Craniofacial Microsomia: A Long-term Outcome of Early Mandibular Distraction Osteogenesis and Comprehensive Care at the Tawanchai Center

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Background: *Craniofacial microsomia (CFM) is a complex, congenital, malformation, primarily involving structures derived from the first and second branchial arches. There is limited information on its long-term management and outcomes.*
Objective: *To present the long-term management and outcome of a patient with CFM treated by early distraction osteogenesis and a protocol of comprehensive care at the Tawanchai Center, Srinagarind Hospital, Khon Kaen University.*
Material and Method: *After reviewing the medical records for the clinical presentations, assessments, and long-term managements and outcomes of patients with CFM at Srinagarind Hospital, we focused on one patient, treated by early surgical reconstruction, mandibular distraction osteogenesis (DO), and comprehensive care according to the protocol developed at the Tawanchai Center.*
Results: *The patient presented normal speech, mouth breathing, normal swallowing, and normal temporomandibular joint function. He had an antimongoloid slant, left malar hypoplasia, a cross bite, occlusal plane canting and a slightly deviated chin to the right, a good mouth opening, and a normal bite pattern. The patient was completely satisfied according to overall satisfaction, nose, and upper lip; and moderately satisfied according to overall face, head shape, and occlusion.*
Conclusion: *Our study suggests that the use of DO in young children with CFM provides good long-term distraction on the growth of the mandible and greater facial symmetry. The study addresses the comprehensive evaluation of the long-term, interdisciplinary, comprehensive care of a patient with CMS. Consideration of the needs and expectations of the patient and his/her family and other involved stakeholders is essential.*

Keywords: *Craniofacial microsomia, Early mandibular distraction osteogenesis, Comprehensive management, Long-term outcome*

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Craniofacial microsomia (CFM) is a complex congenital malformation involving craniofacial structures derived from the first and second branchial arches with highly variable phenotypes, including macrostomia, cleft lip with and/or without cleft palate, pre-auricular appendages or sinuses, ear deformities, hearing loss and orbit, zygomatic, maxilla and mandibular deformity. Syndromic and non-craniofacial anomalies may be findings, including to the cardiac system, the vertebral or central nervous system, the limbs, hemifacial microsomia, first and second branchial arch syndrome, otomandibular dysostosis, oculo-auriculo-vertebral spectrum, facio-auriculo-vertebral syndrome, Goldenhar syndrome, and lateral facial dysplasia[1,2].

The objectives of the study are to review the clinical presentations, assessment, and long-term management and outcome of a patient with CFM, treated by early surgical reconstruction, distraction osteogenesis and comprehensive care as per the protocol of the Tawanchai Center, Srinagarind Hospital, Khon Kaen University[3].

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Material and Method

Study design

From the medical records of patients with

CFM, we reviewed the clinical presentations, assessment, and long-term management and outcome of a patient with CFM, treated by early surgical reconstruction, distraction osteogenesis and comprehensive care per the protocol of the Tawanchai Center[3] seen and managed at Srinagarind Hospital between 1993 and 2011. The patient was treated by mandibular distraction osteogenesis with long-term follow-up.

The protocol of this study was reviewed and approved by the Ethics Committee of Khon Kaen University, using the standards set out in the Declaration of Helsinki. Written, informed consent was obtained for the release of the photograph.

Results

Patient report

A male patient born in Khon Kaen, aged 2 years old, presented with hypoplasia of the left zygoma, orbit, and mandible. Craniofacial microsomia was diagnosed and classified as type IIB based on Mulliken and Kaban's modified Pruzansky classification[4] (Fig. 1 and 2). Distraction osteogenesis of the left mandible was performed when he was 2 years old. Fig. 3 and 4 show the patient during and after the distraction osteogenesis of the left mandible**.** At age 5 years, a calvarial bone graft was used to correct the left floor of orbit and maxilla with scar revision.

At 19 years of age with complete facial skeletal maturation, a bony, soft tissue and dental analysis was performed with panoramic film, 3-D computerized tomography lateral cephalogram, and patient satisfaction evaluated.

The patient had an asymmetrical ovoid facial type with hypoplastic zygoma and maxilla on the left side. The level of his left eye was lower than the right eye, the nasal septum deviated to the left, the chin pointed slightly to the right and there was maxillary and mandibular occlusal canting (Fig. 5). The functional evaluation showed that the patient presented normal speech, mouth breathing, swallowing, and Temporomandibular joint (TMJ) function. The maximum mouth opening was 40 mm with no functional shift or CO-CR discrepancy, skeletal normal bite pattern, or open bite tendency.

Intraoral examination revealed fair oral hygiene with generalized mild gingivitis. Permanent dentitions were present except 18, 24, 25, 26, 27, 28 were missing clinically. Upper dental midline (UDM) shifted to the left 8.5 mm and lower dental midline (LDM) shifted to the right 3 mm (Fig. 6).

The CT scan of the facial bone with 3D reconstruction revealed hypoplasia of the left maxillary bone, left maxillary sinus, left zygomatic arch, lesser wing of sphenoid bone, left nasal turbinates and left mandible, and chronic pansinusitis with probable right antrochoanal polyp (Fig. 7).

Problem lists using the Orbital Mandible Ear Nerve Soft Tissue (OMENS) classification[5] included left craniofacial microsomia ($O_0M_1E_0N_0S_2$), left eye amblyopia, and antimongoloid slant, left malar hypoplasia, cross-bite, occlusal plane canting, and chin deviated to the right. The future surgical and orthodontic management plan included pre-surgical orthodontic management for occlusion, two-jaws surgery (Le Fort I osteotomy + BSSO) for correcting canting and chin deviation, autologous bone graft augmentation of the maxilla and zygoma for contouring,

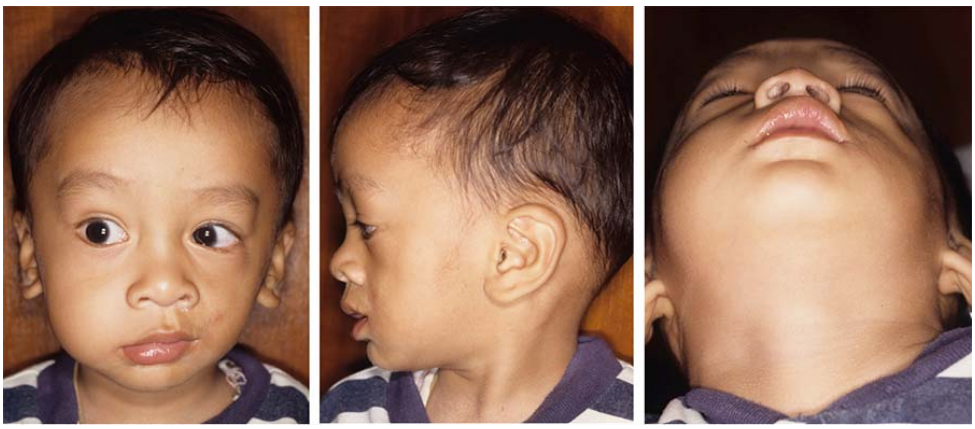

Fig. 1 Two-year-old male presenting with hypoplasia of the left zygoma, orbit, and mandible and diagnosed craniofacial microsomia, type IIB.

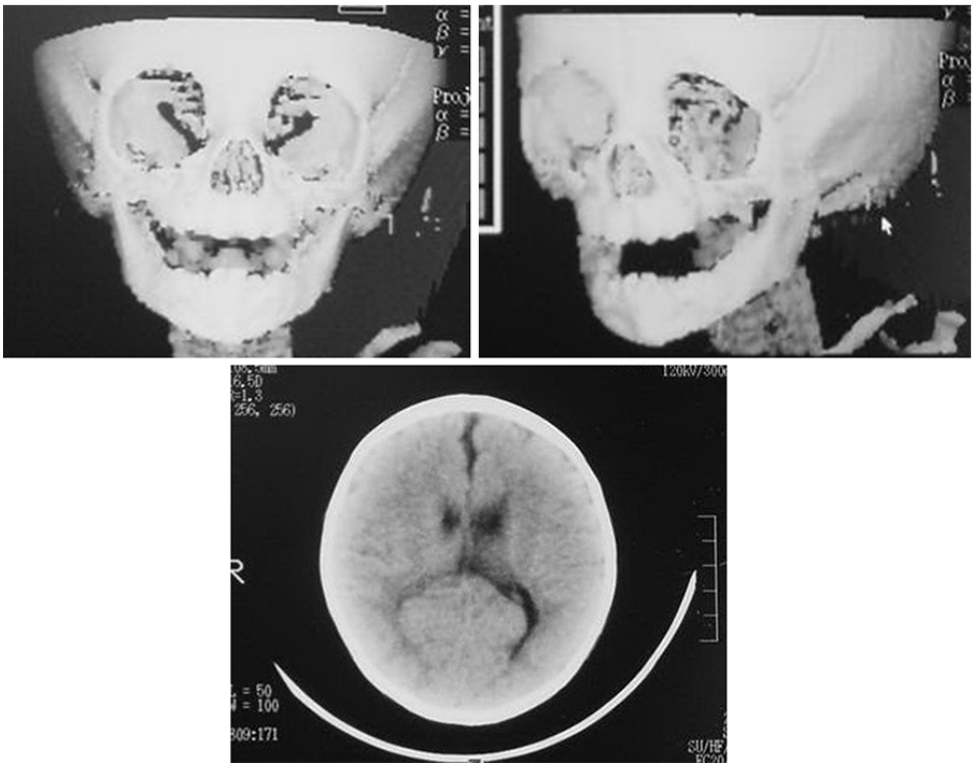

Fig. 2 CT scan of craniofacial skeleton and intracranial structures of the patient reveal hypoplasia of the left zygoma, orbit, and mandible diagnosed as craniofacial microsomia, type IIB with no associated intracranial anomalies.

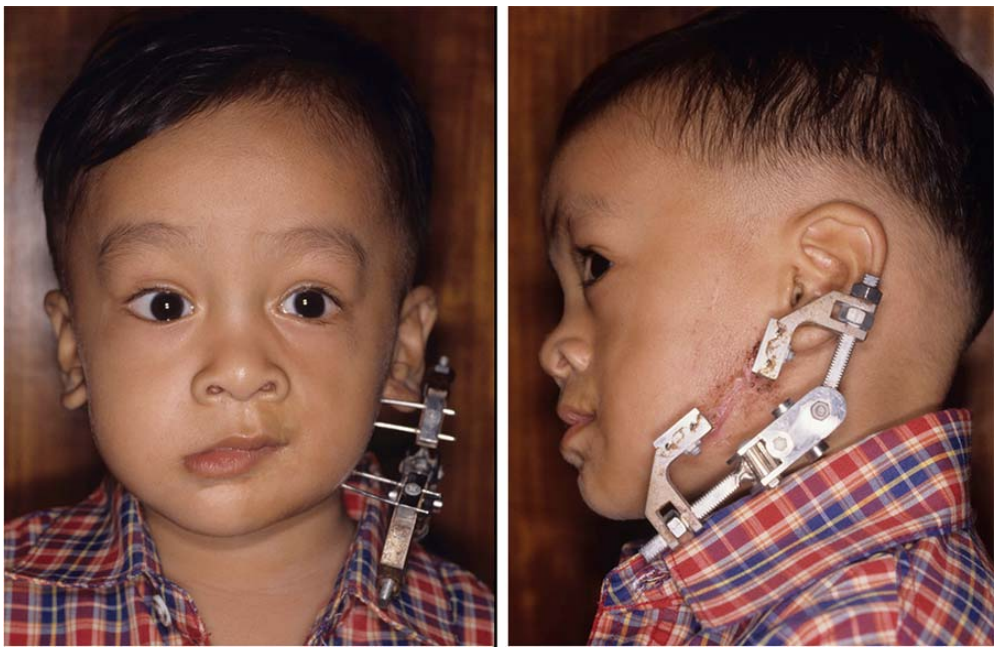

Fig. 3 During distraction osteogenesis of left mandible.

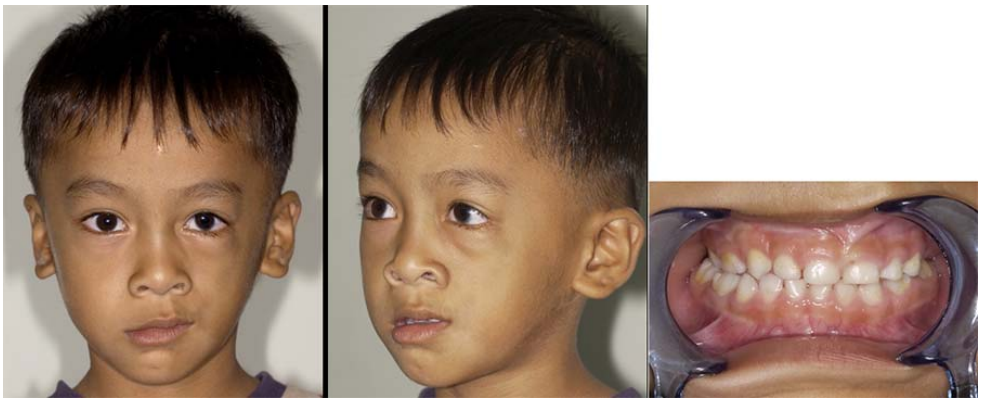

Fig. 4 Patient at age 4 years after distraction osteogenesis.

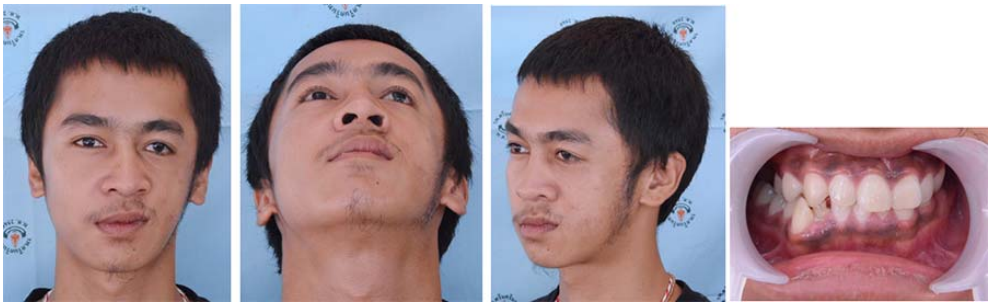

Fig. 5 Patient at 19 and 18 years after DO.

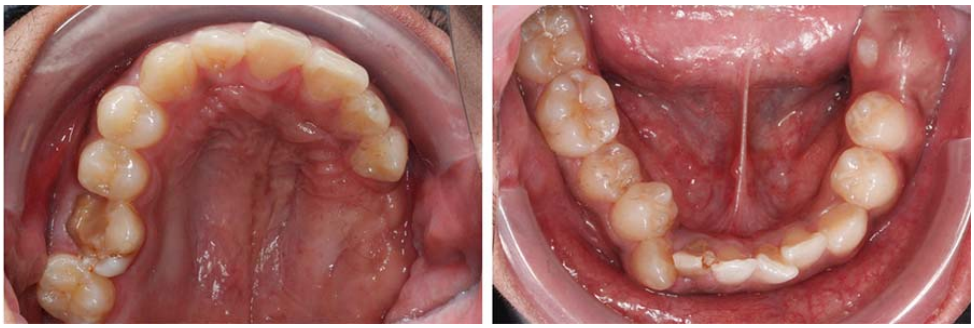

Fig. 6 Intraoral examination.

and autologous fat graft for correcting facial contouring.

The patient scored completely satisfied based on overall satisfaction, nose, and upper lip; and moderately satisfied based on overall face and head shape and occlusion (Table 1).

A psychological adjustment evaluation for quality of life was performed. The patient had no obvious disturbance regarding dental or deglutition functions, confidence, or speech. He was concerned about future operations but confident in the medical team. He had no inferiority complex regarding his face, socialized normally, had good relationships with friends, and was happy with his family. He scored himself 8/10 using the Cantril happiness score.

Discussion

Craniofacial microsomia (CFM) is a unilateral or bilateral congenital deficiency of the affected skeletal and soft tissue structures derived from the first and second branchial arches. The clinical findings of these facial anomalies are important for diagnosis,

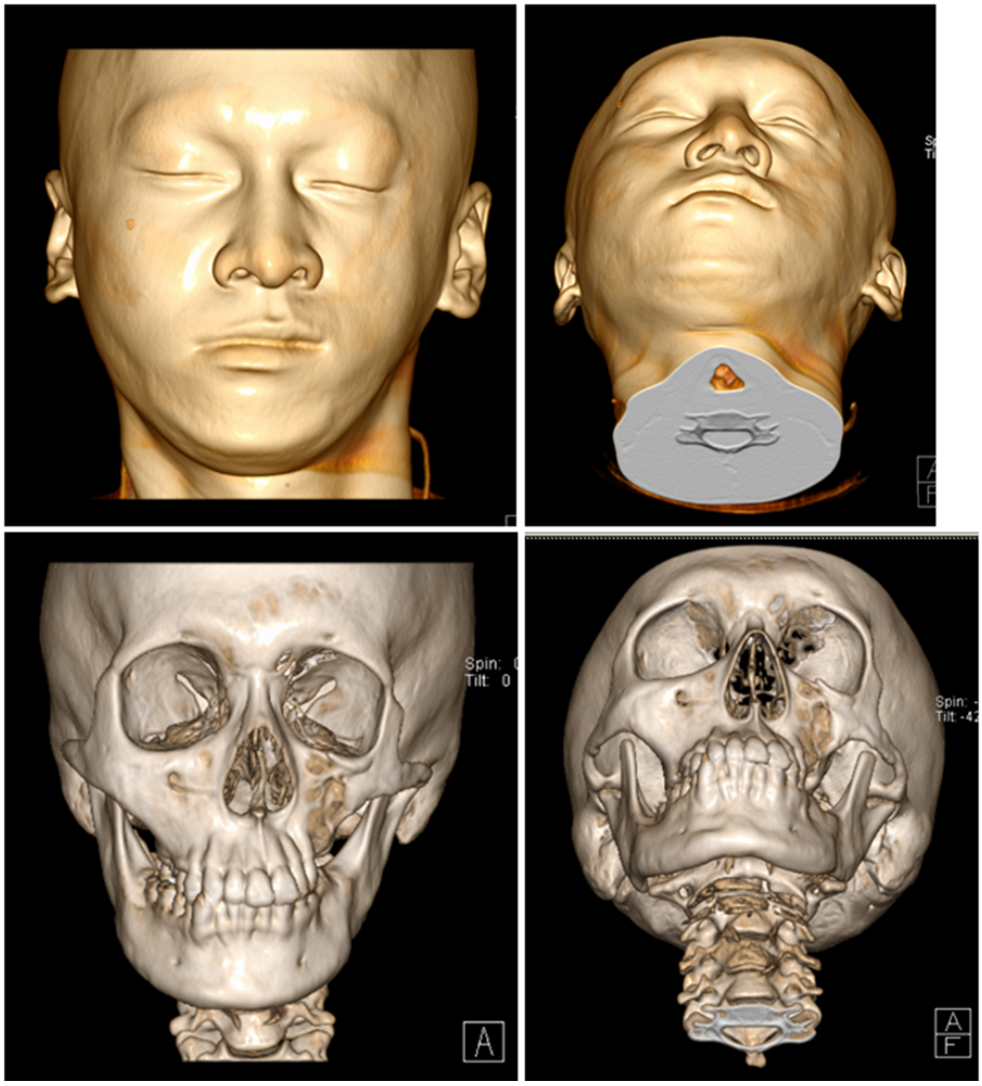

Fig. 7 CT scan of facial bone with 3D reconstruction.

Table 1. Patient satisfaction

Category	Level
Overall satisfaction	5*
Overall face and head shape	3
Nose	5*
Upper lip	5*
Occlusion	3

5 = completely satisfied; 4 = very satisfied; 3 = moderately satisfied; 2 = slightly satisfied; 1 = not at all satisfied

classification, and treatment[6-8]. Microtia is considered as a microform of CFM[7]. Intracranial abnormalities are frequent in CFM with wide variety of anomalies of the nervous system, including, cerebral hypoplasia, epilepsy, hydrocephalus, intracranial lipoma, cognitive delay, and cranial nerve dysfunction. Their incidence represents the second most common craniofacial anomaly after cleft lip and palate (between 1: 5,600 and 1: 26,550 live births). The etiology may be environmental, heritable, multifactorial, and unknown[5,11]. Many syndromes are associated with CFM, including, VATER, CHARGE, MUECS, and OEIS[12]. There are many classifications of patients with CFM, including Pruzanski[13], Kaban's modification[4], OMENS classification[5], the OMENS-Plus[14], and a modified pictorial OMENS-Plus[15].

A multidisciplinary craniofacial team is needed for comprehensive management and treatment

planning[16] with complete analysis of structure, functional requirement, craniofacial development, and other non-craniofacial associated anomalies[17].

A 3-D CT scan is helpful for pre- and post-surgical treatment planning of bony and soft tissue and outcome evaluation, focus on asymmetric hypoplasia of facial skeleton, and temporal bone. Other images may include x-rays of the cervical spine, echocardiogram, and renal ultrasound examination[3]. A CT scan of the temporal bone to assess the external, middle and inner ear structures at 5 and 6 years before microtia reconstruction is helpful to identify patient who need surgical correction to improve hearing.

Long-term protocol of Craniofacial Center is essential[3]. The treatment algorithm is divided into the neonatal period and infancy, early childhood (18 months to 3 years), childhood (4 to 13 years), and adolescence and adulthood (M >16 and F >17). Management during the neonatal and infancy period includes intubation, tracheostomy, and mandibular distraction for respiratory problem, NG tube or gastrostomy for feeding problems, and correction of macrostomia. The management in childhood includes mandibular DO, orthodontic bite block, facial bone reconstruction (s), and ear reconstruction.

Patients who are functionally affected, including mandibular hypoplasia, asymmetry or absence of ramus, condyle and temporomandibular joint fossa require a costochondral bone graft and/or mandibular distraction osteogenesis at the ages of 3 to 4 years[16,17]. Correction of the hypoplastic orbit (or distropia) should be delayed and may be considered when the patient is between the ages of 5 and 7 years with ear reconstruction at 6 to 8 years. Additionally, hearing assessment, the use of hearing aids, close monitoring for speech and language development should be performed. Definitive skeletal reconstruction, including orthognathic surgery for restored and optimal occlusion, genioplasty, facial bone reconstructions should be delayed until complete growth of the facial skeleton in adolescence and adulthood, and requires that the craniofacial team assess the orthodontic and orthognathic deformities. The reconstruction of soft tissue deficiency and asymmetry by fat injections, dermis fat graft, and free vascularized tissue transfers may be performed after the age of skeletal maturity, at the time of, or during, orthognathic surgery or other bony reconstructions[18-21].

Most patients with CFM with mandibular hypoplasia can be managed conservatively before the skeletal maturity. DO is indicated for trachea decannulation, or delayed onset/recurrent OSA in early childhood, and for Pruzanski type IIA and IIB in late childhood and it is an effective technique to reconstruct the hypoplastic mandible and establish more normal skeletal relationships in the growing child[16,22,23]. The goal of its use in growing children with CFM is to increase the vertical or supero-inferior dimension of the ramus and move the chin point to the midline, helping in improving the skeletal and soft-tissue anatomy, reducing the asymmetry early in life, and preventing or reducing the secondary adaptive deformities of the craniofacial skeleton.

There has been concern about the effects of distraction on long-term growth of the mandible in growing children and secondary compensatory growth deformities of the maxilla and zygoma, and facial asymmetry that may reduce the progressive nature of the deformity and secondary deformity[16,24]. In a longitudinal growth study, 5 and 10 years after distraction of the mandible in patients with craniofacial microsomia, the distracted ramus and the average ramus height growth rate on the affected side continued to grow favorably, and DO did not adversely affect the growth potential of the affected mandible[12]. Early DO does not, therefore, affect the growth of the affected mandible; rather it reduces the severity of the deformity, promotes psychosocial functioning, and makes secondary correction a less extensive and challenging procedure[24]. Though there are times for correction of hypoplastic zygoma and maxilla and maxillary and mandibular occlusal canting for final orthognathic treatment, the more, facial symmetry was achieved. Our study suggested that the use of DO in young children with CFM, can provide a good long-term effect on the growth of the mandible.

Comprehensive-based matrix-including health, education, livelihood, social support and empowerment initiated by the WHO following the Declaration of Alma-Ata in 1978-can be used. This strategy was promoted to improve access to rehabilitation services for people with these deformities in low-income and middle-income countries[25].

Conclusion

The current study provides a long-term, comprehensive evaluation, by an interdisciplinary team of a patient with CMS. Such CMS patients require well planned and staged reconstruction plus follow-on care by an experienced multidisciplinary craniofacial team at a craniofacial center. The use of DO in young children with CFM provides good long-term

effect on growth of the mandible which will serve as a platform for further surgical correction. A consideration of the needs and expectation of patient and family and other involved stakeholders is essential.

Limitations of the study

The was a patient report. A study with more patients would be helpful.

What is already known on this topic?

The described deformities and classification of CFM.

What this study adds?

The long-term outcome of a patient with CFM, using mandibular DO, and comprehensive management.

Acknowledgements

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Potential conflicts of interest

None.

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คราniโอเฟเชียลไมโครโซเมีย: ผลลัพธ์ในระยะยาวของการตัดยืดถ่างขยายกระดูกขากรรไกรล่างและการดูแลที่สมบูรณ์แบบตั้งแต่ในระยะแรกโดยศูนย์ตะวันฉาย

บวรศิลป์ เชาวน์ชื่น, พูนศักดิ์ ภิเศก, ปรารถนา เชาวน์ชื่น, อัมพรพรรณ ธีรานุตร

ภูมิหลัง: *คราniโอเฟเชียลไมโครโซเมียเป็นความพิการแต่กำเนิดที่ซับซ้อน เกี่ยวข้องโดยปฐมภูมิกับอวัยวะที่เจริญ จากแนวโค้งเบรเคียลที่หนึ่งและสองมีสารสนเทศที่น้อยที่รายงานการดูแลระยะยาวและผลลัพธ์ของผู้ป่วยเหล่านี้*

วัตถุประสงค์: *นำเสนอการดูแลและผลลัพธ์ในระยะยาวของผู้ป่วยคราniโอเฟเชียลไมโครโซเมีย ที่ได้รับการรักษาโดยการตัดยืดถ่างขยายกระดูกขากรรไกรล่างและแนวทางการดูแลที่สมบูรณ์แบบโดยศูนย์ตะวันฉาย โรงพยาบาลศรีนครินทร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น*

วัสดุและวิธีการ: *นำเสนอผู้ป่วย 1 ราย ที่มีภาวะคราniโอเฟเชียลไมโครโซเมีย ผลการตรวจพบทางคลินิก การประเมิน การดูแลในระยะยาวและผลลัพธ์ผู้ป่วยในวัยผู้ใหญ่ที่ได้รับการรักษาโดยการตัดยืดถ่างขยายกระดูกขากรรไกรล่างและการดูแลที่สมบูรณ์แบบ ผู้ป่วยได้รับการประเมินโดยการวัดสัดส่วนใบหน้า การสบฟัน ภาพถ่าย ภาพเอกซเรย์คอมพิวเตอร์ ภาพเอกซเรย์ด้านข้างของกะโหลกศีรษะ และแบบประเมินความพึงพอใจ*

ผลการศึกษา: *ผู้ป่วยมีการพูด การหายใจ การกลืน และการทำงานของข้อต่อกระดูกขากรรไกรที่เป็นปกติ ส่วนของใบหน้าหางตาด้านซ้ายเอียงลงทางด้านข้าง กระดูกโหนกแก้มด้านซ้ายเล็ก การสบฟันไขว้และเฉียงขึ้นด้านข้าง อ้าปากได้กว้างปกติ ผู้ป่วยมีความพึงพอใจในระดับดีมากและดีทั้งในภาพรวม รูปร่างโดยรวมของใบหน้า จมูก ริมฝีปากบน และใบหน้าด้านตรง*

สรุป: *ผลการศึกษานี้แสดงให้เห็นว่าการรักษาโดยการตัดยืดถ่างขยายกระดูกขากรรไกรล่างในเด็กที่มีคราniโอเฟเชียลไมโครโซเมีย ให้ผลลัพธ์ต่อด้านการเจริญเติบโตของกระดูกขากรรไกรล่างที่ดี ทำให้เกิดความสมมาตรที่สมบูรณ์มากขึ้นของรูปร่างใบหน้า การศึกษาให้ความสำคัญต่อการประเมินที่สมบูรณ์การดูแลระยะยาวแบบทีมสหวิทยาการและที่สมบูรณ์แบบ รวมถึงการคำนึงถึงความต้องการ ความคาดหวังของผู้ป่วย ครอบครัว และผู้ที่เกี่ยวข้อง*