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CLINICAL SECTION

Treatment of hemifacial microsomia in a growing child: the importance of co-operation between the orthodontist and the maxillofacial surgeon

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The treatment of patients with hemifacial microsomia (HM) always requires an interdisciplinary approach including at least maxillofacial surgery and orthodontics. Co-operation not only within the team, but also with the patients and their family is essential in order to achieve the best results. In the case history of the 10-½ year old female patient reported here, three surgical interventions (two with costo-chondral bone grafts) and a 3-year orthodontic treatment have taken place. A harmonious facial and occlusal result was finally reached.

Key words: Orthodontics, hemifacial microsomia

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Introduction

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 HM is between 1 in 3000 and 1 in 5600 births.[1] Males appear to be more frequently affected than females[2] and the right side is affected more often than the left side (3 times compared with 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 (a small blood vessel near the ear) ruptures and bleeds, mice present with a condition that resembles HM.[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.

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 HM 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] classification.

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

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(a) (b) (c)

Figure 1 Records taken at start of the orthodontic therapy, post-surgery (second, successful costo-chondral transplant). Mark the canted occlusal plane and the midline deviation in the lower jaw, away from the affected side

Figure 2 OPT taken before first costo-chondral transplant

Figure 3 OPT taken post-surgery first costo-chondral transplant

(a)

(b)

Figure 4 OPT and RSP taken after removal of the failed transplant

(a)

(b)

(c)

Figure 5 OPT and RSP and RS AP taken before new costo-chondral bone graft

malformations, delay in tooth development and hypoplastic teeth.[12] So far, the patients with HM seen in our hospital present with a higher prevalence of ankylosis of second and third permanent molars, although this has not been described in the literature so far. 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. In these cases, the maxilla is hypoplastic at the affected side.[3]

There are essentially two approaches: either an early (during growth) or a late (after the active growth period) surgical intervention. 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. The problem with some grafts, however, is that they show over-growth.

In some centres the use of the distraction technique is the early procedure of choice. This can increase the


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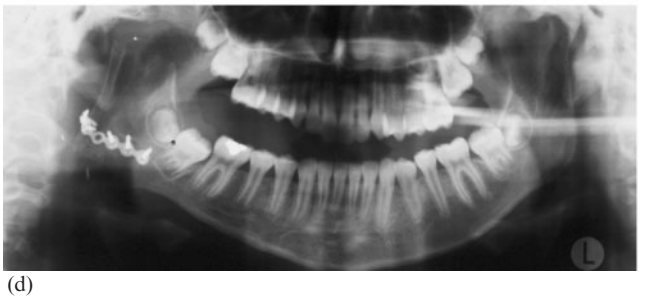
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Figure 6 (a) Post-surgery intermaxillar retainers in place. (b,c) Four weeks post-surgery, inter-maxillary fixation with splint. (d–f) Two months after the second costo-chondral transplant; OPT and RS AP


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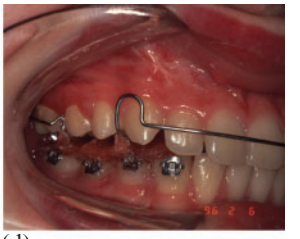
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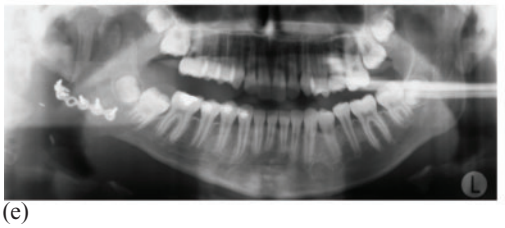
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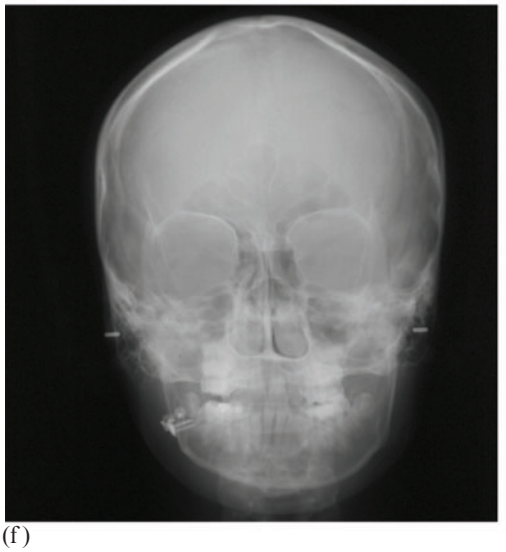
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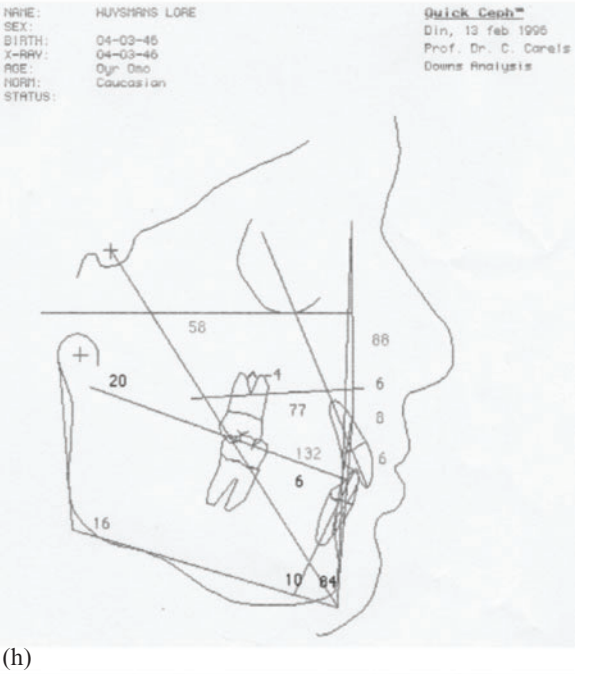

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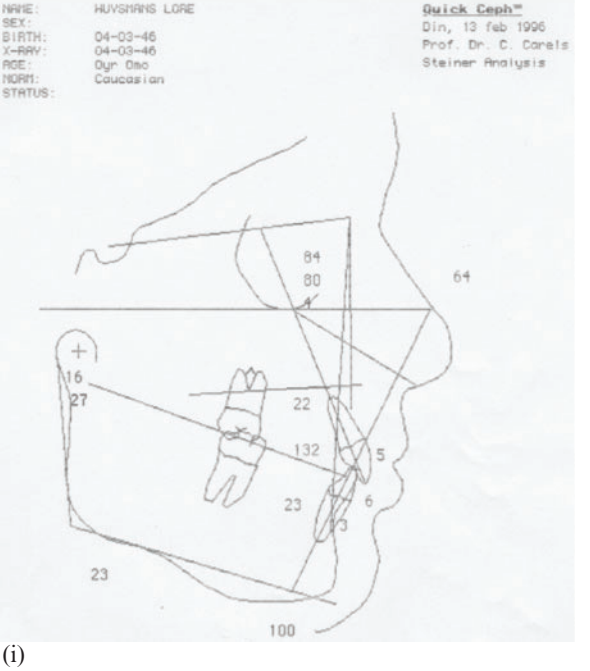


(i)

Figure 7 (a–d) Four months post-surgery with the functional appliance in place. The functional appliance is used as a splint, which is gradually being adjusted. Comparing Figure 2e with Figure 2f shows the extrusion of the lower teeth reached during 4 months of wearing the appliance in combination with intra-maxillary elastics. (e–i) OPT, RSP, RS AP and Downs and Steiner analysis taken before placing a few brackets on the upper premolars and canine at the affected side

(j)

Figure 7 (j) Wearing a removable plate in the upper jaw, which is gradually adjusted to allow extrusion of both the upper and lower teeth in combination with some brackets and inter-maxillary elastics

number of surgical interventions, as later a second operation is often needed to recorrect insufficient growth.

The late procedure consists of either a classical osteotomy (i.e. bimax surgery with canting the maxilla in combination with advancement of the mandible and lengthening the ramus) or a distraction with a surgical intervention.

The timing for surgical procedures to correct HM 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.

Case report

A 10½ year old Caucasian female patient with HM was referred to the University Hospital, Leuven, by her general physician for the asymmetric lower half of her face. She showed a remarkable facial hypoplasia on the right side with the chin deviating to the right also, resulting in a severe asymmetric occlusion (Figure 1a–c). Radiological investigation revealed the presence of a deformed joint at the right side, caused by fibrous ankylosis (Figure 2). The diagnosis of HM was accepted after differential diagnosis with idiopathic juvenile arthritis (medical history and blood investigations). A neonatal trauma had also been excluded as possible cause because there was no known history of trauma in childhood.

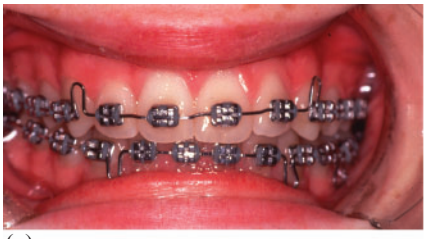
Her treatment history at the University Hospital, Leuven started with a costo-chondral bone graft taken from the fifth left rib that was placed via a pre-auricular approach (02/1994). A wafer was inserted between the dental arches in order to create an asymmetric open bite. Inter-maxillary wire fixation (IMF) was used to secure a stable position (Figure 3). It was removed after a fixation period of 6 weeks. Radiological evaluation revealed that the graft had, however, not maintained its position and had moved to a more antero-medial position. The materials used for osteosynthesis, the major part of the failed bone transplant and the scar tissue were removed, again using a pre-auricular incision in a second operative procedure 4 months after the first one (06/1994; Figure 4a,b).

Nine months later (03/1995), a large bony defect and complete absence of the condyle was assessed on the radiographs (Figure 5a–d). Severe reduction in lower jaw mobility and a deflection in the opening of the jaw to the left was diagnosed clinically. There was a severe asymmetry in the lower posterior face height. The occlusal plane in the upper jaw was inclined upwards to the right and there was marked dentoalveolar compensation. Dentofacial orthopaedic treatment with functional appliances could not be used as there was a fibrous pseudo-arthrosis at the affected side and, thus, absence of condylar growth.

A third surgical intervention was planned 4 months later when the patient was 12 years old (07/1995). A new costo-chondral graft was placed, this time using a retro-mandibular approach, allowing for a dissection of the gonial angle and the ascending ramus; the coronoid process had to be resected in order to allow lengthening of the ramus ascendens by placing the graft. The costo-chondral part of the sixth left rib served as a graft, which was fixed using titanium osteosynthesis plates. A bimaxillary splint was manufactured preoperatively into an over-corrected position. The mandibular dental midline was over-corrected to the left side. At the affected side, an infra-occlusion of the upper and lower posterior teeth was present; a lateral open bite of 12 mm was recorded in the new mandibular position.

Four weeks of inter-maxillary fixation with the splint and intra-oral elastics was applied (Figure 6a–f). The patient was instructed to wear the splint daily for 18 hours and then to remove it for 6 hours, so that unloaded mandibular movements could be performed in order to avoid ankylosis of the joint and to improve the healing. After some weeks the mandibular position seemed stable, and the patient was instructed to wear the splint during the evenings and at nights only. A soft diet was prescribed for 6 weeks and she was not allowed to do any sports.

Two months post-operatively (10/1995), orthodontic treatment was continued aiming to extrude the upper and lower molars, premolars, canines and incisors especially on the affected side, while modifying the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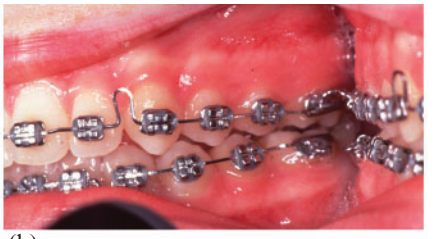
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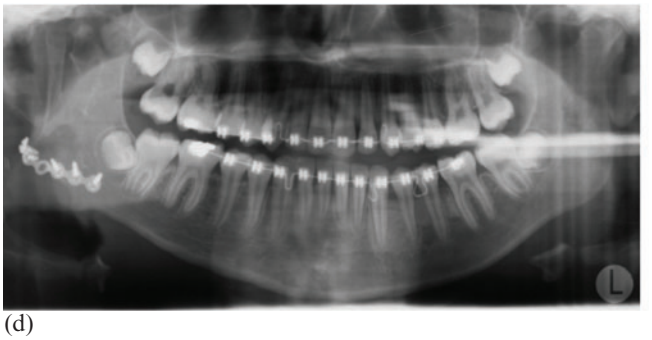
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Figure 8 (a–c) During the fixed appliance therapy. (d,e) OPT and RSP taken during fixed appliance therapy. (f) Correction of the occlusal plane: parallel to the bi-pupilar plane

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) was used in combination with elastics attached to brackets on the premolars (Figure 7a–j). The patient was instructed to wear this appliance day and night except for meals. The lateral open bite was partly closed and the inclined occlusal plane was almost corrected. After this extrusion stage, the lower dental arch was aligned and leveled against the idealized upper occlusal plane using an orthodontic removable appliance.

Throughout this treatment the patient had a markedly reduced mandibular opening. By means of supportive physiotherapy mouth opening could be increased from 2.3 to 20 mm. A few months later, some dysfunction was noticed at the healthy joint, probably due to hypermobility. Further radiographs revealed flattening of the left condyle. Physical therapy and soft diet were prescribed.

At the age of 13 years and 9 months the occlusion was consolidated to a final stage by means of full fixed appliances, using a modified edgewise technique (04/1996; Figure 8a–f). On appliance removal permanent fixed retainers were placed in upper and lower jaw, and a

Figure 9 (a–h) Records taken at the end of treatment, 3 years post-surgery. Patient aged 15½ years. (i,j) OPT and RSP at the end of the orthodontic treatment

Figure 10 (a–j) Records taken at 3 years post-treatment

(k)

(l)

Figure 10 (k,l) OPT and RSP 3 years after orthodontic treatment

functional appliance for night-time wear was added to stabilize the surgical correction and to prevent relapse during eventual further growth. The total orthodontic treatment time was 36 months (Figure 9a–j).

At one of the retention visits a facial asymmetry was mentioned by the patient: on the unaffected side, the gonial angle was more prominent compared with the affected side. The patient, however, rejected another surgical procedure to correct for this asymmetry (Figure 10a–j).

Despite this comment of the patient, the final result was aesthetically acceptable. The patient also demonstrated a good functional occlusion. The goals of the orthodontic treatment were achieved.

Discussion

In the literature, timing of treatment of HM has often been discussed. Different opinions are presented: one group of authors prefers early surgical intervention, 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'.

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 *et al.*[14] claim that an early surgical intervention (between 4 and 9 years old) in patients who need a TMJ-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 3-dimensional growth can also result in a bulk of tissue that can diminish the range of mandibular movements.

In this case, there was a failure of the first intervention, and the graft had to be removed and the environment had to be cleaned out for a second attempt. 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 as claimed by the advocators. One of the important contra-indications, however, is the situation in which TMJ-reconstruction is needed. Distraction can lengthen the jaw and the ramus, but cannot create a normal growing and functioning TMJ. Another disadvantage is the higher risk of infection during the active and passive period of lengthening. At the time this patient was treated there was not yet enough clinical experience built up with gradual distraction osteogenesis in our University Hospital. Therefore, this was not considered an option in this patient.

Conclusion

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. The condyle showed growth capacity and the temporomandibular joint was fully functionally restored.

After 5 years of team treatment by the orthodontist and the maxillofacial surgeon, facial and occlusal symmetry were established. The occlusion appears stable after 3 years of retention. 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-interventions.

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