Maxillary distraction resulting in facial advancement at Le Fort III level in cleft lip and palate patients: A report of two cases

Roger A. Zwahlen MD, DMD [a] and Kurt-W. Bütow MChD, DMD, PhD, DSc (Odont), FCMFOS[b]

[a] Honorary Consultant, Department of Maxillo-Facial and Oral Surgery, Oral and Dental Hospital, Pretoria, South Africa

[b] Head, Department of Maxillo-Facial and Oral Surgery, South Africa

Abstract

An 18-year-old female and a 14-year-old male who had previously received surgery for primary repair of a nonsyndromic cleft lip and palate (including alveolar defect bone grafting) unintentionally developed facial advancement at the Le Fort III level after surgical correction of their maxillary hypoplasia. The Le Fort I osteotomy, originally performed for their maxillary dentoalveolar hypoplasia, was an incomplete osteotomy. It was performed without down-fracture, leaving the pterygomaxillary and septal junctions intact. The gradual advancement of the maxilla during distraction osteogenesis was planned to correct the hypoplastic maxilla, and also prevent subsequent hypernasality; however, during the distraction procedure by means of a rigid external device both patients developed an unintentional facial advancement at the Le Fort III level.

The lack of class III malocclusion in unrepaired cleft patients may suggest a link between cleft surgical procedures and the hypoplastic maxillae.[1.] Maxillary hypoplasia is usually corrected at a later stage and once permanent dentition has been completed. The midfacial appearance and occlusion are corrected by means of a Le Fort I osteotomy, with or without a bone graft. [2. and 3.]

During the past 10 years distraction osteogenesis has become an established alternative treatment modality to conventional orthognathic surgery in the correction of various types of craniofacial dysplasias, one being that of the cleft hypoplastic maxilla. Molina et al[4.] Cohen et al [5.] and Polley and Figueroa [6.] reported their clinical experiences in treating maxillary hypoplasia of cleft palate patients according to the principles developed by Ilizarov. [7., 8. and 9.] Molina et al [4.] emphasized the advantages of the de novo bone regeneration as well as the soft tissue lengthening by means of gradual distraction, which would therefore compensate for the impaired velopharyngeal mechanism.

Another not-to-be-underestimated advantage of distraction osteogenesis is its minimally invasive procedure. The pterygomaxillary junction is not a bony union, according to Molina et al[4.] and therefore a surgical separation of this junction is not indicated as a means of in distracting the maxilla. The very rare adverse event, such as loss of vision [10.] in cleft lip and palate patients, due to an aberrant fracture pattern after a Le Fort I osteotomy, might be prevented.

This report presents 2 patients who had previously undergone both primary cleft lip and palate repairs and bone grafting of the alveolar defects. After an incomplete Le Fort I osteotomy without any down-fracture and with intact pterygomaxillary and septal junctions, these patients showed advancement on a Le Fort III level. A rigid external device (RED System; KLS-Martin LP, Tuttlingen, Germany, as described by Polley and Figueroa [6.]) had been used for the maxillary distraction.

1. Case reports

Patient 1 (Fig 1) was an 18-year-old female who had had a primary repair of a nonsyndromic complete left cleft lip and palate. Patient 2 was a 14-year-old male who had had a primary repair of a nonsyndromic bilateral cleft lip and palate.

Both patients also had a bone grafted alveolar cleft. At a later consultation, both showed obvious maxillary hypoplasia and a preoperative negative overjet of 8 and 9 mm, respectively. They also had borderline impairment of the velopharyngeal mechanism. Patient 1 was 14 years old when Le Fort I and bilateral sagittal split osteotomies were performed. However, the airway could not be maintained and the mandibular osteotomy had to be reversed. When she was 17 years old, a second Le Fort I osteotomy with placement of internal distraction devices was unsuccessful in procuring full advancement of the dentoalveolar arch.

Fig 1. Profile of patient 1 showing the retromaxillary position of the maxilla.

1.1. Surgical technique

In both cases surgery was performed under general anesthesia with nasotracheal intubation. After infiltration of a local anesthetic (lidocaine 1% and epinephrine 1/200,000) an alternating vestibular incision was made extending from the upper right second molar to the upper left second molar. The mucoperiosteum was deflected and the anterior and lateral walls of the maxilla were exposed between the canine roots and the infraorbital nerves. The lateral nasal mucosa was gently elevated on both sides and a high Le Fort I osteotomy was performed using a reciprocating saw, without the separation of the

pterygomaxillary junction and without a down-fracture. Septal disjunction was not done. Two long titanium miniplates (Champy; KLS-Martin) bent and fixed with screws were placed, above the roots and just below the Le Fort I osteotomy incision line on each side. The anterior part of the plates which had been bent before the time, were projected under and in front of the upper lip after penetrating the oral mucoperiosteum. Wound closure was by means of an absorbable suture (Vicryl®, Johnson and Johnson, Gauteng, South Africa). After the wound closure the halo portion of the RED and its vertical bar were rigidly fixated and adjusted. Two traction wires were connected the miniplates to the distraction bolt at the vertical bar.

Distraction was commenced 5 days postoperatively by the patients themselves, who had been instructed to increase the distraction distance by 0.5 mm twice a day. The patients, together with their parents, regularly attended the outpatient clinic at the Department of Maxillo-Facial and Oral Surgery. Both patients initially showed some movement on the Le Fort I level. However, at 22 days (patient 1, Fig 2), and at 18 days (patient 2) after distraction had commenced, both patients complained of bilateral retracted eye globes.

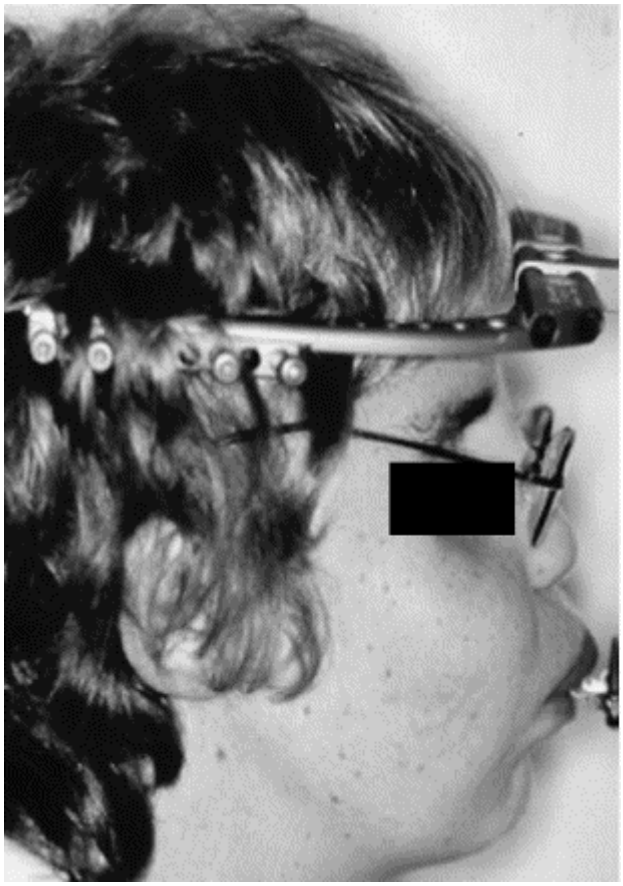

Fig 2. Lateral view of patient 1 showing the “enophthalmos” after Le Fort III osteodistraction.

Apart from bilateral “enophthalmos,” the clinical examinations were as expected. The eye movements in all directions were unrestricted, without any diplopia. Neurological examinations did not reveal any facial numbness or facial nerve damage. On palpation, distended frontozygomatic as well as nasofrontal sutures, could be detected. These presented as demineralised in the following radiographic control of one of the patients (Fig 3). Active distraction was terminated in both patients after the envisaged advancement of the facial skeleton had been achieved. The RED was left in place as retention device for 9 and 10 weeks respectively before being removed. The patients' overall recoveries were uneventful and the bilateral “enophthalmos” slowly improved. Figure 4 is a photograph of patient 1 in a regular follow-up with the facial proportions well balanced.

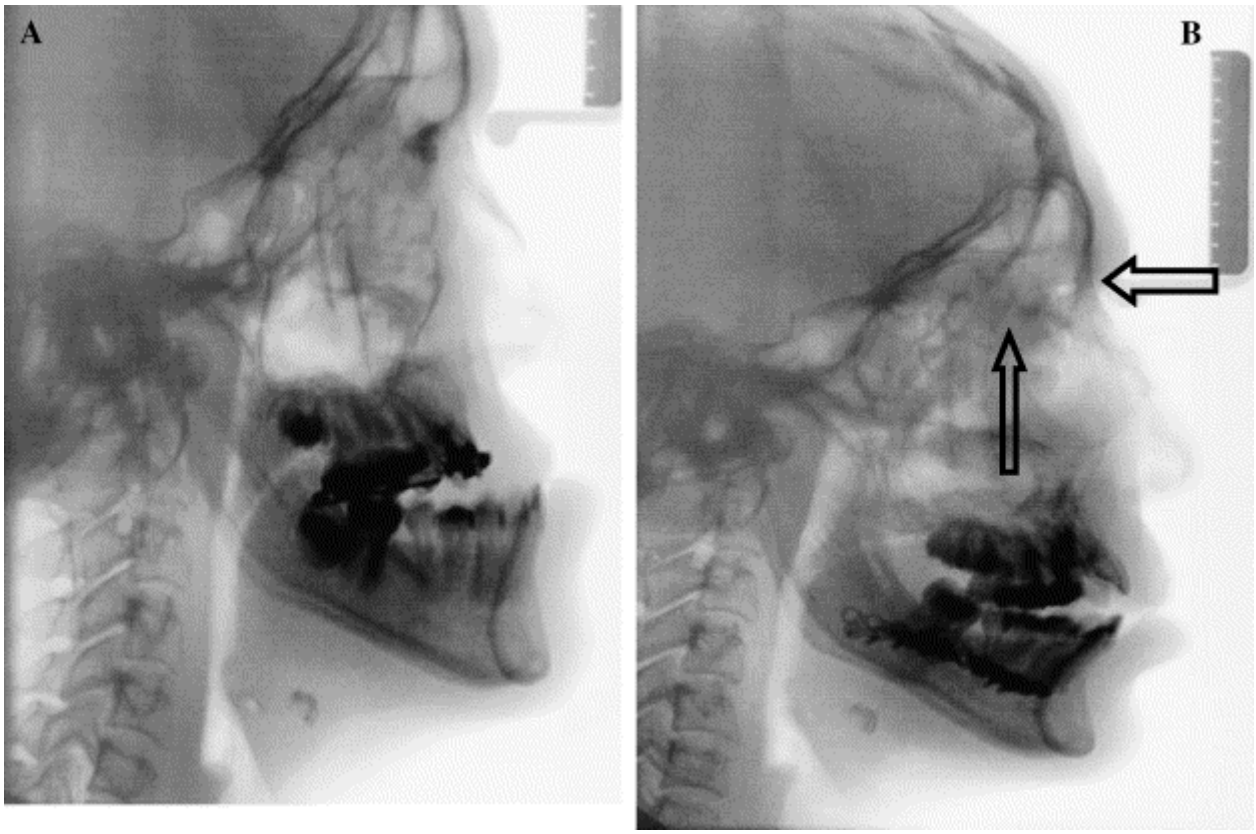

Fig 3. Pre- (**A**) and postoperative (**B**) radiographs of patient 1, indicating the Le Fort III movement after the Le Fort I osteodistraction. Arrows in *B* show demineralization of frontonasal and frontozygomatic sutures.

2. Discussion

Since the 1990's, distraction osteogenesis [4., 5. and 6.] has become a complementary treatment modality to orthognathic surgery (Le Fort I osteotomy, bone grafting and rigid fixation [2. and 3.] for the correction of the hypoplastic cleft maxilla in the adult. The reported relapse tendency of orthognathic surgery may be minimized by the extensive detachment of scar tissue during the mobilization of the maxilla after the down-fracture. [2.] Ross [15.] nevertheless demonstrated in his survey on cleft lip and palate patients that 25% of them developed a maxillary hypoplasia that did not favorably respond to orthognathic surgical treatment.

Fig 4. Clinical view of patient 1 during a follow-up control.

There are certain benefits to distraction osteogenesis for patients who have severe maxillary deficiency that cannot be treated by means of standard orthognathic surgery. The 2 cases reported here also presented preoperatively with impaired velopharyngeal function. In their paper, Molina et al[4.] advocated the use of distraction osteogenesis as a compensation for impaired velopharyngeal function. The same authors [4.] first described the benefits of distraction osteogenesis in cleft patients when a modified “Petit” facial mask, with an intraoral modified quad-helix and elastics, was used for a 2-dimensional distraction device. Rachmiel et al [11.] using a similar method, demonstrated a marked forward advancement of the maxilla. Polly and Figueroa in 1998, [6.] however, introduced the RED system for the first time in cleft patients who experienced insufficient facial movement after the use of the facial mask and elastics. They applied a custom-made intraoral orthodontic splint, which acted as the link between the maxillary skeleton and the external distraction device. Swennen et al [12.] and Harada et al [14.] modified this technique using intraoral titanium chinplates and titanium miniscrews as links between the bone and the RED system. In the 2 cases reported here, the RED system of KLS-Martin was used in combination with titanium miniplates. Buried distraction devices, such as those used by Cohen et al [5.] and Tate et al [13.] are often advocated because they are more acceptable to the patient and leave no extraoral scars. However, there may be difficulty in placing them in the presence of tooth buds or thin bone.

A wide range of devices may be used for osteogenesis distraction. There are also varied methods of doing a Le Fort I osteotomy. The 2 cases reported had an incomplete Le Fort I osteotomy, as suggested by Molina et al[4.] namely a Le Fort I osteotomy enforced from the zygomaticomaxillary buttresses laterally towards the lateral nasal wall of the piriform aperture, without down-fracturing and mobilization. The pterygomaxillary junction and the septal base were therefore left intact.

In contrast, others routinely down-fracture and mobilize the maxilla after pterygomaxillary and septal disjunction,[6., 10., 11., 12. and 14.] Although the “complete” Le Fort I procedure has now most probably

become a standard treatment modality, the significance of its use should not be underestimated. Lo et al [10.] experienced permanent loss of vision as a major complication in 2 of their 94 cleft lip and palate patients who had undergone maxillary distraction osteogenesis. Their literature review detected 5 more patients who had become blind after a standard Le Fort I osteotomy. Two of these Le Fort I osteotomies had resulted in atypical fractures which extended upward into the sphenoid bone, the orbital apex and skull base, therefore causing blindness.

In the cases reported in this paper, distraction was commenced on the fifth postoperative day, which adheres to the time schedule reported in the literature.[4., 5., 6., 10., 11., 12., 13. and 14.] According to Ilizarov, [9.] tissue that is being forced apart responds with primary bone healing and a decrease in the formation of fibrous tissue when the frequency of increasing the distraction forces is accelerated. The distraction device was thus activated 0.5 mm twice daily, instead of 1 mm once daily.

Felemovicius et al[16.] used their clinical study on Tc99 DP to determine the quality and quantity of bone consolidation in patients who had undergone distraction osteogenesis of the craniofacial skeleton. Until the publication of their report, the decision to remove the distraction device had been based upon clinical experience and radiological means only. Felemovicius et al [16.] showed that consolidation occurs earlier in infants than in the older age groups, most probably owing to the higher metabolic rate in these younger patients. In their study, bony consolidation in children younger than 12 months occurred between 4 and 7 weeks, whereas in older children, adolescents, and adults up to the age of 22 years, it occurred between 10 and 14 weeks.

To the knowledge of the authors these are the first 2 reported cases presenting with a facial advancement on the level of a Le Fort III advancement after a Le Fort I osteotomy has been performed for the distraction of a hypoplastic maxilla in cleft lip and palate patients. The pterygomaxillary junction, as stated by Molina et al [4.], is not a true bony union, so the distraction forces will gently enlarge the junction, even if it remains surgically untouched. On the contrary, mobilization of the maxilla by means of Rowe forceps may cause some secondary growth impairment. Some may argue that these unexpected distraction results may be due to an incomplete osteotomy or due to the incomplete separation at the pterygomaxillary junction without down-fracturing. On the other hand, it may be presumed that scar tissue due to the primary cleft surgery generated such resistance contrary to the active distraction forces on the Le Fort I level, that these forces were diverted to the normal chondral sutures such as those of the nasofrontal and frontozygomatic sutures, resulting in an additional movement on the Le Fort III level. Nevertheless, the final distraction result was both esthetically and functionally more than just satisfactory. Both patients are happy with their appearances and no relapses have occurred.

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