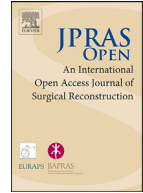




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Case Report

Le fort II distraction osteogenesis with a hybrid system for an Apert syndrome patient: A case report

Shinji Kobayashi^{a,*}, Toshihiko Fukawa^b, Yuichiro Yabuki^{a,c},
Toshihiko Satake^c, Jiro Maegawa^c

^a Department of Plastic and Reconstructive Surgery, Kanagawa Children's Medical Center Japan

^b Fukawa Orthodontic Office Japan

^c Department of Plastic and Reconstructive Surgery, Yokohama City University Hospital Japan

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ABSTRACT

Background: Le Fort II advancement is considered for normalizing the facial appearance in Apert syndrome. When these procedures are performed during growth, overcorrection of midface advancement is required. We developed a system that can control the distance and vector of movement for the central midface to create more normal facial proportions. This case report shows Le Fort II distraction osteogenesis with this hybrid system for an Apert syndrome patient.

Case: The patient was a girl with Apert syndrome with midfacial-nose hypoplasia and skeletal class III malocclusion. She was healthy without respiratory problems and had no learning disabilities. She underwent our Le Fort II distraction osteogenesis with the hybrid system at 10 years and 6 months of age. Her midface was elongated 22 mm at point Or forward and moved 5° downward to the Frankfort horizontal plane compared to the standard position of average Japanese adult women on the cephalogram. Examining the facial image, the midfacial depression was improved 4 years after the operation.

Discussion: Overcorrection of midface advancement is required for patients to reduce the number of procedures during growth. The system that we developed could control the distance and vector of

* Corresponding author.

E-mail address: skobayashi@kcmc.jp (S. Kobayashi).

movement steadily when the central midface was overcorrected to try to create normal adult facial proportions.

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Background

Le Fort advancement is considered for normalizing the facial appearance of patients with Apert syndrome. There are a wide variety of reported procedures, including Le Fort II/III, bipartition method, and simultaneous zygomatic repositioning, for the management of the facial deformity in Apert syndrome.^{1–5} In any case, when these procedures are performed during growth, overcorrection of midface advancement is required for the patients. The aim is that the facial profile be close to the adult one and that repeat operations be avoided, though it would be difficult to avoid Le Fort I/mandibular osteotomy. An overcorrection would need to be advanced to the adult profile as accurately as possible. We developed a system that can control the distance and vector of movement for the central midface to create more normal facial proportions. This case report shows Le Fort II distraction osteogenesis with the hybrid system for an Apert syndrome patient.

Case

This study was approved by Kanagawa Children's Medical Center's institutional review board (Approval number: 61–02). Informed consent from the patient's parents/guardians for undergoing the procedure and publishing images was obtained. The patient was a girl with Apert syndrome. She had midfacial-nose hypoplasia with skeletal class III malocclusion. She was healthy and had no learning disabilities. She had already undergone fronto-orbital advancement twice, and her cranium was sufficient to be expanded. There was marked convexity along the absorbable plates fixed onto her forehead in the second advancement (Figures 1, 2).

She and her parents expressed a strong desire to have surgery to improve her face, including her occlusion, though she had no respiratory problems. Therefore, she underwent Le Fort II distraction osteogenesis with our hybrid system at 10 years and 6 months of age (Figure 3). The Le Fort II osteotomy through the infraorbital rim was performed at the outer side of the infraorbital foramen as much as possible. When the midface was totally freed from the bone attachment, a pair of our own internal distractors was placed. Then, an external distraction device (MEDICAL U&A, Osaka, Japan) was attached, and a total of 6 surgical wires (2 fixed to an internal distraction device, 2 fixed to the piriform aperture, and 2 fixed to the upper portion of the molar) were attached to the external devices.^{6,7} The absorbable plates on her forehead were removed during the operation.

The distance and vector of the midface advancement were determined by reference to the cephalogram of an average Japanese adult.⁸ Finally, her midface was elongated 22 mm at point Or forward and moved 5° downward to the Frankfort horizontal plane compared to the standard position of average Japanese adult women on the cephalogram. Examining the facial image, the midfacial depression including the morphology of the external nose, was improved 4 years after the operation (Figures 1,2,4).

Discussion

Le Fort advancement for Apert syndrome is considered for resolving the exorbitism and improving respiratory problems. Some patients without respiratory problems or with only mild problems might desire an operation to normalize their facial appearance. We consider that this procedure is indicated



Figure 1. A female patient with Apert syndrome at 10 years and 6 months of age.
Left: Preoperative view and occlusion.
Right: Postoperative view and occlusion 4 years after the operation.

when the patient is older than 6 years of age before elementary school. In the process of growing, overcorrection of midface advancement is required for these patients to reduce the number of procedures. In such cases, it would be more desirable that the facial profile be close to the adult one, because repeat Le Fort II/III could be avoided, though it would be difficult to avoid Le Fort I/mandibular osteotomy. The system that we developed could control the distance and vector of movement steadily when the central midface was overcorrected as accurately as possible to create normal adult facial proportions.

As for Apert syndrome, the lateral zygoma and central midface require a different distance and vector of movement to create more normal facial proportions. Therefore, ideally, Le Fort II distraction and simultaneous zygomatic repositioning as a combined procedure can move the lateral orbito-zygomatic complex and central midface an independent distance and vector.^{9,10} In this case, Le Fort II osteotomy was selected because the patient's zygoma developed comparatively more, and repeat Le Fort II osteotomy would not be expected when she reached adulthood. In any case, careful follow-up is needed until she reaches adulthood, especially of her maxillary growth.

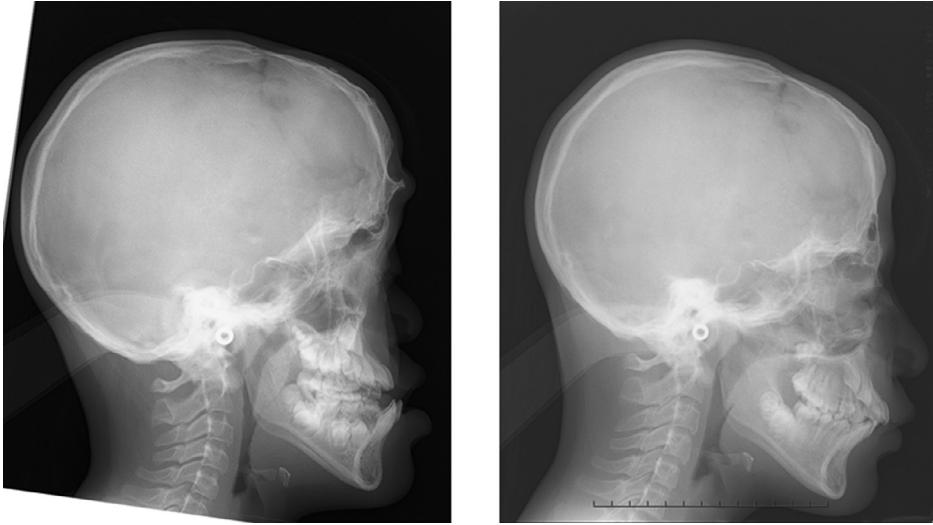


Figure 2. Left: Preoperative cephalogram.
Right: Postoperative cephalogram.

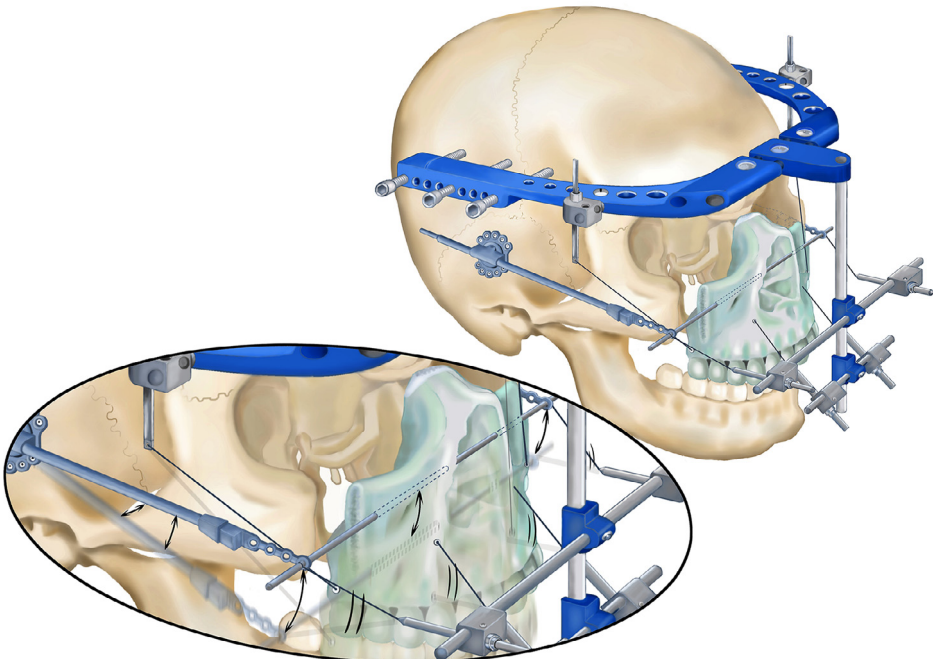


Figure 3. Illustration of our distraction system for Le Fort II.

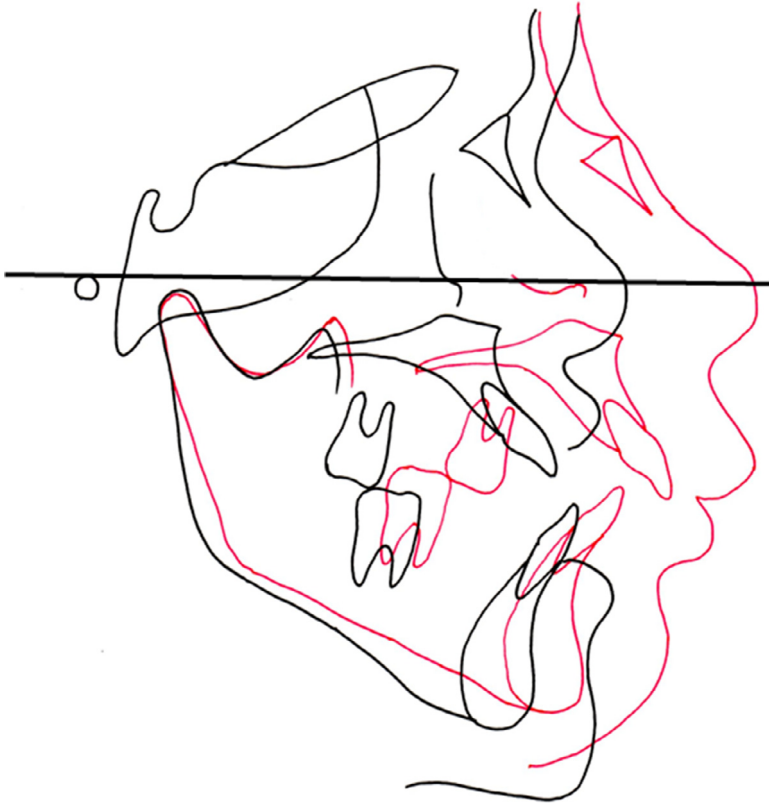


Figure 4. Superimposition of the preoperative and postoperative cephalometrics. Black line: before surgery, Red line: after surgery.

Conclusion

Le Fort II distraction osteogenesis with a hybrid system for Apert syndrome was performed for a patient at the age of 10 years.

Declaration of Competing Interest

None.

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Ethical approval

This study was approved by Kanagawa Children's Medical Center's institutional review board (Approval number: 61–02)

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