

HEMIFACIAL MICROSOMIA: CASE REPORT AND LITERATURE REVIEW

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SUMMARY

Hemifacial microsomia (HFM) is a sporadic congenital malformation of the craniofacial structures derived from the first and second branchial arches. The incidence of HFM has been reported to range from 1 in 3,000 to 1 in 26,000 live births, making HFM the second most common congenital malformation in the face after cleft lip and/or palate.

An 11-year-old girl came at Galeazzi Institute (Milan) in January 2017. She presented left hemifacial microsomia with absence of the left ramus of mandible and the left temporomandibular joint (tmj), part of the zygomatic arch, hypoplasia of the lateral and inferior orbital bone and of the zygomatic bone. She also presented a medial canthal dystopia. She underwent to costochondral bone graft and calvaria bone graft for reconstruction of part of the mandible and the TMJ. An emi-Le Fort I, emi-Le Fort III, and sagittal segmental osteotomy of the right mandible were performed to improve the correct occlusion.

Traditionally, the costochondral graft has been considered the gold standard for ramus-condyle reconstruction in the pediatric mandible when appropriate. Some studies cite growth unpredictability and ankylosis as concerns with rib.

Further studies examining carefully the factors predicting graft growth, such as size of cartilage cap, surgical technique, and postoperative physiotherapy, are warranted.

Key words: hemifacial microsomia, TMJ, bone graft, costochondral graft, rib, mandible.

Introduction

Hemifacial microsomia (HFM) is a sporadic congenital malformation of the craniofacial structures derived from the first and second branchial arches. The incidence of HFM has been reported to range from 1 in 3,000 to 1 in 26,000 live births, making HFM the second most common congenital malformation in the face after cleft lip and/or palate (1-15). The mandible is often the most noticeably affected and, as such, has been extensively studied. However, few studies have addressed the maxillary deformity

seen in this condition. The etiology of HFM continues to be debated, leading to the current existence of multiple competing theories. Early work in the field proposed that cases of HFM are sporadic and tend not to be inherited in a familial fashion (16). However, more recent investigation of rare familial cases has suggested a genetic basis to the disease process (17). Abnormal development and migration of neural crest cells have been associated with the craniofacial deformity in HFM as well as the commonly associated systemic manifestations including vertebral, cardiac, and limb deformities (1, 18). The most widely accepted hypothesis involves in utero in-

jury to the stapedial artery, the main blood supply to the first and second branchial arches (19, 20) resulting in hematoma formation and secondary maldevelopment of the affected area.

There is some evidence that HFM is a progressive deformity, that is, the mandibular and facial asymmetry become worse with age (21). The mandible grows on the unaffected side, but the affected side fails to keep pace, resulting in progressive mandibular asymmetry. Furthermore, the hypoplastic mandible restricts vertical growth of the midface, leading to secondary deformities of the maxilla, nose, and orbit (2). Thus, there may be orbital dystopia, canting of the occlusal plane and piriform rims, and decreased distance between the infraorbital rim and the ipsilateral piriform aperture (22).

These observations have led to the working hypothesis that surgical treatment of HFM in the growing child might provide a more normal “functional matrix,” which would result in improved growth of the mandible and reduction of secondary distortion of the midface (23). The treatment protocol includes mandibular lengthening and rotation. This results in an open bite on the affected side that provides space for vertical midfacial growth. Orthodontically controlled eruption of the maxillary teeth into the space results (24) in vertical midfacial lengthen-

ing. Finally, there is the psychosocial benefit of improved body image.

Materials and methods

A 9-year-old girl came at Galeazzi Institute (Milan) in January 2015. She presented left hemifacial microsomia with absence of the ramus of mandible and the temporomandibular joint (tmj), part of the zygomatic arch, hypoplasia of the lateral and inferior orbital bone and of the zygomatic bone. She also presented a medial canthal dystopia (Figure 1 a, b). The 3D CT scan was manipulated by use of surgical planning software. The mandible was digitally segmented and rotated transversely at the functional right joint, until the chin point was centralized and the dental midlines were coincident. This occlusal position was used to fabricate a surgical splint by use of computer-aided design/computer-aided manufacturing technique. This manipulation created a larger gap at the left ascending ramus that was filled with a digital reproduction of a costochondral graft, placed up to the neo-glenoid fossa (Figure 2).

After tracheostomy, a scalp flap was designed and graven with exposure of the orbits, the max-

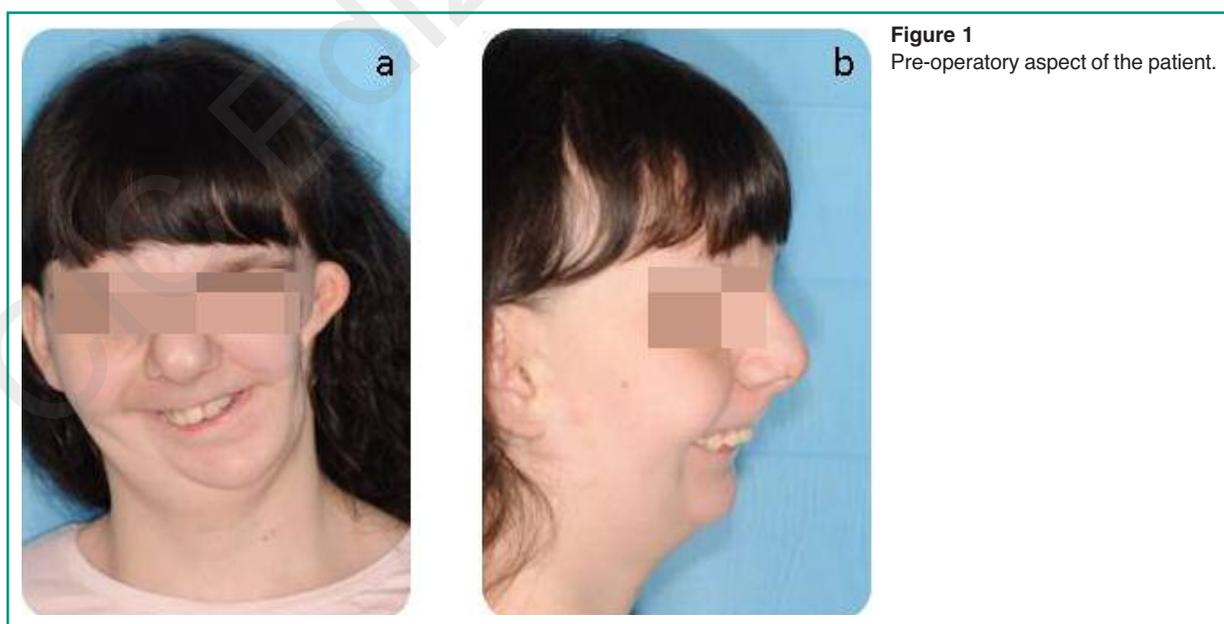


Figure 1
Pre-operative aspect of the patient.

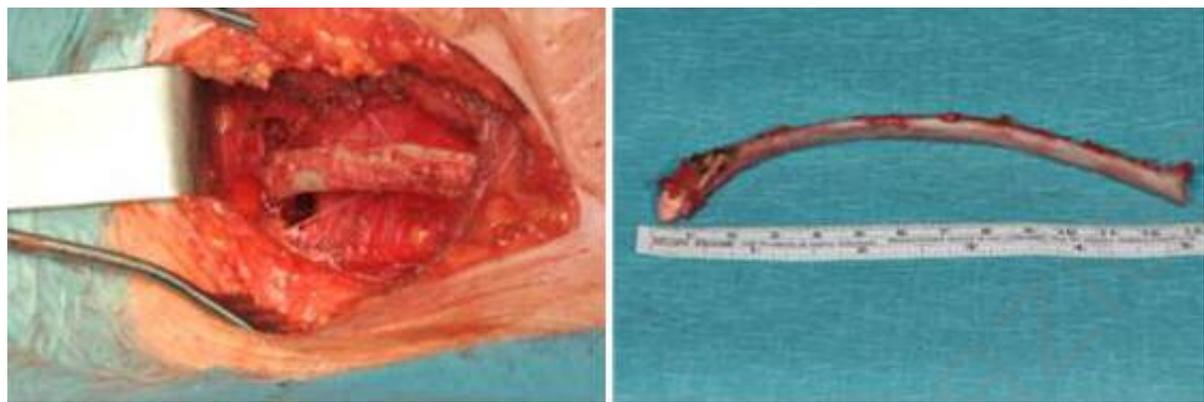


Figure 2
Costochondral graft.

illa and the region of the zygomatic bone. The inferior and lateral frame of the orbit and the zygomatic bone appeared hyperplastic and dislocated considering the contralateral. The zygomatic arch and posterior stop of the neo-TMJ was constructed from above by use of costal bone and cartilage. Fixation occurred anteriorly along the zygomatic bone and posteriorly on the temporal bone. A standard submandibular approach was performed exposing the angle and lateral face of the left mandible. Aberrant, lateral mandibular bone was osteotomized. A pocket contiguous with the neo-glenoid fossa above was dissected bluntly. The oral cavity was accessed, and the previously fabricated dental splint was placed into intermaxillary fixation (IMF) to centralize the chin. A costochondral graft was then fashioned, maintaining 1.5 mm of cartilaginous cap (for the neocondyle), and placed to fill the left ramal gap from the submandibular approach. Once visualized from above as occluding with the skull base and neoglenoid, sterilely along the temporal bone, after partial posterior reflection of the temporalis muscle.

A rib graft is simultaneously harvested from the total right sixth costal cartilage and partial of the eighth through a small transverse incision. After splitting the rectus muscle fibers, the osteocartilaginous junction is exposed (Figure 3). Using a Doyen rib dissector to strip soft tissues, dissection is carried medially and laterally to the osteocartilaginous junction. Laterally, the perios-

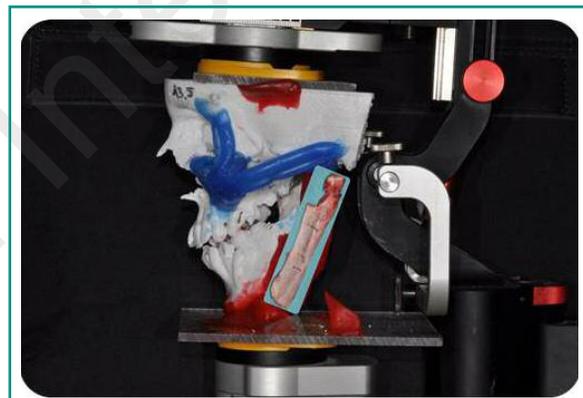


Figure 3
Preoperative planning.

teum is incised and dissected from the bone up to the osteocartilaginous junction, where the periosteum is preserved. The cartilaginous head of the rib graft is rounded off with a knife to give it the desired dimension, then also the eighth rib was carried out without the cartilaginous part. Calvaria graft was obtained (external cortical) (Figure 4). Then we performed osteotomy sec. Le Fort I on the right and sec. Le Fort III on the left. This helped to obtain rotation and climbing of the maxilla of about 4 mm on the right and, at the same time, expansion of the left orbit of 14 mm (Figure 2). Right intraoral retromolar mandibular incision was performed as well as branch exposure under the periosteum and transversal osteotomy of the same. The right



Figure 4
Patient in follow-up.

maxillomalary and nasomalar pillars were fixed with osteosynthesis plates and screws. The left frontomalar pillar was grafted with calvaria bone and then also left maxillomalary and nasomalar pillars were fixed with osteosynthesis plates and screws.

At the end, a z-shaped plastic of the medial chant of the left orbit was made. The intermaxillary block was maintained for three weeks.

Postoperative course was complicated in third day by right facial artery hemorrhage treated by ligation of the same in narcosis. After the removal of the blockade, the re-education of the visus is underway (modest diplopia almost completely regressed) and we started the orthodontic treatment.

The young girl aspect is very increased with increasing also of the function in feeding and speech. Nowadays the patient is undergoing orthodontic therapy for the realignment of the dental arches. The follow-up in still in course and maybe at the end of the growth we will perform a genioplasty and temporal and genien liposculpture.

Discussion

Hemifacial microsomia (HFM) is a sporadic congenital malformation of the craniofacial

structures and has a frequency higher than other craniofacial anomalies (25-30).

The mandible and the maxilla both share an embryologic origin in the first branchial arch, with the arch dividing to form the mandible and maxillary process.

The original classification proposed by Pruzansky focused on the size and shape of the mandible and glenoid (11). A grade I mandible demonstrates mild hypoplasia, a grade II mandible has more severe hypoplasia in addition to malformation of common bony landmarks, and a grade III mandible has complete effacement of common mandibular landmarks. Kaban et al. proposed further stratification of type II patients based on the relationship of the mandibular condyle and glenoid fossa (5) (the Kaban modification of the Pruzansky classification). This system of classification, both in isolation and as a component of the OMENS (orbital asymmetry, mandibular hypoplasia, ear deformity, nerve development, and soft-tissue disease) classification system (12) was based exclusively on clinical and plain radiographic evaluation and remains the most commonly used classification system for clinical and research purposes. Despite widespread use of the Kaban modification of the Pruzansky classification system, recent publications have questioned its reliability, citing its dependence on clinical examination and plain radiographs, when compared with the use of more advanced imaging techniques. Gillies described the first costochondral graft for temporomandibular joint reconstruction in 1920 (31) Since then, multiple autogenous grafts such as clavicle and sternoclavicular joint (23), fibula (32), iliac bone (33), and metatarsal bone (34) have been described for temporomandibular joint reconstruction, particularly in the craniofacial microsomia patient population with mandibular hypoplasia. Generally, bone grafts have several advantages and disadvantages (35-43). The sagittal and vertical mandibular deformity in HFM can also lead to glossoptosis with airway obstruction, especially in bilateral cases. Options to reconstruct the proximal mandible in a type III deformity include transport distraction, vascularized bone transfer, or bone grafting.

Transport distraction, to elongate the ramus, entails movement of the nub of the ramus through space toward the skull base (44). However, this proximal bone transport segment can be difficult to control, and enough bone stock behind the tooth-bearing mandible is a prerequisite. A vascularized osseous transfer (ie, free fibula) is also an effective means to reconstruct a severely deficient proximal HFM mandible (45). This technique is most amenable to cases of nearly total aplasia of the proximal ramus, with little or no structure proximal to the dental-bearing segment. Traditionally, the costochondral graft has been considered the gold standard for ramus-condyle reconstruction in the pediatric mandible when appropriate. Some studies cite growth unpredictability and ankylosis as concerns with rib. However, aberrant growth seems to be minimized by limiting the amount of cartilaginous component to 1 to 2 mm (46, 47).

Conclusions

The costochondral rib graft growth pattern is most likely related to both intrinsic and extrinsic processes. The growth centre theory would imply then that the size of the cartilage cap might affect graft growth. Further studies examining carefully the factors predicting graft growth, such as size of cartilage cap, surgical technique, and postoperative physiotherapy, are warranted.

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