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Technical Note

Virtual planning bone distraction as a golden standard in the treatment of hemifacial microsomia due to Goldenhar syndrome

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Introduction

First described by Von Arlt in 1845, the Goldenhar syndrome was not recognized as an entity until 1952, when the Belgian-American ophthalmologist described the syndrome that bears his name [1]. It has a rare incidence and multifactorial etiology with many prenatal risk factors involved, such as vasoactive drugs, gestational diabetes, twin pregnancies, second trimester bleeding, and artificial reproductive techniques [2].

The syndrome is characterized by peribulbar and/or labial dermoids, atrial appendages and atrial fistulas with a blind bottom located in the pretragus region, microtia and vertebral anomalies [3]. Due to the occurrence of several cases of hemifacial microsomia, this change was considered a distinctive feature of the entity in 1976 [4]. That means the maxillofacial surgeon is under obligation to update and recycle to work with these patients. The syndrome should be diagnosed as soon as possible so the treatment is early and patients do not suffer from physical, psychological, and social developmental delays.

Variouc techniques are used in the treatment of hemifacial microsomia, such as bone grafts, customized prosthesis, and bone distraction [5]. The first report on the use of bone distraction in the treatment of hemifacial microsomia in patients with Goldenhar Syndrome dates back to 1996 and is now, after 25 years, it is one of the treatments with considerable acceptance in the literature, although there are not many publications on the subject. Hemifacial microsomia is the second-highest incidence of congenital craniofacial malformations after cleft lip and palate [5].

The aim of the present work is to report the case of a minor patient who was diagnosed with Goldenhar Syndrome and who underwent bone distraction for the treatment of hemifacial microsomia. The entire planning and production of the distractor as well as the osteotomy and drilling template was carried out virtually.

Innovation report

An eight-year-old patient, accompanied by her parents, attended an appointment at the private clinic Dr. Antônio Targino, located in the city of Campina Grande – PB for investigation. At the first screening visit, the very enlightened parents reported that their daughter had Goldenhar Syndrome and brought tests to confirm the diagnosis. The physical examination reveals a left hemifacial microsomia, especially in the lower jaw area, as well as an appendix in the pretragus area and peribulbar dermoids. The patient was referred without undergoing hemifacial microsomia treatment. Severe malocclusion due to microsomia was observed in addition to the notched tongue on intraoral examination. The three-dimensional tomographic examination was requested for the precise assessment of the dentofacial deformity.

Upon returning with the tomographic examination, the extent of the microsomia was determined (Fig. 1), and some treatment proposals were discussed with those responsible, including bone grafting, customized prosthesis, orthognathic surgery and bone distraction. The doctor's first choice was a bone distraction, but the other options were discussed with caregivers, including orthognathic surgery and bone graft. Bone distraction was the first choice because of its lower morbidity compared to other techniques. While it is an excellent surgical technique, it is not widely due to its higher

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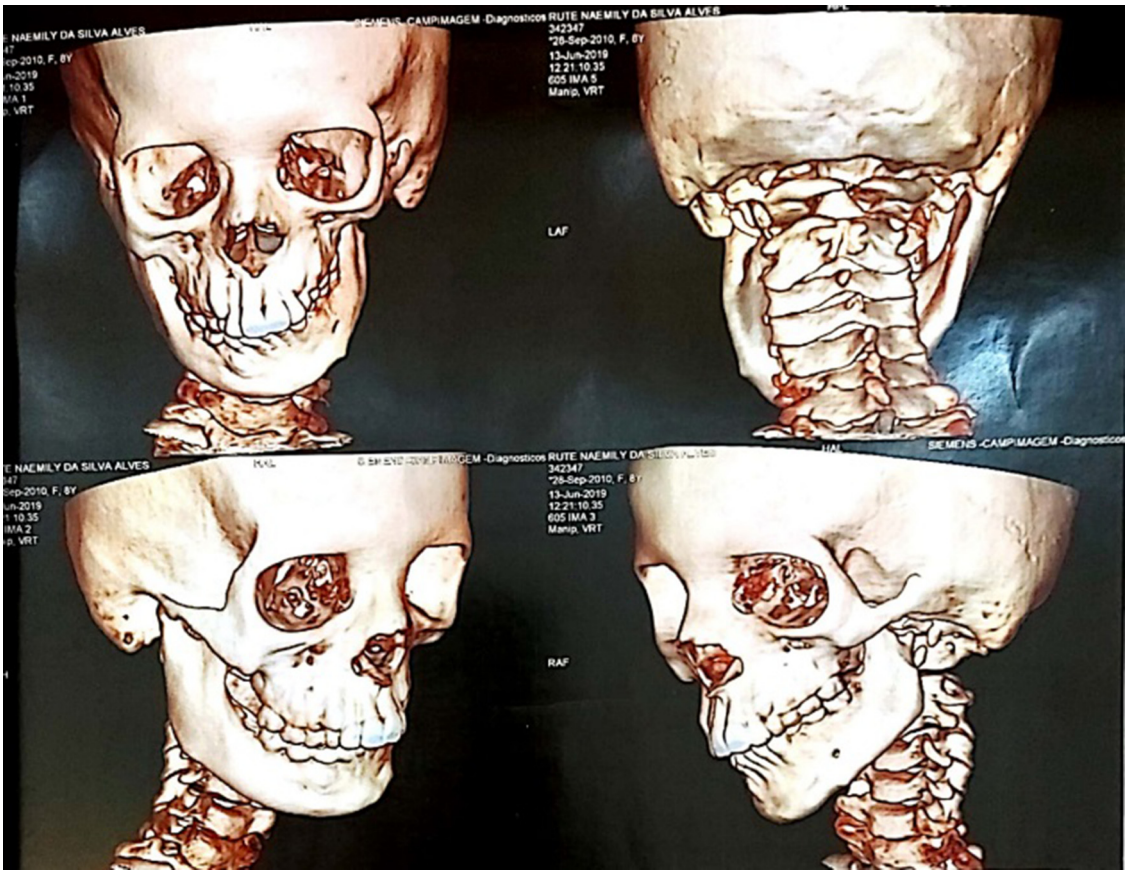

Fig. 1. Preoperative tomographic exam where the extent of hemifacial microsomia can be assessed.

cost and greater difficulty in relation to planning the distraction vectors. The caretakers, who already know the case and the treatment hypothesis, opted for bone distraction.

The digitized three-dimensional tomography was integrated into the system with the digital planning software using Dolphin 5.0® (Dolphin Imaging Systems LLC, USA). The surgeon used the digital images, to plan a tailor-made distractor and sent the image in DICOM format was sent to the company responsible for reproduction (Figs. 2 and 3). After preparing the distractor and the osteotomy and installation templates (Figs. 4 and 5), the operation was planned as the patient did not show any comorbidities. The template was used to perform osteotomy and drillings. After the system was fully positioned (Fig. 6), a light activation was performed to check that it was functioning within the planned vector.

To correct the midline deviation, a contralateral sagittal mandibular osteotomy was performed and fixed with an absorbable plate. Once the vector was confirmed, the distractor returned to the null position. A surgical toilet and suture were performed. The patient was discharged and is being monitored for the previous distraction (Fig. 7). The digital planned device is designed to achieve bone lenghtening in the vestibular and condilar directions, to achieve facial symmetry. Appropriate

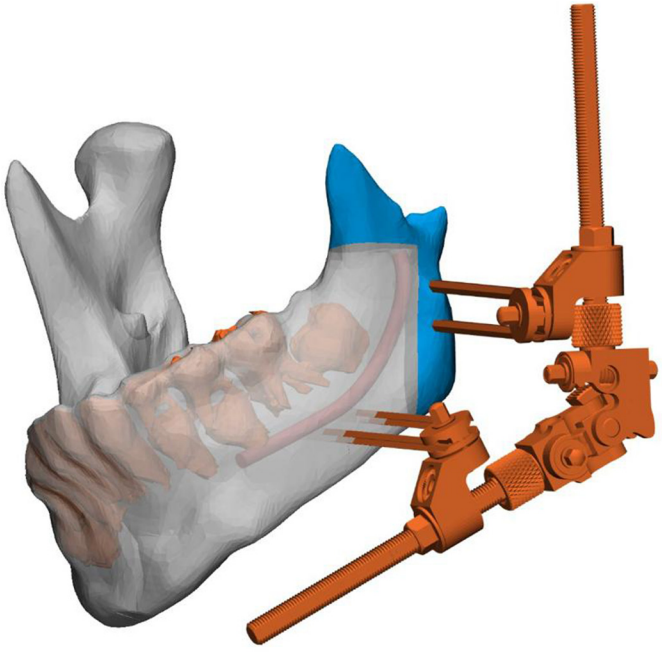

Fig. 2. Virtual distractor planning. Image taken by the Dolphin 5.0® program.

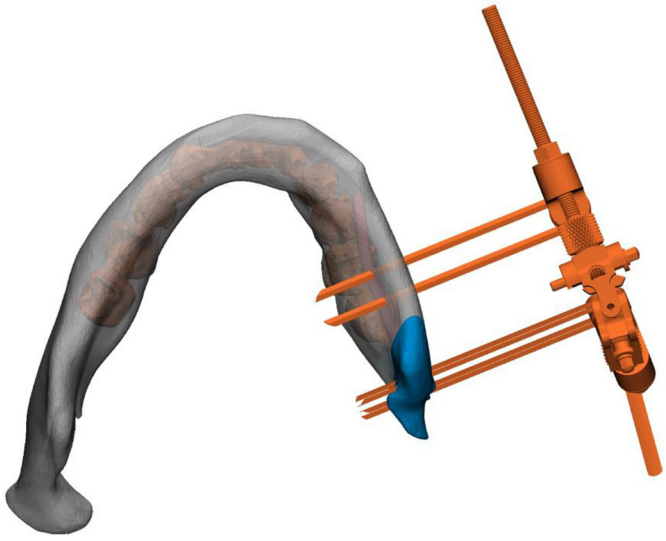

Fig. 3. Virtual planning in bottom view.

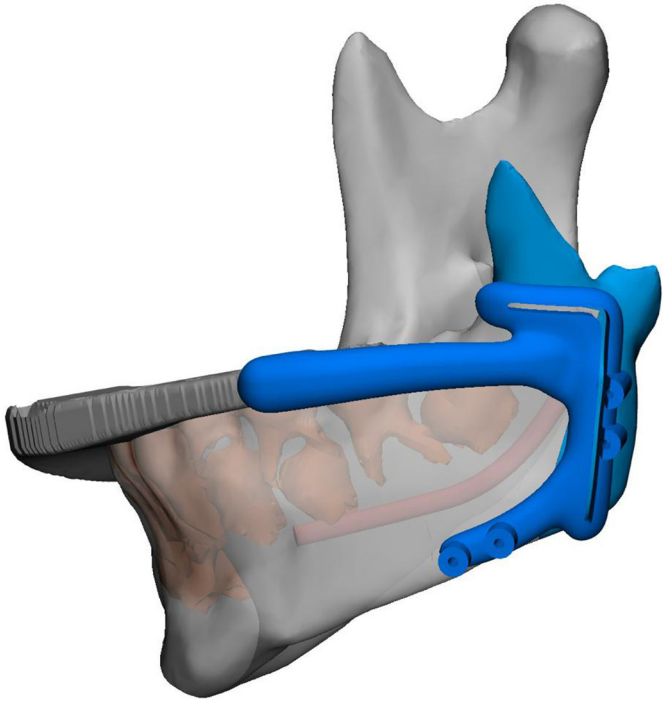

Fig. 4. Virtual planning of the osteotomy guide and installation of the distractor.

three-dimensional bone leghthening was assessed by bone distraction, a very difficul goal to be achieved with any other surgical technique. Due to the multivectorial distraction it is not possible to measure the exact linear amount or the volume of the recovered bone. It shows a significant clinical improvement in the mandibular asymmetry. Both facial aesthetics and occlusion have improved, although occlusion is still far from a reasonable standard. Treatment has yet to continue through orthodontics and other surgical procedures,

Fig. 5. Printed and sterilized virtual guides. These guides were used during the surgery.

Fig. 6. The patient still under general anesthesia, with the distractor installed.

but the bone distraction has been viewed as a successful treatment by the doctor, patient, and parents. Figure 8 shows a comparison before and after the operation, still with the distractor.

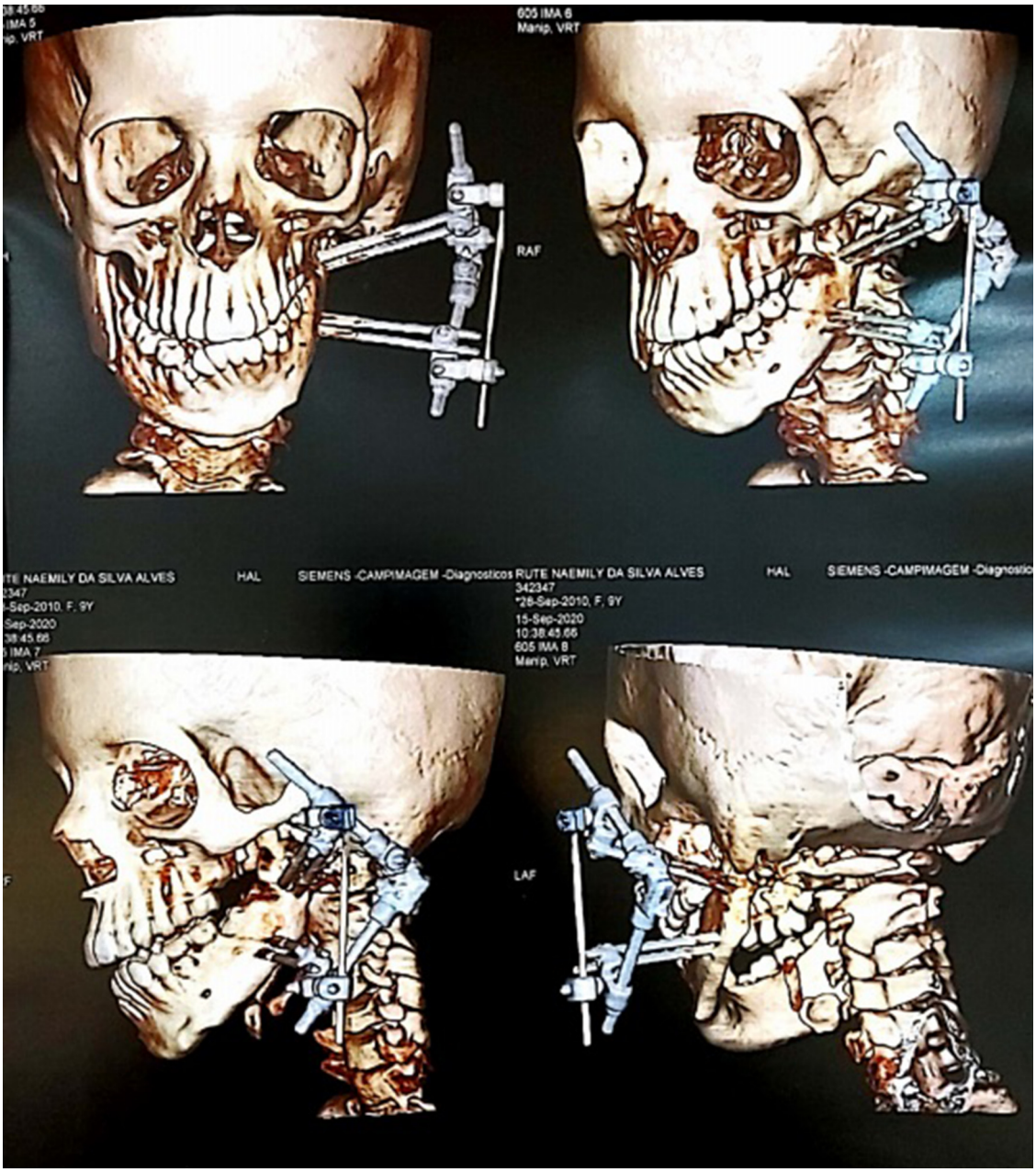

Fig. 7. Postoperative tomography. Possible to notice the distractor in position.

Discussion

One of the desirable features of a bone distractor is ease of use [6]. This detail can have a major impact on the timing of the operation, which is directly related to the postoperative phase. The easier the handling, the faster the operation, the less tissue manipulation. We will have a lighter postoperative period, not free from pain and edema, but at a level that is easier to control.

There is still no consensus in the literature, and although some argue that early interventions in the treatment of hemifacial microsomia are possible and safe, there is no evidence of early bone distraction of the mandible in patients with hemifacial microsomia [5,7]. One of the justifications for an early distraction lies in the psychological condition of the patient and that the psychological damage is reduced by early interventions even in the case of future reinterventions, in the vast majority of cases, necessary interventions [7]. It can be concluded from this that the indication is more related to physical and psychological impairment than to age itself [8]. Another justification is the compensatory growth of the upper jaw due to distraction, which sometimes avoids a second procedure, even without the use of an orthodontic appliance [9].

Fig. 8. Before (A) and after (B) bone distraction. Distractor not removed. Note the symmetry of the face. Photographs are not in the same pattern due to pandemics difficulties. Parents have sent the recent photo from home.

Bone distraction must be multivectoral in order to maintain three-dimensional symmetry and an adequate occlusal plane is obtained [6]. This multivectoral distraction can be achieved in a number of ways, but for this purpose the fabrication of a distractor through digital planning and the use of solid models are essential. The planning and digital production of distractors as well as drilling and installation templates ensure visibility, reproducibility and ease handling [6]. In cases where it is not usual to plan or install the distractors, bone resorption and migration of the distractor to other regions will be reported, creating a life-threatening situation for the patient [10]. Planning and making virtual templates and distractors is essential to obtain a suitable vector that achieves greater accuracy of 93% of the predicted movement for movement obtained in all distraction axes [11]. It is worth mentioning that in most cases bone distractors can be installed intraorally or extraorally, depending on the motion vectors and the availability of sufficient bone tissue in the area of their fixation [12].

The main advantages of digitally manufacturing distractors and templates include correct positioning of the osteotomy line, the angle between the device and the mandibular branch, and the range of motion. This has the effect that the bone regeneration is accompanied by a simultaneous expansion of the soft tissue, which contributes to the stability of the distraction and reduces the risk of recurrence [13]. The osteotomy and drilling lines are drawn according to a perfect geometric plane, which is challenging in the trans-operative phase without previously made templates and distractors [14]. The benefits of bone distraction are closely related to the precise simulation of the distractor's trajectory [14].

The greatest difficulty in virtual planning and manufacturing of distractors lies in muscle strength. Osteotomy changes the balance of muscle strength and, although the distractor has a bone fixation, some of the bone segments (usually the proximal) can be pulled towards the lateral and temporal pterygoid muscles, changing the vector of distraction. For this reason, when planning a bone distraction of the lower jaw, compensation for the medial displacement should be performed [6].

The devices customized by the surgeon makes the procedure cheaper and is available to patients who do not have sufficient income [15]. However, distractors made by digital planning provide superior results because of the complexity of the movements of the bone fragments that can be planned [6].

It is a low complication rate and safe treatment [15]. Presents few life-threatening situations to patients [10]. Patients tolerate the bone distraction process well. It has an excellent ability to increase airway permeability, achieves improvements in the range of 33%, expanding the nasopharynx and oropharynx region, bringing massive benefit to the patient's breathing. It is clear from the literature that there is no perfect treatment for hemifacial microsomias, that is one-step, effective, reliable and definitive [7].

Conclusion

Currently, bone distraction is one of the safest and most efficient therapeutic modalities in the treatment of hemifacial microsomia and a great option for patients with Goldenhar Syndrome. There is still no consensus on the best age group to undergo bone distraction patients, but early intervention has some benefits, even if there is evidence of future reintervention. Virtual planning is the gold standard in the distraction manufacture and installation, largely due to the predictability and ability of multi-vector distraction.

Authors contributions

J. Almeida Jr: Conceptualization, Visualization, Investigation, Methodology, Writing original draft, Writing- Reviewing and Editing, R. Grillo: Investigation, Methodology, Writing original draft, Writing- Reviewing and Editing, R. Teixeira: Visualization, Investigation, Methodology, Writing- Reviewing and Editing.

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Competing interests

The authors have no conflicts of interest to disclose.

Ethical procedures

All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013.

Patient consent

Written informed consent was obtained from patient guardian included in the study to use images and records for publication.

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