Segmental distraction osteogenesis with modified LeFort II osteotomy for a patient with craniosynostosis

Keiichiro Watanabe, a Shingo Kuroda, b Takumi Takahashi, c Takeshi Kijima, d Katsuyuki Torikai, e Keiji Moriyama, f and Eiji Tanaka g
Tokushima, Kagawa, Tokyo, and Kanagawa, Japan

In this article, we report successful orthodontic treatment combined with segmental distraction osteogenesis after a modified LeFort II osteotomy in a patient with craniosynostosis. An 8-year-old boy diagnosed with craniosynostosis had a dished-in face, an anterior crossbite, and a skeletal Class III jaw relationship because of midfacial hypoplasia. At the age of 13 years 6 months, the maxillary second and mandibular first premolars were extracted, and leveling and alignment of both arches was started with preadjusted edgewise appliances. At age 14 years 11 months, the patient had a modified LeFort II osteotomy, and the maxillary segment was advanced 7 mm and fixed to the zygomatic bone. At the same time, segmental distraction osteogenesis was started with a rigid external distraction system, and the nasal segment was advanced for 20 days at a rate of 1.0 mm per day. The total active treatment period was 40 months. As a result of the modified segmental distraction osteogenesis, significant improvement of his severe midfacial hypoplasia was achieved without excessive advancement of the maxillary dentition. Both the facial profile and the occlusion were stable after 1 year of retention. However, the nasal segment relapsed 1.4 mm during the 1.5 years after the segmental distraction osteogenesis. Evaluation of the stability and retention suggests that some overcorrection in midfacial advancement is recommended. (Am J Orthod Dentofacial Orthop 2012;142:698-709)

Craniosynostosis is a congenital malformation characterized by the premature synostosis of the cranial sutures and midfacial hypoplasia causing hypertelorism and exophthalmos. 1, 2 Because of the midfacial hypoplasia, these patients often demonstrate a skeletal Class III jaw relationship with an anterior crossbite and require orthodontic or orthopedic treatment for both functional and esthetic reasons. 3, 4 To treat the midfacial hypoplasia, distraction osteogenesis has been commonly used in the past decade because it could provide significant midfacial advancement with gradual progress compared with traditional orthognathic surgeries. 5-7 Distraction osteogenesis with a LeFort III osteotomy has been preferentially applied in patients with severe hypoplasia and exophthalmos to improve their facial esthetics. 8

On the other hand, the severity of the malocclusion differs greatly in patients with craniosynostosis. Some patients with severe midfacial dysplasia even show a mild Class III occlusion. The discrepancy between the facial profile and the occlusion often causes difficulties in treatment planning. A single osteotomy is occasionally unsuitable for improving the complicated skeletal problems. Polley and Figueroa 9 demonstrated the usefulness of piggyback osteogenesis to move the upper and lower midfacial segments. Additionally, several reports have shown acceptable results of dual segmental distraction osteogenesis after LeFort I and III osteotomies in patients.
Fig 1. Photographs at the initial consultation.

Fig 2. Radiographs and cephalometric tracing at the initial consultation: A, lateral cephalograph; B, panoramic radiograph; C, tracing (black line) superimposed with the mean profilogram of a 6-year-old Japanese boy (gray line) on the sella-nasion plane at sella.
Fig 3. Pretreatment photographs.

Fig 4. Pretreatment dental casts.
with midfacial hypoplasia. However, there are few reports of combining LeFort I and II osteotomies for segmental distraction osteogenesis in patients with craniosynostosis.

This article demonstrates a good prognosis for a patient with craniosynostosis treated with a modified LeFort II segmental distraction osteogenesis by using a rigid external distraction osteogenesis system.

Fig 5. Radiographs and cephalometric tracings at pretreatment: A, lateral cephalograph; B, panoramic radiograph; C, tracing (dotted line) superimposed with the initial tracing (solid line) on the sella-nasion plane at sella.

Fig 6. Photographs after placing the rigid external distraction device system.
DIAGNOSIS AND ETIOLOGY

An 8-year-old boy came to the outpatient clinic of Tokushima University hospital in Japan. His chief complaint was an anterior crossbite. He had been diagnosed with a craniosynostosis subtype at birth in the Hyogo Prefecture Children's Hospital and had an operation for hydrocephalus. Furthermore, he had a medical history of anal atresia, otitis media, cryptorchism testis, hypoplasia of the elbow, and an auditory disturbance. A dished-in face, mild exorbitism, and a short upper lip due to midfacial hypoplasia were noted (Fig 1). He had mild obstructive sleep apnea syndrome in a dorsal position. An anterior crossbite with an overjet of –0.2 mm and an overbite of –1.6 mm were observed. The terminal plane was a mesial step type on both sides. Severe crowding was expected because of the lack of eruption spaces for the permanent teeth (Fig 2). Cephalometric analysis, when compared with the Japanese norm, showed a skeletal Class III jaw-base relationship (ANB, –2.0°) because of a severe maxillary deficiency (SNA, 64.9°). In the mandible, the ramus height was within the normal range, but the body length was significantly short. In the panoramic radiograph, a mediiodens was observed (Fig 2).

TREATMENT OBJECTIVES

The patient was diagnosed as having a Class III malocclusion, with a skeletal Class III jaw relationship caused by midfacial hypoplasia. The treatment objectives were to (1) correct the midfacial hypoplasia and the dished-in face, (2) correct the anterior crossbite and establish ideal overjet and overbite, and (3) achieve
Fig 8. Cephalometric tracings made before (black line) and after (red line) the distraction osteogenesis: A, superimposition on the sella-nasion plane at sella; B, on the palatal contour; C, on the mandibular plane at menton.

Table. Cephalometric summary

<table>
<thead>
<tr>
<th>Variable</th>
<th>Japanese norm*</th>
<th>SD</th>
<th>Initial consultation</th>
<th>Pretreatment</th>
<th>Predistraction</th>
<th>Postdistraction</th>
<th>Posttreatment</th>
<th>1 year posttreatment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Male adult</td>
<td></td>
<td>8 y 7 mo</td>
<td>13 y 6 mo</td>
<td>14 y 11 mo</td>
<td>15 y 2 mo</td>
<td>16 y 10 mo</td>
<td>17 y 8 mo</td>
</tr>
<tr>
<td>Angular (°)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ANB</td>
<td>3.2</td>
<td>2.4</td>
<td>-2.0</td>
<td>-4.1</td>
<td>-3.7</td>
<td>+3.2</td>
<td>+2.6</td>
<td>+2.4</td>
</tr>
<tr>
<td>SNA</td>
<td>81.5</td>
<td>3.3</td>
<td>64.9</td>
<td>65.2</td>
<td>65.4</td>
<td>71.2</td>
<td>70.1</td>
<td>69.9</td>
</tr>
<tr>
<td>SNB</td>
<td>78.2</td>
<td>4.0</td>
<td>66.9</td>
<td>69.3</td>
<td>69.1</td>
<td>68.0</td>
<td>67.5</td>
<td>67.5</td>
</tr>
<tr>
<td>MP-SN</td>
<td>34.5</td>
<td>6.1</td>
<td>56.6</td>
<td>53.0</td>
<td>52.5</td>
<td>53.7</td>
<td>54.0</td>
<td>54.4</td>
</tr>
<tr>
<td>Gonial angle</td>
<td>120.9</td>
<td>6.5</td>
<td>135.3</td>
<td>135.4</td>
<td>133.3</td>
<td>134.4</td>
<td>133.3</td>
<td>134.3</td>
</tr>
<tr>
<td>U1-SN</td>
<td>106.0</td>
<td>7.5</td>
<td>99.7</td>
<td>118.1</td>
<td>101.5</td>
<td>100.9</td>
<td>96.8</td>
<td>98.2</td>
</tr>
<tr>
<td>L1-MP</td>
<td>95.2</td>
<td>6.2</td>
<td>79.4</td>
<td>82.2</td>
<td>92.6</td>
<td>82.3</td>
<td>89.1</td>
<td>87.9</td>
</tr>
<tr>
<td>Convexity</td>
<td>173.2</td>
<td>5.5</td>
<td>179.8</td>
<td>175.2</td>
<td>170.5</td>
<td>172.4</td>
<td>172.5</td>
<td>172.5</td>
</tr>
<tr>
<td>Interincisal angle</td>
<td>124.2</td>
<td>8.6</td>
<td>124.3</td>
<td>106.7</td>
<td>113.4</td>
<td>123.1</td>
<td>121.2</td>
<td>119.9</td>
</tr>
<tr>
<td>Occlusal plane</td>
<td>15.5</td>
<td>4.2</td>
<td>27.9</td>
<td>22.6</td>
<td>28.1</td>
<td>28.9</td>
<td>30.2</td>
<td>30.5</td>
</tr>
<tr>
<td>Linear (mm)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rhinion to VRP</td>
<td>-</td>
<td></td>
<td>52.0</td>
<td>53.8</td>
<td>54.5</td>
<td>70.7</td>
<td>69.1</td>
<td>69.3</td>
</tr>
<tr>
<td>Point A to VRP</td>
<td>-</td>
<td></td>
<td>39.4</td>
<td>40.7</td>
<td>41.9</td>
<td>47.8</td>
<td>47.6</td>
<td>47.4</td>
</tr>
<tr>
<td>Pog to VRP</td>
<td>-</td>
<td></td>
<td>15.9</td>
<td>21.0</td>
<td>21.0</td>
<td>20.4</td>
<td>20.5</td>
<td>20.1</td>
</tr>
<tr>
<td>S-N</td>
<td>72.2</td>
<td>3.3</td>
<td>63.5</td>
<td>66.2</td>
<td>66.7</td>
<td>66.7</td>
<td>69.1</td>
<td>69.8</td>
</tr>
<tr>
<td>N-Me</td>
<td>135.7</td>
<td>4.0</td>
<td>117.4</td>
<td>123.1</td>
<td>123.5</td>
<td>125.3</td>
<td>126.3</td>
<td>126.2</td>
</tr>
<tr>
<td>Me/palatal plane</td>
<td>74.6</td>
<td>3.0</td>
<td>66.0</td>
<td>68.1</td>
<td>67.8</td>
<td>67.8</td>
<td>67.1</td>
<td>67.3</td>
</tr>
<tr>
<td>Ar-Go</td>
<td>53.2</td>
<td>5.7</td>
<td>40.0</td>
<td>46.2</td>
<td>45.6</td>
<td>46.8</td>
<td>45.2</td>
<td>45.4</td>
</tr>
<tr>
<td>Ar-Me</td>
<td>115.6</td>
<td>6.8</td>
<td>84.0</td>
<td>93.1</td>
<td>92.8</td>
<td>94.6</td>
<td>94.3</td>
<td>94.2</td>
</tr>
<tr>
<td>Go-Me</td>
<td>76.6</td>
<td>4.4</td>
<td>49.5</td>
<td>53.4</td>
<td>53.9</td>
<td>54.3</td>
<td>55.1</td>
<td>55.2</td>
</tr>
<tr>
<td>Overjet</td>
<td>3.3</td>
<td>1.0</td>
<td>-0.2</td>
<td>+0.8</td>
<td>-1.3</td>
<td>+5.1</td>
<td>+4.0</td>
<td>+3.9</td>
</tr>
<tr>
<td>Overbite</td>
<td>3.3</td>
<td>1.7</td>
<td>-1.6</td>
<td>-0.4</td>
<td>-1.0</td>
<td>+2.0</td>
<td>+2.6</td>
<td>+2.9</td>
</tr>
</tbody>
</table>

VRP, Vertical reference plane (line perpendicular to SN line through S).
*Wada et al.19
Fig 9. Posttreatment photographs.

Fig 10. Prostreatment dental casts.
an acceptable occlusion with a good functional Class I occlusion.

We planned a segmental distraction osteogenesis with a modified LeFort II osteotomy to improve both the facial profile and the skeletal jaw relationship. The midfacial advancement was performed on 2 levels: the upper segment including the anterior nasal spine and the lower segment consisting of the maxillary dentition.

**TREATMENT ALTERNATIVES**

Several procedures were explored to achieve a proper facial profile and an acceptable occlusion. A conservative treatment of maxillary growth modification by using a maxillary protraction headgear during the growing phase was considered to be effective to modify his skeletal Class III jaw relationship; however, this procedure might not have corrected his severe skeletal disharmony and obstructive sleep apnea syndrome completely, and it seemed to be a compromised treatment. Multi-bracket treatment with tooth extraction after his growth spurt could be useful for correcting his Class III malocclusion with a negative overjet, but it would not improve his concave profile caused by the midfacial deficiency. Therefore, we chose an orthodontic-surgical treatment for this patient.

In growing patients with craniosynostosis, traditional surgical-orthodontic treatments provide good prognoses, but we planned the distraction osteogenesis after his growth spurt for the following reasons: (1) the anterior crossbite was not observed when the first molars erupted, (2) some premolars needed to be extracted to improve the severe crowding, and (3) the distance and direction of the required advancement of the distraction osteogenesis were more predictable in adolescence than in childhood.

**TREATMENT PROGRESS**

After the extraction of the mediiodens, we followed the patient until the end of his active growth (Figs 3-5). At the age of 13 years 6 months, his maxillary
second premolars and mandibular first premolars were extracted to gain eruption space for the canines. An 0.018-in slot preadjusted edgewise appliance was placed in both arches, and leveling and alignment with nickel-titanium archwires were started. He showed a skeletal Class III jaw relationship with an ANB angle of −4.1° at that time.

After 1 year of preoperative orthodontic treatment, segmental distraction osteogenesis was performed at the Department of Plastic and Reconstructive Surgery, Yokohama City University Graduate School of Medicine. After the modified LeFort I and II osteotomies, the maxillary segment was advanced approximately 7 mm, including overcorrection, and fixed to the zygomatic bone. Then a rigid external distraction system was placed on the cranial bone at the same time (Fig 6). The segmental distraction osteogenesis was started 6 days after surgery and continued for 20 days at a rate of 1.0 mm per day. After the osteogenesis, the rigid external distraction device was removed, and Kirschner wires were inserted between the nasal segment and the zygomatic bone for retention.

At the age of 16 years 10 months, the edgewise appliances were removed, and circumferential retainers and lingual bonded retainers were placed in both arches. The total active treatment period was 40 months.

TREATMENT RESULTS

After the segmental distraction osteogenesis, the midfacial hypoplasia and the facial profile were dramatically improved (Fig 7). Cephalometric evaluation after the segmental distraction osteogenesis showed nasal advancement of 16.2 mm at rhinion and maxillary advancement of 5.9 mm at Point A to the reference line, which was defined as a perpendicular line to the sella-nasion plane through sella (Fig 8). A skeletal Class I jaw relationship was also achieved (ANB, 3.2°). Because the maxilla was moved in a forward and downward direction, the mandibular plane angle was increased by 1.2°. The negative overjet was overcorrected to 5.1 mm, and the anterior open bite was improved (overbite, 2.0 mm; Table).

At the end of active orthodontic treatment, at 1.5 years after the segmental distraction osteogenesis, an ideal occlusion having a Class I molar relationship with an adequate interincisal relationship was achieved (overjet, 4.0 mm; overbite, 2.6 mm; Figs 9 and 10). The cephalometric evaluation showed a skeletal Class I jaw relationship (ANB, 2.6°), and the anterior cranial base (S-N) was increased to 70.1 mm (Fig 11). Although the segment moved by the modified LeFort II osteotomy relapsed posteriorly by 1.4 mm at rhinion relative to the reference line, the maxillary segment was almost stable, and the facial profile was maintained (Table).

After 1 year of retention, at 2.5 years after the segmental distraction osteogenesis, both the nasal and the maxillary segments were anteroposteriorly stable relative to the reference line. The skeletal Class I jaw relationship was retained (ANB, 2.4°), and the facial profile and occlusion were acceptable (Figs 12-14; Table).

DISCUSSION

Our patient had a severe midfacial deficiency caused by craniosynostosis. In such patients, midfacial advancement with a LeFort II or III osteotomy has been used for several decades, because it could provide significant improvements of both the facial profile and the occlusion. However, our patient had a mild Class III malocclusion because of an accompanying mandibular deficiency. Therefore, 2-jaw advancement surgery was considered to be the ideal approach. The 2-jaw surgery could provide acceptable results with long-term stability; however, the medical costs are high, and the procedure requires a long surgery and causes severe postoperative discomfort for the patient. 13,14 Additionally, mandibular retrusion is generally more favored than mandibular protrusion, and bimaxillary protrusion is relatively attractive and well accepted in the Japanese population. 15 Consequently, we chose midfacial surgery without mandibular advancement to reduce the strain on the patient.

As for distraction devices, we selected a rigid external distraction system because this device has several advantages compared with the internal device: a simple placement procedure, flexibility in controlling the protraction direction and amount at the various points, and no second operation to remove the device. 16 Several reports have indicated the usefulness of distraction osteogenesis in patients with craniosynostosis. Matsumoto et al 10 treated an adult craniosynostosis patient with LeFort I and III osteotomies using an external distraction device. Satoh et al 11 suggested that, in midfacial distraction for adults and adolescents, mobilizing 2 parts of the midface differently with both internal and external distraction devices instead of using monobloc distraction is preferable both esthetically and functionally.

In our patient, LeFort II osteotomy was the first choice to improve the midfacial hypoplasia. However, the anteroposterior jaw–base relationship might become a skeletal Class II relationship if the nasomaxillary complex were moved anteriorly to obtain sufficient improvement of his facial profile. Then we performed the segmental distraction osteogenesis after the modified LeFort I and II osteotomies, which could provide 2 amounts of advancement in the nasal and maxillary segments, respectively.
Fig 12. Postretention photographs.

Fig 13. Postretention dental casts.
As a result of the modified LeFort II segmental distraction osteogenesis, his facial profile affected by the severe midfacial hypoplasia was improved dramatically. Additionally, the maxillary segment was predictably advanced with the LeFort I osteotomy and fixed to the zygomatic bone, and an acceptable Class I occlusion was achieved. In comparison with the cephalometric tracings before and after the segmental distraction osteogenesis, the maxillary segment was moved in a downward direction, and the mandible was slightly rotated clockwise. Well-balanced facial proportions and a skeletal Class I jaw relationship were achieved without the nasal deformity. These results suggest that segmental distraction osteogenesis after modified LeFort I and II osteotomies is quite useful to treat patients with severe midfacial hypoplasia and mild to moderate Class III malocclusion.

The nasal segment was advanced approximately 20 mm with a rigid external distraction device during the segmental distraction osteogenesis procedure. However, the amount of advancement was reduced to 16.2 mm immediately after removal of the rigid external distraction device. Additionally, the segment showed relapse of about 2 mm during the 1.5 years after the segmental distraction osteogenesis, although it was stable in the succeeding year. These results suggest the requirement of overcorrection in the midfacial distraction osteogenesis to maintain a well-balanced profile. Several previous reports also indicated the necessity of overcorrection of the LeFort II distraction osteogenesis. On the other hand, the maxillary segment that was advanced by the LeFort I osteotomy was nearly stable through all observation periods. Therefore, we propose that this modified midfacial advancement method, which fixes the maxillary segment to the zygoma before the midfacial distraction osteogenesis, provides a more stable and predictable occlusion compared with a single distraction osteogenesis procedure. However, the long-term stability of this procedure is still unknown. Further studies are required to understand its long-term implications.

CONCLUSIONS

We reported the successful treatment of a patient with midface hypoplasia due to craniosynostosis, treated by a modified LeFort II segmental distraction osteogenesis, although it was stable in the succeeding year. These results suggest the requirement of overcorrection in the midfacial distraction osteogenesis to maintain a well-balanced profile. Several previous reports also indicated the necessity of overcorrection of the LeFort II distraction osteogenesis. On the other hand, the maxillary segment that was advanced by the LeFort I osteotomy was nearly stable through all observation periods. Therefore, we propose that this modified midfacial advancement method, which fixes the maxillary segment to the zygoma before the midfacial distraction osteogenesis, provides a more stable and predictable occlusion compared with a single distraction osteogenesis procedure. However, the long-term stability of this procedure is still unknown. Further studies are required to understand its long-term implications.

CONCLUSIONS

We reported the successful treatment of a patient with midface hypoplasia due to craniosynostosis, treated by a modified LeFort II segmental distraction osteogenesis, although it was stable in the succeeding year. These results suggest the requirement of overcorrection in the midfacial distraction osteogenesis to maintain a well-balanced profile. Several previous reports also indicated the necessity of overcorrection of the LeFort II distraction osteogenesis. On the other hand, the maxillary segment that was advanced by the LeFort I osteotomy was nearly stable through all observation periods. Therefore, we propose that this modified midfacial advancement method, which fixes the maxillary segment to the zygoma before the midfacial distraction osteogenesis, provides a more stable and predictable occlusion compared with a single distraction osteogenesis procedure. However, the long-term stability of this procedure is still unknown. Further studies are required to understand its long-term implications.

CONCLUSIONS

We reported the successful treatment of a patient with midface hypoplasia due to craniosynostosis, treated by a modified LeFort II segmental distraction osteogenesis, although it was stable in the succeeding year. These results suggest the requirement of overcorrection in the midfacial distraction osteogenesis to maintain a well-balanced profile. Several previous reports also indicated the necessity of overcorrection of the LeFort II distraction osteogenesis. On the other hand, the maxillary segment that was advanced by the LeFort I osteotomy was nearly stable through all observation periods. Therefore, we propose that this modified midfacial advancement method, which fixes the maxillary segment to the zygoma before the midfacial distraction osteogenesis, provides a more stable and predictable occlusion compared with a single distraction osteogenesis procedure. However, the long-term stability of this procedure is still unknown. Further studies are required to understand its long-term implications.
osteogenesis with a rigid external distraction device. As a result of the modified segmental distraction osteogenesis, significant improvement of severe midfacial hypoplasia was achieved without excessive advancement of the maxillary dentition. In the view of the stability and retention, some overcorrection in midfacial advancement is recommended.

REFERENCES