Orthognathic Surgery in Craniofacial Microsomia: Treatment Algorithm

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Summary: Craniofacial microsomia is a broad term that covers a variety of craniofacial malformation conditions that are caused by alterations in the derivatives of the first and second pharyngeal arches. In general terms, diverse therapeutic alternatives are proposed according to the growth stage and the severity of the alteration. When craniofacial growth has concluded, conventional orthognathic surgery (Le Fort I osteotomy, bilateral sagittal split osteotomy, and genioplasty) provides good alternatives for MI and MIIA type cases. Reconstruction of the mandibular ramus and temporomandibular joint before orthognathic surgery is the indicated treatment for cases MIIB and MIII. The goal of this article is to establish a surgical treatment algorithm for orthognathic surgery on patients with craniofacial microsomia, analyzing the points that allow the ideal treatment for each patient to be chosen. *(Plast Reconstr Surg Glob Open 2015;3:e294; doi: 10.1097/GOX.0000000000000259; Published online 20 January 2015.)*

There are multiple pathologies and causes that can affect facial symmetry. In general, they can be of the congenital type (malformations, deformations, and disruptions), acquired (traumatic), or development related.[1] Of this wide range of causes, craniosynostosis, facial fissures, hemifacial hyperplasia, hemifacial atrophy, hemimandibular hyperplasia, condylar hyperplasia, hamartomas, vascular malformations, neoplasia, and unilateral craniofacial microsomia (CFM), among many others, stand out.[1]

CFM is a broad term that covers a variety of craniofacial malformation conditions that are caused by alterations in the derivatives of the first and second pharyngeal arches. Said alteration is the product of a vascular disruption in the stapedial artery, an altered migration of cells from the neural crest to the pharyngeal arches or a combination of both.[1–4] CFM's clinical presentation reveals alterations on different levels: unilateral or bilateral orbital hypoplasia, maxillary, mandibular, outer ear, cranial nerves, and the associated soft tissue.[1] For this same reason, various terms are used to describe this situation: otomandibular dysostosis, lateral facial dysplasia, malformation syndromes in the first and second pharyngeal arches, temporal oculoauricular dysplasia,[1,4] hemifacial microsomia, and, perhaps the most appropriate, unilateral or bilateral CFM, which considers the variety of structures that are compromised.

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Table 1. Patient Distribution According to Sex, Affected Side, Type of CFM, Surgical Treatment, and Follow-up

Patient	Age (y)	Sex	Type of CFM, Side	Previous Reconstructed Mandibular Ramus	Mandibular Distraction Osteogenesis and Deferred Le Fort I	Bone Grafting and Orthognathic Surgery at the Same Time	Conventional Orthognathic Surgery	Follow-up
1	40	Female	MIIA, bilateral	No	X			5 y
2	33	Female	MIIA, bilateral	No	X			4 y
3	31	Male	MIIA, left	No	X			8 y
4	28	Male	MIII, right	No	X			9 y
5	18	Female	MIIA, right MIII, left	No	X			1 y
6	17	Female	MIII, right	No		X		8 y
7	17	Female	MIII, left	No		X		4 y
8	16	Female	MIIB, right	Yes			X	2 y
9	16	Male	MIII, right	Yes			X	2 y
10	15	Male	MIIA, left	No	X			3 y
11	15	Male	MIII, left	No		X		1 y
12	31	Male	MIII	No	X			5 y
13	28	Female	MIIA, right	No	X			6 mo
14	15	Male	MIIA, right MIII, left	No	X			6 mo
15	17	Female	MIIA	Yes			X	6 mo

The occurrence of CFM is estimated at 1 of 3600–5600 live births, and the ratio of men to women is 3:2. Bilaterals are 10% of cases[5] and unilaterals tend to be located on the right side.[1] There are multiple classification systems to determine the magnitude of the malformation. These are focused on anatomy and the functionality of the temporomandibular joint (TMJ), with the Pruzansky-Kaban classification providing a reconstructive perspective.[1] It classifies them as I, II (A and B), and III, designed to relate to

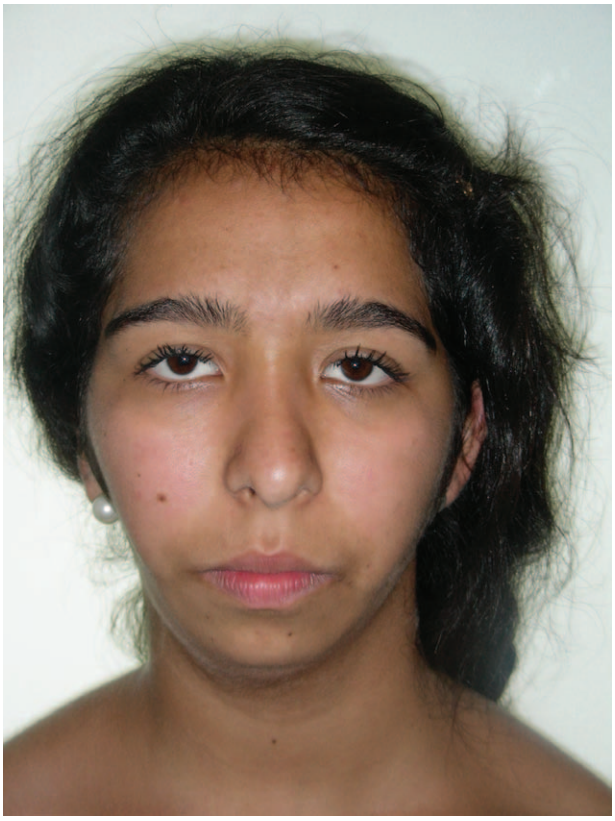

Fig. 1. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Frontal photograph: before surgery.

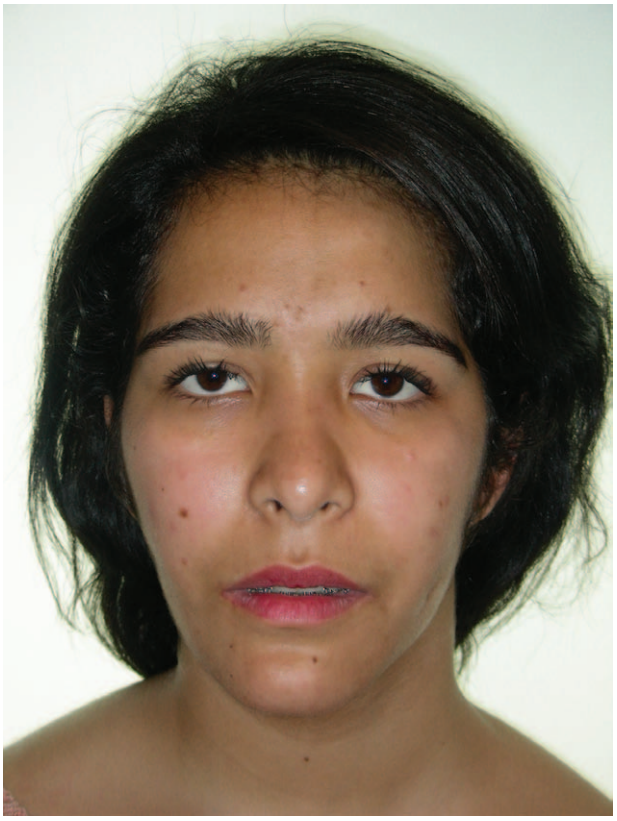

Fig. 2. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Frontal photograph: 1 year post surgery.

the type of skeletal deformity based on the mandible and the TMJ as a point of reference. David in 1987[8] proposed the SAT system, in allusion to the 3 most affected components (Skeletal malformation, Auricular involvement, Tissue defects), which was adapted from the tumor-node-methastasis of neoplasia.[1] Vento in 1991,[7] defined the OMENS system, which expanded the SAT to other affected structures: Orbital distortion, Mandibular hypoplasia, Ear deformity, Nerve defects, Soft-tissue deficiencies. The PLUS was subsequently added when a condition also affected another noncraniofacial structure.[1]

The surgical treatment is fundamentally based on skeletal correction, which in principle is undertaken after growth has finished.[8] However, certain advantages have been detected in early-stage surgical correction, such as improved growth potential by improving the functionality of the affected structures, minimizing secondary alterations from the limited growth of the adjacent structure, and improving patients' esthetic appearance and socialization.[8]

In this context, Kaban et al[8] proposes 4 treatment objectives: increasing the size of the mandible and its associated soft tissue, creating a TMJ if one is lacking, fostering vertical maxillary growth, and obtaining stable occlusion.

In general terms, diverse therapeutic alternatives are proposed according to the growth stage and the severity of the alteration. Thus, in both early temporal or mixed first phase dentition, type I, IIA, and

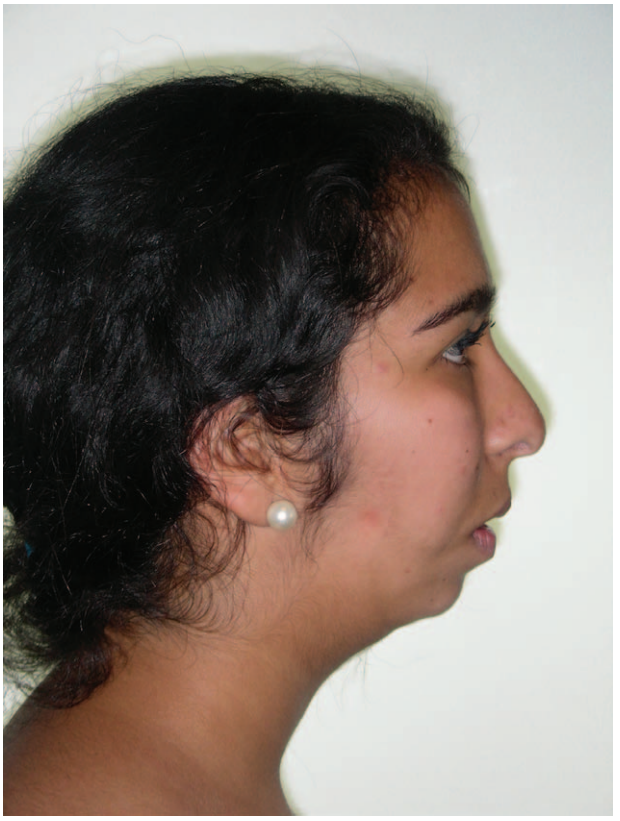

Fig. 3. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Profile facial photograph: before surgery.

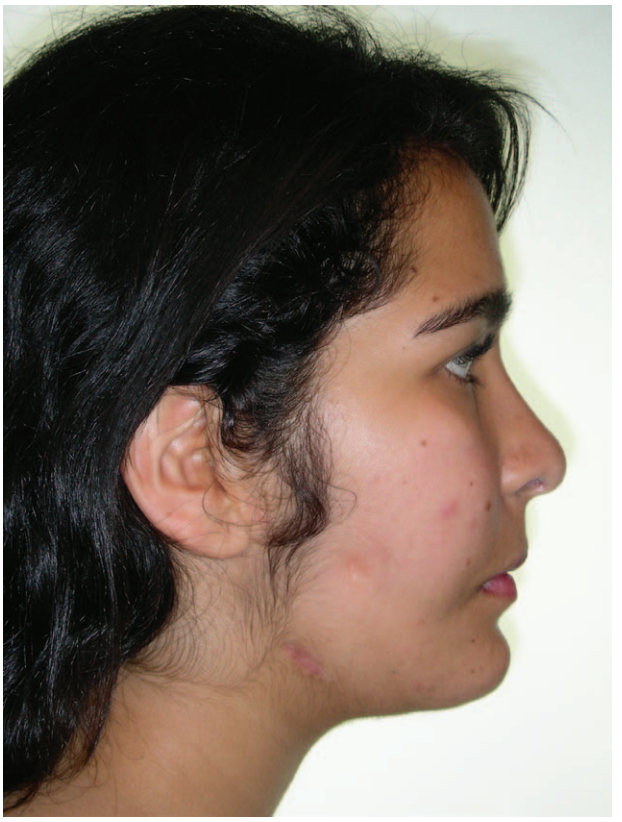

Fig. 4. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Profile facial photograph: 1 year post surgery.

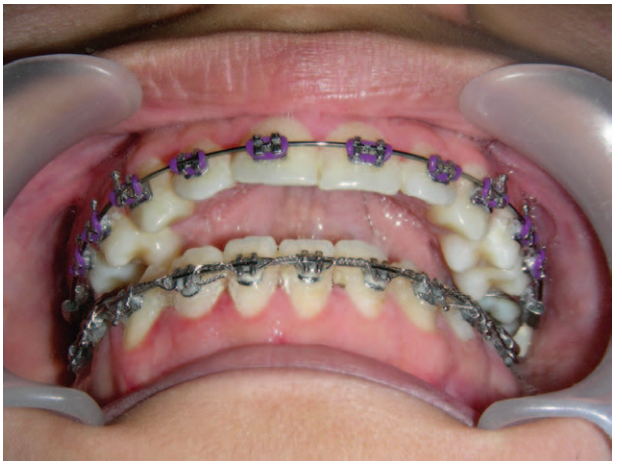

Fig. 5. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Occlusal photograph: before surgery.

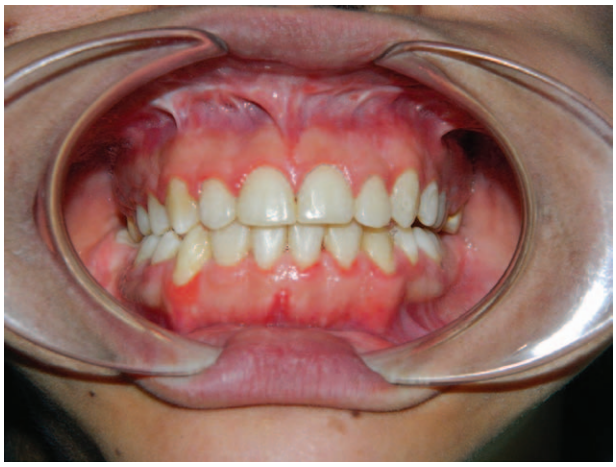

Fig. 6. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Occlusal photograph: 1 year post surgery.

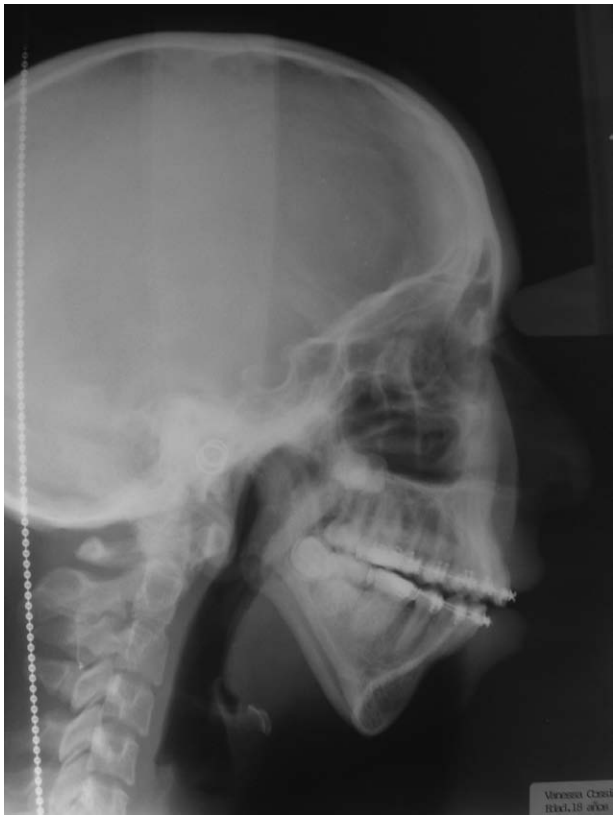

Fig. 7. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Lateral x-ray: before surgery.

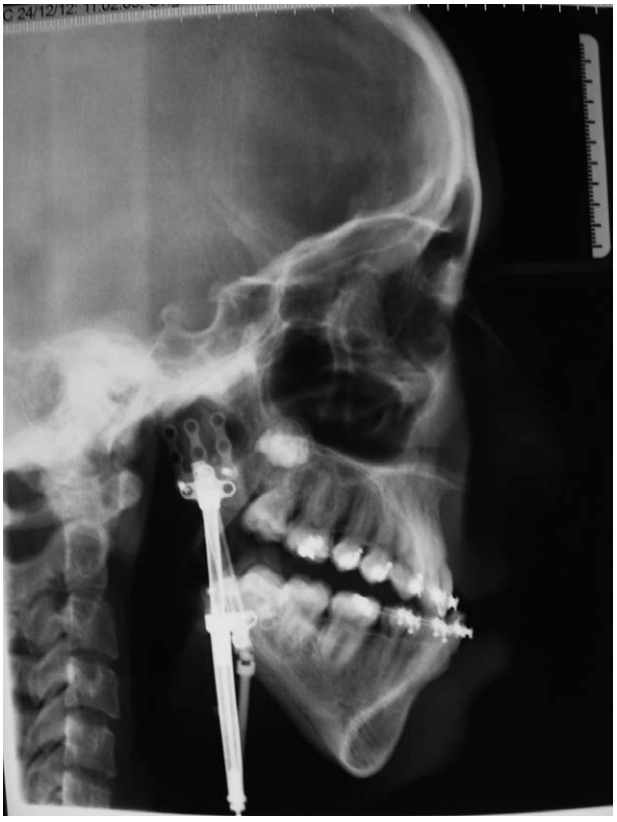

Fig. 8. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Lateral x-ray: at the end of bilateral MDO.

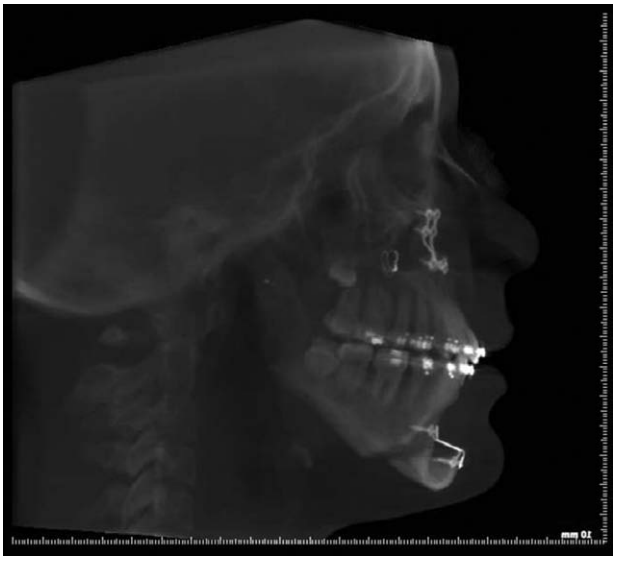

Fig. 9. Bilateral mandibular distraction osteogenesis and deferred Lefort I osteotomy, genioplasty, and rhinoplasty (patient 5). Lateral x-ray: 1 year post surgery.

IIB CFMs require orthopedic treatment to stimulate mandibular growth and to prevent maxillary dentoalveolar compensations.[1] For mixed dentition, in type I and IIA cases, early vertical mandibular elongation might be needed. Cases IIB and III require reconstruction of the TMJ with a costochondral graft or of the iliac crest.

Fig. 10. Reconstruction of right mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 6). Frontal photograph: before surgery.

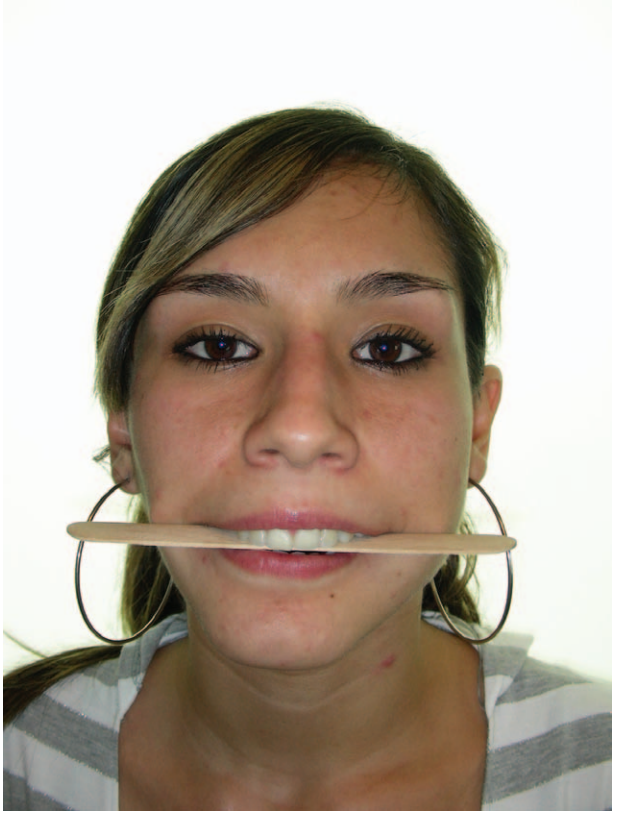

Fig. 11. Reconstruction of right mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 6). Frontal photograph: 3 years post surgery.

When craniofacial growth has concluded, conventional orthognathic surgery (Le Fort I osteotomy, bilateral sagittal split osteotomy, and genioplasty) provides good alternatives for MI and MIIA type cases. Reconstruction of the mandibular ramus and TMJ before orthognathic surgery is the indicated treatment for cases IIB and III.[1,8]

The goal of this article is to establish a surgical treatment algorithm for orthognathic surgery on patients with CFM, analyzing the points that allow the ideal treatment for each patient to be chosen.

MATERIALS AND METHODS

A treatment protocol is proposed using orthognathic surgery on patients with CFM, all from the clinical practice of Fariña (private practice and the maxillofacial service of the Hospital del Salvador, Santiago, Chile) between 2003 and 2013.

This study was approved by the Hospital del Salvador ethics board.

The study includes all patients with CFM who consulted for orthognathic surgery. The group consisted of 15 patients: 8 women and 7 men. The average age was 22.3 years. The group's characteristics are described in Table 1. Orthognathic treatment alternatives are described in Figure 1.

1. Conventional orthognathic surgery: Where after craniofacial growth and development have concluded, there is a reconstructed mandibular ramus or a slight severity (MI).
2. Mandibular distraction osteogenic (MDO) and deferred orthognathic surgery: Given the existence of a mandibular remainder that allows the MDO to be undertaken, an osteotomy of the mandibular ramus is planned (reverse L or horizontal osteotomy) to lengthen the ramus. In cases of MIIA, the vector is planned according to the treatment objectives with regard to the decanting of the occlusal plane, the leveling of mandibular angles, centering of the dental midline, and centering of the chin. In cases of MIIB and MIII, the distraction vector is planned in the direction where the new temporomandibular joint will be established. Once the main objective has been achieved (leveling of the occlusal plane), the consolidation period is awaited (habitually,

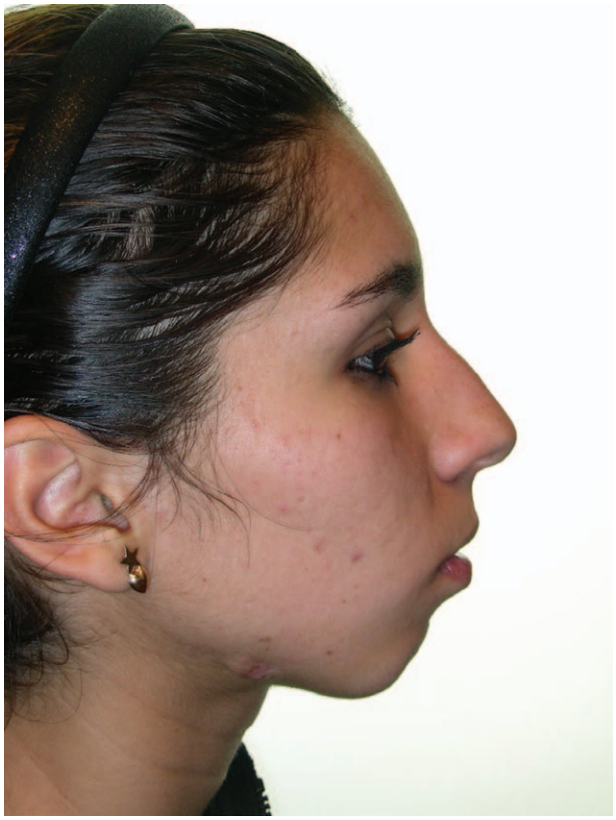

Fig. 12. Reconstruction of right mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 6). Profile facial photograph: before surgery.

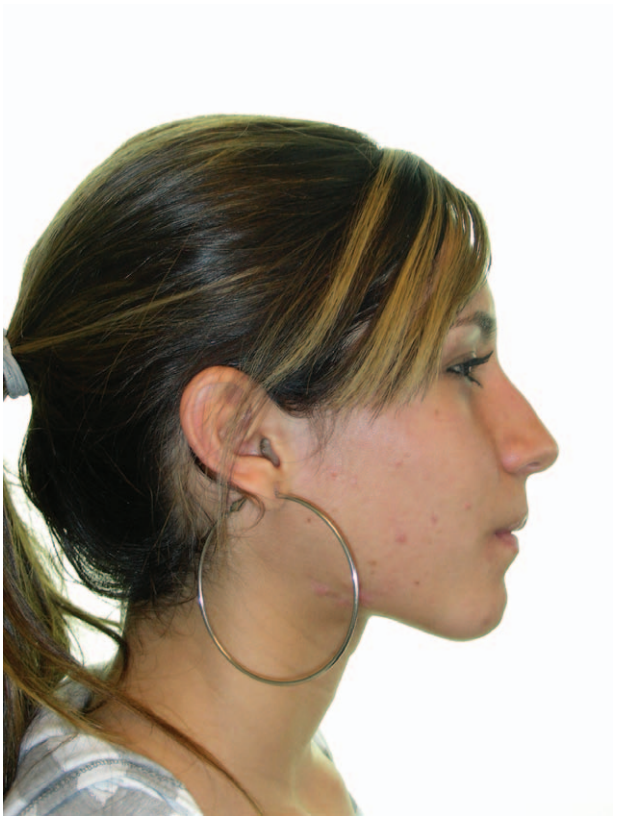

Fig. 13. Reconstruction of right mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 6). Profile facial photograph: 3 years post surgery.

an open ipsilateral bite is produced). Once this period has concluded the distraction, device is removed and a Le Fort I maxillary decanting osteotomy with mandibular self-rotation is performed at the same time. In patients with bilateral craniofacial microsomia, distraction is undertaken until the desired bilateral mandibular ramus elongation is achieved, and the Le Fort I osteotomy and mandibular self-rotation are undertaken at the same time as removing the distractor. When necessary, genioplasty and complementary surgeries are associated (fat graft and rhinoplasty) (Figs. 2–10).

3. Reconstruction of mandibular ramus and orthognathic surgery in one surgical time: If the mandibular ramus remainder is insufficient for undertaking MDO (MIIB, MIII) after craniofacial growth has concluded, orthognathic surgery is undertaken at the same time as reconstruction of the TMJ and the compromised mandibular ramus, using a free graft of the iliac crest, an articular TMJ prosthesis or a fibula free flap.[9] In these cases, planning requires double mounting of the upper maxilla to undertake orthognathic surgery, operating on the mandible first[10,11] (Figs. 11–21).

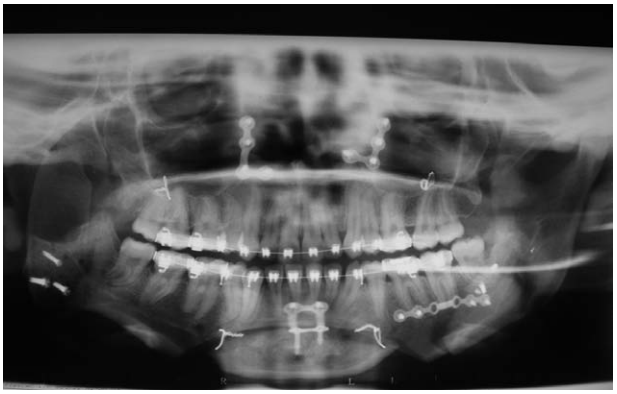

Fig. 14. Reconstruction of right mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 6). Panoramic x-rays: 3 years post surgery.

DISCUSSION

Patients with CFM have multiple facial structures showing reduced growth, which causes diverse

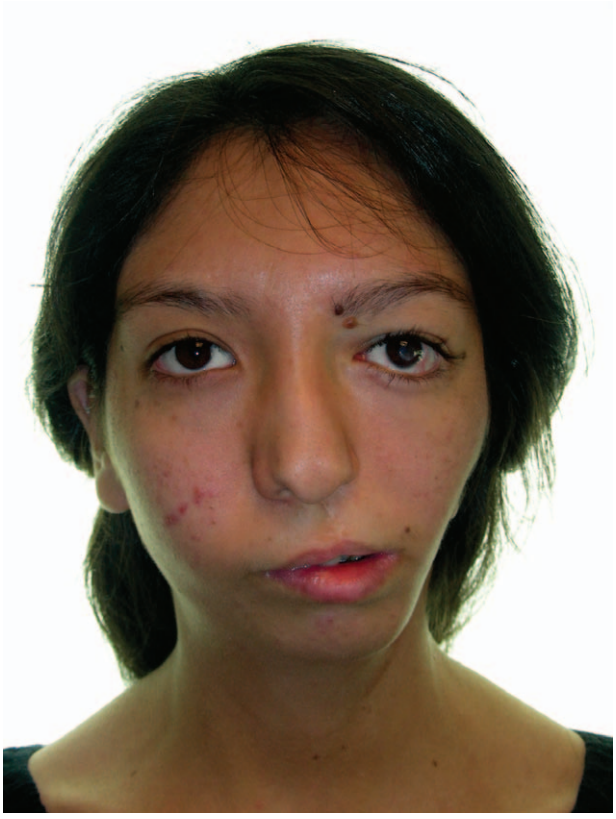

Fig. 15. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Frontal photograph: before surgery.

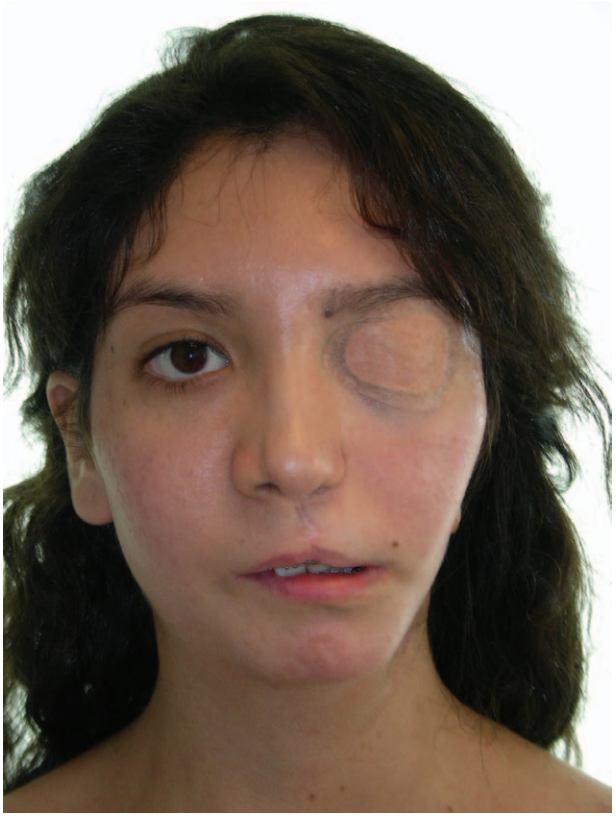

Fig. 16. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Frontal photograph: 4 years post surgery.

degrees of craniofacial asymmetry and is progressive over time.[8] They are complex patients with a 3-dimensional deformation, meaning that it is fundamental that they undergo multidisciplinary treatments according to protocols that must be based on the degree of severity, the level of skeletal maturity, age, and patients' psychosocial needs.

The literature describes a variety of therapeutic focuses, of which not all agree on the age or the types of treatment to be given. During temporal or mixed primary stage dentition, it describes using an orthopedic treatment for patients MI and MIIA to keep the mandible in a low and forward position to produce mandibular elongation and in that way prevent occlusal maxillary canting.[1,8] The TMJ in MIIB and MIII patients is not functional, which is why an orthopedic device is not used and instead the reconstruction of the mandibular ramus must be undertaken. Sándor et al[1] describe using an orthopedic device to prevent dentoalveolar compensations, but they do not undertake reconstructive surgery on the mandibular ramus at that age. Ohtani et al[12] argue that between the ages of 0 and 6, hearing alterations must be evaluated, preauricular papillomas removed, and tooth eruption monitored in MI and MII patients. Ohtani et al[12] propose mandibular reconstruction for MIII patients if there are functional alterations.

For his part, Posnick[5] argues that the most favorable esthetic results are observed in patients who are subjected to mandibular reconstruction at an age when they are closer to skeletal maturity (13–15 years old for girls; 15–16 years for boys). In addition, this is would be optimized if combined with an effective orthodontic treatment. He also argues that MI and MIIA type CFM patients with skeletal maturity are candidates for a bilateral sagittal split osteotomy with a Le Fort I osteotomy. Meanwhile, severe MIIB and MIII cases require mandibular reconstruction, which can be done via distraction of the mandibular ramus (when it is possible to achieve 3-dimensional reconstruction). In MIII cases where there is no condylar-fossa relationship, he argues that the costochondral graft is the best alternative.[5]

From another perspective, Kaban et al[8] say that to optimize growth potential, structures must be in an adequate anatomical position to improve their function, minimizing tissue deformity and distortion. At

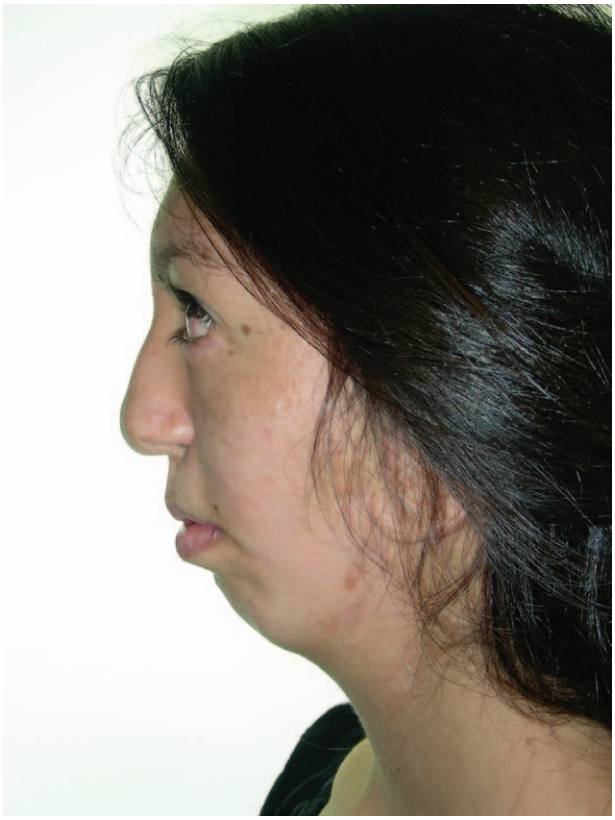

Fig. 17. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Profile facial photograph: before surgery.

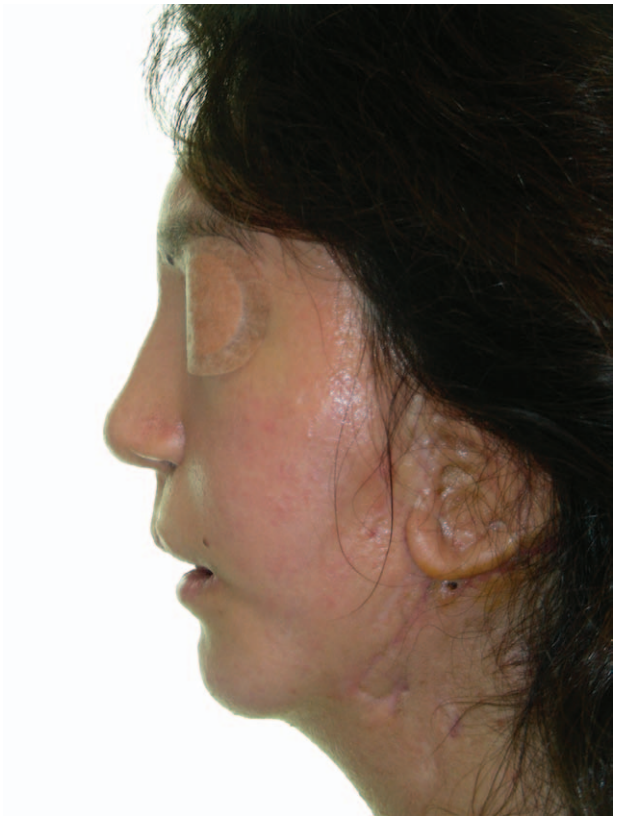

Fig. 18. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Profile facial photograph: 4 years post surgery

the same time, they improve children's appearance and psychosocial development. In this context, they argue that the progressive nature of the disease requires early surgical treatment to optimize growth on the affected side, both in the bone tissue and its adjacent soft structures.

We believe that the ideal development stage for undertaking mandibular ramus elongation (MI and MIIA) or reconstruction (MIIB and MIII) is during second-stage mixed dentition, except in cases that require early treatment due to severe respiratory problems. We argue that distraction is the first choice as long as there is susceptible bone tissue, except in severe MIII with a significant deficit in soft tissue, where we believe that the free flap is the best alternative, as other authors propose, as it contributes soft tissue for the 3-dimensional deficit.[13]

For cases of MI and MIIA, the distraction vector is planned according to the growth pattern of the contralateral mandibular ramus, but in the cases of MIIB and MIII, the first objective of the MDO should be to reach a point in the base of the cranium where the new TMJ will be created, in this way determining the distraction vector.

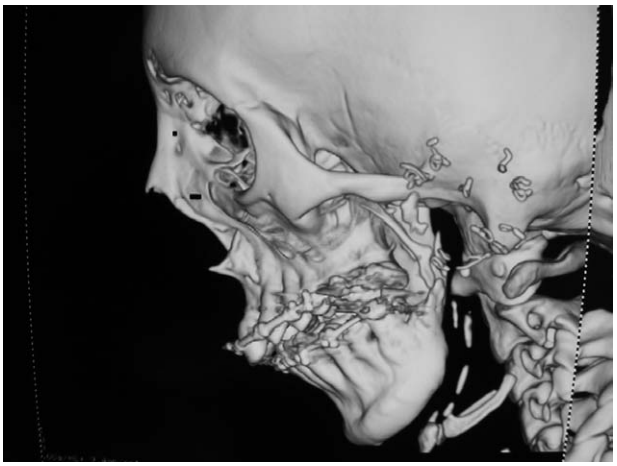

Fig. 19. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Computed tomography scan: before surgery.

CONCLUSIONS

CFM is a development and growth disorder that affects various facial structures, and the mandible becomes the fundamental tissue to base reconstruction on. Initial mandibular alteration

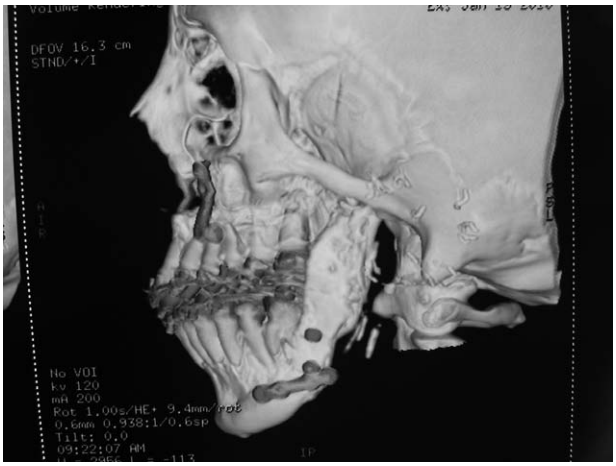

Fig. 20. Reconstruction of left mandibular ramus with iliac crest graft and orthognathic surgery at the same time (patient 7). Computed tomography scan: 4 years post surgery.

causes secondary skeletal alterations that can be avoided or reduced with early reconstruction of the mandible.

In the event of a need for orthognathic surgery, treatment objectives will be both esthetic and functional. There are multiple surgical options, but the one indicated and its justification must be based on the severity and the deficit of mandibular bone stock. In this way, in cases where the mandibular ramus has undergone early reconstruction, conventional orthognathic surgery (Le Fort I osteotomy, bilateral sagittal split osteotomy, and genioplasty) will be undertaken. In cases where there is a ramus remainder that allows mandibular distraction, MDO and deferred orthognathic surgery is a good treatment option. In cases where bone tissue is insufficient for distraction, the most appropriate alternative is orthognathic surgery and mandibular ramus reconstruction with an iliac crest graft during the same surgery.

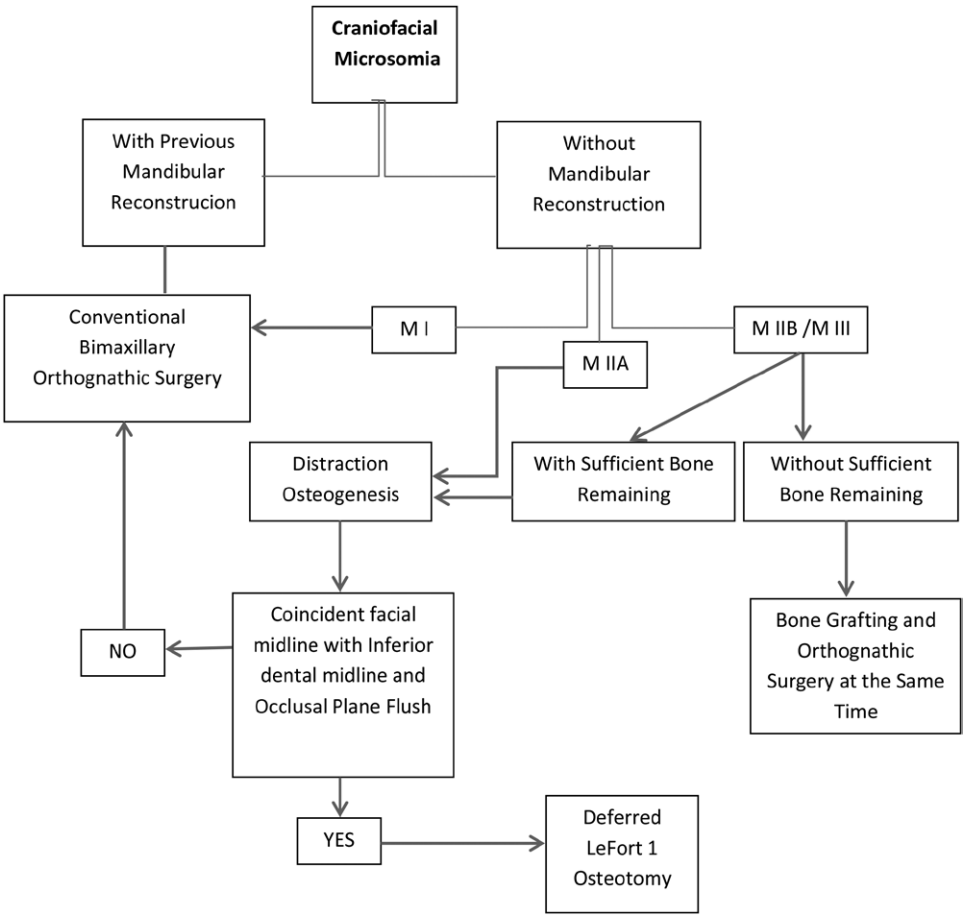

Fig. 21. Treatment algorithm in orthognathic surgery in CFM classification of mandibular severity according to Pruzansky- Kaban classification: MI, MIIA, MIIB, and MIII.

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PATIENT CONSENT

Patients provided written consent for the use of their images.

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