A New Operative Open-Wings Technique to Correct the Frontoforehead Unit in Metopic Synostosis

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Abstract: The technology adoption and creation of a multidisciplinary team have helped to overcome the complexity associated.

Craniofacial surgery has thus emerged from the valuable contributions of neurosurgery, maxillofacial surgery, plastic surgery, eyes, nose, and throat as well as head and neck surgery. A patient with trigonocephaly may present a prominent "keel" forehead, accompanied by recession of the lateral orbit rims, hypotelorism, and constriction of the anterior frontal fossa when the metopic suture fuses before 6 months of age. In a period between 2007 and 2011, in the Salesi Children's Hospital, were treated for nonsyndromic variety of metopic synostosis 11 infants; their ages ranged from 6 months to 9 months, and 7 were males and 4 females. The most important aims of our new surgical technique are the achievement of symmetry as well as normal proportion and reconstruction of the frontoforehead unit but remaining in a very conservative treatment. The morphology and position of the supraorbital ridge-lateral orbital rim region are key elements of upper facial esthetics. This new "open-wings" technique for the reconfiguration of the bilateral emisupraorbital bar requires a midline incomplete osteotomy that involves only the internal cortex of the frontonasal region. Hence, both lateral orbital walls are bent inwardly and tilting forward, as in computed tomographic scan planning, with a greenstick fracture pivoting on the preserved medial frontonasal region. This open-wings conservative technique allows the avoidance of the most important complication that may result in the traditional way such as dead space in the anterior cranial fossa, infections, and blood loss but with an achievement of satisfactory craniofacial form and aesthetic result.

Key Words: Trigonocephaly, supraorbital bar, open-wings technique, craniofacial malformations

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Craniofacial surgery is a relatively new specialty because most of developments have occurred in the 20th century. The technology adoption and creation of a multidisciplinary team have helped to overcome the complexity associated. Advances in anesthesia, surgical techniques, and technology have enhanced good outcomes and safety. Craniofacial surgery has thus emerged from the valuable contributions of neurosurgery, maxillofacial surgery, plastic surgery, eyes, nose, and throat, as well as head and neck surgery.

The term *craniosynostosis* was credited to Otto in 1830 and Virchow in 1831, but the first surgical correction was performed by Lannelongue in 1890. In 1851, Virchow recognized that premature closure of suture line causes limitation of growth perpendicular to that suture and also expansion in the direction of the suture line. When a suture fuses prematurely, the skull restricts the growing brain beneath the suture, thus allowing expansion into regions of less restriction, and this "compensatory" growth of the skull occurs largely in planes parallel to the affected suture, resulting in consistent, recognizable cranial deformity.[1,2]

The incidence of nonsyndromic craniosynostosis has been reported as 0.4 to 1 per 1000 live births. Most of the sutural abnormalities are isolated (nonsyndromic), but approximately 15% are associated with syndromes (syndromic).

The modern era of craniofacial surgery was born with Tessier in the 1960s and 1970s; he established the key principle that large segments of the cranial vault could be completely stripped of blood supply and yet can survive entirely. He asserted several basic principles: (1) correction requires a wide subperiosteal exposure of the face and the orbit; (2) the affected orbit can be moved safely in any direction without risking vision or oculomotor dysfunction; (3) osteotomy and repositioning of the facial structure produce better results than the sole use of bone grafting does; and (4) as many deformities as possible should be corrected during the same operative procedure.[3]

In 1976, Hoffman and Mohr[4] recognized the inadequacy of craniotomy for the correction of craniostenosis and introduced the concept of mobilization of the supraorbital bar with an osteotomy of the orbital process.

The development of power drills, piezosurgery bone scalpel, and craniotomies significantly decreased the operating time, reduced blood loss, and eliminated the cosmetically bad aspect.

Metopic synostosis is often accompanied with a variable degree of phenotypic severity. Metopic synostosis is now the second most common form of craniosynostosis (23.7%–23.3% of cases) and shows a male predominance of 75%. Premature closure of the metopic suture may lead to the formation of a triangular head, otherwise known as trigonocephaly. Patients may present with a prominent "keel" forehead, accompanied by recession of the lateral orbital rims, hypotelorism, and constriction of the anterior frontal fossa when the metopic suture fuses before 6 months of age. If the suture fuses later than 6 months, then the deformity is minimal or not seen at all. Diagnosis is made clinically because the metopic ridge or notch is palpable and the shape of the head is characteristic. Confirmation of the clinical findings is obtained with computed tomography (CT) and three-dimensional reconstructions.[5]

The metopic suture fuses from the glabella and progresses superiorly; as a consequence, premature fusion produces incomplete

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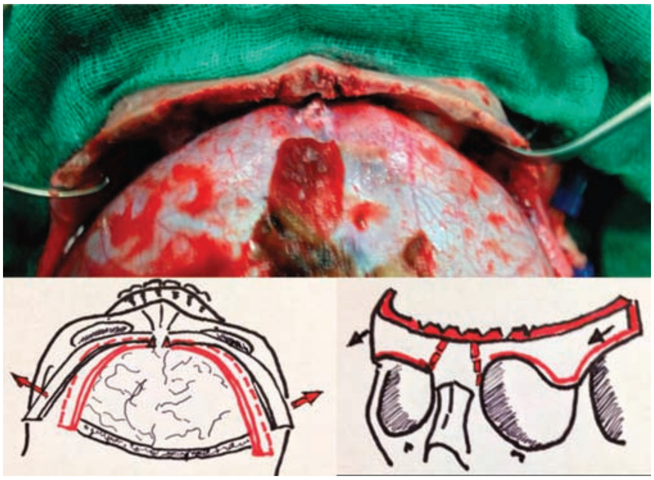

FIGURE 1. Two emisupraorbital bars with an incomplete osteotomy of the internal cortex of the frontonasal region. Both lateral orbital walls are bent and tilted forward according to the CT scan planning. With a greenstick fracture pivoting on the preserved medial frontonasal region.

development of the forehead and orbital spacing. Imaging shows hypotelorism, narrow anterior cranial fossa, hypoplasia of the ethmoid sinuses, and bitemporal narrowing with anterior bowing of the coronal sutures. Distally, there is a compensatory expansion of the occipitoparietal region. The ‘‘dura theory’’ proposes that the dura is associated with a regulatory role in the growth of the skull and fusion of the sutures, acting as an internal periosteum with an osteogenic or directional role. Initially, the craniosynostotic syndromes were classified on the basis of their clinical findings, but now most of the syndromes are defined on a molecular level, according to specific genetic mutations. Mutations in the gene coding for fibroblast growth factor receptor (FGFR1, FGFR2, FGFR3) are responsible for a fraction of the craniosynostotic condition as in trigonocephaly.[6,7]

MATERIALS AND METHODS

In the last 5 years, in a period between 2007 and 2011, in the Ancona Salesi Children's Hospital, were treated for nonsyndromic variety of metopic synostosis 11 infants; their ages ranged from 6 months to 9 months, and 7 were males and 4 were females. All of them underwent a detailed evaluation by a craniofacial team, which includes clinical, morphologic, neurologic, and radiologic assessments. Computed tomographic scanning of the cranial vault, base, and facial structures is very important. Three-dimensional reconstruction of the cranium, orbits, and face is particularly useful in demonstrating the general volume, relationships, and shape of the underlying bony structures as well as in planning the surgery time. The optimal timing of surgical treatment of craniosynostosis is not yet clear; a majority of craniofacial surgeons perform operations on patients between 3 and 12 months of age. Our philosophy, however, is to operate as soon as the patient is able to withstand the stress of surgery to capitalize on the ameliorating effects of brain growth on the overall skull shape. In practice, the timing usually turns out to be approximately 6 to 9 months of age and earlier if there is evidence of increased intracranial pressure (bulging fontanel, progressive optic atrophy).[8,9]

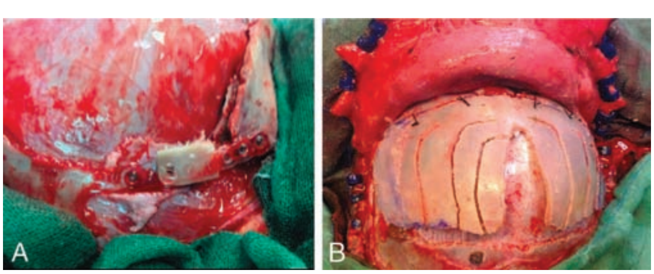

FIGURE 2. A, Frontal bone reconstructed using a remodeled portion of the calvaria. B, Bone graft interposed in the temporoparietal region and fixed with absorbable plates and screws.

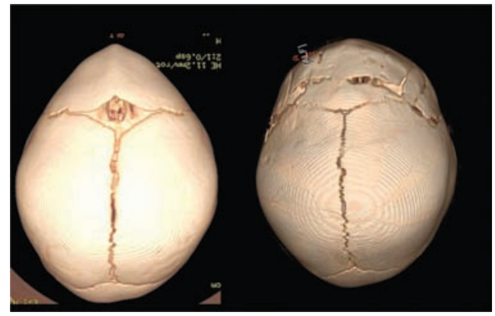

FIGURE 3. Preoperative and postoperative CT three-dimensional scan.

Surgical Technique

A coronal zigzag-line skin incision is made using a No. 15 blade and is followed by electrocautery dissection using a Colorado tip to minimize bleeding. The frontal scalp flap is elevated in the subgaleal plane, and the pericranial flap is elevated separately. This latter flap is saved because it is valuable and very important for protecting the dura mater in the bony defect area. Bony exposure is carried down distal to the nasofrontal junction, to both the zygomas, temporal areas, as well as the superior, medial, and lateral orbital walls. Bifrontal craniotomy is performed by the neurosurgical team. After the craniotomy, the dura mater is cautiously separated from the floor of the anterior cranial fossa, and the supraorbital bar with horizontally long temporal extension up to the medial naso-orbital region is performed bilaterally using a piezosurgery bone scalpel. This new technique for the reconfiguration of the bilateral emisupraorbital bar requires a medial incomplete osteotomy that involves only the internal cortex of the bilateral frontonasal region. Hence, both lateral orbital walls are bent and tilting forward, as in CT scan planning, with a greenstick fracture pivoting on the preserved medial frontonasal region (Fig. 1). The lateral temporal extension is advanced, and calvarial bone graft is interposed and rigidly fixed with absorbable plates and screws to reduce iatrogenic synostosis and leave no residual foreign bodies (Fig. 2).[10] The most important goal of the anterior calvarial remodeling for the correction of trigonocephaly is the reshaping of the orbital and temporal regions.[11,12] Advancement, tilting, and pivoting of the bilateral emibandeau are planned to achieve a symmetric and aesthetically satisfying contour of the supraorbital rim (Fig. 3). Fearon et al[13] demonstrated that intercanthal and interorbital distances increase significantly even if the orbits themselves are not surgically corrected.

The frontal bone is reconstructed using the remaining and remodeled portions of the calvarial bone. It is often possible to reverse the original frontal bone flap (posterior portion now in an anterior position) to obtain an adequate width and contour with the new frontal bone flap.

DISCUSSION

The frontoforehead region is dysmorphic in an infant with metopic synostosis. Defining the normal position of the forehead is critical to achieve overall facial symmetry and balance.[12] The forehead may be considered as 2 separate esthetic components: the supraorbital ridge-lateral orbital rim region and the superior forehead. The supraorbital ridge-lateral orbital rim region includes the glabella and supraorbital rim and extends inferiorly down each frontozygomatic suture toward the infraorbital rim and posteriorly along each temporoparietal region. The morphology and position of the supraorbital ridge-lateral orbital rim region are key elements of upper facial esthetics.[12]

The most important aim of our new surgical technique is the achievement of symmetry as well as normal proportions and reconstruction of the frontoforehead esthetic unit but remaining in a very conservative treatment.

Why this new open-wings surgical conservative technique? Nowadays, the evolution of surgical techniques has included more often conservative treatments of the involved sphenoid bone with the simultaneous correction of the hypotelorism in patients with significant trigonocephaly.

In the metopic synostosis, the most important aesthetic defects are represented by the midline bone keel and hypotelorism, and normally, the midline keel starts above the glabellar-frontonasal junction.[8,14,15]

Hence, this technique wants to preserve the glabellar region and the medial orbital wall with the intercanthal distance not involved in the anomaly growth but contextually to solve the hypotelorism putting forward the tow emisupraorbital bar end lateral orbital wall, not removing the entire supraorbital bone bar as the traditional surgical way.[16–18]

An extensive and traditional complete orbital bandeau may be followed by extraocular imbalance in the postoperative period that normally resolves spontaneously within 6 months.

This open-wings conservative technique allows the avoidance of the most important complication such as dead space[19,20] in the anterior cranial fossa, infections, and blood loss but with an achievement of satisfactory craniofacial form and aesthetic result (Figs. 3–5).[21–24]

Despite the complexity of craniofacial surgery, the infection rate is low in all series reported less than 7%, but a complete orbital bandeau and its forehead advancements create potential dead space in which the risk for epidural abscess is inversely proportional to the rate of brain expansion. Actually, the management of dead space during cranial vault/cranial base expansions is critical to limit complications. The forward advancement of the anterior cranial base, orbits, and midface results in both extradural (retrofrontal) dead space and communication with the nasal cavity. This new anatomic situation may result in hematoma formation, leakage of cerebrospinal fluid, infection, and fistula formation.

Normally, the fronto-orbital advancement performed in infants leads to significant blood loss and 80% to 100% of patients require blood transfusion. In all these children who underwent the standardized fronto-orbital advancement applied to trigonocephaly, the amount of blood loss during operation is ranging from 80 mL to 600 mL, with an average of 220 mL.

In the new open-wings surgical technique, the respect of the frontonasal junction anatomic unit completely avoids the dead space and, with the use of a piezosurgery bone scalpel, allows a more conservative blood loss.

FIGURE 4. Preoperative and postoperative pictures of the patient 2 years after the surgery.

FIGURE 5. A and B, Frontal preoperative and postoperative pictures. C and D, The difference from back view before and 3 years after the surgery.

In our experience with this new conservative technique, we did not have any complications that resulted in dead space in the postoperative period and the average of blood loss was 130 mL; in only 1 case the infants required blood transfusion.

These good results encourage our team to continue in this kind of conservative new surgical way with the awareness that the traditional surgical approach is the basis of the pediatric craniofacial surgery.

SUMMARY

The technology adoption and creation of a multidisciplinary team have helped to overcome the complexity associated. A patient with trigonocephaly may present a prominent "keel" forehead, accompanied by recession of the lateral orbit rims and hypotelorism. In a period between 2007 and 2011 were treated for metopic synostosis 11 infants with ages ranging from 6 to 9 months. The morphology and position of the supraorbital ridge-lateral orbital rim region are key elements of upper facial aesthetics. This new open-wings technique for the reconfiguration of the bilateral emisupraorbital bar requires a medial incomplete osteotomy that involves only the internal cortex of the frontonasal region, in which lateral orbital walls are bent and tilting forward pivoting on the preserved medial frontonasal region. The advantages of the open-wings technique are as follows:

1. Reduce osteotomies, particularly at the level of the anterior cranial base. We avoid some possible complications such as infection, liquoral fistula, and alteration of the olfactory nerve.
2. The frontal bandeau is not completely mobilized. This permits having a central fixed point to start for the reconstruction of frontal bandeau and volet.
3. The main goal of this new procedure is to limit surgical time and facilitate reconstruction. The minor surgical time is an advantage for less bleeding and edema in newborn patients. With this technique, the authors correct at the same time the projection and form of frontal bandeau and volet with good symmetry.

Clinical and development cranial forms demonstrated that this is a useful technique.

The authors added intraoperative pictures and another clinical case with stable results 3 years after surgery.

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