Surgical-Orthodontic Considerations in Subcranial and Frontofacial Distraction

Richard A. Hopper, MD, MS, Hitesh Kapadia, DDS, PhD, Srinivas M. Susarla, DMD, MD, MPH

BACKGROUND

Distraction osteogenesis, initially pioneered for management of limb-length discrepancies, has been a major innovation in craniomaxillofacial surgery. Although craniofacial distraction was initially described for management of mandibular hypoplasia, the technique has been effectively applied to myriad craniofacial differences, including midface deficiency, craniosynostosis, dentoalveolar hypoplasia, and management of hypertelorism. The shortcomings and morbidity of conventional osteotomies for management of complex subcranial and frontofacial deformities

KEYWORDS

- Le Fort III osteotomy
- Le Fort II osteotomy
- Monobloc osteotomy
- Subcranial distraction
- Counterclockwise craniofacial distraction
- Frontofacial advancement
- Syndromic craniosynostosis

KEY POINTS

- Distraction osteogenesis can be safely used for subcranial (Le Fort II, Le Fort III) or frontofacial (monobloc) advancements; proper orthodontic-surgical coordination is critical for a successful result.
- Midface distraction at the Le Fort III level is a valuable tool for managing the midface deficiency when the degree of hypoplasia is uniform across the midface (eg, Crouzon syndrome).
- Midface distraction osteogenesis at the Le Fort II level is useful for management of nasomaxillary hypoplasia (eg, Binder syndrome) or, when done in conjunction with zygomatic repositioning, for management of differential hypoplasia of the central midface relative to the orbitozygomatic complex (eg, Apert syndrome, achondroplasia).
- Frontofacial distraction can be used for management of midface hypoplasia in the context of anterior cranial vault constriction (eg, syndromic midface hypoplasia in the setting of intracranial hypertension). Monobloc frontofacial distraction advances the frontal bone and midface as a single unit.
- Synchronous counterclockwise rotation of the maxillomandibular complex is an emerging technique for management of complex hypoplasia involving the midface and mandible (eg, Treacher Collins syndrome).

ETHICS STATEMENT: All guidelines in the Declaration of Helsinki were followed in the course of this work. Consent was obtained for the use of clinical photographs.

ORCID iDs

a Craniofacial Center, Division of Plastic and Craniofacial Surgery, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105, USA; b Craniofacial Center, Division of Craniofacial Orthodontics, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105, USA; c Craniofacial Center, Divisions of Craniofacial and Plastic Surgery and Oral-Maxillofacial Surgery, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105, USA

* Corresponding author.

E-mail address: SRINIVAS.SUSARLA@SEATTLECHILDRENS.ORG
have been long recognized: length of operations, blood loss, hospital stay, necessity for bone grafting, inadequate soft tissue results, infection, and relapse.\textsuperscript{1–5} There is an increasing volume of data demonstrating the immediate efficacy and stability of subcranial and frontofacial distraction for management of complex morphologic deficiencies affecting the upper two-thirds of the face.\textsuperscript{6–14} Although perioperative morbidity rates for subcranial and frontofacial distraction remain significant due to the complexity of the regional anatomy and procedural details, in many centers, distraction procedures have replaced conventional osteotomies for management of upper and midface deficiency.\textsuperscript{7–17}

**GOALS OF SUBCRANIAL AND FRONTOFACIAL DISTRACTION**

The primary goal of subcranial or frontofacial distraction osteogenesis is to reposition the hypoplastic upper or midfacial skeleton to correct (or overcorrect) the developmental deficiency. Recognition of the specific deficiencies by anatomic region (anterior cranial vault, orbitozygomatic complex, nasal pyramid, and maxilla) is critical for treatment success. Osteotomy patterns are tailored to the morphologic characteristics that need to be addressed (Fig. 1).

In a patient with Crouzon syndrome, there is a proportionate midface discrepancy—the degree of hypoplasia in the orbitozygomatic region matches that of the nasomaxillary complex; the Crouzon phenotype is a morphologically normal midface in an abnormal position (Fig. 2). Correction at this level, via a Le Fort III distraction, is primarily related to improving the relationship between the infraorbital rim and anterior cornea (correcting exorbitism).

In contrast, the patient with Apert syndrome will have a midface deficiency that is disproportionate—the central midface hypoplasia is more profound than the zygomatic hypoplasia. The Apert phenotype is a morphologically abnormal midface in an abnormal position (Fig. 3). Correction in this context is differential between the lateral and central midface. Correction of exorbitism is accomplished with zygomatic repositioning; the central vertical and sagittal midface deficiency is addressed with Le Fort II distraction.

Patients with syndromic midface hypoplasia with concomitant intracranial hypertension related to cranial vault constriction present a more challenging clinical picture. Morphologic characteristics include frontal retrusion, exorbitism, and concomitant midface hypoplasia (Fig. 4). In this context, frontofacial advancement via monobloc distraction is an effective approach for addressing the anterior cranial vault and midface deficiencies simultaneously.

Complex clockwise rotational deformities involving the mid- and lower-facial skeleton are characterized by bimaxillary retrognathism with a class II skeletal pattern, long anterior face height, steep mandibular and palatal planes, and airway obstruction. For this phenotype, synchronous counterclockwise rotation of the maxillomandibular complex is an effective technique for correcting the maxillomandibular position, palatal and mandibular planes, anterior and posterior face heights, and improving airway dynamics (Fig. 5).

**TREATMENT PLANNING**

As with any surgical procedure, appropriate preoperative evaluation, diagnosis, and treatment planning based on sound principles are the keys to a successful result.\textsuperscript{18} A comprehensive dentofacial assessment, including both the hard and soft tissues, is paramount. Objective assessment of cranial morphology, globe position, malar projection, nasal morphology, maxillary and mandibular sagittal and vertical positioning, as well as a

![Fig. 1. Different patterns of osteotomy designs for correction of midface hypoplasia. The Le Fort III osteotomy (left) will result in separation of the entire midface from the cranial base, allowing advancement of the orbitozygomatic and nasomaxillary complexes as a single unit. The Le Fort II osteotomy (middle left) will allow for advancement of the nasomaxillary complex and may be combined with bilateral inverted-L mandibular osteotomies for synchronous counterclockwise craniofacial rotation. Segmentation of the Le Fort III osteotomy (Le Fort II with zygomatic repositioning) can allow for differential movement of the orbitozygomatic complex relative to the nasomaxillary segment (middle right). Severe midface deficiency in the setting of cranial vault constriction can be addressed with simultaneous fronto-orbital and Le Fort III advancement (monobloc advancement, right).](https://clinicalkey.com)
Fig. 2. Midface hypoplasia Crouzon syndrome. The midface deficiency is evenly distributed between the central and lateral midface. Clinically, this will be evident with poor malar support, exorbitism, and a class III profile. Correction of this specific midface morphology can be reliably accomplished with Le Fort III level distraction.

Fig. 3. Midface hypoplasia Apert syndrome. The midface deficiency is differentially distributed. The lateral orbitozygomatic regions are hypoplastic, which is clinically evident as poor malar support and exorbitism. The central midface exhibits vertical and sagittal hypoplasia that is more severe than the lateral midface hypoplasia. Differential correction is indicated with zygomatic repositioning and Le Fort II level distraction with a downward and forward vector.
Fig. 4. Frontofacial dysmorphology in a patient with Crouzon syndrome who presented with multiple cranial de- 
fects, obstructive sleep apnea (AHI 62.6), intracranial hypertension, frontal and supraorbital retraction, malignant 
proptosis, and midface hypoplasia. Management of the midface deformity in the context of anterior cranial vault 
constriction can be accomplished with monobloc frontofacial distraction.

Fig. 5. Complex craniomaxillofacial discrepancy in Treacher Collins syndrome. There is bilateral zygomatic hypo-
plasia, bimaxillary retrognathism, a long anterior face height, and steep palatal and mandibular plane angles. 
This constellation of skeletal differences is associated with clockwise rotation of the mid- and lower-face and 
is amenable to correction with counterclockwise craniofacial distraction (C3DO) using a Le Fort II osteotomy 
with concomitant bilateral mandibular osteotomies and subsequent synchronous maxillomandibular distraction.
detailed intraoral assessment, focusing on hygiene, presence/absence of teeth, state of the dentition, crowding, transverse relationships, and occlusal plane orientation is necessary. Ophthalmologic assessment is an important adjunct in patients with exorbitism and those at risk for intracranial hypertension. Neurosurgical assessment is critical for patients undergoing frontofacial procedures.

Three-dimensional computed tomographic scans are critical for both diagnosis and treatment. Three-dimensional visualization of the specific anatomy is necessary to identify the specific morphologic deficiencies, plan appropriate distraction vectors, safely execute osteotomies and device placement, and overcome anatomic challenges such as persistent bony defects, shunts, sensory nerves, and developing dentition. Dental models (cast or digital) complement imaging modalities in this regard and remain essential for evaluating potential occlusal interferences and for splint fabrication. Virtual surgical planning is used in many cases and affords an opportunity to visualize, in all three dimensions, the proposed skeletal movements and their impact on cephalometric parameters (Fig. 6).

The goals of treatment should be clearly established early in planning, with a focus on treatment plans tailored to the specific characteristics of the midface deficiency. Patients undergoing distraction procedures at the Le Fort II or Le Fort III levels are frequently in the mixed dentition. In this population, maxillary expansion and leveling of the occlusal plane may be required before surgical intervention. Pediatric dental assessment is necessary to manage active decay and reinforce the importance of excellent oral hygiene.

Patients undergoing frontofacial distraction procedures are frequently medically complex and require a collaborative, team-centered approach, including a craniofacial surgeon, neurosurgeon, ophthalmologist, orthodontist, and pediatric...
dentist. Although there remains controversy regarding early monobloc procedures (<2 years of age), older patients undergoing frontofacial advancements have dental needs similar to patients in the mixed dentition undergoing subcranial midface advancements. Preoperative planning for frontofacial distraction should take into account the specific cranial dysmorphology, including the presence of cranial defects and shunts. Coordinated treatment planning frequently allows for custom alloplastic cranioplasty to address cranial defects at the time of frontofacial osteotomies.

DISTRACTION DEVICES

The distraction apparatus (Fig. 7) can be internal (directly affixed to bone) or external (halo device). Internal devices can be used to span the osteotomy gap without a splint, with fixation placed above and below the osteotomy or with a splint, with the inferior plate limb affixed to the splint. Similarly, external devices can be subdivided into those that use tooth-borne splints versus bone-borne fixation to connect to the external framework. Practitioner preferences vary in the choice of device, and each type has specific advantages and disadvantages. Purported advantages of external devices are related to the relative ease of placement, with less extensive subperiosteal dissection necessary, as well as differential forces (can “push” and “pull”), and, perhaps most importantly, 3-dimensional control of movement vectors. These advantages must be weighed against the patient’s tolerance for a cumbersome device that may become loose or dislodged and the risks of pin site infections. Internal devices are generally well hidden and placed closer to the primary osteotomy sites, theoretically reducing torque. However, placement is more challenging and requires extensive periosteal stripping, more significant staged procedures for device placement and removal, and distraction may be limited both in terms of magnitude and direction.

SPLINT FABRICATION AND APPLICATION

With tooth-borne external midface distraction, a prefabricated occlusal splint is necessary (Fig. 8). The splint is fabricated by using an orthodontic headgear bow as a framework within an acrylic occlusal registration. The internal wire is used to conform to the dentoalveolar morphology of the maxilla; the external wire is bent at 90-degree angle to accommodate outrigger hooks. The height of the outrigger hooks depends on both the patient’s age and the planned distraction vector. The transverse dimension between the external wire limbs is slightly wider than the alar base width. The sagittal position of the external framework relative to the acrylic plate is based on the patient’s lip thickness. The internal wire is embedded within a self-curing dental acrylic resin. The completed splint is then polished, both for patient comfort and to remove surface irregularities that are prone to plaque

Fig. 7. Virtual surgical planning is an important adjunct for management of complex maxillomandibular deformities. Precise 3-dimensional visualization of the pertinent anatomy allows the surgeon to plan osteotomies and, when needed, fabricate cutting guides (left: cutting template for inverted-L osteotomies for counterclockwise craniofacial distraction osteogenesis (C3DO), with guide holes for transpharyngeal pin placement). Projected movements along the planned vector can be demonstrated, with associated measurement of cephalometric changes (middle and right: virtual surgical plan depicting pre- and post-distraction positions for C3DO). (From Hopper RA, Kapadia H, Susarla S, et al. Counterclockwise craniofacial distraction osteogenesis for tracheostomy-dependent children with Treacher Collins syndrome. Plast Reconstr Surg. 2018;142(2):447–57; with permission.)
accumulation. Intraoperatively, the splint is cemented to the maxillary dentition using a light-cured glass ionomer cement. In patients with oligodontia or where additional stability is otherwise needed, circum-zygomatic and circum-piriform wires (26-gauge) can be used to reinforce the splint and prevent dislodgement during activation. Once the splint is affixed to the teeth, it is connected to the rigid external device frame via 26-gauge stainless steel wires to the transverse bar. The vertical rod attachment can then be adjusted to achieve the appropriate height for the planned distraction vector.13

With Le Fort II and Le Fort III distraction procedures, the center of resistance for midface advancement is located slightly above the midpoint between the maxillary occlusal plane and nasion.19,20 Application of the distraction force at this level will reliably advance the upper midface without affecting the occlusal plane. Pull of the pterygomasseteric sling tends to inferiorly reposition the posterior aspect of the Le Fort II or Le Fort III segment—if this is not accounted for, an anterior open bite will result from counterclockwise rotation of the midface (ie, if the distraction force is too low). Conversely, a distraction force applied too high will result in clockwise rotation of the midface complex, resulting in overadvancement of the orbitozygomatic regions and possible enophthalmos.

For frontofacial advancements, the center of rotation will be located higher in the midface, resulting in a longer lever arm between the occlusal plane and center of rotation. Rotational movements are less frequently problematic in this group, due to the presence of additional stabilization posts within the lateral midface, but the vector of distraction needs to be continually assessed with serial lateral cephalograms, to ensure a vector parallel to the Frankfort horizontal. In patients undergoing synchronous counterclockwise craniofacial distraction procedures, a Le Fort II osteotomy is performed in conjunction with bilateral mandibular inverted-L osteotomies.14 A custom acrylic occlusal splint, similar to that used for external midface distraction, but

Fig. 8. Splint fabrication for external midface distraction. An orthodontic headgear bow is used for the framework. The internal wire is adapted to the maxillary arch form; the external wire is bent at a 90-degree angle to accommodate the outrigger hooks (top left). Proper orientation of the external framework requires meticulous attention. The width between the vertical limbs should be slightly larger than the alar base width (top right). The sagittal position of the bend should account for the upper lip thickness (bottom left). The final acrylated splint (bottom right).
adapted to include the mandibular dentition, is affixed to the teeth, and the patient is placed into maxillomandibular fixation with transpalatal and circummandibular wires (26-gauge). The splint construct is then wired to the activation arms with a 45-degree upward vector relative to the Frankfort horizontal.

PERIOPERATIVE ORTHODONTIC TREATMENT

During distraction: once the planned osteotomies are completed and the prefabricated occlusal splint is affixed to the midface, the patient will have a latency period of approximately 5 days. The device is then activated at a rate of 0.5 mm twice a day. Distraction is complete when the predetermined endpoints have been achieved. With Le Fort III distraction, this is predicated on the lateral orbitozygomatic contour and correction of exorbitism (Fig. 9). With Le Fort II distraction, this is based on the nasal length and vertical height of the central midface (Fig. 10). For frontofacial advancement, the distraction endpoints are determined by the position of the external orbit relative to the anterior cornea, frontal bone projection, and maxillary incisor position (Fig. 11). In growing patients, a goal overjet of 5 to 6 mm following subcranial or frontofacial advancement may also be considered as a treatment goal. With counterclockwise craniofacial rotation, active distraction is continued until the palatal plane is normalized (sella-nasion to palatal plane angle of 7°, Fig. 12). Following distraction progress with serial lateral cephalograms is critical during the active phase, as incorrect or inadequate vectors and problems with splint adaptation can be addressed. As discussed earlier, incorrect vertical positioning of the distraction force in the Le Fort II or III segment can result in undesirable counterclockwise or clockwise occlusal plane rotations. Placement of temporary anchorage devices in the midline mandible and elastic traction during activation can be helpful to address these specific vector issues. Orthodontic bone anchors are similarly useful adjuncts for vector control in Le Fort II distraction procedures as well as counterclockwise maxillomandibular rotations.

POSTDISTRACTION ORTHODONTIC TREATMENT

Following completion of active distraction, patients will enter a consolidation phase (average 8 weeks). Once formation of bony generate is confirmed radiographically (most evident at the ptterygomaxillary junction), the devices are removed. Postsurgical orthodontic treatment is

Fig. 9. Congruent midface hypoplasia seen in Crouzon syndrome can be managed with Le Fort III distraction.
then undertaken. In many instances, postsurgical orthodontic care for the patient following midface distraction is similar to nonsurgical orthodontic care.

For patients undergoing midface distraction procedures in the mixed dentition, it is paramount to monitor growth of the midface relative to the mandible, as well as to follow dental eruption. There is evidence that subcranial midface osteotomies in younger patients results in disruptions in maxillary molar development and eruption. Families should be counseled regarding this possibility preoperatively, and molar development should be carefully monitored in mixed dentition patients.

**Fig. 10.** Differential midface hypoplasia seen in Apert syndrome is appropriately addressed with Le Fort II distraction and zygomatic repositioning.

**Fig. 11.** This patient with Crouzon syndrome presented with large calvarial defects, intracranial hypertension, malignant proptosis, supraorbital retrusion, midface hypoplasia, and obstructive sleep apnea (AHI 67). The anterior cranial vault constriction and midface hypoplasia were addressed simultaneously with a monobloc frontofacial distraction. Postoperatively, the facial proportions were improved, with adequate globe protection. Postoperative polysomnography demonstrated substantial improvement in sleep-disordered breathing (AHI 3.7).
Fig. 12. The clockwise rotation deformity seen in Treacher Collins syndrome is managed with synchronous counter-clockwise rotation distraction of the maxillomandibular complex, accomplished via Le Fort II and bilateral inverted-L mandibular osteotomies. In this patient, construction of the zygomas with autologous cranial bone graft was completed at the time of device removal. (From Hopper RA, Kapadia H, Susarla S, et al. Counterclockwise craniofacial distraction osteogenesis for tracheostomy-dependent children with Treacher Collins syndrome. Plast Reconstr Surg. 2018;142(2):447–57; with permission.)

Fig. 13. Preoperative occlusion in a patient with achondroplasia (top). Post-distraction occlusion following Le Fort II osteotomy with zygomatic repositioning (middle); the midface position is overcorrected relative to the mandible, with positive overjet at the incisors. At skeletal maturity, the patient underwent Le Fort I and bilateral mandibular sagittal split osteotomies for correction of the residual dentoskeletal discrepancy (bottom). (From Hopper RA, Kapadia H, Susarla SM. Le Fort II Distraction With Zygomatic Repositioning: A Technique for Differential Correction of Midface Hypoplasia. J Oral Maxillofac Surg. 2018 Sep;76(9):2002.e1–14; with permission.)
undergoing subcranial distraction procedures. The available evidence demonstrates that although the distracted midface does not significantly relapse, growth is unpredictable and many patients undergoing subcranial midface advancement in the mixed dentition may require definitive orthognathic surgery at skeletal maturity. In this context, the goals of subcranial advancement established preoperatively are paramount. Anticipation of the need for occlusal and lower midface correction at skeletal maturity is necessary for appropriate treatment planning and patient/family counseling (Fig. 13).

**SUMMARY**

When properly planned and executed, subcranial advancement at the Le Fort II-III levels (including segmental movements and synchronous maxillo-mandibular counterclockwise rotations) and frontofacial distraction can be used to correct a wide array of craniomaxillofacial deformities. Focused attention on the specific morphologic characteristics of the patient, as well as an assessment of growth, and specific goals for distraction endpoints are necessary to ensure an optimal outcome.

**DISCLOSURE**

Dr R.A. Hopper shares patent royalties with KLS Martin. The other authors have nothing to disclose.

**REFERENCES**

21. Caterson EJ, Shetye PR, Grayson BH, et al. Surgical management of patients with a history of early Le Fort III advancement after they have attained


