Review Article

Management of Hemifacial Microsomia: A Review

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Abstract

Hemifacial Microsomia (HM) is a congenital condition in whichthe lower face is unilaterally under-developed. Synonyms for HM include 'otomandibular dysostosis' or 'first and second branchial arch syndrome'. Hemifacial Microsomia is typically diagnosed after a comprehensive medical history and physical examination by a geneticist. There is not a blood test to diagnose the disease. Computed Tomography Scan and X-rays of the face may also be ordered for accurate diagnosis. Treatment of patients with Hemifacial Microsomia (HM) always requires an interdisciplinary approach including at least maxillofacial surgery and orthodontics. Orthodontic treatment is aimed to extrude the upper and lower molars, premolars, canines and incisors especially on the affected side, while modifying the occlusal splint accordingly. Afterwards a functional appliance with very short lingual pelottes is used in combination with elastics attached to brackets on the premolars. The lateral open bite and inclined occlusal plane thus can be corrected with appropriate orthodontic follow up. After this extrusion stage, the orthodontic removable appliance can align and level the dental arches against each other. In the early approach, either the conventional surgical procedure or the distraction techniques are possible. During the conventional surgical procedure, the deficient ramus of the mandible is partly replaced by an autologous costo-chondral bone graft. The chin should be re-positioned in the centre of the face during this procedure. Temporomandibular Joint reconstruction is the best option, as well for the growth as for psychological reasons.

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. The late procedure consists of either a classical osteotomy or a distraction with a surgical intervention. The timing for surgical procedures to correct Hemifacial Microsomia depends on the severity of the condition. Other important surgical interventions, such as the correction of the ear and soft tissues also depend on the severity of the malformation. Co-operation not only within the team, but also with the patients and their family is essential in order to achieve the best result.

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Introduction

The facial deformity caused by Hemifacial Microsomia (HM) is a congenital condition in which the lower half of the face is unilaterally under-developed and does not catch up with normal growth during childhood. The occurrence of Hemifacial Microsomia is between 1 in 3000 and 1 in 5600 births.[1] Males appear to be more frequently affected than females2 and the right side is affected more often than the left side (3 : 2).[3] Until now the cause of HM has been uncertain, although it has mainly been considered to be a developmental abnormality. It was shown in mice that, if the stapedial artery near the ear ruptures and bleeds, mice present with a condition that resembles Hemifacial Microsomia[2, 4]. As results in mice cannot simply be extrapolated to humans, there is no evidence that trauma or excessive motion of the mother might cause such a problem. Until recently it was thought that the defect was due to haemorrhage from the stapedial artery at the time, about six weeks after conception, when the maxillary artery takes over the blood supply to the affected area. More recent work suggests that, although haemorrhage at the critical time may be involved, hemifacial microsomia arises primarily from early loss of neural crest cells[5]. Neural crest cells with the longest migration path, those taking a circuitous route to the lateral and lower areas of the face, are most affected, whereas those going to the central face tend to complete their migratory movement. This explains why midline facial defects including clefts rarely are part of the syndrome. Some degree of asymmetry may be present, but both sides are affected. Neural crest cells migrating to lower regions are important in the formation of the great vessels (aorta, pulmonary artery, aortic arch), and they also are likely to be affected. For this reason defects in the great vessels (as in the tetralogy of Fallot) are common in children with hemifacial microsomia. The spectrum of deformities induced by thalidomide and isotretinoin includes conditions similar to both mandibulofacial dysostosis and hemifacial microsomia.

For unaffected parents with one child affected with HM, the chance that the second child has the same condition appears to be lower than 1%. Parents affected with Hemifacial Microsomia have approximately a 3% chance of passing the condition on to their offspring[5]. The condition seems to have a multifactorial origin and is heterogeneous in its clinical appearance.

Synonyms for HM include 'otomandibular dysostosis' or

'first and second branchial arch syndrome'. The two most frequently used classifications are the-skeletal-auricular-soft tissue (SAT)[6] and the orbital asymmetry-mandibular-hypoplasia-ear malformation-nerve dysfunction-soft tissue (OMENS) deficiency[7].

Although, 'hemifacial' refers to one half of the face, the condition is bilateral in 31% of the cases, with one side being more affected than the other[2, 8, 9]. In 48% of the cases, the condition is a part of a larger syndrome such as Goldenhar Syndrome[10]. The clinical picture of HM varies from a little asymmetry in the face to severe under-development of one facial half with orbital implications, a partially-formed ear or even a total absence of the ear. The chin and the facial midline are off-centred, and deviated to the affected side. Often, one corner of the mouth is situated higher than the other, giving rise to an oblique lip line. Other asymmetric symptoms are the unilateral hypoplastic maxillary and temporal bones, a unilateral shorter zygomatic arch and malformations of the external and internal parts of the ear. Auditory problems (conduction deafness) as a result of malformations in the middle ear and facial nerve dysfunction (temporal and zygomatic branch of the facial nerve[7] are very common in these patients: 30–50% of the patients have auditory problems[11]. Intra-oral structures can also be affected in this condition: agenesis of third molar and second premolar may be present on the affected side, as well as supernumerary teeth, enamel malformations, delay in tooth development and hypoplastic teeth[12]. The masseter, temporal and pterygoid muscles, and the muscles of facial expression are hypoplastic on the affected side. The degree of under-development of the bone is directly related to the hypoplasia of the muscle to which they are attached[13]. In most cases, there is an under-developed condyle, but aplasia of the mandibular ramus and/or condyle, with the absence of one glenoid fossa also sometimes occurs.

Management Approaches

Hemifacial Microsomia is typically diagnosed after a comprehensive medical history and physical examination by a geneticist. There is not a blood test to diagnose the disease. Because the spectrum of severity is so wide, the diagnosis should come from an experienced geneticist skilled in diagnosing craniofacial anomalies. Computed Tomography Scan and X-rays of the face may also be ordered for accurate diagnosis.

In the literature, timing of treatment of HM has often been discussed. Different opinions are presented: one group of authors prefers early surgical intervention (during growth), because they believe that the asymmetry will only increase during growth; others prefer the intervention to be delayed until after growth, because they 'see immediately what they get'.

In the early approach, either the conventional surgical procedure or the distraction technique are possible. During the conventional surgical procedure, the deficient ramus of the mandible is partly replaced by an autologous costo-chondral bone graft. A costo-chondral bone graft is preferred as it still has a growth potential that makes it comparable to the non-affected side. A costo-chondral graft provides length to the ramus, as well as a joint; it also acts as a growth centre. The chin should be re-positioned in the centre of the face during this procedure. For most children, a single operation is sufficient to correct the asymmetry.

When treating the asymmetry with a costo-chondral bone graft, the goal is to replace the distorted or even absent condyle with a new growth centre. This will only give the desired result when there is still some growth left. Therefore, it seems logical that the costo-chondral bone graft is placed before the growth spurt. Munro[14]. claim that an early surgical intervention (between 4 and 9 years old) in patients who need a Temporomandibular Joint reconstruction is the best option, as well for the growth as for psychological reasons.

The costo-chondral bone graft, however, has no growth spurt like the condyle; it grows at another rhythm (slower and irregular), independently from the healthy condyle[15]. Over-growth is often seen at the grafted side[16]. When the costo-chondral graft is growing too much and too fast, this three dimensional growth can also result in a bulk of tissue that can diminish the range of mandibular movements. Like in every surgical procedure where tissue has to be transplanted, there is always a risk of no acceptance of the graft. The reasons of failure are multiple: health of graft and grafted area, the surgical procedure, infection of the surgical wound, unpredictability of acceptance of the graft, location of surgical intervention (condyle and its area are very sensitive to surgical procedures) and the skills of the person performing this surgery.

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 as claimed by the advocators[17]. In some centers the use of the distraction technique is the early procedure of choice. One of the important contra-indications, however, is the situation in which Temporomandibular Joint reconstruction is needed. Distraction can lengthen the jaw and the ramus, but cannot create a normal growing and functioning Temporomandibular Joint. Another disadvantage is the higher risk of infection during the active and passive period of lengthening.

The late procedure consists of either a classical osteotomy (i.e. bimaxillary surgery with canting the maxilla in combination with advancement of the mandible and lengthening the ramus) or a distraction with a surgical intervention[18]. The external ear is usually reconstructed between the age of 6-8 years. This is a multiple stage process with several months between each surgery. Further surgery is the soft tissue of the cheek to increase symmetry or possibly jaw surgery, may be needed when child reaches adolescence.

The timing for surgical procedures to correct Hemifacial Microsomia depends on the severity of the condition. Other important surgical interventions, such as the correction of

the ear and soft tissues also depend on the severity of the malformation.

The Orthodontist has an important role in managing this patient for correction of dento alveolar defects[12]. Orthodontic treatment is aimed to extrude the upper and lower molars, premolars, canines and incisors especially on the affected side, while modifying the occlusal splint accordingly. Afterwards a functional appliance with very short lingual pelottes (in order to be able to insert it in a mouth with limited maximal opening) is used in combination with elastics attached to brackets on the premolars[19]. The patient is usually instructed to wear the appliance day and night except for meals. The lateral open bite and inclined occlusal plane thus can be corrected with appropriate orthodontic follow up [15, 17]. After this extrusion stage, the orthodontic removable appliance can align and level the dental arches against each other.

At the mixed dentition period, the occlusion is consolidated to a final stage by means of full fixed appliances, using several techniques. On appliance removal permanent fixed retainers are placed in upper and lower jaw, and also the functional appliance for night-time wear to stabilize the surgical correction and to prevent relapse during eventual further growth. The total orthodontic treatment may require several years even[17].

Considering the dentofacial point of view, good final results can be achieved with the combined orthodontic and maxillofacial treatment. The condyle can regain growth capacity and the temporo-mandibular joint can be functionally restored. After a long treatment by the orthodontist and the maxillofacial surgeon, facial and occlusal symmetry can be established. The occlusion appears to be stable after prolonged retention period. Craniofacial problems like Hemifacial Microsomia should be treated by craniofacial team with enough clinical experience in treating these dentofacial malformations. This definitely will lead to more predictable and better results with fewer complications.

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