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 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 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.

Many classification systems have been developed.

Methods

A systematic analysis of the materials has been carried out through the search engine PubMed (Medline), 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.

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 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 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.
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 therapeutic 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.

References

Illustrations

Illustration 1

Classification system of Pruzansky (1969)

Illustration 2

Illustration 3

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

Illustration 4

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