Advances in the Treatment of Syndromic Midface Hypoplasia Using Monobloc and Facial Bipartition Distraction Osteogenesis

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Semin Plast Surg 2014;28:179–183.

Abstract

Keywords

- monobloc
- facial bipartition
- syndromic
cranosynostosis
- frontofacial
advancement
- midface hypoplasia
- distraction
osteogenesis

Midface hypoplasia or retrusion remains a persistent feature of syndromic craniosynostosis years after successful treatment of the cranium. Although expansion of the cranial vault in infancy by traditional fronto-orbital advancement, posterior expansion, or both, can treat the immediate intracranial constriction, midface hypoplasia and its stigmata of exorbitism, sleep apnea, central face concavity, and malocclusion remain suboptimally treated. Initial enthusiasm for the procedures was tempered due to a high rate of infectious complications; timing and indications for surgery continue to stir controversy. During the last decade renewed interest with the monobloc and facial bipartition procedure using distraction osteogenesis with either an internal or external distraction system has decreased morbidity significantly. These procedures have re-emerged as powerful and comprehensive tools in the treatment of syndromic midface hypoplasia.

Background

Midface hypoplasia or retrusion remains a persistent feature of syndromic craniosynostosis years after successful treatment of the cranium. Although expansion of the cranial vault in infancy by traditional fronto-orbital advancement, posterior expansion, or both, can treat the immediate intracranial constriction, midface hypoplasia and its stigmata of exorbitism, sleep apnea, central face concavity, and malocclusion remain suboptimally treated.¹ To comprehensively treat both midface hypoplasia and residual cranial constriction, the monobloc operation was devised, reported, and popularized by Ortiz-Monasterio et al (1978).² Further refinement of the monobloc with splitting of facial bone segments in the midline to correct hypertelorism was reported the subsequent year (1979) by van der Meulen et al and further refined by Tessier into the facial bipartition operation after recognizing that the facial deformity particularly in the Apert condition also included a concavity of the central face.^{3,4} The use of the facial bipartition procedure to correct the central

concavity associated with the Apert condition was recently rerecognized and reported by Greig et al.¹ These powerful surgical techniques combining the Lefort III osteotomy with the orbital bandeau with or without bipartition seemed to finally correct the vexing problem of normalizing the face and cranium in complex craniosynostosis patients.

During the 1980s, increasing experience with these procedures at various centers revealed a significant drawback to the monobloc/bipartition operation. The large retrofrontal space (►Fig. 1) created after acute expansion and the subsequent communication between the nasal cavities and the anterior cranial fossa led to a high (> 30%) infection rate that manifested as meningitis, epidural abscess, and/or osteomyelitis of the frontal bones and subsequent bone loss.^{5,6} These significant complications tempered the initial enthusiasm for frontofacial advancement. Polley and colleagues, encouraged by favorable outcomes using distraction osteogenesis (DO) in the mandible, reported in 1995 the application of distraction techniques with the gradual advancement of the monobloc frontofacial segment in an infant.⁷ Their technique using an

Fig. 1 Oblique view of the visible upper orbits after monobloc frontofacial separation. The laminar spreader and pediatric Rowe disimpaction forceps have been used to advance the facial segment prior to internal fixation. Note the significant retrofrontal dead space created by the acute advancement and the communication with the anterior and middle ethmoid sinus.

external distraction device to gradually advance the facial monobloc segment resurrected frontofacial advancement surgery. Subsequent reports by Bradley et al and Arnaud et al in 2006 and 2007, respectively, documented negligible infectious complications, avoidance of frontal bone loss, meningitis, and epidural abscess using internal distraction systems.^{8,9} By 2008, Bradley and colleagues at the University of California Los Angeles reported the successful distraction of the facial bipartition segments after midline osteosynthesis with minimal infectious complications and a low rate of relapse (< 10%).¹⁰ More recently, Dunaway et al have reported on their results at the Great Ormond Street Hospital for Children and performed a review of the literature that confirms the decreasing trend in complications over time with increasing team experience and the use of DO.¹¹

The major advantage of DO using a 1-week latency period followed by a daily advancement of 1 mm (0.5 mm twice daily) was the sealing of the skull base after osteotomy and complete obliteration of the anterior cranial fossa dead space by gradual expansion of the viscerocranium contents mainly by expansion of the cerebrospinal fluid space (hydrocephalus ex vacuo). This process was initially studied and reported by Posnick and colleagues. They demonstrated obliteration of epidural dead space in a patient after frontofacial advancement by 6 weeks.¹² This process is similar to epidural space obliteration after reconstructive cranioplasty. This process, reported by military surgeons Kumar et al during Operation Iraqi Freedom, has physiologic limitations. Epidural dead space > 2 cm in any plane between the undersurface of the cranial reconstruction and the underlying dura immediately after cranioplasty was found to correlate with high rates of infection and loss of the cranioplasty reconstruction.¹³ Initially, the presence of shunts was thought to increase the rate of infectious complications, but both Posnick et al and subsequent researchers have reported favorable results with frontofacial advancement despite the presence of shunts and particularly after the routine use of DO.^{8,12}

Concepts

The monobloc and facial bipartition procedure using DO with an internal or external distraction system has emerged as a powerful and comprehensive tool in the treatment of syndromic synostosis. Despite diligent efforts by various teams to treat the initial cranial deformity of syndromic synostosis with anterior cranial or posterior cranial expansion procedures with and without DO, patients still present to craniofacial clinics in mixed dentition (ages 6–11) with profound facial retrusion. Patients with a retruded brow—defined as a brow to cornea relationship of –5 mm and a retruded inferior orbital rim of –5 mm—and a malocclusion overjet of –5 mm are considered optimal candidates for monobloc or facial bipartition advancement using DO (→Fig. 2). Patients who have had successful advancement of the brow with a brow cornea relationship of (+6–9 mm) are considered optimal candidates for subcranial facial advancement using the Le Fort III osteotomy with DO using either an internal distraction or a rigid external distraction device (RED). Last, patients who need facial concavity correction with or without hypertelorism/dystopia are ideal candidates for the facial bipartition. Although bipartition of the Le Fort III osteotomy has been well described and successfully used by some centers, our preference is to use the facial bipartition selectively due to the increased stability of titanium box plate fixation at the radix/glabella area that is well preserved with adequate bone stock when establishing osteosynthesis of the facial bipartition segments.

Fig. 2 Vertex frontal view of a child in mixed dentition with classic facial features seen in complex syndromic midface hypoplasia. The striking exophthalmos is readily demonstrated as well as the retrusion of the brow relative to the cornea position. The severe associated sleep apnea with midface retrusion was managed with a tracheostomy prior to frontofacial advancement.

Authors' Preferred Technique

At Johns Hopkins Hospital (Baltimore, MD), we prefer a zigzag incision in the parietal and temporal regions of the scalp and a straight incision at the vertex due to the favorable hair growth patterns of the scalp that will ultimately camouflage the incisions. A standard anterior coronal skin flap is developed, with care to preserve tissue for a centrally based pericranial flap. We prefer minimal posterior undermining to minimize hemorrhage. Subperiosteal pockets are created in the temporal regions bilaterally to accommodate the internal distraction systems. Our team prefers and routinely uses a single internal distraction device in each temporal region with the distraction rod placed parallel to the zygomatic arch. This limits the inferior decent and clockwise rotation of the face during subsequent distraction. Control of facial rotation usually in a clockwise orientation is accomplished using orthodontic elastics placed in a class III configuration. We typically use orthodontic bone anchors placed lateral to the permanent canine tooth buds or root. The orbits are dissected in a circular fashion posterior to the lacrimal crest in the proposed osteotomy line typically 2 cm posterior to the orbital rim to control medial and cephalic motion of the functional orbit. The anterior zygomatic arch osteotomy is easily performed using the coronal exposure.

Last, we prefer to maintain the temporal muscle attachments to the cranium and create a minimal keyhole window at the temporal muscle and lateral orbital wing to allow the blade of the reciprocating saw to traverse the inferior orbital fissure.

We prefer a small craniotomy to limit bone loss in the event of infection development. Our dedicated neurosurgery team members perform a bifrontal craniotomy. The anterior and middle cranial fossa are controlled and retracted with coated brain retractors and cottonoids to facilitate easy reciprocating saw placement. The nasal septum and pterygoid plates are exposed through a maxillary sulcus incision. We prefer osteotomy of the pterygoid plates transorally rather than transorbitally. Our team prefers sectioning of the nasal septum and mid palatal split during the bipartition after completion of the down fracture of the intact monobloc segment. The monobloc osteotomy is begun after a zygomatic arch osteotomy by a lateral orbital osteotomy beginning at the inferior orbital fissure. This osteotomy is directed superiorly with care to protect the middle cranial fossa as the saw transgresses the lateral wing of the sphenoid bone in to the anterior cranial fossa. Next, the orbital roof is sectioned lateral to medial with care to remain anterior to the crista galli medially and 2 cm posterior to the orbital rim in the midorbit. Next, the medial and inferior orbital wall osteotomy is completed with a 2-mm osteotomy. After pterygoid osteotomy, the monobloc segment is then down fractured using a pediatric Rowe disimpaction forceps. During down fracture, the perpendicular plate of the ethmoid is sectioned using a double-guarded osteotomy. The segment is then fully mobilized (► **Video 1**). If bipartition is then needed, a V excision of bone is removed centrally and the palate is split between the central incisors similar to a SARPE (supplemental

surgical-assisted rapid palate expansion) procedure. We prefer to bow the face as needed and keep the interdacryon distance less than 18 mm if possible. The face is then brought into its native position with or without bipartition and secured using the internal distractor system (► **Fig. 3**). A pericranial flap is then inset at the skull base and supplemented with human tissue sealant. The flap is used to supplement the skull base seal during distraction. This extra layer of soft tissue coupled with a 5- to 7-day latency period allows the skull base to seal with soft tissue while bone healing as distraction takes place in the following 2 to 3 weeks. We recommend operation at 8 to 10 years of age to avoid additional treatment that is required after the development of the frontal sinus (> 11 years old). If the surgery is performed in the later teen years, we prefer cranialization of the frontal sinus in addition to the other treatments noted above. The craniotomy bone segments are shaped and then plated. We use a latency period of 5 to 7 days and then begin advancement at 1 mm per day usually with a twice daily activation regimen. Average distraction distance in our experience is 15 mm with a range of 10 to 20 mm depending on the needs for advancement. We typically distract between ages 8 to 10 years and prefer to use the orbital volume and correction of exophthalmos as our clinical end points (► **Fig. 4**). Residual occlusal abnormalities are treated later after skeletal maturity at ages 16 to 18 years with a combination of orthodontic treatment and orthognathic surgery. Orthognathic surgery and SARPE at the completion of maxillary and mandible growth is almost always necessary

Fig. 3 Oblique intraoperative view after successful down fracture and mobilization of the monobloc segment. The internal distraction device is being secured anteriorly first. Prior to posterior fixation, the bone gap is obliterated by returning the facial skeletal to its premobilization position. The pericranial flap has been mobilized and inset covering the communication to ethmoid sinus region.

Fig. 4 Vertex frontal view of the same patient in ►Fig. 3 after monobloc distraction and advancement of 17 mm. Note the greatly improved relationship of the brow relative to the cornea, as well as the correction of the pretreatment trigonocephaly.

to completely treat the dentofacial imbalance seen in complex congenital craniosynostosis.

Video 1

A vertex frontal intraoperative view demonstrating complete mobilization of the monobloc segment and movement using minimal traction. Note the large retrofrontal dead space demonstrated with acute advancement.

Online content including video sequences viewable at: www.thieme-connect.com/products/ejournals/html/10.1055/s-0034-1390170.

Current Controversies and Future Directions

Timing and indications for surgery remain controversial. Despite recent evidence suggesting the safety and efficacy of monobloc and facial bipartition surgery using DO, many centers continue to use staged approaches with repeat frontal orbital advancement procedures until the brow is corrected, followed by subcranial Le Fort III advancement surgery in mixed dentition.^{8,14,15} Comparison of clinical outcomes, safety, and cost between one-step facial advancement (monobloc/bipartition) when compared with staged facial advancement (frontal orbital advancement, Le Fort III) remains understudied. Chan et al reported the volumetric changes in the soft tissue after monobloc distraction using three-dimension-

al photography. They noted a volumetric increase of 0.78 cm³ per 1 cm of bone advancement by 6 weeks that was stable at 1-year follow-up.¹⁶ The study of airway changes after monobloc/bipartition and its efficacy compared with Le Fort III remains undercharacterized.¹⁷ Timing of surgery to optimize facial advancement and correction of associated hypertelorism has undergone change in the last two decades. Although Ahmad et al have reported reasonable results using the monobloc frontofacial advancement with distraction in the very young (age < 30 months) with low complication rates, other long-term outcome studies demonstrate lower relapse rates and stable correction of advancements performed after age 6 or during mixed dentition.^{18–23} Virtual surgical planning has emerged as a useful tool in orthognathic surgery, which may streamline preoperative splint fabrication. Its routine use in planning for frontofacial surgery remains less characterized and perhaps underutilized. Gordon and colleagues have described a minimal access technique of monobloc osteotomy. Although attractive in concept, this approach has not been widely employed.²⁴ Although still considered controversial, the monobloc/bipartition procedures remain a powerful tool for complete correction of the residual dentofacial-deformity-associated syndromic craniosynostosis.

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