Improvement of Periorbital Appearance in Apert Syndrome After Subcranial Le Fort III With Bipartition and Distraction

Vaneshri Chetty, MD, Samer E. Haber, MD,† Roman Hossein Khonsari, MD, PhD,‡ and Eric Arnaud, MD†§*

Aim and Scope: Children with Apert syndrome have a characteristic inversion of the orientation of the palpebral fissures, an increase of the inter-orbital distance, telecanthus, and exorbitism. Here, Le Fort III osteotomy with subcranial bipartition and distraction osteogenesis was evaluated as a tool to improve the position of the palpebral fissures in Apert syndrome.

Material and Methods: All patients with Apert syndrome who underwent Le Fort 3 osteotomy with subcranial bipartition and distraction osteogenesis using an external device, with canthopexy, between 2009 and 2014, with available preoperative and postoperative frontal photographs, were included into the study. Palpebral fissure inclination was measured. Ratios of the intercanthal distance (ICD) to the outer-canthal distance (OCD) and the interpupillary distance to the OCD were computed. Preoperative and postoperative values were compared using the Wilcoxon signed-ranks test.

Results: The authors included 15 patients with Apert syndrome. The mean age at surgery was 10 ± 3.4 years and the average follow-up was 7.3 ± 2.9 years. We found normalization of the negative inclination of the palpebral fissures (right eye: 10.7 ± 2.4 degrees preoperatively versus 7.0 ± 3.1 degrees postoperatively, $P < 0.001$; left eye: 12.4 ± 3.9 degrees preoperatively versus 8.7 ± 4.1 degrees postoperatively, $P = 0.01$) and a significant reduction of the interpupillary distance: OCD ratio (0.717 ± 0.027 preoperatively versus 0.699 ± 0.030 postoperatively, $P = 0.03$). These modifications were stable on the long term. There was no significant change of the intercanthal distance:OCD ratio.

Conclusions: Le Fort III facial advancement with subcranial bipartition and distraction improves the position and orientation of the orbital region in children with Apert syndrome

Key Words: Canthal slant, distraction osteogenesis, hypertelorism, Le Fort osteotomy, palpebral fissure, subcranial bipartition

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The normal shape of the eyelid is closely associated with the shape of the orbital rim.[1] The orbits in Apert syndrome are shallow with thin diverging external walls (Fig. 1).[2–4] The reduced sagittal depth of the orbits is accompanied by an increase in the angle between the bony orbits and hypertelorism.[5] In the transverse plane, both the inner and, particularly, the outer interorbital distances are significantly increased in Apert syndrome.[6] The orbits display a retrusion of the upper and lower margins, associated with an abnormal increase in the medial intercanthal distance (ICD), that is telecanthus (Fig. 2). The interpupillary distance (IPD) is also lengthened.[4]

In the normal population, the outer canthus is usually positioned 2 to 3 mm higher than the inner canthus.[7] This vertical relationship is reversed in Apert syndrome: the outer canthus is positioned below the level of the inner canthus.[4,6] The orientation of the palpebral fissure is dictated by the ligamentous attachments of the canthi on the bony orbital rim. The abnormal bony anatomy and ligamentous attachments give rise to the negative canthal slant of the palpebral fissure, characteristically found in the Apert orbit[8] (Fig. 3).

Fronto-facial advancement osteotomies alter the position of the orbital rim. They subsequently impact the position of the eyelid and therefore the shape of the palpebral fissure.[1,9,10] Here, we investigated the effects of sub-cranial facial advancement (Le Fort III) associated with bipartition on the position of the outer and inner canthi, using standard frontal preoperative and postoperative photographs.

METHODS

All patients with Apert syndrome who underwent Le Fort 3 osteotomy with subcranial bipartition, distraction osteogenesis, and canthopexy between January 2009 and April 2014, with available preoperative and postoperative frontal photographs were included into the study. Patients with intrinsic ocular anomalies were excluded.

Surgical Technique

Before Le Fort III osteotomy, all patients had benefited from at least 1 skull expansion procedure to prevent craniocerebral disproportion: fronto-facial advancement and/or posterior expansion by distraction osteogenesis.

From the *University of Witwatersrand, CM. Johannesburg General Hospital, South Africa; †Hôpital Necker - Enfants Malades, Assistance Publique – Hôpitaux de Paris, Service de Neurochirurgie Pédiatrique, Unité Fonctionnelle de Chirurgie Crânio-Faciale; Centre de Référence Maladies Rares CRANIOST, Filière Maladies Rares TeteCou; Université de Paris, Université Paris Descartes; ‡Hôpital Necker - Enfants Malades, Assistance Publique – Hôpitaux de Paris, Service de Chirurgie Maxillofaciale et Chirurgie Plastique; Centre de Référence Maladies Rares CRANIOST, Filière Maladies Rares TeteCou; Université de Paris, Université Paris Descartes, Paris; and §Clinique Marcel Sembat, Boulogne-Billancourt, France.

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Address correspondence and reprint requests to Eric Arnaud, MD, 34 avenue d'Eylau, 75116 Paris, France;
E-mail: drericarnaud@hotmail.com

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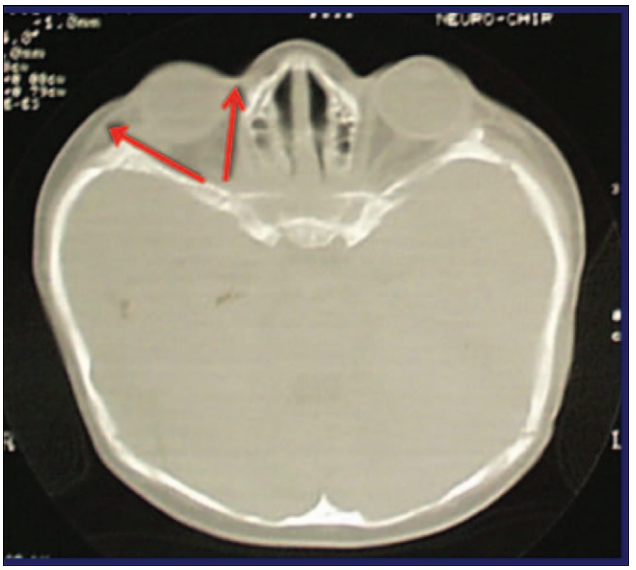

FIGURE 1. Computed tomography scan of an unoperated patient with Apert syndrome demonstrating shallow orbits with thin diverging external walls. Notice the brachycephalic anteroposterior shortening of the skull.

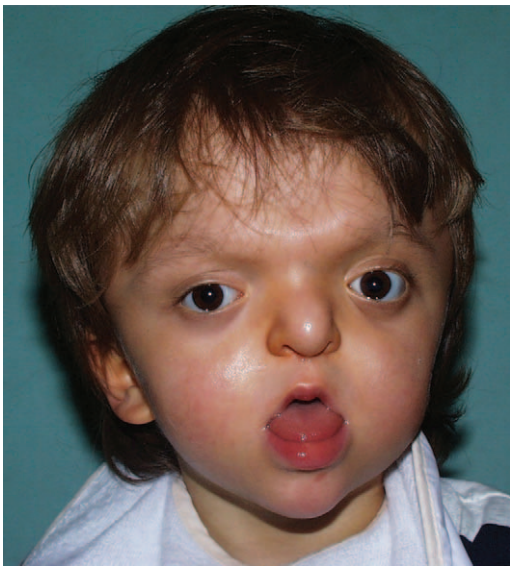

FIGURE 3. Five-year-old girl with Apert syndrome demonstrating exorbitism, hypertelorbitism, and down-slanting palpebral fissures. The lateral canthi are positioned lower than the inner canthi.

Le Fort III osteotomy with subcranial bipartition[11] was performed with inward rotation of both hemi-faces in the frontal plane. This achieved medialization of the orbits and widening and advancement of the maxilla. To get this done, we resected a V-shaped piece of bone superiorly on the nasal bone (Fig. 4) and a triangular bone graft was inserted inferiorly in the gap of the sagittal intermaxillary disjunction. Twelve patients out of 15 presented with a prominent, bossed forehead for which a reduction of the glabellar region was performed simultaneously, by burring or by frontal sinus reduction. Distraction was performed using 2 internal temporo-zygomatic distractors as well as a rigid external distraction device for the maxilla (Fig. 5). Indications were midface retrusion, exorbitism, divergent hypertelorbitism, sleep apnea, and aesthetic enhancement. External and internal canthopexies were performed simultaneously (Fig. 6). The external canthopexy used 4-0 surgical stainless-steel sutures in the lateral edge of the inferior tarsus. It was then anchored to the lateral bandeau or forehead with the right direction of upward pull and overcorrection while preventing visual obstruction at pupillary level. The patients were followed for at least 1 year.

Two-dimensional (2D) frontal photographs of all the face, from all subjects, were acquired pre- and postoperatively (with a minimum follow-up of 12 months) by the same experienced

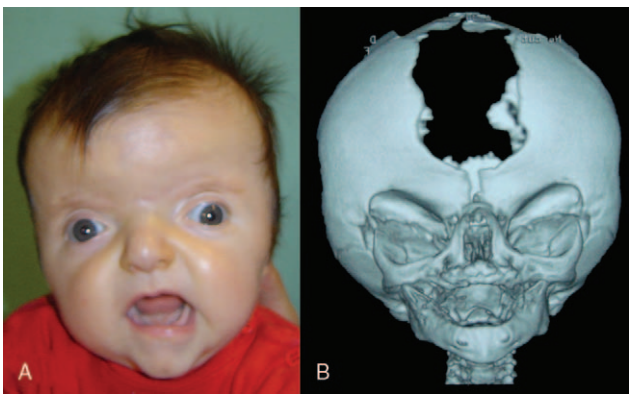

FIGURE 2. (A) Typical facies in Apert syndrome. The intercanthal distance is wide with down-slanting palpebral fissures and maxillary hypoplasia with a high palatal arch. (B) The computed tomography scan confirms the bony deformation namely hypertelorbitism with maxillary hypoplasia. Notice the wide anterior fontanelle that is typical of this deformity.

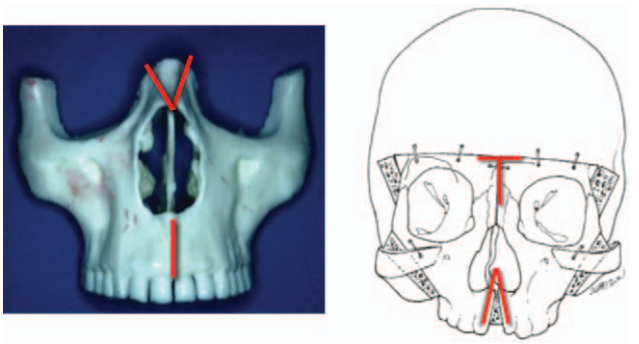

FIGURE 4. Pictorial demonstrating in red the midline fronto-nasal V-shaped osteotomies and the mid-maxillary osteotomy in the Le Fort III segment before (left) and after (right) discarding the upper nasal wedge. Notice how the upper dental arch is splayed in the midline due to the inward rotation of the hemi-Le Fort III segments in the frontal plane.

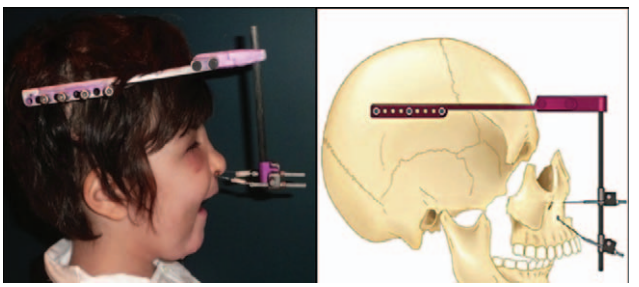

FIGURE 5. Rigid external distraction device anchored directly to the maxilla through wires. Plates and screws are used for wire stability on the anterior surface of the maxilla to re-distribute the anterior traction forces evenly across this surface.

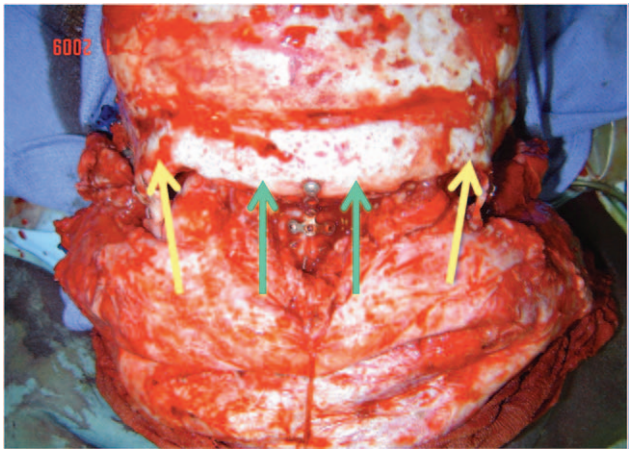

FIGURE 6. Intraoperative bird's eye view demonstrating the locations of the medial (green arrows) and the lateral (yellow arrows) canthopexies.

photographer (EA) using a 5.1-megapixel digital camera. In a standardized fashion, each subject was sitting 1.5 m away from the camera with the visual axis being parallel to the floor of the room (Frankfurt Horizontal Line), with open eyes and a neutral facial expression. The photographic records were stored as JPEG files and further processed through ImageJ software. Linear measurements were performed on reformatted and equal-sized images and ratios were then calculated. Ratios of the ICD to the outer-canthal distance (OCD) and the IPD to the OCD were computed. The palpebral fissure inclination was measured as the angle between the horizontal and the line joining the inner and outer canthi (Fig. 7).

Orbital measurements in Apert syndrome before and after surgery were compared using a 2-sided paired Wilcoxon signed-ranks test. Significance level was set at $P = 0.05$. Statistical analysis was performed using RStudio v. 1.1.453. RStudio, Inc, Boston, MA.

RESULTS

Fifteen patients with Apert syndrome (10 boys and 5 girls) were included. The mean age at surgery was 10 ± 3.4 years. There were no major complications. The mean age at the last postoperative assessment was 18.2 ± 5.8 years. Mean follow-up was 7.3 ± 2.9 years.

The palpebral fissure inclination had an inversed orientation preoperatively in all patients — the lateral canthus was lower than the medial canthus. Mean negative preoperative palpebral fissure inclination was 10.7 ± 2.4 degrees on the right side and 12.4 ± 3.9 degrees on the left side. These angles were significantly reduced postoperatively (7.0 ± 3.1 on the right side, $P < 0.001$ and 8.7 ± 4.1 on the left side, $P = 0.01$).

The ICD:OCD ratio did not show statistically significant variation after surgery. (0.415 ± 0.037 preoperatively; 0.417 ± 0.029 postoperatively, $P = 0.89$). The IPD:OCD ratio was significantly decreased after surgery (0.717 ± 0.028 preoperatively; 0.699 ± 0.030 postoperatively, $P = 0.03$).

DISCUSSION

Tessier first described the use of fronto-facial advancement in Apert syndrome in 1971.[12,13] Van der Meulen used a modified version of the Ortiz-Monasterio monobloc osteotomy to correct orbital hypertelorism in a child with a median facial cleft.[5,10] Tessier later refined the procedure by introducing facial bipartition.[14] It is now known that the Le Fort III with a sub-cranial facial bipartition advances the midface and concurrently narrows the interorbital distance.[7]

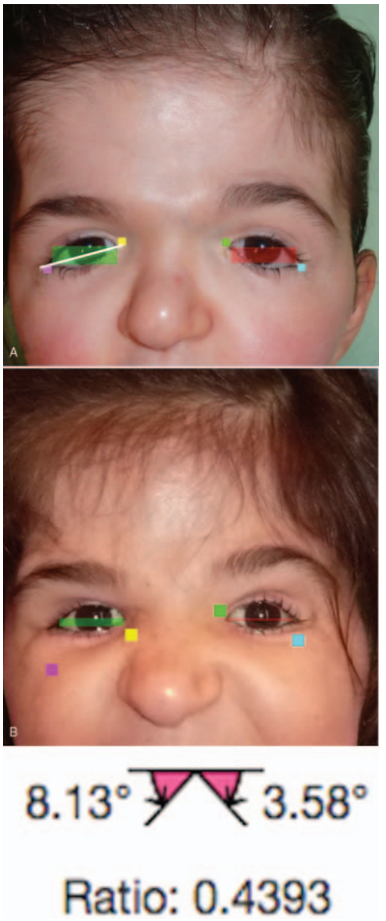

FIGURE 7. Measurement of palpebral fissure inclination (A) pre- and (B) postoperatively as the angle formed by the horizontal and the line joining the medial canthus and the lateral canthus. The measurements were performed by a single examiner in all patients, pre and postoperatively. The canthal positions from (A) were transposed to (B) as colored dots to illustrate the amount of angular correction gained after surgery.

When the lower orbital rim is advanced, the length of the orbit and hence its volume is increased, and exorbitism is decreased. In the same procedure, the orbits are rotated inwards, thus reducing hypertelorbitism.[14]

Prior reports have presented midface advancement as a straight movement with equal advancement at the levels of the nasal root and the incisor tip.[15–17] In 2003 Denny et al[18] first combined the initial principles of Tulasne and Tessier with a rotation of the midface around the transverse axis of the orbits.[19]

The data presented here demonstrates that the palpebral fissures undergo a medial shift and rotation secondary to this "rotation-advancement" orbital displacement, thus reducing the inverted slant and contributing to correct the Apert facies (Figs. 8 and 9). The palpebral fissure inclination was significantly improved bilaterally ($P < 0.05$). While the ICD:OCD ratio variation did not reach statistical significance, the rationale of the facial bipartition procedure dictates that both the ICD and OCD are moved medially together.[9,20,21] Hence, the ICD and OCD are reduced simultaneously, which explains the absence of variation of their ratio postoperatively. On the other hand, the decrease in IPD:OCD ratio reflects the approximation of the eye globes ($P < 0.05$). This correction is statistically significant and long-term.

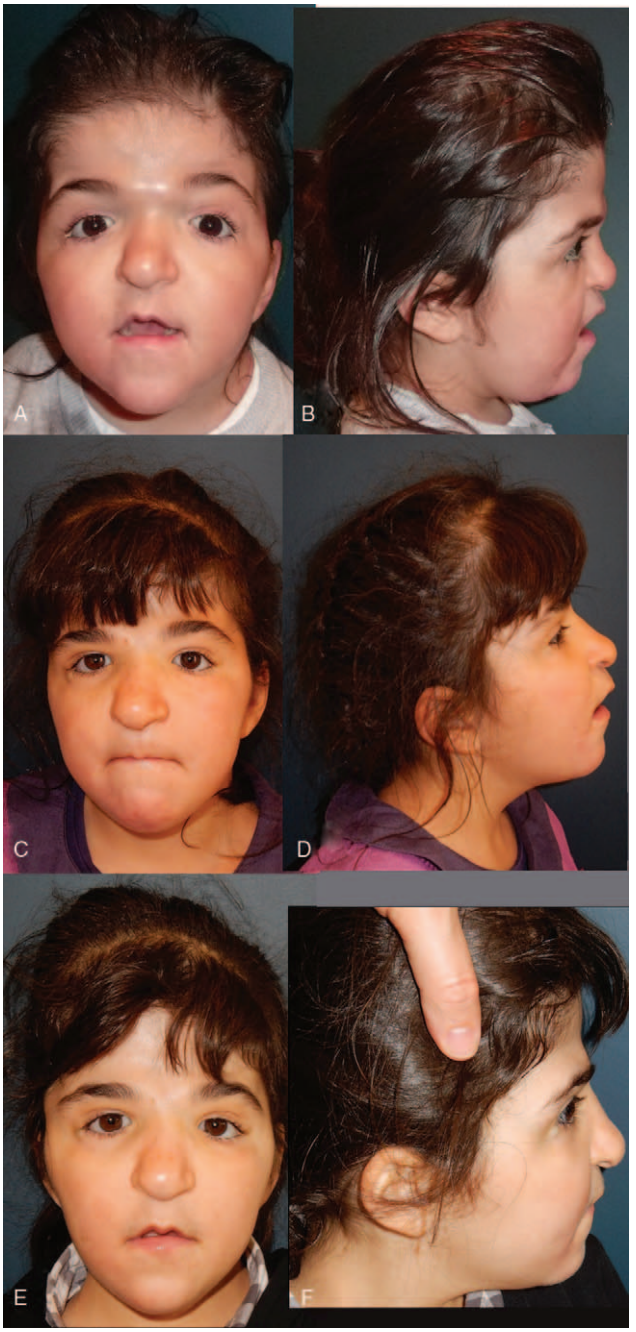

FIGURE 8. Five-year-old girl with Apert syndrome. She had undergone fronto-facial monobloc advancement at age 14 months for correction of exorbitism, sleep apnea, and prevention of turricephaly. She presents at age 5 years with residual sleep apnea and recurrent maxillary deficiency (A, B). Le Fort III osteotomy with bipartition is performed with advancement and medial rotation of both hemi-faces. (C, D) 3-year-postoperative pictures (age 8 years) show adequate correction of the maxillary relationship with residual anterior overcorrection. (E, F) Four-year-postoperative pictures show decreased maxillary overcorrection. The palpebral fissure inclination is still improved.

It is unclear in this study how much improvement was due to bone movements and how much was related to the canthopexies. However, in the senior author's experience, both components are complementary. Moreover, the medialization and inward rotation of the bony orbits requires subperiorbital undermining and separation of the lateral canthus from the lateral orbital rim. If simultaneous lateral canthopexy was to be omitted, then gravity, scarring, and the functional soft tissue forces would contribute to persistence of the lateral canthal descent. The implementation of systematic lateral canthopexy with overcorrection seems to prevent this soft tissue deformity as suggested in the present study.

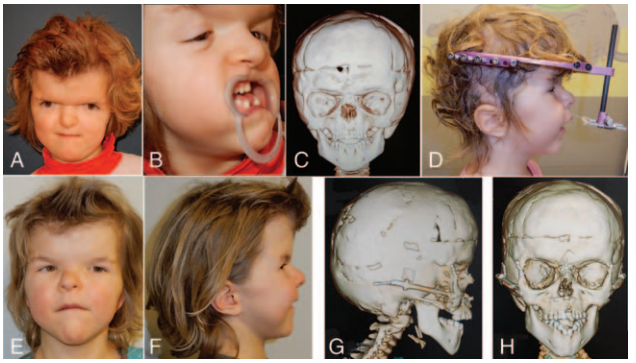

FIGURE 9. (A-C) Seven-year-old girl with Apert syndrome. She previously underwent posterior cranial expansion using translambdoid springs at age 7 months and a fronto-orbital advancement and remodeling at age 2.5 years. She currently displays severe maxillary hypoplasia with class III malocclusion, full crossbite, V-shaped palate, and moderately severe obstructive sleep apnea syndrome (Apnea-Hypopnea Index: 5 per hour). There is also hypertelorbitism: intercanthal distance is 40 mm and the palpebral fissures have an inverted canthal slant. (D) She underwent Le Fort III osteotomy with bipartition and medial rotation of the hemifaces. (E-H) One year after Le Fort III with bipartition and distraction: overcorrection with class II occlusion. Partial but significant improvement of hypertelorbitism and palpebral fissure orientation.

LIMITATIONS

This study is retrospective and the use of ratios as a surrogate for direct ICD measurements are limitations to this study. It does not discern the extent of improvement related to bone movement and that related to the canthopexies. Further improvements should include the use of geometric morphometrics on 3D surface scans of the face of patients with Apert syndrome, to control the biases due to the use of 2D photographs taken in a fixed orientation.

CONCLUSIONS

Le Fort III osteotomy with distraction, associated with bipartition, the removal of a triangular nasal bony wedge and canthopexy, provide satisfactory midfacial advancement and correction of orbital parameters.

While this type of intervention is primarily geared towards fronto-facial functional correction with bony orbital approximation and repositioning, improvement, and correct positioning of the palpebral fissure may be an important aesthetic consideration in the pre-emptive surgical planning stages for children with Apert syndrome.

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