Orthodontic and surgical treatment of a patient with hemifacial microsomia

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This article describes the surgical and orthodontic treatment of a 12-year-old boy with a significant deformity and functional involvement caused by hemifacial microsomia. The left mandibular ramus and condyle were hypoplastic and abnormal in form and location. The lower third of the face was increased, with mandibular retrusion and significant facial asymmetry. He had difficulties in speaking and chewing and problems related to his facial appearance, which caused severe psychosocial disturbances. The patient received orthodontic treatment and temporomandibular joint reconstruction with a costochondral graft on the left side while he was still growing. Three-year follow-up records are presented. (Am J Orthod Dentofacial Orthop 2012;141:S130-9)

The treatment of dentofacial deformities is challenging to orthodontists and maxillofacial surgeons. Attaining good functional, esthetic, and stable results is even more difficult if the patient is still growing and has associated pathologies such as hemifacial microsomia.

Hemifacial microsomia is the second most common facial birth disorder after cleft lip and palate, with an incidence of 1 in 3500 to 6000 live births. The condition is bilateral in about 10% of these subjects. The cause is unknown, but the pathogenesis seems to be attributable to damage to the stapedial artery, which can cause hematoma formation in the first and second branchial arches, resulting in abnormal growth and malformation of the mandible. Another theory suggests that the death of neural crest cells can result in dysmorphology of the branchial arches that is similar to that found in hemifacial microsomia.

The most important clinical findings in hemifacial microsomia are mandibular malformation with facial asymmetry and microtia. Hypoplasia of the soft tissues, orbital involvement, nerve disorders, and other affected anatomic structures are present with a wide range of variations. Therefore, different modalities of treatment might be needed depending on the age of the patient and the severity of the problems.

The purpose of this article was to describe the treatment of a patient with a severe facial deformity due to hemifacial microsomia. Combined orthodontic treatment and orthognathic surgery with a costochondral graft were performed while the patient was still growing. Three-year follow-up records are shown.

DIAGNOSIS AND ETIOLOGY

The patient came to the Oral and Maxillofacial Surgery Department of the State University of Rio de Janeiro, Rio de Janeiro, Brazil, with a severe facial deformity with functional and esthetic involvement. He had been seeing an orthodontist since he was 8 years of age, and facial photographs and dental casts from this time were available (Figs 1 and 2). When he came to our institution, he was wearing fixed orthodontic appliances, which were placed when he was 12 years old. His chief complaints were related to his facial appearance and functional problems that were causing severe psychosocial disturbances. Clinical evaluation showed vertical maxillary excess, mandibular retrusion, and significant facial asymmetry with chin deviation to the left side. A marked occlusal cant in the frontal plane was present with tilting of the corners of the mouth. The left external ear was malformed, with a rudimentary auricle (Fig 3). The most noticeable functional problems were difficulties in speaking and chewing, lack of lip seal,
mandibular deviation during function, and no reproducible centric occlusion. Intraoral photographs showed a Class I malocclusion with a Class III tendency and a mild, lateral open bite on the right side; both dental midlines were deviated (Fig 4). Tomographic and prototyping examinations showed type II-B hemifacial microsomia with a hypoplastic left mandibular ramus and condyle that were abnormal in form and location, and malformation of the glenoid fossa (Figs 5 and 6, Table).

**TREATMENT OBJECTIVES**

Surgical and orthodontic treatment objectives were identified. Because the patient had reasonable leveling, alignment, and coordination of the arches, the main objective was to address his chief complaints and improve his self-esteem. The appliances were maintained, and surgery was performed to correct the deformity. The specific objectives of treatment were (1) asymmetry correction, (2) maxillary impaction and leveling, (3) mandibular advancement with left temporomandibular joint reconstruction, (4) overjet and overbite rectification, and (5) lip seal improvement.

**TREATMENT ALTERNATIVES**

The main alternatives to correct this deformity were (1) orthopedic and orthodontic camouflage, (2) distraction osteogenesis (3) temporomandibular joint reconstruction, and (4) orthognathic surgery.
The first alternative is a more conservative approach, indicated for growing patients with minor deformities. However, in this patient, camouflage of the skeletal problem would be limited to occlusal correction without facial esthetic improvement.

Distraction osteogenesis can be useful when extensive lengthening of the mandible is required. However, this therapy has shown a variable rate of recurrence, problems with intraoral and extraoral devices, and lack of long-term follow-up reports. For these reasons, we did not choose distraction osteogenesis for this patient.

Orthognathic surgery is indicated to obtain adequate jaw relationships and facial symmetry when the temporomandibular joint is functional. In more severe cases, temporomandibular joint reconstruction with orthognathic surgery might be necessary and can be accomplished by

Fig 3. Preoperative facial photographs.

Fig 4. Preoperative intraoral photographs.

Fig 5. Preoperative radiographic films.
placing bone grafts or temporomandibular joint prostheses. A costochondral graft is a common method used to reconstruct the ramus-condyle unit, because it has growth potential. The disadvantages of this method are additional donor-site morbidity, risk of graft resorption and infection, and possible asymmetrical mandibular growth. A temporomandibular joint prosthesis is a predictable

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Fig 6. Preoperative tomographic 3-dimensional reconstructions and prototyping models.

Fig 7. Prototyping models for the surgery.
option for temporomandibular joint reconstruction without donor-site morbidity. However, the financial costs are higher for this treatment, and it is preferably indicated in patients whose growth has been completed.

Considering the severity of the patient’s skeletal deformity, age, chief complaints, and history of psychosocial problems, we chose a treatment plan that entailed orthodontic treatment and orthognathic surgery with temporomandibular joint reconstruction and a costochondral graft.

**TREATMENT PROGRESS**

After the patient was referred to our institution, the full 0.022 × 0.028-in Roth appliance was maintained, and the surgical procedure was indicated because of his lack of motivation with treatment and his psychosocial problems. After complete leveling and alignment of the teeth, presurgical 0.019 × 0.025-in arches were placed, and the patient underwent surgery. Third molars were extracted three months before surgery.

The surgical planning for this patient included using a face-bow in a different way. Because he had an abnormal external left ear, the maxillary models were transferred to the articulator based on the interpupillary and Frankfort horizontal planes. During model surgery, 1 maxillary model was repositioned as planned at the Erickson platform. Subsequently, the mandibular model was fixed at the final occlusion point, allowing for construction of the final splint. The initial maxillary and final mandibular models were then used to obtain the intermediate splint. The traditional sequence of orthognathic surgery was not followed, and the mandible was operated on first because of the lack of a reproducible centric occlusion. The prototyping models were helpful in this phase because they allowed for a preview of the surgical procedure, the prebending of the fixation plates, and a better final position of the chin (Fig 7).

Orthognathic surgery was performed when the patient was 12 years 4 months of age. Maxillary surgery involved an anterior impaction of 2 mm, leveling by inferior reposition of 9 mm on the left side, and a 2-mm rotation to the left to correct the midline. A 4-mm counterclockwise advancement was made on the right side of the mandible with a sagittal split osteotomy. On the left side, a costochondral graft was performed to allow adequate advancement of the mandible and temporomandibular joint reconstruction (Fig 8). An 8-mm advancement, leveling, and a 3-mm lateralization genioplasty were also accomplished.

The postoperative orthodontic phase lasted 6 months and was based on the following: improvement of the right side and the intermaxillary relationship; mild, lateral open bite; and correction of the mandibular midline deviation. Class II and box elastics were used on the right side for this purpose. When the patient turned 13 years old, his braces were removed, and he was instructed to use a maxillary circumferential retainer to maintain the achieved dental results. This decision was made because of the unpredictability of growth of the costochondral graft and the patient’s lack of cooperation and oral hygiene after surgery.

**TREATMENT RESULTS**

The extraoral photographs show better facial symmetry and proportions, a straight soft-tissue profile, adequate lip seal, and a more attractive smile with residual hypoplasia of the lower lip. The mandibular asymmetry was not entirely corrected with a deficient body contour on the left side, but satisfactory facial harmony and a better social relationship were obtained (Fig 9).

The intraoral and final dental cast photographs show good intercuspation, bilateral Class I molar relationships, a 1-mm lower midline deviation to the right, a 2-mm positive overbite, and normal overjet (Figs 10 and 11). Masticatory function had improved, and a reproducible centric occlusion was obtained with incisal guidance and lateral canine discclusion.

Figure 12 shows the posttreatment radiographs, and Figure 13 is an overall superimposition based on the structural method. On the computed tomography images, the cartilage in the costochondral graft immediately after surgery and 3 years later is noticeable (Fig 14). This graft showed similar growth to the mandible, resulting in a slightly lower midline deviation to the opposite side that can be seen on the 3-year posttreatment records (Figs 15-17, Table).

**DISCUSSION**

Hemifacial microsomia is a variable and asymmetric craniofacial malformation. Of all causes of asymmetry, it is the most unpredictable and widely variable in its expression and response to growth modification.
Fig 9. Posttreatment extraoral photographs.

Fig 10. Posttreatment intraoral photographs.

Fig 11. Posttreatment dental casts.
our patient, treatment involved orthognathic surgery and a costochondral graft in the left temporomandibular joint to correct the asymmetry and correlated deformities.

Children with mild deformities might respond favorably to functional appliance therapy, and this more conservative approach should be tried before surgery, because it can improve the esthetics and the stability of the final result. This therapy is indicated in patients from 6 to 10 years old and preferably in the mixed dentition.8,11 Orthodontic treatment is focused on the control of dental eruptions and the correction of dentoalveolar adaptations to the asymmetric position of the jaws.8,12 For this reason, these approaches were not attempted because our patient had a 12-year-old permanent dentition and a significant facial deformity.

In more severe cases, several surgical procedures are indicated, including distraction osteogenesis, costochondral grafts, orthognathic surgery, and temporomandibular joint prostheses. Some of these procedures might be followed by relapse and usually require several operations.6,8,9,13-18

The main advantages of distraction osteogenesis include lack of donor-site morbidity and induction of both bone and soft-tissue generation.6,9,19 Nevertheless, distraction before skeletal maturation has shown variable recurrence of the original deformities20 and problems with extraoral devices, which are socially inconvenient and can leave hypertrophic cutaneous scars.19

The temporomandibular joint prosthesis is a predictable option for total joint replacement. However, these devices have a finite lifespan, do not adapt to facial growth, and are certainly controversial in growing patients.21

In contrast, the costochondral graft is considered the gold standard for temporomandibular joint reconstruction in growing patients and is conventionally used for reconstruction of the ramus and condyle in adults and children.21,22 It can be used on its own or combined with orthognathic surgery.22 In patients in the mixed dentition, mandibular lengthening and creation of an open bite by a costochondral graft might minimize any secondary deformities by the vertical growth potential of the midface.8,22 This does not apply after the permanent teeth have erupted, because vertical midfacial growth is essentially complete. Thus, orthodontically controlled eruption of the permanent teeth into the open-bite space will not be accompanied by vertical growth, and the teeth will simply be extruded.19 In our patient, we decided to
perform orthognathic surgery with a costochondral graft to reconstruct the left temporomandibular joint because of his age, stage of dentition, and the severity of the deformity.

Maintenance after surgery was also a challenge. The growth of the rib appears to be controlled by both intrinsic (growth centers) and extrinsic (functional matrix) factors and is unpredictable, it can be insufficient, adequate, or excessive.

Long-term follow-up is important in these cases, and occasionally additional surgical procedures are needed. The obtained occlusion at the end of treatment was
considered satisfactory with Class I molar and canine relationships. The surgery improved the appearance, body image, and socialization of this patient. Despite the unpredictability of the graft’s growth and his lack of cooperation after surgery, the 3-year postretention records showed that the results were stable (Figs 15-17).

CONCLUSIONS

Hemifacial microsomia can be treated before completion of facial growth to correct facial deformities, solve esthetic problems, and reduce functional and psychological disturbances. Orthognathic surgery and temporomandibular joint reconstruction with a costochondral graft might be successful alternatives for the treatment of this condition. This conclusion is suggested despite the low predictability of the results in growing patients and the possible need for additional surgical procedures.

REFERENCES