

The Role of Distraction Osteogenesis in the Management of Craniofacial Disorders

S R Cohen,*^{MD}, F D Burstein,**^{MD}, J K Williams,**^{MD}

Abstract

Since its introduction in the medical literature in 1992 by McCarthy, distraction osteogenesis of the craniofacial skeleton has become a standard surgical therapy. The present report attempts to trace the development of craniofacial distraction from the perspective of one of the early proponents of the technique. Although the earliest application of distraction was in children with severe unilateral or bilateral mandibular deficiency, its use for functional abnormalities such as apnoea were especially appealing. Distraction osteogenesis for the midface began with external appliances that were attached to the teeth. Newer, buried devices have eliminated the need for external devices in all LeFort III and monobloc cases. For LeFort I and mandibular cases, the ideal internal device has not been manufactured.

Ann Acad Med Singapore 1999; 28:728-38

Key words: Aperts, Crouzons, Distraction osteogenesis, LeFort I, LeFort III, Mandibular hypoplasia, Monobloc, Pfeiffer syndromes

Introduction

The concept of distraction osteogenesis was championed by Ilizarov,¹ beginning as early as 1954, for the treatment of a variety of congenital and acquired deformities of enchondral bone. In 1973, Snyder and associates* reported on gradual distraction of the mandible using an extraoral device in canines. After successfully lengthening the canine mandible,³ McCarthy and his group reported the first clinical application of distraction osteogenesis of the mandible in 4 patients with a variety of congenital craniofacial anomalies in 1992.⁴ Because the technique of gradual osseous distraction of the membranous bones of the human craniofacial skeleton promises major advances in the early reconstruction of severe craniofacial anomalies, numerous reports of clinical mandibular distraction, as well as experimental distraction, in other areas of the craniofacial skeleton have ensued. Osseous expansion of the cranial vault was first reported by Persing and colleagues⁵ in an animal study using spring devices. Distraction of the frontal bone outside the cranial plane was demonstrated by Barone and coworkers.⁶ Successful expansion of the cranial vault using a cranio-tactic device in rabbits was presented by Remmler and his coworkers.⁷ More recently, midfacial advancement was shown to be feasible in adult sheep using lengthening bolts mounted on transversely placed pins.⁸ Using a modified Hoffman

bone-lengthening device, Staffenberg and associates (unpublished data) reported successful midface distraction and advancement in the immature canine without osteotomies. The application of distraction osteogenesis to the human craniofacial skeleton represents a major advance in the treatment of craniofacial-orthognathic deformities. The technique is less invasive than conventional surgery and possibly more stable. Moreover, complex manipulation throughout the craniofacial skeleton can be achieved using buried internal devices. Herein, the clinical applications of craniofacial distraction in the mandible, maxilla, midface, and cranium are discussed. This article also intends to retrace some of the steps taken to develop the Cohen Modular Internal Distraction System (MIDS, Leibinger, Inc).

Our own experience with distraction began in a patient with severe relapse following correction of skeletal deformities associated with a rare craniofacial cleft and anophthalmia and in children with airway disorders. Airway disorders deserve special mention because they can dramatically impact successful functional outcomes in children with craniofacial disorders.⁹ Sleep apnoea, ranging from acute obstructive upper airway collapse, or more insidious variants with noisy breathing at night, frequent wakeful episodes, and daytime somnolence. Poor school performance, deterioration of mental status and, more severe, cardiopulmonary problems can oc-

* Chief

Craniofacial Surgical Services

Children's Hospital of San Diego, San Diego, California

** Co-Director

Craniofacial Surgery, Center for Craniofacial Disorders

Scottish Rite Children's Medical Center, Atlanta, Georgia

Address for Reprints: Dr Steven R Cohen, 8010 Frost St., Suite 412, San Diego, CA 92123, USA.

cur. In longstanding cases, cor pulmonale can be developed from increased pulmonary artery pressures. Children with craniofacial disorders and neurologic problems such as cerebral palsy and Down syndrome are at risk for apnoea.¹⁰ Recognition of apnoea by clinical history is confirmed by quantitative documentation with polysomnography. Twelve-channel overnight polysomnography, often with simultaneous PH probing, is the gold standard for diagnosis of central or obstructive sleep apnoea syndromes. Once diagnosed, the treatment of the anatomic abnormalities can be defined radiologically or endoscopically, and appropriate medical and surgical treatment can be instituted. Medical options include CPAP, intermittent night-time oxygen, and excellent pulmonary toilet.⁹ Conservative surgical options include tonsillectomy/adenoidectomy, uvulopalatoplasty, and nasal septoplasty.⁹ In more severe cases refractory to simple medical or surgical measures, a more aggressive surgical approach is taken.⁹⁻¹² This approach is predicated on the need to expand the craniofacial skeleton, while simultaneously reducing any soft tissue interference such as macroglossia, uvulopalatal excess, and infiltrative processes such as neurofibromatosis of the anterior neck or palate. Distraction has been successfully employed to treat mandibular and maxillary retrusion in younger age groups and represents a less invasive means of surgical treatment.

Mandibular Distraction

Mandibular distraction osteogenesis in the treatment of upper airway obstruction in children with craniofacial deformities was reported by Cohen, Simms, and Burstein¹³ in 16 patients (12 males, 4 females) who had undergone the procedure in conjunction with soft-tissue reduction to treat medically refractory obstructive sleep apnoea. Thirty external distraction devices were placed for bilateral distraction in 14 children and unilateral distraction in 2. The mean age of treatment was 4 years, 8 months (range 14 weeks to 12 years, 8 months). Eight of the patients had failed conventional medical and surgical treatment of obstructive sleep apnoea and were considered tracheostomy candidates, whereas the remaining 8 had tracheostomies placed shortly after birth for upper airway obstruction. These 8 children could not be decannulated by standard protocols. The average distraction distance in our study was 25 mm, with a range of 18 to 35 mm. To date, 7 of the 8 patients with tracheostomies have been decannulated, while one is still in progress (Fig. 1). Clinical improvement in signs and symptoms of sleep apnoea and reduction or elimination of preoperative oxygen requirements occurred in 7 of the 8 children with medically refractory obstructive sleep apnoea. Twelve-channel polysomnograms were obtained preoperatively and postoperatively in each of the 8 patients without tracheostomies. Respiratory dis-

turbance index decreased from a mean of 7.1 to 1.7 after surgery. Lowest oxygen saturation rose from a mean of 0.70 to 0.89 after surgery. Application of mandibular distraction osteogenesis is an important component in the treatment of obstructive sleep apnoea and permits mandibular advancement on the younger child (Fig. 2). As more experience is gained with distraction osteogenesis in the treatment of children with obstructive sleep apnoea, the role of distraction will become better defined. The clinical diagnoses and complications in this group of 16 patients is shown in Tables I and II, while associated soft-tissue procedures are shown in Table III. We have now performed distraction osteogenesis using a modified external device with IC wires through the inferior chin and angle of the mandible in the 35-week-old premature baby with Pierre Robin anomalad. This child tolerated distraction at a rate of 2 mm per day, which permitted extubation in the operating room with fibreoptic endoscopy. The wire piercing the mandibular angles traverses the base of the tongue and needs to be

TABLE I: CLINICAL DIAGNOSIS

	Number
Hemifacial microsomia	7
Pierre Robin Sequence	3
Cerebral palsy	2
Treacher Collins	1
Oromandibular limb hypogenesis	1
Microcephaly	1
Retrognathia	1
Total	16

TABLE II: COMPLICATIONS

	Number
Aspiration	1
Pin site infection	1
Pin loosening	4
Transient marginal mandibular nerve palsy	1

TABLE III: ASSOCIATED PROCEDURES

	Number
Correction of choanal stenosis	2
Septoplasty	1
Turbinectomy	5
Tonsillectomy/Adenoidectomy	7
Tongue reduction	2
Uvulopalatopharyngoplasty	5
Tongue hyoid suspension	7
Correction of nasopharyngeal stenosis	1
Epiglottal suspension	1
Wilkes-Brodie	



Fig. 1a.



Fig. 1b.



Fig. 1c.



Fig. 1d.

Fig. 1. One-year-old girl with right unilateral craniofacial microsomia who required tracheostomy for upper airway obstruction shortly after birth. (a) Preoperative frontal and (b) lateral photograph demonstrating retrognathia with tracheostomy in place. (c) Postoperative frontal and (d) lateral photographs taken one year later, after tracheostomy decannulation.



Fig. 2a. Preoperative lateral photograph of 7-year-old girl with bilateral craniofacial microsomia and obstructive sleep apnoea. Preoperative respiratory disturbance index on 12-channel overnight polysomnogram was 11, whereas lowest recorded oxygen saturation was 59. Fig. 2b. Postoperative lateral photograph following bilateral mandibular distraction for distraction distance of 20 mm. Post distraction respiratory disturbance index was 0.7, whereas lowest oxygen saturation was 87.

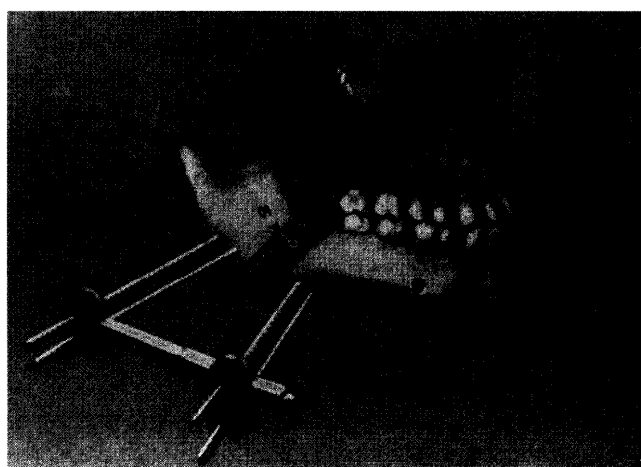


Fig. 2c. Artist depiction of mandibular distraction technique with interposition bone graft to provide immediate sagittal advance in children with obstructive sleep apnoea. (Published with permission from: Cohen S R, Simms C, Burstein F D. Mandibular distraction osteogenesis in the treatment of upper airway obstruction in children with craniofacial deformities. *Plast Reconstr Surg* 1998; 101:312-8.)

the first wire removed to permit easy reintubation, if necessary.

Distraction osteogenesis of the mandible, and occasionally maxilla, fits nicely within the treatment protocol for children with craniofacial microsomia. McCarthy, a leader in this area, has outlined his protocol.¹⁴ He breaks down treatment ages and anatomic classification using the Mulliken and Kaban¹⁵ modification of the original Pruzansky¹⁶ classification:

- Type 1: Hypoplasia of the ramus and the body of the mandible is minimally or slightly affected.
- Type 2: The condyle and ramus are small; the head of the condyle is flattened; the glenoid fossa is absent; the condyle is hinged on a flat, often convex, infratemporal surface; the coronoid process may be absent.
- Type 3: The ramus is reduced to a thin lamina of bone, or it is completely absent.

The Type 2 mandibular deformities are further subdivided into A and B. In Type 2A, there is an abnormality in size and shape of the ramus and the condyle, but the glenoid fossa-condyle relationship is preserved. The glenoid fossa has a normal spatial position, and temporomandibular joint function is almost normal. In Type 2B, the condyle is displaced outside of the vertical plane of that on the contralateral side. Consequently, there is restricted hinge-like functioning of the mandible on the affected side.

McCarthy rarely undertakes mandibular reconstruction under 2 years of age.¹⁴ To prevent tracheostomy, we have utilised distraction osteogenesis in selected infants with craniofacial microsomia and upper airway obstruction. We have also utilised mandibular distraction for tracheostomy decannulation, although this is usually not begun until 2 years of age. From ages 2 to 6 years, the technique of mandibular distraction can be consid-

ered. Not only does distraction expand the affected mandible in all dimensions, it also positively augments the associated soft tissues and muscles of mastication, an effect that probably reduces the rate of relapse and bone graft resorption. In addition, studies¹⁷ of the temporomandibular joint have shown that the distracted ramus and condyle assume a more normal anatomic size, shape, and position. For the child with a Pruzansky Type 1 mandible and a horizontal occlusal plane, distraction would not be recommended, and any type of skeletal reconstruction would be deferred. Molina (unpublished data), however, has shown nice results in children with Type 1 deformities. This is still a controversial issue, but for mild cases, it is probably best to defer surgery until definitive orthognathic surgery can be carried out. For the child with Pruzansky Type 3 deformity, traditional costochondral rib graft reconstruction is performed at about 3 to 5 years of age. Occasionally children will have bilateral craniofacial microsomia with Pruzansky Type 2 and 3 deformity. In these cases, we have used costochondral grafting simultaneously with distraction. Careful planning is necessary to successfully treat these patients. In the child with severe bilateral mandibular deficiency (Pruzansky 2 or 3) with associated sleep apnoea and tracheostomy, bilateral mandibular distraction can be carried out as early as 2 years of age. The technique has distinct advantages in that reconstruction is undertaken earlier and, more importantly, decannulation of the tracheostomy is usually feasible.

The Pruzansky 2 lesions are those most often treated with distraction. Unidirectional or multiplanar distraction can be carried out. Other indications for mandibular distraction include mandibular retrognathia of any cause, especially if associated with upper airway obstruction or sleep apnoea, unilateral or bilateral mandibular hypoplasia secondary to craniofacial microsomia, and retrognathia from any cause producing severe anatomic or functional deformity. In addition, distraction can be applied as a bone transport technique to close segmental defects following cancer resection. The advantage in this application is simultaneous expansion of the gingival-alveolar ridge. Presently, devices for transport are bulky and cumbersome and, in the author's opinion, microsurgical reconstruction is still the technique of choice for the majority of segmental and/or composite mandibular defects.

Midface Distraction

Midface distraction has been applied since at least 1993 or 1994. As early as the fall and winter of 1993, several centres had already performed midface distraction. Even earlier, Molina (unpublished data), working in Mexico City, presented a series of patients who had undergone high LeFort osteotomies and midface distraction by reverse headgear, which essentially was a

tooth-borne dental orthopaedic appliance. In December 1993, at the Scottish Rite Children's Medical Center, our group performed a buried midface distraction in a child with left anophthalmia and craniofacial microsomia.¹⁸ In Munich, Muhlbauer¹⁹ conducted a series of LeFort III and monobloc distractions in a small group of patients with Apert syndrome. His results were reported at the International Meeting of Plastic, Reconstructive, and Aesthetic Surgery in Yokohama, Japan, in 1995. Chin and Toth²⁰ reported on 5 patients who had undergone buried distraction, 2 of whom underwent LeFort III midfacial advancement. Using an external traction appliance, Polley et al²¹ demonstrated a dramatic frontofacial advancement in a newborn with extreme proptosis and upper airway obstruction. In late 1994 and early 1995, we performed buried modified LeFort III advancements in 2 children who had cleft lip and palate with relatively severe midface hypoplasia and Class III malocclusion.²² In each case, transverse maxillary expansion was carried out simultaneously to distraction and, in one case, serial distractors were utilised to provide both vertical and horizontal distraction vectors. In 1996, using specially designed buried midface distraction devices that were easily customised by the surgeon at the operating room table, we performed a subtotal cranial vault reshaping and monobloc facial advancement in a child with Pfeiffer's syndrome with corneal exposure.²³ Following 28 mm of distraction, the proptosis was largely corrected. In 1997, using the Cohen Modular Internal Distraction system, we carried out the first monobloc distraction with facial bipartition. This demonstrated that segmental osteotomies could be utilised, and the newly formed segment could then be distracted into position.²⁴

The indications for midface distraction are currently being established. It is the author's opinion that in the future, monobloc and LeFort III advancement will be done by midface distraction techniques, especially in the younger child. The morbidity of the LeFort III osteotomy and, especially the infectious morbidity of the monobloc, appear to be reduced with distraction. Length of intensive care unit stays, intraoperative blood loss, length of operative times and complications all appear to be reduced using distraction techniques. The conventional LeFort I osteotomy is a simple procedure in the adolescent and adult. In patients, however, with extreme soft-tissue scarring, severe Class III deformities with and without a history of relapse, and in younger children with cleft lip and palate and severe Class III deformities, midface distraction is a promising technique. Molina (unpublished data) and Polley and Figueroa²⁵ have championed external devices. The current consensus is reverse head gear may be suitable for mild deformities, such as 3, 4, 5, and maybe even up to 6 mm underbite, but should not be used in more severe deformities (Fig. 3).



Fig. 3a. Maxillary distraction with reverse head gear and elastic traction following high LeFort I osteotomy.



Fig. 3b. Preoperative lateral and Fig. 3c. postoperative lateral photographs following maxillary distraction.



Polley's device,²⁵ the rigid external distraction (RED) system has shown superb results in cleft patients, but does require the children to wear a bulky and embarrassing external fixation device for up to 6 weeks or longer. The internal distraction devices are easily tolerated by the patient and permit resumption of the majority of school and physical activities. One disadvantage, however, is the need for removal. As much as possible, we have tried integrating midface distraction at the LeFort I level into an overall treatment plan for the cleft child with severe Class III malocclusion. A palatal expander is placed first, and transverse dimensions of the palate are re-established, while simultaneously preparing the bone graft site. Whenever possible, and indeed preferably, orthodontic appliances with surgical hooks are placed and some straightening of the dental maxillary arch is carried out. Hooks are placed on both upper and lower teeth to allow placement of dental elastics at the end of the distraction to mould and manipulate the newly formed regenerative bone. Callus manipulation permits fine-tuning of the occlusion during internal distraction. Midface distraction carried out at whatever level provides the best aesthetic outcome. Once distraction is completed, the patient is returned to the operating room, where alveolar bone grafts are carried out after device removal. In this manner, distraction is integrated into the overall treatment regimen. The indications for midface distraction in children with syndromic craniosynostosis and midface retrusion appear fairly clear cut. In children undergoing monobloc advancement with distraction osteogenesis, gradual expansion of the anterior cranial fossa appears to prevent the immediate development of a large epidural dead space and dimin-

ishes the frequency of epidural infection. In addition, procedure time is shortened with less blood loss, reduced intensive care unit stay, and earlier hospital discharge. Because distraction osteogenesis is a potentially less invasive technique, the mortality and morbidity of the procedure is diminished, permitting earlier frontofacial advancement in infants with syndromic craniosynostosis. Repeat distraction osteogenesis can be carried out, again, more safely than monobloc and/or LeFort III advancement in the 5- to 7-year-old age group. Other indications for midface distraction include treatment of severe maxillary cants in patients with hemifacial microsomia. Molina and Ortiz-Monasterio et al²⁶ have employed a maxillary distraction technique in conjunction with mandibular distraction in patients with hemifacial microsomia to simultaneously correct the occlusal cant. In their technique, a maxillary osteotomy is made, as is a mandibular osteotomy. External mandibular distraction devices are inserted and the patient is placed in maxillomandibular fixation with dental elastics. As the mandible elongates, the maxilla is dragged along with it.

Toth and Chin²⁷ completed 13 LeFort III osteotomies with a technique called continuous midface distraction. In contrast to gradual distraction osteogenesis, a LeFort III osteotomy is performed and an immediate advancement up to 10 mm is carried out. Following this, distraction is carried out over the next several days while the patient is left intubated in the intensive care unit. Distraction is performed with sizeable external pins, and the devices are extremely difficult to remove because of their self-retaining nature. In addition, the devices are not applicable to younger age groups where the poste-

rior aspect of the zygomatic body and arch will not hold the forces required for distraction using the retained devices.

Modular Internal Distraction System (MIDS): Since the first clinical application of distraction osteogenesis to the human craniofacial skeleton was reported by McCarthy¹⁸ in 1992, it was apparent that internal devices would offer certain advantages in terms of patient acceptance, fixation, stability and wound management. An initial prototype of an internal distraction device was fabricated by attaching conventional metallic plates to orthodontic expansion screws. In 1994, midface distraction was reported in a child with craniofacial microsomia.¹⁸ The simplicity of the design permitted attachment of a wide array of plate configurations and sizes, making the system applicable for distraction in any region of the craniofacial skeleton. Moreover, with mild changes in the dimension of the distraction frame, the system can be applied to any shape, size, and age group.

The distraction device was subsequently modified, permitting internal distraction with a small, percutaneous activation screw. Using these new prototypes, monobloc advancement by distraction was successfully carried out on a 9-month-old child with Pfeiffer's syndrome.²³ The profile of the devices was further lowered, and a flexible cable was added to allow percutaneous activation from a distant site. The newest generation of devices have been used for mandibular, LeFort I, LeFort III, monobloc with and without facial bipartition, and cranial distraction. In September 1997, the Federal Drug Administration (FDA) approved the distractor as one of the first internal devices for the midface, mandible and cranium [Modular Internal Distraction (MIDS), Howmedica-Leibinger, Inc.]²⁸

Our design allows the surgeon to fabricate custom internal distraction devices for virtually any region of the craniofacial skeleton. The first-generation system contains expansion screws capable of 15-mm and 30-mm of distraction. Depending on the distraction site and osteotomy, any configuration of titanium plates can be attached to the distraction screw to permit uniplanar, and possibly biplanar, internal distraction. The flexible activation cable is brought out through a distant, inconspicuous stab wound in the hair, behind the ear, or even intraorally.

Mandibular Distraction (Fig. 4): Mandibular distraction is carried out by assembling the devices based on the lateral cephalogram. The appropriate vector is chosen for horizontal, oblique, or vertical mandibular advancement. An acetate overlay template of the 15- and 30-mm devices are available from Leibinger to assist with planning. A long incision is made in the lower buccal sulcus and ramal regions for complete exposure of the mandible. The device is stabilised by first placing the cable

through the percutaneous stab wound. The cable in its track serves as a "third hand," stabilising the device prior to bony fixation. The plates can be cut and adapted to the mandible in situ without the need for complicated reassembly. Once the device is anchored into position, the ramus is cut with a reciprocating saw, proceeding from the buccal to lingual cortex posterior to the nerve. The lingual aspect of the osteotomy may need to be completed with a fine osteotome. Leaving the device open 1 to 2 mm facilitates the osteotomy. Although we have not removed the device for the osteotomy, it is possible that once the drill holes have been made for fixation of the device, the device can be retracted laterally while the osteotomy is conducted from the buccolingual direction. The 15-mm length mandibular distractor is generally chosen. In our series, 7 children have undergone unilateral or bilateral mandibular distraction with the MIDS. The distraction protocol was 1 mm per day, beginning on days 5 to 7. The consolidation period ranged from 2 to 3 months. One of the patients developed granulation tissue along the cable track. This should be obviated by second-generation devices that have removable cables.

Maxillary-LeFort I Distraction (Fig. 5): The LeFort I osteotomy is made as a step, leaving plenty of zygomaticomaxillary buttress available for attachment of the posterior plate. The anterior plate is stabilised along the superior border of the LeFort I, and the cable is brought through a stab wound in the hair, just above the sideburn. By cutting the posterior plate as an upside-down L, a portion of plate superior to the device attachment is made more accessible for screw placement. We have done LeFort I osteotomy with this technique in an 8-year-old girl with maxillary retrusion and bilateral cleft lip and palate. In younger patients, the posterior plate and device may need to be stabilised in the temporal region. Placement in these cases can be accomplished through temporal and upper buccal sulcus incisions. Orthodontic involvement is critical to successful maxillary distraction. In all cases, if possible, orthodontic appliances with surgical hooks are attached to the maxillary and mandibular dentition. At the conclusion of distraction, dental elastics are immediately applied to manipulate the regenerating callus and correct any minor occlusal discrepancies that may have occurred during sagittal and/or vertical advancements of the maxilla. Over the ensuing 6-week period, the dental elastics are gradually weaned, and the patient is allowed to consolidate for 2 to 3 months, depending on the patient's age. The patient then returns to the operating room, where the devices are removed. In children with cleft lip and palate, palatal expansion is carried out prior to, or simultaneous with, maxillary distraction. In addition, hooks are attached to bands on the maxillary molars to permit application of external reverse head gears, if necessary.

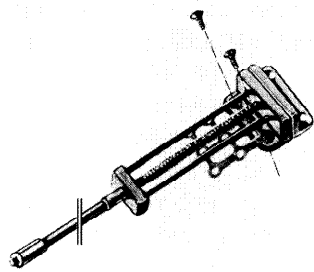


Fig. 4. Artist depiction of mandibular distraction utilising Modular Internal Distraction (MID) system developed by Cohen.

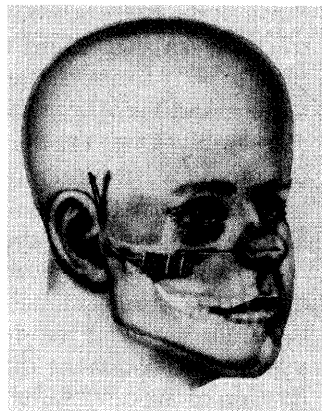


Fig. 5. LeFort I distraction technique, as depicted by artist, utilising Modular Internal Distraction system

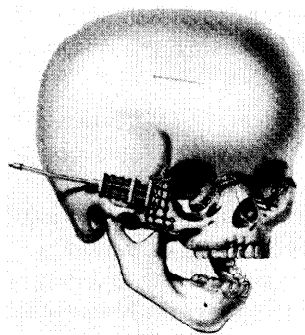


Fig. 6. LeFort III distraction utilising Modular Internal Distraction system.

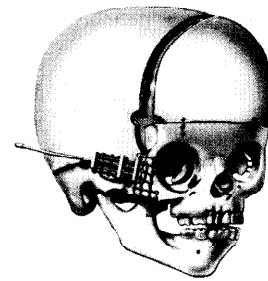


Fig. 7. Monobloc distraction technique utilising Modular Internal Distraction system developed by Cohen.

This can be done if device removal needs to be carried out earlier than originally anticipated. We have now carried out LeFort I distraction with the MIDS on 3 patients. In one patient, distraction was carried out for apnoea, which initially resolved but later recurred for unclear reasons. This was a child with cleidocranial dysostosis, who was substantially overweight, and a Pickwickian component may have played a role. Another patient with acondroplasia and severe midface retrusion is undergoing LeFort I distraction for apnoea. The last patient, an 8-year-old child with bilateral cleft deformity, underwent a successful 15-mm maxillary distraction. At the time of distraction device removal, alveolar bone grafts were carried out. The patient has been followed for 8 months without signs of relapse. It is anticipated, however, that mandibular growth will eventually outpace maxillary position, and the need for conventional orthognathic surgery may still be necessary. Whenever possible, overcorrection is carried out to anticipate the growing mandibular vectors. There were no complications in this group of 3 patients.

LeFort III Distraction (Fig. 6): A standard LeFort III osteotomy is performed. In younger children who are at risk for fracturing the zygomaticmaxillary suture region, the anterior plate is configured and rigidly fixed after the osteotomy, but before the downfracture. In this fashion, inadvertent fracture across the zygomaticmaxillary suture is prevented. The anterior plate wraps around the malar eminence and extends along the inferior orbital rim. The anterior plate is also attached along the lateral and superior orbital rim. When necessary, the plate can be cut and attached to the lateral aspects of the inferior orbital rim to provide increased retention. This, however, has not been necessary in the majority of our cases. The posterior plate is stabilised to the temporal bone underneath the temporal muscle. A 30-mm distraction frame is chosen and attached to the anterior and posterior plates. A flexible cable is brought through a stab wound posterior to the coronal incision. In LeFort

III osteotomy, typically 2 to 4 mm of distraction is performed in the operating room. Distraction is then begun on the postoperative days 5 to 7 at a rate of 1 mm per day. Once the appropriate porion-to-orbitale distance has been reached (approximately 70 to 72 mm) and/or mild enophthalmos has been produced by overcorrection of the malocclusion, distraction is stopped and the distraction gap allowed to consolidate over a period of 2 to 3 months. Lateral and posteroanterior cephalograms are taken to ensure that the distraction devices have opened symmetrically. At the conclusion of distraction, dental elastics are placed to fine tune the occlusion. After consolidation, patients are returned to the operating room, where the devices are removed. The secondary procedure enables the surgeon to correct any residual craniofacial abnormality, such as canthal dystopia or minor facial asymmetries. Four patients have undergone LeFort III osteotomies in our series. The only complication has been related to a small granuloma around the cable track. Stability has been excellent, and the LeFort III segment has come forward without any rotational abnormalities. It is important, however, to make sure the osteotomy is complete and the LeFort III segment is well mobilised.

Monobloc and Facial Bipartition Distraction (Fig. 7): In 1971, Tessier²⁹ was first to describe a single-stage frontofacial advancement in which the orbitofrontal bar was advanced as a separate element, but in conjunction with the LeFort III complex and frontal bone. Seven years later, Ortiz-Monasterio et al³⁰ developed the monobloc to advance the orbits and midface as one unit. In 1979, van der Meulen³¹ described the medial fasciotomy for correction of midline facial clefting. By splitting the monobloc osteotomy vertically in the midline, he moved the two halves of the face together to correct orbital hypertelorbitism. Tessier³² and Ortiz-Monasterio³³ went on to refine the vertical splitting and reshaping of the monobloc segment, thus correcting the midface deformity in three dimensions in a procedure

now known as facial bipartition. Wolfe et al,³⁴ Tessier³⁵ and Kawamoto, while acknowledging the potential risks of the monobloc procedure, continued advocating its use in selected patients since it provides superior results to those obtained with subcranial procedures. Clearly, the monobloc frontofacial advancement, with or without facial bipartition, is not without its problems. Epidural infection, prolonged postoperative ventilation, cerebral spinal fluid leaks, and major haemorrhage can all occur with the monobloc osteotomy. The rationale behind monobloc and facial bipartition distraction is as follows:

- 1) gradual advancement of the monobloc segment is not associated with a substantial retrorobust dead space and probably decreases infectious complications;
- 2) since 5 to 7 days are elapsed before distraction, the nasofrontal opening, in theory, is allowed to remucosalise;
- 3) gradual expansion of the soft tissues takes advantage of creep with gradual skin and subcutaneous tissue relaxation, thereby potentially reducing relapse and permitting greater advancement;
- 4) the procedure appears to be less invasive with decreased blood loss and less operative time, thereby enabling safe application in infants;
- 5) overdistracted of the monobloc complex may eliminate or reduce the frequency of subsequent procedures;
- 6) because of the broad anterior plate attachment, rotation of the monobloc segment with occlusal retraction does not occur; and
- 7) the procedure may be combined with facial bipartitioning and skull vault remodelling, as well as other segmental procedures.

The monobloc distraction is carried out through a

coronal incision. Monobloc osteotomy is performed in a conventional fashion. It is critical that a complete osteotomy be made, and the entire monobloc segment must be mobile prior to activation of distraction devices. As the monobloc is gently rocked forward with Rowe-Killey disimpaction forceps and /or Smith's separators, the vomer and septum are cut with a small osteotome. It is necessary to check both the frontozygomatic and the zygomaticomaxillary sutures. If these are patent or at risk of fracture with mobilisation, the anterior plate is rigidly fixed before the downfracture, securing the sutures. The vector of distraction can be varied from direct horizontal to oblique, depending on the need for both sagittal and vertical advancement. Once the segment is downfractured, a facial bipartition, if necessary, can be carried out and the midline rigidly stabilised with titanium plates and screws. Following this stabilisation, distraction devices are applied. In the 5 patients who have undergone monobloc distraction with or without facial bipartition, there has not been a single epidural infection or cerebral spinal fluid leak. In one patient, a small cable track infection necessitated removal of the devices 2 weeks earlier than originally planned. The longest operative time was the first monobloc distraction (337 minutes). Hospital days ranged from 5 to 12 days, and patients spent a mean of 2.5 days in the intensive care unit. All patients were extubated within 24 hours after the monobloc advancement. Facial swelling was markedly less than with standard monobloc advancement, and recovery in general was much quicker. Perioperative antibiotics were used, and patients were discharged home with oral antibiotics for 7 to 10 days.

Other Types of Distraction

Cranial: Personal communications from Vander Kolk and Patel have confirmed the efficacy of distraction of the cranium. Vander Kolk at Johns Hopkins utilised the

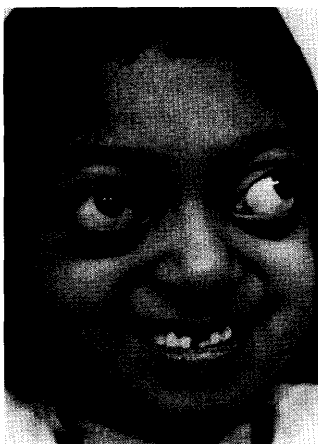


Fig. 8a.



Fig. 8b.



Fig. 8c.



Fig. 8d.

Fig. 8. (a) Six-year-old girl with Crouzon's syndrome and severe midface retrusion. (b) Postoperative photograph following removal of distraction devices. (c) Preoperative lateral photograph. (d) Postoperative lateral photograph following 30 mm of distraction. Patient purposely distracted into Class II malocclusion in anticipation of lateral mandibular growth.



Fig. 9a.



Fig. 9b.



Fig. 9c.



Fig. 9d.

Fig. 9. (a) Preoperative frontal photograph of a y-month-old girl with Pfeiffer's syndrome. (b) Postoperative photograph of same child 3.5 months later, after removal of distraction devices. The patient underwent 28 mm of distraction. (c) Preoperative lateral photograph. (d) Postoperative lateral photograph 3.5 months following removal of devices (Published with permission from: Cohen S R, Boydston W, Burstein F D, Hudgins R. Monobloc distraction osteogenesis during infancy: report of a case and presentation of a new device. *Plast Reconstr Surg* 1998; 101:1919-24.)



Fig. 10a.



Fig. 10b.

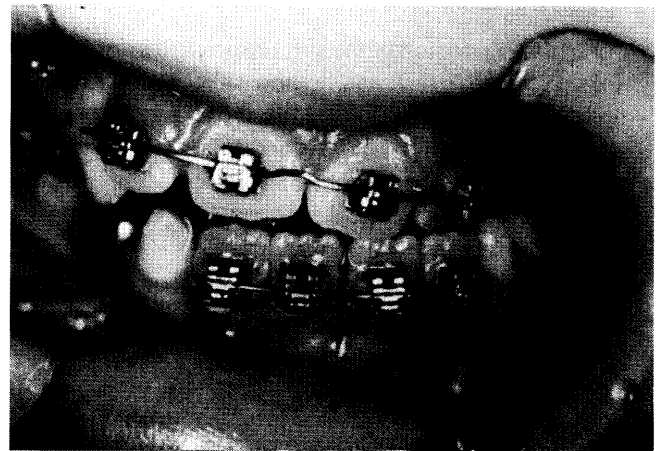


Fig. 10c.

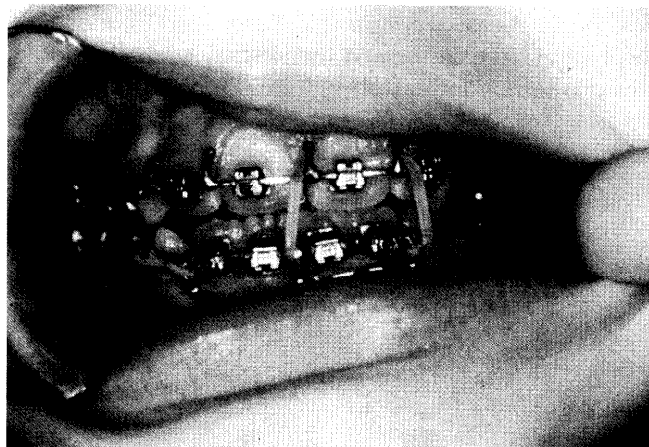


Fig. 10d.

Fig. 10. Seven-year-old girl with neurofibromatosis, left ramal shortening with facial asymmetry, and infiltration of left upper eyelid. (a) Preoperative frontal photograph. (b) Postoperative frontal photograph following 15 mm of left mandibular distraction. (c) Preoperative occlusion. (d) Post distraction occlusion.

MIDS for cranial vault expansion in several patients with slit ventricular syndrome. Patel used the distraction devices in an open fashion, thus producing compression to gradually lower the skull vault in a child with severe turriccephaly and Apert syndrome.

Alveolar: Alveolar devices developed by Chin (personal communication) are now available to distract short alveolar segments, producing increased bone height and expansion of the soft tissue gingival envelope.

Zygomatic: Seigal (personal communication) at the University of Florida has carried out isolated zygomatic distraction in a child with Nagers syndrome.

Case Examples

Case One

NM is a 6-year-old girl with Crouzon's syndrome who underwent 22 mm of LeFort III distraction (Fig. 8). A monobloc osteotomy would have been preferable, but because of enormous emissary veins, the craniotomy could not be safely performed. After 22 mm of horizontal midface distraction, the patient was overcorrected into a

Class II malocclusion in anticipation of later mandibular growth. Two months were allowed to elapse for bony consolidation, and the devices were removed. At the time of device removal, hydroxyapatite cement was employed to augment the frontal bone. She is now 9 months postoperative without evidence of relapse.

Case Two

CF is an 8-month-old girl with Pfeiffer's syndrome (Fig. 9). A monobloc distraction to treat exorbitism with corneal exposure was performed. Bilateral 30-mm distractors were placed. On postoperative day 5, distraction was begun, for a total of 28 mm. After 2.5 months of consolidation, the devices were removed. The child underwent repeat monobloc distraction at age 4 years, again without problems.

Case Three

EK is a 7-year-old girl with neurofibromatosis of the left upper eyelid and significant left mandibular ramal hypoplasia (Fig. 10). Fifteen millimetres of mandibular distraction along an oblique vector was carried out, and a left orbitotomy was performed with upper eyelid debulking and resuspension of the levator apparatus. Orthodontic appliances were placed prior to distraction, and dental elastics were utilised at the conclusion of distraction to manipulate the bony regenerate, fine tuning the occlusion.

Conclusion

Distraction osteogenesis has become an important tool in craniofacial surgery. Its application in the treatment of sleep apnoea and upper airway obstruction has revolutionised the care of these children. It is quite conceivable that as more experience, and thus more confidence, is gained, distraction will eliminate tracheostomy in the majority of cases of acute neonatal and early childhood airway obstruction. Distraction has also played an important role in the treatment of sleep apnoea in children younger than 12 years of age. In patients undergoing LeFort I advancement, excellent options exist for both internal and external devices. It is important to present to the patient several options. Ultimately, we let the family decide the best route after being informed of the relative pros and cons of all techniques. LeFort III advancement in younger children and, for that matter, in adults with severe degrees of midface retrusion are excellent candidates for distraction. Distraction osteogenesis combined with monobloc and facial bipartition osteotomies appear to reduce the infection rate, making this operation safer to perform. The MIDS represents a system that can be utilised throughout the craniofacial skeleton for virtually any site, any shape, and almost any age group. Improving the design and shrinking the size of the distraction frame, along with providing a remov-

able cable and easier device removal, will enhance the MIDS, making it applicable to virtually any situation.

REFERENCES

1. Ilizarov G A. A new principle of osteosynthesis with the use of crossing pins and rings. In: Collection of Scientific Works of the Kurgan Regional Scientific Medical Society. USSR Kurgan, 1954:145-60.
2. Synder C C, Levine G A, Swanson H M, Browne E Z. Mandibular lengthening by gradual distraction: Preliminary report. *Plast Reconstr Surg* 1973; 51:506.
3. Karp N S, Thorne C H M, McCarthy J G, Sissons H A. Bone lengthening in the craniofacial skeleton. *Ann Plast Surg* 1990; 24:231.
4. McCarthy J G, Schreiber J, Karp N, Thorne C H, Grayson B H. Lengthening the human mandible by gradual distraction. *Plast Reconstr Surg* 1992; 89:1.
5. Persing J A, Morgan E I, Cronin A J, Wolcott W I. Skull base expansion: Craniofacial effects. *Plast Reconstr Surg* 1991; 87:1028.
6. Barone C M, Ferder M, Jimenez D F, Grossman L, Hall C, Strauch B, et al. Distraction of the frontal bone outside the cranial plane: A rabbit model. *J Craniofac Surg* 1993; 4:177.
7. Remmler D, McCoy F J, O'Neil D, Willoughby L, Patterson B, Jerald I C, et al. Osseous expansion of the cranial vault by craniostasis. *Plast Reconstr Surg* 1992; 89:787.
8. Rachmiel A, Potparic I, Jackson I T, Sugihara T, Clayman L, Topf J S, et al. Midface advancement by gradual distraction. *Br J Plast Surg* 1993; 46:201.
9. Cohen S R, Ross D A, Burstein F B, LeFaivre J F, Riski J E, Simms C. Skeletal expansion combined with soft-tissue reduction in the treatment of obstructive sleep apnea in children: Physiologic results. *Otolaryngol Head Neck Surg* 1998; 119:476.
10. Burstein F, Cohen S, et al. Surgical therapy for severe refractory sleep apnea in infants and children: Application of the airway zone concept. *Plast Reconstr Surg* 1995; 96:34.
11. Riley R W, Powell N B, Guilleminault C. Inferior sagittal osteotomy of the mandible with hyoid myotomy-suspension. A new procedure for sleep apnea. *Otolaryngol Head Neck Surg* 1986; 94:589-93.
12. Riley R W, Powell N B, Guilleminault C. Current surgical concepts for treating obstructive sleep apnea syndrome. *J Oral Maxillofac Surg* 1987; 45:149-57.
13. Cohen S R, Simms C, Burstein F D. Mandibular distraction osteogenesis in the treatment of upper airway obstruction in children with craniofacial deformities. *Plast Reconstr Surg* 1998; 101:312-8.
14. McCarthy J G. Craniofacial microsomia. In: Cohen S R, editor. *Clinics in Plastic Surgery*. Philadelphia, PA: WB Saunders Co, 1997:459-74.
15. Mulliken JB, Kaban LB. Analysis and treatment of hemifacial microsomia in childhood. *Clin Plast Surg* 1987; 14:91.
16. Pruzansky S. Not all dwarfed mandibles are alike. *Birth Defects* 1969; 5:120.
17. McCormack S V, Grayson B H, McCarthy J G, Staffenberg D A. The effect of gradual distraction of the mandible on the temporomandibular joint. Part II: Clinical study. *J Craniofac Surg* 1955; 6:364.
18. Cohen S R, Rutrick R E, Burstein F D. Distraction osteogenesis of the human craniofacial skeleton: Initial experience with a new distraction system. *J Craniofac Surg* 1995; 368-74.
19. Muhlbauer W. Distraction osteogenesis in the treatment of Apert's syndrome in infancy. In: Harii K, editor. *Transactions of the Scientific Meeting of the International Plastic, Reconstructive and Aesthetic Surgery Society*. Yokohama, Japan: Kugler Publications, 1995.
20. Chin M, Toth B A. Distraction osteogenesis in maxillofacial surgery using internal devices: Review of five cases. *J Oral Maxillofac Surg* 1996; 54:45-53.
21. Polley J W, Figueroa A A, Charbel F T, Berkowitz R, Reisberg D, Cohen M. Monobloc craniomaxillofacial distraction in a newborn with severe craniofacial synostosis: A preliminary report. *J Craniofac Surg* 1995; 6:421-3.

22. Cohen S R, Burstein F D, Stewart M B, Rathburn M A. Maxillary-midface distraction in children with cleft lip and palate: A preliminary report. *Plast Reconstr Surg* 1997; 99:1421-8.
23. Cohen S R, Boydston W, Burstein F D, Hudgins R. Monobloc distraction osteogenesis during infancy: Report of a case and presentation of a new device. *Plast Reconstr Surg* 1998; 101:1919-24.
24. Cohen S R, Boydston W, Hudgins R, Burstein F D. Monobloc and facial bipartition distraction with internal devices. *J Craniofac Surg* 1999; 10:244.
25. Polley J W, Figueroa A A. Management of severe maxillary deficiency in childhood and adolescence through distraction osteogenesis with an external adjustable, rigid distraction device. *J Craniofac Surg* 1997; 181-5.
26. Ortiz-Monasterio F, Molina F, Andrade L, et al. Simultaneous mandibular and maxillary distraction in hemifacial microsomia in adults: Avoiding occlusal disasters. *Plast Reconstr Surg* 1997; 100:852-61.
27. Chin M, Toth B A. LeFort III advancement with gradual distraction using internal devices. *Plast Reconstr Surg* 1997; 100:819-30.
28. Cohen S. Craniofacial distraction with a modular distraction system: Evolution of design and surgical techniques. *Plast Reconstr Surg* 1999; 103:1592-607.
29. Tessier P. The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis: Crouzon's and Apert's diseases. *Plast Reconstr Surg* 1971; 48:419.
30. Ortiz-Monasterio F, Fuente del Campo A, Carillo A. Advancement of the orbits and midface in one piece, combined with frontal repositioning, for the correction of Crouzon's deformity. *Plast Reconstr Surg* 1978; 61:507.
31. van derMeulen J C. Medial fasciotomy. *Br J Plast Surg* 1979; 32:339.
32. Tessier P. Apert's syndrome: acrocephalosyndactyly Type I. In: Caronni E I', editor. *Craniofacial Surgery*. Boston, MA: Little Brown, 1985:280-303.
33. Ortiz-Monasterio F, Fuente del Campo A. Refinements on the bloc orbitofacial advancement. In: Caronni E, editor. *Craniofacial Surgery*. Boston, MA: Little Brown, 1985:263-74.
34. Wolfe S A, Morrison G, Page L I C, Berkowitz S. The monobloc frontofacial advancement: do the pluses outweigh the minuses? *Plast Reconstr Surg* 1993; 91:977.
35. Tessier P. Discussion of the monobloc frontofacial advancement: do the pluses outweigh the minuses? *Plast Reconstr Surg* 1993; 91:988.