Changing Esthetic Paradigm in Hemifacial Microsomia

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Abstract

In hemifacial microsomia (HFM) patients the mandibular deformity is associated with canting of the occlusal plane. It is generally believed that the mandibular hypoplasia inhibits the normal downward growth of the maxilla. Treatment for hemifacial microsomia depends on the child's age and how much is the severity. Mandibular distraction osteogenesis, fibula osteocutaneous free flap for mandibular reconstruction, Composite tissue allotransplantation (CTA) is unique methods for treatment in patients with hemifacial microsomia.


Key words: Cranio-facial, Microsomia, Distraction osteogenesis, Mandibular deformity.

Definition

Hemifacial microsomia was first described by Arlt in 1881. Gorlin used the term hemifacial microsomia to patient with unilateral microtia, macrostomia and malformation of mandibular ramus and condyle whereas; the goldenhar's syndrome was described as a variant, with vertebral anomalies and epibulbar dermoids. The name, craniofacial microsomia, was proposed by Converse. Other synonyms include first arch syndrome, first and second bronchial arch syndrome, otomandibular dysostosis, oculo-auriculo-vertebral dysplasia and lateral facial dysplasia1.

All structures derived from the 1st and 2nd Branchial arches – bones, muscles, cranial nerves, organs and glands can be involved. The collected group of deformities that make up this syndrome may vary greatly in extent and degree covering a wide spectrum ranging from mild underdevelopment of the lower jaw to severe deformity of the skull and face. Characteristically this deformity, as the name implies (hemifacial), involves one side of the face, however involvement of both sides to some degree can be observed in as high as 15% of all cases. The most obvious deformity involves the lower jaw and ear, but soft tissue deficiency, maxillary hypoplasia and
even orbit and skull anomalies may also be present. 

Three percent of all newborns have significant structural anomalies. Hemifacial microsomia, or HFM, is the second most common facial anomaly, second only to cleft lip and palate. Due to a unilateral deficiency of the mandible and lower face, patients who have HFM have specific dental needs that require restorative, orthodontic and surgical correction. Whether the cause is genetic or environmental, there may be a common pathway leading to a disturbance in neural crest cell migration in HFM patients who also have a facial cleft. Transverse oral cleft ("macrostomia") is known to be associated with HFM.

**Classification**

The classification system of mandibular hypoplasia most frequently used is that of Pruzansky:

- Grade 1: mandibles are normal in configuration, but reduced in size.
- Grade 2: mandibles demonstrate hypoplasia plus maldevelopment of the associated condyle and coronoid processes.

The O.M.E.N.S. classification of hemifacial microsomia is the most accepted classification. Each letter of the acronym indicates one of the five major dysmorphic manifestations of HFM:

- **O** - Orbital asymmetry.
- **M** - Mandibular hypoplasia.
- **E** - Ear deformity.
- **N** - Nerve involvement.
- **S** - Soft tissue deficiency.

**Diagnosis**

While its precise etiology is not yet known, it is likely caused by a disruption in the development of the first two branchial arches early in embryologic development. The resulting phenotype is one of varying degrees of unilateral hypoplasia of the mandible, ear deformity, and macrostomia. Early diagnosis of hemifacial microsomia is important not only to plan for surgical reconstruction of affected features, but also to lead the physician in further evaluation for associated abnormalities or disabilities.

A team approach is necessary for the diagnosis of each patient and it must establish the extent of the anatomical deformity and the associated degree of functional impairment. A panoramic radiograph provides an excellent overview of the osseous structures of the mandible and maxillofacial complex. The occlusal radiograph is needed to depict the osseous integrity of the palatal vault. The relationship of the mandible and maxilla to the cranial base can be established initially with a lateral cephalometric radiograph. A posterior-anterior view can be used to depict the degree of osseous asymmetry of the face. Variations expressed clinically and radiographically in hemifacial microsomia preclude classifying all the abnormalities as coming from the first and second branchial arches. Anatomic structures arising from the branchial arches are directly involved but the final expression results from the combined impact of the primary anatomic defect and the secondary effects on contiguous structures.

**Dental considerations**

The primary and permanent molars in hemifacial microsomia were significantly smaller in the mesiodistal dimensions. The most posterior tooth in each arch being the most severely affected and no effect on the canines and the incisors. These findings suggest that the dental lamina in hemifacial microsomia is affected, and support the view that its pathogenesis involves an abnormality of the neural crest.

**Treatment**

From an orthodontic point of view, asymmetries can be gathered in three great clinical entities: mandibular lateral deviations, dental asymmetries without skeletal involvement, skeletal asymmetries. Vertical non-surgical asymmetry may have an obvious local origin. Frequently the occlusal slippage of a severe sagittal or a vertical malformation, which may evolve as a borderline surgery case, is suspected to be the real cause. The treatment needs peculiar strong asymmetric mechanics and, sometimes, unilateral mixed extractions. The post-treatment occlusion can be unstable; for this reason, the finishing steps must be carefully conducted.
The effect of Herbst appliance treatment on facial growth in terms of displacement of the mandible was studied by Sarnäs. During treatment facial growth was redirected and the jaws were displaced anteriorly and to the unaffected side, decreasing the degree of retrognathia and asymmetry. At the same time, however, the tilt of the mandible to the affected side was increased, possibly because of the morphologic and functional conditions of the jaws in hemifacial microsomia. The dental malocclusion was corrected partly through displacement of the jaws and partly through dentoalveolar adaptation13.

Functional appliance therapy with hemifacial microsomia promotes masticatory muscle function on the affected side, thereby stimulating bone growth in the affected condyle over and above what would occur without any treatment intervention. Removable functional appliance therapy, and later, unilateral Herbst appliance therapy, produced a dramatic change in the condylar growth on the affected side. Herbst therapy has the advantage over a removable functional appliance in that patient acceptance is much greater. Lack of patient compliance may be the primary cause of the variable results obtained with functional appliances in hemifacial microsomia cases 14.

Leonardi had treated a case of mandibular asymmetry with a modified activator appliance. The patient was given a Haupl-Andresen activator, which had been modified to reposition the right mandible downward and forward. The functional appliance therapy lasted for approximately 4 years. The affected side showed remarkable condylar growth compared with the normal side. Seven years later, the correction of the mandibular asymmetry was stable and no relapse had occurred15.

A new appliance with samarium cobalt magnets (The propellant unilateral magnetic appliance, PUMA) embedded in unilateral blocks of acrylic is a way of stimulating an autogenous costochondral graft, in hemifacial microsomia patients16.

Distraction osteogenesis is increasingly advocated in treating patients with HM as it is considered as a good alternative for the classical surgical interventions (like osteotomies and bone grafts) and its presumed positive effect on the soft tissue. From an overall dentofacial point of view, good final results were achieved with the combined orthodontic and maxillo-facial treatment of this patient with HM. Craniofacial problems like HM should be treated in craniofacial teams with enough clinical experience in treating these dentofacial malformations. This definitely will lead to more predictable and better results, fewer complications and a smaller number of surgical re-interventions17. The use of a semiburied curvilinear distraction device, with 3D treatment planning, is a potentially powerful tool to correct complex mandibular deformities18.

The latency, rate and rhythm of distraction influence the quality of the regeneration. Most maxillofacial surgeons recommend 4-7 days waiting following corticotomy before initiating distraction. In younger children, the high rate of bone metabolism favour a shorter waiting period. Waiting too long increases the risk of premature bone union. The rate and frequency of distraction are also important. If widening occurs too rapidly, a fibrous non-union results, whereas if the rate is too slow, premature bony union prevents lengthening. The ideal rhythm is a continuous form of distraction. The length of consolidation ranged from 4-6 weeks19.

Since the initial application of distraction osteogenesis to the human mandible by McCarthy, distraction osteogenesis has been used for gradual lengthening of the midface in hemifacial microsomia. Both external and internal devices are available that permit midface distraction. The background of midface distraction and the development of a Modular Internal Distraction (MID) system that permits widespread use of easily customized, buried distraction devices throughout the craniofacial region are available20.

The reconstruction of severe mandibular deformities in patients with hemifacial microsomia (HFM) is difficult. The multiple requirements include temporomandibular joint construction, mandibular ramus and body reconstructions with autogenous bone grafts, and soft tissue facial augmentation. These reconstructions include staged procedures, generally performed at the time
of skeletal maturity. Mandibular distraction osteogenesis has gained popularity as a technique for managing patients with mandibular hypoplasia. However, the use of distraction osteogenesis in HFM patients with severe grade III mandibular deformities. Mandibular distraction osteogenesis has unique advantages for these patients in that it can be performed early in childhood with minimal morbidity.

Wolff's law and the functional matrix theory of Moss established the relationship between bone morphology and the functional and soft tissue forces. A direct relationship between the mandibular deformity and the status of the muscles of mastication has been reported with HFM. The pathogenesis of HFM used as a guide to reconstructing patients, Poswillo reported a direct relationship between the success of a surgical treatment and the ability to reconstruct the components of the functional matrix. Early reconstruction of the facial skeleton is fated to failure because of the impossibility of reconstructing the muscle-periosteal component of the growth force.

The use of the fibula osteocutaneous free flap for mandibular reconstruction in severe hemifacial microsomia patients have been reported in literature. The free flap is safe and effective, and should be considered as a first choice in mandibular reconstruction in severe cases of hemifacial microsomia where distraction osteogenesis is not possible.

Composite tissue allotransplantation (CTA) in children could offer a unique reconstructive opportunity. Treatment of hemifacial microsomia can yield unsatisfactory results, even after multiple surgeries. CTA provides the advantage of intact vascularized bone that would not need to be reshaped to fit the defect, with the correct donor match. CTA also provides reconstruction with similar tissue type in regions of the central midface such as the nose, lips, and eyelids. With advances in transplant immunology to devise mechanisms to decrease immunosuppression and induce donor antigen-specific tolerance, CTA may be a future reality in the pediatric population.

**Conclusion**

Treatment of craniofacial microsomia is individualized. Principles basic to all cases include treating bony tissue deficits first, followed by soft tissue augmentation. The mandible is addressed initially since correction of mandibular malformations often stimulates maxillary growth. Maxillary growth is further enhanced with the use of maxillary activators. Costochondral grafts must be used in TMJ reconstructions. Soft tissue deficits are corrected with local and microvascular free flaps. Facial nerve defects usually are permanent and hearing must be assessed early to allow for hearing augmentation. Reconstruction of middle ear structures is often delayed until craniofacial reconstruction is complete.

**References**


