Part 2: Is the maxillary canting and its surgical correction in patients with CFM correlated to the mandibular deformity?

Pluijmers, B. I.[a*], van de Lande, L. S.[a,b*], Caron, C. J. J. M.[a], Wolvius, E. B.[a], Dunaway, D. J.[c], Padwa, B. L.[b], & Koudstaal, M. J[a,b,c]

a The Dutch Craniofacial Centre, Department of Oral and Maxillofacial Surgery, Erasmus University Medical Center, Sophia's Children's Hospital, Rotterdam, The Netherlands

b The Craniofacial Centre, Boston Children's Hospital, Boston, United States of America

c Craniofacial Unit, Great Ormond Street Hospital, London, United Kingdom

* Both authors contributed equally to this paper.

Head of Department: Professor Eppo B. Wolvius, DDS MD PhD

ABSTRACT

Introduction

Patients with Craniofacial Microsomia (CFM) mandibles Types I/IIa benefit from combined LeFort 1 osteotomy and Mandibular Distraction Osteogenesis (LeFort+MDO); Type IIb from LeFort+MDO or Bimaxillary osteotomy (BiMax); and Type III from BiMax (with 50% of cases having preceding mandibular procedures, including patient-fitted prosthesis) ; as seen in Part 1. This leads to the question how maxillary and mandibular hypoplasia are correlated and influence the types of maxillary correction.

Material and Methods

A retrospective chart study was conducted including patients diagnosed with CFM from 2 large craniofacial units. Radiographic and clinical information were obtained. Unilateral affected patients with available (ConeBeam) CT-scan of the maxillary-mandibular complex, without treatment of the upper jaw prior to the CT-scan were included. A maxillary cant grading system was set up and evaluated. Pearson correlation coefficients were used to correlate the maxillary cant and the severity of the mandibular hypoplasia.

Results

Eighty-one patients were included of whom 39,5% had a Pruzansky-Kaban type III mandible and 42% a mild maxillary cant. There was a significant positive correlation

between severity of the mandibular hypoplasia and the categorized canting (r=0,370; p<0,001; n=81). Twenty-four patients had maxillary surgery, mainly a BiMax.

Conclusion

There is a positive correlation between the severity of mandibular hypoplasia and maxillary cant. The severity of mandibular hypoplasia seems to dictate an intervention for both maxillary and mandibular surgery.

KEYWORDS

Craniofacial microsomia, Oculo-auriculo-vertebral syndrome, Hemifacial Microsomia, OMENS, Maxillary canting, bimaxillary osteotomy, distraction osteogenesis, le Fort I, mandibular distraction osteogenesis

INTRODUCTION

Craniofacial microsomia (CFM) is best described as a congenital malformation of the derivatives of the first and second pharyngeal arches leading to asymmetrical hypoplasia of the facial skeleton and soft tissues. It is generally considered to be the second most common birth defect following cleft lip and palate.(Grabb, 1965; Heike et al., 1993; Birgfeld et al., 2012) The leading hypotheses on the aetiology include local haemorrhage of the stapedial artery(Poswillo, 1975) and disturbed migration of cranial neural crest cells.(Johnston et al., 1995; Tuin et al., 2015; Caron et al., 2017) With dysmorphologies ranging from mild to severe, patients with CFM are phenotypically heterogeneous. Multiple classification systems have been proposed to categorise and report the severity of the different anomalies. The Pruzansky-classification, later subcategorized by Kaban et al.(Pruzansky, 1969; Kaban et al., 1986) describes the mandibular hypoplasia. The O.M.E.N.S., proposed by Vento et al. includes the five major malformations of the craniofacial region, i.e. Orbit, Mandible, Ear, Nerve and Soft-tissue.(Vento et al., 1991) The O.M.E.N.S-classification was expanded to the O.M.E.N.S-plus to encompass the extra-craniofacial anomalies, often seen in CFM.(Horgan et al., 1995) The most recent derivative of the O.M.E.N.S-plus is the pictorial Phenotypic Assessment Tool-Craniofacial Microsomia (PAT-CFM) by Birgfeld et al.(Birgfeld et al., 2011)

In CFM patients asymmetry of, amongst others, the maxillo-mandibular complex is seen. The correction of the mandibular deformity has been reported and reviewed numerously. (Nagy et al., 2009; Pluijmers et al., 2014) In contradiction, as seen in the previous study entitled “Surgical correction of the Maxilla in Craniofacial microsomia

Part 1: a systematic review", the correction of the maxilla has been studied relatively limited and showed that mandibular deviation upwards and towards the affected side is associated with canting of the occlusal plane.(Grayson et al., 1983) [*part1]* A critical step in achieving better facial skeletal harmony is to restore this maxillo-mandibular asymmetry.

The systematic review of the literature reported a possible correlation between the severity of the mandibular deformity and the type of surgery the correct the maxillo-mandibular asymmetry. Namely, patients with a Pruzansky-Kaban type I and IIa were mostly treated with a combined Le Fort I + mandibular distraction osteogenesis procedure (LeFort+MDO), and patients with more severe presentation: Pruzansky-Kaban type IIb and III benefit from bimaxillary osteotomy (BiMax) [*part1]* Although it must be noted that in half of the patients with a type III had undergone multiple mandibular corrections prior to any maxillary surgery. Therefore, the question arose if the severity of maxillary cant was correlated to the mandibular hypoplasia.

Recent studies report a correlation between the different regions of the craniofacial region mentioned above.(Tuin et al., 2015) (Caron et al., 2017)The studies find a stronger correlation between the structures with their origin in the first pharyngeal arch i.e. between the mandible, orbit and soft-tissue and those with their origin in the second pharyngeal arch i.e. nerve and ear. In this analogy one would aspect a positive correlation between the maxillary cant and the Pruzansky-Kaban classification.

In order to study a large group of patients with CFM, a multicenter collaboration was initiated between the craniofacial units of Rotterdam and Boston.

Following the systematic review of the literature, the purpose of this study was to evaluate a possible correlation between the maxillary cant and the mandibular hypoplasia, and to review the types of maxillary correction in our cohort of CFM patients .

MATERIALS AND METHODS

With approval of both Institutional Review Boards (Rotterdam: File number MEC-2013-575; Boston: File number X05-08-058) a chart study was performed on all CFM patients presented at one of the units between January 1980 until January 2016. Unilateral affected patients with available (ConeBeam) CT-scan of the mandible and the medical history were included. CT-scans of patients who had had treatment of the upper jaw prior to the CT-scan were excluded. All charts were reviewed for information on radiographic and diagnostic criteria.

Since there are no standardized measurement tools to retrospectively evaluate the maxillary cant on CT-scan, a classification system was created for CT-scan reconstructions in the coronal plane, capturing 4 categories: normal, mild, moderate and severe. These categories were based on the degree of deviation of the line between the first molar level of the upper jaw and on the orbital rims- selected as landmarks – on a slice of the coronal plane of the CT-scan. (Figures 1-4)

Two observers (BIP, LSvdL) scored the maxillary cant simultaneously 2 times with an interval of 2 weeks apart. The intrarater variability was calculated with the Intraclass Correlation Coefficient Reliability by internal consistency (Cronbach's α) was

examined. A correlation of >0,7 was set as acceptable; a correlation >0,8 was considered good and a correlation >0,9 was marked as excellent.

Descriptive statistics were used to describe affected side, age at time of surgery and diagnostic data. Pearson correlation coefficients were used to correlate the maxillary cant categorized and the severity of the mandibular hypoplasia as proposed by Pruzansky-Kaban.

RESULTS

Of the 492 patients presented at both units, 81 patients with applicable CT-scans could be included for the analysis.

A total of 8 patients had a Pruzanksy-Kaban type I mandible, 11 had a type IIa, 29 were diagnosed with a type IIb and 32 patients had a type III mandible. (Table 1) Overall, most patients had a mild (42%) to moderate (28,4%) cant. There was a significant positive correlation between severity of the mandibular hypoplasia and the amount of canting of the maxilla ($r=0,370$; $p<0,001$; $n=81$). (Table 1) This correlation was seen in the first and in the second analysis. The intraclass variability was 0,868 (CI: 0,797-0,916) and the Cronbach's α was 0,868 which is a good correlation.

Of the 81 patients presented, 24 patients (29,6%) had undergone maxillary surgery. (Table 2&3) A BiMax was the most performed type of surgery (n=13), followed by a Le Fort1 osteotomy combined with a mandibular distraction (n=8) with the help of extra-orally placed distractors with a multivariate vector (in one case the vector control

was not described), a Le Fort 1 osteotomy (n=2) and a Le Fort 1 osteotomy combined with placement of a costochondral graft to elongate the mandible.

Of those 24 surgical patients, 10 patients had preceding mandibular surgery. Six patients had preceding MDO, of whom 3 followed by LeFort+MDO, 2 followed by a BiMax and 1 followed by a LeFort 1 osteotomy. Two patients had preceding mandibular correction with the help of a costochondral rib graft, of whom 1 was followed by a LeFort1 and the other by a LeFort1+MDO. Lastly 2 patients had a bilateral sagittal split osteotomy, before both had a BiMax.

Furthermore 4 of the 24 patients had additional surgery as a ‘finishing touch’ consisting of a genioplasty in all 4. One patient had additional lipofilling and 1 patient received a PEEK implant.

Of the 57 patients who did not had any form of maxillary surgery, 28 did have mandibular surgery. (table 4) Most patients underwent a MDO (n=19), followed by mandibular reconstruction with a bone graft, including calvarial, costochondral and a fibula graft (n=4). In 3 patients it remained unclear which type of mandibular reconstruction had took place. In this group most patients had a moderate (n=11) to mild (n=10) cant. The Pruzansky-Kaban classification was distributed proportionally: most patients had a Pruzanksy-Kaban type 3 (n=14) followed by Pruzansky-Kaban type 2 (n=12) and 2 patients classified with a Pruzanksy-Kaban type 2a mandible.

DISCUSSION

The systematic review of the literature (part one) showed 7 case series on the maxillary correction of asymmetry in patients with CFM. Viable options included LeFort+MDO and BiMax. In type III mandibles a BiMax was the only procedure performed. However, 50% of the patients who underwent BiMax had earlier mandibular reconstruction. Especially, those with a type III had multiple preceding mandibular reconstructions (up to 12 attempts). Furthermore, the LeFort+MDO was, unexpectedly, the most performed type of surgery in a type I mandible. Unexpectedly, since LeFort+MDO is more invasive in terms of post-surgical care with strict distraction protocols and additional removal of the device(McCarthy et al., 1992; McCarthy et al., 2002).

In the cohort presented, the majority of 81 patients had a Pruzanksy-Kaban type III mandible (n=32; 39,5%), suggesting a bias in the study population, since earlier publications suggest an incidence 10-22,9% of type III mandibles(Vento et al., 1991; Poon et al., 2003; Park et al., 2014; Tuin et al., 2015; Caron et al., 2017). The most logical explanation for this bias is the lack of indication for surgery and thus pre-operative scan in patients with a low type Pruzansky-Kaban.

There are no standardized measurement tools to retrospectively evaluate the maxillary cant. The used classification based on the first molars and orbital rim proved to have a good intra-rater variability. The authors acknowledge the subjectivity of the scale. Due to phenotypical heterogeneity of CFM patients and abnormality of development of landmark points such as the skullbase but also of the zygoma and the frontotemporal region; a more objective retrospective measurement tool is challenging to define. (Schaal et al., 2017)

The severity of canting was mild in most of the patients (n=34). There was a positive relationship between the severity of mandibular hypoplasia and maxillary canting, supporting the theory of failed migration of neural crest cells in the first pharyngeal arch (r=0,370; p<0,001; n=81). *(Tuin et al., 2015; Caron et al., 2017)*

In total 24 patients had maxillary surgery. Due to the retrospective nature of the study it was not always clear from the charts if a patient had had orthodontic therapy. However, when reviewing the types of surgery, BiMAx, LeFort+MDO, it is to be expected that nearly all patients had had pre-surgical orthodontic therapy. Most patients had a bimaxillary osteotomy (n=13). Of these 24 patients (41,7%) had a type of mandibular correction earlier in life. The majority (62,5%; n=15) had a type III mandible and a mild cant (50%; n=12). In the systematic review of the literature two types of treatments were seen: LeFort+MDO and BiMax. However, in this study Le Fort I and Le Fort I in combination with insertion of a costochondral graft (CCG) were also seen in a limited number of patients. The majority of the patients were treated with combined Le Fort 1 and mandibular distraction or BiMax. Half of the patients with a mild cant had had previous mandibular surgery. In the group with a severe cant, only one third had a preceding mandibular correction. Nevertheless, in total 8 of the 16 patients (50%) with a severe maxillary cant had had mandibulary surgery versus 16 of the 34 patients (47,1%) with a mild maxillary cant. Moreover, both maxillary and mandibulary surgery, seems to be ‘mandibulary driven’ i.e. the severity of mandibular hypoplasia seems to dictate intervention rather than a severe maxillary cant.

Furthermore, it must be kept in mind that the numbers in this retrospective study are low and the indications for surgery, other than the plausible restoration of facial

harmony, were poorly documented. A future prospective study, as currently is set up between multiple centers with standardized outcome measurement tools should be conducted to identify the optimal treatment strategy.

Comparable with the systematic review, maxillary surgery was performed during the skeletal maturity with a mean age at time of surgery of 18,2 years old. Based on Part One and Part two correction of the maxillo-mandibular asymmetry it is suggested to wait until permanent dentition, if there are no definite indications for early surgery such as functional (e.g. Obstructive Sleep Apnea) and/or psychological impairments.(Murray et al., 1984; Wolford et al., 2001b, a)

CONCLUSION

An additional study was performed on the correlation between the amount of maxillary canting and mandibular hypoplasia. Furthermore, types of surgical correction of the maxilla and mandible in patients with CFM were evaluated. There is a positive correlation between the severity of mandibular hypoplasia and canting of the maxilla. This outcome could support the failed migration of the (neural) crest hypothesis, aiding to further understanding of the pathoetiology of the deformity.

In contrast to the systematic review of the literature the most often performed type of maxillary surgery in the studied group was a Bimaxillary osteotomy. It must be noted that nearly half of these patients had mandibular surgery earlier in life. The majority of them had a more severe phenotype i.e. Pruzansky-Kaban type III mandible, but notably had a mild maxillary cant. However, 50% of the patients with a severe maxillary cant had had mandibulary surgery versus 47,1% with a mild maxillary cant; regardless of (simultaneous) maxillary surgery. Both maxillary and mandibulary surgery seems to be ‘mandibulary driven’ i.e. the severity of mandibular hypoplasia seems to dictate intervention.

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Ethical considerations

This study was approved by the Medical Ethics Review Committee Erasmus MC.

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TABLES

Pruzansky-Kaban classification / Cant	Pruzansky-Kaban type I	Pruzansky-Kaban Type IIa	Pruzansky-Kaban Type IIb	Pruzansky-Kaban Type III	Total
Normal	3	1	4	0	8 (10,0%)
Mild	4	6	11	13	34 (42,0%)
Moderate	1	2	10	10	23 (28,4%)
Severe	0	2	5	9	16 (19,6%)
Total	8 (10,0%)	11 (13,5%)	30 (37,0%)	32 (39,5%)	81 (100%)

Table 1. Type of mandible versus amount of cant. (r=0,370; p<0,001; n=81).

	Pruzansky-Kaban Type IIa		Pruzansky-Kaban Type IIb		Pruzansky-Kaban Type III		Total	
	Number of patients	Mean age in years	Number of patients	Mean age in years	Number of patients	Mean age in years	Number of patients	Mean age in years
BiMax	3	19,0	2[α]	21,9	8[β]	18,6	13[γ]	19,2
Le Fort 1 + MDO	1	11,7	2	18,1	5	16,1	8	16,4
Le Fort 1	n/a	n/a	1	17,2	1	22,0	2	19,6
Le Fort 1 + CCG	n/a	n/a	n/a	n/a	1	18,6	1	18,6
Total	4	17,1	5	19,4	15	18,1	24	18,2
Number of patients with preceding mandibular surgery	1 (25,0%)		2 (40,0%)		7 (46,7%)		10 (41,7%)	

Table 2. Maxillary surgery.

BiMax: bimaxillary osteotomy; CCG: Costochondral graft; MDO: Mandibular Distraction Osteogenesis; n/a: not applicable;[α]:1 following Surgically Assisted Rapid Maxillary Expansion;[β] :2 following Surgically Assisted Rapid Maxillary Expansion; [γ]: 3 following Surgically Assisted Rapid Maxillary Expansion.

	Mild		Moderate		Severe		Total	
	Number of patients	Mean age in years	Number of patients	Mean age in years	Number of patients	Mean age in years	Number of patients	Mean age in years
BiMax	6[α]	17,8	5[α]	19,6	2[α]	22,2	13[γ]	19,2
Le Fort 1 + MDO	5	15,9	n/a	n/a	3	17,3	8	16,4
Le Fort 1	1	22,0	n/a	n/a	1	17,2	2	19,6
Le Fort 1 + CCG	n/a	n/a	n/a	n/a	1	18,6	1	18,6
Total	12	17,4	5	19,6	7	18,9	23	18,2
Number of patients with preceding mandibular surgery	6 (50%)		2 (40%)		2 (28,6%)		10(41,7%)	

Table 3 Maxillary surgery in terms of canting. BiMax: bimaxillary osteotomy; CCG: Costochondral graft; MDO: MandibularDistraction Osteogenesis n/a: not applicable; [α]:1 following Surgically Assisted Rapid Maxillary Expansion; [γ]: 3 following Surgically Assisted Rapid Maxillary Expansion.

Type of surgery	Normal	Mild	Moderate	Severe	**Total**	Pruzanksy-Kaban 1	Pruzanksy-Kaban 2a	Pruzanksy-Kaban 2b	Pruzanksy-Kaban 3
Bone graft	1	2	2	1	**6**	n/a	n/a	2	4
MDO	n/a	6	8	5	**19**	n/a	2	8	9
Unknown	n/a	2	1	n/a	**3**	n/a	n/a	2	1
Total	1	10	11	6	**28**	n/a	2	12	14

Table 4 Patients with mandibular surgery only. n/a: not applicable

CAPTIONS TO ILLUSTRATIONS

Figure 1. Normal

Figure 2. Mild cant

Figure 3. Moderate cant

Figure 4. Severe cant