Orthognathic Surgery and Definitive Revision of the Lip and Nose in Patients with Cleft Lip and Palate

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In patients with cleft lip and palate, a series of treatments have been performed to achieve a normal facial appearance and a good oronasal function. Decisions on timing and the procedure to be performed should always be based on the patient’s growth. Despite successful, well-timed surgery and adequate orthodontic treatment, maxillary hypoplasia appears to be unavoidable in some patients with cleft lip and palate. Class III malocclusion and the accompanying facial deformity are treated with Le Fort I maxillary osteotomy after facial skeletal growth is completed. Maxillary osteotomy provides dramatic improvement in patient’s facial appearance and occlusion; however, there is a possibility of relapse after surgery. In our retrospective evaluation, there are significant correlations between the amount of surgical movement and relapse in all directions. Because the lip and nose cover the maxilla, the movement of the maxilla influences the appearance of the lip and nose. Therefore, definitive revision of the lip and nose in cleft patients should be performed after maxillary osteotomy. At this stage, all procedures that were not done because of their effects on growth can be performed. For definitive management of the nose and lip, all strategies of reconstructive, plastic, and aesthetic surgery should be considered. A series of treatments as the patient grows after the birth is completed by these revisions.

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Introduction

In patients with cleft lip and palate, a series of treatments are performed to achieve a normal facial appearance and good oronasal function. Decisions on timing and the procedure to be performed should always be based on the patient’s growth. Neonates with cleft lip and palate are referred to a plastic surgeon by an obstetrician or a pediatrician. They are then referred to an orthodontist to apply a Hotz’s plate to correct the deformity of the alveolar ridge. Primary repair of the cleft lip is performed at the age of 3 months when the body weight is estimated to be twice that at birth. Subsequently, the surgical repair of the cleft palate is carried out at the age of 18 months old, when the patient begins to speak. At that time, the ears are routinely examined by an otolaryngologist to guard against otitis media which might cause hearing disturbance. In patients with severe disturbance of articulation, speech therapy started at the age of 3 or 4 years old. Secondary repair of the cleft lip is occasionally done before school age in patients in which the appearance of the lip and nose is unpleasant. Orthodontic therapy with exclusive appliances is started at the age of 6 or 7 years, and bone grafting to the alveolar cleft should be done to achieve alveolar continuity at the age of 7 or 8 years, after the dental arch has been corrected. After the canine tooth in the lesser segment is guided to the site of alveolar cleft by orthodontic treatment. Although these surgical procedures have advantages such as adjustment of anatomical structures and positions, they also have disadvantages of surgical stress and scarring which may interrupt facial growth. Furthermore, the growth potential of the facial skeleton in cleft patients might be less than normal because of maxillary bone defect.

Despite successful, well-timed surgery and adequate orthodontic treatment, maxillary hypoplasia appears to be unavoidable in some patients with cleft lip and palate. Anterior cross bite (Class III malocclusion) due to maxillary hypoplasia becomes apparent in preadolescence in some of these patients. Class III malocclusion and accompanying facial deformity are treated with Le Fort I maxillary osteotomy after facial skeletal growth is completed. Because the lip and nose cover the maxilla, the movement of the maxilla influences the appearance of the lip and nose. Therefore, definitive re-
vision of the lip and nose in these patients should be performed after orthognathic surgery. A series of treatments are completed by these revisions as the patient grows after birth. In this article, we report orthognathic surgery and definitive repair of the lip and nose at the final stage of treatment of cleft lip and palate.

Maxillary osteotomies in cleft lip and palate

Maxillary deformities in cleft lip and palate

Congenital and acquired deformities are observed in the maxilla of patients with cleft lip and palate. One congenital deformity is the alveolar cleft which divides the alveolar arch from the dentoalveolar process to the pyriform margin of the maxilla. Another deformity is maxillary hypoplasia that is generally recognized in the anterior-posterior dimension as in class III malocclusion. However, vertical maxillary deficiency, i.e., maxillary growth disturbance in the vertical dimension, is often observed (Figure 1). Palatoplasty is thought to be one of the causes of maxillary hypoplasia. Palatoplasty interrupts normal maxillary growth because of scar formation at the palate and the posterior portion of the maxilla. Tight lip might also influence maxillary growth because protrusion of the maxilla is seen in some cases of untreated cleft lips (Figure 2). Some teeth may be congenitally absent adjacent to the cleft, and some may be lost by dental caries due to poor oral hygiene or by the ectopic eruption unless an alveolar bone graft is performed. These congenital and acquired tooth missing inhibit maxillary growth.

Facial asymmetry is generally observed in patients with unilateral cleft owing to the hypoplasia of the maxilla on the cleft side. In patients with bilateral clefts, alveolar clefts on both sides of the premaxilla are seen. The premaxilla is always protruded and occasionally deviated downward in childhood. Although these deformities tend to improve as the lateral segments grow, the premaxillary protrusion occasionally remains unresolved despite maxillary growth because of the collapse of the lateral segments after palatoplasty.

Maxillary hypoplasia presents both functional and aesthetic problems. The middle third of the face is usually retruded, and this deformity is called mandibular pseudo prognathism (Figure 3). Insufficient vertical growth of the maxilla shortens the maxillary height so that the upper incisors become invisible behind the lip. In the case of deep bite, mandibular prognathism becomes more obvious (Figure 1). The shallow hard palate and teeth crowding may be observed in cases of severe scarring after palatoplasty (Figure 4). Collapse of

Figure 1. A patient with bilateral cleft lip and palate. Note maxillary hypoplasia with vertical maxillary deficiency.

Figure 2. A patient with untreated cleft lip with alveolar cleft. Note maxillary protrusion.

Figure 3. Maxillary hypoplasia due to unilateral cleft lip and palate. (A) It is called pseudomandibular prognathism. (B) Profile view after two-jaw surgeries.
the alveolar arch may also be seen in some cases. According to the extent of mandibular growth, these patients gradually show Class III malocclusion.

**Maxillary osteotomy for maxillary retrusion**

Le Fort I maxillary osteotomy is performed for maxillary deformities of patients with cleft lip and palate. However, according to the amount of maxillary advancement and maxillo-mandibular relationship, two-jaw surgery (maxillo-mandibular osteotomy) is occasionally performed in some cases (Figure 3). Recently distraction osteogenesis after Le Fort I osteotomy has been done for severe maxillary hypoplasia because of the greater advancement that it achieves\(^1\) (Figure 5). Genioplasty, horizontal osteotomy at the mandible symphysis, is sometimes performed to achieve a proportional appearance of the face.

**Problems in maxillary osteotomy of patients with cleft lip and palate**

Problems in maxillary osteotomy for patients with cleft lip and palate are as follows: (1) the speech dysfunction due to velopharyngeal incompetence (VPI) after maxillary advancement; (2) surgical risk; and (3) relapse after maxillary advancement.

(1) Postoperative speech dysfunction

Although VPI never develops after maxillary advancement in non-cleft cases, VPI is observed after maxillary advancement in some of cleft patients.\(^2\) Scarring after palatoplasty in these patients makes the soft palate so stiff that the velopharyngeal space may become wider after maxillary advancement. In our department VPI has been observed after maxillary osteotomy in almost 20% of cleft patients. However, since VPI was present in half of these patients before jaw surgery, VPI after maxillary advancement was estimated 10% among cleft patients. Additionally, all patients who underwent maxillary advancement of greater than 15 mm with distraction osteogenesis showed VPI after surgery. All these patients with VPI, including those treated with distraction, could regain normal speech function after additional surgeries, such as palatoplasty and pharyngoplasty.

(2) Surgical risk

Compared with non-cleft patients, maxillary advancement in cleft patients is more difficult, more complicated, and more restricted.\(^3\) Many complications in addition to VPI are reported during Le Fort I osteotomy.\(^4\) Complications include otitis media, massive bleeding, circulatory disturbance or necrosis of the maxilla, and oculomotor nerve palsy due to cranial base fracture.

Although massive bleeding is rarely seen in non-cleft patients, uncontrolled massive bleeding has been reported in cleft patients be-

![Figure 4. Teeth crowding due to severe palatal scarring.](image)

![Figure 5. Distraction osteogenesis for severe maxillary hypoplasia. (A) Before surgery. (B) During distraction osteogenesis of the maxilla. (C) After surgery](image)
cause of retro maxillary scarring after palatoplasty. Necrosis of the maxilla is one of the most serious complications. Circulatory disturbance might occur by occlusion or rupture of the greater and lesser palatine arteries, which nourish the maxilla, because scar formation from previous surgeries may decrease the elasticity of the arteries needed for maxillary advancement. However, we have rarely encountered this complication, even in cases of ruptured greater palatine arteries. Violent manipulations may cause extensive subperiosteal dissection of the maxilla and disruption of the mucosa of the retromaxillary site, and they may in turn cause circulatory disturbance of the maxilla. The greatest attention must be paid to the blood supply of the premaxilla in cases of bilateral cleft without alveolar bone grafting. Because the blood supply to the premaxilla is limited through the mucosa of the nasal septum and the lip, special care must be taken not to disturb the blood supply.

In Le Fort I osteotomy, the anterior skeletal structure of the maxilla can be directly osteotomized with a surgical saw or a chisel. However, blind dissection and osteotomy must be performed at the pterygopalatine junction, and downward fracture is also done to detach the posterior wall of maxillary sinus. Both insufficient osteotomies and violent manipulations at the downward fracture may trigger rare but serious complications such as skull base fracture with oculomotor palsy.

(3) Relapse after maxillary advancement

The goals of maxillary osteotomy are the return of normal oromaxillary function, the achievement of proportional facial appearance, and long-term stability. Relapse may occur after Le Fort I osteotomy, especially in cleft patients, even if rigid fixation with the miniplate system is performed. Factors suggested to cause relapse include retromaxillary scarring, the muscle pull, soft tissue tension and stability of the occlusion. Functional adjustment and adaptation of occlusion are important to prevent relapse. However, relapse is still difficult to predict before surgery. Therefore, factors related to relapse after Le Fort I maxillary osteotomy were evaluated in our department. Fifty-eight patients with cleft lip and palate who had undergone Le Fort I osteotomy were investigated using cephalograms.

(i) Relapse after advancement

The amount of surgical advancement in the 58 cases ranged from 2 to 13 mm (average, 6.9 mm). Relapse after operation ranged from 1.5 to 7 mm (average, 1.5 mm), and the average rate of relapse was 24.1%, which was similar to that in other reports. The amount of horizontal advancement was correlated with the amount of relapse (Figure 6). Although the correlation between advancement and relapse was disputable because of the small number of cases in previous reports, we believe the large number of cases in our study is a sufficient basis for judging the correlation.

(ii) Relapse after vertical movement

The vertical height of the maxilla is generally underdeveloped in patients with cleft. Half of our patients had vertical maxillary deficiency, but a quarter of our patients had vertical maxillary excess. Therefore, both maxillary advancement and inferior repositioning of the maxilla were needed in a half of our patients, and intrusion of maxillary height was performed in a quarter of our patients. The correlation between the amount of movement and relapse was observed in vertical movement (Figure 7). In comparison between upward and downward movements of the maxilla, the relapse was greater in downward movement than in upward movement.
(iii) Relapse after rotation

Since anterior open bite is usually found in cleft patients with maxillary hypoplasia, clockwise rotation of the maxilla as well as a maxillary advancement was performed in more than a half of our patients, while counter-clockwise rotation was performed in 30% of our patients. The correlation between rotation and relapse was also confirmed (Figure 8).

![Figure 8. Scatter plots of surgical rotation and relapse in 58 patients with cleft lip and palate who underwent Le Fort I osteotomy. A significant negative correlation ($r = -0.51$, $p<0.0001$) was observed between the two variables.]

(iv) Other factors

In addition to the amount of surgical movement, other factors such as relapse rate were evaluated in our patients. The relapse rate was significantly higher in cases of bilateral cleft (37.2%) than in cases of unilateral cleft (19.1%). However, no significant difference was observed in relapse rate between patients undergoing maxillary surgery alone and those undergoing two-jaw surgeries.

(v) Concluding remarks on the relapse after Le Fort I osteotomy

Out of 71 re-examined patients, there were four patients with massive relapse; two with bilateral cleft and two with unilateral cleft. None of the four patients received bone grafts for the alveolar cleft. In the patients with bilateral cleft, the premaxilla was lost or floating and multiple teeth were missed. In conclusion, it became clear that the relapse occurred in all directions. Relapse after maxillary osteotomy may be more frequent in cases with bilateral cleft, multiple missing teeth and shallow bites. Care must be taken to prevent postoperative relapse in these cases, and two-jaw surgery must be available in severe cases to reduce the amount of maxillary advancement.

Rhinoplasty after maxillary osteotomy

Patients with cleft have a characteristic nasal deformity originating from the deformity of the maxilla. The alveolar cleft, accompanied by septal deviation and depression of the pyriform margin, directly influences the appearance of the lower part of the nose. These are the main causes of the cleft-lip nasal deformity.

Malformation of orbicularis oris and levator labii superiors muscles and hypoplastic nasal cartilages also contribute to the nasal deformity. In the unilateral cleft nasal deformity, the nasal spine and the septum composing the central supporter deviated to the non-cleft side, and the alar cartilage composing the lateral supporter was retruded asymmetrically toward the pyriform margin on the cleft side. In bilateral clefts, both supporters are immature and the premaxilla protrudes producing a more severe deformity of the central supporter. In other words, the maxillary deformity must influence the nasal appearance through the nasal cartilages. However, the skeletal correction does not always improve the deformities of cartilages and the nasal appearance. Especially in adult patients, correction including bone grafts to the alveolar clefts seems to be a requirement for achieving better nasal appearance.

Nasal changes after maxillary osteotomy

The movement of the maxilla directly and indirectly changes the facial soft tissues. Because the nose is based on the naso-maxillary skeletal complex, maxillary movement influences the nose through the cartilages. The nasal bone, the nasal septum and the lateral cartilages are never advanced by Le Fort I osteotomy. On the other hand, only the lower half of the pyriform aperture including the anterior nasal spine is advanced with the maxilla, directly lifting the alar base and the columella base and altering the nasal tip through the alar cartilages and soft tissues. At the same time, the ala and the columella are also kept back by forces from the fixed site.

Changes in nasal appearance accompanying Le Fort I osteotomy vary with the direction of motion. Since surgical movement is multidirectional in most cases, changes in nasal shape are complex. Furthermore, vertical or horizontal rotation makes difference in the amount of movement between the pyriform margins and the anterior nasal spine, and this makes predicting facial appearance more complicated. The soft tissue of the canine fossa is advanced and bulged by advancement of the maxilla, and the alar base become wider (Figure 9). The nasal tip is also advanced with maxillary advancement, and the amount of advancement is approximately one fourth of that previously described by Hui. Simultaneously, the nasal tip is rotated upward and the supratip break may be emphasized. The nasal dorsum is shortened in appearance and the nasal cavity can be visible (Figure 9).

In patients with cleft, scar tissues may be present at columella, ala and tip because of previous operations. Since these scar tissues tend to decrease nasal pliability, the nasal appearance in cleft patients reflects skeletal changes by maxillary advancement more than in non-cleft patients. Rhinoplasty before adolescence in
patients with unilateral cleft is limited to nasal tip and ala. This spatial difference rising from scarring by previous rhinoplasties makes the change after maxillary advancement more obvious. Thus, the supratip beak and the upward rotation of the nasal tip become more severe than those in non-cleft patients (Figure 9). The shorter is the interval from the last rhinoplasty to the osteotomy, the more obvious is the deformity of the nose.

Maxillary osteotomy never improves the deviated nasal septum in patients with unilateral cleft. Because patients with bilateral cleft have a wide nasal base and a short columella before surgery, these deformities may become more obvious after maxillary osteotomy (Figure 10). The columella elongated by previous procedure is easily deformed by maxillary advancement.

Definitive rhinoplasty

Rhinoplasty after adolescence in patients with cleft, called definitive rhinoplasty, is an invasive procedure for the part of the nose untreated during the growth phase. Definitive rhinoplasty comprises correction of the deviated nasal septum, the nasal osteotomy, wide dissection and correction of the nasal cartilages, and bone or cartilage grafting. The goals of this procedure are as follows: (1) correction
of nasal disfigurement and achievement of symmetric appearance; (2) correction of the deviated septum and the nasal bone; (3) correction of changes after maxillary osteotomy; and (4) achievement of a proportional relationship between the nose and the rest of the face. Rhinoplasty after maxillary osteotomy should be performed both to improve cleft nose deformity and to achieve a proportional facial appearance. Therefore, both aesthetic and reconstructive considerations are needed for rhinoplasty (Figure 11).

In general, rhinoplasty is rarely performed simultaneously with osteotomy, because nasal intubation for general anesthesia is needed for maxillary osteotomy. Especially, the nasal tip or the nostrils should be operated on secondarily. Only a simple augmentation for nasal dorsum might be applied simultaneously. Rhinoplasty is usually performed more than 6 months after maxillary osteotomy. During this interval, the nasal cartilages deformed by osteotomy recover or become stable and the postoperative effects disappear. Because bone union is completed at that time, the miniplates can be removed. Furthermore, VPI can be assessed and reconstruction for VPI can be performed at the same time, if necessary.

(1) Bone and cartilage graft

The nasal dorsum is often augmented with bone because maxillary advancement results in a nose with relatively low projection. Calvarial bone graft is usually inserted from the supratip to the nasal root. Bone grafting is also used for augmentation around the canine fossa.

Because cartilage is a reliable tissue for correcting and supporting the nasal tip, ala, and columella, it is used for definitive rhinoplasty in many patients with cleft. Cartilages is harvested from the nasal septum or the auricle. In case of unilateral cleft, harvesting the septal cartilage may facilitate correction of the deviated septum. Harvest has additional advantages such as a single operative site and minimal morbidity at the donor site. Cartilage obtained from the nasal septum can be as large as a tip of thumb. The septal cartilage is categorized as a hyaline cartilage which has good supportability. It is used to support the columella and to lift the nasal tip, and to augment the nasal dorsum. However, its use for augmentation of the nasal dorsum is limited to minor deformity.

Cartilage from the ear is an ideal material because it is elastics and is easily harvested. The size of the graft depends on the size of patient's ear. In cases requiring elasticity or thickness, the cartilage is folded and formed into an L shape by suturing with an absorbable suture, and thus processed cartilage is particularly useful for augmentation and support of the nasal tip.10

(2) Lateral osteotomy of the nose

Patients with unilateral cleft have an asymmetric nose, and some have a distinctive twisted nose. In our experience, lateral nasal osteotomy was performed for 10 of 113 patients treated with maxillary osteotomy. Lateral osteotomy is usually carried out at secondary revision of the nose after maxillary osteotomy.

Definitive revision of cleft lip

The lip is less affected by maxillary osteotomy in its shape than nose. However, many patients with cleft desire labial scar revision even at the final stage of a series of cleft lip surgery. It is important that the lip is revised with all procedures and harmonized to the face. The goals of definitive lip revision include (1) correction of
the remaining deformity, (2) correction of changes after maxillary osteotomy, (3) achievement of symmetry, and (4) a proportional labiofacial relationship.

In some patients with bilateral clefts, the upper and lower lips are still imbalanced after maxillary advancement. In patients with a short lip, the cross-lip flap from the lower lip is especially effective (Figure 12). Almost all patients wish for revision of the residual scar on the lip; however, it is difficult to make the scar completely invisible. In adult male patients, hair can be transplanted to conceal the hairless scar. In patients with a thin vermilion, fascial grafting can be used for augmentation.

Conclusion

Because the mandible has the latest growth spurt of all bones, jaw surgery is generally performed after adolescence when mandibular growth has been completed. The purpose of jaw surgery is to achieve normal oromandibular function and to improve facial appearance; however, treatment is not always completed with this operation. Some aspects of facial appearance are improved with osteotomies but some are not. The nose and the lip covering the maxilla are greatly affected by maxillary osteotomy. Patients with cleft originally have some deformities of the lip and nose. Thus, correction of the nose and lip may be required. At this stage, all procedures that were not done because of their effects on growth can be performed. For definitive management of the nose and lip, all strategies of reconstructive, plastic and aesthetic surgery should be considered.

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