CONCLUSIONS

- The use of BTDO technology helps to increase the success rate of treating the alveolar cleft.
- Better cosmetic aspects are achieved, as they generate hard and soft tissues together, which cannot be achieved by other techniques.
- This technique is more acceptable for patients, as it avoid donor site morbidity bone. Also, device is well tolerated by patients.

REFERENCES

1. Agurto P, Castellon L, Fuenzalida C, et al. Management of severe midface retrusion with distraction osteogenesis in patients with cleft lip and alveolus. *J Craniofac Surg* 2020;31:1551–1555
2. Mossaad AM, Al Ahmady HH, Ghanem WH, et al. The use of dual energy X-ray bone density scan in assessment of alveolar cleft grafting using bone marrow. *J Craniofac Surg* 2021:1–4Ahead-of-print
3. Sun XC, Wang H, Li JH, et al. Repair of alveolar cleft bone defects by bone collagen particles combined with human umbilical cord mesenchymal stem cells in rabbit. *Biomed Eng Online* 2020;19:1–19
4. Sakamoto T, Ishii T, Sueishi K, et al. Interdental distraction osteogenesis in unilateral cleft lip and palate patient with wide alveolar cleft. *Bull Tokyo Dent Coll* 2020;61:201–209
5. Liou EJW, Chen PKT. Intraoral distraction of segmental osteotomies and miniscrews in management of alveolar cleft. *Semin Orthod* 2009;15:257–267
6. Singh S, Mehrotra D, Gupta C. Wide alveolar cleft and midface distraction: report of a case. *J Oral Biol Craniofac Res* 2012;2:123–125
7. Zhang J, Zhang W, Shen SG. Segmental maxillary distraction osteogenesis with a hybrid-type distractor in the management of wide alveolar cleft. *Cleft Palate Craniofac J* 2018;55:1133–1137
8. Binger T, Katsaros C, Rücker M, et al. Segment distraction to reduce a wide alveolar cleft before alveolar bone grafting. *Cleft Palate Craniofac J* 2003;40:561–565
9. Rachmiel A, Emodi O, Gutmacher Z, et al. Oral and dental restoration of wide alveolar cleft using distraction osteogenesis and temporary anchorage devices. *J Cranio Maxillofac Surg* 2013;41:728–734
10. Al-Ahmady HH, Abd Elazeem AF, Bellah Ahmed NE, et al. Combining autologous bone marrow mononuclear cells seeded on collagen sponge with Nano Hydroxyapatite, and platelet-rich fibrin: Reporting a novel strategy for alveolar cleft bone regeneration. *J Cranio Maxillofac Surg* 2018;46:1593–1600
11. Sunitha C, Gunaseelan R, Anusha V, et al. Maxillary movement in cleft patients treated with internal tooth borne distractor. *J Maxillofac Oral Surg* 2013;12:266–272
12. Seifeldin SA. Is alveolar cleft reconstruction still controversial? (Review of literature). *Saudi Dent J* 2016;28:3–11
13. Seifeldin SA. Is alveolar cleft reconstruction still controversial? (Review of literature). *Saudi Dent J* 2015;28:3–11
14. Yu H, Wang X, Fang B, et al. Comparative study of different osteotomy modalities in maxillary distraction osteogenesis for cleft lip and palate. *J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg* 2012;70:2641–2647
15. Ghasemianpour M, Ehsani S, Tahmasbi S, et al. Original article: Distraction osteogenesis for cleft palate closure: a fi nite element analysis. *Dent Res J (Isfahan)* 2014;11:92–99
16. Liang L, Liu C. Trans-sutural distraction osteogenesis for alveolar cleft repair: an experimental canine study. *Cleft Palate Craniofac J* 2012;49:701–707
17. Laster Z, Reem Y, Nagler R. Horizontal alveolar ridge distraction in an edentulous patient. *J Oral Maxillofac Surg* 2011;69:502–506
18. Pektas ZÖ, Kircelli BH, Bayram B, et al. Alveolar cleft closure by distraction osteogenesis with skeletal anchorage during consolidation. *Int J Oral Maxillofac Implant* 2008;23:147–152
19. Tae KC, Gong SG, Min SK, et al. Use of distraction osteogenesis in cleft palate patients. *Angle Orthod* 2003;73:602–607
20. Shahab N, Sar C, Sarac M, et al. Reconstruction of premaxilla with alveolar distraction osteogenesis in a patient with complete cleft lip and palate: a case report. *Cleft Palate Craniofac J* 2018;56:534–537
21. Hegab AF. Closure of the alveolar cleft by bone segment transport using an intraoral tooth-borne custom-made distraction device. *J Oral Maxillofac Surg* 2012;70:e337–e348
22. Erverdi AN, Yilmaz B, Motro M, et al. Simultaneous alveolar cleft closure and dental midline correction with curvilinear intraoral distraction. *Cleft Palate Craniofac J* 2013;51:344–349
23. El-Sayed KM, Khalil H. Transpalatal distraction osteogenesis prior to alveolar bone grafting in cleft lip and palate patients. *Int J Oral Maxillofac Surg* 2010;39:761–766
24. Dolanmaz D, Karaman AI, Durmus E, et al. Management of alveolar clefts using dento-osseous transport distraction osteogenesis. *Angle Orthod* 2003;73:723–729
25. Choi S-H, Park JH, Cha J-Y, et al. Intraoral premaxillary distraction in a patient with maxillary retrognathic cleft lip and palate: a case report. *Cleft Palate Craniofac J Off Publ Am Cleft Palate Craniofac Assoc* 2018;56:827–830
26. Zemann W, Pichelmayer M. Maxillary segmental distraction in children with unilateral clefts of lip, palate, and alveolus. *Oral Surg Oral Med Oral Pathol Oral Radiol Endodontol* 2010;111:688–692
27. Pichelmayer M, Mossböck R, Droschl H. Maxillary segmental distraction in a patient with bilateral cleft lip and alveolus with subsequent tooth transplantation: a preliminary case report. *Cleft Palate Craniofac J* 2008;45:446–451
28. Shadia E, Abdelaziz B, Najla Dar-Odeh AA. Intraoral wound dehiscence after open reduction internal fixation of mandibular fractures: a retrospective cohort study. *Wounds* 2021;33:60–64
29. Perez D, Ellis E, Vega OA. Distraction osteogenesis for craniomaxillofacial problems. *Tex Dent J* 2011;128:1159–1170
30. Liao L-S, Tan Z, Zheng Q, et al. Animal experimental study on repairing alveolar clefts by using rectilinear distraction osteogenesis. *J Plast Reconstr Aesthetic Surg* 2008;62:1573–1579
31. Erverdi N, Küçükkeleş N, Şener C, et al. Interdental distraction osteogenesis for the management of alveolar clefts: archwise distraction. *Int J Oral Maxillofac Surg* 2011;41:37–41

Correction of Midface Deficiency in Patient With Crouzon Syndrome by Orthognathic Surgery and Patient Specific Facial Implant

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Abstract: Crouzon syndrome, also known as craniofacial dysostosis, is an autosomal dominant inherited disease characterized by

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early cranial fusion and consequential craniofacial malformations. In patients with Crouzon syndrome, the growth of the midface is affected due to early fusion of the cranial base, which results in exophthalmos, ocular ptosis, midface deficiency, and maxillary retrognathism. Frontofacial advancement using Le Fort III osteotomy is the conventional method for treating patients with Crouzon syndrome. However, this procedure has drawbacks such as extensive operation field and high possibility of serious complications (eg, meningitis). For patients with mild symptoms, facial esthetics and proper occlusion can be promoted through conventional orthognathic surgery, but midface deficiency cannot be completely resolved. Thus, in this case report, midface augmentation was performed for a patient with Crouzon syndrome by undergoing conventional orthognathic surgery, along with patient-specific implants made using a 3D virtual technique. Implants were 3D printed using polyetherketoneketone and simultaneously implanted during the orthognathic surgery. After the surgery, not only were the patient's occlusion and facial esthetics improved, but also exophthalmos and ocular ptosis were reduced by the midfacial augmentation effect of patient-specific implants placed in the midface. Since the implants were made exactly as what surgeons have intended through computer-aided design (CAD)/computer-aided manufacturing (CAM) and 3D printing techniques, problems such as under-/over-correction were avoided. In addition, the possibility of implant malpositioning was minimized using surgical stents as implants were passively fitted on the patient's bone surface. This case verified that the application of 3D technology to the field of oral and maxillofacial surgery can reduce the invasiveness of surgery and improve the accuracy of the operation. Therefore, by using cutting-edge technologies, the field of oral and maxillofacial surgery is expected to be developed further in the near future.

Key Words: 3D virtual technology, Crouzon syndrome, midface augmentation, patient-specific implant

Crouzon syndrome is an autosomal dominant genetic disorder, which was described in 1912 by Octave Crouzon. Due to the *Fibroblast Growth Factor Receptor 2* gene mutation, an abnormal bone metabolism pattern is shown, causing characteristic dysmorphism of the bones forming the craniofacial area. This has led Crouzon syndrome to be called "craniofacial dysostosis."[1] Patients with Crouzon syndrome characteristically show early fusion of the cranial suture, mainly that of the coronal and sagittal sutures, resulting in craniosynostosis such as brachycephaly. Furthermore, the growth of the midface is affected due to the failure of cranial growth from early cranial anastomosis, resulting in midface deficiency, high palatal vault, and narrow maxillary dental arch. It has also been reported that exophthalmos or lagophthalmos occurs from the small size of the orbit, causing ocular ptosis, and frequent inflammation occurs in the cornea and conjunctiva, the most anterior regions of the eyeball.[2]

Frontofacial monobloc advancement including distraction osteogenesis can be performed for facial esthetics and proper occlusion accompanied by severe midface deficiency in patients with Crouzon syndrome to move the maxilla and frontal bone anteriorly. Since this surgical method requires coronal approach and osteotomy of the skull bone, including Le Fort III osteotomy, the surgical field is extensive and serious complications such as cerebrospinal fluid leak, meningitis, and impaired visual acuity may occur.[3,4] It has been reported that the facial appearance and occlusion state of patients with mild midface deficiency can be improved by orthognathic surgeries using only Le Fort I osteotomy. However, after the surgery, since the facial area below the orbital rim cannot be advanced due to the position of Le Fort I or II osteotomy lines, ocular ptosis, and exophthalmos are highly likely to remain.[5]

Thus, in this study, we presented a case of a patient with Crouzon syndrome that is characterized by mild midface deficiency, whose facial esthetic improvement and occlusal recovery were achieved through orthognathic surgery involving Le Fort I osteotomy and bilateral sagittal split ramus osteotomy (BSSRO), along with volume enhancement of the infraorbital area, using the patient-specific implant, which was made using CAD/CAM and 3D printing technology.

CLINICAL REPORT

A 20-year-old male patient, presenting with midface deficiency, anterior open bite, and skeletal class III malocclusion, was referred for orthognathic surgery after presurgical orthodontic treatment from the Department of Orthodontics at Seoul National University Dental Hospital. The patient was diagnosed with Crouzon syndrome due to midface deficiency and characteristic features such as exophthalmos. The clinical and radiographic examination revealed a high palatal vault, extensive midface deficiency from lower infraorbital rim to zygoma area, exophthalmos, and ocular ptosis (Fig. 1).

To treat this patient, 2 surgical methods were considered. The first method is midface advancement by Le Fort III osteotomy to correct midface deficiency and then simultaneously or sequentially correct dentofacial deformity via Le Fort I osteotomy. However, in this case, Le Fort III osteotomy was not needed as the midface deficiency was not severe and lagophthalmos was not observed. Moreover, simultaneous Le Fort III and I osteotomy in 1 operation can lead to a decline of surgical accuracy; thus, 2-staged surgical intervention is required, leading to a longer treatment period. The second method is setting proper occlusion through conventional orthognathic surgery with Le Fort I osteotomy and BSSRO and placing facial implants to the midface deficient area. Thus, the facial appearance of the lower-midface area can be improved through maxillary advancement via Le Fort I osteotomy, and the deficiency correction of the upper-midface depression from the infraorbital area to pyriform aperture area can be attained using CAD/CAM assisted patient-specific facial implant. The second method was chosen for treatment this patient.

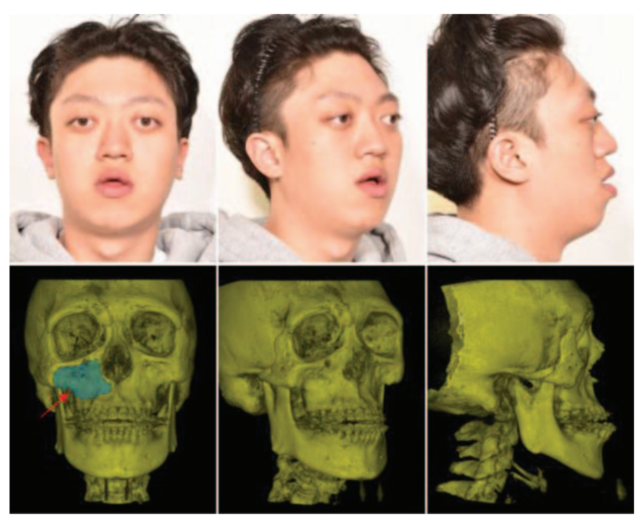

FIGURE 1. Facial photographs and radiographs taken after preoperative orthodontic treatment (physical growth completed state). The patient is diagnosed with Crouzon syndrome and shows only mild symptoms. Extensive midface deficiency is recognized from clinical photos and CBCT images (red arrow). CBCT, cone-beam computed tomography.

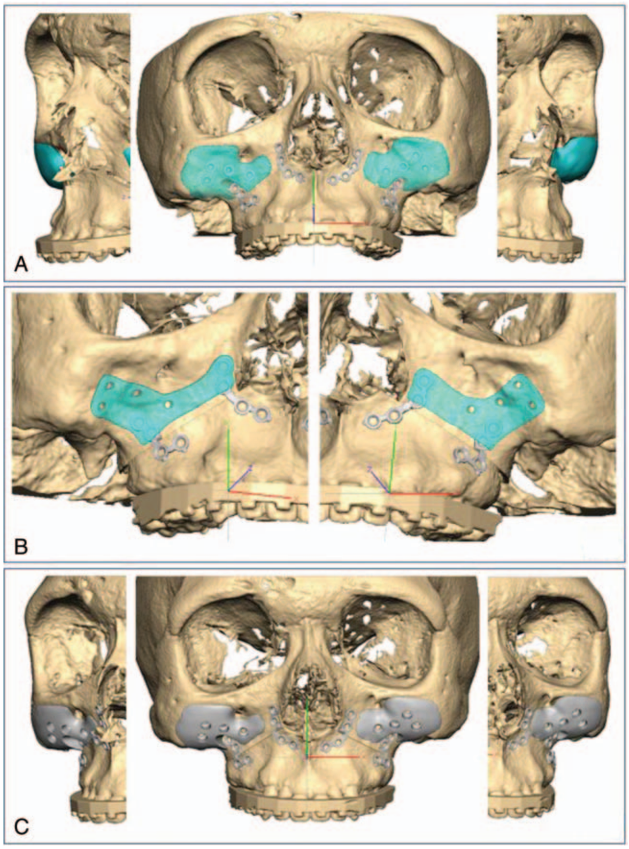

FIGURE 2. Design of patient-specific implant (A). Surgical stent design for screw hole formation (B). The expected result from virtual surgical planning after orthognatic surgery and implant placement (C). The position and shape of the customized plate are designed using CAD/CAM technology and 3D virtual simulation, and the implant was designed based on it. Surgical stents for implant fixation screw hole formation were designed to be supported by maxillary plate so that its size can be reduced, with better surgical accuracy and usage simplicity.

To do virtual surgical planning and to design patient-specific implants, the following method was implemented. 3D image of the skull bone was obtained by converting the digital imaging and communications in medicine file, acquired from taking 3D facial cone-beam computed tomography (CBCT) of the patient before surgery, into a surface tessellation language (STL) file. This STL file was superimposed with the STL file of the dentition area from scanning a dental cast. Using these data, virtual surgery was performed using the FACEGIDE system (Megagen Implant, Daegu, Korea) based on the orthognathic surgical plan. After determining the postoperative position of the maxillary segment, the position to fix the maxillary segment was selected. Then the implants were designed to cover from the infraorbital depressed area to the osteotomy line.

The patient-specific implants were designed in such a way that their outline continues smoothly from the lower edge of the orbital rim to the osteotomy line inferiorly and to the zygoma area laterally, and screw fixation holes were set for fixing implants in the area with sufficient bone thickness (Fig. 2A). The plate-supported surgical stents that reflect the position of the screw holes were fabricated using CAD/CAM technique, and patient-specific implants that can be passively seated on the patient's bone surface were fabricated using 3D printing technology (Fig. 2B-C). Polyetherketoneketone that is 3D printable and has better antibacterial properties than polyetheretherketone, was used for the implant.[6]

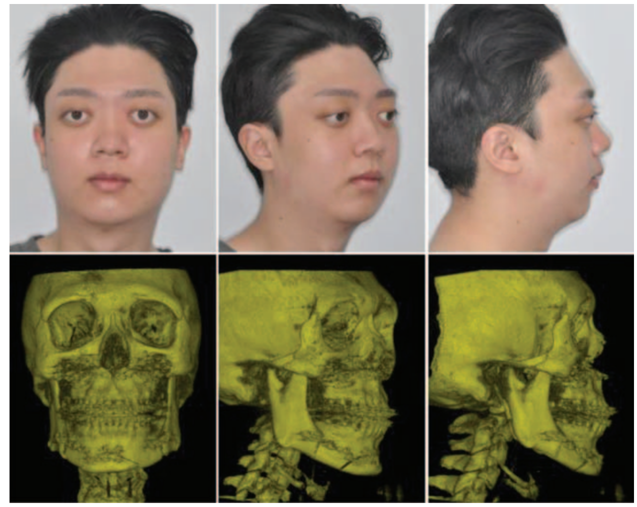

FIGURE 3. Clinical photos and radiographs after orthognathic and patient-specific implant graft surgery. Proper occlusion was achieved through orthognathic surgery, and midface deficiency along with exophthalmos and ocular ptosis was improved through a radiolucent patient-specific implant.

Two months after surgery, the anterior open bite and crossbite were resolved through orthognathic surgery, and the midface deficiency was significantly improved. Furthermore, exophthalmos was significantly improved through the infraorbital area volume enhancement (Fig. 3).

DISCUSSION

In patients with Crouzon syndrome, the development of the nervous system is affected due to the early fusion of cranial sutures. The length of anterior cranial base is shortened as sphenoid bone is compressed, and the volume of the orbit is decreased as the middle cranial base anteroinferiorly deviates due to the consequential increase in intracranial pressure. Furthermore, maxillary hypoplasia and resultant decreased orbit size and midface deficiency often result in unesthetic facial features and malocclusion in patients with Crouzon syndrome due to the cranial base growth failure. Thus, due to intracranial pressure increase, patient with severe symptoms need to undergo craniosurgery during their early ages and need midface advancement through Le Fort III osteotomy if lagophthalmos is observed.[3,4]

In early childhood and growth period, craniofacial surgery such as craniectomy is required to resolve life threatening symptoms. Meanwhile, treatment after the growth period is more focused on improving esthetics and setting proper occlusion. Moreover, midface advancement using Le Fort III osteotomy can be considered if the patient shows symptoms such as severe midface deficiency or lagophthalmos. However, this procedure often leads to high possibility of serious complications such as cerebrospinal fluid leak, meningitis, and massive bleeding. Therefore, meticulous diagnostic planning before surgery, high level of concentration during the surgical process, and scrupulous postoperative care after surgery are required.[7,8] Furthermore, for patients with mild symptoms, proper occlusion and improved esthetics can be achieved through undergoing orthognathic surgery after their physical growth is completed. Conventionally, maxillary advancement through Le Fort I osteotomy compensates for deficiency growth of the midfacial area, and BSSRO of the mandible can obtain appropriate occlusion by repositioning the distal segment along with maxillary dentition.

However, only the lower midfacial area can be advanced, and it is impossible to resolve the overall midface deficiency due to the position of the Le Fort I osteotomy line.[5,9] Thus, facial implant placement can be considered because the upper-midface deficiency cannot be resolved through Le Fort I osteotomy.

Various types of prosthesis, including silicone (polydimethylsiloxane), GoreTex (expanded polytetrafluorethylene, WL Gore & Associates Inc, Flagstaff, AZ), and MedPor (high-density porous polyethylene, Porex Industries, Fairburn, GA), have been used to alleviate facial depression. Since these prostheses are supplied by sizes, premanufactured prostheses of an appropriate size should be selected during the surgery and need to be carved before placing them. Consequentially, the prostheses are not passively fitted on the bone surface, and the gap is formed, leading to a risk of prothesis migration to an unwanted area. Also, since prostheses are often placed based on the surgeon's tactile sense and experiences, there are possibilities of malpositioning or under-/overcorrection.[10]

However, the prosthesis, manufactured using CAD/CAM and 3D printing technology can be passively seated on the patient's bone surface without gap. By forming screw fixation holes in advance during the manufacturing procedure, the position of the prosthesis can be confirmed during the fixation process, and the migration of the prosthesis can be prevented by stably fixing it. Furthermore, the surgeon can decide the thickness of the prosthesis based on the patient's facial contour, and the possibility of under-/overcorrection can be significantly minimized. Finally, since the fixation holes of the prosthesis are designed and set to avoid major anatomical structures such as nerve trunk, there is little possibility of complications such as nerve damage.[11]

Here, implants made of polyetherketoneketone were used to relieve the midface deficiency of a patient with Crouzon syndrome. First, through virtual surgery, the postoperative position of the patient's maxilla and mandible was checked, and then the implants covering the patient's midface were designed accordingly. After designing customized plates for maxillary fixation, plate-supported surgical stents were designed to indicate the holes where the implant fixation screws would be located. Before placing implants using the prepared surgical stents, screw holes were formed in the maxillary anterior wall, and the location of the implants was double-checked by placing them in accordance with these holes.

To shorten the operation time and improve the accuracy and predictability of surgeries, the development of 3D virtual surgery software, CAD/CAM and 3D printing technology has been applied in various fields of surgery. In the field of maxillofacial surgery, 3D technology has been applied to maxillofacial reconstruction and orthognathic surgeries to obtain more esthetic surgical outcomes. Even with this case presented, if the surgery were performed by the conventional method, the operation might not have been done as planned. Particularly, the application of new technology can be advantageous to the operator when it comes to the prediction of hard tissue movement through virtual simulation during patient diagnosis or treatment planning, not to mention the utilization of 3D virtual surgery program and surgical materials fabricated using CAD/CAM and 3D printing technology.

In this case report, patient-specific facial implants using 3D virtual surgery, CAD/CAM and 3D printing technology were reviewed. As shown in the postoperative cone-beam computed tomography data and clinical photos, the application of these latest technologies certainly increases the surgical accuracy and guarantees better results. Although long-term stability studies have not been conducted, the latest technology applied to surgery is expected to be better than the 1 with the conventional method. Thus, it is necessary to strive to pursue better surgical outcomes using various cutting-edge technologies in the field of oral and maxillofacial surgery.

REFERENCES

1. Lu X, Sawh-Martinez R, Forte AJ, et al. Classification of subtypes of Crouzon syndrome based on the type of vault suture synostosis. *J Craniofac Surg* 2020;31:678–684
2. Lu X, Forte AJ, Sawh-Martinez R, et al. Orbit, zygoma, and maxilla growth patterns in Crouzon syndrome. *Int J Oral Maxillofac Surg* 2019;48:309–321
3. Hu CH, Wu CT, Ko EW, et al. Monobloc frontofacial or Le Fort III distraction osteogenesis in syndromic craniosynostosis: three-dimensional evaluation of treatment outcome and the need for central distraction. *J Craniofac Surg* 2017;28:1344–1349
4. Engel M, Berger M, Hoffmann J, et al. Midface correction in patients with Crouzon syndrome is Le Fort III distraction osteogenesis with a rigid external distraction device the gold standard? *J Craniomaxillofac Surg* 2019;47:420–430
5. Sancar B, Tanisik BH. Treatment of the patient with Crouzon syndrome with orthognathic surgery. *J Craniofac Surg* 2020;31:806–808
6. Khashaba MM, Shaheen HA, Ibrahim WH, et al. Accuracy of patient-specific temporal implants using PEKK. *J Craniomaxillofac Surg* 2021S1010-5182(21)00133-5
7. Raposo-Amaral CE, Denadai R, Pereira-Filho JC, et al. Serious complications after Le Fort III distraction osteogenesis in syndromic craniosynostosis: evolution of preventive and therapeutic strategies. *J Craniofac Surg* 2018;29:1397–1401
8. Raposo-Amaral CE, Denadai R, Zanco GL, et al. Long-term follow-up on bone stability and complication rate after monobloc advancement in syndromic craniosynostosis. *Plast Reconstr Surg* 2020;145:1025–1034
9. Fattahi T, Salman S, Steinberg B. Augmentation of the infraorbital rim in orthognathic surgery. *Int J Oral Maxillofac Surg* 2017;46:1315–1318
10. Patel K, Brandstetter K. Solid implants in facial plastic surgery: potential complications and how to prevent them. *Facial Plast Surg* 2016;32:520–531
11. Cheng KJ, Liu YF, Wang R, et al. Topological optimization of 3D printed bone analog with PEKK for surgical mandibular reconstruction. *J Mech Behav Biomed Mater* 2020;107:103758

Long-Term Stability of Alveolar Bone Graft in Cleft Lip and Palate Patients: Systematic Review and Meta-Analysis

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Objective: Alveolar bone grafts are the golden standard in treating patients with oral cleft and hence, the long-term success of this

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