# Distraction Osteogenesis

# 34

Raymond Harshbarger and Patrick Kelley

Distraction osteogenesis (DO) of the craniomaxillofacial skeleton has been an adaptation of the original studies on lengthening posttraumatic long bone injuries. Using the principles elucidated by Ilizarov, and building on the work of McCarthy, the scope of DO in the head and neck region has expanded dramatically. Capitalizing on the extensive body of work by orthopedic surgeons in fracture repair, those interested in the congenital and acquired conditions of the head and neck have been able to adapt the technique for bone lengthening throughout the craniofacial skeleton. Using the concepts of tension, stress, and blood supply, the process of distraction osteogenesis is now well anchored in the armamentarium of craniofacial surgeons. Deficiencies of soft tissue, scarring, and limited bone stock can now be overcome by gradual lengthening of the soft callus, along a vector with semirigid fixation; scarless bone formation can lend to the improvement of airway/breathing, masticatory function, eye protection, brain function, and finally craniofacial form.

## History

The first recorded attempts at manipulating bone segments for the purpose of elongation dates back to the time of Hippocrates. External traction devices were used in the posttraumatic axial skeleton. In the early 1800s, continuous traction was used in posttraumatic extremities with the introduction of a formal osteotomy. Codivilla, in 1905, further refined these earlier processes reporting femur elongation at the site of former trauma through a process of external traction, using an osteotomy. Russian orthopedic surgeon, Gavril Ilizarov, in the 1950s developed techniques for treating posttraumatic tibial defects which form the scientific basis for modern-day process of distraction osteogenesis. Ilizarov's observations included use of "mechanical tension," a key signal during natural bone growth, and the relationship of blood supply and loading on maintenance of bone. Through Ilizarov's work, the basic principles of distraction osteogenesis were formalized, including osteotomy, latency, and rate/rhythm of distraction. Distraction has since been adapted for use in other regions of the body including the craniofacial skeleton. During the nineteenth and twentieth centuries, early attempts at bone lengthening in the craniofacial skeleton centered on mandibular lengthening. Various osteotomies, combined with acute advancements, and limited ability for skeletal fixation tended to result in variable bone formation, partial relapse, and overall instability (see Fig. 34.1). From 1970 to 1991 investigators began to apply Ilizarov's techniques to the canine mandible. Following this, McCarthy reported the first use of distraction osteogenesis in the human craniofacial skeleton, by successfully lengthening the mandible in a child with craniofacial microsomia (see Fig. 34.2). Since that time, use of DO in the bones of the face and skull has increased dramatically. Currently, DO has been described in the cranium, orbits, midface, and mandible for use in a variety of conditions both congenital and acquired.

## Biology and Biomechanics

The principles of bone formation in distraction osteogenesis mirror that of basic fracture repair. Steps to heal a fracture include (1) traumatic impact to bone, (2) induction/inflammation, (3) callus formation, and (4) remodeling (see Table 34.1). After fracture, a hematoma consolidates and is replaced with vascular proliferation, inflammatory cells, and fibroblasts, with recruitment of osteoprogenitor cells from the periosteum. Over time extensive capillary ingrowth occurs with formation of a soft callus made of fibrous tissue,

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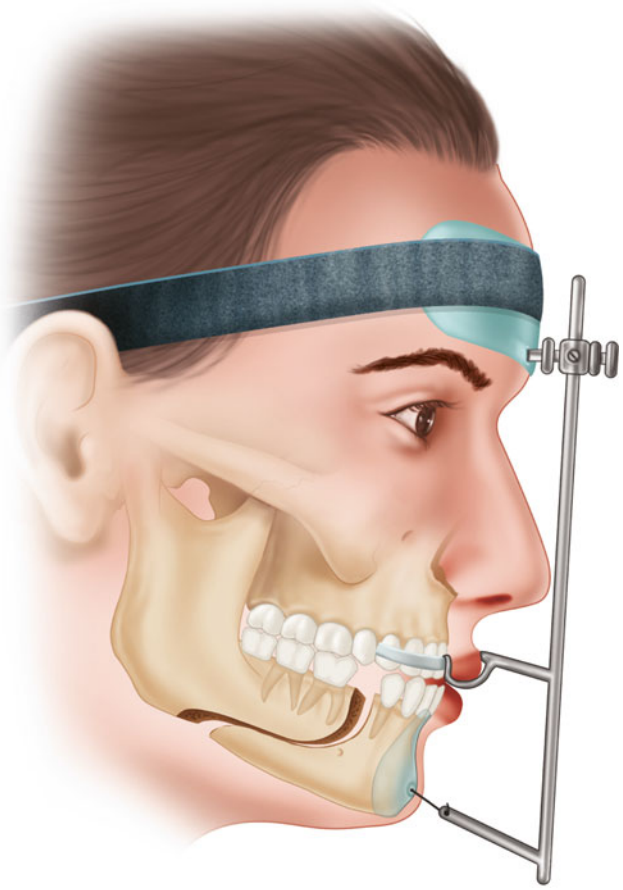


**Fig. 34.1** Kasanjian's external "over the face" mandible traction device

the result of the presence of osteoprogenitor cells signaled by factors released at the time of the fracture. Eventually, the soft callus is replaced by hard callus as osteoblasts induce mineralization of bone. Following this, remodeling occurs. The formal stages of DO include (1) osteotomy with application of distraction device, (2) latency, (3) distraction, (4) consolidation, and (5) remodeling (see Fig. 34.3). After osteotomy (instead of traumatic fracture), a latency period ensues, essentially to allow the formation of a soft callus. Through the work of Ilizarov, latency was established at about 1 week after osteotomy. This principle, however, was founded on long bones, with endochondral bone formation, in a posttraumatic state. Through experiments on the canine mandible, and McCarthy's early work in children, the concept of latency of 7 days was transferred for use in the craniofacial skeleton. Formal studies regarding the latency period of various regions of the craniofacial skeleton have not been reported. Alterations in the latency period may be done based on the age of the patient, i.e., less time in younger patients (neonatal) and longer in older patients (adults). Latency must be gauged properly, given that too short a waiting period before active distraction would result in a fibrous distraction versus too long a latency with premature bony consolidation.

A successful active distraction phase relies on the presence of a soft callus, which can be slowly elongated with traction forces to achieve bone lengthening. Within the distraction phase, rate and rhythm of distraction must be established. By convention, with Ilizarov's pioneering work as a foundation, a standard rate of 1 mm/day was determined

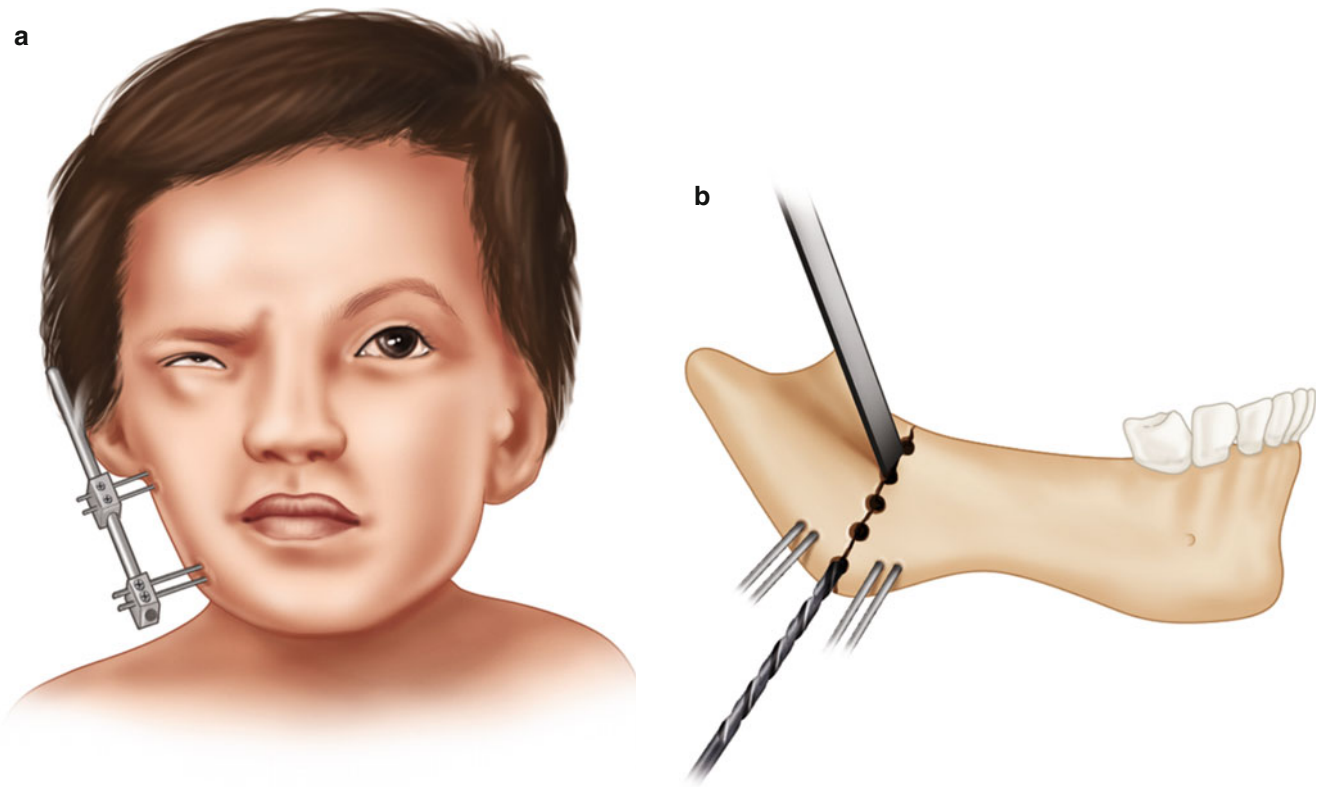



**Fig. 34.2** (**a**) McCarthy's patient with hemifacial microsomia undergoing external mandibular distraction with a Hoffman device (Stryker Leibinger, Kalamazoo, MI). (**b**) Schematic showing placement of osteotomy and external traction pins

**Table 34.1** Distraction osteogenesis impacts the soft callus phase of typical fracture healing

| Fracture healing |
|---|
| Impact |
| Induction |
| Inflammation |
| Soft callus |
| Gradual traction (distraction) |
| Hard callus |
| Remodeling |

to produce bone in the distracted gap. Rhythm refers to the frequency of activation (turning) of the device. Many protocols include a rhythm of twice daily activation. Both the rate and rhythm of distraction may vary depending on the age and region to be distracted. Neonatal mandibular distraction may succeed using 2 mm/day, with twice daily device activation, while cranial distraction in the adult may require 0.5 mm/day with once daily activation. After the active distraction phase is completed, consolidation begins. The hallmark of this phase, in which the devices remain for stability, is a transition

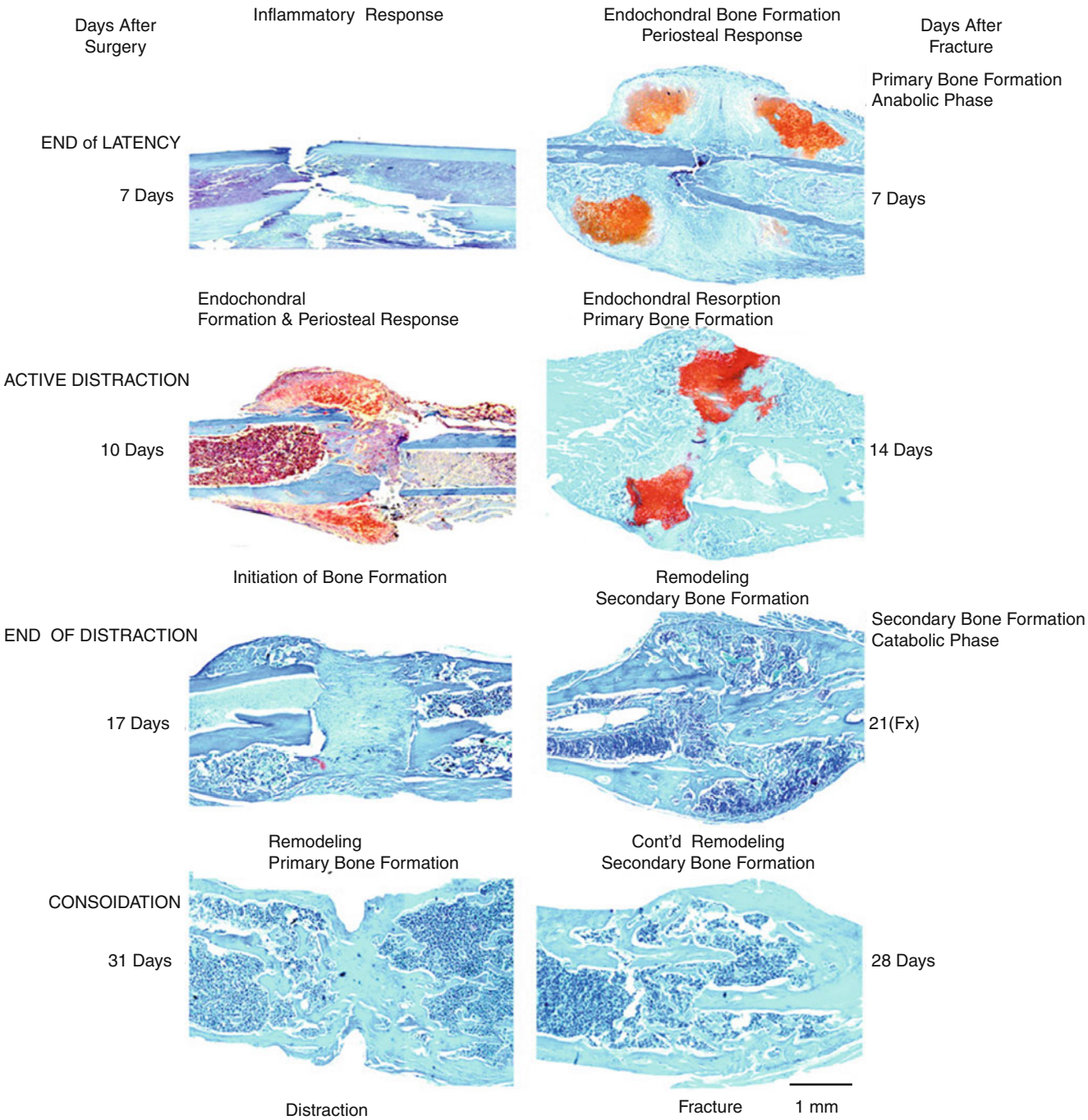



**Fig. 34.3** Histologic comparison of healing by distraction osteogenesis (*left*) and healing after fracture (*Right*)

of the fibrous interzone into mineralized bone. Ossification occurs in parallel to the distraction vector. Experimental studies in a rat model have shown the presence of both intramembranous and endochondral bone formation in a distracted mandible. In craniofacial DO cases which involve occlusion, devices may be removed prior to complete ossification to allow for molding of the generate and a more ideal bite relationship. The time to ossification and device removal is not precise, but generally 8 weeks after cessation of active distraction, stable bone has formed, range 6–12 weeks. Again, neonates form mandibular bone faster perhaps in the 4–6-week range, while adults may require longer periods of consolidation. Long-term remodeling of the ossified generate may occur with strengthening of the initial bony scaffold by addition of parallel-fibered lamellar bone.

For most of the indications of DO in the craniofacial skeleton, simple DO would be performed with a single osteotomy between two vascularized regions of bone and lengthening occurring as the segments are moved apart. Occasionally, transport DO is required. Here, an osteotomized segment of bone is transported across a bone gap. For example, transport of a mandibular segment for reconstruction of the ramus/condyle, or across an acquired gap from trauma or cancer resection. Also important to successful outcomes in DO are the vectors of distraction which are vertical, horizontal, and oblique.

## Distraction Osteogenesis of the Mandible

The proof of concept of craniofacial DO began experimental animal models and was first shown in humans clinically in a patient with craniofacial microsomia. After McCarthy showed success in lengthening the human mandible through DO, this concept has been applied to a variety of both congenital and acquired conditions involving mandibular deficiencies.

### Neonate

The indications for DO of the mandible are numerous and relate to both the age of the patient and particular condition being treated. In the neonatal population, micrognathia with tongue-based airway obstruction may prompt bilateral mandibular DO to prevent tracheostomy in an acutely unstable airway or to reverse severe obstructive sleep apnea, often accompanied by feeding difficulties, gastroesophageal reflux, and general failure to thrive (see Table 34.2). Most frequently, this would be encountered in the patient with Pierre Robin sequence but can be seen in a variety of conditions including Treacher Collins syndrome, Nager syndrome, and craniofacial microsomia. If micrognathia with an unstable airway (unable to undergo polysomnography) is detected in the neonate, operative airway evaluation is indicated to confirm the tongue-based airway obstruction and rule out any other airway anomalies. Bilateral mandibular osteotomies with distractor placement are then performed to prevent a tracheostomy. Neonates, who are stable enough to undergo polysomnography, should do so, and if severe obstructive sleep apnea is detected, they may benefit from mandibular DO to prevent the long-term effects of sleep apnea on the developing brain and to avoid failure to thrive.

**Table 34.2** Evaluation of neonate with micrognathia

| Neonatal evaluation |
|---|
| Failure of proper respiratory function |
| Neonatal sleep study (polysomnography) |
| Feeding evaluation |
| Genetics |
| MRI (for central apnea) |
| Muscular tone |
| Airway evaluation (direct laryngo-bronchial exam) |
| 3D CT scan |

The technique for neonatal DO involves exposure of the lateral mandibular border through a small submandibular incision. In the neonate, the marginal mandibular nerve is typically located along the mandibular border, so an incision 1.5–2 cm below the border should help to avoid this singular structure. Dissection is carried through the investing fascia of the submandibular gland, and then cranially, to the mandibular border. Subperiosteal exposure is undertaken, visualization of the border, angle, ramus, sigmoid notch, coronoid process, and condylar process is important to correctly position the osteotomy. Our preference is for the inverted L osteotomy, beginning on the anterior border of the ramus above the lingula extending horizontally and turning vertically posterior to the nerve and parallel to the posterior ramus down to the border. Sufficient space should be left on the proximal segment of the bone for placement of the footplate. This will avoid injury to tooth buds and to the inferior alveolar nerve. A sagittal saw is used (smaller kerf) to complete 90 % of the osteotomy. A microdistractor with ratchet mechanism (to prevent turning the wrong way) is placed across the osteotomy (see Fig. 34.4). The device is planned so that the turning handle is brought out anteriorly through the incision or posteriorly behind the ear through a separate stab incision. High profile 5 mm screws are placed in each footplate on either side of the osteotomy. The vector of distraction is typically horizontal to slightly inferior in the Pierre Robin patient (see Fig. 34.5a, b) but may be much more vertical in patients without much ramus, i.e., Treacher Collins syndrome. And in those cases, the osteotomy may be modified to a stairstep to allow the device to orient vertically across the posterior aspect of the osteotomy. After device fixation, the remaining 10 % of the anterior osteotomy on the ramus is completed with a micro-osteotome. The turning arm is attached to the device, and it is tested, and then returned to zero position.

Neonates will heal much faster than children or adults, so their overall course of DO is shorter. Latency is typically one night, with an accelerated rate/rhythm of 0.9 mm twice daily.

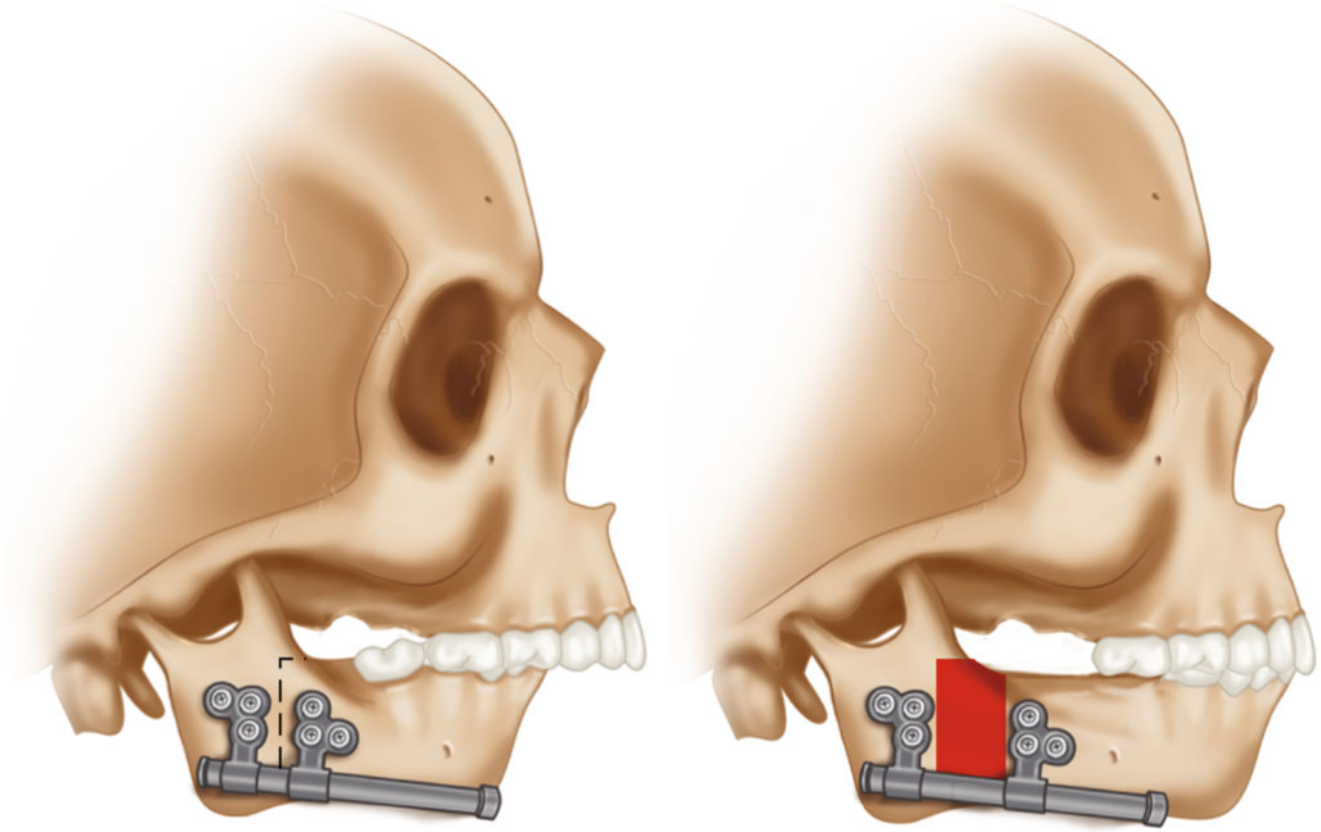


**Fig. 34.4** Schematic showing placement of inverted L osteotomy and internal distractor. *Red* represents distractate

In 7–14 days, distraction is completed. Overcorrection is sought with the patient in a class III skeletal relationship at the end of distraction (mainly for airway improvement). Patients may be kept intubated for the first few days after device placement to allow for diminished swelling and increased airway dimension through distraction. Extubation occurs in the OR with nasopharyngoscopy to judge improved airway space. Oral feeding can begin within a day or two after extubation. Consolidation is typically 6 weeks. Postdistraction polysomnography is conducted 2 months after distraction. Limited evidence exists as to the long-term growth of distracted neonatal mandibles.

## Childhood

Older children may also benefit from mandibular DO. Some are tracheostomy dependent patients from infancy with micrognathia. Others include micrognathic patients without a tracheostomy but with evidence of severe obstructive sleep apnea on polysomnography (see Fig. 34.6) not tolerating CPAP, or craniofacial microsomia patients with severe mandibular occlusal cants. In evaluating these patients, CT scans are helpful for surgical planning. With all mandibular distraction candidates, the surgeon must ensure that there is sufficient bone stock to place a distraction device. A severity level beyond the Pruzansky 2b category is typically not a distraction candidate. Using a 3D CT scan, the planned osteotomy, distraction vector, and result can be simulated with modern software techniques (see Figs. 34.7 and 34.8).

As with the neonate, mandibular osteotomies in a child must avoid tooth buds and the inferior alveolar nerve. A similar surgical approach is undertaken to access the mandible, except that the submandibular incision should be made lower on the neck. With growth the marginal mandibular nerve migrates inferior to the mandibular border. In children, we also prefer the inverted L osteotomy. Given the greater distance between the Risdon incision and the anterior ramus, an intraoral incision can be used to access the anterior ramus and to visualize the lingua (entrance of the inferior alveolar nerve). When using VSP, footplate and osteotomy guides can be used to precisely execute the bone cuts and position the internal distractors to achieve the desired vector. Internal devices are preferred by many but external devices using pins on either side of the osteotomy can also be used, as was favored early on. The latter offer the theoretical advantages of less dissection, which has not been shown that this would lead to better ossification of the generate. They can be used in a multiplanar fashion and in complex anomalies, although controlling multiple vectors can be difficult. Less desirable aspects of the external devices would include pin loosening and pin track scars on the face (see Fig. 34.9).

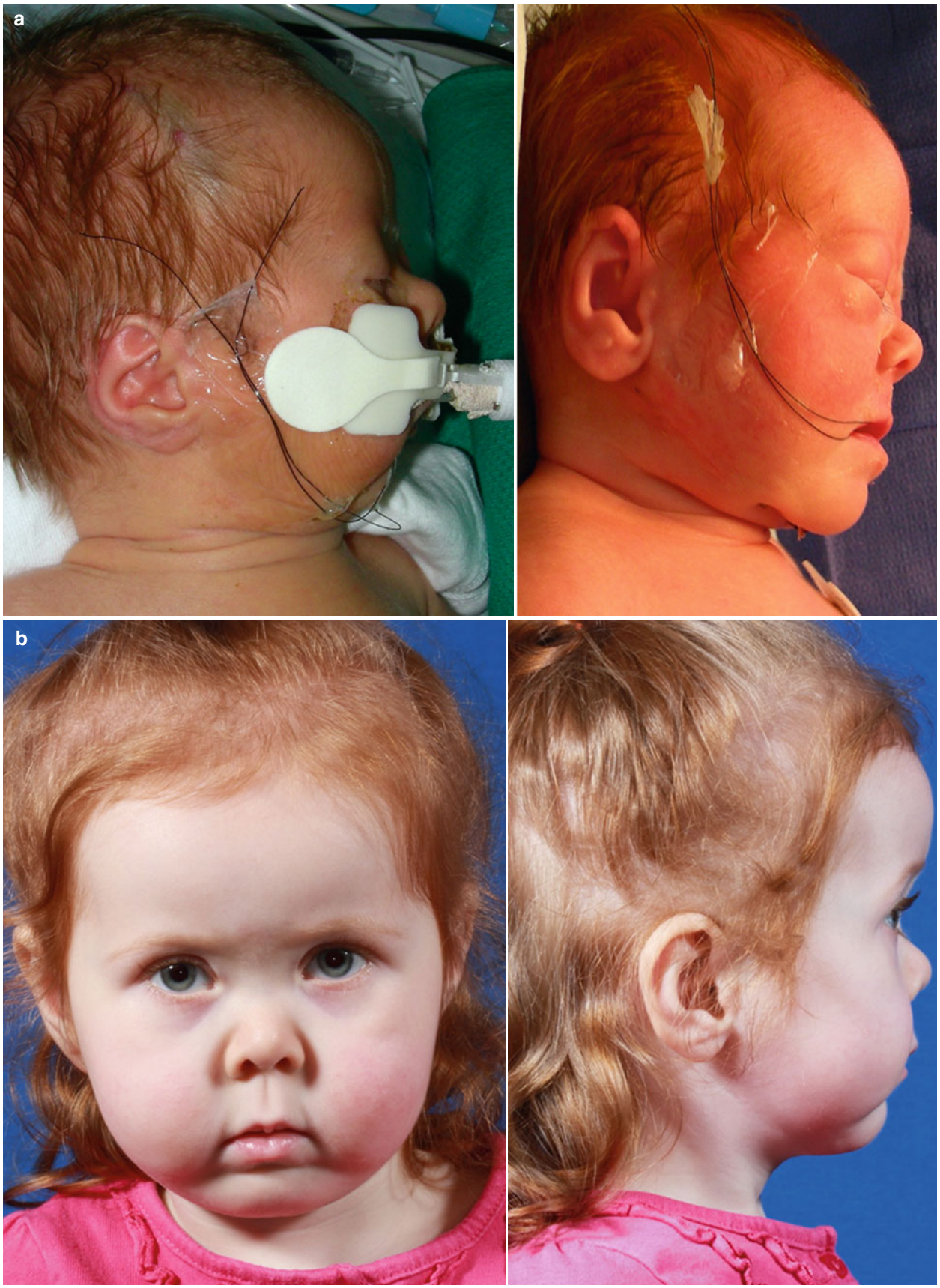



**Fig. 34.5** (**a**) Six day old infant with Pierre Robin Sequence prior to mandibular osteotomies and device placement (*Left*), and 1 week later, immediately after extubation upon completion of 14 mm of horizontal mandibular distraction. (*Right*). (**b**) 22 months after bilateral mandibular distraction osteogenesis

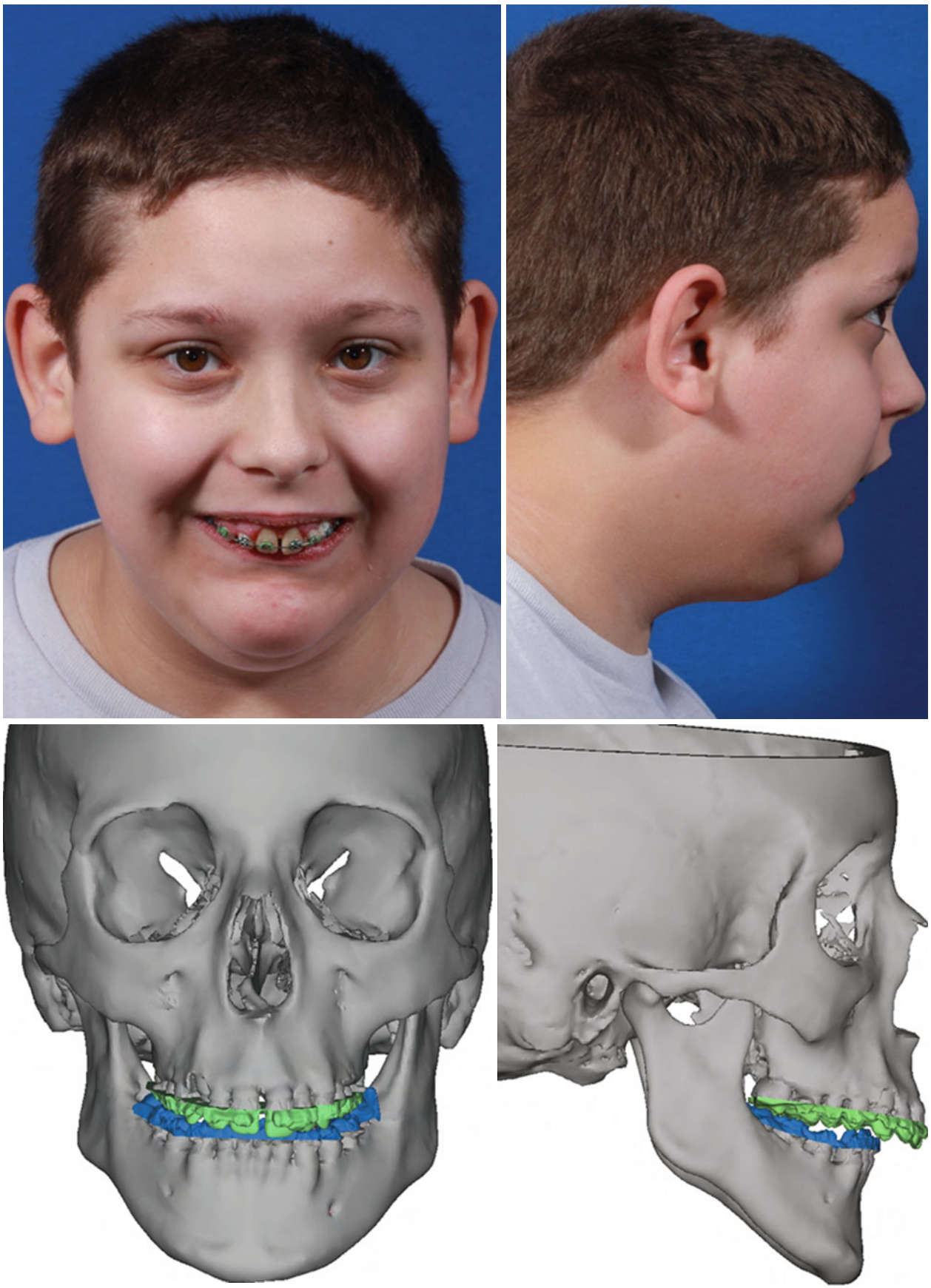


**Fig. 34.6** A 9-year-old boy with micrognathia, severe obstructive sleep apnea, and drooling

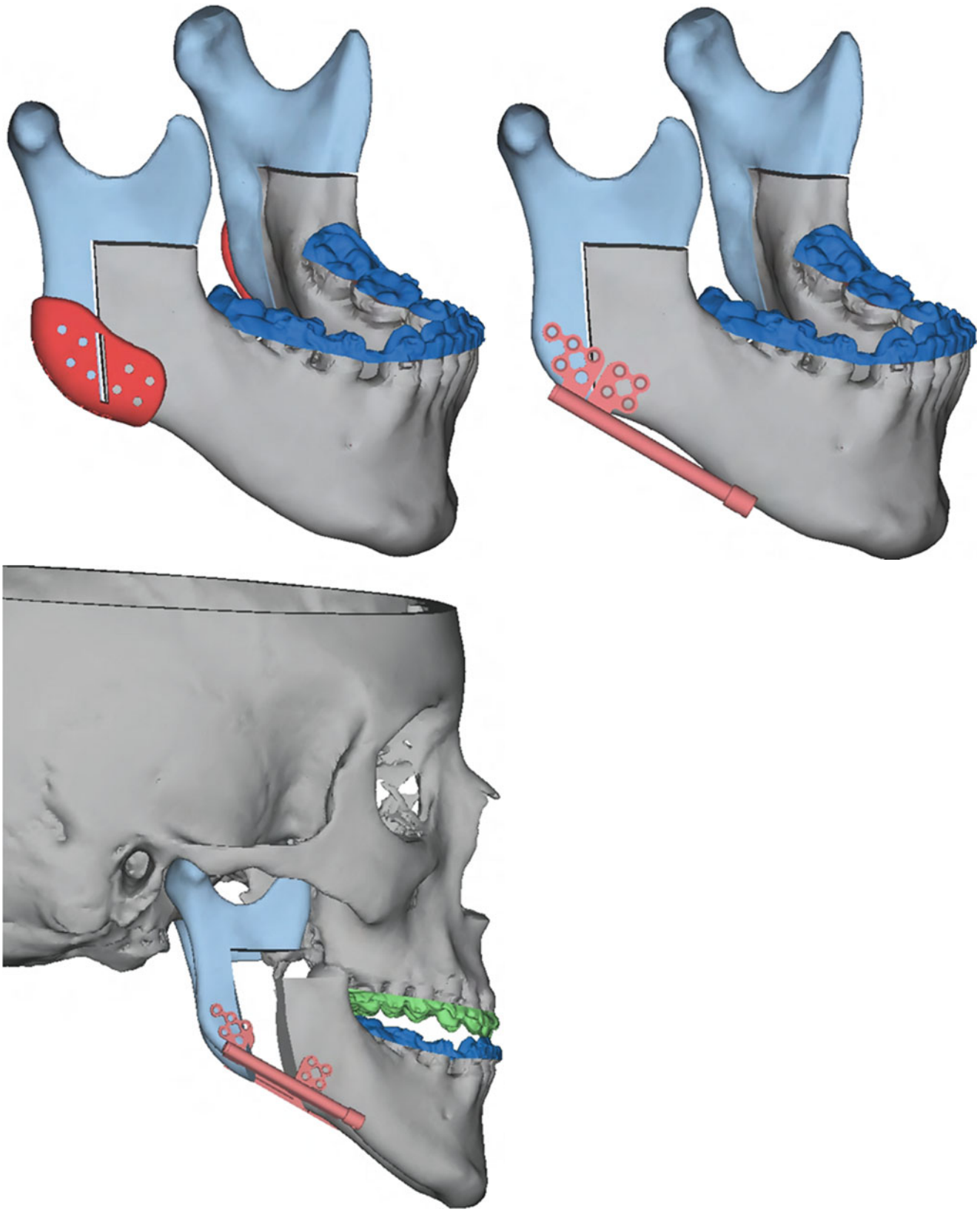


**Fig. 34.7** Preoperative CT showing plan for inverted L osteotomy, device footplate guides, device positioning with vector planning, and below, computer simulation of completed distraction (expected mm of distraction) (Medical Modeling, Golden, CO)

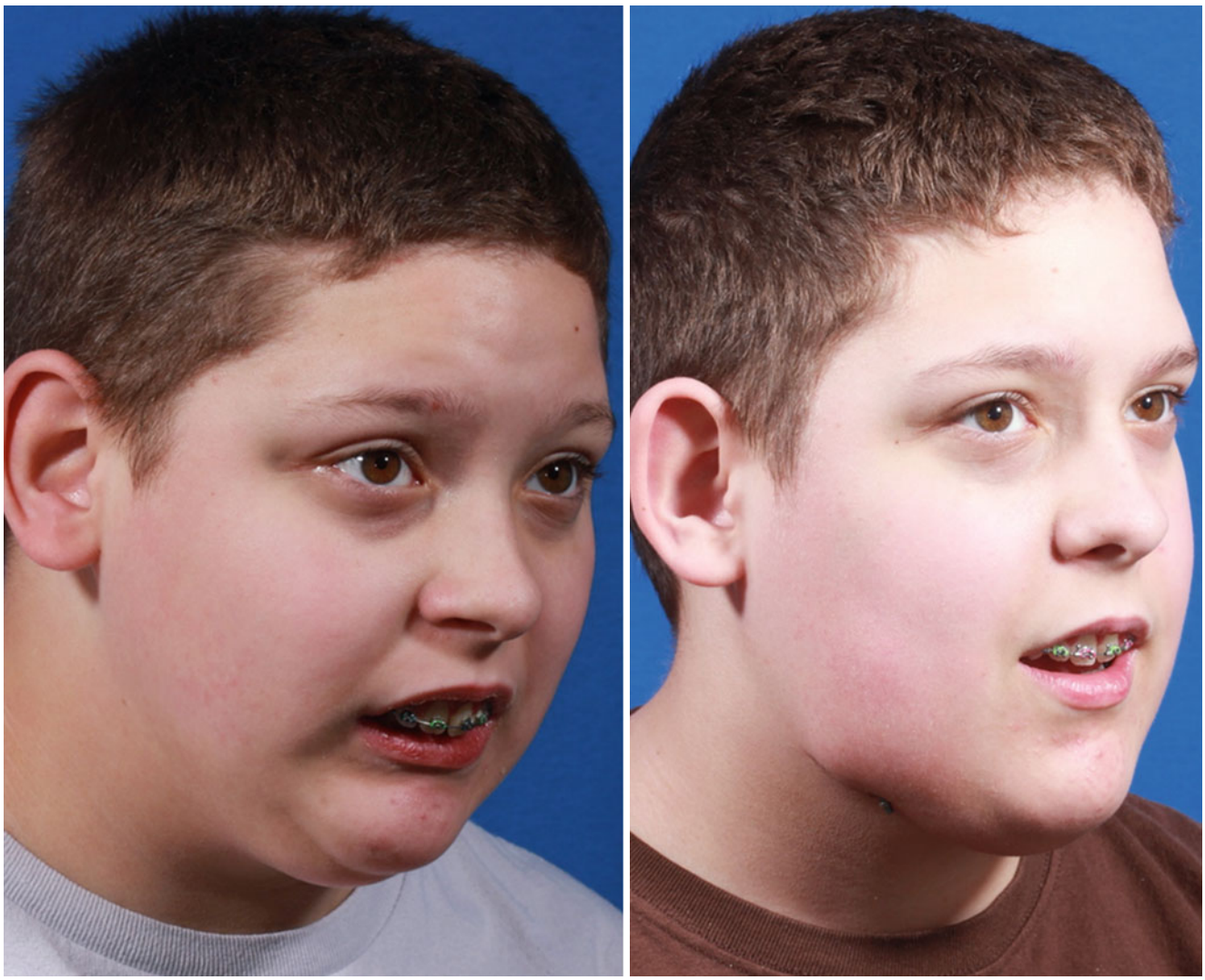


**Fig. 34.8** Same patient preoperative oblique view (*left*) and (*right*) early consolidation phase with device present

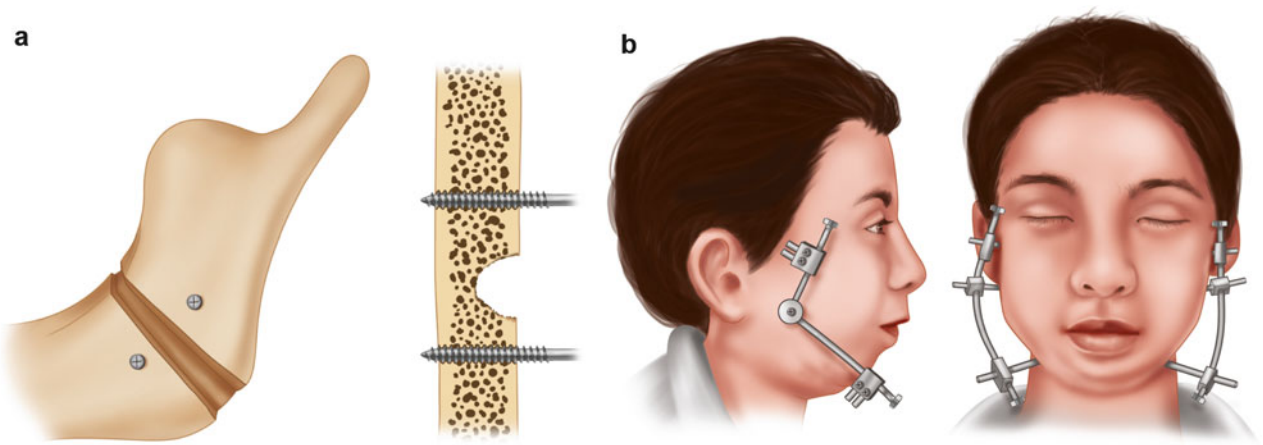



**Fig. 34.9** Molina's patient with corticotomy technique (**a**) and (**b**) external distractors in place

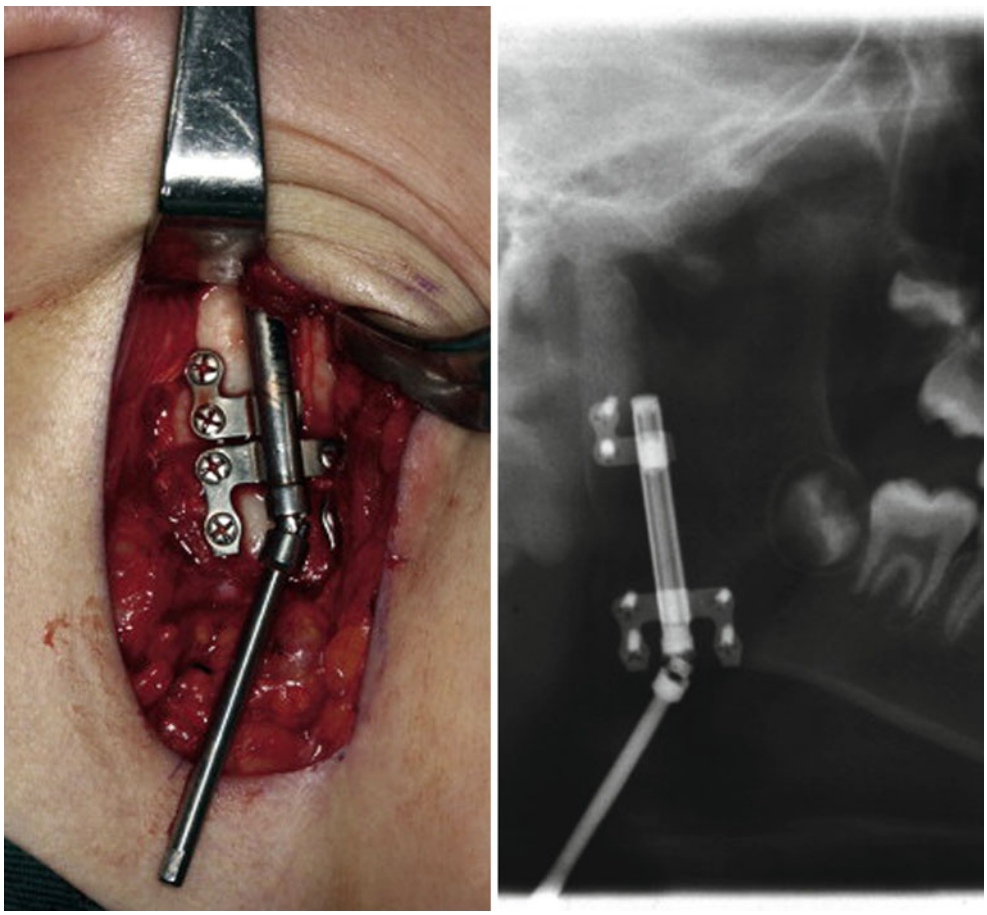


**Fig. 34.10** Example of treatment of condylar hypoplasia with transport distraction. Schematic on *left*, with cephalogram on *right* illustrating movement of transport disk

Once the osteotomies are made and the devices placed, the total distraction course is longer than the neonate. Typically a 5–7-day latency is observed prior to device activation. Depending on the patient and indication for mandibular lengthening, a portion of the distraction may be able to be completed at home by the parents. Active distraction is 1 mm/day. Consolidation lasts 8 weeks but may be shortened slightly to allow for molding of the generate through use of orthodontic appliances and elastics. The end point of DO is skeletal class I or slightly class III.

## Skeletally Mature Patient

Once growth has completed, certain patients have a significant class II malocclusion with or without the presence of severe obstructive sleep apnea. If patients with severe OSA present prior to completed skeletal growth, DO of the mandible may be an option to treat the OSA, knowing that a completion orthognathic procedure may be needed after growth completion. With skeletal maturity, traditional orthognathic techniques are preferred to correct mandibular hypoplasia with class II malocclusion. However, if there is a significant positive overjet (10 mm or more) and or deficient mandibular ramus to allow a traditional sagittal split osteotomy, or a significantly tight soft tissue envelope, DO would be preferred to achieve lengthening without a risk of relapse. Virtual planning can be used to plan precise osteotomies, vectors, and device placement. Typically, as in childhood mandibular DO, a combined intra/extraoral approach to osteotomy and internal device placement is used. Latency in this population is between 7 and 10 days, with a 1 mm/day rate of distraction and a 2-month consolidation period. Devices can be removed prior to complete consolidation for molding of the generate to achieve a better bite relationship. End point of DO is skeletal class I.

## Transport Distraction of the Mandible

There are circumstances when a bone gap is present in the mandible. Congenitally, this may occur with severe hemifacial microsomia, with a Pruzansky 2b or 3 configuration (see Fig. 34.10). Posttraumatic or postresection bone gaps may also exist. Transport DO can successfully bridge segmental gaps, or help to create a neo-functioning TMJ in the case of severe hemifacial microsomia. An internal or external device can be used to achieve transport DO. The concept of transport DO is to liberate a small moveable segment of bone called the transport disk (see Fig. 34.11). This segment will move along a prescribed vector to bridge to another area of

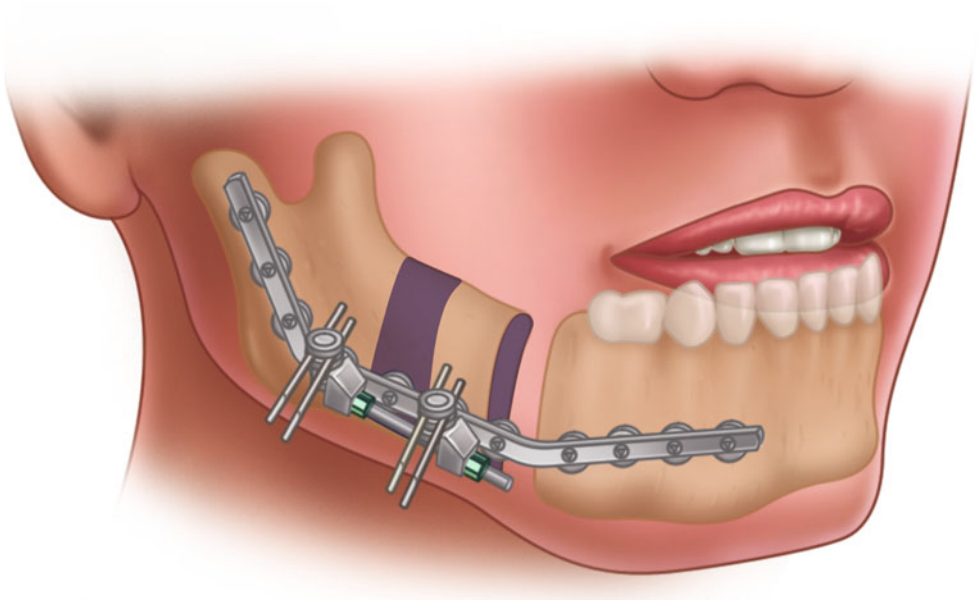


**Fig. 34.11** Schematic of posttrauma/resection segmental mandibular defect treated with transport distraction. Transport disk is moving distally along the device, with new bone forming in the pink region

**Table 34.3** Advantages of Le Fort I DO

| |
|---|
| Overcome soft tissue tightness (scar, hypoplasia) |
| Stable advancement |
| Preservation of speech |
| No bone graft |

bone, or in the case of a congenital absence of the condyle, up the neo-TMJ. Complications include failure to align the transporting segment with the opposite stable bone and nonunion at either the starting or ending points of transport.

## Distraction Osteogenesis of the Maxilla

Patients with maxillary deficiency may also benefit from DO. Common diagnoses treated include clefts of the lip and palate, congenital maxillary hypoplasia, and Binder syndrome. Most of these patients require osteotomy at the Le Fort I level; however, patients with Binder syndrome require osteotomy at a higher Le Fort II level. When maxillary growth is complete, around age 12, the deficiency can be treated. When the patient's class III malocclusion (negative overjet) is less than 10 mm, a standard Le Fort I osteotomy with one step correction can be performed. However, with more severe conditions, such as those with a negative overjet of greater than 10 mm, overcoming soft tissue and scar limitations and creation of a stable advancement without relapse can be difficult. In these instances, Le Fort I by DO is preferred. In addition, bone grafting is not necessary when distraction is used. Past reports have shown that large negative overjets can be overcome with Le Fort I DO without degrading speech (see Table 34.3). Of importance when selecting a candidate for Le Fort I DO is that the maxilla is unified. Cleft lip/palate patients who have not undergone alveolar bone grafting to unite the segments are not candidates for DO.

There are several methods to achieve advancement by distraction. An external device fixed to the skull can be used (see Fig. 34.12a) that pulls the maxillary segment anterior via attachments to the bone or to an intraoral splint. The external distraction method moves the Le Fort I segment well and has the advantage of vector control, but the child must wear a large external rig during the consolidation period. Advocates of this technique typically remove the halo prior to complete consolidation (approximately one month after active distraction) and guide the generate with class III elastics. A completely internal device can also be used to achieve Le Fort I DO (see Fig. 34.12b). This device spans the osteotomy and pushes the inferior, mobile segment anteriorly. It is well tolerated by the patients but is more challenging to use in patients with asymmetric hypoplasia and limited bone inferior to the osteotomy above the tooth roots for footplate positioning. A third method involves a partial bone-borne, partial orthodontic-borne device.

Planning for patients undergoing Le Fort I DO with a partially orthodontic and partially bone anchored device involves virtually planning and possibly fabrication of a model. Presurgical planning includes the anticipated amount of distraction, the vector of distraction, and guides for precise placement of preoperatively constructed partial orthodontic, partially bone born distraction devices. The surgical plan may include overcorrection of the existing deformity into a slight class II relationship (see Fig. 34.13a–c).

Intraoperatively, with the patient under anesthesia via nasotracheal intubation, standard subperiosteal exposure of the maxilla is obtained. Positioning guides for the zygoma-borne footplates of the distractors are placed. After predictive footplate holes are drilled, a standard Le Fort I

osteotomy is completed. Downfracture at the Le Fort I level is performed with Rowe disimpaction forceps. Any posterior bony interferences are taken down with rongeurs. The Le Fort I segment is reduced. Zygomatic footplates of the devices are mounted using the predrilled predictive footplate holes. The lower device footplates rest along the orthodontic appliances and are secured to the orthodontics with multiple 28-gauge wires. Turning arms are attached to the devices and the distractors are tested and returned to the zero position.

The distraction course for Le Fort I DO is as follows. Standard latency period is 7 days. Devices are activated at 1 mm/day until the desired correction is achieved. Consolidation is typically 6–8 weeks. Molding of the generate can be performed if need be with earlier device removal. Using the partially orthodontic bone device with presurgical virtual planning lends itself to the need for less generate molding, however.

## Combined Maxilla/Mandible

In patients with both maxillary and mandibular deficiency, simultaneous DO of both jaws has been described. Skeletally

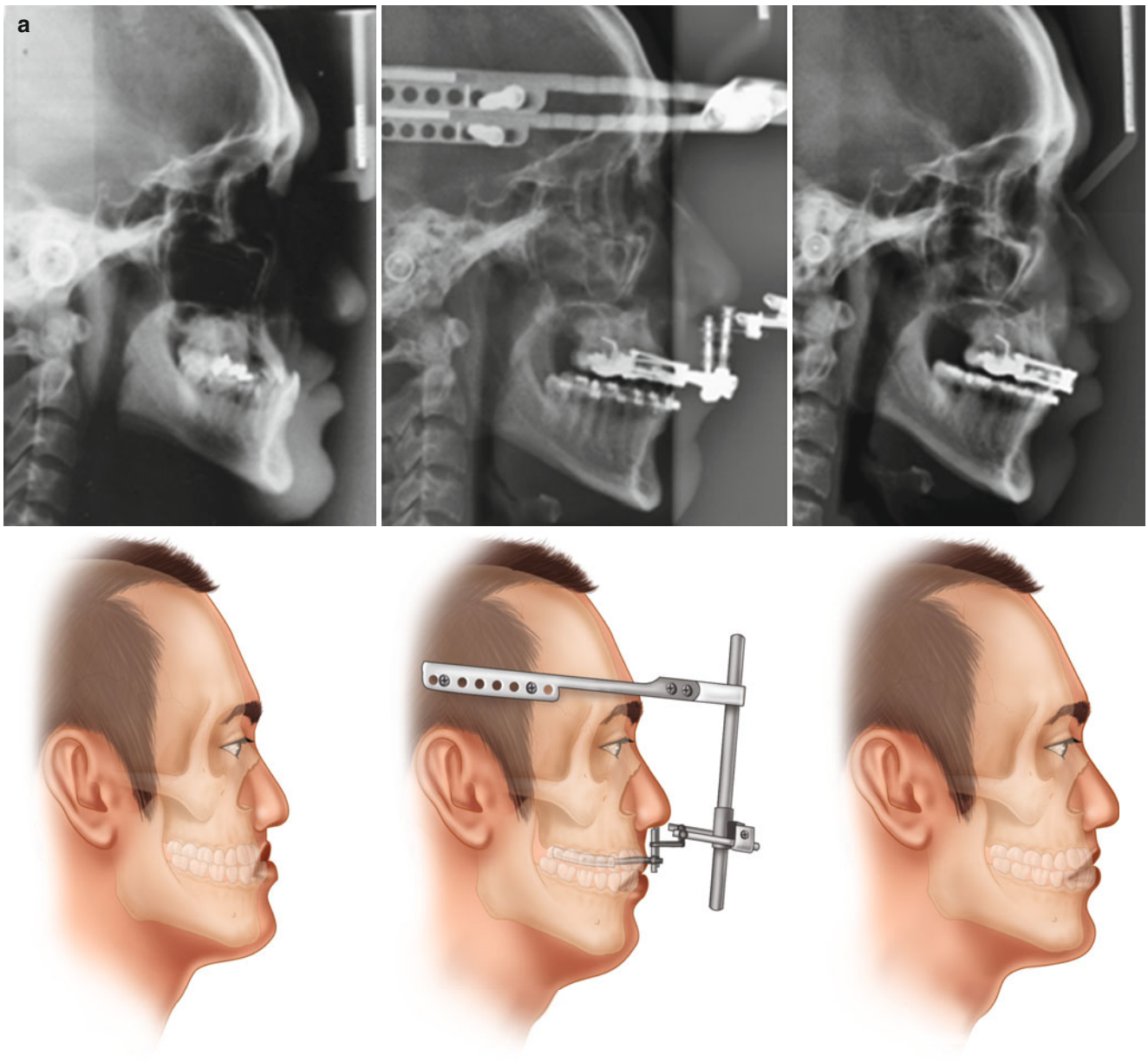
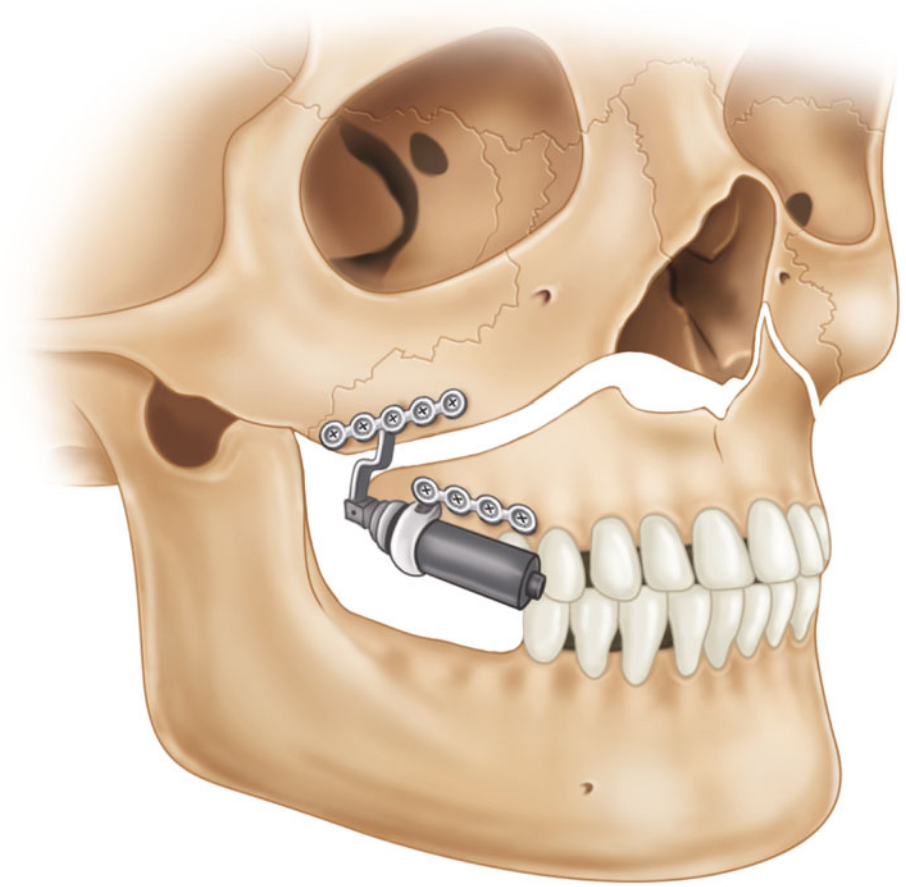



**Fig. 34.12** (**a**) Patient with cleft lip/palate and maxillary hypoplasia treated with RED (rigid external device) (KLS Martin Group, Jacksonville, FL). (**b**) Drawing of internal maxillary distraction device

**Fig. 34.12** (continued)




mature patients with severe hemifacial microsomia have roll deformities of both the maxilla and mandible. With limited vertical growth of the mandibular ramus in a patient with severe hemifacial microsomia, who has not undergone mandibular DO in childhood, the maxillary growth on the affected side is limited. These occlusal cant deformities can present challenges when attempting to achieve skeletal correction with traditional orthognathic surgical movements. In addition to the skeletal deformity, soft tissue is limited on the affected side. Monasterio and Molina have addressed this two jaw and soft tissue deformities with bimaxillary DO in patients with Pruzansky 1 or 2 hemifacial microsomia (see Fig. 34.14). After osteotomy of the mandibular ramus on the affected side and Le Fort I osteotomy, the patient is placed in maxillomandibular fixation (MMF). External distraction of the affected mandibular ramus is conducted with the Le Fort segment moving passively in the MMF.

## Cranium

Distraction is used in the cranium less frequently than other areas and no real clear indications have developed. Lauritzen performed much of the pioneering work on this concept using springs. It appears that distraction is mostly being used to solve unique and less commonly encountered diagnoses and clinical circumstances. The cranium is different than many other bones in which distraction has developed as a predominant treatment modality. The bones are relatively thin in youth and intimately related to the CSF space presenting the challenge of achieving stable fixation without injuring the meninges and increasing the risk of life-threatening meningitis. Additionally, the cranium does not incur a great deal of stress forces that stimulate bone hypertrophy in healing, yet the cranium has demonstrated remarkable ability to regenerate in gaps in ages below 18 months.

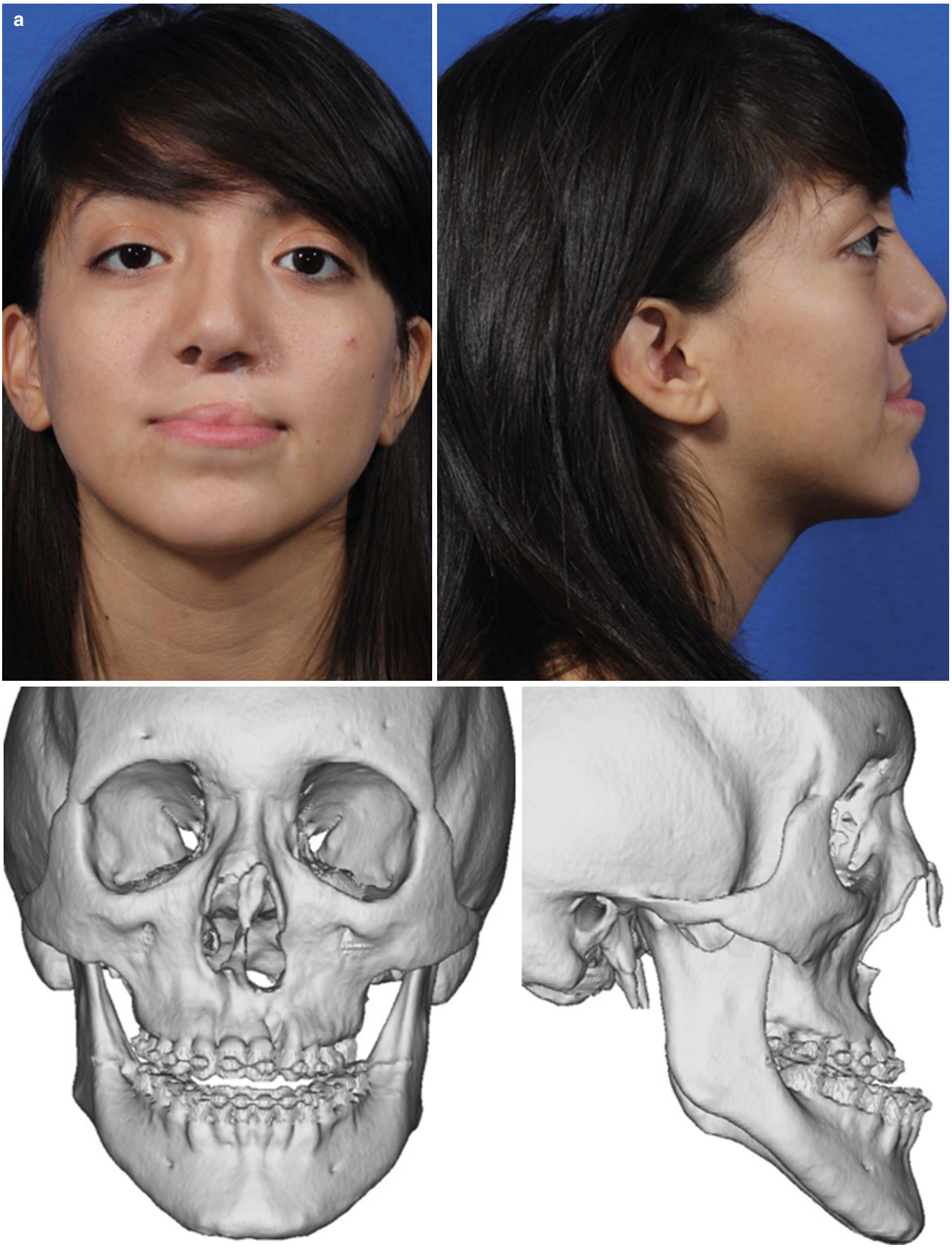



**Fig. 34.13** (**a**) Patient with cleft-related maxillary hypoplasia and class III malocclusion. (**b**) *Upper* – Computer-simulated Le Fort I osteotomy with maxillary distractor (Medical Modeling, Golden, CO) (partial bone, partial orthodontic-borne device, KLS Martin Group, Jacksonville, FL). (**c**) Predistraction and 1-year postdistraction cephalograms

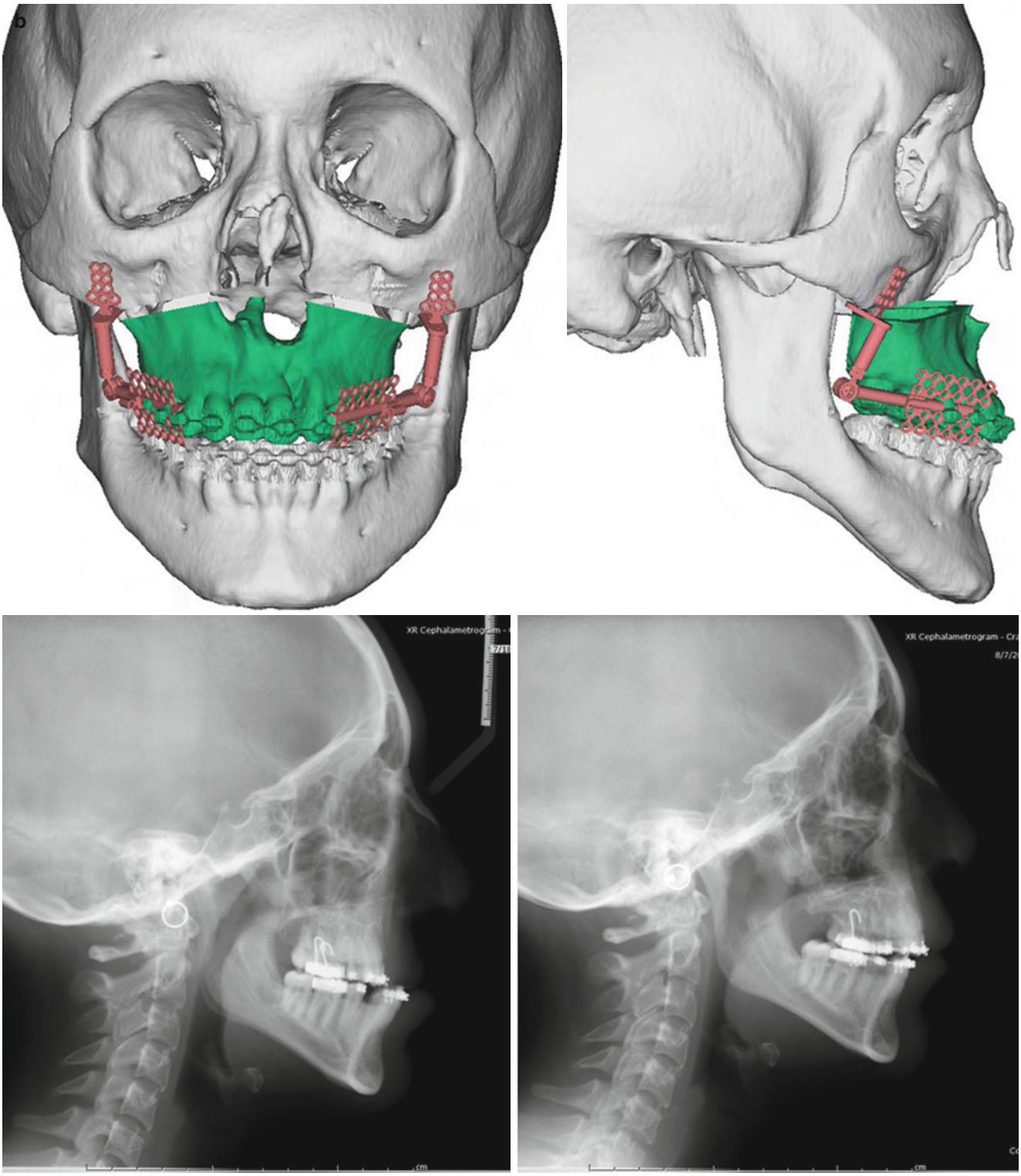



**Fig. 34.13** (continued)

**Fig. 34.13** (continued)


Distraction employed as a method to expand the cranium has several advantages over traditional expansion procedures theoretically. Distraction allows for a slower, gradual expansion, which avoids the epidural dead space with its inherent risks created in procedures that acutely expand the cranial volume (see Fig. 34.15). Slower expansion also allows the scalp to more slowly accommodate expansion and therefore great degrees of scalp expansion can be achieved akin to the degrees of expansion afforded by a tissue expander in the scalp versus a large rotation flap of the scalp. This is an

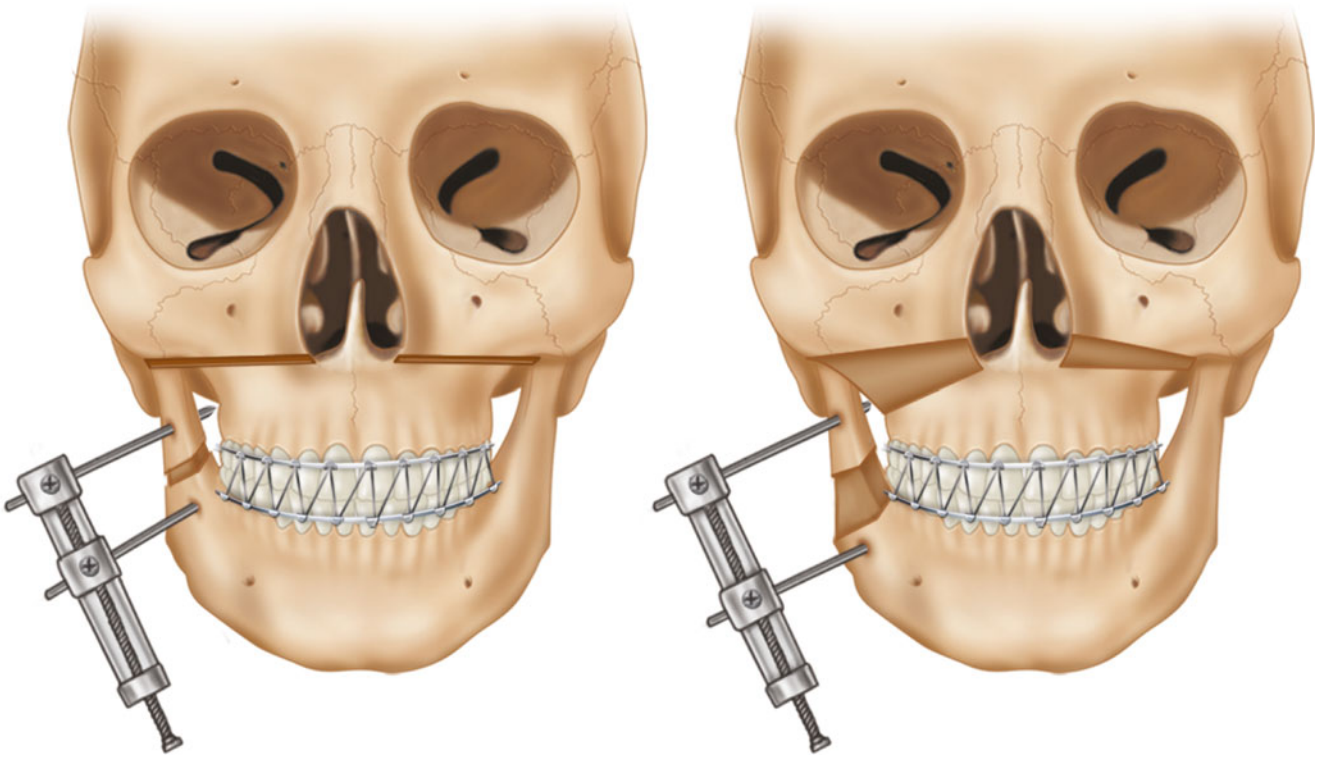


**Fig. 34.14** Schematic of Monasterio/Molina simultaneous maxillary and mandibular distraction, treating R-sided hemifacial microsomia. Le Fort I osteotomy and R mandibular osteotomy, placement of maxillomandibular fixation, and mandibular distractor application. Distraction of the mandibular device corrects the roll deformity of both jaws

important factor since it is often the scalp that limits the volume expansion achieved in traditional single-staged expansion procedures. Additionally, there are more opportunities to maintain vascularity of the cranial bone segments using a distraction method, whereas traditional procedures turn the cranial bone into bone grafts. Distraction procedures are designed to create bone, therefore theoretically reducing reliance on donor bone grafts and minimizing long-term bone defects.

On the other hand there are several disadvantages to cranial distraction. The greatest limitation is that the cranium is a complex three-dimensional structure reconstruction which often requires complex three-dimensional movements which is not easily achieved by bulky uniplanar distractors available on the market today (see Fig. 34.16). The cranium is thin in youth, which challenges adequate fixation of distractors, which need to bear the force of expansion. Lastly, secondary procedures are now required to remove the device, the inconvenience of which may be overcome by enhanced bone formation and minimizing secondary cranial defects themselves requiring additional procedures and donor sites.

Patients with multiple-suture synostosis including syndromic patients often experience elevated intracranial pressure and its sequelae early and ideally benefit from total cranial vault expansion. These patients are often remarkable for a progressive turricephaly. There is questionable utility to repeated expansions of the anterior cranial fossa when an

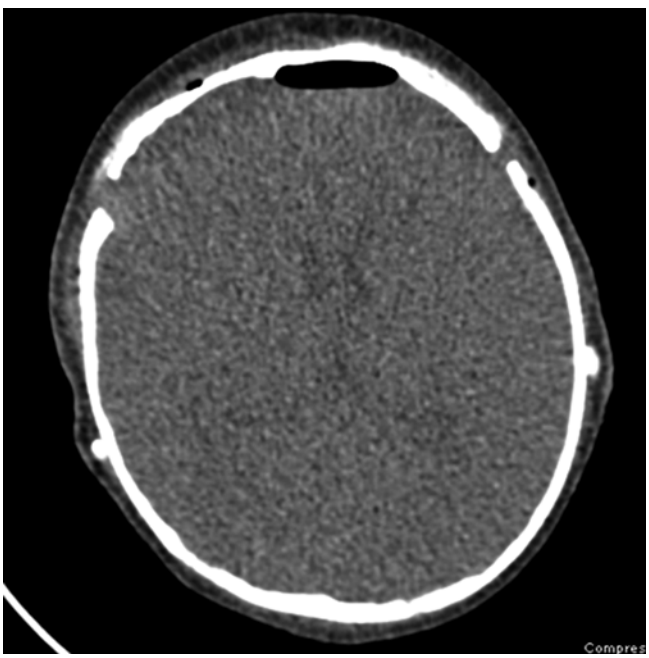


**Fig. 34.15** Epidural dead space created by traditional cranial expansion procedures that increases risk of infection and compromises blood flow to cranial bone secondary to lack of contact

underdeveloped cranium restricts other regions. This is the reason that Chiari malformations are treated with a posterior vault expansion and not an anterior vault expansion. Our

protocol has been to expand the posterior cranial vault as an initial procedure beginning when the patient first starts to demonstrate signs and symptoms of elevated pressure (usually before 6 months of age) (see Fig. 34.17). Traditional expansions are challenged in this early age group by the fact that the bone is thin and soft and does not lend itself to rigid fixation. The posterior vault often relapses under the pressure of the expanded scalp and with recumbent positioning of the child during sleep, which is difficult to avoid in this age group. Also pertinent here is the fact that these patients have an extreme undersupply of cranial bone (reason for their symptoms) and bone graft donor material. Often the patient has a large number of defects and a fairly weak posterior and mid-vault to base the second-stage anterior cranial vault expansion.

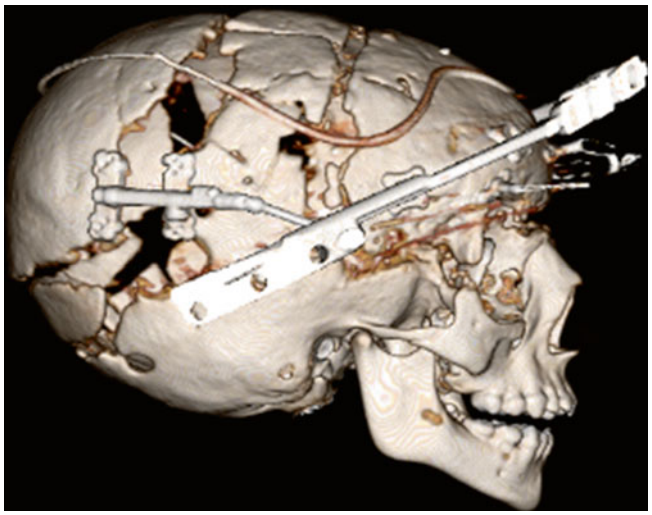


**Fig. 34.16** Scaphocephaly induced by distraction secondary to the limited vector of expansion afforded by today's devices

Posterior vault distraction can be used to overcome the limitations of thin immature cranial bone and the late sequelae of cranial defects and its impact on the stability of anterior cranial vault expansion. Expanding the posterior vault at a reasonably early age will also limit the degree of turricephaly that develops in these patients. A traditional zigzag, coronal scalp incision is employed to expose the posterior two-thirds of the cranium. Cranial cuts are made with a side-cutting craniotome leaving the bone attached to the dura in all areas. The posterior cut is made inferior to the transverse sinus at the torcula but traverses it in the region of the asterion. Greenstick out-fracture of the base of the occiput prevents any step-off from developing as the posterior vault is expanded by the distractors. We employ two cranial distractors (KLS Martin), which have ball joints strategically incorporated to relieve stress on the distractors which inevitably occurs as a result of the inability to achieve a single harmonious vector between the distractors due to the asymmetry inevitably present. Distractor arms emerge through the incision anteriorly.

Standard latency protocol for this procedure is 7 days. Once initiated, the devices are turned 0.5 mm twice daily for a rate of 1 mm/day. Consolidation period is limited to 2 months. Some modification of these time frames and rates can be made based on the age and bone regenerating capacity of the patient.

Another group of patients for which cranial distraction has been used are those with a history of multiple cranial expansion procedures yet still need more expansion in context of a heavily scarred and tight scalp. The slow and gradual expansion of the

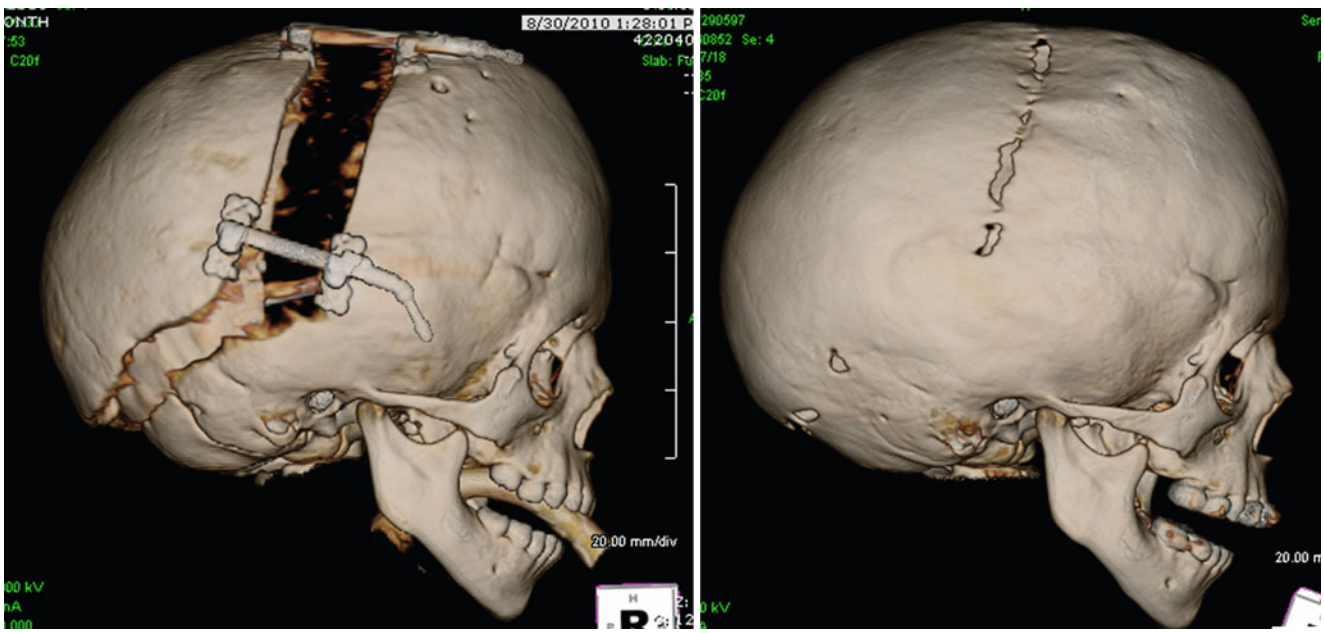



**Fig. 34.17** Posterior cranial vault distraction in a patient with multisuture synostosis as an initial stage to prevent progressive turricephaly. Note the amount of generate bone in this early consolidation image

cranium prevents any further deterioration of the scalp. This strategy should only be applied to patients that need expansion in a single plane. The last group of patients where distraction has offered a benefit is patients with late-onset single- or multiple-suture synostosis who present with a normal cranial morphology but inadequate volume and elevated pressure. These patients can usually be treated by a single vector movement, and by using distraction, donor sites are not required and the generate bone can result in a bilaminar skull with excellent quality avoiding long-term activity restriction.

## Orbit/Upper Midface

The application of distraction to the orbital and upper midface region has significantly improved outcomes of the procedures used to address exorbitism and midface hypoplasia, when severe presents as obstructive nasal breathing and sleep apnea. Distraction applied to the Le Fort I segment was discussed earlier but will be addressed here as well, as it applies to the movement of the maxillary dentition along with the orbits. The two primary procedures are the Le Fort III and Monobloc. These procedures are usually not attempted until the patient has at least completed growth in the orbital region, which is around 7 years of age in boys and girls.

The major functional purpose of the Le Fort III procedure is to advance the lower half of the orbital rim to achieve greater depth to the orbit to accommodate the globe and position the lower eyelid such that it can reach the upper lid for closure and protection of the cornea. There is a strong tendency for function to follow form in this region so reestablishing a normal appearance is often accompanied by a return to functional globe protection. It must be remembered that these operations are first and foremost orbital in nature. Attempts to prioritize the occlusion over the orbit can lead to disastrous results with either under-correction and persistent exorbitism or more commonly enophthalmos which is very difficult to correct once established. The Monobloc procedure additionally aims to increase the intracranial volume in the anterior cranial fossa. Usually an adequate expansion is associated with a precise correction of the orbital position so positioning the lateral orbital rim at the mid-axis of the globe is a good technique for judging the end point of advancement.

The great number of bony interferences and mass of soft tissue that has to move with these procedures generates a great deal of resistance. This is especially true with movements beyond 3–5 mm; distraction with its slow and steady movements is much more effective with much less relapse potential than a single-staged advancement. Additionally, a pulling force on the central face with the lateral components "riding along" is much more reliable and effective for overcoming the resistance than any technique that pushes from the outside margins. This observation comes from the high rate of failures generated by internal distractors pushing from the temporal fossa region relative to the very reliable RED distractor (KLS Martin), which pulls from an anterior and more central vector. Our preference is to use the RED almost exclusively. Although this is an external distractor, the transcutaneous anchoring points are in line with the vector of distraction so pin track scars are not an issue with the use of this device. The patient is left with very little permanent hardware and the device can be removed without making any surgical incisions other than minor pin site suturing. The most important advantage of the RED is that the surgeon maintains complete three-dimensional control of movement. This device does require sturdy bone in the supra-auricular region for securing the device and is almost impossible to use when not present.

### Le Fort III

The Le Fort III procedure is completed in the same fashion as the traditional procedure and with the same degree of mobilization of the mobile segment. Failure to mobilize the segment adequately is the most common reason for failure to advance sufficiently. Our most common approach to the orbital and upper midface is via a coronal scalp incision although a subcranial approach using a series of palpebral incisions has been described. It is best to avoid lower eyelid incisions and complete the orbital osteotomies via the coronal and a complete degloving of the orbit. Care is taken to avoid the detachment of the medial canthal tendon. The nasoglabellar osteotomy must be completed in such a manner that it avoids entrance into the anterior cranial fossa and CSF violation and leak. The point of osteotomy of the lateral orbit depends on where the orbit becomes hypoplastic. If the entire lateral orbit is underdeveloped, then the superior-lateral orbit can be included with a greenstick in the mid-lateral orbit so that a gradual movement occurs superiorly and a greater advancement occurs inferiorly. Every attempt should be made to avoid any stairstepping at the mid-lateral orbit where the soft tissue envelope is thin and the step-off will look odd. If the lateral orbit is in reasonable position, then a lower osteotomy can be performed in the region of the upper edge of the inferior orbital rim and the superior edge of the arch. The soft tissues are thicker there so any stairstep is easily concealed. Osteotomy of the arch is best made as posterior and possible to avoid and mid-lateral cheek depression. The temporalis muscles should be completely mobilized if the osteotomy is anywhere above the mid-lateral orbital rim to avoid any soft tissue contour depression. While mobilizing the temporalis the surgeon should pay careful attention to avoid injuring the deep temporal motor nerve on its deep surface to avoid any atrophy. The muscle and canthal tendon are reattached to their respective places on the transport segment to maintain the normal soft tissue-bone relationships.

**Fig. 34.18** Preoperative and postoperative lateral photos of a patient who underwent a distraction Monobloc procedure to treat exorbitism and elevated intracranial pressure. Note the natural appearance around the orbit because the orbit is kept as a single unit

Inclusion of the Le Fort I segment requires a pterygoid dysjunction; exclusion requires a septal osteotomy at the maxillary crest. It cannot be stressed enough that the Le Fort III segment must be mobilized very well as if the segment was going to be advanced without distraction; it should then be allowed to settle back into its native location to initiate callus formation during the latency period.

After the segment is mobilized and settled back into position, a decision is made about the location of the wire attachment. A minimum of four wires with two on the upper portion and two on the lower portion of the transport segment ensures true three-dimensional control of the movement. Areas with thick bone where pin-retaining plates can be located include the mid-inferior orbital rim, the body of the zygoma, and the inferior pyriform margin. If the Le Fort I segment is transported, then wire attachment to a dental splint or firm arch wire is arranged to avoid any contact or displacement of the lip, which can easily be ulcerated over time.

Latency for Le Fort III procedure is 7–10 days with transport of 1 mm/day divided twice daily. Of course alterations need to be made as discussed previously. Consolidation for these patients is usually as short as possible since the cranial halo is so uncomfortable and makes it difficult to function and sleep. The advancement can be considered stable when the transport wires become loose. This usually occurs three to six weeks into consolidation. Early in consolidation, the segment is still very easily moved especially by any occlusal forces at play. Therefore, care must be taken to avoid the influence of these forces on the final position especially if the advancement results in a malocclusion which will try to work itself out while the generate bone is still moldable. Either longer consolidation or the strategic use of bite splints can be employed to prevent this effect. Strategic use of non-chew diets is also very effective and awareness of grinding preoperatively is very important in this situation. If circumstances allow the bite to become a priority, then early removal and strategically applied elastics will mold the generate bone into a favorable occlusal relationship if these forces are applied very early in consolidation.

## Monobloc

The Monobloc advances the entire orbit so it is a particularly effective and stable way of treating a patient whose orbits are hypoplastic in all dimensions (see Fig. 34.18). Our center uses the Monobloc primarily in syndromic diagnoses who suffer from multiple sutures fusing. We attempt to expand the posterior fossa as much as possible early in life in an

attempt to preserve the integrity of the orbits, essentially buying time until a single Monobloc procedure can be performed to address the anterior cranial fossa and the orbits in one operation. The bones must be sufficiently mature and calcified to accept rigid fixation for a Monobloc to be successful.

Distraction of the Monobloc segment is very similar to the Le Fort III with the exception that it requires an intracranial and extracranial approach to mobilize the segment. The addition of the entire circumferential orbit and a variable portion of the forehead to the Le Fort III segment constitutes the Monobloc. In addition, the forehead is turned into a bone graft by the bifrontal craniotomy so it is subject to all issues of a graft especially resorption. By expanding the anterior cranial fossa gradually, there really is very little issue with epidural dead space. This issue is the primary reason why distracting the Monobloc reduced the complications associated with the procedure which were unacceptably high and morbid including a reasonably high risk of death. The primary risk surrounding the Monobloc procedure is the osteotomy in the anterior portion of the floor of the anterior cranial fossa which creates a communication between the nasal cavity and the anterior cranial fossa. This osteotomy violates the integrity of this separating barrier, and distraction seems to prevent both acute and long-term risk of meningeal infections originating in the nose.

The surgical access to the Monobloc procedure is the same as the Le Fort III with the addition of a bifrontal craniotomy. Again, the inferior eyelid access can and should be avoided to prevent degloving if possible. If required, soft tissue suspension should be performed (ala Gruss). Just like the Le Fort III procedure, the Monobloc is primarily an orbital and cranial volume procedure and not an occlusal procedure, so if the Le Fort I segment is included, some degree of malocclusion should be anticipated, understanding that a Le Fort I (and possible a mandible procedure) will be required closer to the time of skeletal maturity to resolve malocclusion. As with the Le Fort III, care should be taken to avoid detaching the medial canthal tendon. The temporalis muscle and the lateral canthal tendon are handled in the same fashion.

Special attention must be focused on the osteotomy through the cribriform plate. We use cortical bone grafts from the inner table of the frontal bone which are fixated to the Monobloc segment with intracranial plates and designed to override the cribriform posteriorly so that as the Monobloc segment advances the bone gradually slides in to fill the void created. Additionally, a large pericranial flap is raised at the time of the coronal exposure. It is used to interpose between the thin dura of the cranial base and the bone to create an additional layer of protection from the nasal cavity. The Monobloc especially requires very aggressive mobilization. Because it is such a large transport segment, it is very susceptible to interferences and resistance. Once allowed to settle back into its native position, a decision about the location of wire attachment is made. Ideally, six wires (two on superior orbit, two on inferior orbit/zygoma, two on maxillary dentition) are employed which will give perfect control over the segment.

Reducing the risk of cerebral and meningeal infection is paramount. While distraction has reduced complication by eliminating the acute creation of a large epidural dead space, complication still can occur and be very severe. Several points on this issue will be discussed. If the patient has a frontal sinus and it is violated during the surgery, cultures should be taken as documentation of resident flora. Strong consideration should be given to cranializing the sinus especially if the drainage of the sinus is damage or compromised. In addition to the bone grafts and pericranial flap, we manage patients with broad-spectrum antibiotics for 72 h postoperatively and maintain intubation for at least 72–96 h to prevent coughing and other causes of elevated positive airway pressure from causing material in the nose from being forced into the anterior cranial fossa, allowing a strong fibrin seal to form. Additionally, this is a very important aspect of safe airway management since nasal swelling and tongue swelling often put the airway at risk for obstruction and reintubation is very difficult in these patients postoperatively especially with a RED device interfering. Temporary tarsorrhaphy should also be considered especially if exorbitism is severe to protect the corneas during the acute swelling phase.

Latency is usually 7–10 days with a rate of 1 mm/day divided twice daily. Position of the lateral orbit again is a good measure for the end point of distraction. Loosening of wires again can be used to determine when to remove the device. The same occlusal concerns as the Le Fort III exist with the Monobloc. The Monobloc is a very big endeavor with significant risks and perioperative changes that the patient and family should be prepared for. The craniofacial team should be ready for supporting the family during the recovery process. Most patients are happy that they had the procedure but would not be willing to experience it again if required so care must be taken to make sure the initial procedure is successful.

## Complications

Complications during the distraction process can be broken down into those related to adequate bone generation and those related to effects on the surrounding soft tissue envelope. Inadequate generation of bone is caused by either device failure, errors in technique, inappropriate distraction protocol, or poor environment for bone healing. Choosing the correct device for the forces at play is critical to preventing device failure and providing adequate stability to prevent

disruption of the generate bone during distraction and consolidation. While it is often true that greater movements can be achieved with distraction over traditional osteotomies, there are limits. Failure to realize the limits of a particular region or device can lead to treatment failure.

To maximize transport and bone generation, the osteotomy must be performed in such a way that it is complete yet performed using strategies that preserve bone viability at the margins of the osteotomy and minimize any gapping. Overheating the bone using drills and saws or allowing a gap to persist will lead to delays in bone healing (secondary bone healing vs. primary healing) and affect the quality of generate bone that can be produced.

The distraction protocol employed must be adjusted to accommodate the bone healing capacity of the patient and the particular region and condition of the region being distracted. Factors that affect bone healing affect distraction in much the same way. For example, prolonged latency in very young, naïve patients can often lead to premature consolidation, while failure to prolong the latency and slow distraction timelines in a wound that is heavily scarred, or in an older patient, for example, will likely lead to poor bone formation. Age, previous trauma or surgery, radiation, and poor bone stock affect bone healing and therefore require adjustments in the distraction protocol. Use of BMP with proper patient consent in the case of off-label use or import of healthy soft tissues can help overcome some of the negative effects of a poor healing bed.

Distraction procedures have a significant impact on the surrounding soft tissues. The negative effects can often obscure the perceived outcome by the patient despite a successful bone generation. Devices that rely on external pins that are perpendicular to the axis of distraction can cause particularly poor scarring because they drag through the tissues creating a scar track. Preoperative planning should be attentive to placement of any portion of the distractor that traverses the skin to the most concealed area possible so as to minimize the impact of scarring.

Bone is the only tissue capable of significant regeneration capacity. The response of soft tissues in the region of distraction can best be described as an accommodation. As the distraction lengths increase, the ability of the soft tissue envelope to accommodate can be a limiting factor and one can expect neuropractic injuries and pain to be experienced. Most nerve injuries recover, but pain should be perceived as an indication that the soft tissue envelope is reaching its limits.

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