the frontal bone flap, (D) deformed supra-orbital bar, (E) median osteotomy was carried out and bone fractured outward and secured with titanium miniplates and screws, and (F) post-operative head shape.

cases of metopic synostosis. Fusion of the metopic suture on computed tomography scan before 1–2 years of life has generally been considered consistent with the diagnosis of metopic synostosis [5,6].

1.3. Management

Given the variability in presentation, a variety of surgical techniques exist, ranging from synostectomy [15], the "floating forehead" (*le front flottant*) approach to cranial vault expansion [16], to fronto-orbital reshaping and expansion with interpositional bone grafts. The concept that surgery may improve cognitive function [17,18] and esthetic outcome, early surgical intervention is unequivocally beneficial in this patient population. Surgical reconstruction for metopic synostosis was carried out using an approach that involves bifrontal craniotomy. After frontal craniotomy, a standard fronto-orbital osteotomy with an orbital osteotomy at the frontozygomatic suture was carried out in all patients. The fronto-orbital bar was then split in the midline, and any prominence along the fused metopic suture was blurred away. The fronto-orbital bar was expanded and plated with microtitanium/biodegradable plates and screws. The frontal bone was recontoured, widened and plated to the newly positioned fronto-orbital bar with microplates. The scalp flap was returned to position and closed.

2. Case reports

2.1. Case I

A 10-month-old female baby was referred to our unit with the complaint of a congenital bony swelling in the middle of the forehead. She was a first born child of non-sanguineous marriage following a LSCS of a normal term delivery. The swelling was noticed by her mother immediately after her birth that progressively increased in size (presently 3 cm × 4 cm). On examination the anterior fontenell remained open and collapsed. Head circumference was 44 cm. Her cardiologic report was suggestive of congenital heart disease, situs solitus, and small ostium secundum ASD L to R shunt. Gessels children behavior schedule (GCBS) findings suggested that the child had acquired motor characteristics, adaptive behavior and personal–social behavior of 9 months and language skills of 6 months age level. CT head showed fused/absent metopic suture with prominence of frontal bone (Fig. 1).