Results of Contemporary Craniofacial Osteotomies in Treating Craniosynostotic Patients

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ABSTRACT
Craniosynostosis denotes the premature pathological fusion of one or more of the cranial sutures. The treatment of these cases has advanced rather dramatically over the past thirty years. A better understanding of the neurocranial development and calvarial osteogenesis, the various osteotomies and procedures utilized, rigid internal fixation, use of resorbables and team approach has played a role in the advances of management of these cases. This article presents our surgical results of thirty-seven craniosynostotic patients of different age groups and different suture involvement treated by contemporary craniofacial osteotomies over the past three years at Ain-Shams University Hospital. The same craniofacial team treated all cases. All patients underwent in-depth clinical evaluation including anthropometric craniofacial measurements and received CT scans prior to surgery. Preoperative ophthalmologic evaluation, genetic counseling and intelligence quotient (I.Q) tests were conducted routinely. Surgical planning and osteotomies were designed to meet the specific goals for each patient according to the site and number of sutures as well as patients’ age. Normalization of the cranial shape and the cephalic index was achieved following the surgical correction in all cases with only minor complications reported.

INTRODUCTION
Premature closure of any of the calvarial sutures prevents separation of the calvarial bones. Inevitably, this produces a restriction on growth vectors perpendicular to the affected suture with compensatory growth parallel to them, leading to a morphologic change in calvarial shape. These changes are specific and characteristic for every type of craniosynostosis [1-3]. Accordingly, an increase in the anteroposterior length of the calvarium accompanied by a reduction in the bitemporal width denotes sagittal suture synostosis or scaphocephaly. On the other hand, a reduction in the anteroposterior length characterizes coronal synostosis or brachycephaly. While unilateral affection of the coronal suture, known as frontal plagiocephaly, appears as unilateral flattening of the forehead with recession & elevation of the brow and supralateral aspect of the orbit on the affected side.

Pathological fusion of the metopic suture, known as trigonocephaly, presents as a keel-shaped deformity of the forehead with bitemporal narrowing and associated hypotelorbitism. The sequence of events leading to premature ossification of sutures is unknown. Biomechanical forces and genetically determined local expression of growth factors have been implicated in the etiology of craniosynostosis [4-9]. Animal experiments and a recent interest in molecular biology point toward interplay between the dura and the underlying brain. This interaction occurs by means of a local alteration in the expression of transforming growth factor, MSX2, fibroblast growth factor receptor, and TWIST. The dura also serves as an intermediary source of signaling, which is mediated by transforming growth factor, fibroblast growth factor receptor, TWIST, and MSX2. Genetic studies have now determined that mutations within these factors are responsible for various types of craniosynostosis [7,10-12]. Fibroblast growth factor receptors 2 and 3 mutations were present in all patients with syndromic craniosynostosis [13]. The specific source generating these signals and gene amplification is not yet understood. Furthermore, the genetic causes of single-suture craniosynostosis are still largely unknown, as mutations in common craniosynostosis-associated genes and structural chromosomal aberrations have been rarely found in these cases [14]. The growing study of genetics, biotechnology, the influence of growth factors, and stem cell research provide additional opportunities of innovation for the future [15]. Uncorrected synostosis, quite frequently, is associated with a deformity that progresses to involve the facial skeleton resulting in asymmetry of the face and malocclusion. Asymmetry of the orbit leads to ocular dystopia and consequent strabismus. Moreover, if the synostosis goes un-
corrected, the intracranial pressure increases. This has been documented in the animal model and in humans [16,17]. Failure to remove the synostosis would lead to unfortunate consequences, including hydrocephalus, blindness, mental retardation, and premature death. Therefore excision of the synostosed suture and maintenance of the suturectomy site as a nonossified zone into childhood is necessary for normal cranial growth and development. Currently, the principles of surgical intervention are not only to excise the fused suture(s) but also attempt at normalizing the calvarial shape [18]. Minimally invasive procedures such as endoscopic suturectomies, spring-mediated cranioplasties, and distraction osteogenesis are being discussed in the recent plastic and neurosurgical literature. However, there are disagreements among craniofacial centers on the most optimal timing and best operative procedure [19]. While spring-mediated cranioplasty is advocated by one group [20], it is reported to be associated with higher complications by another group [21]. Despite this controversy, it is agreed upon that the surgical goal is to increase the intracranial volume, especially under the fused suture, and prevent any long-term complications. Normalization of the calvarial shape successfully using variations of calvarial vault reconstruction and fronto-orbital advancement remain the gold standard in achieving this goal.

Aim of work:

The aim of this work is to present the results of using contemporary craniofacial osteotomies in treating craniosynostotic patients of variable age groups with different suture involvement.

PATIENTS AND METHODS

Patients diagnosed with craniosynostosis presenting to the craniofacial clinic at the Plastic Surgery Department, Ain-Shams University, Cairo, Egypt were included in this follow-up study. A craniofacial team composed of the same craniofacial plastic surgeon, neurosurgeon and pediatric anesthetist treated all cases. As a standard care of practice, all patients underwent in-depth clinical evaluation including anthropometric craniofacial measurements and calculations of the cephalic index (the cranial index is the ratio of maximum calvarial width divided by maximum calvarial length multiplied by 100) [22]. CT scans were performed on all cases prior to surgery. Genetic counseling was done consistently. Preoperative ophthalmologic examination including fundus examination to detect any raised intracranial pressure was conducted routinely. Intelligence quotient (I.Q) tests were carried out to evaluate mental functioning. Antibiotics were started the night before surgery and continued for one week.

The surgical procedure was carefully explained to all parents and to the adolescent patients. Surgery was performed between six and twelve months of age whenever patients presented early. Exposure was carried out through a coronal approach. Metallic plates were still used mostly in older patients, although the majority of younger patients received resorbable plates & screws for fixation.

Patients underwent variations of calvarial vault reconstruction and fronto-orbital advancement, which are the mainstays of treatment for craniosynostosis. Surgical planning and osteotomies were designed to meet the specific goals for each patient according to the site and number of sutures as well as patients’ age. For sagittal synostosis, a modified “pi”-type strip craniectomy on either side of the sagittal & coronal sutures along with vertical barrel stave osteotomies on the parietal & lambdoid bones were performed (Fig. 1). For patients older than one year, total vault remodeling was carried out. Recontouring the supraorbital bandeau was done for brachcephalic cases. If these cases were accompanied by compensatory bossing, flattening was achieved by flipping of the temporal bone (Fig. 3). Advancing the frontoparietal bones and the supraorbital bandeau was done for brachcephalic cases. If these cases were accompanied by compensatory temporal bossing, flattening was achieved by flipping of the temporal bone (Fig. 3). In cases with plagiocephaly, treatment was individualized according to the extent of hemicoronal ring involvement [23]. If confined to the frontoparietal suture with moderate deformity then they were treated unilaterally in the form of unilateral fronto-orbital advancement and forehead remodeling (Fig. 4). If Fusion involved the whole coronal hemiring (i.e., frontoparietal, frontosphenoid, frontothmoid sutures) indicating a more extensive deformity, it was corrected bilaterally. Orbital reconstruction was also carried out to mimic the contralateral normal orbital measurement (Fig. 5). In older patients, facial asymmetry and deviation was corrected by performing an adjuvant naso-orbito-ethmoidal osteotomy and a sliding genioplasty (Fig. 6). In craniosynostotic patients accompanied by midface retrusion, monoblock advancement was considered starting from around the age of six years of age (Fig. 7).
Patients were transferred to the ICU for at least 24 hours postoperatively, and a CT scan was obtained within the first week as part of the protocol. Any intraoperative or postoperative complications in recovery or any later complications including readmission to hospital and reoperation were recorded.

RESULTS

Thirty seven patients were surgically treated over the past three years. They were sixteen males and twenty one females. Twelve patients presented before they reached one year of age while sixteen patients were between one and four years at the time of presentation. The remaining nine patients presented later on with four of them in their late teens. The average age of the patients at the time of the operation was two years and three months (range: 6 months to 18 years). Isolated single suture synostosis comprised twenty six patients; three with sagittal synostosis, another five with coronal synostosis, eight with unilateral coronal synostosis, and ten with metopic synostosis. Eleven patients presented with multiple suture synostosis, eight of them were syndromic cases.

Intelligence quotient (I.Q) values ranged from 39 to 109 (average 84). These values did not show significant changes from the preoperative to the six months postoperative values for each patient. Ophthalmological examination confirmed starting mild papilleodema in five cases that warranted urgent surgical interference. Only in one case of a cloverleaf deformity that papilloedema was severe and the patient was losing her sight because of the profound increase in intracranial pressure.

Normalization of the cranial shape and the cephalic index was achieved following the surgical correction. In cases of scaphocephaly, reducing the anteroposterior length and increasing the transverse width improved the calvarial contour. In treating trigonocephaly, the surgical aim was achieved by repairing the misshapen keel-shaped forehead and increasing the volume of the anterior cranial fossa laterally (Fig. 2). In cases of brachycephaly, improving the calvarial shape was achieved by the ventrally advancing the frontoparietal bones and the fronto-orbital bar (Fig. 3). In cases of frontal plagiocephaly, excising bone at the frontozygomatic and nasomaxillary sutures reduced the height of the elongated ipsilateral orbit, while inserting a cranial bone graft into the supraorbital rim increased the width of the narrow ipsilateral orbit. Further symmetry was achieved by reconturing of the forehead and orbital bandeau (Figs. 4,5,6). Simultaneous advancement of the forehead and the face done in older patients with midfacial retrusion corrected the associated exorbitism and improved both the forehead and facial contours (Fig. 7).

Minor dural tears occurred intraoperatively in two cases. They were repaired prior to fixing the osteotomized segments and passed uneventful in the postoperative period. Partial disruption of the coronal wound occurred in one case and was managed by secondary sutures. There were no complications related to using the resorbables for fixation. In one older patient, the metal hardware was visible and was removed upon her parents request. Neither disastrous complications such as meningitis, sinus hemorrhage and brain cortical damage occurred nor morbidity related to intraoperative blood loss.

Fig. (1): A modified “pi”-type strip craniectomy on either side of the sagittal & coronal sutures along with vertical barrel stave osteotomies on the parietal & lambdoid bones are performed on sagittal synostosis. Note that the osteotomy does not cross the normal coronal suture and that the parasagittal strips are advanced by wires which will compress the skull in the antero-posterior dimension (and expand transversely).
Fig. (3): A four-year-old male child presenting with brachycephaly.
(A) Intraoperative view demonstrating the turn over of the temporal bone to compensate for the increase of bitemporal width. (B) Preoperative lateral view. (C) Six months postoperative lateral view. (D) Preoperative front view. (E) Six months postoperative front view.

Fig. (4): A one-year-old female child presenting with right frontal plagiocephaly. The synostosis is confined to the frontoparietal suture with moderate deformity hence the unilateral frontoorbital advancement and forehead remodeling. (A) Preoperative front view. (B) Preoperative top view. (C) Three months post-operative front view.
Fig. (5): A child with left sided frontal plagiocephaly with fusion involving the whole coronal hemiring. (A) Immediate preoperative front view at 6 months of age. (B) An intraoperative view demonstrating bilateral frontorbial advancement and forehead remodeling performed with orbital reconstruction carried out to mimic the contralateral normal orbital measurement. Note the use of resorbable plate and screws. (C) Three years postoperative front view with minimal residual right temporal hollowing.

Fig. (6): A case of left-sided plagiocephaly presenting late. (A) Front view demonstrating the twist involving the whole craniomaxillofacial skeleton and not confined to upper third. (B) Cephalometric tracing showing the difference in orbital dimensions as well as the midline shift. (C) Preoperative worm’s view. (D) One year postoperative worm’s view following cranial remodelling, naso-orbito-ethmoidal osteotomy and a lateral sliding genioplasty.

Fig. (7): A sixteen-year-old female with Crozon syndrome. (A) Intraoperative view demonstrating a monoblock advancement secured with miniplates at the zygomatic arch. (B) Preoperative front view demonstrating exorbitism. (C) One and half years postoperative front view. (D) Preoperative oblique view (E) One and half years postoperative oblique view. Note the fullness of the malar eminence and the maintenance of the nasofrontal suture.
DISCUSSION

Lannelongue [24] performed the first reported surgical procedure for correction of craniosynostosis in 1890 through releasing, but not resecting, the fused suture. Two years later, Lane [25] described the first strip craniectomy, with extraction of the fused suture, but unfortunately, with death of the child postoperatively. The reported series of complications and deaths associated with craniosynostosis surgery led to its suspension. Thirty years later, Mehner (1921) [26] described the first successful use of strip craniectomy to remove a fused suture. The aim has been to excise only the fused suture in the hope that the secondary changes would automatically correct themselves as the brain grows in the first year of postnatal life [27,28]. Over time, reossification at the site of suturectomy became problematic, leading to interventions that included repeat osteotomy, adding chemical agents to the dura to prevent reossification, or addition of mechanical barriers of alloplastic materials to prevent this problem [29]. More radical craniectomies were also discussed by Faber and Towne [30], but it was not until the work of Tessier [31,32] in the late 1960s and early 1970s that cosmetic objectives began to receive emphasis. Although strip craniectomy was used often, it lost much support with the advent of cranial vault reconstruction.

The quality of care given to patients in this study met the international standards set by benchmarking on craniosynostosis [33,34,35]. The same craniofacial team that composed of a craniofacial plastic surgeon, a neurosurgeon and a pediatric anesthetist treated all cases. This contributed to greater operative efficiency, better perioperative anaesthetic techniques & diminished intraoperative blood loss as documented by other studies [36,37]. CT scans were performed on all cases prior to surgery. Genetic counseling was done consistently. Antibiotics were started the night before surgery and continued for one week. All patients were transferred to the ICU for at least 24 hours postoperatively. Furthermore, in this study, preoperative fundus examination along with intelligence quotient (I.Q) tests were conducted routinely for all patients.

Increased intracranial pressure in children with nonsyndromic synostosis has been reported and has been assumed to affect mental development [16,38,39]. A number of studies have suggested that children with nonsyndromic craniosynostosis are at increased risk for learning disorders and mental retardation, regardless of whether surgery is performed [40]. There has been considerable debate regarding the hypothesis that surgical treatment of single-suture craniosynostosis prevents retardation or improves global cognitive functioning at a later age. Cohen & colleagues [41] documented mild delay in mental and motor ability before surgery that was improved for motor but not for mental ability following surgical repair of single-suture craniosynostosis. At present, multicenter studies are ongoing to better elucidate this question, but current opinion seems to find little support for the contention that surgery is therapeutic in this regard [40]. In this study, intelligence quotient (I.Q) did not show differences between the preoperative and the six months postoperative values. However, because of the small sample size in this study, relatively short term follow-ups, together with the diversity in age and type of sutures involved, it was difficult to draw any conclusions in this regard.

It is interesting to observe that unlike other studies, [42-44] sagittal suture synostosis did not present in this series as the most common form of nonsyndromic single suture synostosis. This could be explained by the fact that the majority of these children are still treated at an earlier stage in life by strip craniectomies by the neurosurgeons in our country. On the other hand, metopic suture synostosis comprised the largest number of patients in this series accounting for ten out of twenty six patients. Although this observation is dissimilar to classic literature [45,46] it concurs with Selber’s et al. [47] findings that metopic synostosis is on the rise. This could be explained in part by the fact that metopic synostosis is the only form of single suture synostosis that has an association with family history [48,49] and that increasing proportions of syndromic patients may be clues to the etiology of this epidemiologic event [47].

There are many variations of calvarial remodeling and orbitofrontal advancement that are considered the mainstay surgical techniques for treating craniosynostosis [50-53]. These techniques have been refined considerably and the selection from them is usually based on the surgeon’s preferences, training, and collegial interactions. Experience has shown that more extensive reshaping yields excellent results, particularly in older children with moderate to severe deformities [54]. In agreement with other craniofacial surgeons, patients in this study underwent more aggressive remodeling procedures to excise the fused suture(s), correct the associated deformities and promote normal calvarial growth.

Specific findings tend to occur in particular malformations, but each patient is unique. Reconstruction of the specific aesthetic units of the
frontoforehead, posterior cranial vault, orbito-naso-zygomatic and maxillary-nasal base to its normal proportions is essential in achieving normal morphological appearance of patients with craniosynostosis [55]. Establishing the normal position of the supraorbital ridge-lateral orbital rim region and the superior forehead aesthetic units is critical to the overall facial symmetry and balance [56]. In this series, the eyebrows with overlying supraorbital ridge were always placed in a plane anterior to the cornea and arced gently posteriorly into the temporal fossa. The forehead component was remodeled and positioned so that it has a gentle posterior curve that levels out at the coronal suture region when seen in profile. Surgical advancement of the retruded midface with simultaneous reduction of the increased transverse width restored the balance of the orbito-naso-zygomatic aesthetic unit.

Because the face grows as a template off the cranial base, uncorrected synostosis leads to asymmetry of the face and occlusal plane. An older child with uncorrected unicoronal synostosis demonstrates asymmetry of the facial skeleton. This is associated with a reduction in height of the ipsilateral maxilla and mandible, resulting in an oblique occlusal cant [57]. These findings were evident in two of our older patients presenting with frontal plagiocephaly. In addition to performing adjuvant naso-orbito-ethmoidal osteotomies at the time of cranial surgery, these patients received sliding genioplasties later on as part of their staged reconstruction plan.

In addition to methods of treatment, the timing of intervention is controversial, without much scientific evidence to support early surgical treatment in infancy versus late treatment [58]. Practitioners who prefer early intervention (e.g. before 6 months of age) do so for several reasons, believing that the operation is technically easier secondary to the plasticity of the calvaria, the osseous defects created by cranial vault reconstruction are more reliably healed with earlier treatment, and the morphologic results will be superior. Others believe surgery should be delayed until a year or more of life, thinking that operating on more mineralized bone present at that age will prevent recurrence of the deformity. A delay in surgery beyond the first 9 to 12 months of life leads to progressive deformity of the cranial base, resulting in abnormal facial growth and asymmetry of the maxilla and mandible [13]. It has been our protocol to operate around 9 months whenever these kids presented early. Earlier intervention was done on selected cases for functional reasons.

Bioreabsorbable fixation has been widely used for cranial reconstruction in the pediatric population. It avoids the complications of metal hardware migration and imaging artifacts [59-61]. However, concerns of inflammatory reactions, granuloma formation, and incomplete resorption have been lately raised [62]. A recent study conducted on pediatric patients under the age of 2 years with follow up to seven years confirmed the safety of the use of bioresorbables with low morbidity rates [63]. The results of this work coincide with this conclusion regarding the safety of using biodegradables with minimal morbidity in the pediatric population.

In conclusion, contemporary surgical management of craniosynostotic patients requires the collaborative work of a craniofacial team composed of a craniofacial plastic surgeon, a neurosurgeon, and a pediatric anaesthetist. The goals of surgical intervention should be individualized for each case and include both the release of the affected suture(s) to allow for unrestricted cranial development and the establishment of normal aesthetic units of the craniofacial skeleton. Surgeons caring for patients with craniosynostosis must maintain a thorough understanding of the 3-D anatomy, characteristic dysmorphology associated with different types of synostosis, and the variable osteotomies available for surgical correction.

REFERENCES

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