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Clinical Case Report

Virtual planning for orbital reconstruction in a craniofacial fibrous dysplasia surgical approach – Case report

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ABSTRACT

The present case report aims to describe a surgical approach procedure for an orbital reconstruction in a patient with craniofacial fibrous dysplasia, planned with virtual mirroring and simulation using a 3D-printed realistic model. The patient was followed up by the neurosurgery and the oral and maxillofacial surgery teams and was diagnosed with monostotic fibrous dysplasia in the right frontal bone, affecting the right orbital roof. Our team performed surgical planning through virtual mirroring using the non-affected side. The surgical procedure was then performed with surgical access through the frontal bone to the orbital roof and reconstruction using a titanium plate. Both surgery teams monitored the patient, who presented no postoperative complications. (Rev Port Estomatol Med Dent Cir Maxilofac. 2024;65(3):142-147)

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Planeamento virtual para reconstrução da órbita numa abordagem cirúrgica de displasia fibrosa craniofacial – Caso clínico

Palavras-chave:
Displasia fibrosa craniofacial
Displasia fibrosa óssea
Impressão tridimensional

RESUMO

O presente relatório de caso tem como objetivo descrever um procedimento cirúrgico para reconstrução orbitária, através de planeamento por espelhamento virtual e simulação com modelo realístico impresso em 3D, utilizado para abordagem cirúrgica de displasia fibrosa craniana. Paciente sob acompanhamento pelas equipas de Neurocirurgia e Cirurgia e Traumatologia Oromaxilofaciais, diagnosticada com displasia fibrosa monostótica frontal à direita, com acometimento de teto orbitário direito. Foi realizado planeamento cirúrgico por meio de espelhamento digital com o lado não patológico e, posteriormente, procedimento cirúrgico com acesso cirúrgico pelo osso frontal ao teto da cavidade orbitária e reconstrução com placa de titânio. A paciente foi monitorizada por ambas as equipas e não apresentou complicações cirúrgicas pós-operatórias. (Rev Port Estomatol Med Dent Cir Maxilofac. 2024;65(3):142-147)

Introduction

Fibrous dysplasia (FD) is a genetic and non-hereditary disease in which normal bone tissue is replaced by immature and randomly distributed fibro-osseous tissue, resulting in deformity, fractures, pain, and functional impairment.[1-3] The term "craniofacial fibrous dysplasia" (CFD) describes the FD that affects the cranial and facial bones.[1]

FD represents 5% to 7% of all benign bone tumors and is classified as monostotic if only one bone is involved or polyostotic if multiple bones are involved.[2] Unlike FD in extremity bones (commonly with fractures), CFD usually presents with slow-growing lesions, causing gradual and painless swelling and facial asymmetry.[2,4] Lesion growth can be external (and visible) or internal, filling facial sinuses, invading the orbital region, or both.[4] Due to the high concentration of vital structures in this region, these lesions can result in severe complications, and their treatment presents unique challenges.

CFD management represents a dilemma for surgeons due to the orbit and skull base anatomic boundaries. The CFD treatment goal is to correct or prevent functional problems while reaching aesthetic improvement. The decision stands between performing total excision of the lesion (and sometimes causing high deformity) or excision of just the aesthetic contouring of the craniofacial skeleton and further monitoring.[5] Virtual planning techniques help determine with better precision up to which point we should remove the lesion, besides being extremely helpful in the subsequent planning of the necessary facial reconstruction in the intraoperative period.

In this scenario, we present a case report of a patient diagnosed with CFD in the right frontal bone affecting the orbital roof. We performed surgical planning for the orbital reconstruction through virtual mirroring and simulation using a 3D-printed realistic model. Despite the high complexity of the case, we achieved positive outcomes with this surgical procedure without complications.

Case Report

A 39-year-old female patient under treatment by the Oral and Maxillofacial Surgery Residency Program of the Dentistry School of Ribeirão Preto of the University of São Paulo (USP), São Paulo, Brazil, was first referred by the neurosurgery team of the Beneficência Portuguesa Hospital of Ribeirão Preto, São Paulo, Brazil, with the diagnosis of CFD of the right frontal bone affecting the right orbital roof.

The patient presented with the main clinical complaints of blurred vision, reduced vision field, and right superior eyelid ptosis for the past 3 years. She presented a positive medical history of pituitary adenoma diagnosed ten years earlier, a hypothesis previous to hormonal repercussions. Additionally, she had chronic asthma and used continuous medications consisting of inhaled salbutamol sulfate and inhaled beclomethasone dipropionate during asthma attacks. She also reported surgery for myopia correction on both ocular globes 18 years earlier with no apparent complications.

During the clinical exam, we observed facial asymmetry in the right orbital region, a solid volume increase during palpation in the right frontal area that extended to the right superior periorbital region, and no pain during palpation. We observed orbital dystopia between the right and left ocular globes and ptosis of the right superior eyelid. During the visual acuity test, the patient reported a reduced visual field of the right ocular globe in all directions and a slight motility impairment of ocular movements to the right side. The patient also reported paresthesia in the right ocular globe and right frontal bone areas.

We performed the preoperative evaluation through imaging exams using a multislice helical computerized tomography (CT) scan of the skull and face. CT scan was executed through volume acquisition in a multidetector dispositive, with no contrast. We observed a lesion in the right frontal bone area that caused thickening and heterogeneity of the medullary bone of the region and had a "ground glass" appearance, extending to the orbital roof and the right frontal sinus (Figure 1A-D). There was no evidence of the lesion extending beyond the bone tissue or periosteal reaction, and the other bone structures were preserved, such as brain ventricles, sulci, basal cisterns, and fissures. Moreover, there were no extra-axial collections nor any midline shifts.

Magnetic resonance imaging (MRI) evaluation was conducted using a 3 Tesla MRI scanner, acquiring multiplanar and multisequence images with specific coils, T2-weighted, FLAIR, T1-weighted, pre and post-contrast, SWAN, and diffusion. We observed thickening and heterogeneity of the medullary bone of the right frontal bone area, which also affected the orbital roof. It presented T2-hyperintensity and irregular structure more prominent in the medial region of the variation after contrast medium injection, showing imprecise limits and measuring approximately 5.6 x 3 centimeters (Figure 2A-D), in agreement with the previous CT scan report. Other structures showed regular shape and density.

After analyzing the preoperative image exams, we imported the CT scan DICOM files (Digital Imaging and Communication in Medicine) to a 3D surgery planning software (Mimics 19.0, Materialize NV, Leuven, Belgium). We performed the virtual mirroring (Figure 3A) of the non-affected side (left side) to use it as a model (Figure 3B) for the side that was affected by the FD (right side). After the mirroring and rendering, we printed a 3D model in resin (Figure 3C). The surgery planning was made through the 3D printed mode, allowing us to visualize and correct the titanium plate folding in the orbital roof for the right-side orbital reconstruction during surgery.

After finishing the surgical planning, we performed the surgery under general anesthesia alongside the neurosurgery team. We elevated the coronal flap and exposed the disease-affected side in the right frontal bone area. Following previous planning, we used drills, chisels, and surgical saws to perform the frontal bone craniotomy. We initially removed the bone affected by CFD and sent it for anatomopathological examination. Then, we created a window to the orbital roof (Figure 4A-B). Using surgical drills, we performed bone grinding until close to the orbital roof (next to the superior orbital fissure, optic canal, and the right frontal sinus) and then removed the bone tissue that compressed the remaining structures located in the foramina. The previously folded titanium plate was placed and adapted in the orbital roof for the orbital reconstruction and fixed

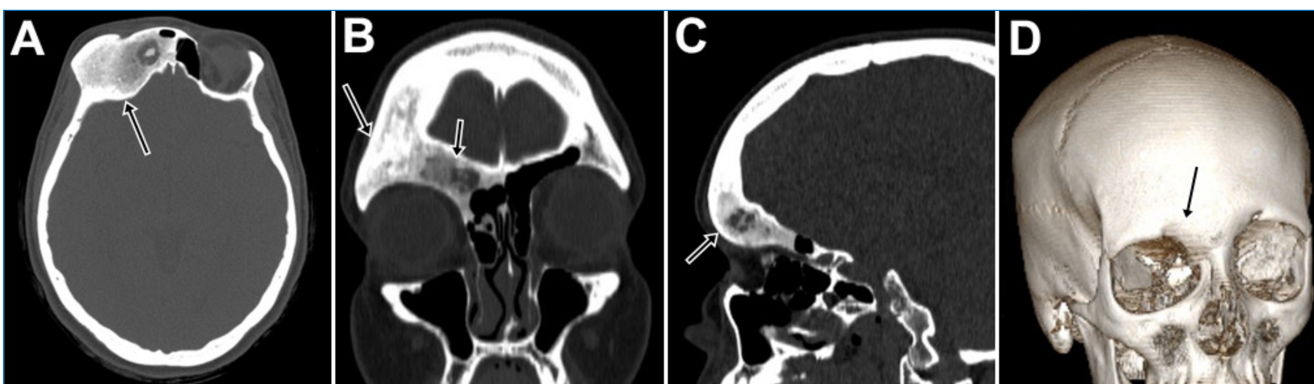

Figure 1. Preoperative computerized tomography (CT) scan images of the skull and face. (A) Axial cut; (B) Coronal cut; (C) Sagittal cut; (D) 3D reconstruction. The arrows point to the bone lesion in the right frontal bone area, causing the thickening and heterogeneity of the region's medullary bone and showing a "ground glass" appearance, extending to the orbital roof and the right frontal sinus.

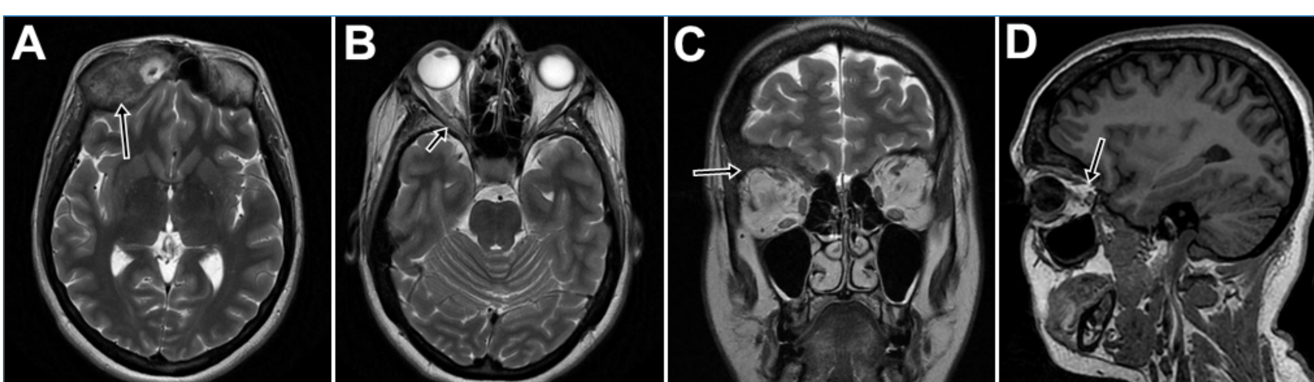

Figure 2. T2-weighted magnetic resonance imaging (MRI) images. (A) and (B) Axial; (C) Coronal; (D) Sagittal. The arrows point to the medullary bone's heterogeneity in the right frontal bone area and the affected orbital roof, presenting T2-hyperintensity and irregular structure in the medial area after contrast medium injection.

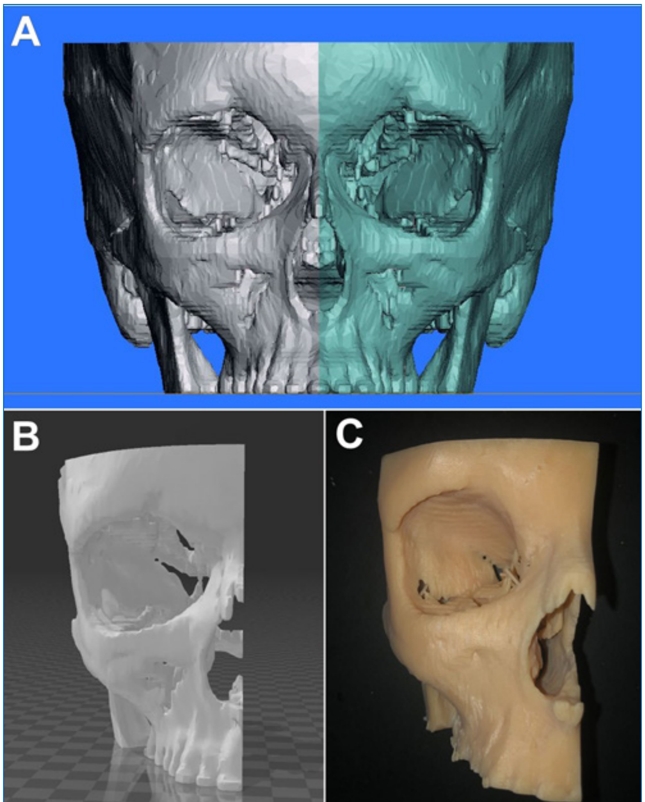

Figure 3. Virtual planning and the 3D-printed model. (A) Virtual mirroring of the non-affected side; (B) Non-affected side model mirrored on the fibrous dysplasia side; (C) 3D-printed model.

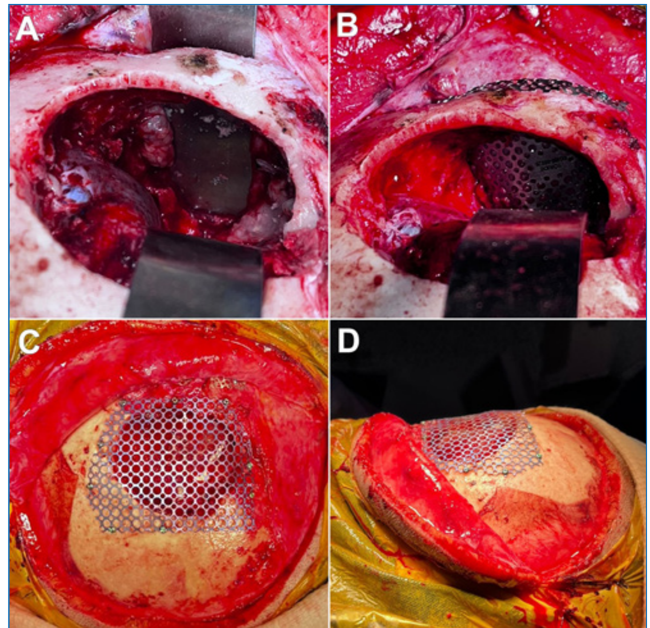

Figure 4. Surgical procedure pictures. (A) Resection of bone margin affected by fibrous dysplasia and window for the right orbital roof; (B) Titanium plate previously folded and adapted to the orbital roof for the orbital reconstruction and attached with two monocortical screws; (C) and (D) Titanium plate adapted to the bone defect created in the frontal bone area and attached with two monocortical screws.

with two monocortical screws (Figure 4C-D). After the surgical procedure under abundant 0.9% saline irrigation, we applied closed suction drainage using a flat drain from the posteroinferior area to the scalp incision. Finally, we repositioned the coronal flap using resuspension sutures of soft tissues and made a compressive bandage in the scalp and right periorbital areas.

We requested a control and immediate postoperative CT scan, with no contrast medium injection due to a medical request. In the CT scan, we observed the frontal bone craniotomy, the right orbital roof resection, and reconstruction using the titanium plate well adapted in both structures (Figure 5A-D). We also observed a discreet zone of emphysema around the titanium plate and in all the surgical sites, corresponding to the surgical procedure performed. After partial resection, we noticed a sclerotic and diffuse bone lesion with imprecise margins, but it was more located in the frontal bone area and the remaining frontoethmoidal suture. Other structures, such as brain ventricles, sulci, basal cisterns, and fissures, remained preserved.

One day after surgery, clinical examination revealed exophthalmos on the right side, with the right superior eyelid ptosis still present, reduced ocular motility on the right side, and diminished visual acuity to all directions, but with preserved vision and no signs of amaurosis.

The bone fragments removed during the surgical procedure using drills were sent for biopsy and underwent anatomopathological examination. The anatomopathological examination used the calvaria oval fragments measuring 3.6 x 3.4 x 1.0 centimeters and multiple irregular bone fragments from the right orbital roof measuring 3.0 x 2.5 x 2.0 centimeters altogether. In microscopical analysis, the histological cuts showed calvaria made of compact and mature cortical bone tissue and medullary components exhibiting hematopoietic tissue with cellular atypia. However, in the multiple bone tissue fragments removed from the orbital roof, we observed medullary parts with a storiform-pattern fibroblastic proliferation in between irregular trabeculae made of nonlamellar bone tissue and flanked by osteoblasts, with bone maturation toward its borders. The histological features, correlated to clinical and radiographic findings, were consistent with the clinical hypothesis of CFD but with no atypia. Due to a medical request, an immunohistochemistry test was performed for a more precise lesion phenotypic analysis. The histological cuts were incubated with AE1, AE3, P63, Galectin-3, and CD163 antibodies, and the reactions were analyzed by a detection technique using a secondary antibody integrated polymer and peroxidase. The immunohistochemistry results were sensitive to distortions in the histological processing, especially during the fixation step. Finally, the immunohistochemical findings associated with the morphological features corroborated the clinical hypothesis of CFD.

Both teams monitored the patient during the immediate postoperative period, and she developed no postoperative complications. After the edema and emphysema regression, besides suture and bandage removal, the patient presented

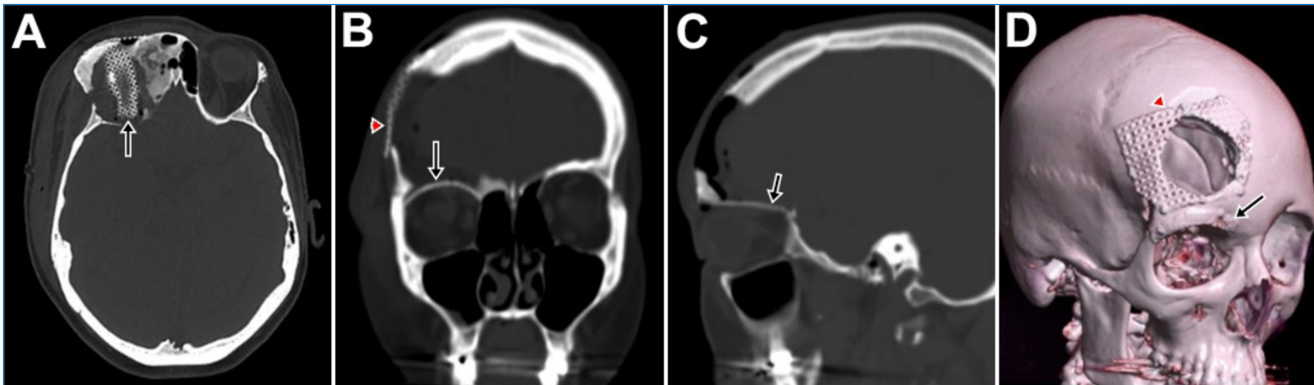

Figure 5. Immediate and postoperative CT scan images with no contrast medium injection. (A) Axial cut showing the titanium plate well-adapted in the right orbital roof and right frontal bone area; (B) Coronal cut showing the titanium plate well-adapted in the right orbital roof and right frontal bone area; (C) Sagittal cut showing the adaptation of the titanium plate in the right orbital roof with discrete zones of subcutaneous emphysema surrounding it; (D) 3D reconstruction of both well-adapted titanium plates.

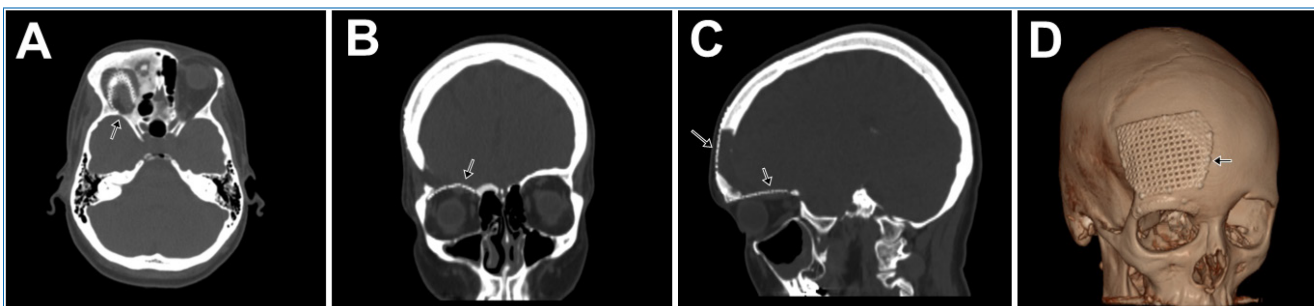

Figure 6. Two-year follow-up CT scan images with no contrast medium injection. (A) Axial cut showing the titanium plate well-adapted in the right orbital roof and the right frontal bone area; (B) Coronal cut showing the titanium plate well-adapted in the right orbital roof and right frontal bone area; (C) Sagittal cut showing the adaptation of the titanium plate in the right orbital roof; (D) 3D reconstruction of both well-adapted titanium plates.

good clinical improvement in the follow-up, with no cerebrospinal fluid fistula, preserved neural motricity surrounding resection areas, and continuous clinical and physical improvement in the postoperative period.

At the 6-month follow-up, the patient exhibited paresis of the right levator palpebrae superioris nerve due to manipulation of the region during surgery. Physiotherapy sessions were initiated following this observation, and improvement in the condition was noted during subsequent follow-up visits, with significant regression 12 months postoperatively, with good operative evolution. Additionally, the patient did not present any ocular impairments, particularly in extrinsic motility, with significant improvement observed in the 2-year follow-up after the surgical procedure (Figure 6A-D).

Discussion and Conclusions

FD is a disease that causes a significant and visible asymmetry when involving the craniofacial area. Besides aesthetic deformities, other symptoms associated with lesions next to vital structures may occur.[2] The most frequent findings of CFD around the eyes are orbital hypertelorism due to the involvement of frontal bone, sphenoid, ethmoid, and proptosis. Other features include difficulty during eyelid closure, crossed eyes, optic neuropathy, nasolacrimal duct obstruction, tearing, and trigeminal neuralgia.[4,5] Among the cited features, the patient presented eyelid ptosis, difficulty during eyelid closure, crossed eye, and altered visual acuity as a result of ocular structures compression by the CFD, confirming the most common findings of this particular disease.

The surgical approach of CFD is always a challenging choice for the surgeon. The surgical management is specific for each condition and case. However, there are three treatment options in CFD management: monitoring as a conservative treatment, pharmacotherapy using bisphosphonates or steroid therapy, and surgical treatment by conservative removal or radical resection with immediate reconstruction.[1] The treatment chosen for this patient was a mix of partial resection with conservative removal surrounding vital structures and immediate reconstruction of the bone defect, performed by both the oral and maxillofacial team and the neurosurgery team.

Reconstruction is an essential step of the surgical procedure, and the material chosen to replace the bone defect is primordial for the surgery's success. Autologous bone grafts (from calvary, ribs, iliac crest, revascularized free-flaps) or titanium plates can be used to avoid functional and aesthetic impairments.[6] Most authors suggest a conservative surgical approach, with partial resection of the dysplasia-affected sites besides bone contouring,[6] just as performed in this case, in which we preserved essential structures around the right orbit.

The use of technology is vital for successful surgical planning, both in simple and complex procedures. The CFD surgical resection can be assisted by tridimensional models, image planning, and a navigation device for better security and precision in locating the orbit.[6-8] The decompression in CFD next to the optic canal is made through neurosurgery to preserve the structures around this foramen.[6] The present case corroborates the findings that the approach through virtual surgical planning, such as the non-affected side mirroring technique, is essential for maintaining bone conformation during surgery. Additionally, the 3D-printed model is very helpful for a good adaptation of the titanium plate, increasing surgical precision and the surgery success rate while preserving important structures next to the orbit.

The CFD presents significant management challenges due to its rarity and lack of standardized treatment. Bouet et al.[9] proposed in their systematic review that radical surgery, when feasible, is the gold standard for treating CFD involving the orbital and facial bones. Optic nerve decompression should be reserved for cases with impaired visual acuity and not performed prophylactically. Regular monitoring is essential in asymptomatic, stable cases to detect any changes. Bisphosphonates are effective for pain management and complement surgical treatment. Therefore, the authors emphasize the importance of adopting a tailored treatment strategy based on individual patient needs and clinical circumstances.[9]

In conclusion, the present case report draws attention to virtual planning using mirroring and its positive outcomes. Using a 3D-printed model guarantees better visualization during surgery, and better adaptation of the fixation device reduces the chances of complications during and after surgery, directly helping the bone resection planning when necessary for FD treatment.

Conflict of interest

The authors have no conflicts of interest to declare.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed their work center protocols on access to patient data and for its publication.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

CRediT authorship contribution statement

Marcelo Santos Bahia: Data curation, Methodology, Writing – original draft, Writing – review & editing. **Marcella Yumi Kadooka:** Data curation, Formal analysis, Writing – review & editing. **Breno Nery:** Conceptualization, Data curation, Methodology, Supervision, Writing – review & editing. **Cassio Edvard Sverzut:** Conceptualization, Data curation, Methodology, Supervision, Writing – review & editing. **Alexandre Elias Trivellato:** Conceptualization, Data curation, Methodology, Supervision, Writing – review & editing.

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