The Use of Virtual Surgical Planning in Total Facial Skeletal Reconstruction of Treacher Collins Syndrome: A Case Report

Yifan Guo, MD[1] Joseph Lopez, MD, MBA[2] Robin Yang, MD, DDS[2]
Alexandra Macmillan, MA (Cantab), MBBS[2] Amir H. Dorafshar, MBChB[2]

[1] Department of Plastic and Reconstructive Surgery, Brody School of Medicine at East Carolina University, Greenville, South Carolina
[2] Department of Plastic and Reconstructive Surgery, JHMI, Baltimore, Maryland

Address for correspondence Amir H. Dorafshar, MBChB, Department of Plastic and Reconstructive Surgery, JHMI, 601 N. Caroline St., Suite 8161, Baltimore, MD 21231 (e-mail: adorafs1@JHMI.EDU).

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Abstract

Facial skeletal reconstruction of patients with severe Treacher Collins syndrome (TCS) requires correction of both midface and mandibular deficiencies. Implementing virtual surgical planning can provide an accurate three-dimensional analysis of craniofacial abnormalities, creating calvarial donors that match the anatomy of the desired malar augmentation and facilitating bimaxillary movements, positioning, and fixation in orthognathic surgery. We present a case of an 18-year-old patient with TCS, who underwent staged zygomaticomaxillary reconstruction and double-jaw osteotomy with sliding genioplasty, using computer-assisted surgical planning. Following these operations, the patient achieved not only improved facial harmony but also class I occlusion.

Keywords
- CAD/CAM
- virtual surgical planning
- Treacher Collins syndrome
- midface hypoplasia
- mandibular distraction

Treacher Collins syndrome (TCS) management requires multiple complex operations to address orbitomalar and maxillomandibular abnormalities as well as reconstruction of soft-tissue abnormalities to achieve the ever elusive aim of normalcy. Goals of facial skeletal reconstruction in patients with TCS are (1) to establish anatomic midfacial projection, (2) to correct facial convexity, and (3) and to improve facial skeletal discrepancy to relieve airway obstruction and occlusal relationships.[1] Early techniques of surgical reconstruction have been described by Tulasne and Tessier.[2] The initial stage of reconstruction usually involves mandibular distraction during infancy, if significant airway obstruction is present.[3] Orbitozygomatic reconstruction is then performed at a second stage, typically after 5 years of age.[4] Complex orthognathic surgery, including Le Fort I osteotomy, bilateral sagittal-split osteotomy, and genioplasty, is indicated to improve maxillomandibular relationships and to improve mandibular retrusion.[5]

Virtual surgical planning (VSP) provides surgeons with an opportunity to preoperatively design osteotomies, simulate skeletal movements with visualization of operative options in multistage procedures, and evaluate three-dimensional (3D) bony reconstruction.[6,7] It has been well embraced across multiple specialties.[8–12] Surgical cutting guides and occlusive splints can be fabricated by computer-aided design and computer-aided manufacturing (CAD/CAM), based on patient-specific anatomy and desired outcome. This case illustrates the advantages of using VSP in an adult patient with TCS, who presented in a delayed fashion, to help design anatomically shaped and sized midfacial bone grafts, and for accurate orthognathic surgical planning.

Case Report

An 18-year-old female patient with history of TCS was referred to our institution to discuss the surgical options for improvement in facial appearance. On craniofacial examination, the patient exhibited severe midface hypoplasia, down-slanting palpebral fissure, and a retromicrognathic jaw (►**Fig. 1**). Of note, her past surgical history was noted for

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Fig. 1 Preoperative photos with three-dimensional computed tomography. Patient has malar hypoplasia with complete absence of zygomatic arch and malar prominence, hypoplastic lateral orbital wall, and inferior orbital rim. Patient also has retromicrognathia with anterior open bite.

a tracheostomy and percutaneous endoscopic gastrostomy tube placement while in infancy, and bilateral microtia reconstructions as a child.

Stage 1: Zygomaticomaxillary Complex Reconstruction

Given the extent of her mid- and lower facial hypoplasia, a multistage approach for surgical reconstruction was designed to augment her midface and to advance her retrognathic jaw position. The first stage focused on midface reconstruction. Her primary concern was midface hypoplasia and we wished to set the midface in projection prior to performing orthognathic surgery. Using VSP, we realized that a Le Fort II osteotomy would not have given sufficient bone stock for a subsequent bilateral split sagittal osteotomy. We thus elected to use full-thickness autologous calvarial bone as an onlay augmentation. Computed tomography (CT) of the face and brain with 1-mm-thin cuts and 3D reconstruction were performed. The imaging data were then submitted (3D Systems; Rock Hill, SC) for VSP. The location of calvarial bone graft harvest was selected based on the desired contour, shape, and optimal thickness of the final malar autologous implant (►**Fig. 2**). Cutting guides were designed to allow craniotomy to be performed in a safe location, away from the sagittal sinus. The craniotomy cutting guide was designed so that it would allow adequate amount of split-thickness calvarial bone graft to reconstruct the donor site (►**Fig. 2**).

Once the temporoparietal region was exposed through a coronal incision, the cutting guides were placed and donor sites were marked, and the craniotomy was then performed. The calvarial bones were then marked following the contour of zygoma using the cutting guides. After the osteotomy was performed, the remains of the calvarial bone were then split to perform a cranioplasty of the donor defect.

A subperiosteal pocket was dissected in the midface, and the temporalis muscle was reflected off the infratemporal fossa to allow exposure of the zygomatic process of the temporal bone. Medially, the inferior orbital rim and anterior maxillary wall were fully exposed with maxillary buccal incisions, and the inferior orbital nerve was protected. Once the calvarial bone graft was anatomically placed into the subperiosteal pocket in the midface, it was secured

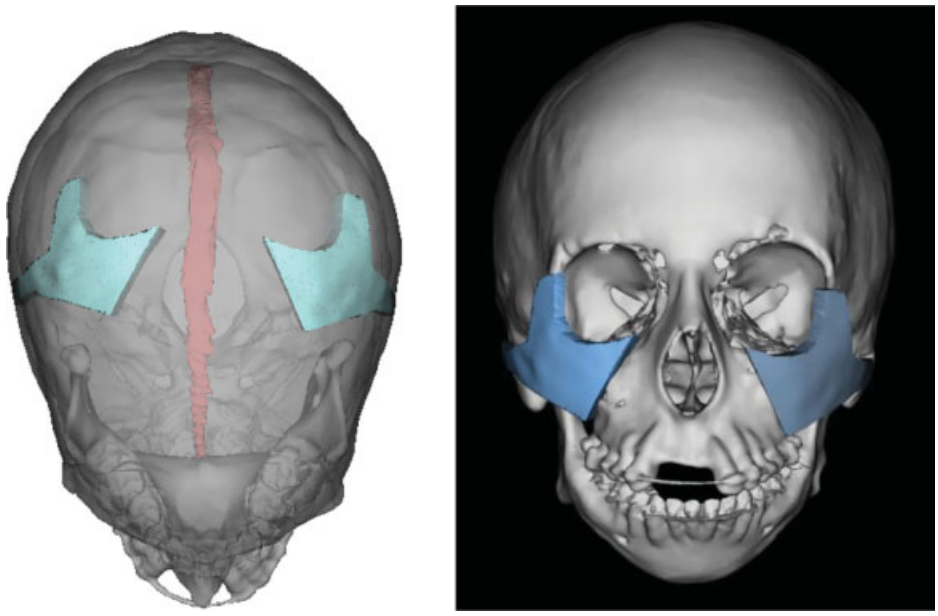


Fig. 2 Computer-aided design and computer-aided manufacturing. Zygoma templates were designed based on normal anatomy. The templates were transposed to parietal bone, where ideal calvarial donor sites were chosen based on shape, contour, and thickness.

laterally to the zygomatic process of the frontal bone using rigid fixation. The midface soft tissue was re-elevated to the lateral orbital rim with suspension sutures. To improve the position of the lower eyelids, bilateral canthopexies were secured to the bone superior and medial to the zygomaticofrontal suture. After the temporalis muscle was resuspended, the coronal incision was closed over a temporary drain. A postoperative CT scan on postoperative day 1 demonstrated excellent zygomatic reconstruction with satisfactory anatomic positioning and symmetric projection (►**Fig. 3**).

Stage 2: Orthognathic Surgery

The patient underwent a complete preoperative orthodontic assessment prior to orthognathic surgery. Given the degree of lower facial asymmetry and the complexity anticipated with bimaxillary movements, we utilized VSP to design the operation. Standard cephalometrics were performed based on 3D CT (►**Table 1**). Computer-aided simulation surgery was performed to first advance and rotate the mandible in the counter-clockwise direction to decrease the obtuse gonial angle, lengthen the horizontal body, and correct the class II malocclusion. This was all followed by performing a Le Fort I osteotomy to correct the resulting posterior open bite and to elongate the shortened posterior maxilla (►**Fig. 4**). Intermediate and final dental splints were manufactured based on occlusive patterns. A sliding genioplasty was designed to correct her retrusive chin and a cutting guide was fabricated.

The patient subsequently underwent bilateral sagittal-split osteotomy to allow horizontal advancement and counter clockwise rotation of the mandible. The intermediate occlusive splint was then placed to allow for proper positioning of the mandible against the stable maxilla to ensure centric relation. A standard Le Fort I osteotomy was then performed followed by a genioplasty using standard VSP cutting guides. Overall, these orthognathic procedures decreased the facial vertical height, corrected the retrusive chin, and closed the previously open bite.

Postoperatively, the patient completed final orthodontic adjustments with class I occlusion (►**Table 1**). She has symmetric and excellent malar projection. Her facial convexity is now corrected with much improved facial aesthetics (►**Fig. 5**).

Cephalometric Analysis

Preoperative and postoperative cephalometric analysis was performed digitally using Dolphin imaging software (Dolphin Imaging & Management Solutions, Chatsworth, CA). Standard cranial, maxillary, mandibular, and dental landmarks were identified in the usual fashion. An appropriate analysis was performed to determine the change in craniofacial relationships postsurgically.

The hallmark dysmorphology of TCS is well described. The goal of cephalometric analysis in this patient population is to quantify the powerful changes that can be made with maxilla-mandibular surgery with regard to both function (airway and occlusion) and overall facial harmony. Preoperative analysis revealed the usual craniofacial abnormalities associated with TCS (►**Fig. 6**, ►**Table 2**). Preoperative lateral cephalometric analysis revealed maxillary deficiency in the anterior-posterior dimension as well as the vertical dimension when compared with the cranial base (sella–nasion–A angle [SNA], posterior nasal spine to nasion [PNS to N]). The mandible was also globally deficient in most vectors. There were both mandibular body and ramus height deficiencies (gonion to pogonion [Go to Pg] and anterior nasal spine to gonion [ANS to Go]). There was also a significant difference between the position of the maxilla and the mandible (A–nasion–B angle [ANB]). Postsurgically, there was significant return-to-normal values of

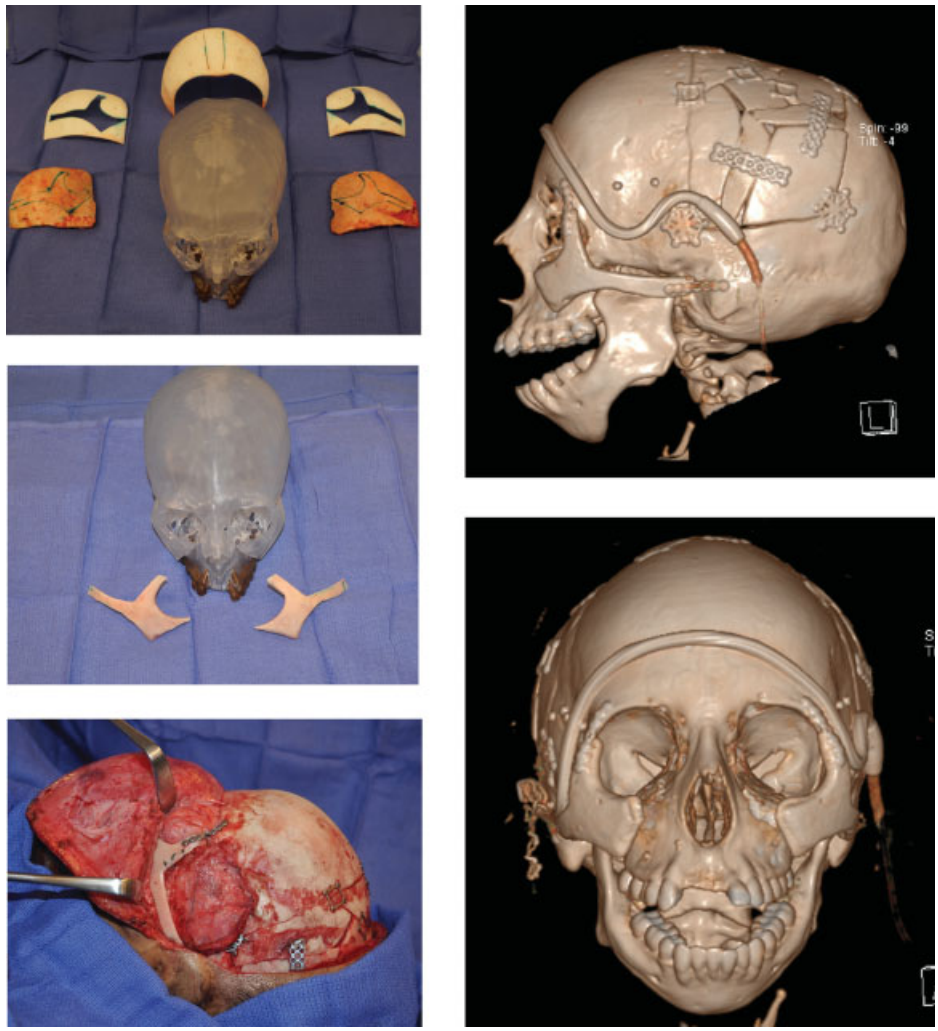


Fig. 3 Intraoperative photos and postoperative imaging demonstrate the positioning of the zygoma implants and total coverage of the donor sites using split parietal bone.

Table 1 Planned movements of cephalometric landmarks as determined in the virtual forum in the planning stage, using 3D CT scans.

Point	Name	Anterior/Posterior	Left/Right	Up/Down
ANS	Anterior nasal spine	0.39 mm posterior	2.01 mm left	0.29 mm up
A	A point	0.52 mm anterior	1.14 mm left	0.41 mm down
ISU1	Midline of upper incisor	1.91 mm anterior	1.39 mm left	0.11 mm down
U3L	Upper left canine	0.05 mm posterior	1.13 mm left	0.44 mm down
U6L	Upper left anterior molar (mesiobuccal cusp)	1.71 mm posterior	1.78 mm right	3.02 mm down
U3R	Upper right canine	4.71 mm anterior	1.15 mm left	0.17 mm down
U6R	Upper right anterior molar (mesiobuccal cusp)	5.54 mm anterior	1.50 mm right	2.41 mm down
ISL1	Midline of lower incisor	8.15 mm anterior	2.53 mm right	2.26 mm up
L6L	Lower left anterior molar (mesiobuccal cusp)	7.78 mm anterior	1.40 mm right	3.51 mm down
L6R	Lower right anterior molar (mesiobuccal cusp)	6.12 mm anterior	1.53 mm right	0.60 mm down
B	B point	12.41 mm anterior	2.93 mm right	1.13 mm down
Pog.	Pogonion	22.03 mm anterior	0.91 mm right	4.64 mm down

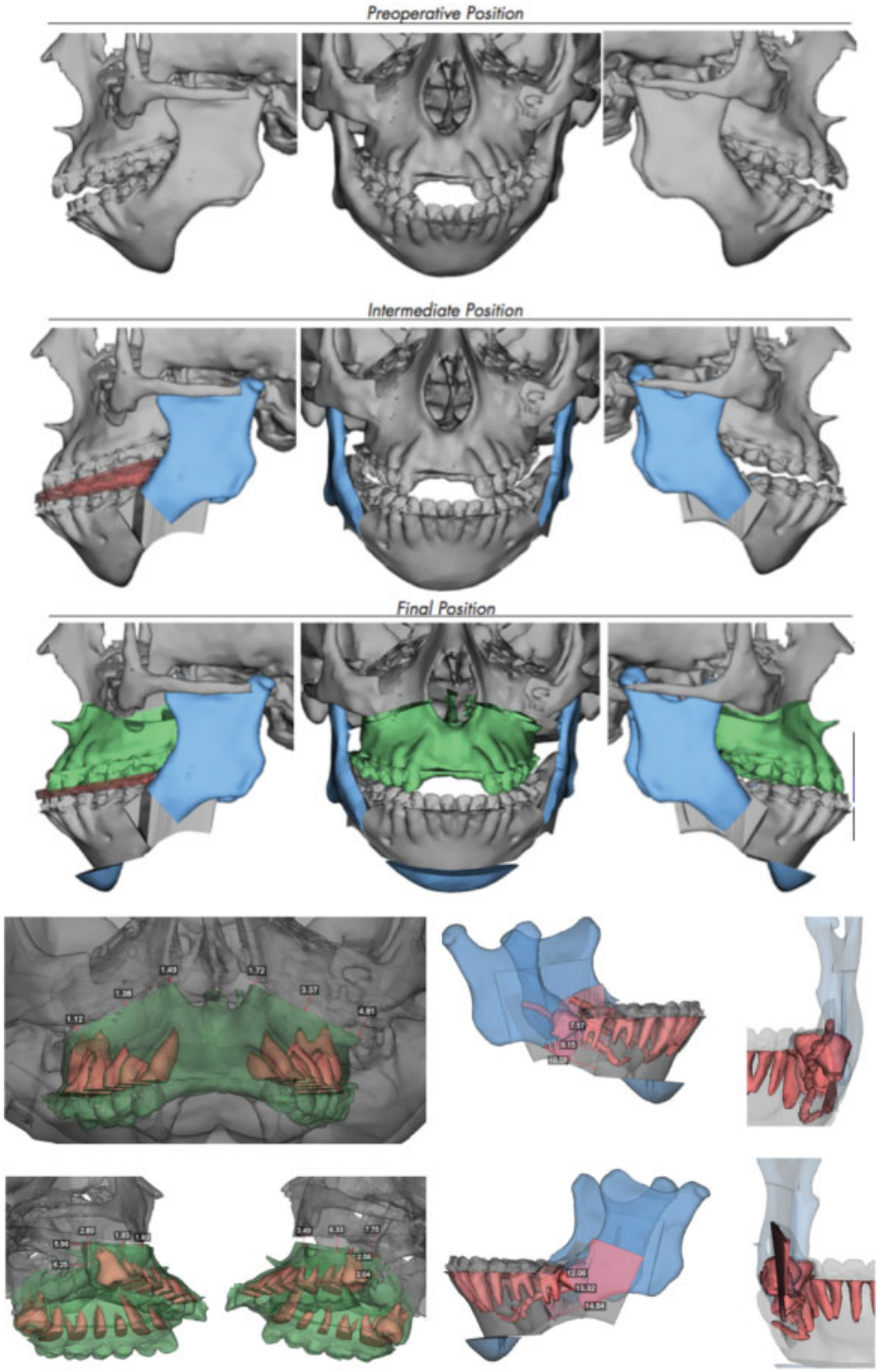


Fig. 4 Computer-aided design and computer-aided manufacturing for orthognathic surgery. Bilateral sagittal split osteotomy with advancement and counter-clockwise rotation of the mandible, Le Fort I osteotomy to lengthen the posterior maxilla, and sliding genioplasty to correct the retrusive chin position. The dental anatomy and inferior alveolar nerves were marked to allow osteotomies to be performed in a safe and predictable fashion.

the maxilla and the mandible when compared with the cranial base (sella–nasion–B angle [SNB] and SNA), with a better balance between the two structures (ANB). Mandibular body length was also lengthened to a more normal value. Utilization of the bilateral sagittal split for the mandible did not have significant effects on the ramus height of the mandible. There was no significant change in the occlusal plane measurements as well as the posterior/anterior facial heights.

Discussion

We describe the use of VSP in the management of a case of an adult with delayed presentation of TCS. Since the primary



Fig. 5 Postoperative photos and three-dimensional computed tomography. Patient now has improved malar projection. The severe facial convexity has been corrected with improved facial aesthetics.

concern of the patient was midface hypoplasia, we addressed this in the first instance. VSP enabled choice of optimal surgical technique. We used calvarial split graft to reconstruct anatomically individualized zygomas, followed by reconstruction of soft tissue and bilateral canthopexies. VSP allowed planning of the orthognathic stage, which comprised bilateral sagittal-split osteotomy, a Le Fort I osteotomy, and genioplasty. Postoperative cephalometric analysis demonstrates excellent outcome with much improved anthropometric measurements (►**Table 2**).

Typically, bony reconstruction is required to restore malar projection. Autologous calvarial onlay bone grafting is the most commonly used technique.[1,13] Other techniques including fat grafting and alloplastic materials are of limited use or associated with complications.[14–16] Bone grafting is ideally performed at an older age for two reasons. As the diploic space and skull bicortical thickness become fully developed after 9 years of age, splitting of the calvarial bone of adequate thickness can be performed with relative ease for reconstruction of the donor site.[17] In addition, in younger children, the surgeon must take into account growth of the facial structures. Malar osteotomy with lateral rotation of the zygoma has been previously described.[18] However, this requires additional calvarial bone graft for zygomatic arch reconstruction and bridging of osteotomy gaps. Le Fort II osteotomies have also been described with subsequent bilateral sagittal split osteotomy (BSSO) of the mandible; however, the VSP technique helped us to recognize that the BSSO required to achieve normal occlusion would not have been anatomically possible in our patient.

VSP enables safe identification of calvarial graft of appropriate thickness and contour to achieve malar projection. It is difficult to accurately create the desired 3D shape and contour of the calvarial bone graft using traditional techniques. Without VSP, after the calvarial donor graft is osteotomized to the shape of zygoma, additional bone bending is required to match the contour of the malar region. This weakens the strut and can contribute to relapse and the need for further reconstruction. With computer-assisted presurgical planning, the desired zygomaticomaxillary complex is designed first, and then the calvarial donor site can be selected matching both the shape and contour of the implant.

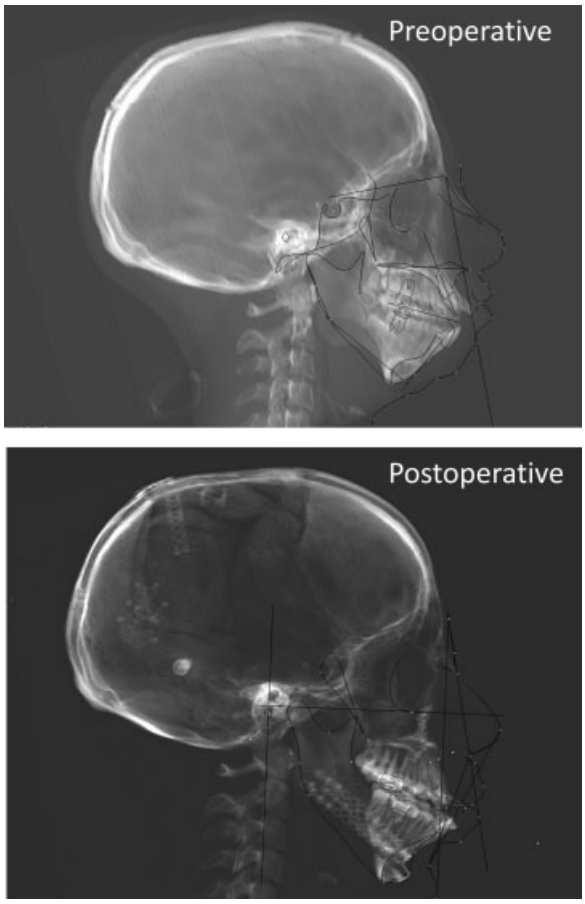


Fig. 6 Cephalometric images pre- and postoperatively showing an improvement in mandibular body length and an improvement in positions of maxilla and mandible.

Table 2 Pre- and post-operative cephalometric measurements and normal values. An improvement in measurements after operative intervention trending towards the normal range is evident.

	Pre-op	Post-op	Normal
SNA (mm)	78.6	82.4	82 ± 2
SNB (mm)	67.0	75.2	80 ± 2
ANB (°)	11.5	7.3	2 ± 2
PNS to N (perp HP)	41.6	41.6	51 ± 2
N to ANS	53.2	48.8	50.0 ± 2.4
Mandibular plane	55.8	50.6	24 ± 5
Gonial angle (Ar–Go–Gn)	151	148	122 ± 7
Go–Pg (mm) (mandibular body length)	55.1	66.8	74 ± 6
Ar to Go (mm) (mandibular ramus)	35.5	36.2	46.8 ± 2.5
Occlusal plane	32	32	14 ± 3

Abbreviations: ANB, A–nasion–B angle; ANS, anterior nasal spine; Ar, articulare; Gn, gnathion; Go, gonion; N, nasion; Pg, pogonion; PNS, posterior nasal spine; SNA, sella–nasion–A angle; SNB, sella–nasion–B angle.

The patient-specific cutting guides can be produced through the CAD/CAM process, making this previously difficult process relatively straightforward.

Orthognathic surgery is challenging in TCS, as it requires bimaxillary movements and rotations in multiple planes. Although traditional model surgery provides excellent correction at the occlusal level, the planning process can be time consuming and optimal outcomes can be difficult to achieve when significant facial asymmetry is present.[19] VSP is particularly useful in such cases. Standard cephalometric analysis can be accurately performed on 3D CT images. Virtual surgery can be performed to simulate desired repositioning of the maxilla and mandible segments. Cutting guides, positioning guides, and oral splints can be fabricated. This allows greater accuracy in performing the osteotomy and in positioning the osteotomized segments according to predetermined bony movements.[20]

In conclusion, VSP is a reliable, accurate, and safe surgical tool in midface and orthognathic reconstruction of late-presenting TCS. Based on our early experience, both aesthetic and functional results are highly encouraging, demonstrated by cephalometric improvements postoperatively. VSP allowed consideration of various surgical options for all parts of this multistage procedure, allowing identification of challenges in advance. This case demonstrates the advantages of using VSP to visualize and choose optimal procedures prior to surgery, design anatomically individualized midfacial bone grafts, and accurately plan orthognathic surgery in a delayed presentation of TCS.

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