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Correction chirurgicale de l'hypoplasie mandibulaire dans les microsomies hémi-faciales : Une étude rétrospective de 39 cas

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ABREVIATIONS

ATM	Articulation Temporo-Mandibulaire
CAE	Conduit Auditif Externe
CCG	Costo-Chondral Graft
CD	Chin Deviation
DO	Distraction Osteogenesis
HFM	HemiFacial Microsomia
IVP	Insuffisance Verticale Postérieure
LAST	LAST (follow-up)
MHF	Microsomie hémi-faciale
MMO	Maximal Mouth Opening
OAVS	Oculo-Auriculo-Vertebral Spectrum
OSBM	Ostétomie Sagittale de la Branche Montante
OVRS	Ostéotomie Verticale RétroSpigienne d'allongement de la branche montante
PA	Postero-Anterior (cephalograms)
POSTOP	POSTOPerative (time)
PREOP	PREOPerative (time)
PVI	Posterior Vertical Insufficiency
SD	Standard Deviation
SM	Distance entre la ligne supra-orbitaire et le point occlusal mandibulaire côté sain
SM'	Distance entre la ligne supra-orbitaire et le point occlusal mandibulaire côté atteint
SO	Distance entre la ligne supra-orbitaire et le point occlusal maxillaire côté sain
SO'	Distance entre la ligne supra-orbitaire et le point occlusal maxillaire côté atteint
SSO	Sagital Split Osteotomy
TMJ	Temporo-Mandibular Joint
VRO	Vertical Ramus Osteotomy

INTRODUCTION

La microsomie hémifaciale (MHF) est une anomalie du développement touchant les structures cervico-faciales dérivant des premier et deuxième arcs branchiaux. Cette pathologie fait partie d'une entité plus large appelée Spectre Oculo-Auriculo-Vertebral (OAVS) (1). Ce syndrome est caractérisé par la variabilité des atteintes cliniques (2), les trois anomalies les plus communes étant l'atteinte de l'oreille, de la mandibule et l'hypoplasie des tissus mous (figure a) (3). La malformation est le plus souvent unilatérale mais une atteinte bilatérale symétrique ou asymétrique peut survenir (4). D'autres malformations générales ou cranio-faciales sont associées dans plus de 50% des cas et doivent être recherchées (5). A l'étage cranio-facial les anomalies les plus fréquemment rencontrées sont les chondromes pré-tragiens (44,4%), l'aplasie du conduit auditif externe (25,1%) et les atteintes oculaires (24,3%). Les malformations médullaires cervicales (24,3%) et les pathologies cardiaques congénitales (27,8%) représentent les anomalies extra-crâniennes les plus fréquentes dans cette pathologie (1).

L'incidence de la MHF de 1 pour 5600 naissances rapportée dans la littérature (6,7) a probablement été surestimée ; des études récentes ont rapporté une prévalence des syndromes OAVS de 3,8 pour 100 000 naissances en Europe (1) faisant de la MHF l'anomalie congénitale la plus fréquente après les fentes labio-alvéolo-palatines (8).

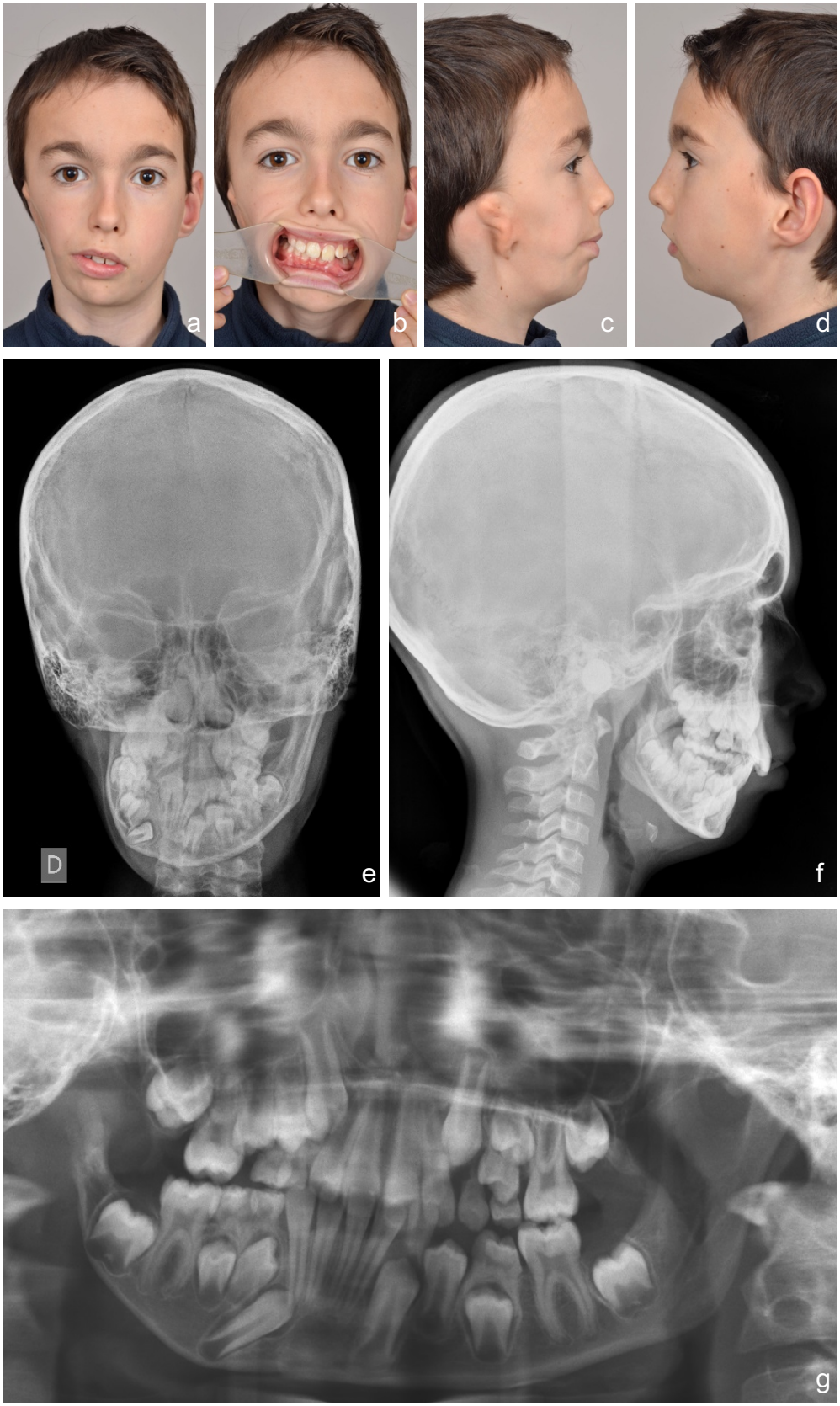


Figure a. Enfant porteur d'une microsomie hémi-faciale droite grade III de Pruzansky. Photographies de face (a), en occlusion (b), de profil (c et d). Téléradiographies de face (e), de profil (f) et panoramique dentaire (g). Noter l'obliquité du plan d'occlusion en haut à droite et la déviation du menton du côté de l'atteinte. Hypoplasie sévère de l'oreille externe et atteinte des tissus mous de l'hémiface droite. Hypoplasie sévère de la branche montante droite. Source : Pr Mercier.

De nombreuses classifications de la MHF ont été proposées (4,9,10) mais la plus utilisée est la classification décrite par Pruzansky qui distingue trois grades de l'atteinte mandibulaire allant de la simple diminution de taille à l'agénésie complète du ramus (11). Cette classification a été secondairement modifiée par Kaban et al. séparant le grade II en IIA et IIB en fonction de l'atteinte de l'articulation temporo-mandibulaire (ATM) (tableau a) (12).

La classification OMENS proposée par Vento *et al*. dissocie les cinq atteintes faciales majeures rendant compte de la grande hétérogénéité de la maladie (tableau b) (13).

Le grand nombre de classifications existantes dans la MHF et leur échec dans la collecte de la variabilité phénotypique de la maladie rend difficile la définition d'un protocole thérapeutique standardisé guidé par une classification.

Classification de Pruzansky modifiée par Kaban *et al.*	
Grade I	Ramus hypoplasique à un degré variable mais anatomie mandibulaire conservée
Grade II	IIA – Le ramus mandibulaire, le condyle et l'ATM sont présents mais hypoplasiques et de forme anormale IIB – Ramus hypoplasique, de forme anormale et en position antéro-médiale ; absence d'articulation avec l'os temporal
Grade III	Déformation sévère de la branche montante avec absence d'ATM

Tableau a. Classification de Pruzansky modifiée par Kaban et al.; ATM : Articulation Temporo-Mandibulaire.

Classification OMENS			
Anomalies de l'orbite		**Anomalies de l'oreille**	
O_0	Orbite de taille et de position normales	E_0	Oreille normale
O_1	Anomalie de taille	E_1	Hypoplasie auriculaire, toutes les structures sont présentes
O_2	Anomalie de position ($O_2\uparrow$ ou $O_2\downarrow$ selon la position horizontale de l'orbite)	E_2	Absence de CAE et hypoplasie du pavillon
O_3	Anomalies de position et de taille	E_3	Absence d'oreille et malposition du lobule
Anomalies de la mandibule		**Anomalies nerveuses (VII)**	
M_0	Mandibule normale	N_0	Nerf facial normal
M_1	Diminution de la taille du ramus	N_1	Atteinte des branches supérieures
M_2	Anomalie de la forme du ramus (M_{2A} si la cavité glénoïde est dans la même position que le côté sain, M_{2B} si l'ATM est déplacée avec une hypoplasie condylienne sévère	N_2	Atteinte des branches inférieures
		N_3	Atteinte de toutes les branches du nerf facial
		Atteinte des tissus mous	
M_3	Absence complète de ramus, de cavité glénoïde et d'ATM	S_0	Pas d'atteinte musculaire ou des tissus mous
		S_1	Déficit minime
		S_2	Déficit modéré
		S_3	Déficit sévère

Tableau b. Classification OMENS ; ATM : Articulation Temporo-Mandibulaire ; CAE : Conduit Auditif Externe.

La prise en charge de la MHF requiert une approche multidisciplinaire. La reconstruction de la mandibule hypoplasique demeure controversée et l'âge de la première chirurgie diffère en fonction des centres (14). Pour résumer, les équipes qui considèrent que la malformation ne subit pas d'aggravation avec la croissance recommandent de réaliser la reconstruction primaire en fin de croissance (15–17). A l'inverse, les équipes considérant que l'atteinte est progressive sont en faveur d'un traitement chirurgical précoce pour prévenir et limiter l'aggravation de l'asymétrie faciale (12,18). De nombreuses solutions chirurgicales existent et peuvent être combinées successivement chez le même individu pendant et après la croissance (14).

Les objectifs du traitement sont de restaurer et de préserver la symétrie du visage et les fonctions oro-faciales (19). Grace à son potentiel de croissance intrinsèque, le greffon chondro-costal (GCC) représente le traitement de référence pour la reconstruction du ramus et de l'ATM dans les grades IIB et III de la

classification de Pruzansky-Kaban (14,20). La chirurgie orthognathique d'élongation et de rotation mandibulaire a longtemps été proposée pour les grades I et II mais tend à être remplacée par la distraction osseuse (DO) mandibulaire, avec des résultats variables (21). Quelle que soit la technique chirurgicale employée chez l'enfant, une dégradation clinique du résultat est communément observée nécessitant des procédures additionnelles en fin de croissance dont les parents doivent être prévenus (22,23).

Depuis plus de 30 ans nous utilisons un protocole unique de chirurgie primaire avec pour objectifs :

(1) d'augmenter la taille de la mandibule hypoplasique,
(2) créer une ATM fonctionnelle,
(3) stimuler la croissance du maxillaire
(4) et restaurer une occlusion fonctionnelle (19,24,25).

La GCC pour les stades évolués ou l'ostéotomie verticale rétrospigienne d'allongement de la branche montante (OVRS) pour des stades modérés permettent d'atteindre ces objectifs en créant un ramus fonctionnel articulé avec la base du crâne.

Du fait de la faible prévalence de la maladie et des difficultés de suivi, il existe peu d'articles rapportant les résultats à long terme de la chirurgie primaire mandibulaire chez les jeunes patients souffrant de MHF (26). La plupart des études sont constituées de petits échantillons et le suivi atteint rarement la fin de la croissance chez ces enfants (21). L'objectif de notre étude était d'évaluer les résultats à long terme de la chirurgie primaire mandibulaire sur le plan esthétique, architectural et fonctionnel à partir d'une série de jeunes patients atteints de MHF.

ARTICLE

ABSTRACT

Introduction: Hemifacial microsomia (HFM) is a developmental disorder affecting head and neck structures derived from the first and second branchial arches. Repair of the mandibular deformity remains controversial and there is a poor literature regarding the late outcomes. The aim of this study was to evaluate architectural and aesthetical long-term results of the mandibular primary surgery in patients with HFM.

Methods: Thirty-nine patients with types II and III HFM were included in this retrospective study. Depending on the mandibular deformity assessed by the Pruzansky classification, patients were treated using a costochondral graft (CCG) or a vertical ramus osteotomy (VRO). Architectural and aesthetical parameters were evaluated using standardized frontal cephalometric and photographic measurements preoperatively, postoperatively and at the end of the follow-up.

Results: The mean age of primary mandibular surgery was 13.1 ±3.5 years (7-21) and the mean follow-up duration 70.4 ±57.1 months (0-252). Thirty-three patients (82.5%) were considered as having reached full growth at last follow-up. A CCG was given in 24 cases mainly in types IIB and III while VRO was realized in 15 IIA cases. The architectural analysis showed the restoration of a levelled occlusal canting at the end of follow-up ($p<0.0001$); the chin deviation was immediately corrected by surgery but a trend to recurrence was noted at the last follow-up ($p<0.0001$). The aesthetical evaluation revealed a significant improvement of the chin deflection and the correction of the lip commissural line tilting ($p<0.0001$). A non significant correlation was found between the soft tissue statute and the trend for the deformity to relapse particularly regarding the maxillary occlusal canting ($p=0.06$). Twenty-three percent of patients required an additional orthognathic revision. No severe complication was reported after the procedure.

Conclusion: CCG and VRO remain interesting and safe procedures with good outcomes allowing a one stage correction of occlusion and preservation of mandibular growth in young patients with types II and III HFM.

INTRODUCTION

Hemifacial microsomia (HFM) is a developmental disorder affecting head and neck structures derived from the first and second branchial arches. It is part of a larger entity called Oculo-Auriculo-Vertebral Spectrum (OAVS) (1). The syndrome is characterized by the variability of clinical findings (2), the three major features are auricular, mandibular and soft tissues hypoplasia (3). With an overall incidence of 1 in 5600 births (6,7), HFM is the second most common craniofacial defect after cleft lip/palate. The deformity is usually unilateral but symmetrical or asymmetrical bilateral cases can exist (4). In more than 50% of cases others craniofacial and/or general anomalies are associated and have to be looked for (5).

Many classifications of HFM have been proposed (4,9,10) but the most commonly used is the Pruzansky one categorizing three grades of mandibular deformity (11). This classification has been later modified by Kaban *et al*. separating grade II in IIA and IIB according to the presence or absence of temporo-mandibular joint (TMJ) (table 1) (12). An other interesting classification is the OMENS score dissociating the Orbital, Mandibular, Ear (external), cranial Nerve and Soft tissues deformities, reflecting the high heterogeneity of the disease (27).

Pruzansky classification modified by Kaban	
Type I	All mandibular and TMJ components are present and normal in shape but hypoplastic to a variable degree
Type II	IIA – The mandibular ramus, condyle and TMJ are present but hypoplastic and abnormal in shape IIB – The mandibular ramus is hypoplastic and markedly abnormal in form and location, being medial and anterior; there is no articulation with the temporal bone
Type III	The mandibular ramus, condyle, and TMJ are absent. The lateral pterygoid and temporalis muscles, if present, are not attached to the mandibular remnant

Table 1. Pruzansky modified Kaban classification; TMJ: Temporo-Mandibular Joint.

HFM requires a multidisciplinary approach. Repair of the mandible anomalies remains controversial; timing of treatment differing largely among centres (14). To summarize, authors considering that facial asymmetry does not change with growth, recommend to delay primary surgery at the end of growth (15–17). In the other hand, authors considering HFM as a progressive disease correlated to the skeletal involvement, advocate for an early surgical treatment to prevent worsening of facial

asymmetry (12,18). Numerous surgical options exist that can successively been combined in the same individual during and/or after growth (14). The goals of treatment are to achieve and to maintain optimal facial symmetry and to improve facial functions after completion of craniofacial growth (19). Thanks to an inherent growth potential, costochondral graft (CCG) remains the gold standard for the TMJ and ramus reconstruction in types IIB and III of the Pruzansky-Kaban classification (14,20). Elongation and rotation orthognathic procedures have long been proposed for types I and II, but tend to be replaced by mandibular distraction osteogenesis (DO) with various results so far (21). Whatever the surgical technique used in children, clinical deterioration is commonly observed, requiring further procedures which parents have to be informed (22,28).

Because of the low prevalence of the deformity and the difficulties of follow-up, there are very few papers relating the long term results of primary mandibular surgery in HFM young patients (26). Most of studies are based on few patients and the mean follow-up rarely reaches the end of growth (21). The purpose of this study was to retrospectively evaluate long-term aesthetic, architectural and functional results of the primary mandibular surgery in young patients with HFM. We have been using a unique primary surgery protocol for more than 30 years with the objectives of (1) increasing the size of underdeveloped mandible, (2) creating a functional TMJ, (3) releasing restricted growth of maxilla and (4) restoring a functional occlusion (19,24,25). We assume that CCG or vertical ramus osteotomy (VRO) for moderate stages could achieve these goals as it allows creating a functional ramus articulated with temporal bone.

MATERIALS AND METHODS

Patients presenting from 1981 to 2015 with hemifacial microsomia in the Maxillofacial Surgery and Stomatology Department of the Nantes University Hospital, France, were included in the study and retrospectively analysed. The exclusion criteria were non operated patients, primary mandibular surgery realized in another centre, operated patients without primary mandibular surgery.

Data collection:

Patient's chart was reviewed and data were completed documenting date of birth, side of involvement (right or left), dates and types of operations. The Pruzansky-Kaban grade for the mandibular hypoplasia and the soft tissue involvement according to the OMENS classification were assessed for each patient. The others associated cranio-facial and visceral anomalies were also collected.

Primary surgery protocol:

When possible the primary mandibular surgery was realized between 9 and 12 years old, after the permanent mandibular canine teeth have beginning their eruption, to perform an associated genioplasty if needed. The correction of mandibular hypoplasia was achieved using CCG or VRO based on Pruzansky-Kaban grade. All procedures were performed by the same senior author (J.M.) under general anaesthesia with nasotracheal intubation, using a low submandibular incision, often completed with an intraoral approach. No pre-auricular approach was needed. For the CCG harvesting, the contralateral sixth or seventh rib was collected through a sub-mammary incision and a perichondrium strip was maintained on the costochondral junction. An average 5mm of carved cartilage portion was left in place on the graft. Then the CCG was placed in contact with the skull base, and fixed laterally to the mandibular stump thanks to bicortical screws. VRO was realized according to the Caldwell-Letterman technique (29). Briefly, an extra-oral approach was done in all patients. After protecting the mandibular nerve, the ramus was sectioned from the sigmoid incisura to the pre angular notch. The ramus could be elongated while functional mandibular condyle stayed in the same location. Osteosynthesis was performed with 0.8 or 1mm thickness miniplate. In all cases a posterior open bite was created on the affected side and an interocclusal splint was positioned and progressively reduced in size to promote secondary maxillary teeth

egression. The primary surgery was sometimes associated with conventional orthognathic procedure such Le Fort I osteotomy (LF1) in eldest patients, as well as contralateral sagittal split osteotomy (SSO) or genioplasty. Some patients required an additional procedure at the end of growth consisting in VRO, LF1, SSO or genioplasty. An orthodontic preparation was most often needed and maxilla-mandibular elastic therapy was performed postoperatively for at least six weeks. In cases of CCG, active TMJ physiotherapy was prescribed (30,31) to promote early recovering of motion.

Clinical evaluation:

Frontal standardized photographs were analysed preoperatively, postoperatively 6 months after the intervention and at the end of the follow-up. To assess the chin deviation, the angle α between the facial median line and a line from the glabella to chin was measured. The angle β of the tilted lip commissure plane was measured using a perpendicular line from the facial median line (Fig. 1).

Radiographic evaluation:

A cephalometric analysis (32–34) was performed on Postero-Anterior (PA) cephalograms preoperatively, postoperatively in the few days following surgery and at the last time of the follow-up. Two reference lines were used for the frontal analysis. The supra-orbital line joining the top of the orbital roofs served as the horizontal reference line, while a perpendicular line defined by the median axis of symmetry passing through the cristagalli served as the vertical reference line. In complex cases presenting with orbital asymmetry, a horizontal reference line was drawn using the landmarks of the petrous part of the temporal bones or the zygomatic arches.

SO was defined as the distance measured between supraorbital line and the most prominent cusp of the second maxillary molar or occlusal point on the normal side. SM was defined as the distance between supraorbital line and the most prominent cusp of the second mandibular molar or occlusal point on the normal side. The affected side was designed SO’ and SM’. A ratio of SO’/SO assessed the maxillary canting of the occlusal plane (Fig.1) while SM’/SM represented the mandibular occlusal plane canting.

The chin deviation (CD) assessed the laterogenia on frontal X-ray and was defined as the distance between the projection point of the axis of lower incisors on the chin symphysis and the median facial line.

For patients receiving a secondary procedure, the preoperative parameters of the second intervention were defined as the last follow-up data of the primary mandibular surgery. Data from secondary surgery were not collected.

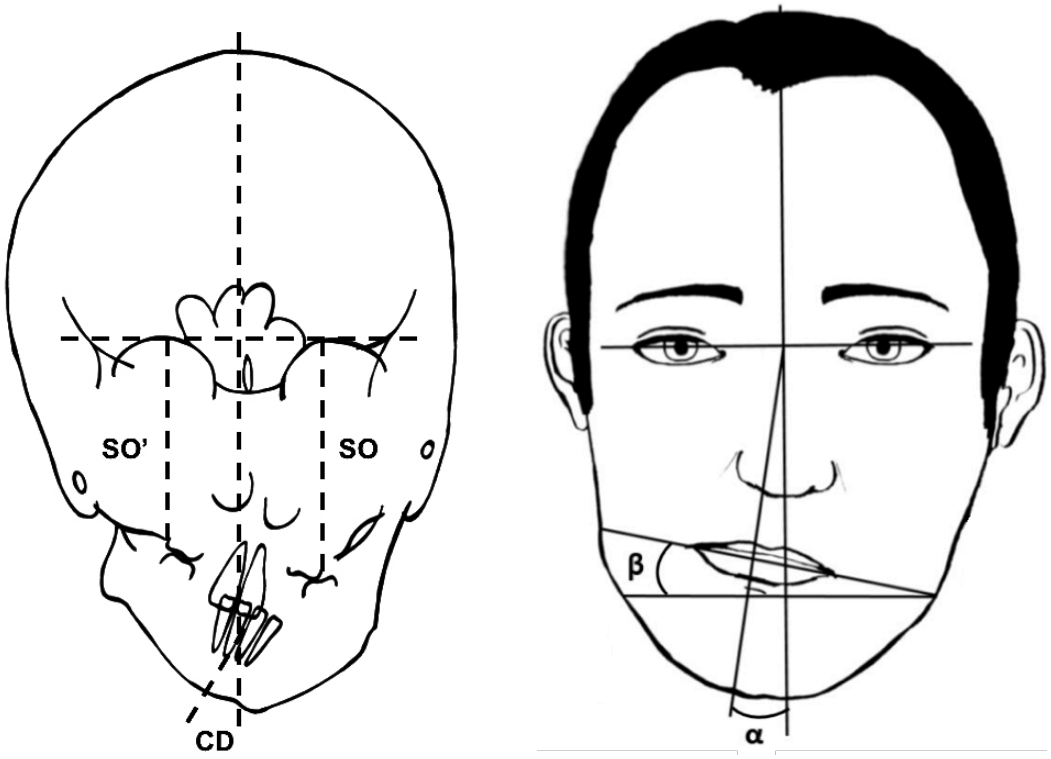


Figure 1. A) Cephalometric analysis on frontal x-rays. SO: Distance between supra-orbital line and maxillary molar occlusal point on the non affected side. SO': Distance SO on the affected side. CD: distance between the projection point of the axis of lower central incisors on the chin symphysis and the median facial line. B) Clinical evaluation of the facial soft tissues. Angle α: Chin deviation. Angle β: Bi-commissural line tilting.

Secondary endpoints:

Both surgical procedures CGG and VRO were compared according to the aesthetical and architectural parameter. Correlation between aesthetic and architectural results and age of surgery, soft tissue statute and Pruzansky gravity score was tested. Surgical complications and impact of surgery on TMJ function were collected.

Statistical analysis:

The methodical error of the cephalometric and facial measurements was assessed by Dahlberg's formula (mean square error ($S.E^2$)=$d^2/2n$ where d is the difference between the first and the second measurements, and n the number of double measurement) (35,36). To determine the intra-observer error, cephalometric lengths and facial angles were measured twice at 2-weeks intervals by the same investigator in 15 random patients.

Statistical analysis was achieved thanks to GraphPad Prism 5.0 for Mac (GraphPad Software, La Jolla, CA, United-States). Quantitative data were analysed using a paired t-test when there were more than 30 replicates values and a Wilcoxon test when there were less than 30 paired observations. Complementary analysis was performed for paired observations using a one way ANOVA with Bonferroni's post test. Qualitative data were compared using Chi2 or Fisher exact test. Secondary endpoints were studied using a linear regression. A p-value lower than 0.05 ($p<0.05$) indicated a statistical significance.

RESULTS

Epidemiologic data:

Forty-eight patients presenting with HFM were reviewed in the study. Nine patients were excluded according to the exclusion criteria (4 patients were not operated, 4 others did not receive a primary mandibular surgery and one patient was operated in another centre). On the 39 remaining patients, 32 (82%) were considered as having reached full growth, with a mean age at the end of follow-up of 20.8 ±3.9 years. Seven (18%) patients were growing children, with a last follow-up mean age of 13.2 ±0.6 years.

Mean age of primary mandibular surgery was 13.1 ±3.5 years in our series, while secondary mandibular intervention was realized at 17.4 ±2.7 years. The mean follow-up duration after the primary procedure was 70.4 ±57.1 months reaching 81.9 ±58.5 months in the full growth group (Table 2).

Patients characteristics	
Sex: Females / Males, n (%)	21 (53.8) / 18 (46.2) (p=0.65)
Affected side: Right / Left, n (%)	25 (64.1) / 14 (35.9) (p=0.01)
Pruzansky-Kaban classification: IIA / IIB / III, n (%)	17 (43.6) / 12 (30.8) / 10 (25.6)
Soft tissue statute (OMENS classification): 0 / 1 / 2 / 3, n (%)	3 (7.7) / 17 (43.6) / 12 (30.8) / 7 (17.9)
Age of primary intervention (years), mean ± S.D. (range)	13.1 ±3.5 (7-21)
Age of secondary intervention (years), mean ± S.D. (range)	17.4 ±2.7 (13-21)
Follow-up duration (months), mean ± S.D. (range)	70.4 ±57.1 (0-252)

Table 2. Patients characteristics; n: Number of patients; SD: Standard deviation.

A significant predominance of the right affecting side was found (64.1% vs 35.9%, p=0.01), 4 cases were bilateral with a right prominence. The Pruzansky-Kaban modified classification of the mandibular hypoplasia revealed a predominance of types IIA compared with types IIB and III (43.6%, 30.8% and 25.6%).

All the patients analysed presented with mandibular hypoplasia. Ear abnormalities were present in 97.4% of cases, ocular anomalies in 25.6%, facial nerve palsy and macrostomia in 15.4%. Other frequent anomalies were velopharyngeal insufficiency (17.9%), and vertebral malformations (25.6%). All the associated abnormalities are presented in table 3.

Associated anomalies		
Main abnormalities	Mandibular hypoplasia, n (%)	39 (100)
	Ear abnormalities (microtia, preauricular tags, conductive hearing loss), n (%)	38 (97.4)
	Ocular (orbital dystopia, epibulbar dermoids, coloboma, strabism), n (%)	10 (25.6)
	VII nerve palsy, n (%)	6 (15.4)
	Macrostomia, n (%)	6 (14.4)
Associated craniofacial anomalies	Velopharyngeal insufficiency, n (%)	7 (17.9)
	Cleft lip and/or palate, n (%)	2 (5.12)
	Ankyloglossia, tongue hypoplasia, n (%)	2 (5.12)
	Skull base asymmetry, n (%)	1 (2.6)
	Peritonsillar cyst, n (%)	1 (2.6)
	Oesophagus atresia and tracheo-oesophageal fistula, n (%)	1 (2.6)
	Choanal atresia, n (%)	1 (2.6)
	Dental agenesia, n (%)	1 (2.6)
	Endobuccal adhesions, n (%)	1 (2.6)
	Temporo-maxillo-sphenoidal ankylosis, n (%)	1 (2.6)
	Meningocele, n (%)	1 (2.6)
	Arachnoid cyst, n (%)	1 (2.6)
Others	Vertebral abnormalities, n (%)	10 (25.6)
	Thumb hypoplasia, n (%)	3 (7.7)
	Heart defect, n (%)	2 (5.12)
	Pylore stenosis, n (%)	1 (2.6)

Table 3. Associated anomalies; n: Number of patients.

Primary surgery protocol:

According to the primary intervention, 24 patients received a CCG (61.5%) and 15 patients were given a VRO (38.5%). Type of mandibular surgery was significantly associated with the Pruzansky-Kaban severity score, with a predominance of CCG in types IIB and III ($p<0.0001$) while a VRO was given in types IIA. The mandibular surgery was most often associated with other orthognathic procedures in three-quarters of the cases, with non-statistical correlation with the severity score ($p=0.54$). A contralateral sagittal split osteotomy and/or a genioplasty were given in more than half cases (table 4).

		Pruzansky-Kaban classification		
Primary surgical procedure	Total, n=39	IIA, n=17	IIB, n=12	III, n=10
Costochondral graft (CCG), n (%)	24 (61.5)	3 (17.6)	11 (91.7)	10 (100)
Vertical ramus osteotomy (VRO), n (%)	15 (38.5)	14 (82.3)	1 (8.3)	0 (0)

		Pruzansky-Kaban classification		
Associated procedures	Total, n=39	IIA, n=17	IIB, n=12	III, n=10
Isolated mandibular procedure, N (%)	8 (20.5)	2 (11.8)	4 (33.3)	2 (20)
Sagittal split osteotomy, N (%)	23 (59.0)	11 (64.7)	5 (41.7)	7 (70)
Genioplasty, N (%)	22 (56.4)	9 (52.9)	6 (50.0)	7 (70)
Le Fort I osteotomy, N (%)	10 (25.6)	6 (35.3)	0 (0)	4 (40)

Table 4. Primary surgical procedure: Type of interventions, associated orthognathic procedures; n: Number of patients; N: Number of procedures.

The secondary mandibular surgery was realized at a later stage in nine patients (23%) and consisted in bilateral SSO and genioplasty for 7 cases. Only one patient qualified for a mandibular VRO revision, and a LF1 osteotomy was required in two cases (table 5). No correlation was found with the severity score of the Pruzansky-Kaban classification (p=0.63).

		Pruzansky-Kaban classification		
Secondary surgical procedure	Total, n=9	IIA, n=2	IIB, n=4	III, n=3
Vertical ramus osteotomy, N (%)	1 (11.1)	1 (50.0)	0 (0)	0 (0)
Sagittal split osteotomy, N (%)	7 (77.8)	2 (100.0)	4 (100)	1 (33.3)
Genioplasty, N (%)	7 (77.8)	1 (50.0)	3 (75.0)	3 (100)
Le Fort I osteotomy, N (%)	2 (22.2)	0 (0)	1 (25.0)	1 (33.3)

Table 5. Secondary surgical procedure; n: Number of patients; N: Number of procedures.

Aesthetic evaluation:

The mean Dahlberg standard error for α and β angles measurement was 0.34° (0 - 2.25°).

Primary mandibular surgery allowed immediate and significant improvement in chin deflection and correction of the lip commissural line tilting in all the patients whatever the growth statute (3.36° ±2.42 pre-operatively vs 1.60° ±1.50 post-operatively for angle α; 4.19° ±3.03 pre-operatively vs 1.83° ±1.79 post-operatively for angle β; $p<0.0001$). Surgical results were stable over time regarding the chin

deviation while a 32.6 percent deterioration rate was observed in the lip commissural tilting with time (p=0.01) (figure 2).

Associated genioplasty and Le Fort I osteotomy did not affect the correction of the chin asymmetry nor the commissural canting between preoperative and last follow-up times (p=0.83 and p=0.27 respectively). Results of the aesthetical parameters measured at the three times of evaluation are presented in table 6.

	Preoperative	Postoperative	Last follow-up
Chin deviation α (°), mean ± SD			
All the patients (n=39)	3.36 ± 2.42	1.60 ± 1.50	1.78 ± 2.03
End of growth (n=32)	3.03 ± 2.46	1.46 ± 1.53	1.45 ± 1.84
With genioplasty (n=19)	3.00 ± 2.59	1.73 ± 1.49	1.61 ± 1.46
Without genioplasty (n=13)	3.09 ± 2.34	1.36 ± 1.63	1.18 ± 2.40
Lip commissural line tilting β (°), mean ± SD			
All the patients (n=39)	4.19 ± 3.03	1.83 ± 1.79	2.58 ± 2.47
End of growth (n=32)	3.79 ± 3.13	1.71 ± 1.78	2.48 ± 2.56
With Le Fort I osteotomy (n=10)	3.00 ± 2.40	1.71 ± 1.25	1.78 ± 2.28
Without Le Fort I osteotomy (n=22)	4.21 ± 3.07	1.79 ± 1.78	2.50 ± 2.57

Table 6. Chin deviation and lip commissural line tilting analysis on standardized photographs, depending on growth, completion of genioplasty or Le Fort I osteotomy; n: Number of patients; SD: Standard Deviation.

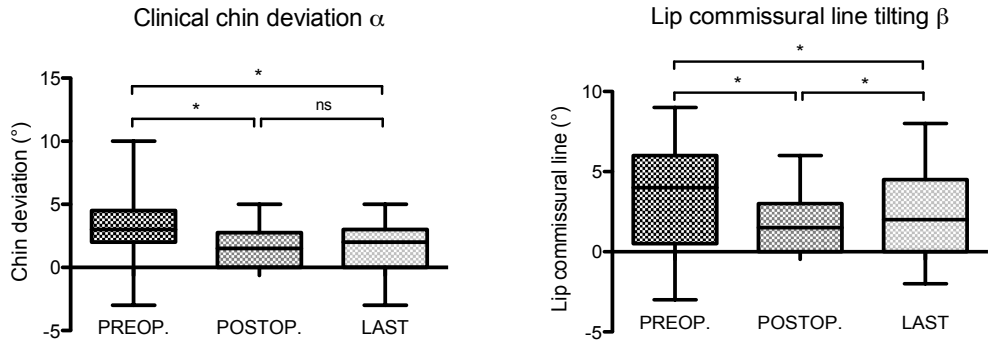


*Figure 2. Variation of chin deviation and lip commissural line tilting at preoperative (PREOP.), postoperative (POSTOP.) and last follow-up (LAST) times in patients having reached the end of growth; * p<0.05; ns: Non significant.*

Architectural results:

The mean Dahlberg standard error for distance measurement was 1.43mm (0 – 16) for SO', 1.50mm (0 – 9) for SO and 4.74mm (0 – 42.25) for CD. The mean Dahlberg score was $1.69.10^{-4}$ (0 – $1.13.10^{-3}$) for the SO'/SO ratio.

The primary mandibular surgery did not impact immediately the maxillary occlusal canting reflected by the SO'/SO ratio (0.939 ±0.039 in preoperative time vs 0.944 ±0.068 in postoperative period; p=0.54). Restoration of a levelled occlusal canting was observed at the end of follow-up for all patients (0.939 ±0.039 pre-operatively vs 0.972 ±0.041 at last follow-up; p<0.0001) (fig.3). Logically associated Le Fort I surgery immediately restored a normal maxillary canting (0.967 ±0.044 post-operatively vs 0.938 ±0.039 pre-operatively; p=0.004). No difference was observed in the maxillary flattening at the end of follow-up whatever the achievement of Le Fort I osteotomy (0.972 ±0.042 with associated Le Fort I osteotomy vs 0.973 ±0.041 Without Le Fort I osteotomy ; p=0.56). Results of architectural parameters on cephalometric analysis are listed in table 7.

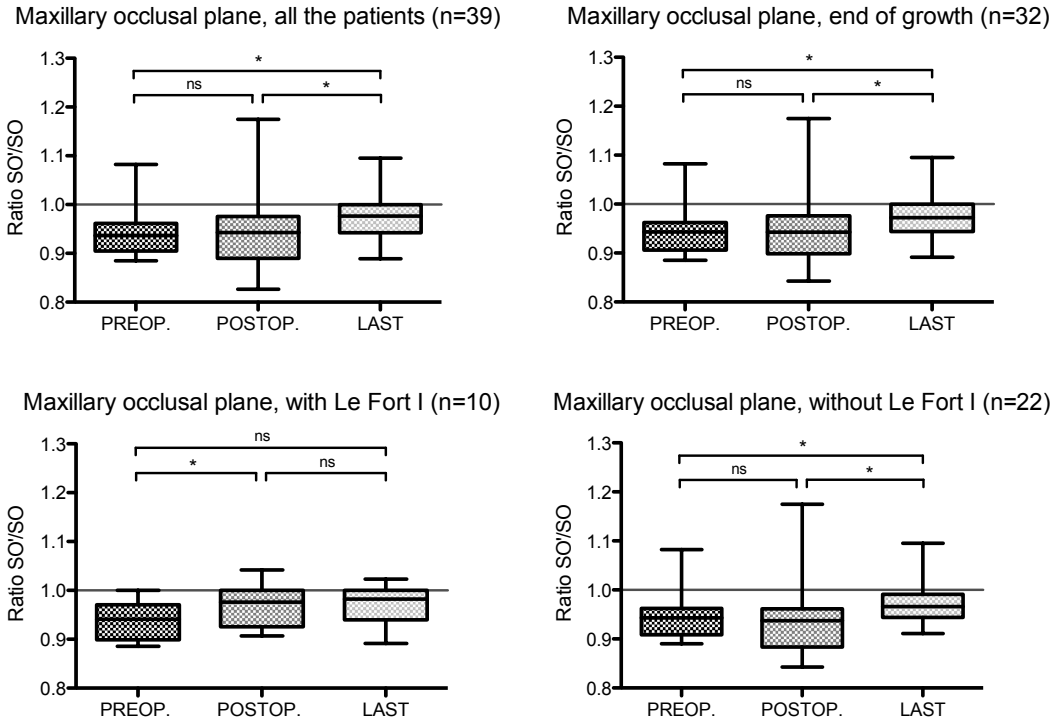


*Figure 3. Variation of the maxillary occlusal plane at preoperative (PREOP.), postoperative (POSTOP.) and last follow-up (LAST) times depending on growth and completion of Le Fort I; * p<0.05; ns: Non significant.*

The mandibular occlusal canting was significantly improved with the primary surgery in all patients. By lowering the affected side while creating a posterior open bite, an overcorrection was noted (SM'/SM ratio 0.939 ±0.039 in preoperative period vs 1.036 ±0.040 in postoperative time; p<0.0001). Results remained good with a slight trend to recurrence of the mandibular asymmetry at the last follow-up evaluation (p<0.0001).

	Preoperative	Postoperative	Last follow-up
Maxillary canting SO'/SO (ratio), mean ± SD			
All the patients (n=39)	0.939 ± 0.039	0.944 ± 0.068	0.972 ± 0.041
End of growth (n=32)	0.942 ± 0.041	0.946 ± 0.063	0.973 ± 0.040
With Le Fort I (n=10)	0.938 ± 0.039	0.967 ± 0.044	0.972 ± 0.042
Without Le Fort I (n=22)	0.943 ± 0.042	0.936 ± 0.069	0.973 ± 0.041
Mandibular canting SM'/SM (ratio), mean ± SD			
All the patients (n=39)	0.939 ± 0.039	1.035 ± 0.040	0.972 ± 0.041
End of growth (n=32)	0.941 ± 0.041	1.026 ± 0.033	0.973 ± 0.040
Chin deviation (mm), mean ± SD			
All the patients (n=39)	11.13 ± 8.23	3.35 ± 4.07	6.30 ± 5.09
End of growth (n=32)	9.97 ± 8.52	2.84 ± 3.93	5.54 ± 5.21
With genioplasty (n=19)	8.74 ± 9.88	3.32 ± 4.47	4.82 ± 4.49
Without genioplasty (n=13)	11.92 ± 5.63	2.08 ± 2.91	6.89 ± 6.43

Table 7. Maxillary and mandibular canting, and chin deviation analysis on frontal cephalometric X-rays, depending on growth, completion of Le Fort I or genioplasty; n: Number of patients; SD: Standard Deviation.

The primary surgery as well immediately corrected the chin position in all the patients (3.35mm ±4.07 post-operatively vs 11.13mm ±8.23 pre-operatively; p<0.0001) but the results did not remain stable in time with partial recurrence of the deviation (6.30mm ±5.09 at last follow-up vs 3.35mm ±4.07 post-operatively; p=0.02). The achievement of a concomitant genioplasty did not improve the immediate position of the chin (3.32mm ±4.47 post-operatively for the group with simultaneous genioplasty and 2.08mm ±2.91 for the group without genioplasty; p=0.24) but seemed to stabilize the chin position at the end of growth (figure 4).

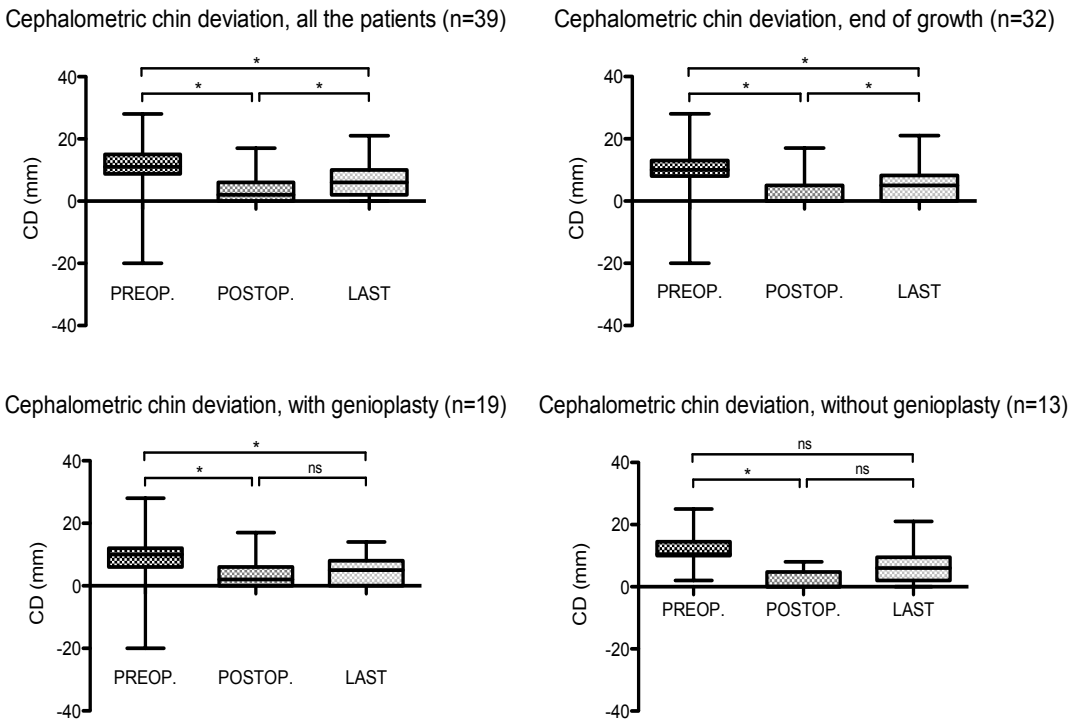


*Figure 4. Variation of the cephalometric chin deviation at preoperative (PREOP.), postoperative (POSTOP.) and last follow-up (LAST) times depending on growth and completion of genioplasty; * p<0.05; ns: Non significant.*

Secondary endpoints results:

Both VRO and CCG groups were compared according to the aesthetical and cephalometric parameters. There were no differences in the mean age of primary mandibular surgery (13.4 ±3.4 vs 12.9 ±3.6; p=0.79) nor in the mean follow-up time (76.9 ±37.9 vs 67.2 ±67.7; p=0.23). Logically, the preoperative parameters as a reflexion of the initial severity were significantly higher in the CCG group, while there was no significant difference at the last follow-up (table 8).

	Vertical ramus osteotomy (n=15)	Costochondral graft (n=24)	p
Chin deviation α (°), mean ± SD			
Preoperative	2.00 ±2.25	4.23 ±2.14	0.01
Postoperative	1.00 ±0.96	2.00 ±2.67	0.06
Last follow-up	1.47 ±1.30	1.86 ±1.91	0.75
Lip commissural line tilting β (°), mean ± SD			
Preoperative	3.07 ±2.67	4.91 ±3.09	0.05
Postoperative	1.43 ±1.60	2.10 ±1.90	0.26
Last follow-up	2.00 ±1.96	2.55 ±2.40	0.49
Maxillary canting SO'/SO (ratio), mean ± SD			
Preoperative	0.949 ±0.027	0.932 ±0.045	0.04
Postoperative	0.940 ±0.050	0.947 ±0.078	0.71
Last follow-up	0.982 ±0.049	0.973 ±0.032	0.75
Chin deviation (mm), mean ± SD			
Preoperative	7.13 ±9.90	13.64 ±5.79	0.01
Postoperative	1.80 ±2.88	4.41 ±4.47	0.04
Last follow-up	3.92 ±4.46	5.59 ±4.78	0.30

Table 8. Comparison of clinical and cephalometric parameters between vertical ramus osteotomy and costochondral graft; n: Number of patients; SD: Standard Deviation.

Variation of clinical and radiological parameters between last follow-up and postoperative time have been correlated to different endpoints (figure 5). It seemed to exist a correlation between the soft-tissue statute and the trend for the deformity to relapse, particularly regarding the maxillary occlusal canting SO'/SO (r^2=0.11; p=0.059). In the same way, a non significant correlation was found between the Pruzansky score and the recurrence of the mandibular deformity measured on the SO'/SO ratio (r^2=0.06; p=0.19). Age of primary surgery seemed to influence the stability of the results regarding the esthetical results; an early mandibular surgery before the age of 13 years being significantly correlated with the recurrence of lip commissural line tilting (p=0.03). No correlation was found between age of surgery and the radiological parameters.

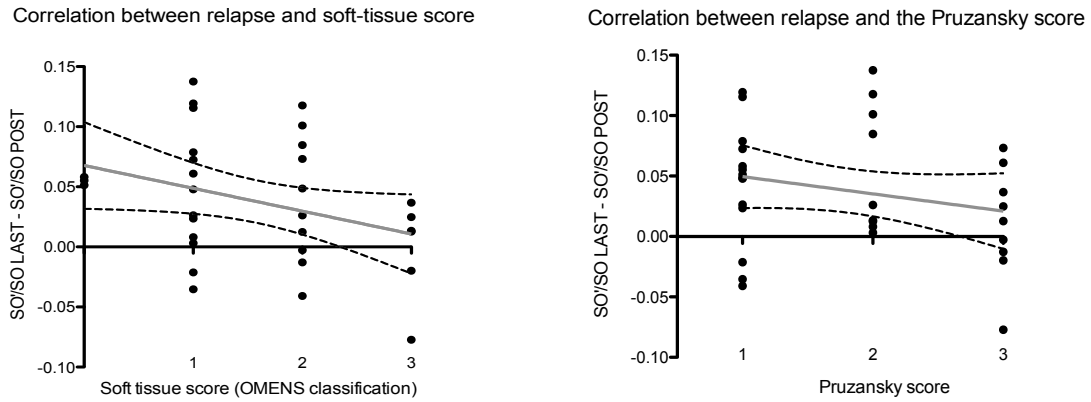


Figure 5. Linear regression between the relapse in radiological maxillary occlusal canting (ratio SO'/SO) and the OMENS soft tissue score or the Pruzansky score.

Surgical complications:

A temporary hypoesthesia in the inferior alveolar nerve territory was noted in 5 cases (12.5%), consecutive to CCG (n=4), or VRO (n=1). Two patients (5.0%) presented a facial nerve paresis after CCG, upon them, one presented with a permanent paresis in the lower labial branch territory. Two immediate post-operatory infections requiring a surgical drainage were reported (5.0%). One patient showed a hypertrophic scarring at the submandibular incision. A CCG overgrowth was observed in two cases (5.0%), only one needed a corrective secondary mandibular surgery. No complication was reported at the rib harvesting site, except a hypertrophic scarring in 1 patient.

Functional results:

Joint amplitudes values at the end of follow-up were known for 17 patients. Majority of subjects (82.4%) had considered normal mouth opening (Open bite ≥35mm), with a mean opening of 36.9 ±6.4mm. Others articular movements are shown in table 8.

Articular movements, end of follow-up	
MMO ≥35mm, n(%)	14 (82.4)
MMO (mm), mean ± S.D. (range)	36.9 ±6.4 (25-48)
Controlateral diduction (mm), mean ± S.D. (range)	8.7 ±2.2 (4-10)
Homolateral diduction (mm), mean ± S.D. (range)	9.5 ±1.1 (7-10)

Table 8. Articular movements analysis; n: Number of patients; MMO: Maximal Mouth Opening; SD: Standard Deviation.

DISCUSSION

Restoration of a facial symmetry in HFM remains challenging for cranio-maxillo-facial surgeons. Whatever the surgical procedure and planning used, it is essential to first restore the skeletal support of the face in a normal configuration (37) to adopt a normal shape of the soft tissues (38). As recommended by most of authors (19,25,33), we argue for an early correction of the mandibular deformity in childhood to repair the “functional matrix” (39) of the mandible and to allow a symmetric facial growth. A primary mandibular surgery proposed in mixed dentition between 9 and 12 years is interesting as the first molars have erupted and can be used to stabilize the orthodontic device; furthermore the mandibular growth is less active at this stage. Although the age of primary surgery is still debated according to the progressive nature of the deformity, the psychosocial dimension of these children has to be considered before delay the surgery until the completion of growth.

We have presented our experience with 39 growing as well as skeletally mature MHF operated patients. A significant right predominance of the disease was noted with a 2/3 ratio, as commonly described in the literature (5). All the patients studied were grade II or III of the Pruzansky-Kaban modified classification. For type IIA patients, VRO was preferred to distraction osteogenesis (DO), that represents an increasingly popular alternative to this procedure; a recent meta-analysis reporting a 40.3% rate of DO procedures in patients with type I and II HFM (21). VRO was chosen for several reasons. At first, it allows a one-stage correction of the posterior vertical insufficiency (PVI) with consequent lengthening of the mandibular ramus. A recent unpublished work in 35 patients with unilateral PVI reported a mean 15.4mm mandibular elongation after VRO procedure with stable results at one year postoperatively (40). Moreover, VRO represents a safe procedure but raises the issue of external approach. Therefore an intraoral strategy is commonly used by various teams to prevent any cutaneous or nervous damage (41,42). Despite substantial advantages and recent improvement of DO (intra-oral distractor, minimally invasive surgery, absence of donor site morbidity and hypothetical expansion of soft tissue) (43–46), a brief literature analysis shows a lack of long-term data on efficacy (15,47,48). In addition a 90 percent rate of long-term relapse of the deformity has been reported after DO requiring further procedures or orthognathic revision (49). For types IIB and III, patients were given a CCG as commonly described in the literature (21). Padwa *et al.* were the first to document long-term

midfacial growth after CCG in 33 children with types IIB and III HFM (33). They showed that a good long-term result could be obtained in patients operated at old age and presenting with a low OMENS score. To our knowledge, there are very few papers relating the long term efficacy of CCG as a primary mandibular surgery in HFM patients (21,26).

Our mean follow-up time (6.8 years after primary surgery) is one of the most important reported in the literature (21). This follow up duration allow us to affirm that a significant and durable improvement of the maxillary occlusal canting and the clinical chin deviation could be obtained after a one stage procedure. While a trend to the recurrence of the deformity was noted for some parameters, most of the results remained satisfactory. It is now established that the constructed ramus does not grow as well as the normal side and leads to slight to moderate relapse (20,50,51). Wan *et al.* described a 56% rate of undergrowth with CCG in series of 31 Pruzansky III graded patients followed-up for more than 13 years (52). Sagittal osteotomy or DO on the constructed CCG can be proposed for the late correction of this undergrowth (52). Aesthetical and cephalometric parameters have been compared in our study between VRO and CCG primary procedures. Despite a more severe deformation at the preoperative time in CCG group, there was no difference at the end of growth suggesting a same growth potential of the constructed ramus with both processes. We believe that the costochondral grafting in childhood in patients with HFM allows to create a functional ramus with a cranial support facilitating a secondary procedure (53). The glenoid fossa can be missing in severe deformities making difficult to obtain this cranial support (54) and leading to internal or upward shifts of the graft.

Thanks to the mandibular lengthening, the normalisation of the maxillary cant, reflected by the increase of the SO'/SO ratio, could be performed in most of patients. This reminds again the remarkable vertical growing potential of the maxillary dento-alveolar portion as a response to the generated open-bite (55). These results highlight also the importance of the maxilla-mandibular elastic therapy consisting in nocturnal fixation and of the use of a bite occlusal splint to allow a slow vertical movement of the maxilla. Perfect symmetry in the occlusal canting could not be obtained since the SO'/SO ratio remained inferior to 1 at the end of growth, which not necessarily leaded to a late correction; a slight occlusal cant tilting can be misrecognized by the majority of observers (32).

A concomitant Le Fort I osteotomy was performed in ten patients in our series permitting an immediate improvement of the facial appearance. This approach has to

be reserved to adolescent or adult populations (4) when an inter-incisors point deviation has to be corrected and when the maxillary growth potential is less effective to correct the vertical dimension. In the same way a genioplasty was associated to the ramus reconstruction in 56% of cases with a good esthetical impact in the patient; that was only possible because of the delayed primary surgery to 10-12 years of age when the permanent mandibular canine has started its eruption. In majority of cases, a sagittal split osteotomy was realized on the contralateral side from the age of 11 to carry out a derotating osteotomy of the mandibular ramus.

In the light of the surgical complications reported in this study, VRO and CCG represent safe procedures. Furthermore, a normal mouth opening was obtained in 82.4% of patients. As TMJ ankylosis is described after CCG reconstruction of the mandibular ramus (50,56), we did not report this complication.

Nine patients (23%) received an orthognathic revision at a later stage after completion of growth with a mean age of 17.4 years. There is no data found in the literature about the number of mandibular procedures required in HFM. A recent systematic review of mandibular reconstruction in the growing patient with HFM reported a secondary procedure needed in 18 patients on 285 analysed, which is probably largely underestimated (21). Wan et al. reported an orthognathic correction after CCG in 13 patients on 16 having reached skeletal maturity (52).

The main drawback of our study is the great heterogeneity of the studied population in terms of severity of the mandibular hypoplasia, soft tissue status and the age of primary surgery. We used standardized PA cephalograms for our analysis for more reliability in measuring distances (57). The supraorbital line was selected as horizontal reference since these points are not altered during surgery, while the vertical line from the crista galli represented the best approximation of the midline (58). The SO'/SO ratio reflecting the maxillary occlusal cant and the mandibular offset was used to eliminate the variability factors between radiographs. A three-dimensional analysis would be more relevant to assess the cephalometric changes but was not realized because of the long inclusion period and because of the radiation generated.

Despite the absence in our study of significant correlation between the soft tissue statute and the relapse of the deformity, it is clearly established that hypoplasia of the subcutaneous and muscular tissues as well as the VIIth nerve involvement are responsible for less favourable response to the primary mandibular surgery (33,59,60). Likewise by the contralateral displacement of the lower jaw and the soft

tissues pulling on the mandible, the bone surgery can accentuate the observed deficiency of soft tissues on the affected side (19). Osteocutaneous free flap (61,62) and in the near future tissue engineering technique (63) would represent interesting alternatives for secondary reconstruction of severe deformities in HFM.

If the CCG represents the gold standard for repair of the hypoplastic ramus in types III HFM, there is no consensus for types II. Many surgical procedures have been described and can be combined in patients (DO, VRO, SSO, inverted-L osteotomy). More study with standardized measurements are needed to compare DO and VRO and to assess their efficacy at the end of growth in patients with hemifacial microsomia.

CONCLUSION

Costochondral graft and vertical ramus osteotomy remain interesting and safe procedures for the primary mandibular reconstruction in children with types II and III hemifacial microsomia. We describe good aesthetical and cephalometric results at the end of growth after a one stage correction. A secondary orthognathic procedure was needed in 23% of patients after completion of growth because of a trend to relapse of the chin deviation and maxillary occlusal canting, probably as a consequence of the soft tissue hypoplasia.

DISCUSSION

La restauration d'une symétrie faciale dans les MHF demeure difficile. Quels que soient les procédures et le planning chirurgical employés, il est essentiel de restaurer en premier lieu le squelette facial dans une configuration normale (37) avant d'envisager la correction des tissus mous (38). Comme recommandé par la plupart des auteurs (19,25,33), nous prônons une correction précoce de la malformation mandibulaire dans l'enfance pour rendre à la mandibule sa « matrice fonctionnelle » (39) et ainsi permettre une croissance faciale symétrique. Ainsi nous proposons une chirurgie primaire mandibulaire en denture mixte entre 9 et 12 ans, après l'éruption des premières molaires afin de stabiliser l'appareillage orthodontique ; de plus la croissance mandibulaire est moins active durant cette période. Le caractère progressif de la malformation et l'âge de la chirurgie primaire demeurent très débattus à l'heure actuelle, mais la dimension psychosociale chez ces enfants atteints doit être considérée avant de reporter l'intervention primaire en fin de croissance.

Nous rapportons notre expérience de 39 patients atteints de MHF opérés pendant l'enfance et en fin de croissance. Une prédominance droite de l'atteinte a été rapportée avec un ratio de 2/3, comme couramment décrit dans la littérature (5). Tous les patients étudiés présentaient une atteinte de grade II ou III de la classification de Pruzansky-Kaban. Dans les types IIA, une OVRS a été pratiquée préférentiellement par rapport à la DO, intervention en plein essor ; une méta-analyse récente décrivant 40,3% de procédures de distraction chez des patients porteurs de grades I et II de MHF (21). L'OVRS constitue à notre sens une procédure de choix pour plusieurs raisons. Tout d'abord elle permet une correction en une fois de l'insuffisance verticale postérieure (IVP) par l'allongement du ramus mandibulaire. Un travail récent non publié réalisé dans notre service chez 35 patients atteints d'IVP unilatérale a montré un allongement moyen du ramus de 15,4mm et un maintien du résultat à un an après la chirurgie (40). Par ailleurs l'OVRS est une procédure sûre mais pose le problème de la voie cutanée externe pouvant conduire à des cicatrices inesthétiques et à des atteintes du rameau mentonnier du nerf facial. De nombreuses équipes utilisent ainsi une approche endo-buccale pour éviter ces complications liées à la voie d'abord (41,42). Malgré des avantages non négligeables de la DO et l'amélioration récente des dispositifs (distracteur intra-oral, chirurgie mini-invasive, absence de morbidité de site donneur et expansion des parties molles) (43–46), il

existe un manque de données sur son efficacité en fin de croissance (15,47,48). Un taux de 90% d'échecs à long-terme a été avancé avec la récidive de la déformation conduisant à la réalisation de procédures de distraction additionnelles ou à une chirurgie orthognathique conventionnelle (49). Une GCC a été réalisée majoritairement dans les grades IIB et III (21). Padwa *et al.* ont été les premiers à rapporter les résultats tardifs de la croissance faciale après GCC chez 33 patients atteints de MHF grades IIB et III (33). Ils ont ainsi montré de bons résultats à long-terme sur la symétrie faciale d'autant plus que les patients étaient opérés tardivement et qu'ils présentaient un faible score OMENS. A notre connaissance, il existe peu d'articles traitant des résultats en fin de croissance de la chirurgie primaire mandibulaire chez des enfant opérés de MHF (21,26).

Notre suivi moyen atteignant 6,8 ans après la chirurgie primaire est un des plus importants décrits dans la littérature (21). Cela nous permet d'affirmer qu'une amélioration significative et durable du plan d'occlusion du maxillaire et de la déviation clinique du point menton a pu être obtenue après une seule intervention. Malgré une tendance à la dégradation de la symétrie faciale observée dans l'analyse des paramètres, les résultats demeurent satisfaisants. Il a été clairement établi que la croissance du ramus reconstruit demeure inférieure à celle du côté sain conduisant à la récidive de la déformation (20,50,51). Wan *et al.* ont décrit un taux de 56% d'hypo-croissance costale dans une série de 31 patients atteints de MHF grade III et suivis plus de 13 ans après une GCC (52). L'ostéotomie sagittale de la branche montante (OSBM) ou la DO du greffon chondro-costal (52) représentent des alternatives intéressantes pour la correction tardive de l'hypocroissance. Nous avons comparé les paramètres esthétiques et céphalométriques entre les groupes opérés d'une GCC et d'une OVRS mettant en évidence une déformation initiale plus sévère dans le groupe de patients ayant reçu un greffon costal. En revanche il n'existait aucune différence statistiquement significative en fin de croissance suggérant un potentiel de croissance identique entre la GCC et l'OVRS. Nous pensons que la GCC chez l'enfant porteur de MHF permet de créer un ramus fonctionnel avec un appui crânien qui facilite la réalisation d'une chirurgie secondaire (53). Par ailleurs la cavité glénoïde peut être absente dans les cas sévères de malformation rendant difficile l'obtention de cet appui crânien (54) et pouvant conduire à la fuite vers le haut ou en interne du greffon.

Grace à l'allongement chirurgical de la branche montante, la normalisation du plan d'occlusion maxillaire reflétée par l'augmentation du rapport SO'/SO a pu être

obtenue chez la plupart des patients. Ceci rappelle l'incroyable potentiel de croissance verticale dentoalvéolaire du maxillaire en réponse à l'infraclusie postérieure créée (55). Ces données soulignent également l'importance du blocage élastique maxillo-mandibulaire nocturne et l'utilisation d'une gouttière occlusale progressivement réduite sagittalement pour permettre l'égression lente des dents maxillaires. L'horizontalisation complète du plan d'occlusion maxillaire n'a pas pu être obtenue en fin de croissance sur le plan céphalométrique puisque le rapport SO'/SO est resté inférieur à 1. La persistance d'une inclinaison légère n'a pas nécessairement conduit à une chirurgie tardive dans la mesure où celle-ci demeure le plus souvent imperceptible sur le plan clinique (32).

Une ostéotomie de Lefort I concomitante à la chirurgie mandibulaire a été réalisée chez 10 patients conduisant à une amélioration immédiate de la symétrie faciale. La chirurgie du maxillaire est néanmoins réservée à l'adolescent ou à l'adulte (4) lorsque la déviation du point inter-incisif médian doit être corrigée et lorsque le potentiel de croissance attendu du maxillaire n'est pas suffisant pour corriger la dimension verticale. Une génioplastie d'avancée et de recentrage a pu être réalisée dans 56% des cas avec un fort impact esthétique chez ces patients ; ce geste n'étant rendu possible que par le report de l'intervention primaire après l'âge de 9 ans lorsque les canines permanentes mandibulaires ont fait leur éruption. Chez la majorité des patients étudiés, une OSBM controlatérale de dérotation mandibulaire a été effectuée dès l'âge de 11 ans.

A la lumière du faible taux de complications rapporté dans cette étude, l'OVRS et la GCC représentent des interventions sûres pour la reconstruction du ramus hypoplasique des MHF. De plus une ouverture buccale considérée comme normale (>35mm) a été obtenue chez 82,4% des patients. Des cas d'ankylose temporo-mandibulaire après GCC ont été décrits dans la littérature (50,56) mais aucun n'a été rapporté dans ce travail.

Neuf patients (23%) ont bénéficié d'une chirurgie mandibulaire secondaire en fin de croissance avec un âge moyen d'intervention de 17,4 ans. Il existe peu de données dans la littérature sur le nombre moyen d'interventions chirurgicales mandibulaires chez les patients atteints de MHF. Une étude récente de la reconstruction mandibulaire dans les MHF rapportait la réalisation de 18 chirurgies secondaires chez les 285 patients analysés (21), nombre probablement sous-estimé. Wan *et al.* ont rapporté le recours à une intervention secondaire en fin de croissance après GCC chez 13 des 16 patients étudiés (52).

Le principal défaut de notre étude est la grande hétérogénéité de la population étudiée en termes de sévérité de l'hypoplasie mandibulaire, de l'atteinte des tissus mous et de l'âge de la chirurgie primaire. Les analyses de distances ont été effectuées sur des téléradiographies de face standardisées pour une meilleure fiabilité des mesures (57). La ligne supra-orbitaire a été sélectionnée comme ligne horizontale de référence puisque les points qui la constituent restent constants durant la chirurgie ; la ligne de référence verticale passant par le processus crista-galli est quant à elle la meilleure approximation de l'axe médian de la face (58). Le rapport SO'/SO qui reflète le plan d'occlusion du maxillaire et le décalage mandibulaire a été utilisé pour éliminer la variabilité entre les clichés téléradiographiques. Une analyse céphalométrique tridimensionnelle serait plus pertinente en terme de précision des mesures et d'étude de la croissance dans les trois dimensions, mais n'a pas pu être réalisée compte tenu de la longue période d'inclusion des patients et de l'irradiation qu'elle génère.

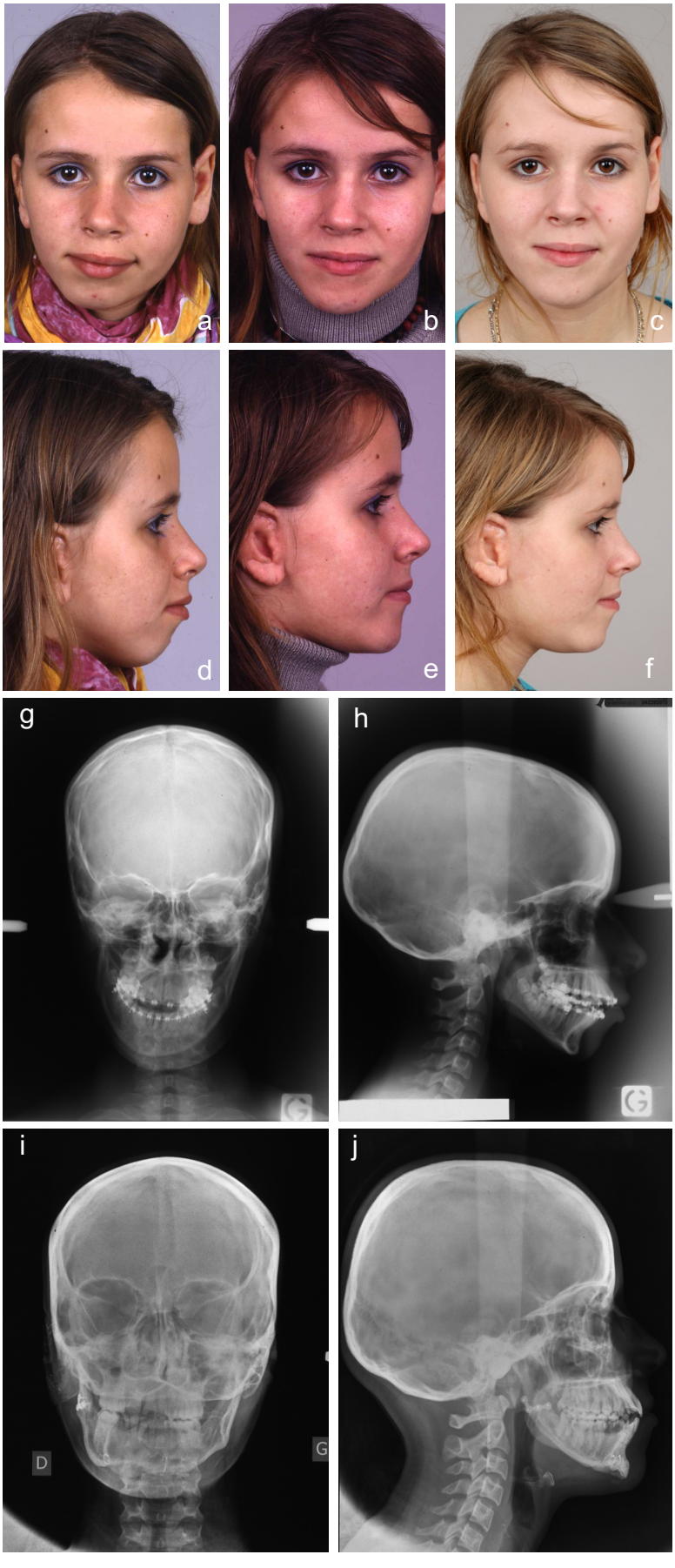
Nous n'avons pas mis en évidence de corrélation statistiquement significative entre les tissus mous et la tendance à la dégradation du résultat chirurgical. Il est tout de même clairement établi que l'hypoplasie des tissus musculaires et sous-cutanés de la face, de même que l'atteinte du nerf facial, participent à la récidive lente de la malformation (33,59,60). Par ailleurs, de par le déplacement de la mandibule et des tissus mous insérés sur celle-ci, la chirurgie mandibulaire peut accentuer le déficit en parties molles du côté atteint (19). Les lambeaux libres ostéo-cutanés (61,62) et dans un futur proche les techniques d'ingénierie régénérative (63) représentent des alternatives intéressantes pour la reconstruction secondaire des déformations sévères dans les MHF.

Si la GCC représente la technique de choix pour la réparation du ramus hypoplasique dans les grades III, il n'existe aucun consensus pour les types II de Pruzansky. De nombreuses procédures chirurgicales ont été décrites et peuvent être successivement utilisées chez les patients (DO, OVRS, OSBM, ostéotomie en L inversé). Des études complémentaires avec des critères d'évaluation standardisés sont nécessaires pour comparer la DO et l'OVRS en terme de résultats en fin de croissance chez les patients porteurs de microsomie hémi-faciale.

CONCLUSION

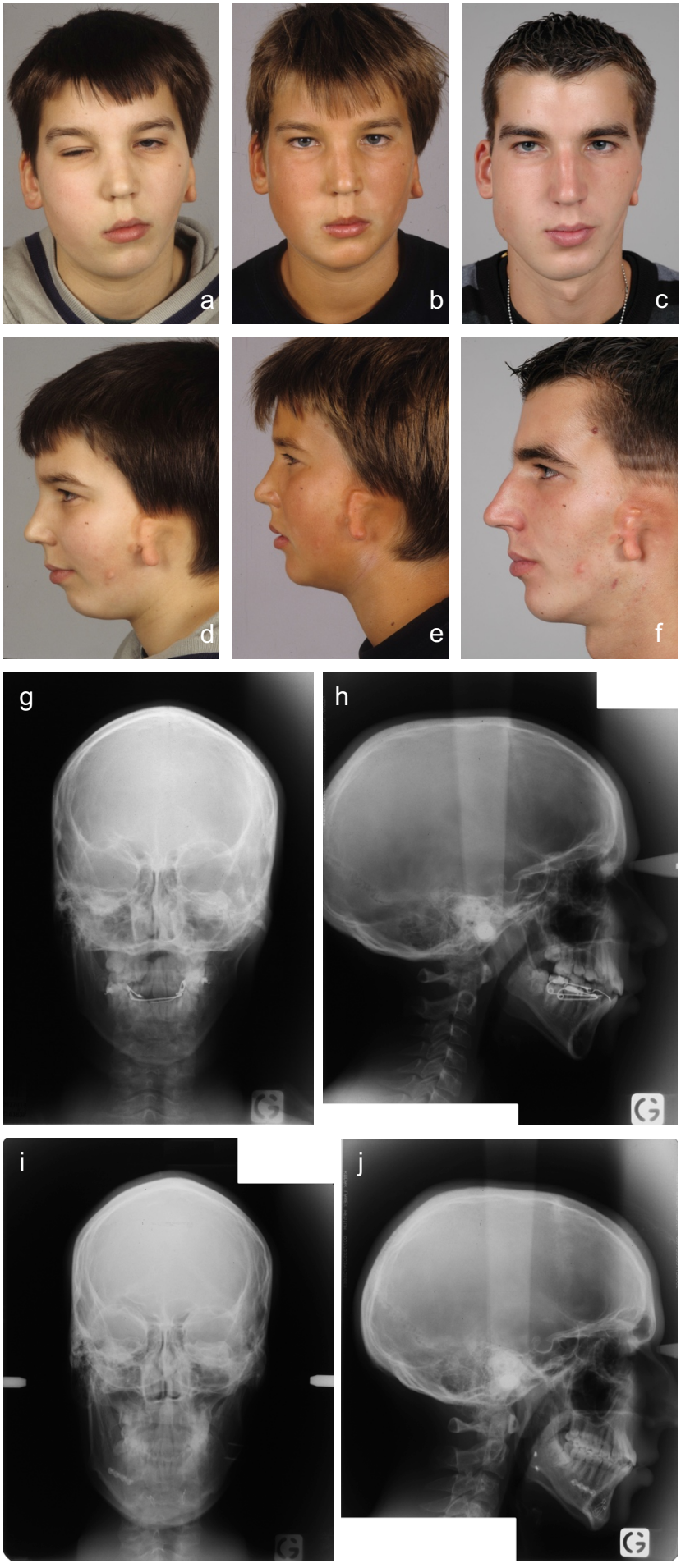
La greffe chondro-costale et l'ostéotomie rétrospigienne d'allongement du ramus mandibulaire demeurent des procédures efficaces et sûres pour la reconstruction mandibulaire primaire chez les enfants porteurs de microsomie hémi-faciale de grade II et III. Nous décrivons des résultats esthétiques et céphalométriques satisfaisants en fin de croissance après une seule intervention mandibulaire chez la majorité des patients. Une intervention de chirurgie orthognathique secondaire a été nécessaire dans 23% des cas. La dégradation des résultats a concerné la déviation clinique du menton et le plan d'occlusion du maxillaire, et représente probablement la conséquence de l'hypoplasie des tissus mous faciaux.

CASUISTIQUE



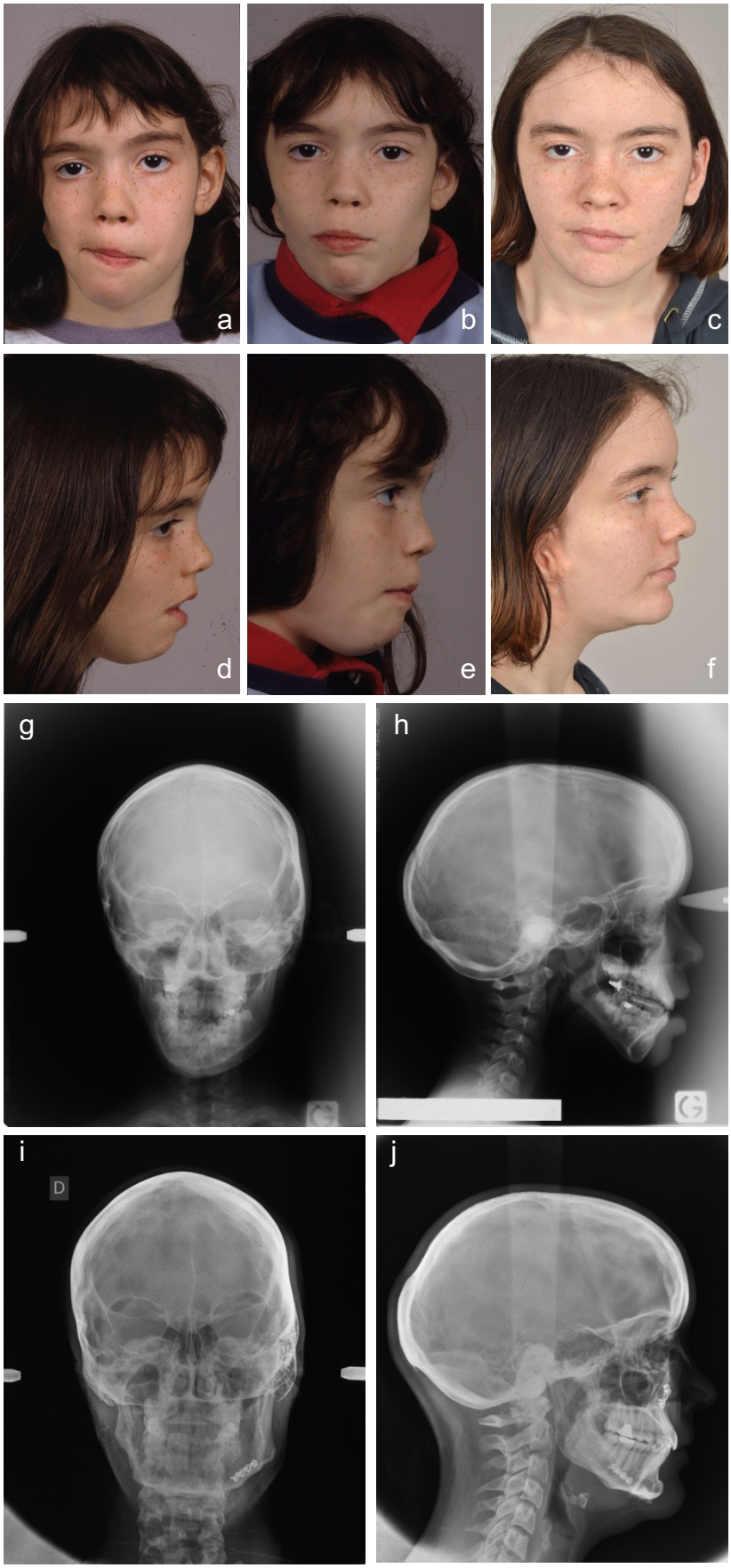
CAS NUMERO 1

Patiente porteuse d'une microsomie hémi-faciale droite de type IIA opérée à l'âge de 14 ans d'une OVRS droite associée à une OSBM gauche de dérotation et génioplastie. Suivi de 92 mois. Photographies de face et de profil en période préopératoire (a, d), postopératoire (b, e) et lors du dernier suivi (c, f). Téléradiographies de face et de profil en période préopératoire (g, h) et lors du dernier suivi (i, j).
Source : Pr Mercier.



CAS NUMERO 2

Patient porteur d'une microsomie hémi-faciale gauche de type IIB, opérée à l'âge de 11 ans d'une GCC gauche associée à une OSBM droite de dérotation et à une génioplastie. Suivi de 64 mois. Photographies de face et de profil en période préopératoire (a, d), postopératoire (b, e) et lors du dernier suivi (c, f). Téléradiographies de face et de profil en période préopératoire (g, h) et lors du dernier suivi (i, j).
Source : Pr Mercier.



CAS NUMERO 3

Patiente porteuse d'une microsomie hémi-faciale droite de type III, opérée à l'âge de 8 ans d'une GCC droite. Chirurgie secondaire réalisée à l'âge de 15 ans consistant en OSBM gauche associée à une ostéotomie de Lefort I d'horizontalisation et de recentrage et à une génioplastie. Suivi de 120 mois. Photographies de face et de profil en période préopératoire (a, d), postopératoire après chirurgie primaire (b, e) et lors du dernier suivi (c, f). Téléradiographies de face et de profil en période préopératoire (g, h) et lors du dernier suivi (i, j).
Source : Pr Mercier.

BIBLIOGRAPHIE

1. Barisic I, Odak L, Loane M, Garne E, Wellesley D, Calzolari E, et al. Prevalence, prenatal diagnosis and clinical features of oculo-auriculo-vertebral spectrum: a registry-based study in Europe. Eur J Hum Genet. août 2014;22(8):1026‑33.
2. Bettega G, Morand B, Lebeau J, Raphaël B. Morphological alterations of oto-mandibular syndromes. Ann Chir Plast Esthét. oct 2001;46(5):495‑506.
3. Cousley RR, Calvert ML. Current concepts in the understanding and management of hemifacial microsomia. Br J Plast Surg. oct 1997;50(7):536‑51.
4. Converse JM, Coccaro PJ, Becker M, Wood-Smith D. On hemifacial microsomia. The first and second branchial arch syndrome. Plast Reconstr Surg. mars 1973;51(3):268‑79.
5. Burglen L, Soupre V, Diner PA, Gonzalès M, Vazquez MP. Oto-mandibular dysplasias: genetics and nomenclature of syndromes. Ann Chir Plast Esthét. oct 2001;46(5):400‑9.
6. Grabb WC. The first and second branchial arch syndrome. Plast Reconstr Surg. nov 1965;36(5):485‑508.
7. Gorlin R, Cohen MJ, Levin L. Branchial arch and oro-acral disorders. In: Syndromes of the Head and Neck (3rd ed). Oxford University Press; 1990. p. 641‑9.
8. Charrier JB, Bennaceur S, Couly G. Hemifacial microsomia. Embryological and clinical approach. Ann Chir Plast Esthét. oct 2001;46(5):385‑99.
9. Madrid JRP, Montealegre G, Gomez V. A New Classification Based on the Kaban's Modification for Surgical Management of Craniofacial Microsomia. Craniomaxillofacial Trauma Reconstr. mars 2010;3(1):1‑7.
10. David DJ, Mahatumarat C, Cooter RD. Hemifacial microsomia: a multisystem classification. Plast Reconstr Surg. oct 1987;80(4):525‑35.
11. Pruzansky S. Not all dwarfed mandibles are alike. In: Birth Defects original Article Ser V,2. 1969. p. 120‑9.
12. Kaban LB, Moses MH, Mulliken JB. Surgical correction of hemifacial microsomia in the growing child. Plast Reconstr Surg. juill 1988;82(1):9‑19.
13. Vento AR, LaBrie RA, Mulliken JB. The O.M.E.N.S. classification of hemifacial microsomia. Cleft Palate-Craniofacial J Off Publ Am Cleft Palate-Craniofacial Assoc. janv 1991;28(1):68‑76; discussion 77.
14. Birgfeld CB, Heike C. Craniofacial microsomia. Semin Plast Surg. mai 2012;26(2):91‑104.
15. Meazzini MC, Mazzoleni F, Bozzetti A, Brusati R. Comparison of mandibular vertical growth in hemifacial microsomia patients treated with early distraction or not treated: follow up till the completion of growth. J Cranio-Maxillo-fac Surg Off Publ Eur Assoc Cranio-Maxillo-fac Surg. févr 2012;40(2):105‑11.
16. Polley JW, Figueroa AA, Liou EJ, Cohen M. Longitudinal analysis of mandibular asymmetry in hemifacial microsomia. Plast Reconstr Surg. févr 1997;99(2):328‑39.
17. Rune B, Selvik G, Sarnäs KV, Jacobsson S. Growth in hemifacial microsomia studied with the

aid of roentgen stereophotogrammetry and metallic implants. Cleft Palate J. avr 1981;18(2):128‑46.

18. Kearns GJ, Padwa BL, Mulliken JB, Kaban LB. Progression of facial asymmetry in hemifacial microsomia. Plast Reconstr Surg. févr 2000;105(2):492‑8.

19. Kaban LB, Padwa BL, Mulliken JB. Surgical correction of mandibular hypoplasia in hemifacial microsomia: the case for treatment in early childhood. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. mai 1998;56(5):628‑38.

20. Ross RB. Costochondral grafts replacing the mandibular condyle. Cleft Palate-Craniofacial J Off Publ Am Cleft Palate-Craniofacial Assoc. juill 1999;36(4):334‑9.

21. Pluijmers BI, Caron CJJM, Dunaway DJ, Wolvius EB, Koudstaal MJ. Mandibular reconstruction in the growing patient with unilateral craniofacial microsomia: a systematic review. Int J Oral Maxillofac Surg. mars 2014;43(3):286‑95.

22. Suh J, Choi TH, Baek S-H, Kim JC, Kim S. Mandibular distraction in unilateral craniofacial microsomia: longitudinal results until the completion of growth. Plast Reconstr Surg. nov 2013;132(5):1244‑52.

23. Posnick JC. Hemifacial Microsomia: Evaluation and Treatment. In: Orthognathic Surgery. St. Louis: W.B. Saunders; 2014. p. 1095‑158.

24. Vargervik K, Ousterhout DK, Farias M. Factors affecting long-term results in hemifacial microsomia. Cleft Palate J. déc 1986;23 Suppl 1:53‑68.

25. Mulliken JB, Ferraro NF, Vento AR. A retrospective analysis of growth of the constructed condyle-ramus in children with hemifacial microsomia. Cleft Palate J. oct 1989;26(4):312‑7.

26. Kumar P, Rattan V, Rai S. Do costochondral grafts have any growth potential in temporomandibular joint surgery? A systematic review. J Oral Biol Craniofacial Res. déc 2015;5(3):198‑202.

27. Cousley RR. A comparison of two classification systems for hemifacial microsomia. Br J Oral Maxillofac Surg. avr 1993;31(2):78‑82.

28. Posnick JC. 28 - Hemifacial Microsomia: Evaluation and Treatment. In: Orthognathic Surgery. St. Louis: W.B. Saunders; 2014. p. 1095‑158.

29. Caldwell JB, Letterman GS. Vertical osteotomy in the mandibular rami for correction of prognathism. J Oral Surg. juill 1954;12(3):185‑202.

30. Delaire J, Le Roux J, Tulasne JF. Functional treatment of fractures of the mandibular condyle and its neck. Rev Stomatol Chir Maxillofac. juin 1975;76(4):331‑50.

31. Mercier J, Huet P, Perrin JP. Functional management of fractures of the mandibular condyle. Rev Stomatol Chir Maxillofac. oct 2000;101(4):203‑6.

32. Padwa BL, Kaiser MO, Kaban LB. Occlusal cant in the frontal plane as a reflection of facial asymmetry. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. août 1997;55(8):811‑816; discussion 817.

33. Padwa BL, Mulliken JB, Maghen A, Kaban LB. Midfacial growth after costochondral graft construction of the mandibular ramus in hemifacial microsomia. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. févr 1998;56(2):122-127-128.

34. Lippold C. Treatment of hemimandibular hyperplasia: The biological basis of condylectomy.

35. Dahlberg G. Statistical Methods for Medical and Biological Students. Br Med J. 14 sept 1940;2(4158):358‑9.

36. Kamoen A, Dermaut L, Verbeeck R. The clinical significance of error measurement in the interpretation of treatment results. Eur J Orthod. oct 2001;23(5):569‑78.

37. Ortiz-Monasterio F. Early mandibular and maxillary osteotomies for the correction of hemifacial microsomia. A preliminary report. Clin Plast Surg. oct 1982;9(4):509‑17.

38. Munro IR. One-stage reconstruction of the temporomandibular joint in hemifacial microsomia. Plast Reconstr Surg. nov 1980;66(5):699‑710.

39. Poswillo D. The pathogenesis of the first and second branchial arch syndrome. Oral Surg Oral Med Oral Pathol. mars 1973;35(3):302‑28.

40. Leveau S. Evaluation de l'ostéotomie verticale rétrospigienne d'allongement de la branche montante de la mandibule dans les insuffisances verticales postérieures unilatérales. Thèse pour le diplôme d'état de Docteur en Médecine. Nantes; 2009.

41. Ghali GE, Sikes JW. Intraoral vertical ramus osteotomy as the preferred treatment for mandibular prognathism. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. mars 2000;58(3):313‑5.

42. Choi YJ, Ha Y-D, Lim H, Huh J-K, Chung CJ, Kim K-H. Long-term changes in mandibular and facial widths after mandibular setback surgery using intraoral vertical ramus osteotomy. Int J Oral Maxillofac Surg. 2 mai 2016;

43. Baek S-H, Kim S. The determinants of successful distraction osteogenesis of the mandible in hemifacial microsomia from longitudinal results. J Craniofac Surg. juill 2005;16(4):549‑58.

44. McCarthy JG, Katzen JT, Hopper R, Grayson BH. The first decade of mandibular distraction: lessons we have learned. Plast Reconstr Surg. déc 2002;110(7):1704‑13.

45. McCarthy JG, Stelnicki EJ, Mehrara BJ, Longaker MT. Distraction osteogenesis of the craniofacial skeleton. Plast Reconstr Surg. juin 2001;107(7):1812‑27.

46. Molina F. Mandibular elongation and remodeling by gradual distraction. An experience of 277 cases. Ann Chir Plast Esthét. oct 2001;46(5):507‑15.

47. Meazzini MC, Mazzoleni F, Gabriele C, Bozzetti A. Mandibular distraction osteogenesis in hemifacial microsomia: long-term follow-up. J Cranio-Maxillo-fac Surg Off Publ Eur Assoc Cranio-Maxillo-fac Surg. déc 2005;33(6):370‑6.

48. Bartlett SP. No evidence for long-term effectiveness of early osteodistraction in hemifacial microsomia. Plast Reconstr Surg. mai 2010;125(5):1567‑8.

49. Ascenço ASK, Balbinot P, Junior IM, D'Oro U, Busato L, da Silva Freitas R. Mandibular distraction in hemifacial microsomia is not a permanent treatment: a long-term evaluation. J Craniofac Surg. mars 2014;25(2):352‑4.

50. Munro IR, Phillips JH, Griffin G. Growth after construction of the temporomandibular joint in children with hemifacial microsomia. Cleft Palate J. oct 1989;26(4):303‑11.

51. Perrott DH, Umeda H, Kaban LB. Costochondral graft construction/reconstruction of the ramus/condyle unit: long-term follow-up. Int J Oral Maxillofac Surg. déc 1994;23(6 Pt 1):321‑8.

52. Wan DC, Taub PJ, Allam KA, Perry A, Tabit CJ, Kawamoto HK, et al. Distraction osteogenesis

of costocartilaginous rib grafts and treatment algorithm for severely hypoplastic mandibles. Plast Reconstr Surg. mai 2011;127(5):2005‑13.

53. Mercier J-M, Perrin J-P, Longis J, Arzul L, Corre P. Facial asymmetries and their skeletal component. Rev Stomatol Chir Maxillo-Faciale Chir Orale. sept 2014;115(4):219‑28.

54. Tulasne JF. Oto-mandibular dysplasias: indications for conventional osteotomies and costochondral graft. Ann Chir Plast Esthét. oct 2001;46(5):527‑37.

55. Mercier J, Gordeeff A, Delaire J. Syndrome of unilateral posterior vertical insufficiency of the face. Clinical and therapeutic aspects. Acta Stomatol Belg. juin 1989;86(1):13‑32.

56. Saeed NR, Kent JN. A retrospective study of the costochondral graft in TMJ reconstruction. Int J Oral Maxillofac Surg. déc 2003;32(6):606‑9.

57. Kaban LB, Mulliken JB, Murray JE. Three-dimensional approach to analysis and treatment of hemifacial microsomia. Cleft Palate J. avr 1981;18(2):90‑9.

58. Fattah AY, Caro C, Khechoyan DY, Tompson B, Forrest CR, Phillips JH. Cephalometric outcomes of orthognathic surgery in hemifacial microsomia. J Craniofac Surg. sept 2014;25(5):1734‑9.

59. Huisinga-Fischer CE, Vaandrager JM, Prahl-Andersen B. Longitudinal results of mandibular distraction osteogenesis in hemifacial microsomia. J Craniofac Surg. nov 2003;14(6):924‑33.

60. Scolozzi P, Herzog G, Jaques B. Simultaneous maxillo-mandibular distraction osteogenesis in hemifacial microsomia: a new technique using two distractors. Plast Reconstr Surg. 15 avr 2006;117(5):1530‑1541; discussion 1542.

61. Santamaría E, Morales C, Taylor JA, Hay A, Ortiz-Monasterio F. Mandibular microsurgical reconstruction in patients with hemifacial microsomia. Plast Reconstr Surg. déc 2008;122(6):1839‑49.

62. Warren SM, Borud LJ, Brecht LE, Longaker MT, Siebert JW. Microvascular reconstruction of the pediatric mandible. Plast Reconstr Surg. févr 2007;119(2):649‑61.

63. Henkel J, Woodruff MA, Epari DR, Steck R, Glatt V, Dickinson IC, et al. Bone Regeneration Based on Tissue Engineering Conceptions - A 21st Century Perspective. Bone Res. sept 2013;1(3):216‑48.

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Correction chirurgicale de l'hypoplasie mandibulaire dans les microsomies hémi-faciales : Une étude rétrospective de 39 cas

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RESUME

Introduction : La microsomie hémifaciale (MHF) est une anomalie du développement touchant les structures cervico-faciales dérivant des premier et deuxième arcs branchiaux. La prise en charge chirurgicale de l'hypoplasie mandibulaire demeure controversée et il existe peu de données dans la littérature en ce qui concerne les résultats en fin de croissance. L'objectif de notre étude était d'évaluer les résultats à long terme de la chirurgie primaire mandibulaire sur le plan esthétique, architectural et fonctionnel dans une série de jeunes patients atteints de MHF.

Méthodes : Trente-neuf patients atteints de MHF grades II et III de la classification de Pruzansky ont été rétrospectivement inclus dans cette étude. En fonction de la déformation mandibulaire les patients ont bénéficié d'une greffe chondro-costale (GCC) ou d'une ostéotomie verticale rétrospigienne d'allongement de la branche montante (OVRS). Une analyse des paramètres architecturaux et esthétiques a été réalisée à partir des clichés téléradiographiques et photographiques frontaux en préopératoire, postopératoire et en fin de suivi.

Résultats : L'âge moyen de la chirurgie primaire mandibulaire a été de de 13,1 ans ±3.5 ans (7-21) et le suivi moyen de 70.4 ±57.1 mois (0-252). Trente-trois patients (82.5%) ont été considérés comme ayant atteint la fin de leur croissance lors du dernier suivi. Une GCC a été pratiquée dans 24 cas principalement dans les types IIB et III tandis qu'une OVRS a été réalisée chez 15 patients majoritairement de grade IIA. L'analyse architecturale a montré la restauration d'un plan d'occlusion horizontal à la fin du suivi ($p<0.0001$) ; la déviation mentonnière a été immédiatement corrigée par la chirurgie mais une tendance à la récidive a été notée en fin de croissance ($p<0.0001$). L'analyse esthétique a révélé une amélioration significative de la déviation mentonnière et la correction de la ligne bi-commissurale ($p<0.0001$). Une corrélation non significative a été trouvée entre le degré d'atteinte des tissus mous et la tendance à la récidive du plan d'occlusion maxillaire ($p=0.06$). Vingt-trois pourcents des patients ont nécessité une seconde intervention de chirurgie orthognathique.

Conclusion : La CCG et l'OVRS sont des procédures chirurgicales sures et fiables permettant la correction en une fois de l'occlusion dentaire et de la croissance mandibulaire chez les jeunes patients atteints de MHF de grades II et III.

MOTS-CLES

Microsomie hémi-faciale, chirurgie primaire mandibulaire, greffe chondro-costale, Ostéotomie d'allongement vertical, croissance