3. Kelly A, Bough ID Jr, Luft JD, et al. Hairy polyp of the oropharynx: case report and literature review. *J Pediatr Surg* 1996;31:704–706
4. Nicastro V, Zagaria A, Abita P, et al. Association between obstructive sleep apnea and hearing loss: a literary review. *Acta Medica Mediterranea* 2019;35:3411
5. Freni F, Galletti B, Nicastro V, et al. Treatment of paradoxical vocal fold motion in a 12-year-old patient with Patau's syndrome. *EuroMediterranean Biomed J* 2019;14:168–171
6. Gazia F, Abita P, Alberti G, et al. NICU infants & SNHL: experience of a Western Sicily tertiary care centre. *Acta Medica Mediterranea* 2019;35:1001–1007
7. Conti A, Capasso E, Casella C, et al. Blood transfusion in children: the refusal of jehovah's witness parents'. *Open Med (Wars)* 2018;13: 101–104
8. Kalcioglu MT, Can S, Aydin NE. Unusual case of soft polyp causing airway obstruction and review of literature. *J Pediatr Surg* 2010;45:e5–e8
9. Wu J, Schulte J, Yang C, et al. Hairy polyp of the nasopharynx arising from the eustachian tube. *Head Neck Pathol* 2016;10:213–216
10. Tariq MU, Din NU, Bashir MR. Hairy polyp, a clinicopathologic study of four cases. *Head Neck Pathol* 2013;7:232–235
11. Eichel BS, Hallberg OE. Hamartoma of the middle ear and eustachian tube. Report of a case. *Laryngoscope* 1966;76:1810–1815
12. Galletti B, Gazia F, Freni F, et al. Endoscopic sinus surgery with and without computer assisted navigation: a retrospective study. *Auris Nasus Larynx* 2019;46:520–525
13. Holt GR, Holt JE, Weaver RG. Dermoids and teratomas of the head and neck. *Ear Nose Throat J* 1979;58:520–531
14. Calcaterra T. Teratomas of the nasopharynx. *Ann Otol Rhinol Laryngol* 1969;78:165–171
15. Chakravarti A, Vishwakarma SK, Arora VK, et al. Dermoid (hairy polyp) of the nasopharynx. *Indian J Pediatr* 1998;65:473–476
16. Kelly AB. Hairy or dermoid polypi of the pharynx and naso-pharynx. *J Laryngol Rhinol Otol* 1918;33:65–70
17. Galletti B, Gazia F, Galletti C, et al. Endoscopic treatment of a periorbital fat herniation caused by spontaneous solution of continuity of the papyracea lamina. *BMJ Case Rep* 2019;12:e229376
18. Freni F, Galletti B, Bruno R, et al. Multidisciplinary approach in the removal of post-trauma foreign bodies in the head and neck district: Cases report and review of literature. *Acta Medica Mediterranea* 2019;35:405–410
19. De Luca F, Alessi L, Bruno E, et al. Graves' disease in childhood: new epidemiological, pathophysiological and therapeutic insights. *Minerva Pediatr* 2016;68:66–69
20. Pacino GA, Salvatore C, Antonino M, et al. Advanced olfactory neuroblastoma in a teenager: a clinical case and short review of literature. *Childs Nerv Syst* 2020;36:485–489
21. Serra A, Caltabiano R, Giorlandino A, et al. Nasal metastasis as the first manifestation of a metachronous bilateral renal cell carcinoma. *Pathologica* 2017;109:421–425
22. Anastasi G, Bramanti P, Di Bella P, et al. Volume rendering based on magnetic resonance imaging: advances in understanding the three-dimensional anatomy of the human knee. *J Anat* 2007;211:399–406

Fronto-Orbital Advance in a Patient With Roberts Syndrome

Jaime Andres Jimenez Alvarez, DDS,[*]
Leonardo Andres Chacon, DMD,[†]
Jesus Andres Duque Montealegre, DDS,[‡]
José Manuel Valdés Reyes, DDS,[§]
and Carlos Perez Cataño, DMD[||]

Abstract: Roberts Syndrome is an extremely rare syndrome reporting about 150 cases in the literature, with a very low survival rate. The authors present a case of a female patient with Roberts Syndrome who also had a coronal craniosynostosis. The aim of this case report is to present a case of a patient with Roberts Syndrome with a brachycephaly that required management of fronto-orbital advancement. In conclusion Roberts Syndrome is a rare disease, which can have different skeletal variations. This syndrome can manifest itself with craniosynostosis, with the requirement of a comprehensive management to correct it and avoid compression of the brain with endocranial hypertension.

Key Words: Craniosynostosis, fronto-orbital advancement, Roberts syndrome

Roberts Syndrome is a rare syndrome with few cases in the literature. This syndrome was first described by John Bingham Roberts in 1919.[1] In 1969, it was called a "pseudothalidomide" syndrome because of the similarity with the malformations that occurred in patients who mature when they took thalidomide during pregnancy.[2] Hall and Greenberg[3] called it hypomelia-hypotrichosis syndrome and facial hemangioma in 1972.

Roberts Syndrome is an autosomal recessive disorder, the gene responsible is the *ESCO2* gene, described in 2005. The causative gene for Roberts Syndrome is the establishment of cohesion 1 homologue 2 (*ESCO2*), which encodes a protein comprising 601 amino acids belonging to the Eco1 family of acetyltransferases.[4] The definitive diagnosis is made by cytogenetic tests.[5]

Clinically, Roberts Syndrome is characterized by growth retardation, limb malformations that include symmetric mesomelic shortening and anterior-posterior axis involvement in which the frequency and degree of involvement of long bones is, in decreasing order: radii, ulnae, and humeri in the upper limbs; fibulae, tibiae, and femur in the lower limbs. Hand malformations that include brachydactyly and oligodactyly. Craniofacial abnormalities that include, cleft lip and/or cleft palate, encephalocele, premaxillary prominence, microbrachycephaly, midfacial capillary hemangioma, malar flattening, downslanted palpebral fissures, widely spaced eyes, exophthalmos resulting from shallow orbits, corneal clouding, underdeveloped ala nasi, beaked nose, and ear malformations.[6]

A case of Roberts Syndrome with craniosynostosis has not been reported, in which a fronto-orbital decompression is required. Given the low frequency described in the literature, the objective of this article is to present a clinical case of Roberts Syndrome with brachycephaly that required fronto-orbital advancement.

CLINICAL REPORT

This is a 2-year-old female patient who came to the Oral and Maxillofacial Surgery Unit and Neurosurgery Unit of the Roosevelt

From the [*]Department of Oral and Maxillofacial Surgery, Roosevelt Institute; [†]Department of Neurosurgery, Roosevelt Institute, Nueva Granada Militar University, Militar Central Hospital; [‡]Department of Oral and Maxillofacial Surgery, Roosevelt Institute, Bogotá, Colombia; [§]Department of Oral and Maxillofacial Surgery, Pontifical Xavierian University, Havana, Cuba; and [||]Department of Neurosurgery, Nueva Granada Militar University, Militar Central Hospital, Bogotá, Colombia.

Received February 16, 2020.
Accepted for publication March 16, 2020.

Address correspondence and reprint requests to José Manuel Valdés Reyes, DDS, Department of Oral and Maxillofacial Surgery, Pontifical Xavierian University, Street 19 between 10 and 12, Havana, Cuba; E-mail: estomatojose@gmail.com, jose_valdes@javeriana.edu.co

The authors report no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0000000000006577

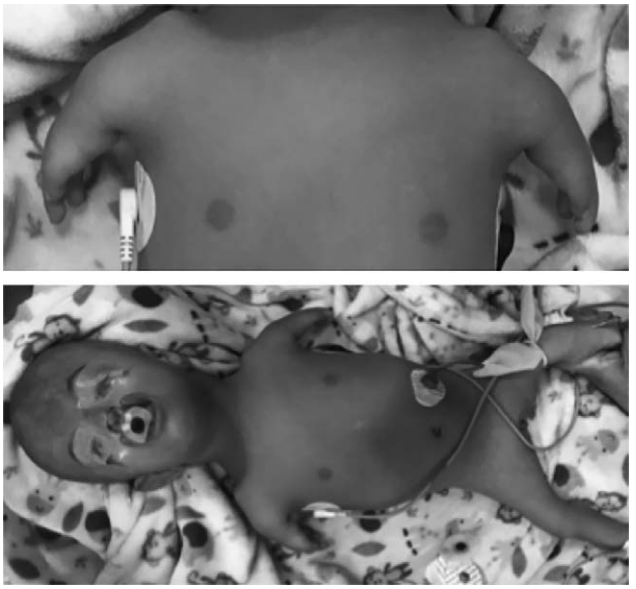

FIGURE 1. This is a 2-year-old female patient with Roberts Syndrome. The patient presented growth retardation and limb malformations.

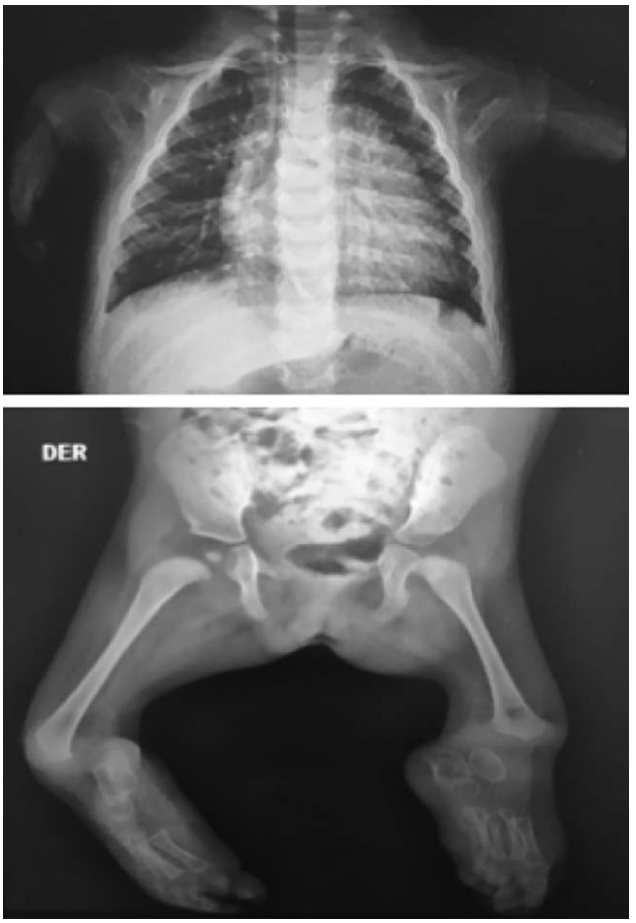

FIGURE 2. X-ray that manifests a deficiency or shortening of the proximal to mid portions of the limbs.

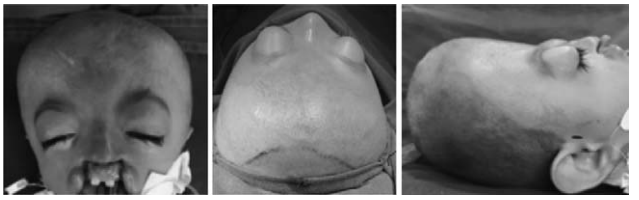

FIGURE 3. The craniofacial anomalies presented were cleft lip and cleft palate, premaxillary prominence, malar flattening, frontal flattening, exophthalmos resulting from shallow orbits, ear malformations, and frontal hemangioma.

Institute, presenting a Roberts Syndrome diagnosticated by genetics in the second week of life with mutations in the *ESCO2* gene [8p21.1] (Fig. 1).

The patient presented growth retardation and limb malformations include symmetric mesomelic shortening and anterior-posterior axis involvement, hand malformations include brachydactyly and oligodactyly (Fig. 1). In the skeletal x-ray, it characteristically manifests as a deficiency or shortening of the proximal to mid portions of the limbs (Fig. 2).

The craniofacial anomalies (Fig. 3) presented were cleft lip and cleft palate, premaxillary prominence, malar flattening, frontal flattening, exophthalmos resulting from shallow orbits, ear malformations, frontal hemangioma and bilateral craniosynostosis of the coronal suture or brachycephaly (Fig. 4A–C).

The patient comes with signs of endocranial hypertension, gyral impressions on the inner table of the skull, and bilateral craniosynostosis of the coronal suture (Fig. 4), the management with a craniotomy leaves a strip of bone over the vertex and 2 lateral struts from the vertex to the temporal squamosal region and this includes supraorbital rim advancement in conjunction with bifrontal craniotomy in this case of bilateral coronal synostosis, for which the deformity involves the fronto orbital región (Fig. 5), we leaving the periosteum attached to the underlying bone also assists in holding the bone together if it fractures during later remodeling. The orbital osteotomy is performed after a bifrontal craniotomy, which extends approximately 5 mm above the supraorbital rim, the frontal bone then is remodeled. Barrel osteotomy was performed in the orbits for resolve the exophthalmos.[7]

Bilateral supraorbital rim advancement and bilateral temporalis composite advancement are performed. The struts extending from the vertex are reduced in height and shifted posteriorly. Wires are passed between the inferior tip of the struts and the posterior basal temporal bone and gradually cinched down to reduce the height of

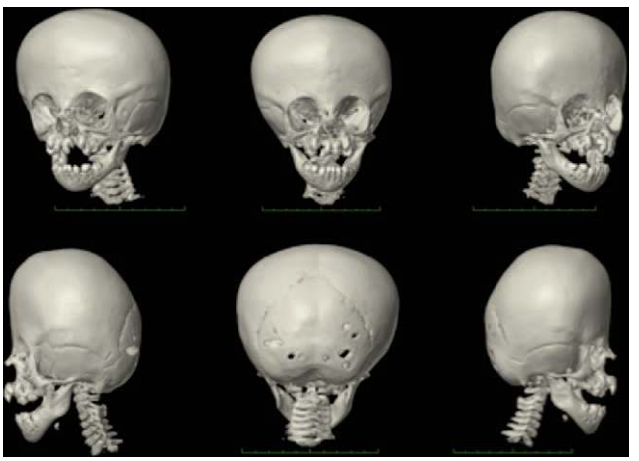

FIGURE 4. Bilateral craniosynostosis of the coronal suture.

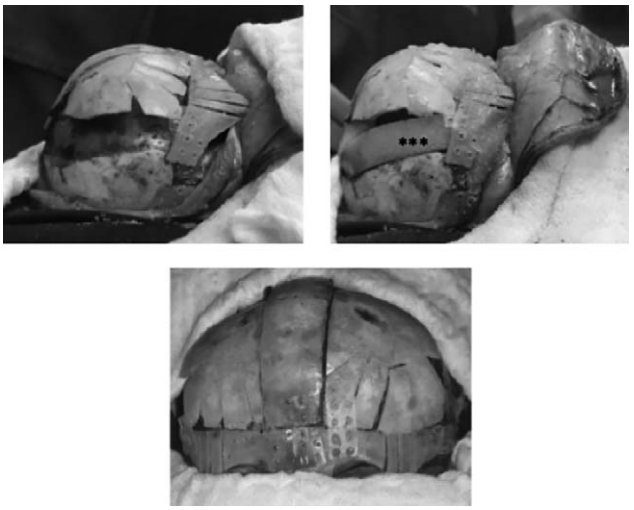

FIGURE 5. Bilateral supraorbital rim advancement and bilateral temporalis composite advancement. Bone grafts ∗∗∗were performed in gaps of the skull.

the skull. Bone grafts ∗∗∗ were performed in gaps of the skull (Fig. 5).

The procedure improved intracranial hypertension, frontal projection and exophthalmos, postoperative tomography is presented (Fig. 6). The procedure was performed without complications, Figure 7 shows the evolution of before and after surgery. In a second time, the surgery of the cleft lip and cleft palate will be done.

DISCUSSION

The patient displayed premature separation of the centromeres and craniofacial anomalies and limb deformities, findings that served as confirmation of the diagnosis of Roberts Syndrome including bilateral coronal craniosynostosis. The diagnosis of craniosynostosis is mainly given by the shape of the newborn's head and is corroborated by complementary images. Approximately 8% of craniosynostosis are syndromic and the rest is due to malpositions of the child during its growth stage.[8,9]

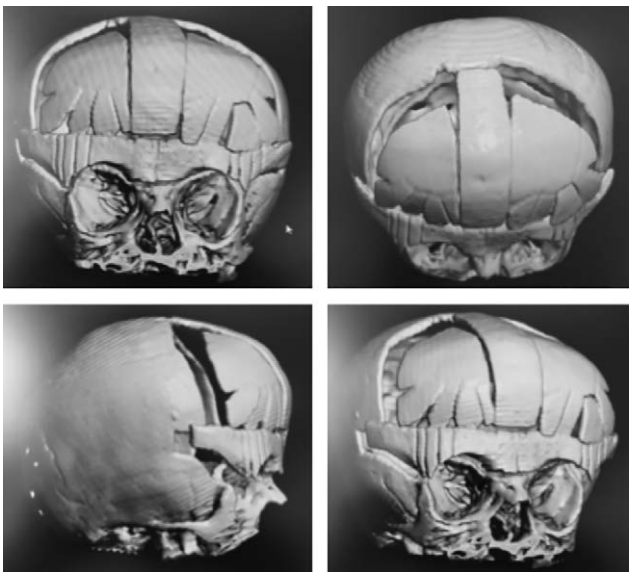

FIGURE 6. Postoperative tridimensional tomography.

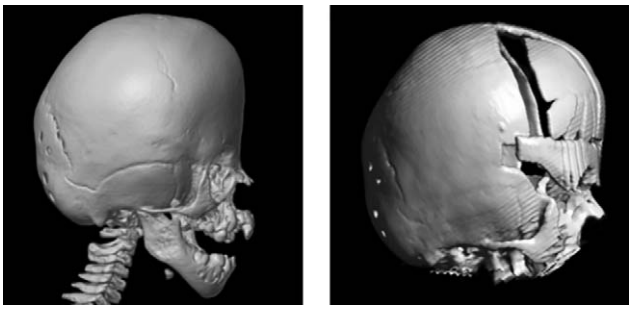

FIGURE 7. Evolution before and after surgery.

The management for craniosynostosis is very varied, osteogenic distraction and fronto-orbital advancement have been used specially in the anterior craneal vault deformities. This procedure is generally used in plagiocephaly, in turricephaly, and in the metopic synostosis (trigonocephaly). This technique, which is carried out by means of a coronal approach, is complemented by an advance of the fronto-orbital bar and a fixation with resorbable plates, achieving an adequate frontal projection and reducing the exophthalmos present and increasing the anteroposterior diameter of the skull.[10]

The fronto-orbital advancement was described by Tessier for the treatment of craniosynostosis. It has been described mainly in the Crouzon syndrome and Apert syndrome.[11] In the literature their arent case reports of this procedure in Roberts Syndrome.

In bicoronal synostosis, correcting bilateral forehead and supraorbital rim width and retrusion (brachycephaly) is necessary to normalize fronto-facial balance and to afford orbital protection. Current techniques involve a coronal craniotomy for resolve the first problem limited cranial volume-restricting central nervous system development.[7] In this uncommon case of Roberts Syndrome associated with coronal craniosynostosis, a fronto-orbital advance was made without complications, the patient continued her neurodevelopment without complications and the endocranial hypertension was eliminated.

CONCLUSION

Roberts Syndrome is a rare disease, which can have different skeletal variations. This syndrome can manifest itself with craniosynostosis, with the requirement of a comprehensive management to correct it and avoid compression of the brain with endocranial hypertension.

REFERENCES

1. Roberts JB. A child with double cleft lip and palate, protrusion of the intermaxillary portion of the upper jaw and imperfect development of the bones of the four extremities. *Ann Surg* 1919;70:252–253
2. Herrmann J, Feingold M, Tuffli GA, et al. A familial dysmorphogenetic syndrome of limb deformities, characteristic facial appearance and associated anomalies: the "pseudothalidomide" or "SC-syndrome." *Birth Defects Orig Art Ser* 1969;V:81–89
3. Hall BD, Greenberg MH. Hypomelia-hypotrichosis-facial hemangioma syndrome (pseudothalidomide, SC syndrome, SC Phocomelia syndrome). *Am J Dis Child* 1972;123:602–604
4. Zhou J, Yang X, Jin X, et al. Long–term survival after corrective surgeries in two patients with severe deformities due to Roberts syndrome: a case report and review of the literature. *Exp Ther Med* 2018;15:1702–1711
5. Vega H, Waisfisz Q, Gordillo M, et al. Roberts syndrome is caused by mutations in ESCO2, a human homolog of yeast ECO1 that is essential for the establishment of sister chromatid cohesion. *Nat Genet* 2005;37:468–470

6. Gordillo M, Vega H, Jabs EW. Roberts Syndrome. 2006 Apr 18 [Updated 2013 Nov 14]. In: Adam MP, Ardinger HH, Pagon RA, et al. eds. GeneReviews. Seattle, WA: University of Washington, Seattle; 1993–2019. Available in: https://www.ncbi.nlm.nih.gov/books/NBK1153/
7. Cochrane DD. Operative techniques in pediatric neurosurgery. 2000. By A. Leland Albright, Ian F. Pollack, P. David Adelson. Published by Thieme. 312pages. C$263.13 approx. Can J Neurol Sci. 2001;28:377–378.
8. Governale L. Craniosynostosis. *Pediatr Neurol* 2015;53:394–401
9. Mendonca D, Gejje S, Kaladagi N. Fronto-orbital advancement: revisited. *J Cleft Lip Palate Craniofac Anomal* 2015;2:20–26
10. Ching H, Wu C, Wen-Ching E, 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;00:00–10
11. Tessier P. Total facial osteotomy. Crouzon's syndrome, Apert's syndrome: oxycephaly, scaphocephaly, turricephaly. *Ann Chir Plast* 1967;12:273–286

New Solutions to Improve the Accuracy of the Navigation-Guided Foreign Body Removal in Craniomaxillofacial Deep Space

Chuxi Zhang, MD,† Jinyang Wu, DDS,*† Chengshuai Yang, MDS,*† Nian Liu, DDS,*† Wenyu Hui, MDS,*† Yong Zhang, DDS,*† and Shilei Zhang, DDS, MD*†*

Objective: Surgical navigation-guided removal of foreign bodies in the craniomaxillofacial region has been proven to be an effective method. However, there have been some unsuccessful patients due to reduced navigation accuracy or complicated and undetectable anatomy. This article summarizes the experience and proposes some solutions to achieve better results.

Study design: Two solutions were proposed to optimize the surgical navigation procedure: using a 3-dimensionally printed customized mandible retainer to indirectly maintain the consistency of the foreign body's visual images of preoperative planning and intraoperative navigation and importing real-time endoscopic imaging during surgery to provide vision under complex anatomy. Two patients were selected for each method.

Results: The foreign bodies were successfully and minimally invasively removed in all patients assisted by optimized surgical navigation. During follow-up at 3 to 6 months postoperatively, no complications were found.

Conclusion: Improving navigation accuracy and providing real vision might be effective at compensating for insufficient navigation due to navigation positioning errors or the interference of imperceptible and complicated anatomy.

Key Words: Craniomaxillofacial, 3-dimensional printing, endoscope, foreign body, surgical navigation

Foreign bodies retained in the craniomaxillofacial region often cause local infections, pain, and even life-threatening effects, which have serious effects on the aesthetics, function, and psychology of the patient. The maxillofacial region has a complex anatomical structure which brings great difficulties for foreign body removal surgery.

The key to removing foreign bodies is precise localization and reasonable surgical approaches.[1] An image-guided navigation system allows the surgeon to synchronize the intraoperative position of the instruments with computed tomography (CT) imaging of the anatomy of the patient[2] and gives the surgeon continuous real-time orientation intraoperatively,[3] which has become an effective and widely accepted method to improve outcomes, reduce operative time and minimize the possibility of craniocerebral injury.[4–9]

Between 2008 and 2019, 30 patients underwent foreign body extraction with optical navigation-assisted systems, but 2 of them were unsuccessful (Supplementary Digital Content, Table 1, http://links.lww.com/SCS/B529). According to preoperative CT, the location of the foreign bodies in both patients was in the pterygopalatine space. The positioning error of the navigation caused by image shift and the interference of complex anatomy may be the main causes of these instances of surgical failure. To solve these problems, we propose 2 new solutions and summarize the relevant experience.

METHODS

Patients

Four patients (Supplementary Digital Content, Table 2, http://links.lww.com/SCS/B529) with foreign bodies retained in the oral and maxillofacial deep space after trauma were admitted to the Department of Oral and Cranio-maxillofacial Surgery, Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, from September 2016 to July 2019. The patients underwent

From the *Department of Oral and Cranio-maxillofacial Science, Shanghai Ninth People's Hospital, College of Stomatology, Shanghai Jiao Tong University School of Medicine; and †National Clinical Research Center, Shanghai Key Laboratory of Stomatology and Shanghai Research Institute of Stomatology, Shanghai, China.

Received October 13, 2019.
Accepted for publication March 19, 2020.

Address correspondence and reprint requests to Professor Shilei Zhang, DDS, MD, and Yong Zhang, DDS, Department of Oral and Cranio-maxillofacial Science, Shanghai Ninth People's Hospital, College of Stomatology, Shanghai Jiao Tong University School of Medicine; National Clinical Research Center, Shanghai Key Laboratory of Stomatology and Shanghai Research Institute of Stomatology, Shanghai, China; E-mails: leinnymd@hotmail.com, zhangyong1362@163.com
CZ and JW contributed equally and are co-first authors.

This work was supported by the National Natural Science Foundation of China (81701034, 81671035), the Project from Cooperative Innovation Center of Translational Medicine (TM201717), Shanghai Municipal Health Commission (201840075), the Shanghai Ninth People's Hospital Affiliated Shanghai JiaoTong University School of Medicine Technology Innovation Fund (CK2018003), and the Project from Shanghai Key Laboratory of Medical Imaging Computing and Computer Assisted Intervention (17DZ2272300-1, 17DZ2272300-2, 17DZ2272300-3)
The authors report no conflicts of interest.
Supplemental digital contents are available for this article. Direct URL citations appear in the printed text and are provided in the HTML and PDF versions of this article on the journal's Web site (www.jcraniofacialsurgery.com).

ISSN: 1049-2275
DOI: 10.1097/SCS.0000000000006584