Surgical Correction of Craniofacial Microsomia: Evaluation of Interventions in 565 Patients at Three Major Craniofacial Units

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Abstract

Aims

Craniofacial microsomia is characterized by an asymmetric, hypoplasia of derivatives of the first and second pharyngeal arch, leading to a variety of phenotypic presentations. Studies on surgical correction

of patients with CRANIOFACIAL MICROSOMIA have small cohorts, leaving controversial opinions on the optimal treatment modality, the indication for surgery and the optimal timing of surgery. The purpose of this study was to evaluate the types of, timing of, and total number of surgical corrections performed and the number of surgical procedures in correlation to the severity of the phenotype.

Patients and Methods

A retrospective chart study was conducted including patients diagnosed with CRANIOFACIAL MICROSOMIA from three large craniofacial units. Demographic, radiographic, and clinical information was obtained, including type and number of surgical procedures and age at the time of surgery.

Results

A total of 565 patients were included. In total, 443 (78.4 percent) of all patients underwent some form of surgery during their life, varying from skin tag removal to major craniofacial operations. The number of surgical interventions was higher with increasing severity of phenotype, bilateral presentation, and a younger age at the first intervention.

Conclusions

Multiple surgical corrections are frequently seen in patients with a more severe or bilateral presentation. Furthermore, those who are treated earlier in life for correction of asymmetry of the mandible will undergo significantly more surgical procedures to correct the asymmetry later on, independent of the Pruzansky-Kaban type mandible. A prospective international multicenter study is designed with a uniform registration and outcome measurement tool to identify the optimal treatment strategy.

Introduction

Craniofacial microsomia is best described in its embryologic origin: a developmental defect of any structure of the first and/or second pharyngeal arch leading to a predominantly asymmetric, hypoplasia of their derivatives.[1-3] The phenotypical presentation and therefore, the severity of the deformity can vary significantly.[2,4-6] Furthermore bilateral involvement is reported in 2.5-34% of cases [7,8]. Starting early in life, patients can encounter several functional and/or aesthetic problems. Orbital malformations can include epibulbair dermoids, eyelid coloboma, orbital dystopia, and micro- or anophthalmus.[4,9,10] Furthermore, hypoplasia of the jaw may vary from a normally shaped but smaller sized mandible to an abnormally shaped mandible with absence of the condyle and ramus leading not only to functional problems such as a malocclusion, airway problems or ankylosis; but also, to a distinct facial scoliosis.[8,11]. External ear problems, occurring in the majority of patients with ra, ranges from microtia to anotia with atresia of the auditory canal. [4,9,10] Another aspect frequently seen in patients with craniofacial microsomia is the presence of preauricular or facial tags and/or pits with or without cartilage remnants. Besides, soft-tissue problems due to muscle and/or fat underdevelopment or atrophy are described. Macrostomia, (Tessier 7 cleft) can be part of the phenotype of craniofacial microsomia. Finally, facial nerve palsy of either a part of or all branches is observed in 10-45% craniofacial microsomia patients.[12]

Due to the wide phenotypic spectrum of craniofacial microsomia, an internationally accepted classification and/or grading system is essential for the communication between different centers with regards to patient care and research. One of the first grading systems used is the Pruzansky classification,[13] later adjusted by Kaban et al.[14,15] Vento et al. further extended the classification system with inclusion of classifications for Orbital asymmetry, Ear deformity, Nerve dysfunction and Soft-tissue deficiency, to the orbital, mandibular, ear, neural,
and soft-tissue classification;[10] modified by Horgan et al. to the orbital, mandibular, ear, neural,
and soft-tissue -plus which covers additional extracranial features.[4] The most recent derivative is the pictorial Phenotypic Assessment Tool-Craniofacial Microsomia by Birgfeld et al.[5]
Craniofacial microsomia is regarded to be the second most common congenital facial condition following cleft lip and palate.[11] Because of the variable presentation of craniofacial microsomia a wide range of treatment options are available. [16-49] Studies on surgical treatment in craniofacial microsomia

are limited to small cohorts and expert opinions, with significant differences. There is no consensus on indications, the optimal treatment modality and optimal timing of surgery.

In order to study a large group of patients with craniofacial microsomia, a collaboration between the craniofacial units of Erasmus University Medical Center (EMC) Rotterdam, Great Ormond street Hospital NHS Foundation Trust (GOSH) London and Boston's children's Hospital (BCH) Boston, was initiated. Because of this collaboration it was possible to analyze a large data set of patients with craniofacial microsomia. The purpose of this retrospective study was to evaluate the type of surgical corrections of the craniofacial anomaly in patients with craniofacial microsomia. Additional objectives were to evaluate the timing of the procedures and the total number of surgical corrections performed**.** Lastly, the number of surgical procedures in correlation to the severity, including a unilateral versus bilateral phenotype, was evaluated.

Patients and Methods

Study design

With approval of the institutional medical ethics board of all three centers (i.e., Medical Ethics Review Committee Erasmus MC, file number MEC-2013-575; IRB Medical Ethics Review Committee Boston Children's Hospital, file number X05-08-058; and the R&D Medical Ethics Review Committee, Great Ormond Street Hospital NHS Foundation Trust, file number 14 DS25), a retrospective cohort study was conducted. Patients' charts from January of 1980 until January of 2016 of patients diagnosed with craniofacial microsomia were reviewed. Patients in whom adequate diagnosis based on the orbital, mandibular, ear, neural, and soft-tissue–plus classification or phenotypic assessment tool for craniofacial microsomia was not able to be determined were excluded. The Boston Children's Hospital has a history of reporting the orbital, mandibular, ear, neural, and soft-tissue– plus classification in patients' charts. At the Great Ormond Street Hospital and the Erasmus University Medical Center, all patients with both clinical and radiographic images were included and scored with the help of the phenotypic assessment tool for craniofacial microsomia by two trained researchers at each location. If there was any disagreement, the score was discussed if needed with a researcher from the other location, until agreement was achieved. Throughout time, different treatment strategies have been used at each center.

Variables

Baseline characteristics included sex and diagnosis. All surgical notes were reviewed; type and number of surgical procedures, indication, and age at the time of surgery were registered. Surgical procedures included any surgical procedure to correct a functional or aesthetic problem related to craniofacial microsomia (e.g., osteotomies, lipofilling, ear reconstructions). Removal of hardware (such as distraction devices) or surgical treatment to address complications such as infections or device failure was not counted as a surgical correction. Complications following surgery were annotated.

All information was anonymized before it was imported into IBM SPSS Version 24 (IBM Corp., Armonk, N.Y.) for analysis. Descriptive statistics were used to describe sex, laterality, and diagnostic data. Pearson correlation coefficients were used to correlate the severity and laterality. An independent *t* test was performed to compare the mean age between intervention groups. A linear regression was calculated to predict the number of operations based on the age at first surgery and Pruzansky-Kaban type.

Results

Patient demographics

Craniofacial microsomia was diagnosed in 955 patients. A total of 565 patients met the inclusion criteria. (Table 1)

The male to female ratio was 1.2:1. A total of 496 (87,8%) patients had a unilateral presentation of craniofacial microsomia. The distribution between right and left-sided craniofacial microsomia was 1.2:1 (n=496) as well. (Table 1)

Most patients with unilateral craniofacial microsomia had a Pruzansky-Kaban mandible Type I (26.8%) or IIa (26.6%) followed by the Pruzanksy-Kaban IIb (23.2%) and III (15.9%). In 40 patients from the BCH the data on the mandible was inconclusive with notes in the charts doubting 2 different scores for example 1 or 2a. The Pruzansky-Kaban classification of the more severely affected side in patients with bilateral craniofacial microsomia was significantly more frequently scored as IIB or III compared to the Pruzansky-Kaban classification of the unilateral affected patients ([Pearson chi-square

(3) = 26,227, p < 0.001]). (Tables 2 & 3) The classification of the nerve proved to be challenging in this retrospective cohort, we were unable to score the nerve function reliably.

	EMC	GOSH	BCH	Total
Number of patients	152	134	279	565 (100%)
Male	79	70	157	306 (54.2%)
Female	73	64	122	259 (45.8%)
Right sided CFM	75	70	127	272 (48.1%)
Left sided CFM	67	46	111	224 (39.6%)
Bilateral CFM	10	18	41	69 (12.2%)

Table 1. Patient Demographics of the Total Population
EMC, Erasmus Medical Center; GOSH, Great Ormond Street Hospital;
BCH, Boston Children's Hospital; CFM, craniofacial microsomia.

	EMC	GOSH	BCH	Total
Pruzansky- Kaban I	45	27	61	133 (26.8%)
Pruzansky- Kaban IIa	39	44	49	132 (26.6%)
Pruzansky- Kaban IIb	32	26	57	115 (23.2%)
Pruzansky- Kaban III	26	19	34	79 (15.9%)
Inconclusive data	-	-	37	37 (7.5%)

Table 2. Pruzansky-Kaban Classification of 496 Unilateral Craniofacial Microsomia Patients
EMC, Erasmus Medical Center; GOSH, Great Ormond Street Hospital;
BCH, Boston Children's Hospital; CFM, craniofacial microsomia.

	Bilateral most severe	Bilateral less severe
Normal		1 (1.4%)
Pruzansky- Kaban I	9 (13%)	18 (26.1%)
Pruzansky- Kaban IIa	11 (15.9%)	21 (30.4%)
Pruzansky- Kaban IIb	18 (26.1%)	10 (14.5%)
Pruzansky- Kaban III	28 (40.6%)	16 (23.2%)
Inconclusive data	3 (4.3%)	3 (4.3%)

Table 3. Pruzansky-Kaban Classification in 69 Bilateral Patients with Craniofacial Microsomia

Correction of:	Number of patients (n=443)
Orbito / Zygomatic complex	53 (12%)
Eye†	55 (12.5%)
Mandible	189 (42.7%)
Maxilla	73 (16.5%)
Ear	228 (50.2%)
Nerve	2 (0.5%)
Soft-tissue	230 (51.9%)
Macrostomia	61 (13.8%)
Cleft lip and/or palate	72 (16.3%)

Table 4. Number of Patients Treated Surgically per Region*
*n = 443.
†Correction of eye deformities (e.g., ptosis, epibulbar dermoid).

Orbito-zygomatic-complex

With regards to the orbito-zygomatic-complex; 53 of the 565 patients had surgery involving these structures. Forty-nine patients had a unilateral presentation of craniofacial microsomia. More than half of the patients had an abnormal orbital size and/or displacement (Table 5). Most patients received an

alloplastic malar implant (n=23), followed by bone grafts including costochondral (n=8), iliac (n=5) cavarial (n=5) and fibula (n=1) bone grafts. Eleven patients had a variety of orbito-zygomatic osteotomies. Only four patients had a bilateral presentation. Two received correction of the asymmetry by alloplastic malar implants. The other two bilateral patients received a bone graft of either a fibula graft or an iliac bone graft.

Eye

In total 55 patients with craniofacial microsomia underwent surgery to the eye. In most patients this consisted of removal of an epibulbar dermoid (Table 6).

Type of surgery	Unilateral CFM (n=40)	Bilateral CFM (n=15)
Removal epibulbar dermoid	18	7
Exo/esotropia correction	9	1
Ptosis correction	8	2
Coloboma repair	6	4
Canthoplasty	3	5
Probing of nasal lacrimal duct	3	1
Prosthesis	2	2
Cornea correction	2	
Dacrocystorhinostomy	1	
Entropion correction	1	

Table 6. Eye Surgery
CFM, craniofacial microsomia.

Mandible

Of the 443 patients who received a form of surgery to correct the deformity, 42.7 percent had a mandibular correction or reconstruction.

Most patients had a Pruzansky-Kaban type III mandible, followed by the type IIb, type IIa and type I (Table 7). The patients with inconclusive data on the Pruzansky-Kaban classification were left out (n=17).

	P-K I (n=2)		P-K IIa (n=9)		P-K IIb (n=17)		P-K III (n=21)		Indecisive P-K	
	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral
Orbit classification	O4	O0	O0: n=6 O1: n=1 O3: n=2	n/a	O0: n=6 O1: n=2 O2: n=6 O3: n=2 O4: n=1	n/a	O0: n=4 O1: n=5 O2: n=4 O3: n=1 O4: n=3	O0: n=3	O0: n=4	n/a
Number of patients	1	1	9	n/a	17	n/a	18	3	4	n/a
Alloplastic implant	n=1	n/a	n=7	n/a	n=3	n/a	n=6	n=2	n=3	n/a
Bone graft	n/a	n=1		n/a	n=8	n/a	n=9	n=1	n=1	n/a
Osteotomy	n/a	n/a	n=2	n/a	n=6	n/a	n=3	n/a	n/a	n/a
*Mean age at time of first surgery (median)	15.5	n/a	12.9 (14.7)	n/a	14.7 (15.8)	n/a	12.3 (11.5)	15.7 (16.8)	13.5 (15.1)	n/a
Mean number of surgical procedures	1	1	1	n/a	1.1	n/a	1.2	2	1.3	n/a

Table 5. Orbitozygomatic Reconstructions*

P-K, Pruzansky-Kaban; O0, normal orbit; O1, abnormal orbital size; O2, inferior orbital displacement; O3, superior orbital displacement; O4, abnormal orbital size and displacement; n/a, not applicable.

	P-K I (n=7)		P-K IIa (n=33)		P-K IIb (n=65)		P-K III (n=71)		Overall(n=176)	
	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral
Number of patients	7	n/a	29	4	59	6	54	17	149	27
Most performed surgery	Alloplastic graft.	n/a	MDO	Osteotomy	MDO	MDO	CCG	CCG	MDO	BG
*Mean age at time of first surgery (median)	16.40 (17.51) (n=5)	n/a	12.14 (11.61)	18.77	13.13 (11.31) (n=51)	8.70 (10.11) (n=4)	10.67 (9.4) (n=49)	7.04 (6.31) (n=7)	12.12 (sd 6.07)	7.45 (sd 4.61)
Mean number of surgical procedures	1	n/a	1.41	2	1.75	2.5	2.3	2.5	1.85	2.44

Table 7. Mandibular Reconstructions

P-K, Pruzansky-Kaban; n/a, not applicable; MDO, mandibular distraction osteogenesis; CCG, costochondral rib graft; BG, bone graft (including CCG).

Unilateral craniofacial microsomia

Six patients with a Pruzansky-Kaban type I mandible underwent surgery, including an alloplastic graft, genioplasty, distraction osteogenesis, costochondral graft and osteotomy. Patients with a unilateral Pruzansky-Kaban type IIa most often received distraction osteogenesis to lengthen the underdeveloped mandible. In the group of patients with a unilateral Pruzansky-Kaban type IIa 10 of the 19 patients underwent multiple surgeries to correct the deformed mandible. Most often the additional surgery consisted of a genioplasty (n=5). In the unilateral Pruzansky-Kaban type IIb group, 26 patients underwent distraction osteogenesis followed by reconstruction of the mandible with the help of a costochondral graft (n=17). However, in 29 of 59 (49.2%) unilateral Pruzansky-Kaban type IIb patients, an additional type of surgery was carried out. Most patients (n=15) underwent additional osteotomies including bimaxillary osteotomies, unilateral or bilateral sagittal split osteotomies and genioplasties.

In the group with a unilateral Pruzanksy-Kaban type III, most patients underwent a reconstruction of the absent condyle with the help of a bone graft (n=30). Most often, a costochondral graft was used, followed by an iliac bone graft and calvarial bone graft. Additional surgery to correct the deformity in this group was performed in 61percent of the patients. Most of these patients had additional osteotomies including bimaxillary osteotomies, unilateral; or bilateral sagittal split osteotomies and genioplasties (n=13), followed by distraction osteogenesis of the bone graft (n=10).

Bilateral craniofacial microsomia

The same trend found in unilateral patients can be found in the bilateral group. Osteotomies are most often performed in the Pruzansky-Kaban IIa group (n=4) and distraction osteogenesis is performed most in the bilateral Pruzanksy-Kaban type IIb group (n=4). The need of reconstruction of the mandible using bone grafts is most frequently seen in the Pruzanksy-Kaban type III group (n=11). Furthermore, 75 percent of all bilateral patients who underwent surgery to correct the deformity needed additional surgery with frequencies ranging from 1 to 5 surgical procedures and an average of 2.44.

Number of surgical procedures.

As mentioned above, in bilateral patients the average number of surgical procedures is 2.44, in comparison to unilateral patients who will undergo an average of 1.85 procedures [Pearson chi-square (5) = 16.037, p = 0.007]. The age of patients at their first mandibular procedure, who underwent 1-2 surgical procedures compared to those with 3 or more, drops from a mean of 12.18 years (n= 116) to 9.73 years (n=35) ($t_{(149)}$=2.11, p=0.036).

Finally, a linear regression was calculated to predict the number of surgeries based on the age at first surgery and Pruzansky-Kaban. The linear regression model indicated that for every year increase in age a lower number of operations was performed, independent of the Pruzanksy-Kaban type mandible (coefficients -0.033, SE 0.016, p=0.042).

Maxilla

Seventy-three patients underwent correction of the maxilla (Table 8). The interventions included bimaxillary osteotomies, with a “classic” bilateral sagittal split osteotomy of the mandible and a distraction osteogenesis of the hypoplastic mandible or bone graft with a unilateral sagittal split osteotomy on the “normal” side (Le Fort plus mandibular distraction osteogenesis procedure); and single Le Fort osteotomies with or without a preceding surgically assisted rapid maxillary expansion and recontouring of the maxilla. A surgically assisted rapid maxillary expansion followed by a Le Fort I osteotomy was not counted as an additional correction, because this is regarded as a preparative step.

In a total of 42 patients, classic bimaxillary osteotomies were performed, followed by a single Le Fort I procedure (n = 7) and a Le Fort plus mandibular distraction osteogenesis procedure (n = 5). In eight patients, a surgically assisted rapid maxillary expansion was performed preceding the definite osteotomy. All patients were treated at the end of puberty. Additional maxillary surgery to reach the desired facial symmetry was rarely seen.

	P-K I (n=1)		P-K IIa (n=14)		P-K IIb (n=26)		P-K III (n=27)		Overall(n=73)	
	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral
Number of patients	1	n/a	12	2	24	2	21	6	63*	10
Most performed surgery	unknown	n/a	BiMax	BiMax$^{\alpha}$	BiMax±	BiMax	BiMax$^{\alpha}$	BiMax$^{\alpha}$	BiMax∞	BiMax$^{\pm}$
*Mean age at time of first surgery (median)	unknown	n/a	16.7 (18.1)	21.3	18.6 (17.6)	16.8	16.3 (16.7)	17.5 (16.8)	17.2 (17.2)	18.4 (16.8)
Mean number of surgical procedures	1	n/a	1	1	1.1	1	1.1	1	1.1	1

Table 8. Maxillary reconstructions.

BiMax: Classic bimaxillary osteotomy; n/a: not applicable; P-K: Pruzansky-Kaban;

α: 1 following surgically assisted rapid maxillary expansion.

±:2 following surgically assisted rapid maxillary expansion.

∞:4 following surgically assisted rapid maxillary expansion..

* 5 unilateral patients had indecisive Pruzansky-Kaban classifications, all of whom had a bimaxillary osteotomy.

	E0: Normal ear		E1: All parts present. mild deformity		E2: Auricle ½- 2/3 of predicted size Not all parts present		E3: Severely malformed, often peanut shaped		E4: Anotia	
	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral	Unilateral	Bilateral
Number of patients	2	n/a	9	n/a	37	1	144	17	2	1
Most performed surgery	Otoplasty	n/a	Otoplasty	n/a	Reconstruction with CCG	Reconstruction with CCG	Reconstruction with CCG	Reconstruction with CCG	n/a	Reconstruction with CCG
Mean age at time of first surgery (median)	10.7 (7.5)	n/a	6.2 (5.6)	n/a	9.6 (9.5) (n=35)	11.7	10.4 (10.1) (n=127)	11.5 (11.4) (n=13)	30.4 (30.4)	n/a
Mean number of surgical procedures	1	n/a	1.4	n/a	2.4 (n=36)	2.5 (per ear)	2.6	2.7 (per ear)	2.5	1

Table 9. Ear reconstructions.

CCG: Costochondral graft; n/a: not applicable.

Ear

A total of 228 patients (40.4 percent) underwent reconstruction of the ear. Of 10 patients, there were insufficient data on the type of surgery. These were left out of the analysis.

In the normal to mild cases (E0 to E1), patients underwent otoplasty, most often accompanied by other soft-tissue reconstruction in the area (Table 9). A total of 37 unilateral E2 ears were corrected. Most ears were corrected with the help of a costochondral graft (n = 23) followed by a “regular” otoplasty (n = 14). One bilateral case had rib-graft reconstructions for both ears.

In the unilateral patients with type E3 ears, 120 patients underwent multistage reconstruction with the help of a costochondral graft. Three of these patients required a preceding tissue expander treatment. Nineteen patients had a “regular” otoplasty; however, in five cases, a reconstruction with the help of a costochondral graft was performed later. In one patient, a prosthetic ear was used. Furthermore, one patient’s ear was reconstructed with a free vascular skin graft. In the bilateral patients with type E3 ears, all patients underwent multistage reconstruction with a costochondral graft. Two patients needed a tissue expander before the definitive treatment. Three patients received a neoear for their anotia (E4). Two patients, one of whom was a bilateral patient, received a reconstructed ear using a costochondral graft. One unilateral patient received an implant-supported epithesis.

Nerve

One patient underwent a cross-facial nerve grafting at age 15. Another patient received gold weights to correct the lagophthalmos caused by facial nerve impairment.

Soft Tissue

In total, 230 patients underwent soft-tissue correction. In most cases (n = 139), it concerned the removal of a skin tag, dermoid or duct cyst, skin pits, or cartilage remnant in the embryologic pathway of the first or second pharyngeal arch. Most patients had incomplete data on the age at first treatment other than a notion of treatment during infancy. If we set these procedures apart, 91 patients underwent surgery involving soft tissue primarily. The most common procedure was lipofilling, with an average of 1.2 procedures per patient, followed by a type of free autologous tissue transfer (Table 10).

Clefts

Macrostomia correction was performed in 61 patients, of whom six also had a cleft lip and palate. Nine patients were treated for their cleft lip. A total of 24 patients had a cleft palate, which needed correction. Furthermore, 32 patients underwent cleft lip and palate surgery; eight of these patients had a bilateral presentation.

Type of procedure	Number of patients	Mean age*
Total	230	14.7 (n=91)
Skintag, dermoid or duct cyst, skin pits or cartilage remnant removal	139	Infancy
Lipofilling	57	14.7
Autologous tissue flap	19	16.7
Scar revision	15	9.6
Browlift	5	14
Botulinum toxin type A	6	9.5
Tissue reduction	11	16.8
Tissue expander	6	13.3

Table 10. Soft-Tissue Reconstructions*
*n = 91.

Complications

Complications were annotated in 5.7 percent of patients (n = 3) that underwent an orbitozygomatic complex reconstruction, all of which were (autologous) graft infections. One complication (1.8 percent) was registered in the eye correction: a corneal melt. In 63 patients that underwent mandibular reconstruction (33.3 percent), a complication occurred. These complications included autologous graft infections (n = 12), postoperative hypoesthesia (n = 9), and ankylosis or graft ankylosis (both n = 8). Furthermore, wound infection (n = 6), infected osteosynthesis material and malunion/nonunion (n = 5), malpositioning of the distraction device and overgrowth of the rib graft (n = 3), infected Medpor (Porex Surgical, Newnan, Ga.) (n = 2), and graft dehiscence and hypertrophic scars (n = 1) were annotated. In the maxillary surgery group, three patients (4.1 percent) encountered a complication: one patient had a wound infection, one patient had a sinusitis, and one patient had hypoesthesia postoperatively. In 27 patients who underwent ear reconstruction (11.8 percent), notation of a complication was made in the charts. Most had a wound infection (n = 15), followed by an infection of the autologous graft and hypertrophic scars (n = 4), graft dehiscence (n = 3), and a malfunctioning of the hardware (n = 1). In three of the 230 patients with a soft-tissue correction (1.3 percent), a notation was made of a complication: two wound infections and one infected autologous graft. There were no complications mentioned in the patients' charts after the nerve reconstruction, the correction of the macrostomia, or the cleft lip and/or palate repair.

Discussion

Craniofacial microsomia has a heterogeneous presentation, demanding a multiangle approach.[46-48] Studies on surgical correction of patients with CFM until now have small cohorts restricted to expert

opinions, with significant differences on not only the optimal treatment modality but also indication of surgery and optimal timing of surgery. In this study the surgical corrections performed at three large centers were evaluated. Because of the retrospective nature of the study indications were not always stated and therefore left out of this study. Furthermore, the complications and treatment of complications were poorly documented. The presented data might be an under-reporting of the true numbers.

For the evaluation of the patients both the phenotypic assessment tool for craniofacial microsomia and the orbital, mandibular, ear, neural, and soft-tissue–plus classification were used.[4] The authors are supportive of the phenotypic assessment tool for craniofacial microsomia, despite the tool's convenience and ease of use, some data including nerve function, and radiographic orbit position were left out since it was impossible to gather these specific data from the historic charts.

This study finds a ratio of 1.2:1 for both the male-to-female ratio as for the right-to-left ratio, which represents a more right sided affected deformity. Some authors report a 3:2 predominance for the male gender and for a right-side presentation of the deformity, whereas other studies report a more equally distribution by sex and side.[4,6,50-52] Furthermore 87.8 percent had a predominantly unilateral presentation compared to 2.5 to 34 percent reported in literature.[53-55]

Throughout the years, different treatment modalities and paradigms have been proposed for the correction of the asymmetry of the deformity.[24,32,56,57] In this cohort 78.4 percent of the patients underwent surgery related to their craniofacial deformity.

The Pruzansky-Kaban type III mandible patient is the most challenging in terms of correction of the facial asymmetry.[26,29,57] Patients with a unilateral type IIb mandible are in need of multiple operations in 49.1 percent. A unilateral type III mandible, however, would need multiple operations in 61 percent of the cases; however, the most challenging patients are the bilateral patients, who are in need of multiple surgical procedures in 83 percent (Pruzansky-Kaban type IIb) and 76 percent (Pruzansky-Kaban type III) of all cases.

With regard to mandibular surgery, age at the first surgical procedure is shown to have an influence on the number of surgical procedures needed throughout life. Patients, both unilateral and bilateral, who underwent three or more surgical corrections of the mandible were treated significantly earlier at a mean age of 9.73 years (n = 35), compared with those who underwent "only" one or two surgical procedures of the mandible, who were on average treated at 12.81 years (n = 116; t_{149} = 2.11; p =

0.036). Also, the linear regression model indicated that for every year decrease in age, the number of operations performed went up, independent of the Pruzansky-Kaban type mandible. In other words, those who are treated earlier in life for correction of asymmetry of the mandible will undergo more surgical procedures to correct the asymmetry, possibly suggesting that the operations might be responsible for impaired growth, which then increases the need for more operations. This reinforces the policy of correcting mandibular asymmetry at an older age unless there are significant functional problems (e.g., obstructive sleep apnea).[28,58]

Surgical interventions regarding the maxilla were most often single procedures. The treatment outcome, measured as the number of sur- gical procedures/the need of additional surgery, might be better because of the skeletal maturity of the patients at the time of surgery; the mean age at the time of intervention was 17.2 to 18.4 years.

Half of all patients underwent ear reconstruction, with the Nagata two-step otoplasty most frequently performed. However, in 40.1 percent, the number of surgical procedures was more than two. Concurrent with the literature, patients on average underwent a total of 2.5 instead of two procedures, with one case requiring nine procedures because of skin necrosis or inflammation.[59,60]

Facial nerve weakness could possibly contribute to a negative surgical outcome in terms of patient satisfaction and quality of life. Cline et al. recently reviewed the literature concerning the prevalence of facial nerve palsy, which is found to range between 10 and 45 percent.[12] In comparison with other surgical corrections, little has been written on facial nerve reanimation, especially with other surgical techniques, for the correction of asymmetry in craniofacial microsomia.[61] However, reconstruction of the nerve is comprehensive as a result of agenesis and/or underdevelopment of the overlying structures. [62] To evaluate the influence of the facial nerve in corrective surgery, it would be recommended to look into a grading system that includes both dynamic and static symmetry in patients.

Based on the data presented, we advocate postponing corrective surgery until a more mature age unless there are significant functional problems. This approach will most likely reduce the number of surgical interventions and burden of care for these patients and their caretakers.

Conclusion

Although craniofacial microsomia is considered to be the second most common craniofacial defect after cleft lip and palate,11 the numbers of patients in case series in the literature are low. We present the surgical data of 565 craniofacial microsomia patients. Most patients had a unilateral Pruzansky-Kaban type I (26.8 percent) or IIa (26.6 percent) mandible, followed by the Pruzansky-Kaban IIb (23.2 percent) and III (15.9 per- cent) mandible. In this large cohort, 78 percent of the patients underwent surgery related to their craniofacial deformity.

Concurrent with other studies, the most challenging patients in terms of correction of asymmetry have Pruzansky-Kaban type III mandibles. However, we found that another subgroup consists of bilateral craniofacial microsomia patients in whom one could assume the bilaterality would lead to more symmetry but who would face an even larger number of surgical procedures—on average 1.75 times more—than unilateral craniofacial microsomia patients. Furthermore, those who are treated earlier in life for correction of asymmetry of the mandible will undergo significantly more surgical procedures to correct the asymmetry later, independent of the Pruzansky-Kaban type mandible. Prospective studies are essential and patient outcome measurements are needed to be able to truly compare patient outcomes between different treatment modalities to further improve care.

Currently, a prospective international multicenter study is designed with uniform registration and outcome measurement tools. To reduce the number of surgical interventions, the clear indications for surgery should be defined.

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Introduction

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