Mandibular Distraction Osteogenesis in Unilateral Craniofacial Microsomia : Preliminary Report†

CHARAN MAHATUMARAT, M.D.*,
PRAYUTH CHOKRUNGVARANONT, M.D.*,
NOND ROJVACHIRANONDA, M.D.*

Abstract

At present, Ilizarov's distraction principle becomes applicable in craniofacial surgery. We would like to present a report of mandibular lengthening by distraction osteogenesis that has been performed in 4 Thai children with unilateral craniofacial microsomia in King Chulalongkorn Memorial hospital from 1996 to 1997. The distraction process was composed of latency, distraction, and consolidation phases. After the latency period, the distraction was performed by a patient's family member at home at the rate of 1 millimeter per day. Facial asymmetry and malocclusion were improved in all cases after the process was completed. No complication was experienced. No relapse or complication was detected after a mean follow-up period of 99.5 weeks. However, more cases and longer follow-up are needed before any conclusion can be made.

Key word : Mandibular Lengthening, Distraction, Osteogenesis, Ilizarov, Hemifacial, Craniofacial Microsomia

MAHATUMARAT C,
CHOKRUNGVARANONT P, ROJVACHIRANONDA N
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Since Professor Gavriil Abramovich Ilizarov's Distraction OsteoGenesis (DOG) technique has been refined to successfully lengthen endochondral bones and the surrounding soft tissue matrix for more than 50 years(1), the technique was just recently applied to the human facial skeleton(2).

* Plastic and Reconstructive Surgery Unit, Department of Surgery, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

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Table 1. The SAT multisystem classification proposed by David DJ, Mahatumarat C, Cooter R in 1987[5].

Skeletal Categories (S)
S1 = Small mandible with normal shape.
S2 = Condyle, ramus, and sigmoid notch identifiable but grossly distorted; mandible strikingly different in size and shape from normal.
S3 = Mandible severely malformed, ranging from poorly identifiable ramal components to complete agenesis of ramus.
S4 = An S3 mandible plus orbital involvement with gross posterior recession of lateral and inferior orbital rims.
S5 = The S4 defects plus orbital dystopia and frequently hypoplasia and asymmetrical neurocranium with a flat temporal fossa.
Auricle Categories (A)
A0 = Normal.
A1 = Small, malformed auricle retaining characteristic features.
A2 = Rudimentary auricle with hook at cranial end corresponding to the helix.
A3 = Malformed lobule with rest of pinna absent.
Soft-Tissue Categories (T)
T1 = Minimal contour defect with no cranial nerve involvement.
T2 = Moderate defect.
T3 = Major defect with obvious facial scoliosis, possibly severe hypoplasia of cranial nerves, parotid gland, muscles of mastication; eye involvement; clefts of face or lips.

Fig. 1. External bi-directional distractor used in this study.

Several reports later demonstrated good short-term results in patients with craniofacial pathology from various diseases[3,4].

Among congenital disorders with craniofacial asymmetry, Craniofacial Microsomia (CM) is the second most common after cleft lips and palates. Patients usually present with unilateral involvement of mandibular hypoplasia, microtia, and facial soft tissue hypoplasia. Conventional surgical correction of the mandible is delayed until the maximum of mandibular growth is gained at about 18-20 years old while most soft tissue surgery can be commenced as early as infancy. However, it is generally accepted that early restoration of the mandible will alleviate the impact of facial deformities on the child. Final dentoalveolar relationship should be better if the jaw relationship is corrected as early as possible.

This report therefore studied outcomes of the mandibular DOG technique in Thai children with the Craniofacial Microsomia (CM).

SUBJECTS AND METHOD

From 1996 to 1997 children with CM classified by SAT classification[5] were treated by the mandibular DOG technique in King Chulalongkorn Memorial Hospital (Table 1). Their parents all agreed with our treatment protocol after details, benefits, and possibilities of the outcome of the procedure had been explained.

Preoperative assessments included complete physical examination, severity of the disorder assessed by the SAT classification, cephalography, three-dimensional computerized tomographic (3D-CT) scan of facial bones, and preoperative photography. Dental occlusion was evaluated by an orthodontist of the Chulalongkorn Craniofacial Team who took care of the occlusion both in the pre-surgical and post-distraction period.

Our mandibular distractor is an external bi-directional device made in Germany (Fig. 1). Under general anesthesia with local injection of 1 per cent xylocaine and 1:100,000 adrenaline solution, intraoral incisions were made over mandibular rami to expose both outer and inner surfaces of the rami, angles, and bodies subperiosteally. The dissection

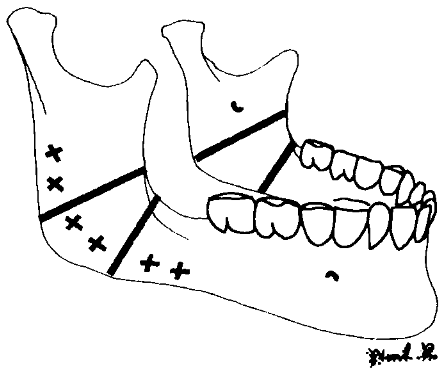

Fig. 2. Markings of osteotomy and K-wire insertion sites.

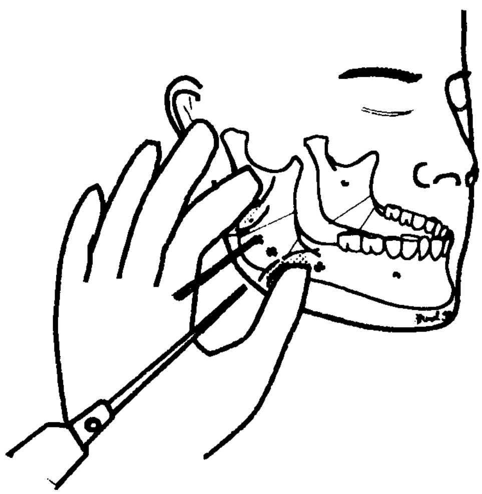

Fig. 3. Percutaneous K-wires were inserted before osteotomy, with cheek skin being pinched. Notice one K-wire which was already in place.

was always limited to only the area of expected osteotomy. Two appropriate osteotomy lines were chosen and marked both proximal and distal to the mandibular angle (ramus and body). They would divide the mandible into 3 segments (proximal, middle, and distal). Each must be wide enough for holding subsequent K-wire insertion. The distal line should be posterior to the last molar (Fig. 2).

The next step was percutaneous insertion of 2 K-wires (2.0-mm in diameter) at each segment. Position of the K-wires was very critical to achieve desired lengthening, which would lead to a good distractor axis at the time of distractor assembly. We pinched the covering skin prior to the wire insertion to compensate for future distraction. This might help to prevent high continuous tension and resulting scars at the pin sites during the distraction phase. (Fig. 3)

Outer mandibular corticotomy was then performed with a saw followed by inner corticotomy with an osteotome. Care should be taken in order to preserve the inferior alveolar nerve. We completely osteotomized the mandible after all K-wires were inserted. The distraction pins later replaced the K-wires one by one. Finally, the distraction device was assembled over the pins.

By the end of the procedure, the mandible should be in its original pre-osteotomy condition with the distractor arms and pins in place. Vector of the distractor must also be correct as planned. A closed suction drain was laid and kept in place for 24-48 hours postoperatively. (Fig. 4)

The distraction process was started between 6 to 14 days after the operation (latency phase). Rate of distraction of both ramus and body was 1 mm per day as an outpatient basis until facial symmetry and designed mandibular length were achieved (distraction phase). A single person, either the patient's father or mother, would execute the distraction after our initial instruction. Endpoint is when mandibular height and antero-posterior length were comparable to the normal contralateral side and planned occlusion was achieved. The distractor would be left in situ for another 8 weeks so as to permit bone healing of the fractured sites (consolidation phase or stabilization phase).

At the time of distractor removal, the device was disassembled from the distractor pins which were then unscrewed from the mandible. All were done under local anesthesia. Orthodontic treatment was completed to achieve good occlusion.

Our follow-up periods were 6, 12, 24, 52 weeks, and then yearly with clinical and radiological assessment of facial asymmetry, surgical scars, bony and occlusal change, and complications (Table 2).

Relapse was one of our observations. It meant radiological reoccurrence of mandibular asymmetry with resulting occlusal distortion or facial asymmetry, compared to the status at the end of the distraction and orthodontic treatment.

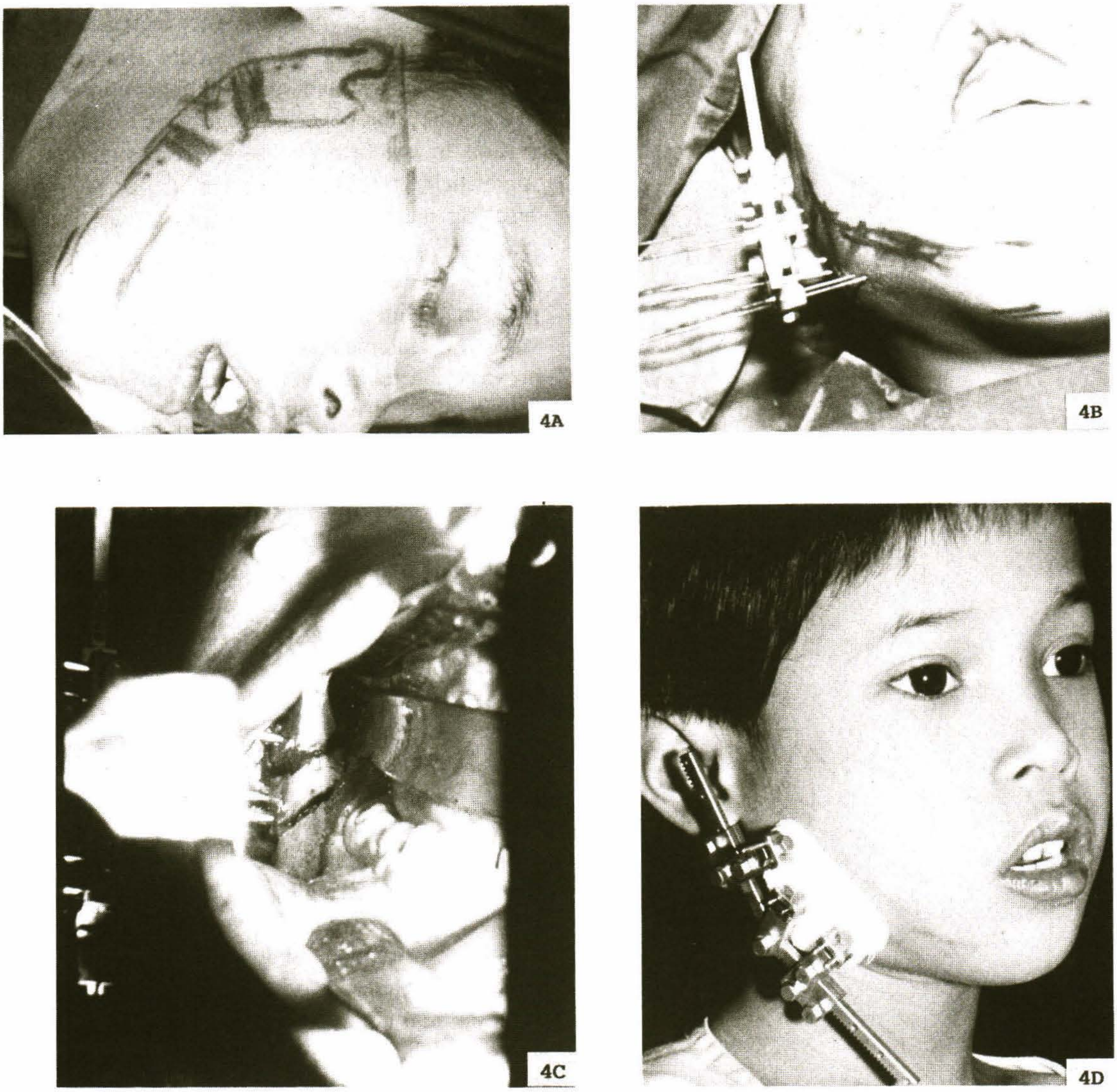

Fig. 4. **Surgical techniques A) Skin marking on the mandible. Important landmark is the mandibular angle. Observe the location for pin insertion and osteotomy at each segment (body, angle, and ramus), B) Percutaneous K-wire insertion after subperiosteal exposure of the mandible, C) 2 osteotomy lines are shown between the K-wires, D) During latency phase (1 week after the surgery, but before starting daily distraction).**

RESULTS

From January 1996 to February 1997, we treated 4 Thai patients with this technique (Table 3).

They all presented with the most common clinical findings, i.e. mild skeletal deformity, severe auricular defect, mild-to-moderate facial soft tissue defect without cranial nerve dysfunction. (Fig. 5-8)

Mean increases in the mandibular length were 12.0 mm at ramus, 9.0 mm at body, and 21.0 mm totally (Table 4).

Mean follow-up period was 99.5 weeks. No complication or relapse was found. Revision of facial scars was not needed. The sensation supplied by inferior alveolar nerve could not be evaluated in

Table 2. Complications monitored during the distraction treatment protocol.

Infection : around pin sites, operative sites, osteomyelitis
Vascular injury : hemorrhage, hematoma
Neurological injury : marginal mandibular nerve, inferior mandibular nerve
Axial deviation with resultant unplanned occlusion
TMJ location seen by X-rays
TMJ stiffness
Premature consolidation : failure of the osteotomy to open after the initiation of distraction
Delayed consolidation : absence of new bone formation
Fracture at the osteotomy site

TMJ = temporomandibular joint

Table 3. Patients included in this study (Age and severity and duration of distraction process in days)

Name	Patient 1	Patient 2	Patient 3	Patient 4
Diagnosis	Rt. CM	Rt. CM	Rt. CM	Rt. CM
Age (yr.)	7	12	13	10
SAT classification	$S_1A_3T_1$	$S_2A_3T_2$	$S_1A_3T_2$	$S_1A_3T_2$
Latency				
Average = 12.75	6	14	14	14
Distraction				
Average = 24.00	16	46	23	11
Consolidation				
Average = 72.25	62	64	89	74
Total				
Average = 109	84	124	129	99

Table 4. Increase in mandibular length after the distraction process (millimeter).

Name	Ramus	Body	Total
PN	8.0	7.0	15.0
AW	27.0	15.0	42.0
CT	7.0	8.0	15.0
AC	6.0	6.0	12.0
Avg	12.0	9.0	21.0

these children with reliability. Skeleton and occlusion have been stable since the operation.

DISCUSSION

Distraction osteogenesis (DOG) was conceived by Ilizarov in Kurgan, Siberia in 1951 when one patient accidentally turned the connection rods between rings in distraction rather than compression and he observed new bone formation radiographically[1]. Since then it has become a treatment popular in orthopedic and later craniofacial surgery. Possible advantages over conventional orthognathic osteotomy and immediate bone movement are: -

1. Simpler operative technique.
2. Less chance of bony nonunion.
3. No need for bone grafts, certainly less donor morbidity.
4. Less chance of bony relapse, theoretically, due to gradual expansion of covering soft tissue.
5. Expected less bleeding from osteotomy site because cut bone edges are usually left at original position and compacted to each other.
6. More bone lengthening can be achieved, limited only by instrument design.

In CM, distraction has been done in more than 150 patients in the last 10 years[3,4,6,7,11]. There is no report of long-term follow-up from large numbers of patients[8].

For patients with minimal bony deformity, there is no controversy in that soft tissue correction can be proceeded while the jaws are left to fully grow.

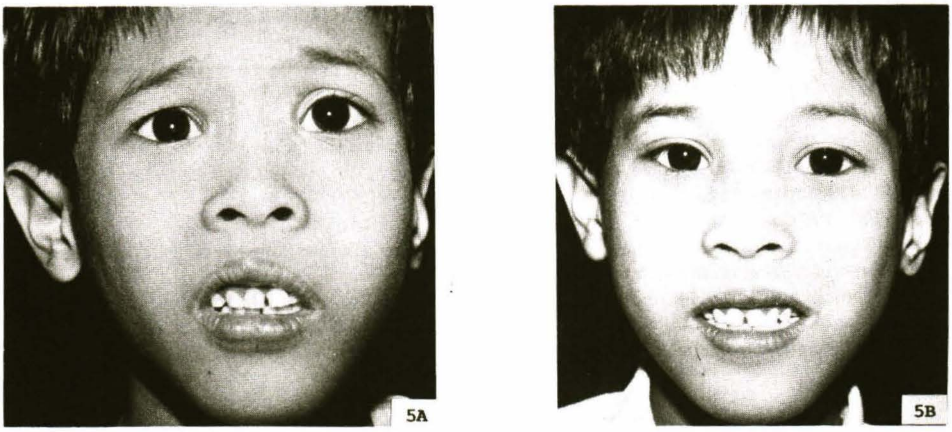

Fig. 5. **Case 1; PN: A) Preoperative AP view, B) Post distraction view.**

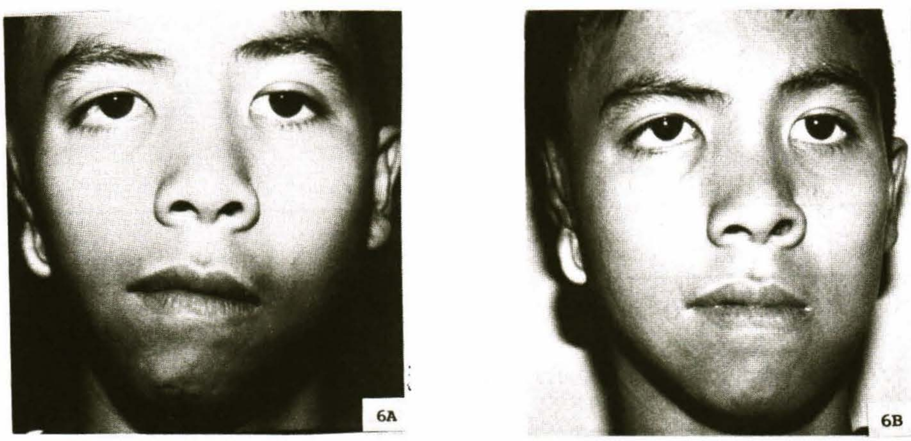

Fig. 6. **Case 2; AW: A) Preoperative AP view, B) Post distraction view.**

For untreated patients with severe mandibular deformity at an early age, abnormal bony structure will act as a malformed template for all surrounding bones and soft tissues. If this is left until bony maturation, final facial appearance, soft tissue asymmetry, mastication muscle and facial expression muscle will change in its anatomy and distorted jaw relationship will be severe and permanent. To avoid this, reconstruction of the mandibular condyle-ramus complex is necessary before bony maturation. Recommended temporomandibular joint (TMJ) reconstruction is by autogenous tissue, e.g. costochondral graft, metacarpal bone graft. DOG is claimed to be feasible in this situation by some surgeons(3,9,10) but a long-term study with more patients is required.

For patients with moderate bony deformity, as in this study, it is nevertheless controversial. The risk of growth inhibition from conventional surgery has to be balanced with benefit that will be gained from early establishment of normal bony structure and as a result, reduction in psychological stress. Moreover, there is a chance of having relapse of the deformity after early conventional reconstruction. DOG has obvious theoretical superiority, as mentioned earlier, as it can be performed even in young children with small mandible full of unerupted teeth.

There are currently 2 osteotomy techniques for mandibular distraction, incomplete osteotomy (outer corticotomy)(3) and complete osteotomy(11).

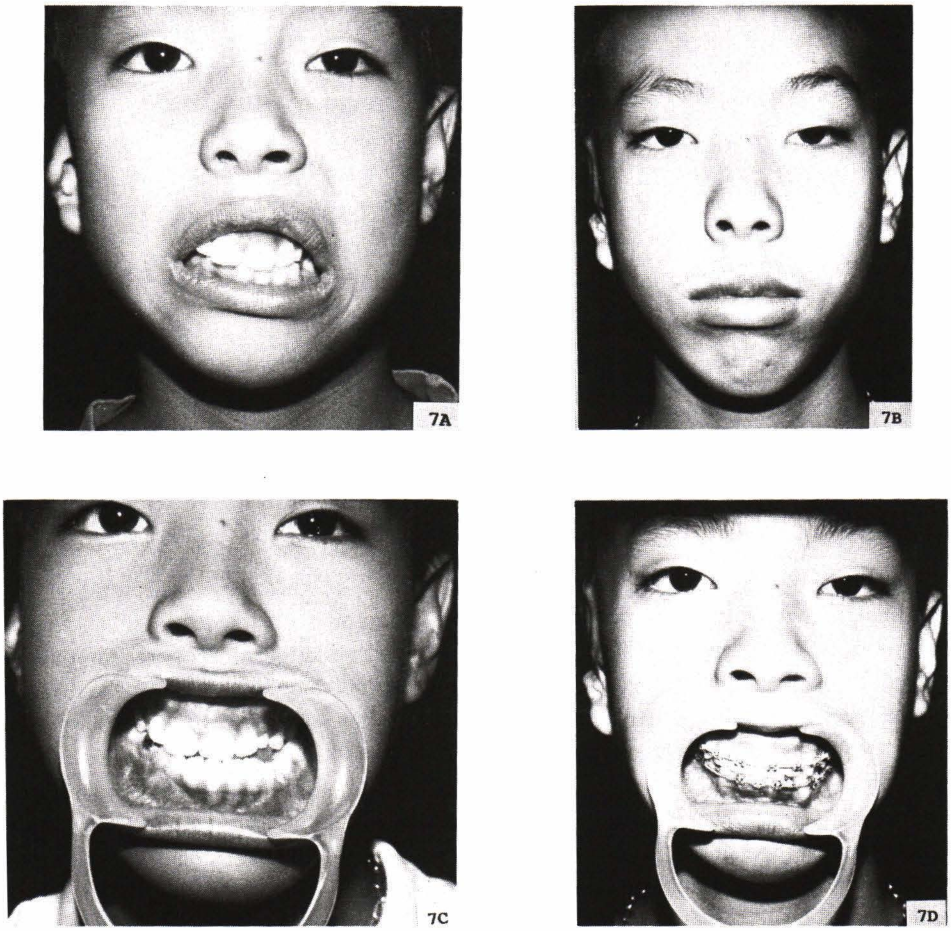

Fig. 7 **Case 3; CT: A) Preoperative view demonstrating facial asymmetry with right cheek smaller than the left, B) After distraction, face regains the symmetry with chin at midline, C) Preoperative occlusal view shows anterior open bite and midline mandibular shift to right, D) After distraction, anterior open bite is corrected with midline adjusted plus overcorrection.**

We selected the complete osteotomy technique because we believed that the basic principles of DOG must be followed to achieve a good, comparable result as in long bone distraction unless proved otherwise. We, therefore, followed a strict protocol of distraction process, from latency to consolidation phase. The distraction device must be able to stabilize the mandibular segments so that the process of bone healing can start.

To overcome the problem of lengthy treatment, and mental stress to our pediatric patients, decreasing the duration of these phases has been tried. In our opinion, the latency phase is critical to initiation of the bone healing process as it allows proper revascularization disrupted during the surgery and essential inflammatory process to complete. In trying to shorten it, local damage by the surgery has to be taken into account. Experiments in long bone distraction recommended 5-7 days latency period but also showed varied latency from 0 to 21 days. If the cortex is gently separated with preservation of the entire medullary system and periosteal sleeve, distraction can begin immediately (12).

Until now no complication has been experienced (Table 2) and no revision of facial scars was needed, but we are not going to conclude the procedure is free of problems. Though separate

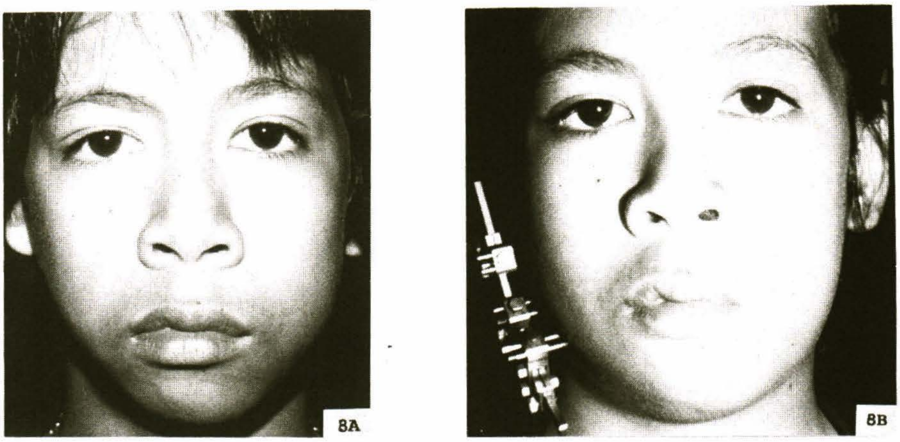

Fig. 8. Case 4; AC: A) Preoperative AP view, B) During distraction AP view.

inner and outer corticotomy were performed, the possibility of inferior alveolar nerve injury still exists and can not be checked until the patients grow up. Not mentioning the whole cost of a lengthy treatment process, the instrument itself is quite expensive and unaffordable by most patients in our society.

Changes could also be made to the distraction rate and duration of consolidation phase by the fact that healing of facial bones differs to long bones. All of these need firm scientific proof before clinical application.

The next subject to be considered is how to objectively evaluate the treatment result with accuracy. Each surgeon had his own endpoint before stopping the distraction, sometimes not clearly defined. Satisfaction at the endpoint depended on individual evaluation, from both surgeons and patients or their relatives. Previous authors have tried to measure the change of facial soft tissue landmarks(3), but in our practice we found this so subjective and invalid that it was abandoned during the study. On the other hand, tracings and measurements of bony change from radiological studies, e.g. cephalography, CT scan, are not questionable. Recently, Roth DA et al used computerized digitization of 3D CT scan to present a quantitative analysis of volumetric changes after mandibular DOG in 10 patients(13).

As a result, determination of both short- and long-term outcome is very difficult. More confusion is caused with the term "relapse". Is it the soft tissue or skeleton, reoperation or occlusal change that indicates the "relapse"? With an occlusal change, how bad will the "relapse" be. We do not think that the definition is similar in previous reports. It is our rational to use the skeletal and occlusion after the distraction and orthodontic treatment as the baseline. Worsening of this is regarded as relapse, no matter how long it happens after the surgery. The mandible and covering soft tissue are already prone to abnormal development even before the surgery. Different surgical techniques certainly affect later growth. The pathology of disease is another variable. Unilateral CM and bilateral mandibular hypoplasia as in several syndromes, e.g. Treacher-Collins syndrome, Nager syndrome, have a different natural history and background genetic drive. Mandibular growth after the distraction in these diseases cannot be assumed to be similar. The normal non-operated side of the mandible will affect final shape and size as well. Hollier LH et al found from nine children with unilateral CM that the affected side always grew at a slower rate than the contralateral side after the distraction process was complete(14).

SUMMARY

After 10 years of development, mandibular DOG is currently acceptable among craniofacial surgeons. Unfortunately, we still lack adequate experimental and clinical evidence for standardization.

Within a group of Thai children with unilateral CM of uniform deformities, mandibular lengthening by DOG could reverse the facial asymmetry to the planned skeletal and occlusal endpoint without complications, at least in the short term. However, we need more cases and longer follow-up before any conclusion can be made.

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REFERENCES

1. Maiocchi AB. Historical review. In: Maiocchi AB, Aronson J. Operative principles of Ilizarov: fracture treatment, nonunion, osteomyelitis, lengthening, deformity correction. New Design PAC. Milan: Italy, 1991: 3-8.
2. McCarthy JG, Schreiber J, Karp N, Thorne CH, Grayson BH. Lengthening the human mandible by gradual distraction. Plast Reconstr Surg 1992; 89: 1-8, Discussion 9-10.
3. Molina F, Ortiz Monasterio F. Mandibular elongation and remodeling by distraction: a farewell to major osteotomies. Plast Reconstr Surg 1995; 96: 825-40.
4. Polley JW, Figueroa AA. Distraction osteogenesis: its application in severe mandibular deformities in hemifacial microsomia. J Craniofac Surg 1997; 8: 422-30.
5. David DJ, Mahatumarat C, Cooter RD. Hemifacial microsomia: a multisystem classification. Plast Reconstr Surg 1987; 80: 525-35.
6. Carls FR, Sailer HF. Seven years clinical experience with mandibular distraction in children. J Craniomaxillofac Surg 1998; 26: 197-08.
7. Ortiz Monasterio F, Molina F, Andrade L, Rodriguez C, Sainz Arregui J. Simultaneous mandibular and maxillary distraction in hemifacial microsomia in adults: avoiding occlusal disasters. Plast Reconstr Surg 1997; 100: 852-61.
8. McCarthy JG. The role of distraction osteogenesis in the reconstruction of the mandible in unilateral craniofacial microsomia. Clin Plast Surg 1994; 21: 625-31.
9. McCormick SU, McCarthy JG, Grayson BH, Staffenberg D, McCormick SA. Effect of mandibular distraction on the temporomandibular joint: Part 1, Canine study. J Craniofac Surg 1995; 6: 358-63.
10. McCormick SU, Grayson BH, McCarthy JG, Staffenberg D. Effect of mandibular distraction on the temporomandibular joint: Part 2, Clinical study. J Craniofac Surg 1995; 6: 364-7.
11. McCarthy JG, Stelnicki EJ, Grayson BH. Distraction osteogenesis of the mandible: a ten-year experience. Semin Orthod 1999; 5: 3-8.
12. Aronson J. The biology of distraction osteogenesis. In: Maiocchi AB, Aronson J. Operative principles of Ilizarov: fracture treatment, nonunion, osteomyelitis, lengthening, deformity correction. New Design PAC. Milan: Italy, 1991: 42-52.
13. Roth DA, Gosain AK, McCarthy JG, Stracher MA, Lefton DR, Grayson BH. A CT scan technique for quantitative volumetric assessment of the mandible after distraction osteogenesis. Plast Reconstr Surg 1997; 99: 1237-47.
14. Hollier LH, Kim JH, Grayson B, McCarthy JG. Mandibular growth after distraction in patients under 48 months of age. Plast Reconstr Surg 1999; 103: 1361-70.

การเพิ่มความยาวกระดูกขากรรไกรล่าง โดยวิธียืดกระดูกในผู้ป่วยโรค Unilateral Craniofacial Microsomia

จรัญ มหาทุมะรัตน์, พ.บ.,*
ประยุทธ โชคสุ่งวรานนท์, พ.บ., นนท์ โรจน์วชิรนนท์, พ.บ.**

ในปัจจุบันการเพิ่มความยาวกระดูกด้วยการตัดแล้วยืดกระดูกโดยใช้หลักของอิลิซารอฟ (Ilizarov's distraction principle) เริ่มมีการนำมาประยุกต์ใช้ในการรักษาความพิการของกะโหลกศีรษะและใบหน้า รายงานนี้นำเสนอผลของการยืดกระดูกกรามล่าง โดยใช้วิธีการยืดกระดูกตามหลักของอิลิซารอฟ ในผู้ป่วยเด็กไทย 4 รายที่เป็นโรค Unilateral Craniofacial Microsomia ในโรงพยาบาลจุฬาลงกรณ์ ตั้งแต่ปี พ.ศ. 2539-2540 กระบวนการยืดกระดูกประกอบไปด้วยขั้นตอนต่าง ๆ ดังนี้ คือ การตัดกระดูก (osteotomy) แล้วใส่อุปกรณ์ยึดกระดูกโดยที่ยังไม่ยืดกระดูก ขั้นตอนนี้เรียกว่า latency phase ขั้นตอนต่อไปก็ทำการยืดกระดูกจนได้ความยาวตามต้องการ เรียกว่า distraction phase หลังจากนั้นรอให้มีการสร้างกระดูกใหม่เกิด ขึ้นสักระยะหนึ่งก่อนที่จะทำการเอาอุปกรณ์ยึดกระดูกออก เรียกว่า consolidation phase หลังจากผ่านระยะ latency phase แล้วการยืดกระดูกกระทำโดยบุคคลในครอบครัวผู้ป่วยเองที่บ้าน โดยหมุนเครื่องมือให้ยืดกระดูกให้ยาวขึ้นวันละ 1 มิลลิเมตร หลังจากเสร็จสิ้นทุกขั้นตอนแล้วพบว่าใบหน้าที่ไม่สมดุล และการสบฟันที่ผิดปกติมีลักษณะดีขึ้นในผู้ป่วยทุกราย โดยไม่พบภาวะแทรกซ้อน ไม่พบการกลับเป็นซ้ำ หรือภาวะแทรกซ้อน จากการติดตามผู้ป่วยต่อไปเป็นเวลาเฉลี่ย 99.5 สัปดาห์ อย่างไรก็ตามควรต้องศึกษาต่อไปในผู้ป่วยจำนวนมากขึ้นและติดตามผลการรักษาให้นานขึ้นก่อนที่จะสรุปผลการรักษาใด ๆ

คำสำคัญ : ขากรรไกรล่าง, ยืดกระดูก, Distraction, Craniofacial, Microsomia

จรัญ มหาทุมะรัตน์, ประยุทธ โชคสุ่งวรานนท์, นนท์ โรจน์วชิรนนท์
จดหมายเหตุทางแพทย์ ฯ 2544; 84: 811-820

* หน่วยศัลยศาสตร์ตกแต่งและเสริมสร้าง, ภาควิชาศัลยศาสตร์, คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย, กรุงเทพ ฯ 10330