Distraction Osteogenesis of the Craniofacial Skeleton

Learning Objectives: After studying this article, the participant should be able to: 1. Review the biomechanical principles and pertinent cellular and molecular biology of distraction osteogenesis of the craniofacial skeleton. 2. Describe the clinical indications and applications of distraction osteogenesis of the craniofacial skeleton. 3. Describe maxillary, mandibular, midface, and calvarial procedures in distraction osteogenesis. 4. Discuss the clinical outcomes and complications of distraction osteogenesis of the craniofacial skeleton.

The year 2002 marked the end of the first decade in clinical distraction osteogenesis of the craniofacial skeleton. In this short period, its application has increased exponentially. More than 3000 cases have been performed according to a recent survey, and more than 700 articles have been written on this subject in the MEDLINE database since 1996. It is a powerful surgical tool and enables surgeons to achieve results not previously attainable. Despite all this, distraction osteogenesis is practiced by only a small number of plastic surgeons. This article reviews the biomechanical principles; the pertinent cellular and molecular biology; and the clinical indications, applications, controversies, and complications of distraction osteogenesis of the craniofacial skeleton. (Plast. Reconstr. Surg. 114: 1e, 2004.)

Plastic surgeons alter the product of morphogenesis. The natural attainment of body form is a multifactorial, polygenic process. A key ingredient of this complex process is mechanical force. Force is completely ubiquitous; it can originate from local growth, the earth’s gravitation, muscular contraction, and surface tension, to name just a few sources. Cellular response to force is therefore a very ancient and critical part of the biotic process. Distraction osteogenesis, like soft-tissue expansion, taps into this ancient, universal property; grow if stretched. Initially used in orthopedic surgery by Codivilla in 1905, it was systematically developed and refined by Ilizarov. Ilizarov’s meticulous work definitively established the fact that bone will form in response to tension. This apparently contradicted Wolff’s law regarding bone remodeling, which associates bone formation with compression and bone resorption with tension. In 1992, McCarthy et al. reported in the English literature the first application of distraction osteogenesis to lengthen the human mandible. This method is now used extensively at every level of the craniofacial skeleton. There are three main phases to distraction osteogenesis: latency, activation, and consolidation. Latency is that period immediately following the osteotomy and application of distractor; it ranges from 1 to 7 days. After the latency phase is the activation phase. During this phase, the distraction device is activated by turning some type of axial screw, usually at 1 mm/day in four equal increments of 0.25 mm each. Once activation is complete, the third and final phase is the consolidation phase. Typically, the consolidation phase is twice as long as the time required for activation. The above three phases constitute the Ilizarov protocol designed for lengthening the long, endochondral bones of the lower extremity. Whether this is the optimal protocol for the...
craniofacial skeleton as well is not completely clear. Today, many different devices are being used clinically, with many different distraction protocols.

This review article describes the biomechanical, cellular, and biomolecular events that occur during distraction osteogenesis. The indications, clinical applications, controversies, outcomes, and potential complications of distraction osteogenesis in the craniofacial skeleton are discussed at each level, from the mandible to the forehead.

**Biomechanics of Distraction Osteogenesis**

Distraction osteogenesis can be considered as a very special, altered form of fracture healing. It represents an effective and long-term augmentation of the human morphology by using mechanical force to induce and direct bone and soft-tissue formation. Unlike expansion of the soft tissue by tissue expanders, the osseous tissue, once produced, does not contract over time after the removal of the expander device. This is because bone is rigid, and it responds to the mechanical demands placed on it. Bone is the only living tissue that can effectively withstand both tensile and compressive loads, with a tensile strength of 12,000 psi and a compressive strength of 15,000 psi. To achieve targeted bone growth, a rigid stretching device delivers tensile force to the developing callus at the site of the bone cut (periosteum- and marrow-sparing corticotomies in the original Ilizarov protocol; complete osteotomies in most craniofacial centers now), a process known as callotasis. In response to this force, the callus elongates. The amount of elongation as a fraction of the original length is known as tensile strain. In distraction osteogenesis, the typical protocol is 0.25 mm at four times per day, or 1.0 mm/day. In most cases, the osteotomy creates an initial defect of approximately 1.0 mm. Thus, the strain is 100 percent during day 1 of activation and drops to 50 percent for day 2 and 33 percent for day 3. By day 10, the theoretical strain induced by 1.0 mm of elongation in a 10-mm callus is decreased to 10 percent. This reduction in strain as distraction progresses is inevitable given a constant distraction rate. Bone tissue as a material can tolerate only 1 to 2 percent of tensile strain, a parameter known as ultimate tensile strain. Thus, no bone tissue can exist if the load environment produces more than 1 to 2 percent tensile strain. In normal fracture healing, ossification is seen when the interfragmentary strain is below the ultimate strain. Not surprisingly, by week 4 of distraction, with the tensile strain approaching or below the ultimate strain level, bone formation starts. On microscopy, the classic description is that there are five histologic zones: one central zone of fibrosis bordered on either side by the two transitional zones, which are themselves bordered by the remodeling zones. The central zone is better described as the central zone of mesenchymal proliferation. The current concept also has five zones but adds four transitional areas between the zones. It assigns two paracentral zones, one on each side of the central zone, joined by the transitional area of vasculogenesis. Peripherally, the paracentral zones border the proximal-distal zone, separated from them by another transition area: the area of mineralizing fronts where the highest ratio of cell division was observed throughout the activation phase (Fig. 1). Apoptosis is present in the paracentral zones. Woven bone is the first type of bone to appear. It is not clear at present which of the five zones or four transitional areas are actually structurally the most likely to undergo tensile strain in response to the tensile stress imparted by the distractor. This will depend on the elastic modulus of the various zones and transitional areas, which has not yet been measured or reported. Very limited direct biomechanical characterization is available even from animal experiments. Mofid et al. reported recently that, using a standard distraction protocol in New Zealand White rabbits, the mandibular regenerate after 8 weeks of consolidation had a bending stiffness of 200 N/mm, which was approximately 50 percent of the intact mandible. The test was three-point bending and the load rate was 0.1 mm/second. Robinson et al. reported that the average torque required to distract human mandibles at 0.5 mm/day was 4.2 ± 1.6 Ncm, which could be converted to an estimated linear tensile force of 35.6 N. The distractor for that study had a failure, or yield, force of 235.8 N.

All distractors have the following three components: an intraosseous component, to transmit the displacement to bone/callus; an anchorage component, to push or pull against; and some type of axial screw which, when turned, generates the primary displacement. The system is configured in such a way that it is only as strong as the weakest link: any single component failure will result in the failure of
the distraction process. There are two major types of distractors: internal and external. An inherent difference in force delivery between the internal and external distractors is the distance from the callus surface to the activating axial screw. The closer this activating axial screw is to the central (neutral) axis of the bone/callus, the more effective the stretching. This is because whenever the force vector is not directly coaxial, or in line, with the central axis there will be a turning moment. The external devices rely on intraosseous pins to transmit the force. The longer the distance from the axial screw of the distractor to the callus, the less effective the distraction. Internal distractors thus enjoy reduced perpendicular distance from the callus to the activating axial screw. However, this advantage comes at increased difficulty at the time of device removal that can add significantly to the overall morbidity. The external distractors allow for easier adjustment of the direction of the distraction. Of the two principal means of delivering the tensile force in achieving distraction—push or pull—internal distractors are limited to only pushing apart the bone segments. The external distractor with half-halo–type anchorage achieves distraction by pulling. The actual magnitude of force required to elongate the callus is unknown and is likely to vary from site to site and from individual to individual. Polley and Figueroa\textsuperscript{27} reported using a 10-N force by means of heavy elastics to gradually distract the maxilla. Using torque wrench measurements, turning moments from 14 to 18 Ncm were delivered to the activating screw of the distractor in one center.\textsuperscript{28} Because it is difficult to predict precisely what the total resistance is, the planned distraction trajectory may differ from the actual trajectory obtained during distraction. This has been confirmed in simulated internal mandibular distraction with and without soft tissues such as masseter, temporalis, and the suprahyoid muscles.\textsuperscript{29}

**CELLULAR AND MOLECULAR BIOLOGY OF DISTRACTION OSTEOGENESIS**

Bone is a highly specialized connective tissue. It differs from all other nonmineralized, connective tissue in that it is hard (Vickers hardness of 30 kg/mm\textsuperscript{2} for young human bones and 38 kg/mm\textsuperscript{2} for mature human bones when measured wet).\textsuperscript{20} This hardness is attributable to the mineralization of the fibrillar extracellular matrices. There are many reports based on animal models of the cellular and molecular events that occur during the distraction of a healing bone callus.\textsuperscript{30-32} For obvious reasons, there are no comprehensive comparable human data. Immediately after the osteotomy, the formation of hematoma and inflammatory infiltrates is exactly the same as in any standard osteotomy or low-energy fracture. There is a decrease in oxygen tension and local pH and a reversal of the electric field potential, with the fractured bone ends attain-
ing a more positive charge, approximately 1 mV, whereas the fracture hematoma demonstrates a sharp increase in electronegativity to −6 mV. Toward the end of the latency period, by day 5 after osteotomy, the site is filled with granulation tissue packed with round, cuboidal mesenchymal cells and nascent capillaries. The orientations of the cells, capillaries, and matrices at this time are isotropic (random orientation). There is a rapid decrease in the level of mRNA of bone-specific proteins and extracellular matrices such as osteocalcin and type I collagen, respectively. This is not surprising, because the tissue being assayed by Northern blotting is not really “bone” but rather granulation tissue. Of note was the rapid increase of the transforming growth factor-β1 mRNA level to 2.5-fold of the normal bone by day 3 of the latency period. As distraction progresses, the differences between fracture healing and distraction become more obvious. Mechanical strain is a critical factor. Ilizarov asserted that the alteration of the mechanical environment within the distraction gap led to “stimulation of both proliferative and biosynthetic cellular functions.” More recent research has focused on precisely what these events are. There is increased cell division with the proliferation index, as measured by proliferating cell nuclear antigen immunohistochemistry, being highest at the mineralization fronts. The cell types begin to appear more fusiform in shape. The more central area, however, retains the more cuboidal, primitive mesenchymal appearance. As distraction progresses, the most striking feature is the tremendous degree of anisotropy at the junction of the osteotomy site and mineralization front; the orientation is now parallel to the line of tension. In the region immediately adjacent to the central zone, there are many apoptotic figures. Here, in the paracentral zones, the cellularity decreases and ground substances accumulate. Between the central zone and the paracentral zone is the transitional area, where much vasculogenesis occurs. The mRNA profile at this early to mid-activation phase is a rapid and sustained increase of the transforming growth factor-β1 mRNA level, with a slow but steady increase in the mRNA of type I collagen. Lagging behind is the bone-specific osteocalcin mRNA. At the end of the activation period, there is clear anisotropy, with the bone trabeculae in perfect alignment with the direction of the distraction.

Not all distractions require an osteotomy first. In the distraction of the immature cranial sutures, sutures may serve as the “callus.” Distraction of the cranial sutures has been shown to reduce the level of some key signaling peptides that are very important during embryogenesis and development. The three proteins of the hedgehog family, Shh (Sonic hedgehog), Ihh (Indian hedgehog), and Dhh (Desert hedgehog), are such examples. These signaling proteins bind to a class of transmembrane receptors, patched-1, and alter the activity of protein kinase A, which sets off a cascade of intracellular molecular events. Tensile stress produced by the distraction appears to reduce the level of the hedgehog proteins and their receptor, patched-1, in all regions of the periosteal tissue, especially the periosteum. Of particular interest is the observation that such reduction renders a synostosing suture more like a normal one. However, how the applied tensile force actually causes the observed alteration in cellular and molecular activities is obscure.

To understand how distraction forces regulate the creation and differentiation of new bone, it is critical for an experimental model to distinguish new bone formation attributable to distraction from secondary bone formation resulting from unaided fracture healing. A rat model of distraction osteogenesis that does differentiate between those two fundamental mechanisms of new bone formation has been established. The model documented a critical defect size in the rat mandible for discrimination of new bone formation attributable to distraction from unaided fracture healing at the site of the osteotomy. The distraction protocol was then performed with a defect larger than that critical size. Determination of a critical-size defect allowed clear identification of new bone formation attributable to distraction processes alone. The model also included analysis of a subcritical-size defect that uniformly heals without difficulty, allowing a comparison of normal fracture healing at the same anatomic site, in the same animal model. This model has permitted isolation of the variables necessary for identification of the significant differences between distraction osteogenesis and osteotomy alone and to more accurately attribute the changes to the specific stimuli produced by the process of distraction. It has been hypothesized that mechanical forces created during distraction osteogenesis are responsible
for the osteogenic responses, and that these changes arise through integrin-dependent mechanotransduction. Mechanotransduction is the process by which mechanical forces are converted to cellular signals. These forces exert their effects by means of many pathways. One such pathway is integrin-dependent signal transduction. The integrin-mediated signal transduction cascade has been proposed as a primary pathway by which mechanotransduction occurs. Many in vitro experimental studies focusing on cells subjected to mechanical loading have investigated signal transduction during bone growth and adaptation. Within the integrin-mediated signal transduction cascade, focal adhesion kinase, c-Src (pp60c-src) and mitogen-activated protein kinase are believed to be key molecular mediators. Although these mediators have been studied extensively in vitro, insufficient research had been completed to date evaluating their role and the molecular mechanisms of integrin-mediated mechanotransduction in vivo. Using the rat model of distraction osteogenesis, the expression of focal adhesion kinase, c-Src, and mitogen-activated protein kinase in critical-size and subcritical-size defects was examined. Findings demonstrated immunolocalization of all three molecular mediators in mandibles undergoing distraction osteogenesis but not in the critical-size or subcritical-size defects, despite varying degrees of bone formation in the latter two groups. Furthermore, the mRNA in situ hybridization patterns of bone-specific proteins, such as bone sialoprotein, were found to mirror focal adhesion kinase immunolocalization patterns in mandibles undergoing distraction osteogenesis, demonstrating an association of focal adhesion kinase expression with the osteogenic process specific to distraction osteogenesis. These findings support the belief that bone formation in distraction osteogenesis is regulated by mechanical forces and these forces act at the cellular and molecular level by means of integrin-mediated signal transduction pathways. Investigation of the cellular and molecular biology of distraction osteogenesis is still in its infancy, but new discoveries show promise to significantly increase our understanding of the intricacies of this fascinating process.

**Mandibular Distraction Osteogenesis**

The mandible was the initial site of application of distraction osteogenesis in the face. The mandible’s structure is similar to the tubular structure of the long bones of the skeleton. Principles learned by orthopedic surgeons over the previous 80 years from distraction of the long bones of the lower extremity were rapidly adapted to this new location. Distraction osteogenesis has provided a powerful tool for treatment of many mandibular deformities that previously could not be successfully treated by the “conventional” methods of orthognathic surgery, free tissue transfer, or nonvascularized bone grafts. In addition, the use of distraction osteogenesis has been extended into applications that have been previously treated by one of these other conventional approaches to optimize outcome.

The two major strengths of distraction osteogenesis in mandibular reconstruction are the ability to provide strong bone with an excellent blood supply and the ability to provide effective expansion of the soft-tissue envelope. The importance of expansion of the soft-tissue envelope cannot be stated strongly enough, because the gradual expansion of the soft tissue over an extended period of time is much more effective than that which can be obtained in the short time window of a single operation. These two factors together allow a much greater skeletal advancement than can be obtained using conventional techniques and allow the creation of a construct that is stable in the advanced and reconstructed position. This is in contrast to the considerable relapse that can often be seen after conventional procedures in larger-scale advancements.

One of the primary planning considerations in mandibular distraction osteogenesis is the use of either an external distraction framework or an internal device. Critical to this decision is an evaluation of the goals of the distraction process. The external devices have the powerful advantages of allowing bone distraction in three planes and allowing the surgeon to alter the direction, or vector, of the distraction process while the distraction is proceeding. Pensler et al. first reported this principle of “molding the regenerate.” The “molding” takes advantage of the ability to manipulate the semisolid state of the nonmineralized, and hence nonrigid, bone in the distraction gap. This allows for “fine-tuning” of the distraction process while the distraction is proceeding, and thus permits dental relationships to be adjusted before the patient enters the consolidation phase of bone healing.
framework also allows greater amounts of ultimate expansion length. Expansions of 40 mm or greater have been reliably obtained. The disadvantages of an external frame distractor are the creation of a facial scar and the increased distance from the body of the distractor to the bone surface, leading to a longer "moment arm" at the pin-bone interface and an increased possibility of pin loosening. In addition, there is the need for "pin care" by the patient at the percutaneous pin sites.50

The child in Figure 2 has several congenital problems, including VATER association (vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, and radial and renal anomalies) and a severely hypoplastic mandible. She required tracheostomy during infancy, and she remained tracheostomy-dependent at 9 years of age. An external multiaxial distraction technique was selected because of the need to provide for stable fixation in the severely hypoplastic mandible and limited jaw opening and small mouth that precluded consideration of an internal device. After distraction of approximately 38 mm, the child's tracheostomy was successfully decannulated. Her preoperative and postoperative photographs are shown. Her advancement has been stable, without relapse, for the past 5 years.

An internal distraction framework may be used if the goal of distraction is to provide a moderate gain in length in only one direction. The osteotomy is made transorally, and the distraction frameworks are placed using a transbuccal percutaneous technique. The goal of distraction with internal devices is generally more modest, in the range of 25 mm or less. This is a consequence of the constraints placed on the physical size of the device and the ability to fit it within the mouth. In addition, the direction of the distraction cannot be altered after the device is placed. This inability to alter the vector of distraction dictates that unless the distraction frameworks are placed with a perfect vector of distraction initially, larger-scale distractions will lead to larger and larger discrepancies as the axial length of the distraction vector proceeds. The child in Figure 3 has Moebius syndrome, cleft palate, and a severely hypoplastic mandible. She had a tracheostomy placed shortly after birth and remained tracheostomy-dependent at 3 years of age. Preoperative planning revealed the need for single-vector sagittal advancement of the mandible. Bilateral mandibular osteotomy and placement of internal framework bone distraction devices were performed using a percutaneous technique. The child underwent bilateral distraction of 25 mm on each side, with a linear advancement of the mandible. Note that the advancement has led to an improvement in facial and dental relationships and minimal scarring. The technical considerations in this type of procedure are significant. After osteotomy and "greenstick" fracture of the mandibular bone, the surgeon is essentially trying to control three separate components (fixator, proximal, and distal bone segments) simultaneously through the mouth and percutaneously through the cheek. This is compounded by the fact that in these congenital cases, the mandibular segments are often small, and there is the need to obtain favorable alignment to allow the establishment of an appropriate vector of distraction. When the operation is
completed with appropriate rigid bone fixation, the results of distraction osteogenesis can be quite dramatic.

In addition to the decision regarding internal or external distraction frameworks, other crucial parameters are the selection of osteotomy site and pin site and the consideration of the location of the inferior alveolar nerve and the location of the teeth. In planning the osteotomy site, distraction osteogenesis of the mandible follows the principles of long-bone distraction. The cross-sectional area of the bone formed in the distraction process (and to a great extent the strength of the bone generated) is directly related to the cross-sectional area at the site of the osteotomy of the mandible. The 4-year-old girl in Figure 4 with a hypoplastic mandible that was originally thought to be attributable to “congenital” facial microsomia had been followed since the age of 1 year. Her mother had undergone a series of reconstructive procedures for a hypoplastic mandible, including costochondral grafting for mandibular hypoplasia. Figure 4, above, also shows the congenital ear deformity. She also had a limitation in mandibular opening. Surprisingly, computed tomography scanning of the temporomandibular joint showed sagittal fractures through the condylar head on the left and right, with a hyperplastic response on the right leading to bony ankylosis. The distraction was performed with an osteotomy through the angle of the mandible and an external device, with a gain of approximately 40 mm. The postoperative computed tomography scan (Fig. 4, below) shows the dramatic difference between bone formation on the hyperplastic and normal sides and illustrates the principle of bone formation being linked to the cross-sectional area of the osteotomy. The child’s facial scars have faded significantly.

These distraction cases highlight the application of bone distraction in the mandible with a goal of relieving airway obstruction that had persisted into childhood. This is a more common problem than had been previously recognized. The advent of sophisticated polysomnographic instruments has made this problem more widely appreciated. It is unlikely that these cases could have been treated effectively without the use of mandibular distraction osteogenesis, considering the limited magnitude.
of the scalar movements that can be obtained by conventional procedures. Nonetheless, these cases represent only one indication for distraction osteogenesis of the mandible. Two special situations merit specific discussion: hemifacial microsomia (oculoauriculovertebral spectrum) and mandibular distraction for upper airway obstruction in Pierre Robin syndrome.

Hemifacial microsomia is one of the most common congenital anomalies, with an incidence of approximately one in 5000 births. Mandibular distraction lengthening has been used in many cases of hemifacial microsomia to provide jaw lengthening and even to create a “pseudotemporomandibular joint.” The results in treatment of this disorder have been rewarding. The treatment is limited to patients with appropriate bone stock distal to the dentition for creation of an osteotomy (Pruzansky grades I and II). The primary advantages are the creation of strong, well-vascularized bone and the expansion of the soft-tissue envelope. This bone is much better for providing satisfactory and durable expansion without resorption and loss of projection, as is often seen when costochondral grafts are used to reconstruct the mandibular ramus. It is essential that a glenoid fossa or shelf be present to provide a buttress for the distraction process; otherwise, one must be created. It is important to also appreciate that the distraction process will not provide complete correction of the deformity of hemifacial microsomia, and appropriate planning for maxillary osteotomy must be considered in the overall treatment-planning process. Furthermore, milder cases of skeletal discrepancy in hemifacial microsomia can be appropriately treated successfully with conventional orthognathic surgery.
Pierre Robin syndrome is the combination of a hypoplastic mandible, cleft palate, and glossoptosis. Glossoptosis is literally the tongue falling backward into the pharyngeal space, and many of these infants develop ventilatory obstruction. Experienced authors from several centers have reported the management of upper airway obstruction using distraction osteogenesis. Perhaps no other application of mandibular distraction osteogenesis has been as controversial. The proponents of distraction osteogenesis in this application state that it provides effective management of the relatively acute airway obstruction in these infants.65 The opponents of distraction osteogenesis in Pierre Robin syndrome raise several arguments. First, there is the concept of “catch-up” growth of the mandible. This principle suggests that the mandible in Pierre Robin syndrome grows rapidly during the first year of life and minimizes the magnitude of the skeletal discrepancy, and as such would obviate the need for distraction osteogenesis (it should be noted that this principle of catch-up growth itself is not clearly established and is somewhat controversial). Second, they cite the risk of injury to the dental follicles within the mandible by the osteotomy and distraction process in the infant. Finally, the majority of Pierre Robin syndrome patients can be effectively treated by conservative measures such as proper positioning or other limited techniques such as lip-tongue adhesion and glossoxy. These approaches will fail for only a few patients, and whether mandibular osteotomy and distraction osteogenesis are preferable to tracheostomy is subject to debate. Ortiz Monasterio and colleagues have provided additional insight into this issue in their recent report that distraction osteogenesis of the mandible not only corrects the airway obstruction of Pierre Robin syndrome but also has a beneficial effect on swallowing and reflux.66 If these findings are validated in other centers, the debate will not be between mandibular distraction osteogenesis versus tracheostomy, but will be between mandibular distraction osteogenesis versus tracheostomy and glossoptomy and Nissen fundoplication.

Mandibular distraction can also be used in treating segmental mandibular defects from oncologic resections, or segmental defects from trauma or infections.67 This type of distraction is known as transport, or trifocal, distraction. It is well described in the orthopedic literature in treating segmental or intercalary bone defects.68,69 A similar application of this transport technique has been reported in treating patients with temporomandibular joint ankylosis. The fused proximal mandibular condylar segment is resected and a second osteotomy made near the mandibular angle. The ramal segment is then transported posterosuperiorly to reconstitute mandibular condyle, restoring occlusal vertical dimension and mandibular range of motion. Other than these examples, its use in the craniofacial skeleton has been more limited. In these applications, the strengths of distraction osteogenesis as compared with conventional techniques (well-vascularized bone and expansion of the soft-tissue envelope) may be outweighed by the liabilities of an extended period of treatment time (distraction and bone consolidation) and the lack of a need for expansion of the soft-tissue envelope. For example, after a composite mandible resection for cancer, the defect can be reliably reconstructed in a single-stage reconstruction with an osteomyocutaneous microvascular free tissue transfer (e.g., fibula, iliac, scapula). Perhaps the role of distraction osteogenesis in these clinical settings is limited to the salvage of failed primary reconstructive attempts, in which there already exists a situation in which wound contraction has occurred and expansion of this tissue is required.

Despite the unique power of this technique in treating facial deformity, few plastic surgeons currently use mandibular distraction osteogenesis as a therapeutic approach. Perhaps the problems that have been cited by some have limited widespread enthusiasm for the technique.70 This should not be the case. Appropriate planning and meticulous technique are essential for consistent success. In the application of the technique of distraction lengthening to the mandible, the surgeon must carefully consider the analogy of the mandible to long bones and the similar clinical circumstances that exist in these situations. The principles of mandibular fracture management should also be carefully remembered and respected. The mandible has considerable range of motion. Other than these examples, its use in the craniofacial skeleton has been more limited. In these applications, the strengths of distraction osteogenesis as compared with conventional techniques (well-vascularized bone and expansion of the soft-tissue envelope) may be outweighed by the liabilities of an extended period of treatment time (distraction and bone consolidation) and the lack of a need for expansion of the soft-tissue envelope. For example, after a composite mandible resection for cancer, the defect can be reliably reconstructed in a single-stage reconstruction with an osteomyocutaneous microvascular free tissue transfer (e.g., fibula, iliac, scapula). Perhaps the role of distraction osteogenesis in these clinical settings is limited to the salvage of failed primary reconstructive attempts, in which there already exists a situation in which wound contraction has occurred and expansion of this tissue is required.

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fracture of the angle of the mandible are substantial. The demands on a framework used in distraction osteogenesis of the mandible are even more substantial, and the use of three monocortical screws on either side of the osteotomy is unlikely to be sufficient in most cases. In addition, the technique used during placement and fixation of the distraction devices must be precise and pristine. Whereas the conventional techniques used in facial fracture management are useful as underpinnings, the technique used in placing distraction frameworks (internal or external) must be rigorously applied. The basic principles of using new fresh burrs, using constant irrigation during the drilling process, and minimizing thermal injury to the bone must be strictly followed in this technique. Furthermore, the actual placement of the pins and/or screws should be meticulous. If a pin or screw needs to be “backed out,” it is often better to drill a new hole and place the pin/screw with a “fresh” placement than to risk unstable and inadequate fixation that will loosen and lead to failure of the distraction process.

In summary, distraction osteogenesis provides a powerful and reliable technique for managing well-vascularized bone in mandibular reconstruction and providing simultaneous expansion of the facial soft-tissue envelope. These attributes have been used effectively in treating disorders that have been previously not optimally managed using the conventional techniques of orthognathic surgery, microvascular transfer, and nonvascularized bone grafting. The most frequent application has been in treating congenital deformities. The technique is reliable, but strict attention to technical details of bone fixation should be observed.

Maxillary Distraction at the Le Fort I Level

Maxillary hypoplasia frequently occurs in patients with cleft lip and palate. In approximately 25 percent of these cases, the class III malocclusion is severe enough to require surgical intervention. Distraction at the Le Fort I level has become the workhorse for managing these severe maxillary retrusions commonly associated with cleft lip and palate. Before the advent of conventional Le Fort I osteotomy, this difficult condition was treated for decades by orthodontists using reverse face gears with heavy elastics. The result is usually disappointing, with advancements only in the range of 3 to 4 mm. The rate-limiting factor is the extensive palatal scaring. This means that to advance the maxilla, significant tensile force is required. This tensile force on the maxilla necessarily produces corresponding pressure on the anchoring pads over the forehead and chin. The tensile force necessary to achieve maxillary advancement appears to be high enough to produce sufficient pressure to cause skin necrosis under the anchoring pads. To reduce this tensile force, the resistance to anterior translation of the maxilla was decreased by complete Le Fort I osteotomy. This surgically assisted maxillary advancement used a combination of face mask, heavy elastics, and Le Fort I osteotomy and improved the magnitude of advancement to the 5-mm range measured at the maxillary incisor edge, which is comparable to conventional Le Fort I osteotomy and advancement. The next improvement came about by providing a rigid anchorage directly to the temporal region of the cranial skeleton using pin-retained hemi-halo and screw-generated pull in lieu of the elastics. This resulted in the current, versatile, external maxillary distraction system, which is capable of advancing maxilla for more than 30 mm. Polley’s early reports showed average advancement of 11.6 mm with minimum relapse. One key component is the solid osseous anchorage. By using a torque wrench to tighten the transcutaneous anchoring screws, the stress levels of 8 psi for adults and 4 psi for children were provided as a guide. Because there is no need to establish maxillomandibular fixation, the operation can be performed with oral intubation. Because there is no need for plate-and-screw fixation, the osteotomy can be made very high, at the level of the infraorbital foramen. This has two very significant salutary effects: avoiding damaging the developing permanent tooth follicles and providing high-level central midfacial advancement (Fig. 5). In addition, because no plating is necessary, the operative time is reduced. The conventional Le Fort I osteotomy with plate-and-screw fixations has been used extensively before distraction, but it is limited by the amount of advancement possible and a significant relapse rate. The average advancement achieved by experienced surgeons using conventional Le Fort I varies from 4.5 mm to 7.8 mm for unilateral clefts and an average relapse of 4 to 40 percent, with larger relapses seen in longer follow-ups.

The need and the desire to close the anterior open bite during the advancement neces-
sitate a downward vector. If the osteotomy is performed at the Le Fort III level, the inevitable result is vertical elongation of the orbit. However, at the Le Fort I level, the downward movement is only limited by the aesthetics. Occasionally, too much gingival show renders further clockwise rotation of the maxilla unacceptable. This occurs when the posterior aspect of the maxillary segment has rotated inferiorly because of the center of resistance being higher than the line of pull, thus creating a turning moment rotating the maxilla counterclockwise. To lessen this, the vector of pull should be made high. The risk of Le Fort I distraction has all the risks of conventional Le Fort I osteotomy and the added potential problems related to the distractor.\textsuperscript{76,79} Significant hemorrhage can occur during Le Fort I osteotomy, especially when the bone cut is carried high posterolaterally into the pterygomaxillary fissure. Blindness following Le Fort I osteotomy for distraction has also been reported.\textsuperscript{80} The external device is bulky and has been associated with compound cranial fractures as a result of minor trauma.\textsuperscript{81} In summary, there are five significant advantages for distraction osteogenesis of the maxilla at the Le Fort I level: large advancements, low relapse rates caused by simultaneous soft-tissue expansion, decreased operating time, the ability to keep the osteotomy high, and low incidence of detrimental speech outcome resulting from velopharyngeal insufficiency.\textsuperscript{82}

**Maxillary Distraction at the Le Fort III Level**

Distraction techniques were first adapted to the midface by craniofacial surgeons treating children with craniofacial dysostosis–associated maxillary hypoplasia. Using a device that penetrated the skin in the malar region, Chin and Toth\textsuperscript{11,83} were the first to
report distraction of the maxilla. Their device was used to rapidly expand the Le Fort III osteotomy gap and then was left in place for 6 months before removal. This initial report was notable for the use of rapid expansion (other surgeons using distraction techniques were typically using a 1-mm/day expansion rate). Despite this accelerated advancement, new bone formation was clinically evident after a 6-month consolidation period. The authors did observe significant bradycardia during the distraction phase in two of their initial series of nine patients, presumably secondary to the oculocardiac reflex.

Shortly after this report, other surgeons adapted similar distraction devices and rotated them 180 degrees to allow the devices to exit in the less noticeable preauricular area. More importantly, the speed of distraction was slowed down, which permitted faster bony consolidation, shortening the time needed for rigid retention. Additional experience with Le Fort III distraction revealed that these devices were not without some significant downsides. One common problem is the difficulty in finding a reliable point of attachment, distraction will either occur asymmetrically, lagging on the side that is not stable, or not occur at all (if both sides are unstable). Longer devices, with a broader plate for fixation, are a potential solution to this predicament, but their use is not always possible in younger children because screw placement in the maxilla may damage permanent tooth follicles. Another problem with the use of bilateral buried devices is the inability to change the vector of distraction once the buried plates are in place. Difficulties frequently arise with the removal of the devices, which tend to become imbedded in bone during the distraction process.

While many surgeons were in the process of experimenting with Le Fort III distraction, Polley and Figueroa were working on an external halo distraction device, based on the orthodontic face mask popularized by Delaire. This device was developed to treat the difficult advancements of the Le Fort I segment associated with a cleft lip and palate. Attachment of the external device to the Le Fort I segment was accomplished through the use of heavy orthodontic wires. Their technique was found to be extremely effective at advancing the midface, despite a scarred palate (average reported advancement, 11.7 mm). Moreover, these advancements were accomplished with an extremely low complication rate. Of all the available distraction devices, the hemi-halo–type external distractors are the most adjustable. Unhappy with the results of bilateral internal distraction devices, Fearon adapted this hemi-halo distraction device for the treatment of children with craniofacial dysostosis–associated midfacial deficiency. Modifications were made in the standard Le Fort III osteotomy to permit greater advancement, and this external device was attached to the maxilla through the use of a dental splint secured with maxillary drop-wires. When compared with a cohort of age-matched controls who had undergone the standard Le Fort III procedure, this initial series of Le Fort III hemi-halo distraction patients was shown to have a significantly greater advancement of the maxilla (average advancement, 19 mm), without any increase in complications. Computed tomographic scan analysis suggested that the facial profile of patients who had undergone hemi-halo distraction was preferable to those who had undergone a standard advancement. This improved appearance is believed to derive from the midline vector of traction associated with the use of the external device (by pulling the centrally depressed face forward). The bilateral zygomatic-based, internal devices advance the lateral aspects of the midface, potentially exacerbating the centrally deep midface. Other advantages of the halo distraction Le Fort III over the standard procedure included better correction of sleep apnea and shorter operative times (secondary to the elimination of the need for rigid fixation and the need to harvest cranial bone graft). As with distracting other sites of the facial skeleton, the results achieved with midfacial distraction are critically dependent on the vector of distraction. The external halo distracter permits changes in the vector of distraction after placement, unlike buried devices. The halo distraction device has also been reported to be a successful salvage technique for complications arising from bilateral buried subcutaneous devices.

Unlike distraction at the Le Fort I level, the Le Fort III distraction procedure is not an orthognathic procedure; it is a technique that should be used to reposition the malar emi-
nences. It is important to avoid the temptation to try to close the anterior open bite that is typically seen in the craniofacial dysostoses, because this can only be accomplished at the expense of unnaturally lengthening the vertical orbital distance. The primary indication for distraction of the Le Fort III is for the treatment of the hypoplastic maxilla in children who have not completed facial growth (where some degree of overcorrection is desired) (Figs. 6 and 7). Distraction is seldom indicated in the mature facial skeleton and should be reserved for planned advancements in excess of 1 cm in patients in whom scarring may prevent accomplishing this with a standard procedure. Lengthening of the maxilla (with

Fig. 6. (Above, left) A 7½-year-old boy with Apert syndrome seen before distraction. (Above, right) Preoperative lateral view. Notice the significant midfacial retrusion. (Below, left) Frontal view of the patient after Le Fort III osteotomy, with a rigid eternal distractor in place. (Below, right) Lateral view of Le Fort III distraction in progress. Notice the overcorrection.
closure of the associated open-bite deformity) is best delayed until the middle to late teenage years, when facial growth is complete. At this time, the maxilla may be both advanced and lengthened with a combination standard Le Fort III with Le Fort I.

In summary, distraction of the midface offers numerous advantages: this technique permits gradual expansion of the surrounding soft-tissue envelope, permitting a greater advancement than could be achieved with traditional procedures. Although the treatment time is longer, distraction is a shorter operation that eliminates the need for internal fixation and bone graft harvesting. With no metal plates left behind, secondary surgery is greatly facilitated, should the need for it arise. The disadvantages of midfacial distraction include the following: a second procedure is needed to remove the device, distraction is not always symmetric, the postoperative orthodontic work is more challenging, and the maxilla cannot be simultaneously vertically lengthened if the distraction is at the Le Fort III level.

FRONTAL FACIAL ADVANCEMENT AND DISTRACTION OSTEOMESIS

An extremely important technique for reconstructing patients with syndromal midfacial deformities is the frontal facial advancement or monobloc osteotomy. Frontal facial or monobloc advancement for a patient with a syndromal midface deformity is often the most important procedure that a patient may undergo for rehabilitation of their congenital facial disfigurement. During this procedure, the forehead and the face are literally sectioned or separated from the skull base and repositioned three-dimensionally to correct the midface and frontal deficiencies. For appropriately selected patients, this is an outstanding procedure that can yield dramatic morphological and functional results. The frontal or forehead advancement expands the anterior cranial vault, releasing intracranial hypertension, which is common as a result of the bicoronal synostosis in these patients. The forehead advancement is also used for normalization of the frontal bone and forehead aesthetic projection. The orbital advancement with the monobloc osteotomy moves the entire “functional orbit” forward, normalizing its position, giving appropriate support to soft tissues about the orbital rims and eyelids, and repositioning the orbital walls, allowing normalization of vectors of the extraocular musculature. In this procedure, exorbitism and exophthalmos are corrected, normalizing orbital aesthetics and improving extraocular muscular imbalance. The monobloc osteotomy also advances the entire nose.
including the nasal dorsum. This repositioning of the nasal structures in the sagittal plane normalizes nasal aesthetics and opens the nasal airway passage. In addition, advancement of the palate, maxilla, and zygomatic bones corrects a myriad of functional problems for these patients, including opening the oral airway, correcting class III skeletal and dental relationships, and creating the proper oral and nasal cavities for improvement in articulation and speech resonance. Although the functional and morphological gains from this operation are essential for these patients, the monobloc osteotomy is perhaps the most feared osteotomy in all craniomaxillofacial surgery. Standard monobloc advancement procedures include the frontal facial disjunction, intraoperative repositioning of the entire face and forehead complex, extensive interpositional bone grafting, and many points of rigid internal fixation. In addition, in this operation, the midface is often stabilized with intermaxillary fixation as well. The procedure can be a long one and requires extensive bone grafting and carries the potential for major blood loss. The greatest problem with the monobloc osteotomy with the traditional approach has been the extremely high incidence of postoperative infection. This incidence can range anywhere from 10 to 50 percent, even in experienced hands. When an infection does occur, it is not typically a small problem but can include major intracranial abscesses, extensive bone destruction, and even mortality. It is for this reason that in the past many excellent centers worldwide have avoided the monobloc osteotomy.

The application of distraction osteogenesis for the monobloc osteotomy has revolutionized this technique and has enabled this outstanding procedure to now be performed by many surgeons worldwide.87 Frontal facial advancement through distraction osteogenesis offers many advantages over the traditional monobloc procedure. Some of these advantages include a decreased operative time, the fact that bone grafting is no longer required, elimination of internal fixation, decreased blood loss, and decreased hospitalization. The greatest advantage for distraction osteogenesis and the monobloc osteotomy is the potential reduction of the infection rate with this procedure.88 With monobloc distraction, the osteotomized frontal and facial bones are not advanced on the table at the time of the surgery. This means there is no significant dead space created in the anterior cranial fossa at the time of the operation. The slow, gradual, rhythmic distraction of the frontal facial region occurs at a rate that does not create a significant open dead space communicating the anterior fossa with the nasal pharynx.89 In this fashion, ascending oral pharyngeal contamination into the intracranial space can be greatly reduced and controlled.

Monobloc midface distraction can be performed with an external distraction device or with an internal device anchored along the zygomatic arch.90 Although both techniques are used, many centers prefer an external distraction device for the monobloc advancement. External distraction allows complete control over the advancing midface and frontal bone. Typically, with external monobloc distraction, anchorage points to the skeleton following osteotomy are in the frontal bones bilaterally and to the maxillary dental splint. Two points of fixation to the strong frontal bone and two points of fixation to the intraoral splint allow four points of excellent control for advancing and repositioning of the midface. The distraction procedure allows basically unlimited sagittal advancement of the midface, and rotational and vertical changes can be made as the distraction process continues. Final positioning of the midface can be titrated on the basis of individual aesthetic requirements for the patient. The patients typically wear the external halo for approximately 3 months. After a 1-week latency period, activation begins and continues over the next 2 to 3 weeks. Consolidation of the advanced segment is over the next 6 to 8 weeks. Confirmation of consolidation should be performed on a clinical basis, according to physical examination (Fig. 8).

The experience with internal distraction of midface monobloc osteotomies has been disappointing in some centers. The main problem is the lack of control over the midface section. With internal distraction, the distraction devices are secured at the time of surgery and the final position of the midface cannot be adjusted during the course of distraction. Gauging precise vectors intraoperatively for placement of the distraction devices is difficult. Internal distraction osteogenesis in the monobloc setting may have its greatest application for those uncommon instances where midfacial advancement is required in infancy. One
additional disadvantage of the internal distractor is the need for and difficulty of removal.

Much still needs to be analyzed and learned regarding monobloc distraction osteogenesis. As experience continues to grow, the technique’s reliability, predictability, and very low complication rate promise to be even more impressive. With greater experience, the precise indications for monobloc midface advancement with distraction osteogenesis will become elucidated.

**Calvarial Distraction Osteogenesis**

The calvaria is one of the last regions of the craniofacial skeleton to enter clinical distraction. However, the concept of expanding the cranium by tensile force has been reported since 1986. Several groups have adapted absorbable plates and screws for the anchorage while using high-strength, nonabsorbable axial screws for activation. This has made device removal less cumbersome. Because soft-tissue constraint is one of the theoretical rate-limiting factors in the correction of craniosynostosis by conventional fronto-orbital advancements, distraction is ideal for overcoming this particular problem. Research efforts have concentrated over the past several years on the development of totally implanted distraction devices. However, to date, there remains no approved, completely submerged cranial distraction system for clinical use in this country. A group in Sweden led by Lauritzen has developed and deployed implanted springs to achieve distraction osteogenesis of the cranium. They used compressed springs to push apart the bone segments following osteotomies. They reported the tensile force delivered by a spring to be as high as 20 N. This type of distraction differs from all other types of distraction osteogenesis in that the force is continuous and ever reducing. Insufficient data exist at the present to evaluate this type of distraction osteogenesis. The indications for distraction os-
teogenesis of the cranium are not as clearly defined as those for the other levels of the craniofacial skeleton. One concern is the compressive force that is necessarily placed across the patent sutures located anterior and posterior to the fused one undergoing distraction. For example, in placing the distractor across the coronal region, the ipsilateral lambdoid, frontozygomatic, and frontonasal sutures must necessarily undergo compressive strain. How this affects the behavior of these patent sutures has not been well documented. The experience with intracranial constraint models certainly indicates that, given enough compression, sutures will fuse.96,97 The compressive effects on the temporomandibular joint have been reported.98 Similar concerns about the bordering joints have been expressed in the orthopedic literature.99

**CLINICAL OUTCOME AND COMPLICATIONS**

Distraction osteogenesis is not performed by a large number of plastic surgeons. In a recent survey of 2476 surgeons in the United States and other countries, only 274 (11 percent) responded, and of this group, only 148 stated that distraction is part of their practice.70 The ability of distraction to provide superior advancements is clear. These advancements are much more resistant to relapse. Perhaps the most significant point is that distraction osteogenesis represents an emerging enabling technology. It is a potent surgical tool and can produce stable advancement exceeding 20 mm at every level of the craniofacial skeleton and soft-tissue envelope, advancements that could not be easily achieved without distraction. However, at present, it is associated with an impressive complication rate. The reported complication rates vary from 0 percent to 35.6 percent to 60 percent.70,82,100 The potential complications include the following: device failure; pin pullouts; infection; hardware exposure; damage to vital structures and adjacent joints; inappropriate consolidation (premature consolidation or fibrous nonunion); and inappropriate vector of distraction resulting in asymmetry, less-than-ideal occlusion, or frank malocclusion. The orthodontic work after distraction is more extensive than after conventional orthognathic surgery. Movements that can be satisfactorily achieved without distraction osteogenesis should be treated with conventional, single-stage operations.102 Faced with a rapidly increasing number of new distraction devices, surgeons must be reminded of the learning curve so as to balance between adopting newer, more efficient devices with which he or she has little experience and accumulating sufficient cases with the existing systems to gain proficiency.

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