



## Hemifacial Microsomia and Its orthodontic-Surgical Treatment. Ten Years Follow Up.

<b>Noemi LeivaV.</b>	Orthodontist. Assistant Professor, director of the department of craniofacial malformations. Faculty of Dentistry, University of Chile. Luis Calvo Mackenna Hospital.
<b>Sebastian VelizM.</b>	DDS, Academic. Faculty of Dentistry, University of Chile.
<b>Macarena Rotman S.</b>	DDS, Department of craniofacial malformations. Fellow. Faculty of Dentistry, University of Chile.
<b>Loreto Castellón Z.</b>	Maxillofacial Surgeon, Universidad Mayor. Luis Calvo Mackenna's Hospital.

**ABSTRACT**

Hemifacial Microsomia (HFM) is the second most prevalent congenital craniofacial malformation after cleft lip and palate. It is an alteration in the spectrum that mainly affects the structures derived from the first and second branchial arch, compromising structures as the jaw, orbit, external and middle ear, facial nerve and soft tissues. It presents a variable phenotypic expression, affecting from only one structure, to some cases in which severe alterations of multiple organs have been observed. Because of this, treatment depends mainly on the structures involved, the patient's age and the severity of these alterations. The aim of this paper is to present a case of unilateral hemifacial microsomia treated and monitored for 10 years, with an orthodontic - surgical focus.

**KEYWORDS**

Hemifacial microsomia, Congenital Abnormalities, costochondral graft.

**Introduction.**

Hemifacial Microsomia (HFM) corresponds to a congenital condition in spectrum that affects the development of the structures derived from the first and second branchial arch. It is the second most prevalent craniofacial malformation, after cleft lip and palate, with an estimated incidence of 1/5600 live births (LB)<sup>(1)</sup>. Being an anomaly in spectrum, it presents a great phenotypic and severity variation of the alterations that presents the committed structures<sup>(2)</sup>. Some of these structures are committed, being 5 the most affected:

1. Orbit: Mainly Orbital Dystopia (Alteration in position)<sup>(3)</sup>.
2. Jaw: The main affected structure. It can present an asymmetric development caused by hypoplasia, which ranges from a mild decrease in body size or mandibular branch to no branch condyle and coronoid fossa on the affected side<sup>(4-7)</sup>. A classification used in these patients is Pruzansky's amended by Kaban, which describes the different degrees of severity in the jaws compromised (Table No. 1).
3. Pinna (pabellon auricular): From mild microtia to anotia in correlation with facial deformity. You can also find malposition of the ear, preauricular tags of skin and cartilage and commitment of the middle and inner ear.
4. Facial Nerves: Commitment facial nerve, in a lesser extent trigeminal and hypoglossal commitment.
5. Soft Tissue: Deficiency of facial soft tissue, subcutaneous tissue, macrostomia, masticatory muscle hypoplasia, facial clefts and velopharyngeal insufficiency<sup>(8, 9)</sup>.

It is generally accepted that the ideal treatment for these patients is through multidisciplinary teams, where a variety of specialists allow assess and treat integrally each of the alterations of the patient, according to the degree of severity that are affected the different structures, taking into account the expectations of the patient and his family<sup>(10)</sup>. The aim of this paper is to present the case of a patient treated as orthodontic-surgical, with a follow up of 10 years.

**Case report.**

Female patient, 12 years old, diagnosed with right hemifacial microsomia type III (According to Kaban), which is derived to an orthodontics and maxillofacial surgery team of the Luis Calvo Mackenna Hospital (HLCM). No other morbid history. Among her surgical history, she went through a surgery for macrostomia and right costochondral graft for branch, at 6 years old which had an overgrowth and caused a severe mandibular lateral deviation. Presents an obvious facial asymmetry with further development of right side of the jaw, ocular dystopia with left eye orbit in caudal position, skeletal Class III, inverted bite and occlusal plane severely uneven (Figure 2).

**The treatment plan contemplated:**

- Initial analysis with cephalometric and dental study models.
- Orthognathic surgery; mandibular osteotomy with modified technique of rib. Orthognathic surgery consisted on Le fort I osteotomy, bilateral sagittal mandibular osteotomy of the branch and genioplasty advance. (Figure 4)
- Postsurgical orthodontic treatment (Figure 5).

**Discussion**

Hemifacial microsomia is a congenital craniofacial malformation characterized by hypoplasia of tissues derived from the first and second branchial arch. It affects 70% of the cases unilaterally, while in 30% of the cases the compromise is bilateral. There is always one side more affected than the other<sup>(1, 2)</sup>. Disproportionately it affects the right side to the left at a ratio of 3: 2; and in the same proportion it affects men and women<sup>(11)</sup>.

The event is highly variable between patients according to severity, and mainly affects the jaw, orbital's development and external ear; as well as it can submit extracranial alterations that can affect: skeletal, cardiac, renal and gastrointestinal systems.

Treatment planning depends on the type of malformation and its severity of it. Generally, transdisciplinary teams perform treatment and the main objective is to improve esthet-

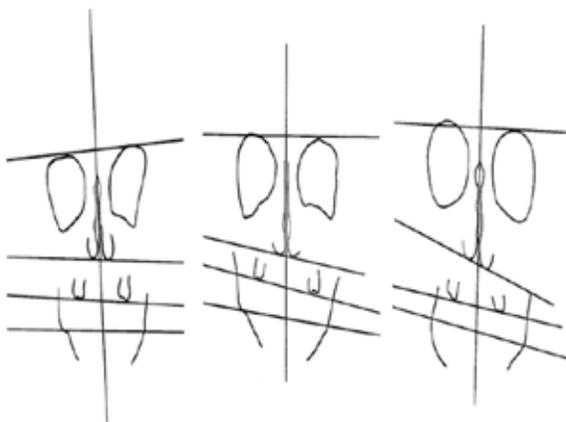
ic and functionality of facial symmetry. Through orthognathic surgery , plastic surgery and orthodontic surgery, the team seeks to increase the affected size of the jaw and its associated soft tissue, creating a functional temporomandibular joint that allows mobility, laterality and opening in cases where it is absent, establishing a functional dental occlusion, a facial's esthetic improvement and correction of secondary deformities in the jaw<sup>(12, 13)</sup>.

In patients with HCM type III, surgical treatment may require a costochondral graft to shape the hypoplastic mandible<sup>(14)</sup>. In this case, the patient developed a poorly controlled growth of the graft, which led to further accentuate facial asymmetry, being necessary to perform a second operation to compensate for changes caused by this complication of rib grafts, as is the unpredictable growth graft.

**Table No. 1**

Classification of Pruzansky amended by Kaban <sup>(15)</sup> .	
I	Jaw with normal morphology. but small
IIA	Short mandibular branch of abnormal size, glenoid in proper position and functional.
IIB	Glenoid in altered position: inferior, medial and anterior position.
III	Absence of temporomandibular Mandibular Joint (TMJ)

**Figure 1: Schematic of the variations that occur in maxillofacial bone growth.**



**Figure 2: Facial asymmetry with further development of right side of the jaw, skeletal Class III, inverted bite and occlusal plane severely uneven.**



**Figure 3: Initial lateral and panoramicradiography.**



**Figure 4: Post orthognathic surgery images; Orthognathic surgery consisted on Le fort I osteotomy, bilateral sagittal mandibular osteotomy of the branch and genioplasty advance.**



**Figure 5: Postsurgical orthodontic treatment**

## REFERENCES

1. Cohen MM, Jr., Rollnick BR, Kaye CI. Oculoauriculovertebral spectrum: an updated critique. *Cleft palate J.* 1989;26(4):276-86. | 2. Heike CL HA. Craniofacial Microsomia Overview. In: Pagon RA, Adam MP, Bird TD, et al, editors *GeneReviews™* [Internet] Seattle (WA): University of Washington, Seattle, 1993-2013 Available from: <http://www.ncbi.nlm.nih.gov/books/NBK5199/>. 2009 | 3. Gorlin RJ C MJ, Hennekam RCM. *Syndromes of the Head and Neck*, 4th edition. New York: Oxford University Press. 2001:790. | 4. Sze RW, Paladin AM, Lee S, Cunningham ML. Hemifacial microsomia in pediatric patients: asymmetric abnormal development of the first and second branchial arches. *AJR Am J Roentgenol.* 2002;178(6):1523-30. | 5. Ongkosuwito EM, van Neck JW, Wattel E, van Adrichem LN, Kuijpers-Jagtman AM. Craniofacial morphology in unilateral hemifacial microsomia. *Br J Oral Maxillofac Surg.* 2012. | 6. Ongkosuwito EM, van Vooren J, van Neck JW, Wattel E, Wolvius EB, van Adrichem LN, et al. Changes of mandibular ramal height, during growth in unilateral hemifacial microsomia patients and unaffected controls. *J Craniomaxillofac Surg.* 2013;41(2):92-7. | 7. Kitai N, Murakami S, Takashima M, Furukawa S, Kreiborg S, Takada K. Evaluation of temporomandibular joint in patients with hemifacial microsomia. *Cleft palate Craniofac J.* 2004;41(2):157-62. | 8. Fan WS, Mulliken JB, Padwa BL. An association between hemifacial microsomia and facial clefting. *J Oral Maxillofac Surg.* 2005;63(3):330-4. | 9. Funayama E, Igawa HH, Nishizawa N, Oyama A, Yamamoto Y. Velopharyngeal insufficiency in hemifacial microsomia: analysis of correlated factors. *Otolaryngol Head Neck Surg.* 2007;136(1):33-7. | 10. Association AC-P-C. *Parameters for the Evaluation and Treatment of Patients with Cleft Lip/Palate or Other Craniofacial Anomalies.* Revised Edition 2009. Original Publication *Cleft Palate Craniofac J.* 1993;30 (Suppl 1). | 11. Mielnik-Blaszczak M OK. Hemifacial Microsomia - Review of the Literature. *Dent Med Probl.* 2011;40(1):80-5. | 12. Ohtani J, Hoffman WY, Vargervik K, Oberoi S. Team management and treatment outcomes for patients with hemifacial microsomia. *Am J Orthod Dentofacial Orthop.* 2012;141(4 Suppl):S74-81. | 13. Vargervik K, Oberoi S, Hoffman WY. Team care for the patient with cleft: UCSF protocols and outcomes. *J Craniofac Surg.* 2009;20 Suppl 2:1668-71. | 14. Liu SY-C, Good P, Lee JS. Chapter 94 - Surgical Care of the Hemifacial Microsomia Patient. *Current Therapy In Oral and Maxillofacial Surgery.* Saint Louis: W.B. Saunders; 2012. p. 828-34. | 15. Kaban LB, Moses MH, Mulliken JB. Surgical correction of hemifacial microsomia in the growing child. *Plastic and reconstructive surgery.* 1988;82(1):9-19.