Corrected Cephalometric Analysis to Determine the Distance and Vector of Distraction Osteogenesis for Syndromic Craniosynostosis

Shinji Kobayashi, MD*
Toshihiko Fukawa, DDS*†
Takashi Hirakawa, DDS‡
Toshihiko Satake, MD§
Jiro Maegawa, MD§

Background: The purpose of this study was to confirm the utility of a corrected cephalometric analysis to facilitate the planning of distraction osteogenesis with Le Fort III osteotomy for syndromic craniosynostosis.
Methods: This prospective study involved 4 male and 2 female patients (mean patient age, 8 years 9 months; age range, 4 years 6 months to 13 years 2 months) with Crouzon syndrome who were treated with Le Fort III maxillary distraction using our previously described system of analysis of a corrected cephalogram and who underwent clinical follow-up. Lateral cephalograms were obtained immediately after device removal.
Results: Distraction of orbitale moved the vector downward to the adult profile, but there was slightly less elongation than the adult profile for the distraction distance. The desired and real mean angles after distraction of point A were 29.2 ± 7.9° and 6.1 ± 8.5°, respectively, and the desired and the real mean distances after distraction of point A were 30.6 ± 12.7 mm and 29.4 ± 4.1 mm, respectively.
Conclusions: Using the corrected cephalometric analysis, the distance and vector of distraction osteogenesis with Le Fort III osteotomy could be determined in patients with syndromic craniosynostosis. The distraction system brought the patients' facial bones to the planned position using controlling devices. *(Plast Reconstr Surg Glob Open 2017;5:e1482; doi: 10.1097/GOX.0000000000001482; Published online 6 September 2017.)*

BACKGROUND

Treatment of syndromic craniosynostosis is critically dependent on the successful advancement of the midface. Distraction osteogenesis for syndromic craniosynostosis has been applied extensively to all components of the craniofacial skeleton with favorable results since its first application to lengthen the human mandible.[1–11]

*From the *Department of Plastic and Reconstructive Surgery, Kanagawa Children's Medical Center, Yokohama, Japan; †Fukawa Orthodontic Office, Kamakura, Japan; ‡Hirakawa Orthodontic Clinic, Yokohama, Japan; and §Department of Plastic and Reconstructive Surgery, Yokohama City University Hospital, Yokohama, Japan.*
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However, the distance and direction of overcorrection for the midface are difficult to determine accurately in younger patients, although there are some reports that recommend as anterior an overcorrection as possible in younger children.[12–14] The usual cephalometric analysis, which was used to assess changes in the position of the midface, was based on the use of a reference like the sella or the sella-nasion line. However, such an analysis does not provide an adequate evaluation of craniofacial advancement relapse because these parameters are variably altered in cases of relapse and not stable during distraction.[15,16]

We developed a corrected cephalometric analysis to determine the distance and vector of distraction osteogenesis to overcome this problem. The purpose of this study was to confirm the utility of corrected cephalometric analysis to move the profile as close to an adult profile as possible, to reduce the number of distraction osteogenesis with Le Fort III osteotomy throughout life for syndromic craniosynostosis and to evaluate the stability and/or resultant change in the facial features over a period of 1 year or more after removal of the distraction device.

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METHODS

This prospective study was approved by Kanagawa Children's Medical Center's institutional review board (Approval number: 61-02). Informed consent from the patients' parents/guardians for undergoing the procedure and publishing images was obtained. Only patients who were treated from 2010 to 2014 with Le Fort III maxillary distraction using our previously described system of analysis of a corrected cephalogram and who underwent clinical follow-up were included in the study (Fig. 1).[17,18] Four male and 2 female patients with Crouzon syndrome were treated during the study period. Mean patient age was 8 years and 9 months at the time of osteotomy and distraction (range, 4 years and 6 months to 13 years and 2 months; Table 1).

Corrected Cephalometric Analysis

Manually traced lateral cephalograms were obtained preoperatively. Twelve cephalometric landmarks [Point A (A), anterior nasal spine (Ans), articulare (Ar), point B (B), gonion (G), lower incisor incisal edge (L1), menton (Me), nasion (N), orbitale (Or), pogonion (Pog), sella turcica (S), and upper incisor incisal edge (U1)] situated in the midsagittal plane were identified and digitized parallel to the Frankfort horizontal plane (FHP). Lateral cephalograms of average normal Japanese adults (CANJA) are generally used as the standard at the relevant departments by most orthodontists in Japan, with an average age for males and females of 23 years and 7 months, and 19 years and 7 months,[19] respectively, and patients' lateral cephalograms obtained not more than 1 month before distraction (T0) were superimposed. The x axis was oriented parallel to the FHP, and the y axis was perpendicular to the x axis. The tracings were positioned along the x and y axes, with the sella registered at 0 in this coordinate system. After T0 was superimposed on CANJA at the sella, T0 was moved back parallel to the FHP, to the position of Ar` where Ar of T0 corresponded to a perpendicular line parallel to the y axis at Ar of CANJA. The newly moved back T0 was defined as the corrected T0 (cT0). Then, the distance and the vector between cT0 and CANJA at Or and A were measured directly. Finally, the distance and vector of distraction osteogenesis were determined by referring to the vector from Or of cT0 to Or of CANJA and the distance between them (Fig. 2).

Midface Advancement

All patients underwent Le Fort III osteotomy with our midface advancement system using a rigid external and internal distraction device.[17,18] The device was elongated at a rate of 1 mm per day. The anteroposterior and vertical positions of the maxilla were controlled by differentially activating the superior, inferior, and vertical wire attachments on the device, and check lateral cephalograms were obtained every week. The distraction was concluded by the average duration of distraction from 4 to 6 weeks after sufficient overcorrected maxillary advancement was achieved relative to the planned cephalometric position.

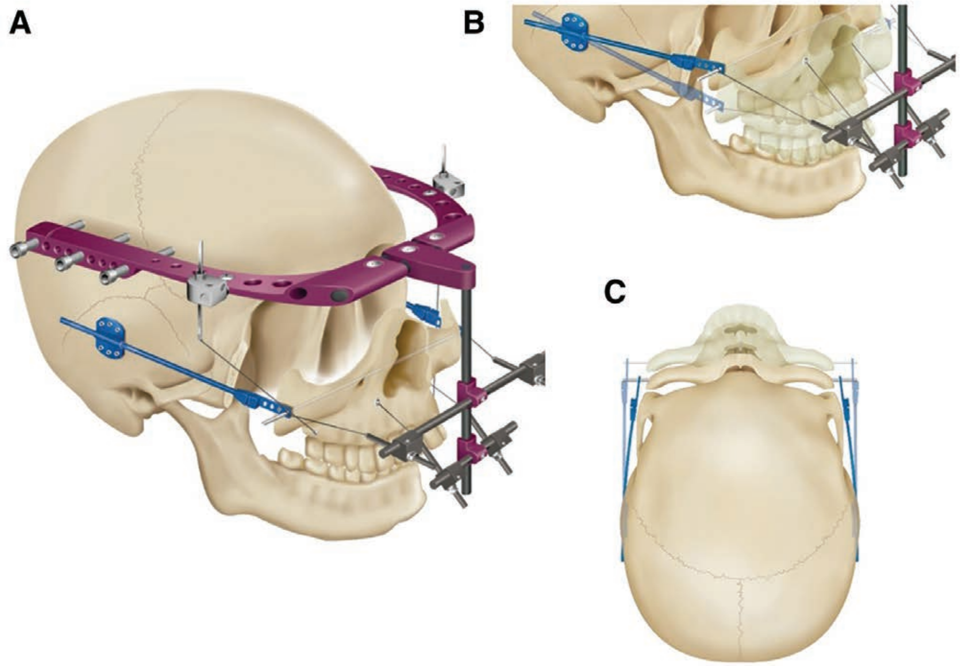

Fig. 1. A, Maxillary distraction technique with internal and external devices. B, The internal distraction device has an adjustable angle, while the external distraction device enables control of the distraction distance via the surgical wires. Additionally, the device that can control the direction of the advanced maxilla vertically is attached to the external distraction device. C, The angle of the internal distraction device's fixation position on the temporal bone can be altered by 5–15°.

Table 1. Patients' Data

Number	Gender	Age of Le Fort III
1	M	9 years and 6 months
2	M	5 years and 0 months
3	M	6 years and 3 months
4	M	12 years and 9 months
5	F	13 years and 2 months
6	F	4 years and 6 months

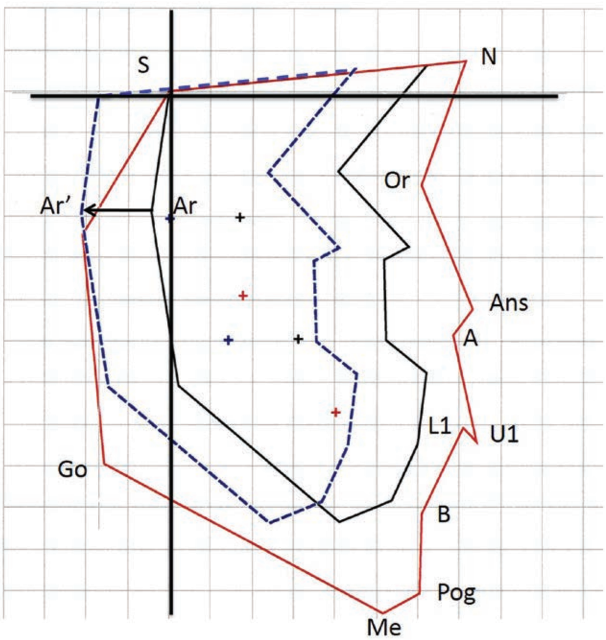

Fig. 2. A corrected cephalometric analysis. Lateral cephalogram (T0, *black*), corrected lateral cephalogram (cT0, *dotted blue*), and a cephalogram averaged from normal Japanese adults (CANJA; *red*) were superimposed. Black arrow: the distance from Ar to Ar' where Ar of T0 corresponds to a perpendicular line parallel to the *y* axis at Ar of CANJA. A: point A. The most posterior midline point in the concavity between the ANS and the prosthion (the most inferior point on the alveolar bone overlying the maxillary incisors). Ans: anterior nasal spine. The anterior tip of the sharp bony process of the maxilla at the lower margin of the anterior nasal opening. Ar: the intersection of the posterior margin of the mandibular condyle and the inferior border of the temporal bone. B: point B. The most posterior midline point in the concavity of the mandible between the most superior point on the alveolar bone overlying the mandibular incisors (infradentale) and pogonion (the most anterior point on the chin). Go: Gonion. Point created on the mandibular angle by a line bisecting the angle formed by planes tangent to the branch and to the body of the mandible. L-1: the incisal tip of the mandibular central incisor. Me: menton. The lowest point on the symphyseal shadow of the mandible seen on a lateral cephalogram. N: nasion. The most anterior point on the frontonasal suture in the midsagittal plane. Or: orbitale. Lowest point of orbit. Pog: Pogonion. The most anterior point on the contour of the bony chin, as determined by a tangent through the nasion. S: sella. The geometric center of the pituitary fossa. U-1: the incisal tip of the maxillary central incisor.

Postoperative Cephalometric Analysis

Lateral cephalograms were obtained twice after removing the devices: immediately after removing the devices (T1) and 1 year after removing them (T2).

The cT0, T1, and T2 cephalometric tracings were superimposed at the sella, the anterior and posterior cranial bases, and the anterior contours of the middle cranial fossae. All relevant clinical data were obtained from our medical center.

RESULTS

The mean differences in the 12 points were directly measured, including Or and A, between lateral cephalograms of cT0 and CANJA, cT0 and T1, and T1 and T2. Linear displacement of the points during the period between 2 measurements was also calculated (Tables 2, 3; Figs. 3–18).

cT0 and CANJA Measurements

The mean distances that Or and A were expected to advance between cT0 and CANJA were 25.4 ± 7.8 mm and 30.6 ± 12.7 mm, respectively. The angle viewed from the FHP showed that the Or of cT0 was expected to move downward to the Or of CANJA by 2.8 ± 15.9° and that A of cT0 was expected to move downward to A of CANJA by 29.2 ± 7.9°.

cT0 and T1 Measurements

The mean distances that Or and A were actually clinically advanced between cT0 and T1 were 19.8 ± 4.0 mm and 29.4 ± 4.1 mm, respectively. The angle viewed from the FHP showed that the Or of cT0 was moved downward to the Or of T1 by 3.3 ± 8.9°, and A of cT0 was moved downward to A of T1 by 6.1 ± 8.5°, which was significantly different from the planned change of 29.2 ± 7.9°.

T1 and T2 Measurements

The mean distances by which Or and A changed between T1 and T2 were 0.8 ± 1.6 mm and 2.1 ± 2.9 mm, respectively. The angle viewed from FHP showed that the Or of T1 changed downward to the Or of T2 by 1.3 ± 2.1°, and A of T1 changed downward to A of T2 by 2.6 ± 3.5°. The midface grew slightly downward at A.

DISCUSSION

We developed a corrected cephalometric analysis to superimpose the patient's cephalogram to the normal child's one to determine the distance and vector of distraction osteogenesis for craniosynostosis syndrome. However, it was difficult to advance the patients' faces along the planned position accurately. On the other hand, maxillary retrusion was not recognized over a period of 1 year or more after removal of the distraction device.

The distance and direction of overcorrection of the midface required in patients with craniosynostosis syndrome has not been known. From a psychological perspective, it is recommended to not distract to the final adult maxillary position.[20] Patients might not be able to accept their facial appearance, which would probably change dramatically. Instead, the midface should be advanced as much as possible, especially at the level of the orbitozygomatic components, while avoiding a signifi-

Table 2. Differences in Cephalometric Measurements between CANJA, cT0, and T1 at Or

Case	Distance from cT0 to CANJA at Or (mm)	Distance from cT0 to T1 at Or (mm)	Distance from T1 to T2 at Or (mm)	cT0 to CANJA Downward Angle at Or (degree)	cT0 to T1 Downward Angle at Or (degree)	T1 to T2 Downward Angle at Or (degree)
1	21.5	21	1	23.5	9.5	2.5
2	28	14.5	4	6.5	6	5
3	37	21.5	0	5.5	-11.5	0
4	16.5	23	0	-9	2	0
5	19	15	0	-21.5	0	0
6	30.5	23.5	0	12	14	0
Mean ± SD	25.4±7.8	19.8±4.0	0.8±1.6	2.8±15.9	3.3±8.9	1.3±2.1

Table 3. Differences in Cephalometric Measurements between CANJA, cT0, and T1 at A

Case	Distance from cT0 to CANJA at A (mm)	Distance from cT0 to T1 at A (mm)	Distance from T1 to T2 at A (mm)	cT0 to CANJA Downward Angle at A (degree)	cT0 to T1 Downward Angle at A (degree)	T1 to T2 Downward Angle at A (degree)
1	30	32	2	34	4	1.5
2	42.5	34	7	35.5	0	8
3	38	32	4	29	-4	6
4	19	24	0	35.5	14.5	0
5	12	25	0	15	4	0
6	42	29.5	0	26	18	0
Mean ± SD	30.6±12.7	29.4±4.1	2.1±2.9	29.2±7.9	6.1±8.5	2.6±3.5

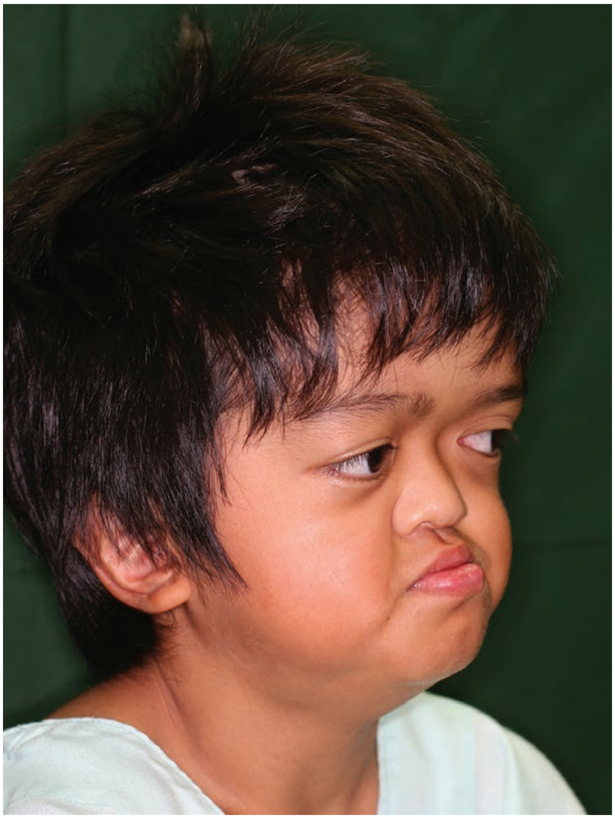

Fig. 3. A 9-year-old male patient with Crouzon syndrome. Preoperative view. He has severe maxillary retrusion.

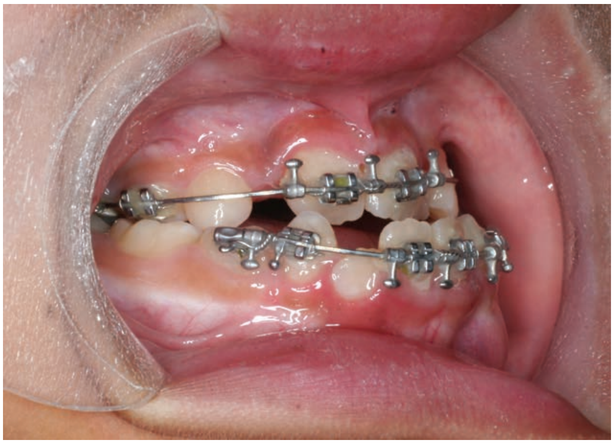

Fig. 4. Preoperative view. He has a class III malocclusion.

cantly abnormal appearance.[14] Some authors use Baumrind's longitudinal data (porion to orbitale distance) to determine the postoperative position of the orbit.[21] We believe that the patients' profiles should be overcorrected as anterior as possible and moved anteriorly to as close as possible to an adult profile within the range that does not result in any problems for eating and the eyelids, to reduce the number of distraction osteogenesis with Le Fort III throughout life.

However, there are no established standards indicating the degree of overcorrection required in children. The reason for this is that the patient's cephalogram cannot be superimposed to the normal child's one accurately because the lengths of the anterior cranial base (the distance between S and N) and the lengths of the posterior cranial base [the distance between S and basion (Ba), which is situated at the back of the posterior cranial base] in patients with craniosynostosis syndrome are shorter than those of normal children, and the lengths of the anterior cranial base and posterior cranial base are not sufficiently developed and grown.[15,16,22–25] Therefore, it may be difficult to accurately determine the distance and the vector of elongation, especially in younger patients. We needed

Fig. 5. One year postoperatively. Point Or is advanced by 29 mm.

Fig. 7. Three years postoperatively.

Fig. 6. One year postoperatively. Point A is advanced by 31 mm.

Fig. 8. Three years postoperatively.

to superimpose the patient's cephalogram on the normal child's cephalogram without the influence of the shorter anterior cranial base and posterior cranial base in the patient than in the normal child. We focused on Ba, which was positioned at the back of the anterior cranial base and posterior cranial base to solve this problem because Ba was not affected by the insufficient growth of the anterior cranial base and posterior cranial base. Then, Ar, which was positioned at the nearest point to Ba, was used as a substitute for Ba that was not defined on the cephalogram of CANJA. We were able to get the value of the predicted distance and vector to be close to the adult profile as accurately as possible without the influence of insufficient growth of the anterior cranial base and posterior cranial base by moving the patient's Ar close to CANJA's Ar.

However, it was difficult to advance the patients' faces along the planned position accurately when we advanced the faces in practice. The desired mean angle before distraction and the real mean angle after distraction of Or were $2.8 \pm 15.9°$ and $3.3 \pm 8.9°$, respectively, and the desired mean distance before distraction and the real mean distance after distraction of Or were 25.4 ± 7.8 mm and 19.8 ± 4.0 mm, respectively. That meant that distraction of Or was able to move the vector downward to the adult profile, but there was slightly less elongation than the adult profile for the distraction distance. Meanwhile,

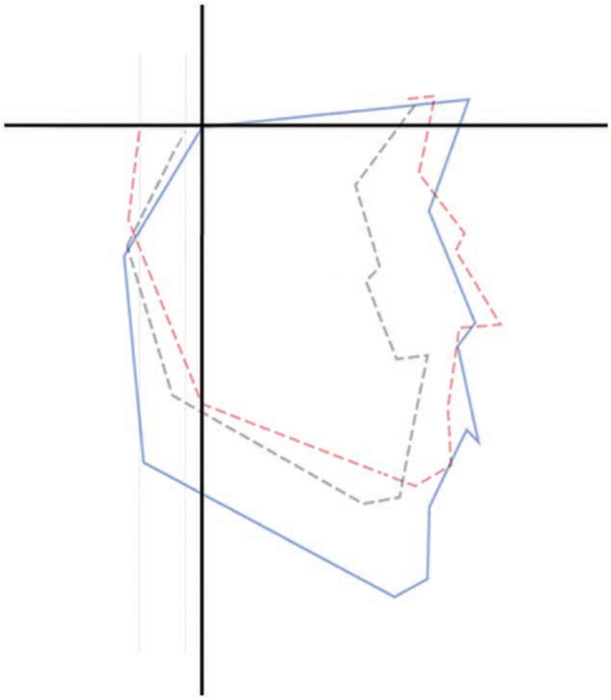

Fig. 9. The patient's preoperative corrected cephalogram (cT0, *black*), the cephalogram immediately after removing the devices (T1, *red*), and the cephalogram averaged from normal Japanese adults (*blue*) are superimposed.

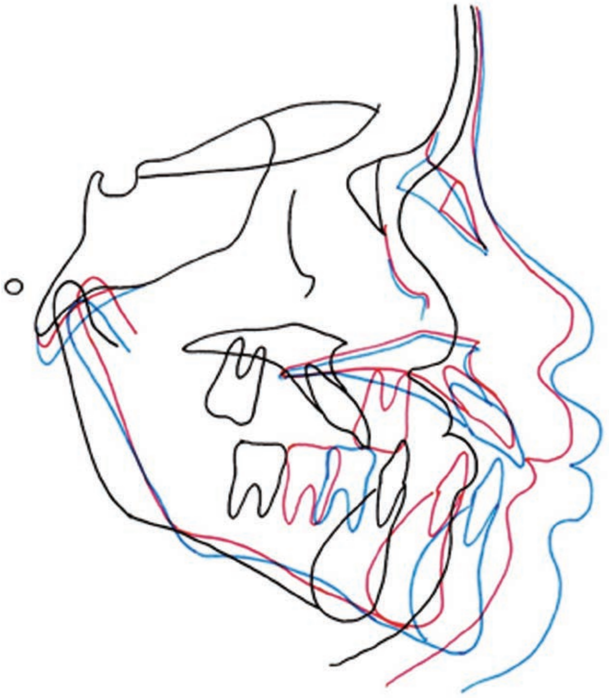

Fig. 10. Cephalogram with tracings superimposed on the anterior cranial base and posterior cranial base. Preoperative (*black*), immediately after removing the devices (*red*), and 3 years after surgery (*blue*).

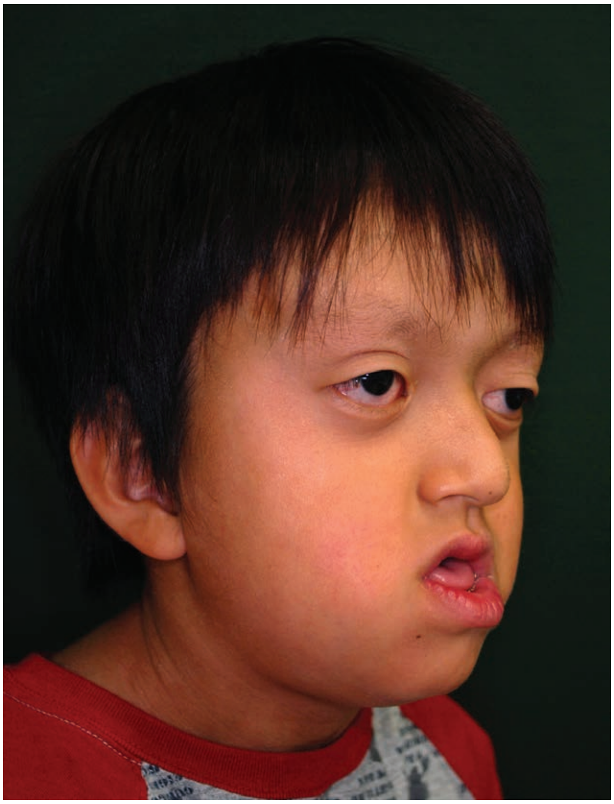

Fig. 11. A 6-year-old male patient with Crouzon syndrome. Reoperative view. He has severe maxillary retrusion.

Fig. 12. Preoperative view. He has a class III malocclusion.

the desired mean angle before distraction and the real mean angle after distraction of A were 29.2 ± 7.9° and 6.1 ± 8.5°, respectively, and the desired mean distance before distraction and the real mean distance after distraction of A were 30.6 ± 12.7 mm and 29.4 ± 4.1 mm, respectively. That meant that A was elongated to the extent of the adult profile for the distraction distance, but it was not advanced to the planned position for the distraction vector. The reason for this was that the desired distance and vector of Or were different from those of A, and the greater the distraction distance, the bigger was the differ-

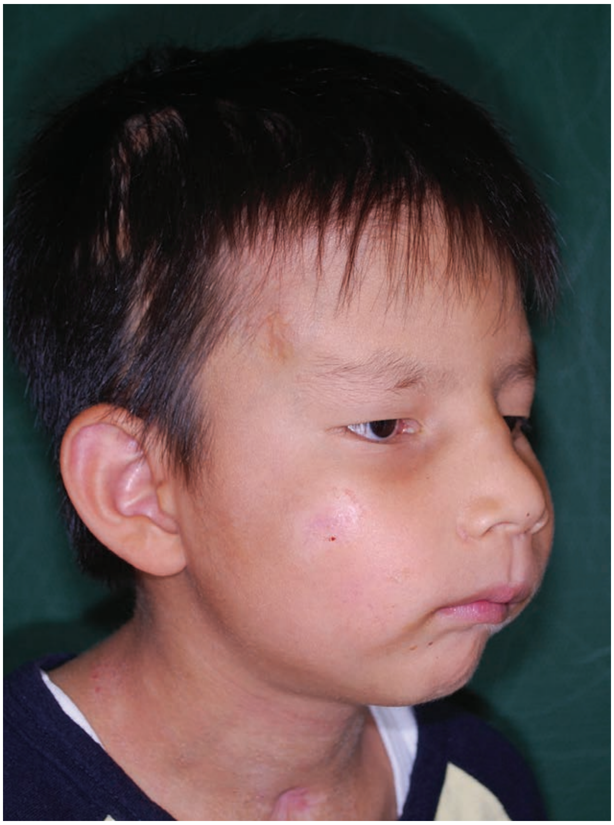

Fig. 13. One year postoperatively. Point Or is advanced by 35 mm.

Fig. 14. One year postoperatively. Point A is advanced by 36 mm.

Fig. 15. Five years postoperatively.

Fig. 16. Five years postoperatively.

ence in the vectors of Or and A when overcorrection was performed in younger patients.[14,26] We finally gave first priority to alignment of Or rather than alignment of A to advance the midfacial segment as a compromise proposal to avoid a second distraction osteogenesis with the Le Fort III procedure because we could not achieve control of both Or and A. We might avoid a second distraction osteogenesis with Le Fort III in the future, because after removal of the distraction device, the facial features were maintained and changed little over a period of 1 year or more. However, we may not avoid Le Fort I and/or mandibular osteotomy because we could not put A in the desired position.

In this study, we described the use of our corrected cephalometric analysis for distraction osteotomy in patients with Crouzon syndrome. Its use in patients with Apert syndrome, who have a short face, may be more difficult. Such patients may require other procedures, such as Le Fort II and zygomatic osteotomy, in addition to Le Fort III surgery.[26] Ultimately, careful long-term follow-up is required in these patients.

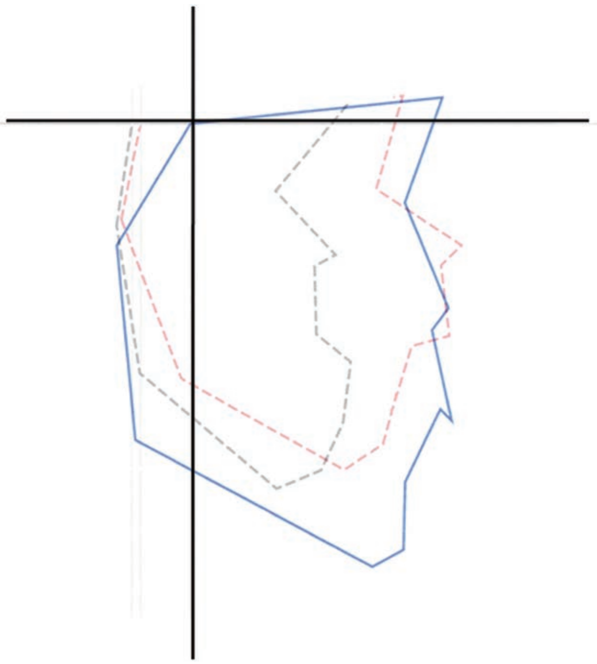

Fig. 17. The patient's preoperative corrected cephalogram (cT0, *black*), the cephalogram immediately after removing the devices (T1, *red*), and a cephalogram averaged from normal Japanese adults (*blue*) are superimposed.

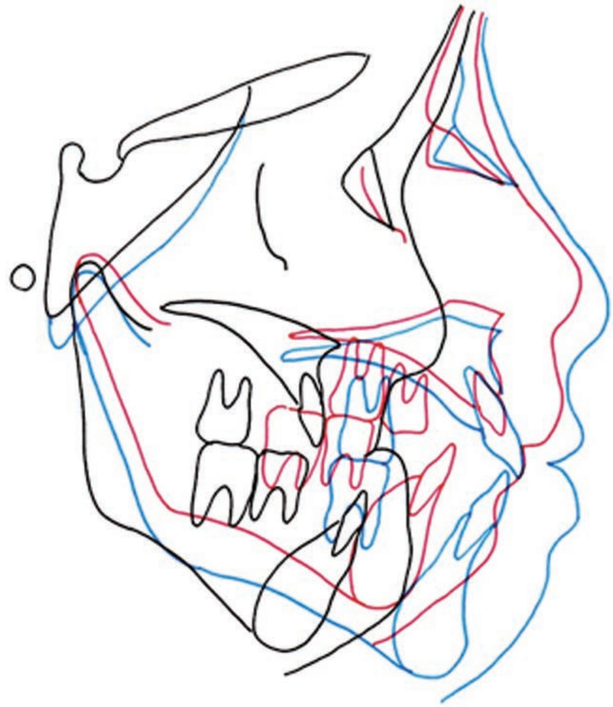

Fig. 18. Cephalogram with tracings superimposed on the anterior cranial base and posterior cranial base. Preoperative (*black*), immediately after removing the devices (*red*), and 5 years after surgery (*blue*).

CONCLUSIONS

Using our corrected cephalometric analysis, we were able to determine the distance and vector of distraction osteogenesis with Le Fort III osteotomy in patients with syndromic craniosynostosis. Our distraction system allowed the patients' facial bones to be brought into the planned position using controlling devices. We believe that this method might be effective in infants.

Shinji Kobayashi, MD
Department of Plastic and Reconstructive Surgery
Knagawa Children's Medical Center
Mutsukawa 2-138-4
Minami-ku
Yokohama
Kanagawa
Japan 232–8555
E-mail: skobayashi@kcmc.jp

PATIENT CONSENT

Parents or guardians provided written consent for the use of the patients' image.

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