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Long-term follow-up of a patient diagnosed with Crouzon syndrome who underwent Le Fort I and III distraction osteogenesis using a rigid external distractor system

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ABSTRACT

This case report describes the successful treatment of Crouzon syndrome in a patient with severe midfacial deficiency and malocclusion, including reverse overjet. In Phase I treatment, maxillary lateral expansion and protraction were performed. In Phase II treatment, after lateral expansion of the maxilla and leveling of the maxillary and mandibular dentition, an orthognathic approach including simultaneous Le Fort I and III osteotomies with distraction osteogenesis (DO) was used to improve the midfacial deficiency. After DO, 12.0 mm of the medial maxillary buttress and 9.0 mm of the maxillary (point A) advancement were achieved, which resulted in a favorable facial profile and stable occlusion. Even after eight years of retention, the patient's profile and occlusion were preserved without any significant relapse.

KEY WORDS: Crouzon syndrome; Craniofacial surgery; Distraction osteogenesis; Rigid external distractor system; A long term follow up

INTRODUCTION

Crouzon syndrome is an autosomal dominant disorder characterized by multiple premature fusions of craniofacial sutures that cause secondary alterations in facial bones and facial appearance. It is a rare entity that occurs in 1 in 60,000 newborns.¹ Partial or complete premature fusion of cranial and/or facial sutures, as well as synchondrosis, causes typical clinical features of this syndrome, such as a risk of developing raised intracranial pressure, which has the potential to impair both vision and neurocognitive development. Crouzon syndrome also results in characteristic facial appearances including hypertelorism, exophthalmos, external strabismus, parrot-beaked nose, short upper lip, and mid-facial deficiency with a hypoplastic maxilla. Early prophylactic cranial vault expansion is advised to alleviate the pathological symptoms associated with increased intracranial pressure, such as ocular complications, including optic atrophy or potential impairment of neurocognitive development.² Additionally, patients with Crouzon syndrome often present with Class III malocclusion, anterior crossbite, and midface concavity due to maxillary deficiency, which requires orthognathic treatment. Le Fort III osteotomy is frequently used to successfully treat craniofacial deformities.³ Furthermore, DO in combination with Le Fort III osteotomy is an alternative treatment for craniofacial problems in Crouzon syndrome. Both approaches are effective in providing favorable craniofacial function and aesthetics in Crouzon syndrome.⁴⁻⁷ However, further clinical evidence is necessary to select a treatment protocol for correcting craniofacial features and malocclusion in patients with Crouzon syndrome.⁸

Studies have reported successful treatment of patients with midfacial hypoplasia involved in Crouzon syndrome by Le Fort III DO using a rigid external distractor system (RED) in mixed dentition.⁹⁻¹¹ However, a limited number of studies have explored the long-term stability of these treatments, especially the treatment outcomes of Le Fort III DO. In this report, a patient with Crouzon syndrome was treated through an interdisciplinary approach combining Le Fort I and III DO with orthodontic treatment, leading to a favorable facial appearance and occlusive stability in the long run.

Diagnosis and Etiology

A 5-year-old girl with Crouzon syndrome was referred to the clinic because of midface deficiency and an anterior crossbite (Figures 1 and 2). Before visiting the hospital, fronto-orbital advancement with Le Fort III osteotomy and strabismus surgery was performed at the age of four years to improve her intracranial pressure and exorbitism.¹² At the time of the first visit to the hospital, the chief complaint was a concave facial profile and an anterior reverse overjet. The patient exhibited severe midfacial deficiency with skeletal Class III malocclusion and total crossbites. During the first phase of orthodontic treatment at five years of age, maxillary lateral expansion and protraction using a reverse headgear were performed to improve midfacial deficiency for four years. However, limited forward advancement of the maxilla was observed on the superimposition of lateral cephalograms (Supplemental Figure 1).

Before beginning Phase II treatment, an extraoral examination of the patient at the age of 14 years and 10 months revealed severe midfacial deficiency, moderate exorbitism, and a concave facial profile with a protruding forehead. The occlusion consisted of anterior and posterior crossbites. The occlusion was classified as Class III dental relationships on both sides, with an overjet of -3.8 mm and an overbite of 3.9 mm. The maxillary dental arch showed lateral constriction and severe crowding, with a labially blocked right canine, whereas the mandibular dental arch exhibited moderate crowding. Dental tubercles were observed on the palatal sides of the maxillary lateral incisors. The maxillary and mandibular skeletal and dental midlines coincided with the facial midline. Additionally, the patient had no symptoms of sleep-disordered breathing. Panoramic radiography revealed congenitally missing bilateral second and third molars on the maxillary arch. Cephalometric analysis showed a skeletal Class III relationship (ANB, -8.9°) with a retrusive maxilla (SNA, 74.0°). The maxillary incisors were proclined (U1-FH, 129.1°), and the mandibular incisors showed normal inclination (L1-MP, 90.5°) (Figure 3; Table). Neither the maxilla nor the mandible showed further growth from the end of Phase I

treatment, which allowed us to initiate Phase II treatment at this time (Supplemental Figure 2).

Treatment Objectives

The treatment objective was to improve the midfacial deficiency with a concave-type facial profile associated with a skeletal Class III jaw deformity. Lateral expansion of the maxillary dentition was required to harmonize the maxillary and mandibular dental arches. Additionally, rotation of the maxillary first molars and crowding in both arches required correction during preoperative orthodontic treatment. The following treatment plan was proposed: (1) maxillary lateral expansion with a quad-helix appliance, (2) placement of the preadjusted edgewise appliances in both dental arches to level and align the dentitions, (3) simultaneous Le Fort I and III osteotomies with DO, (4) obtaining ideal occlusion by detailing, and (5) retention. A plan was made to move the upper and lower halves of the midface by 12.0 mm and 10.0 mm, respectively.

Treatment Alternatives

Several alternative treatment options were available. These included: (i) extraction of the first maxillary and mandibular premolars and Le Fort III DO. This may be combined with the advancement of mandibular bilateral sagittal split osteotomy (BSSO) advancement to potentially improve the airway. The maxillary premolars were retained due to the congenital absence of the maxillary second molar. However, maxillary incisor proclination persisted until the end of the treatment. Maxillary premolars could be extracted or temporary anchorage devices could be used to incline the maxillary incisors. (ii) Le Fort III DO or osteotomy for acute midface advancement was considered if there was no need to differentiate between the advancement of the orbital rim and maxilla. In this case, the objective was to improve the patient's exorbitism and midfacial deficiency, while less advancement was necessary for proclined maxillary incisors. (iii) Orthodontic camouflage would provide positive overjet and retain skeletal discrepancies.

Recently, maxillary anterior segment distraction osteogenesis (MASDO) has been developed to advance the anterior maxillary segments and improve the retrusion of the maxilla without worsening the velopharyngeal function.^{13,14} However, the effects of improving exorbitism have been limited.

Treatment Progress

At five years of age, a reverse headgear was used to protract the maxilla to correct a skeletal discrepancy and midfacial deficiency for four years.

Phase II orthodontic treatment was initiated at 14 years and 10 months of age by lateral expansion of the maxilla using a quadhelical appliance. The intermolar width was increased by 3.0 mm by improving the mesial rotation of the maxillary first molars. The mandibular third molars were removed and the dental tubercles on the palatal side of the maxillary lateral incisors were reduced. Subsequently, 0.022-in pre-adjusted fixed appliances were placed on the maxillary and mandibular dentitions for leveling and alignment (Figure 4). The maxillary incisors showed proclination prior to orthognathic surgery. After one year of presurgical orthodontic preparation, combined Le Fort I and III DO was performed at the age of 15 years and 10 months to improve exorbitism by forward movement of the orbital rim, while limiting forward movement of the maxillary incisor (Figure 5). Distraction was performed at two levels to produce different advancements in the orbital rim and maxilla. Both segments were performed 1.0 mm / day. After 12 days, the lower half of the midface reached its planned position with a positive overjet. Four days later, the upper half of the midface reached the planned position, resulting in the preferred facial profile with improved exorbitism and midfacial deficiency. With periodic assessment of facial and intraoral occlusions, the position of the device was adjusted to change the vector of bone movement. The use of extraoral devices may cause significant discomfort; however, they greatly facilitate the manipulation of the vector direction. The intermaxillary elasticity can also be used to change the direction of bone movement. After active distraction,

intermaxillary consolidation with an occlusal splint was performed for one week. A slight enlargement of the upper airway was observed in the superimposition of the presurgical and postsurgical cephalograms (Figure 5). After two years of postoperative orthodontic treatment, all appliances were removed and replaced with Begg-type retainers in both the arches. No obvious root resorption was detected on the panoramic radiographs (Figure 6). The facial profile and occlusion did not undergo significant relapse and maintained a favorable status even eight years after DO surgery (Figures 7 and 8).

Treatment Results

The concave facial profile and midfacial deficiency showed substantial improvement with anterior movement of the midfacial bones and combined Le Fort I and III DO. In the present report, combined Le Fort I and III DO resulted in forward movement of the medial maxillary buttress and point A as 12.0 mm and 9.0 mm, respectively, which substantially improved the facial profile and occlusion. Post-treatment facial photographs revealed a straight facial profile. Intraoral photographs showed normal overjet and overbite with favorable occlusion. The molar relationship was Class I on both sides. Maxillary and mandibular crowding were eliminated to achieve proclination of the incisors. Post-treatment cephalometric analysis revealed a skeletal Class I relationship with an ANB angle of -0.7°. The interincisal angle (110.8°) was smaller than the ideal value at the end of the treatment (Figure 9, Table). Postsurgical CT was not performed to reduce the radiation dose.

DISCUSSION

Crouzon syndrome is associated with a wide range of craniofacial deformities and severe malocclusion resulting from premature fusion of cranial sutures and synchondroses. Advancement of the frontal bone at an early age is frequently performed to prevent or improve the intracranial pressure.¹⁵ Furthermore, Crouzon syndrome can result in skeletal hypoplasia of the midface and

severe malocclusion, such as reverse overjet.¹ Orthodontic treatment was performed in different phases to correct skeletal and dental discrepancies. In this case, a reverse head gear **was used** to improve midfacial deficiency, which resulted in limited forward movement of the maxilla. A recent study revealed that patients with syndromic craniosynostosis, including **those with** Crouzon syndrome, exhibit early radiological fusion of the circummaxillary suture.¹⁶ **These results indicate that an orthopedic approach for correcting the maxilla in patients with syndromic craniosynostosis should be considered with caution. Additionally, meticulous assessment of follow-up radiographic examinations is highly recommended to evaluate the efficacy of treatment and avoid unnecessary interventions.** When severe skeletal deficiency persists even after adolescence, surgical intervention is required to achieve normal occlusion.

Due to severe midfacial deficiency and exophthalmos, surgical intervention, including Le Fort III osteotomy, is frequently used in the treatment of malocclusion. **This case report demonstrates** the long-term results of comprehensive orthodontic treatment in a patient with Crouzon syndrome treated with **a combination of** Le Fort I and III DO using an RED system.

Le Fort III osteotomy was first described by Gillies in 1950 and **was** successfully performed by Tessier in 1971.^{4,17} Ortiz-Monasterio et al¹⁸ introduced **the** craniofacial monobloc Le Fort III osteotomy as an improved surgical procedure for the treatment of patients with craniosynostosis. DO with Le Fort III osteotomy has also been used as a treatment protocol for patients with severe midfacial deficiencies and exophthalmos.^{19,20} Several case series have shown an average forward movement of the midface following Le Fort III DO of 12-20 mm²¹ or 12-22 mm²⁰. In some studies, Le Fort III DO has been shown to be stable for more than five years.²²⁻²⁴

In contrast, combined Le Fort I and Le Fort III osteotomies have been **used** in cases **requiring** differential correction of the orbital rim and maxillary component.^{25,26} Le Fort I and Le Fort III

distraction osteotomies have been successfully performed in patients with syndromic craniosynostosis; **however**, their long-term stability remains largely elusive.²⁷⁻³⁰

In the present case, eight years after distraction, **minimal** relapse of maxillary advancement was observed. Patients with syndromic craniosynostosis, including **Crouzon's** syndrome, show a higher frequency of sleep apnea due to midface or mandibular hypoplasia.³¹ Surgical mandibular advancement should be considered in such cases. In the present case, **no sleep disorders were detected** and the size of the mandible was within the normal range; thus, **the patient was treated** without mandibular advancement. This case report, along with existing evidence, suggests the advantage of Le Fort I and III DO as an option for orthognathic treatment of malocclusion and **for** achieving long-term stability in patients with Crouzon syndrome and severe midfacial deficiency.

CONCLUSIONS

- **A patient diagnosed with Crouzon syndrome was effectively treated using a RED system during concurrent Le Fort I and III DO procedures. The patient's exorbitism and midfacial deficiency were notably ameliorated, and her Class III malocclusion was successfully corrected. Eight years after DO, there was minimal relapse of maxillary advancement.**
- Considering **the** present clinical results, simultaneous Le Fort I and III DO is suggested **as** an efficient treatment with good long-term stability in patients with Crouzon, which requires different advancements between the orbital rim and maxilla.

SUPPLEMENTAL DATA

Supplemental Figures 1 and 2 are available online.

Supplemental Figure 1. Superimposed cephalometric tracings: start of Phase I treatment (black) **and** end of Phase I (**gray**).

Supplemental Figure 2. Superimposed cephalometric tracings: end of Phase I treatment (black) and start of Phase II (gray).

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FIGURE LEGENDS

Figure 1. Initial records (age, 5 years and 8 months): A, Facial and intraoral photographs; B, Radiographs and cephalometric tracing.

Figure 2. Initial dental models.

Figure 3. Pretreatment records (age, 14 years and 10 months): A, Facial and intraoral photographs; B, Radiographs and cephalometric tracing.

Figure 4. Preoperative of sagittal split ramus osteotomy records (age, 15 years and 9 months): A, Facial and intraoral photographs; B, Radiographs and cephalometric tracing.

Figure 5. Postoperative simultaneous Le Fort I and III DO records (age, 15 years and 10 months): A, Profile photograph, lateral and posteroanterior cephalograms; B, superimposed cephalometric tracings: presurgery (black), postsurgery (blue).

Figure 6. Posttreatment records (age, 17 years and 8 months): A, Facial and intraoral photographs; B, Radiographs and cephalometric tracing.

Figure 7. Postretention records (age, 26 years and 0 months): A, Facial and intraoral photographs; B, Radiographs and cephalometric tracing.

Figure 8. Postretention dental models.

Figure 9. Superimposed cephalometric tracings: pretreatment (black), posttreatment (red),
postretention (green).