Distraction Osteogenesis for Micrognathia in Cipto Mangunkusumo Hospital : A Case Series

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Background: Micrognathia is usually associated with genetic syndromes, characterized by mandibular hypoplasia causing a receding chin. The overall incidence of micrognathia was 1 per 1600 births, makes it a rare case. Severe micrognathia can be a neonatal emergency due to airway obstruction by the tongue in the small oral cavity. One method for correcting micrognathia is distraction osteogenesis. Lack of experience due to rare incidence of case, expensive cost of distraction device and technical complexity of the operation can be obstacles to this management.
Patient and Method: We report two cases of micrognathia corrected with distraction osteogenesis conducted in Cipto Mangunkusumo Hospital from 2011-2012. The method consists of implantation of bilateral distraction device to the inferior border of the mandibular body. The patients then followed postoperatively.
Result: Mandibular lengthening by gradual distraction is a proper method for young patients with micrognathia. Despite our minimal experience and intricate kind of method, we are trying to improve our skill in the future.
Summary: Distraction osteogenesis is one method for correcting congenital mandibular hypoplasia.
Keywords : *Distraction osteogenesis, micrognathia*
Latar Belakang: Mikrognatia adalah suatu kelainan genetik yang berupa hypoplasia mandibular. Insidens hipospadia adalah 1 dari 1600 kelahiran hidup. Mikrognatia yang parah dapat merupakan kasus gawat darurat akibat sumbatan jalan nafas oleh lidah dalam rongga mulut yang sempit. Salah satu metode untuk koreksi mikrognatia adalah distraksi osteogenesis. Hambatan dalam penanganan kasus ini adalah kurangnya pengalaman karena jumlah kasus yang sedikit, alat yang mahal dan teknik yang sulit.
Pasien dan Metode: Laporan dua kasus koreksi mikrognatia dengan distraksi osteogenesis yang dilakukan di RS Cipto Mangunkusumi dari tahun 2011-2012. Metode tindakan adalah pemasangan alat distraksi di kedua sisi mandibula.
Hasil: Pemanjangan mandibular dengan distraksi bertahap adalah metode yang tepat untuk koreksi mikrognatia.
Ringkasan: Distraksi osteogenesis adalah suatu cara untuk koreksi hypoplasia mendibula kongenital.
Kata Kunci : *Distraction osteogenesis, micrognathia*

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Micrognathia is characterized by mandibular hypoplasia causing a receding chin. Surgical management by mandibular distraction is indicated when respiratory or feeding difficulties persist despite positioning maneuvers.[1,2]

Distraction osteogenesis (DO) is a biologic process of new bone formation between the surfaces of bone segments that are gradually separated by traction. A callus forms between the separated bone segments and as long as the traction proceeds, callus tissues are stretched inducing the new bone formation.[3] DO was first introduced by Codivilla at the beginning of twentieth century and during 1950s. DO has been applied to craniofacial region since McCarthy et al reported the first clinical application of the technique in the treatment of four children with either unilateral or bilateral mandibular hypoplasia.[4]

The underlying principle of DO, as described by Ilizarov, is "the mechanical induction of new bone between bony surfaces that are gradually distracted." The process of DO begins with careful preoperative assessment and planning are performed and then the distraction device is inserted. Awaiting period (latency phase) is allowed to elapse so that osseous healing is initiated at the bony gap, periosteal integrity is restored, and callus formation begins. The bone segments at either end of the gap are then progressively distracted

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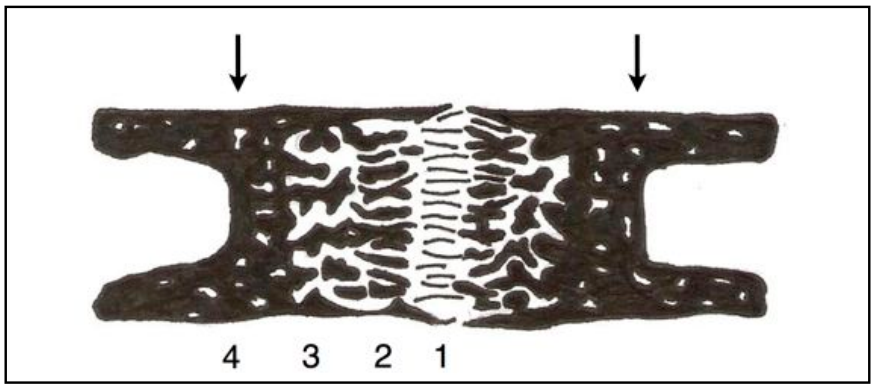

Figure 1. Schematic representation of the stages of bone formation during distraction osteogenesis: (1) zone of fibrous tissue, (2) zone of bone mineralization, (3) zone of bone remodeling, and (4) zone of mature bone.

over several days (distraction phase) during which osteogenesis is induced, producing a regenerate of immature bone laid down between the cut bone ends. With time, the bone remodels into a more mature state (consolidation phase), and the surrounding soft tissues accommodate to their new positions and lengths.[5,6]

During the distraction phase, bone formation occurs in response to the tension-stress forces exerted on the regenerate, and healing proceeds primarily by a reparative membranous ossification process. The middle of the regenerate consists of a fibrous central zone where osteoid is deposited with collagen fibers oriented parallel to the direction of distraction (Figure 1). Ossification occurs as a primary mineralization front advances from either end of the fibrous central zone, resulting in a bridge of immature bone across the distraction gap. Although the volume and architecture of the new bone are comparable to the adjacent bones, animal studies have shown that mineral content and radio density are less. In addition to bony changes, there are effects on the adjacent soft tissues that occur in response to osseous distraction. Muscle and soft tissue mass increases via a process referred to as distraction histogenesis. Clinically, this offers a distinct advantage since several craniofacial anomalies have soft tissue hypoplasia in addition to deficient bony structures. Neurovascular elements contained within distracted bony segments are also stimulated to elongate.[5,6]

PATIENT AND METHOD

In this case report, we intended to present the treatment of two patients with micrognathia conducted in Cipto Mangunkusumo Hospital from 2011-2012.

The first patient was a 9-months-old boy (Figure 4a,b). The second patient was a 7-months-old girl (Figure 5a,b). Both patient admitted to the plastic surgery division for facial cleft no. 7 (Bilateral Goldenhar Syndrome). Bilateral macrostomia reconstruction was done in both patient, but the patient still presented with micrognathia which cause their difficulty in swallowing. Therefore, we conducted the distraction osteogenesis for fixing the micrognathia for both patients.

An internal resorbable mandibular distractor device was used bilaterally and the patients then followed postoperatively.

Surgical Procedure

The surgical procedures conducted as follows: (1) Bilateral incisions 2 cm below the angle of the mandible were carried out and scissor dissection below the level of the platysma was performed until the inferior border of the mandible was reached. (2) Choose the appropriate size distractor and drive-screw extension for the patient. Using the drive-screw extension, determine the desired vector of distraction then mark the proposed osteotomy. Insert drive-screw in desired vector and thread drive-screw into distractor (Figure 2a,b). (3) Fixate the distractor directly to the bone with the drive-screw extension in place. (4) Back the drive-screw extension out of the distal plate so the osteotomy can be performed. To performed monocortical osteotomies, begin with a reciprocating saw and finish with an osteotome. (5) The osteotomy is carefully completed with an osteotome and a bone spreader. Neurovascular bundle is preserved, care is taken to slowly

divide the cortices circumferentially, gently spreading the bone edges (Figure 2c). (6) Once the osteotomy has been completed the drive screw is threaded through the proximal plate into the receiving compartment of the distal plate and the device is activated at least 5.0 mm

The devices were activated after latency period of 4-7 days. In the following days, distraction continued 1 until 2 mm/day at each side depends on amount of distraction needed. The devices were removed following the consolidation period of 6-12 weeks.[7,13,14]

There are three main phases to distraction osteogenesis: latency, activation, and consolidation. Latency is the period immediately following the osteotomy and application of distractor; it ranges from 1 to 7 days. After the latency phase is the activation phase. During this phase, the distraction device is activated by turning some type of axial screw, usually at 1 until 2 mm/day. Once activation is completed, the third and final phase is the consolidation phase. Typically, the consolidation phase is twice as long as the time required for activation.[6]

RESULT

In the first patient, one year after mandibular distraction, micrognathia is still noted (Figure 4c,d). In second patient, after

Figure 2. Incisions, dissection below the level of the platysma and determine the desired vector of distraction.

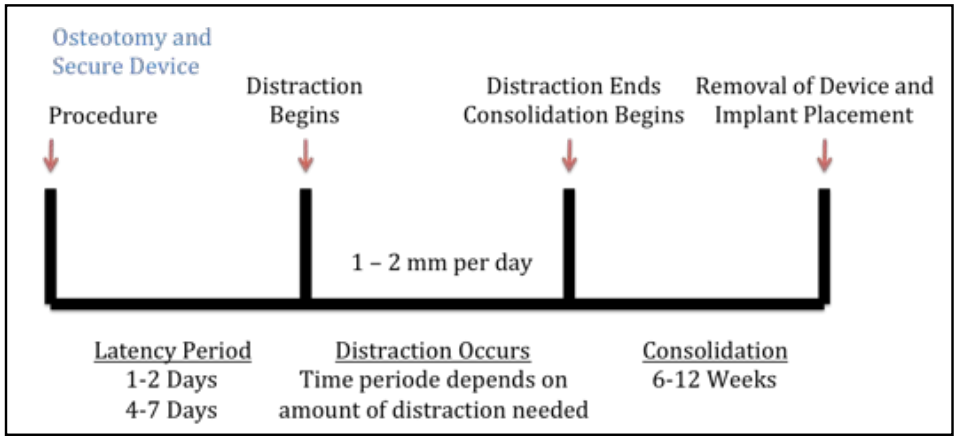

Figure 3. The protocol of distraction

Figure 4. Figure of the first patient. **Left:** Pre-operative condition, anterior view. Patient presented with micrognathia. **Right:** Postoperative anterior and lateral view one year after mandibular distraction. Micrognathia is still noted in this patient.

Figure 5. Figure of the second patient. **Left:** Pre-operative condition, lateral view. Patient presented with micrognathia. **Right:** Postoperative lateral view after mandibular distraction.

mandibular distraction patient was at the consolidation phase (Figure 5c,d). Facial scarring in both patients are cosmetically acceptable and do not need further revision.

DISCUSSION

There is no data about incidence of micrognathia in Indonesia. The overall incidence of micrognathia in a study population evaluated and delivering at one institution in Michigan, USA was 1 per 1600 births, makes it a rare case. Based on statistic data of Indonesian population, Crude Birth Rate on 2010 was 18,4, so it can be generally concluded that incidence of micrognathia was approximately about 4308 in a year.[1,8]

Previous studies done by Schaefer et al, have proposed a mandibular-maxillary discrepancy greater than 8 to 10 mm as an indication for surgical management, although all aspects of examination and diagnostic studies should be included in the decision to proceed with surgical procedures.[2,9] In this case series, feeding difficulties are the surgical indication for both patients.

Reported complications from the use of mandibular Distraction Osteogenesis (DO) in children have included penetration of the floor of the mouth with a pin or loosening of a pin after a fall, development of an abscess at the pin

site, inadequate distraction requiring a second DO procedure, and facial scarring requiring revision.[7]

In this case series, first patient showed relapse on his micrognathia due to growth rapidity of his mandibula unable to match the growth of his maxilla. Therefore, a secondary DO is needed. The second patient showed a good improvement, patient is still in consolidation phase. The follow-up period for the second patient was too short to allow definitive conclusions, and for this reason, long term observation is necessary Facial scarring in both patients are cosmetically acceptable and do not need revision.

One of the difficulties of distraction osteogenesis, however, is that accurate positioning of the proximal segment can be difficult to achieve either because of an inaccurate displacement vector or because of an unpredictable soft tissue influence on the immature regenerate. It has been shown in an animal model and in clinical case reports that post-distraction regenerate can be molded by external forces. Huisinga-Fischer et al., in their 3-year follow-up study, claimed that 50 percent of cases showed relapse at the end of the first year and that this relapse had a progressive character when studied at 3 years after distraction.[10] The soft tissue is known to remodel around newly formed bone. This occurs over an undetermined period of time. However, studies have shown that suprahyoid muscle complex forces play lengthening in up to 1 year of follow-up. After removal of the distraction device at the end of the consolidation period of 8 weeks, the soft tissue and muscle continue to exert posteriorly directed forces on the distracted bone.[11,12]

In general, the procedure of distraction osteogenesis still a difficult technique. The problems that occurred in our hospital are lack of experience due to rare incidence of case, expensive cost of distraction device and technical complexity of the operation which can be obstacles to this management.

Although our surgical experience is still limited, here in Cipto Mangunkusumo hospital, we are trying to improve it to overcome those difficulties.

SUMMARY

Distraction osteogenesis is a good technique for correcting micrognathia. However we find some difficulties: (1) lack of experience due to rare incidence of case, (2) adaptation to new instrumentations, (3) expensive cost of distraction device, (4) need more extensive cooperation with other specialty such as orthodontics, pediatrics, that can determine the result.

In summary, distraction osteogenesis provides a powerful and reliable technique for providing well-vascularized bone in mandibular reconstruction for micrognathia.[6] It is an effective technique for treating micrognathia. However, it must be borne in mind that the successful of this technique need a lot of factors, from preoperative (preoperative planning and extensive cooperation with orthodontics), intraoperative (adaptation to new instrumentation and doing the osteotomy monocortically) and postoperative (evaluation, cooperation with other specialty such as orthodontotics and paediatrics, and also parents involvement).

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