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## Orthodontic/Orthopedic Treatment of Hemifacial Microsomia: a review.

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# Orthodontic/Orthopedic Treatment of Hemifacial Microsomia: a review.

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## Abstract

Hemifacial microsomia is the second most frequent craniofacial birth defect after cleft lip and palate. Reported incidence varies from 1 case every 3000 to 1 case every 5600 newborns. The cause is unknown, but the pathogenesis seems to be attributable to damage to the stapedia 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 clinical presentation is variable and 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. Many classification systems have been developed. Treatment planning depends on type of malformation and severity in its expression.

## Introduction

Hemifacial microsomia is the second most frequent craniofacial birth defect after cleft lip and palate. Reported incidence varies from 1 case every 3000 to 1 case every 5600 newborns. Males and females are not equally affected: females are less frequently affected than males with an estimated ratio of 2 : 3. There is also a difference between the affected sides: a right malformation is more frequent than the left one (ratio: 3 : 2). The cause is unknown, but the pathogenesis seems to be attributable to damage to the stapedia 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 condition is believed to be sporadic, although there are documented examples of familial transmission. A causal gene was mapped to 14q32 in 1 family but not in another affected family, suggesting genetic heterogeneity.

The clinical presentation is variable and 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.Â

Many classification systems have been developed. (Illustration1,2,3,4)

## Methods

A systematic analysis of the materials has been carried out through the search engine PubMed (Mediline), with the use of the following key words "Hemifacial Microsomia treatment". 701 articles have been identified. Articles published from 2012 and 2019 were selected. Original articles, literature reviews, randomized studies, case-control studies were included. Only articles about "Orthodontic, Orthopaedic and surgical Treatment of Hemifacial Microsomia:were included.

## Results

As results, 9 articles were considered valid. Mainly through plastic/orthognathic surgery and orthodontics, the treatment seeks to improve functionality, along with optimum facial symmetry, in order to:

1. Increase the size of the affected mandibular side and its associated soft tissue.
2. Create a joint simulating the TMJ in cases where it is absent.
3. Correct secondary deformities in maxilla.
4. Achieve functional occlusion, as well as aesthetic facial and dental appearance.
5. Improve and horizontalize the occlusal plane.
6. Achieve mouth opening if it is limited.

Conventional orthodontic treatment may initially include functional appliances with the use of rigid acrylic activators, which are individualized according to each case. These devices allow for expansion of

affected tissue, taking advantage of patient's physiological growth. Sometimes they can have height planes on the healthy side, allowing for vertical compensation of the affected area, always bearing in mind that facial midline should be centered with tooth midline. This can later be complemented with conventional fixed orthodontics.

An example of functional appliance is the asymmetrical functional activator (AFA). The AFA has a hybrid design, being a combination of the two following functional appliances: biteblock components of the bionator and the vestibular shields of the Frankel appliance on the affected side. On the affected side, it is necessary to free the vertical growth of the maxilla, maintaining upper and lower teeth apart; thus, the ideal appliance is the Frankel I function regulator. This device maintains the vertical dimension by the means of the buccal shields, avoiding any occlusal contact. Allowing the passive vertical eruption of the upper teeth, the appliance corrects the occlusal plane canting. The soft tissue tension due to the buccal shields improves the stretching and lengthening of the soft tissues too. Buccal shields are supplied with a screw, which is progressively activated in order to increase the vertical dimension.

On the healthy side, an Andresen functional appliance is indicated both to avoid dental eruption and to guide the mandible in the therapeutical position improving the chin symmetry.

The acrylic and the wire elements contribute to create the correct setting for skeletal and dentoalveolar correction by forcing the mandible in the correct position in the three dimensions of the space.

Surgical management of the mandible is almost always essential also because asymmetry tends to recur after orthopaedic treatment. Some patients may later require orthognathic surgical correction of skeletal deficiencies.

## Conclusions

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. Orthodontic treatment is focused on the control of dental eruptions and the correction of dentoalveolar adaptations to the asymmetric position of the jaws.

Hemifacial microsomia is a heterogeneous, variable

disease of unique expression in each subject, both in its etiology and severity and therefore in its treatment. Being an alteration of wide spectrum, it affects various structures of the individual according to its severity. This is why a very well coordinated interdisciplinary work is vital in these patients, since they may even have psychosocial and extracranial alterations which should be timely explored and treated.

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## Illustrations

### Illustration 1

Classification system of Pruzansky (1969)

A. Classification of Pruzansky <sup>10</sup>	
Grade I	Minimum mandibular hypoplasia with all structures present.
Grade II	Condyle, ramus and sigmoid notch are present, but with serious alteration in shape and size.
Grade III	Mandibular ramus can be reduced to a small thin layer of bone, or does not exist.

### Illustration 2

Pruzansky's classification modified by Kaban et al (1988)

B. classification of Pruzansky modified by Kaban <sup>10</sup>	
I	Mandibular morphology is normal but small.
IIA	Mandibular ramus is short but of abnormal size; glenoid cavity in right position and functional.
IIB	Glenoid cavity is in an altered position, in inferior, medial, and anterior position.
III	Absence of temporomandibular joint (TMJ)

### Illustration 3

OMENS: Treatment planning depends on type of malformation and severity in its expression

<b>C. OMENS<sup>11</sup></b>	
<b>O: Asymmetry of the orbit (Orbit)</b>	
O0	Orbit with normal size and position.
O1	Abnormal orbital size.
O2	Abnormal orbital position (place a position arrow, e.g.: O2 ↑ for upper, O2 ↓ for lower).
O3	Abnormal orbital size and position.
<b>M: Mandibular hypoplasia (Mandible)</b>	
M0	Normal mandible.
M1	The mandible and glenoid fossa are small, with a short ramus.
M2A	The glenoid fossa has an anatomically acceptable position with reference to the opposite TMJ.
M2B	The TMJ is displaced in a lower, medial and anterior way, with a severely hypoplastic condyle.
M3	There is a complete absence of ramus, glenoid fossa and TMJ.

### Illustration 4

OMENS: Treatment planning depends on type of malformation and severity in its expression

<b>E: Deformity in the outer ear (Ear)</b>	
E0	Normal ear.
E1	Mild hypoplasia, but all structures are present.
E2	Absence of the external auditory canal with variable hypoplasia of the shell.
E3	Lobe poorly positioned, with absence of ear. Lobar remnant is generally moved towards a lower anterior position.
<b>N: Nerve involvement (Nerve)</b>	
N0	There is no facial nerve involvement.
N1	Upper involvement of facial nerve (temporal and zygomatic rami).
N2	Lower involvement of facial nerve (buccal, mandibular and cervical rami).
N3	All rami of facial nerve are affected. Other nerves can be involved, such as trigeminal N V (sensory), hypoglossal N XII, and the rest of cranial nerves with their own numbers.
<b>S: Deficiency in soft tissue (Soft tissue)</b>	
S0	There is no deficiency of soft tissue nor muscle deficiency.
S1	Minimal soft tissue and minimal muscle deficiency.
S2	Moderate – between both extremes, S1 and S3.
S3	Severe soft tissue deficiency due to hypoplasia of subcutaneous tissue and muscle.