

Surgical management of hemifacial microsomia in a growing patient: Systematic review of the literature

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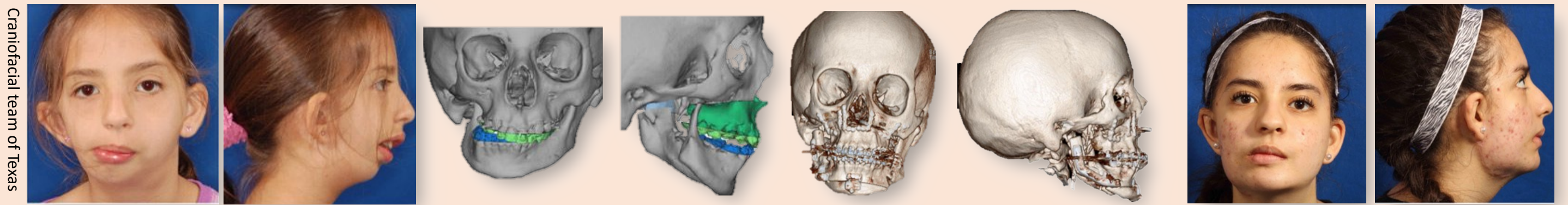
Hemifacial microsomia

Hemifacial microsomia (HFM) (OMIM N°164210) is the second most common disorder that affects craniofacial region after cleft lip/palate. Clinically, HFM is characterized by defects of facial growth, most often asymmetrical, in which facial bones (maxilla, mandible, zygoma or temporal bone), facial muscles and nervous tissues are affected.

The main etiology of HFM is not well established; it is considered as a complex genetic and environmental based embryonic pathology. Most reported cases of HFM are sporadic; some were autosomal recessive or dominant inherited cases. Whole-exome or genome sequencing have shown that HFM may be caused by heterozygote mutations of SF3B2 gene.

Surgical repair of HFM

Therapeutic options at our disposal are the mandibular osteogenic distraction (MOD) and costochondral grafts (CCG). These procedures would allow the early correction of mandibular asymmetry, enhance facial growth which results in the improvement of associated dysfunctions.



Systematic review of literature

Therapeutical protocols vary between authors. Besides, the success and the recurrence rates and the postoperative complications are still difficult to compare. Moreover, there is a debate concerning the decision-making criteria for the ideal age of intervention

Objectives: To perform a systematic review of literature to identify and update the current data concerning surgical protocols used to repair mandibular asymmetry in Hemifacial microsomia (HFM) in growing patients. More precisely, we aimed to compare relapses, complication rates, short- and long-term results of Mandibular Osteogenic Distraction (MOD) and costochondral grafts (CCG), two main techniques used to repair HFM.

Results

1. Data analysis and interpretation

Out of 23 included studies. 6 studies had OCEBM level of evidence of 5, 16 studies with OCEBM of 4 and only one study with OCEBM level of evidence of 3b. No statistical analysis was intentionally performed in this systematic review.

2. Age of surgical repair analysis and interpretation

For MOD, the average age of surgical repair was 8.8-year-old; For CCG, in our review, the average age of surgical repair was 7.02-year-old.

3. Clinical diagnosis and severity of HFM (Pruzanski-Kaban type modified by Madrid)

Type I: Mandible ramus TMJ were morphologically normal while present variable hypoplastic forms.

Type IIa: Ramus and TMJ are hypoplastic and present morphological deformities.

Type IIb: Severe type IIa and is associated with displacement of mandible medially and anteriorly.

Type III: Mandible ramus and TMJ are completely absent while temporalis and lateral pterygoid muscles (when existed) are not inserted to the remaining mandible mass.

Type IV: It is a type III associated to mandibular body hypoplasia.

4. Immediate and long-term post-operative results

All studies reported an immediate post-operative improvement of mandibular deficiency and hypoplasia.

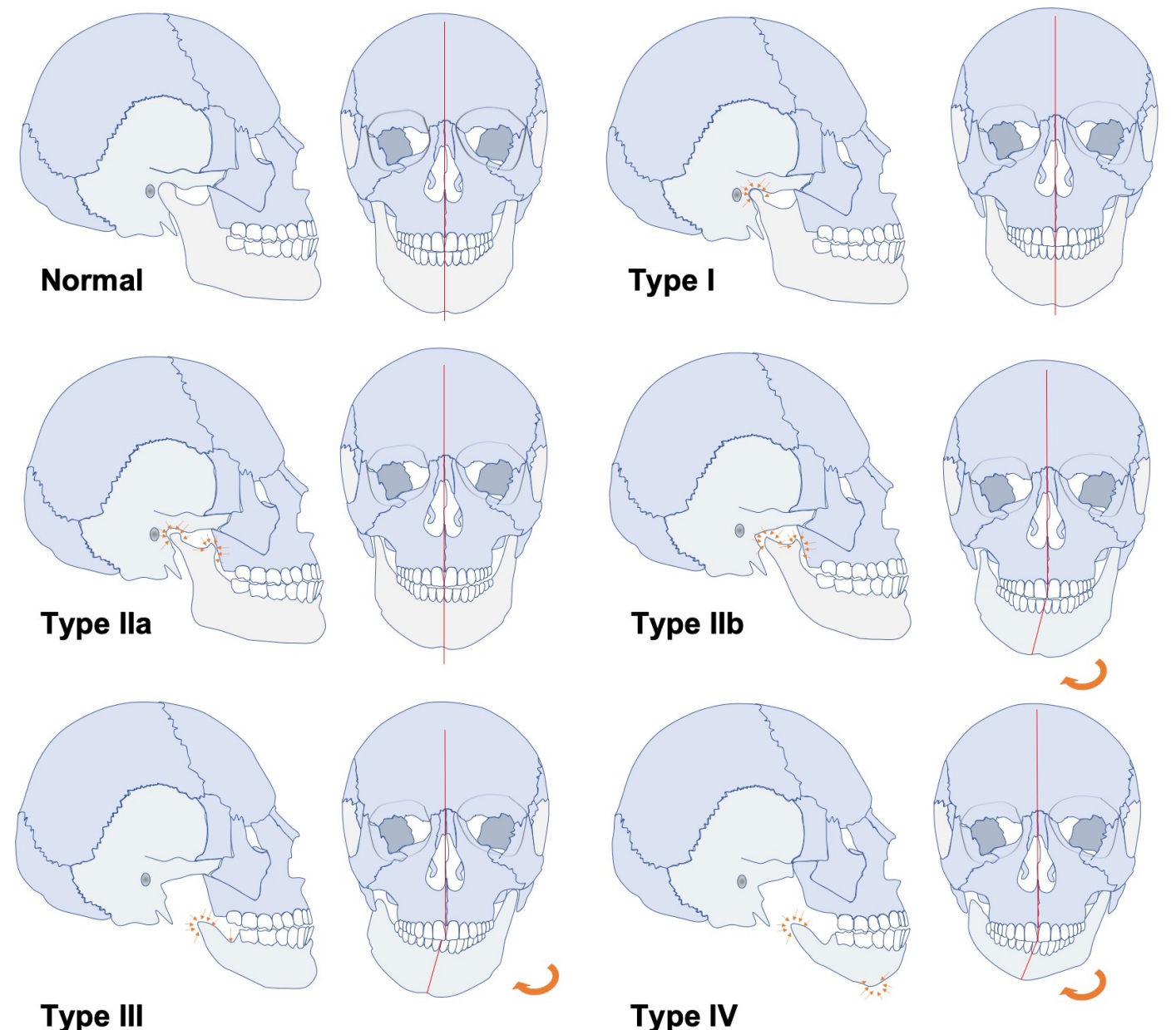
5. Associated Orthodontic treatment

Orthodontic and/or orthopedic treatments before distraction would allow the correction of occlusal canting, coordinate the width of dental arches by enhancing maxillary growth on the affected side. This would allow the establishment of a stable occlusion after surgical repair, reducing the risk of recurrence.

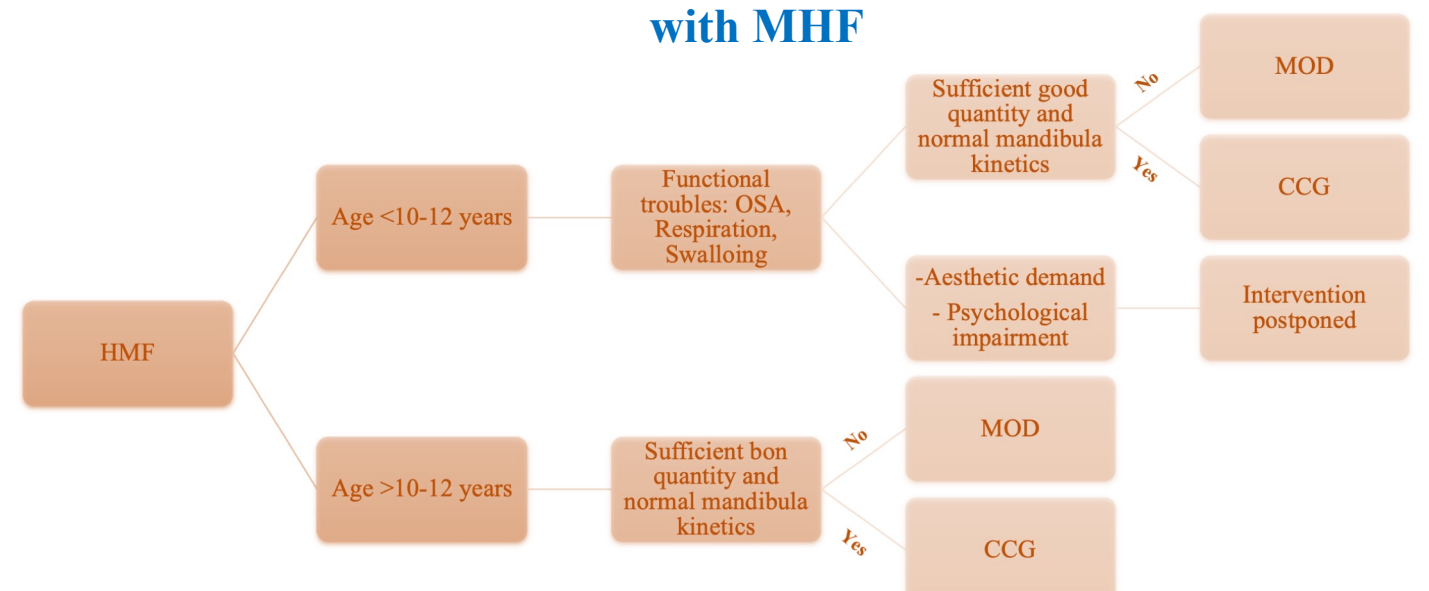
6. Psychological factors and quality of life:

Early MOD was beneficial both psychosocially and for cost-effectiveness or QALY score.

Severity of HFM (Pruzanski-Kaban type modified by Madrid)



Proposal of decision making for the management of a patient presenting with MHF



Conclusions

This systematic review identified 2 points: firstly, performing an MOD or an early CCG improves functional and aesthetic parameters in the short term, but recurrence is very common, secondly, there is no consensus allowing to candidate a gold standard protocol. Evaluating the quality of life of HFM patients is necessary and should be mainly considered when facial asymmetry is repaired.