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# Multidirectional cranial distraction osteogenesis technique for treating bicoronal synostosis

Ataru Sunaga [a, b, *], Yasushi Sugawara [c], Akira Gomi [d], Daekwan Chi [b], Hideaki Kamochi [e], Hirokazu Uda [b], Kotaro Yoshimura [b]

[a] Department of Pediatric Plastic Surgery, Jichi Children's Medical Center Tochigi, 3311-1, Yakushiji, Shimotsuke, 329-0498 Tochigi, Japan
[b] Department of Plastic Surgery, Jichi Medical University, 3311-1, Yakushiji, Shimotsuke, 329-0498, Tochigi, Japan
[c] Lilla Craniofacial Clinic Tokyo, 1-7-17, Ginza, Chuo-ku, 104-0061, Tokyo, Japan
[d] Department of Pediatric Neurosurgery, Jichi Children's Medical Center Tochigi, 3311-1, Yakushiji, Shimotsuke, 329-0498, Tochigi, Japan
[e] Department of Plastic Surgery, Shizuoka Children's Hospital, 860, Urushiyama, Aoi, Shizuoka, 420-8660, Shizuoka, Japan

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ABSTRACT

Fronto-orbital advancement by distraction osteogenesis is a useful means of surgically correcting bicoronal synostosis. However, the scope for morphological revision is limited. To address this issue, we developed a multidirectional cranial distraction osteogenesis (MCDO) technique that we quantitatively assessed in patients with bicoronal synostosis.

In this case series, five patients with bicoronal synostosis were treated with MCDO at a mean age of 13.4 months (range 9–22 months). Distraction started 5 days after surgery and the activation period was 11.2 days (range 10–14 days). The distraction devices were removed 47.2 days (range 33–67 days) after completing distraction. Improved cranial shape was confirmed by CT data. Mean preoperative CI, APL, and ICV readings of 102.1%, 13.5 cm, and 1179.4 ml, respectively, had reached 94.0%, 14.9 cm, and 1323.9 ml, respectively, upon device removal. These values were well preserved at 1 year (90.4%, 15.8 cm, and 1461.3 ml, respectively).

In conclusion, MCDO successfully enables both cranial expansion and correction of a flat forehead, constituting a valid treatment alternative for patients with bicoronal synostosis.



## 1. Introduction

Current surgical treatments for bicoronal synostosis remain controversial. Although one-stage, fronto-orbital advancement (FOA) is widely used (Jane and Persing, 2000), anterior–posterior relapse is problematic (Lwin et al., 2011). In addition, avascularized bony pieces, dead space between reshaped bone segments, and dura mater can increase the likelihood of extradural abscess, late resorption, and postoperative bone defects.

The development of fronto-orbital advancement by distraction osteogenesis (FODO) (Sugawara et al., 1998), a method entailing internal distraction, which some craniofacial surgeons have applied to bicoronal synostosis (Jeong et al., 2016; Mundinger et al., 2016; Nonaka et al., 2004; Satoh et al., 2015), offers several advantages over one-stage FOA. The incremental gains in intracranial volume (ICV) afforded by its gradual expansion of the cranium and soft-tissue envelope are far greater. Because dural attachments to bone flaps are preserved, leaving the blood supply intact, early postoperative bone formation and proper cranial growth are anticipated, as well as a reduced risk of extradural abscess.

Despite various advantages over FOA, the limited capacity of FODO to direct the movement of bone flaps is clearly a drawback. Movement occurs solely along the axis of the distraction device; thus, correcting a flat forehead is generally difficult. In fact, FODO could exacerbate such distortions in patients with open metopic sutures and anterior fontanels (Fig. 1).

To overcome this disadvantage, we have developed a new distraction osteogenesis technique for the treatment of craniosynostosis. Our multidirectional cranial distraction osteogenesis (MCDO) method is valid for all phenotypes of craniosynostosis, offering a means of achieving any desired cranial shape (Sugawara et al., 2010; Gomi et al., 2016; Sunaga et al., 2017a,b). Here, we present a series of five patients with bicoronal synostosis treated by

* Corresponding author. Department of Pediatric Plastic Surgery, Jichi Children's Medical Center Tochigi, 3311-1, Yakushiji, Shimotsuke, 329-0498, Tochigi, Japan.
*E-mail address:* atarusuna@me.com (A. Sunaga).

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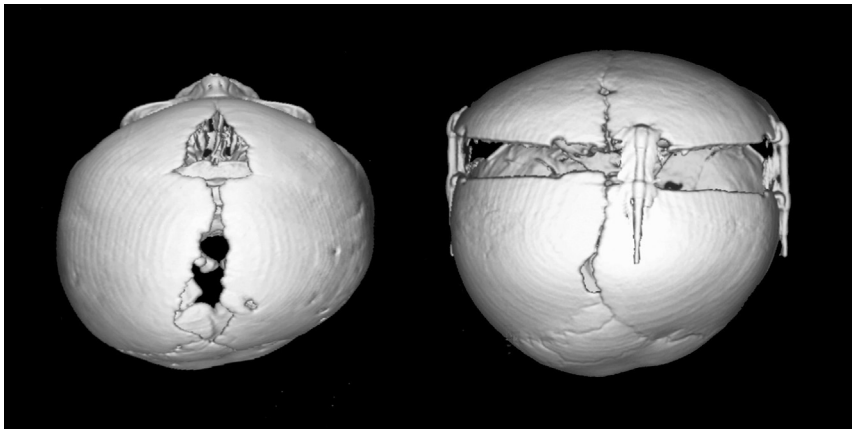

**Fig. 1.** 'Gullwing deformity' resulting from FODO.

MCDO, analyzing procedural efficacy through quantitative cranial assessments.

## 2. Case series

### 2.1. Patient selection

From 2013 to 2014, five patients (mean age 13.4 months; range 9–22 months) presenting with bicoronal synostosis underwent MCDO at Jichi Children's Medical Center Tochigi (Table 1). Two patients were nonsyndromic, but the other three patients had Apert syndrome. All were clinically diagnosed with bicoronal synostosis, confirmed by preoperative three-dimensional (3D) CT scans.

### 2.2. Surgical technique

The MCDO surgical procedure has been previously described (Sugawara et al., 2010). Briefly, the anterior cranium is osteotomized to produce 6–8 rectangular or triangular pieces and a supraorbital bar, using an ultrasonic bone scalpel (Sonopet; Stryker Corp, Kalamazoo, MI, USA) to facilitate safe osteotomy without dural tears. Individual bones (except for the supraorbital bar) are not dissected from the dura, leaving their vascularity intact. Traction pins are placed in each segment of bone. The frame is fixed on anchor pins set in temporal bones after wound closure. Wires secured in the traction pin-holes are then passed through openings in the frame, fixing them to frame-based distractors (Fig. 2).

Five days after surgery, distraction is started at a rate of 1.5 mm/day. This rate is later modified in accord with 3D CT scan data generated on postoperative day 12.

In our patients, desired skull shapes were generally achieved within a 14-day activation period. Because bony segments were closely approximated, early bone formation and fusion were fully expected. Thus, the frame and all pins could be removed under sedation after a relatively brief consolidation period of around 6 weeks.

### 2.3. Quantitative assessment

Consistent with the clinical practice protocol of Jichi Medical University, all patients underwent 3D CT scans preoperatively, upon device removal (postoperative month 0), at 6 months, and at 1 year. Pre- and postoperative cranial morphology was chronicled by recording cranial index (CI), anteroposterior cranial length (APL), and intracranial volume (ICV) values. The latter were plotted with the ICV growth curve for normal Japanese children (Kamochi et al., 2017). All measurements were obtained using a DICOM image viewer (OsiriX; Pixmeo, Bernex, Switzerland).

### 2.4. Results

There were no major complications within the follow-up period. In one patient, a minor infection developed at the anchor pins during the consolidation phase, but was resolved with antibiotic therapy. Anchor pins loosened in one patient, prompting accelerated device removal. Operative times ranged from 191 to 290 min (mean 230.8 min) and transfused blood volume ranged from 18.1 to 35.9 ml/kg body weight (mean 25.9 ml/kg). Mean postoperative hospital stay was 16.4 days (range 15–17 days). Planned distraction programs were completed in all patients, with activation phases ranging from 10 to 14 days (mean 11.2 days), and consolidation periods ranging from 33 to 67 days (mean 47.2 days) (Table 1).

Cranial shapes in all patients showed improvement (Figs. 3–5). CI declined from baseline (mean 102.1%), averaging 94.0% upon device removal, 91.5% at 6 months postoperatively, and 90.4% at 1

**Table 1**
Patient data.

| Case No. | Age (months) | Sex | Syndrome | Operating time (min) | Bleeding (ml) | Blood transfusion per body weight (ml/kg) | Complication | Activation period (days) | Consolidation period (days) | Hospital stay (days) |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | 10 | F | Nonsyndromic | 290 | 30 | 31.6 | None | 11 | 33 | 16 |
| 2 | 9 | M | Nonsyndromic | 214 | 60 | 18.1 | Minor infection | 14 | 67 | 19 |
| 3 | 13 | F | Apert | 224 | 280 | 35.9 | None | 10 | 41 | 15 |
| 4 | 13 | F | Apert | 191 | 250 | 21.4 | None | 10 | 55 | 15 |
| 5 | 22 | M | Apert | 235 | 80 | 22.6 | Anchor pins loosening | 11 | 40 | 17 |
| Ave | 13.4 | | | 230.8 | 140 | 25.9 | | 11.2 | 47.2 | 16.4 |

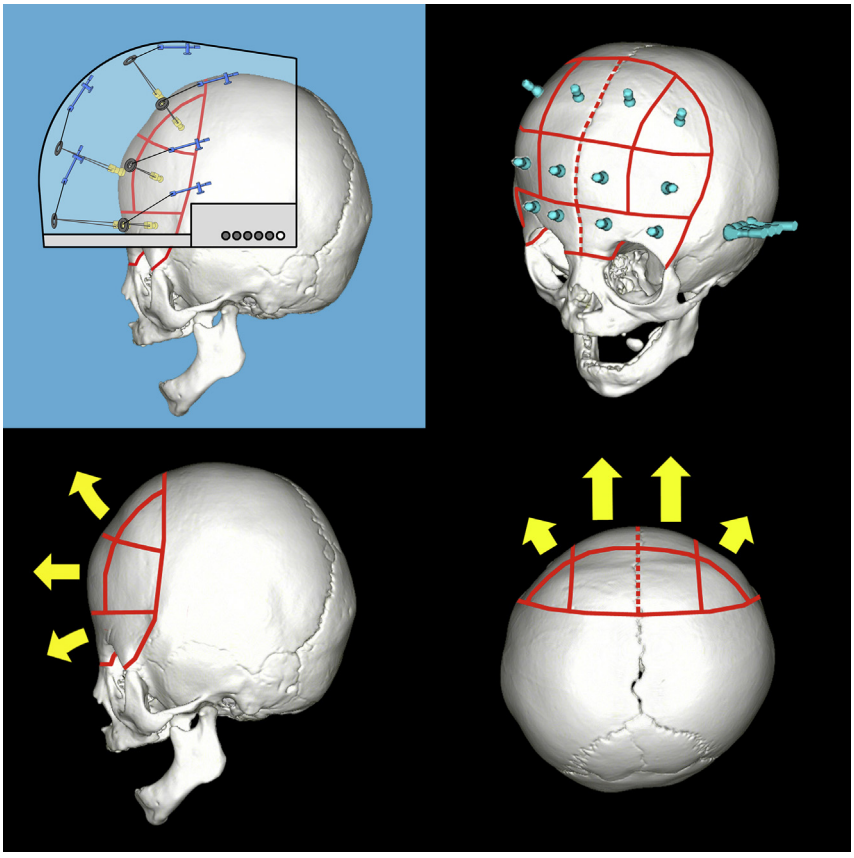

**Fig. 2.** The schemas of MCDO. Vertical distraction of each bone flap enables remodeling of the skull to the desired shape.

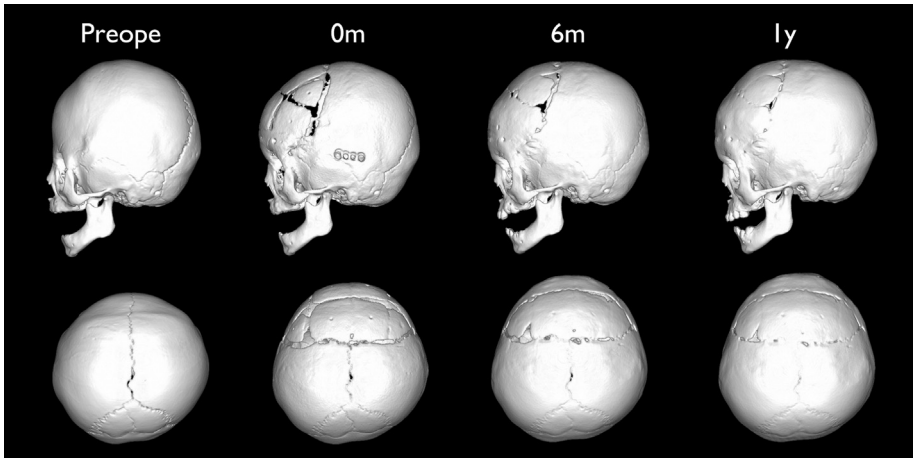


**Fig. 3.** Pre- and postoperative three-dimensional computed tomographic images of Patient 1.

year. APL and cranial width exceeded baseline levels (means: APL 13.5 cm; width 13.8 cm), averaging 14.9 cm and 14.0 cm, respectively, upon device removal, 15.5 cm and 14.2 cm at 6 months, and 15.8 cm and 14.3 cm at 1 year. ICV also increased from baseline (mean 1179.4 ml), averaging 1323.9 ml upon device removal, 1410.6 ml at 6 months, and 1461.3 ml at 1 year (Table 2). Comparison with the normal ICV growth curve for Japanese children showed that postoperative cranial growth was not hindered, at least in the first year (Fig. 6).

*2.4.1. Patient 1*

A 9-month-old boy with nonsyndromic bicoronal synostosis presented with brachycephaly and a flat forehead (CI, 104.2%). MCDO was uneventfully performed (operative time 214 min). Distraction was initiated 5 days after surgery and completed on postoperative day 19. During the consolidation period, a minor infection developed at the site of the anchor pins, which resolved following antibiotic treatment. All devices were removed 67 days after finalizing the distraction. There was a clear improvement in forehead contour, and the corrected CI (97.1% at device removal) continued to progress (93.0% at 1 year) (Fig. 3).

*2.4.2. Patient 2*

A 13-month-old boy presented with Apert syndrome. A subsequent 3D CT scan showed bicoronal synostosis with open anterior fontanel and metopic suture (CI, 103.3%). The MCDO procedure was

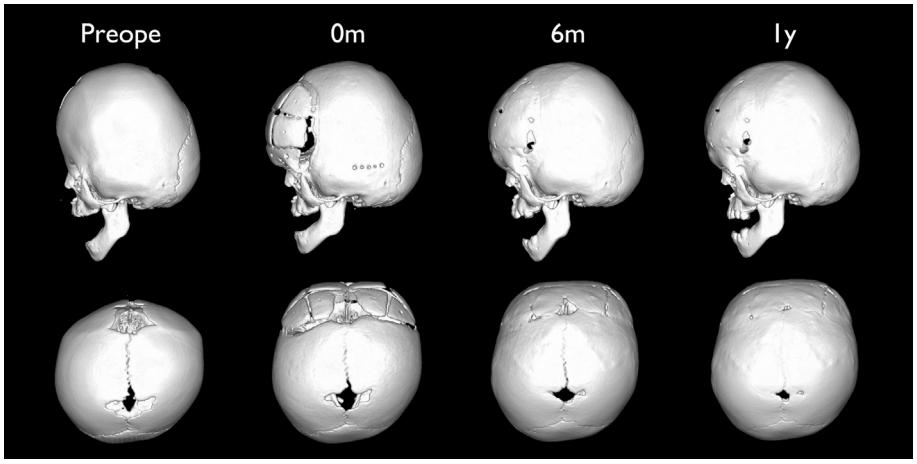


**Fig. 4.** Pre- and postoperative three-dimensional computed tomographic images of Patient 2.

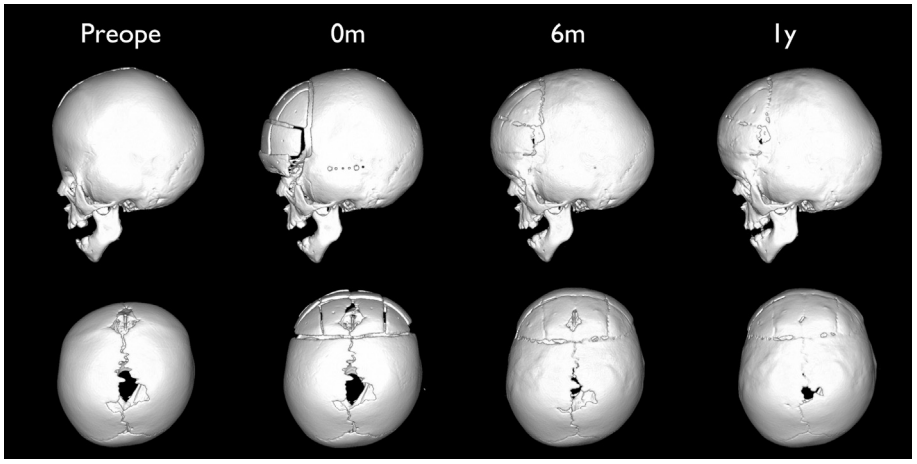


**Fig. 5.** Pre- and postoperative three-dimensional computed tomographic images of Patient 3.

**Table 2**
Pre- and postoperative cephalic indexes, anterio-posterior lengths of the cranium, and intracranial volumes. Values are means (standard deviations).

| | Pre | 0 m | 6 m | 1 y |
|---|---|---|---|---|
| CI | 102.1 | 94 | 91.5 | 90.4 |
| AP distance | 13.5 | 14.9 | 15.5 | 15.8 |
| ICV | 1179.4 | 1323.9 | 1410.6 | 1461.3 |

performed (operative time 191 min), with an uneventful postoperative course. Distraction started 5 days after surgery and was completed on postoperative day 15. All devices were removed 55 days after distraction was finalized. The CI (93.5% at device removal) was even better at 1 year (91.5%).

*2.4.3. Patient 3*

A 22-month-old boy with Apert syndrome showed bicoronal synostosis, with open anterior fontanel and metopic suture. MCDO was performed (operative time, 235 min). During the consolidation period, the patient accidentally fell, loosening the anchor pins. As a result, the device was removed prematurely, 40 days after completing distraction. Nevertheless, a good cranial shape was achieved and sustained at 1 year postoperatively (Fig. 5).

## 3. Discussion

The surgical goals for bicoronal synostosis are to expand the cranium in the anteroposterior plane and correct a flat forehead. Although FOA is sufficient for most cases of non-syndromic craniosynostosis, the extent of expansion is limited, and postoperative relapse frequently occurs due to the high tension of the skin envelope. According to Lwin et al. (2011), 65% of patients undergoing FOA experience anteroposterior relapse, with 48% of operative advancement lost in the first 5 months.

On the other hand, distraction osteogenesis serves to minimize postoperative relapse owing to bone regeneration during the consolidation period. Quantitative analysis has shown that there is no relapse after FODO (Yamaguchi et al., 2014). In our case series, not only was anteroposterior relapse not observed after MCDO, but declines in CI also continued after device removal, and cranial growth potential was unhindered in the first postoperative year.

The MCDO technique has several advantages over FODO. Most importantly, each bone flap can be moved in any desired direction, as opposed to the single direction in FODO (i.e., along the axis of the distractor). Consequently, the contour of a flat forehead can be improved via MCDO by distracting multiple segments centrifugally, even in the presence of open metopic sutures or anterior fontanels. Additionally, the bone flaps lie in closer approximation, so that earlier bone formation and fusion are likely, enabling a shorter

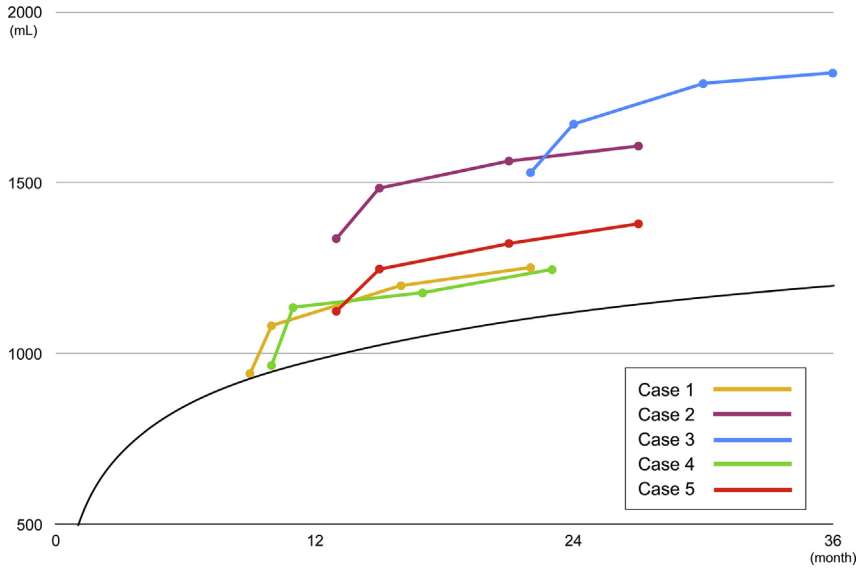


**Fig. 6.** Pre- and postoperative intracranial volumes. The black line depicts the normal ICV growth curve for Japanese children.

consolidation period. The reported average consolidation period for FODO procedures is 10 weeks (range 6.8–32.5 weeks) (Mundinger et al., 2016), whereas 6 weeks of consolidation proved sufficient to avoid relapse in our patients. Finally, the distraction devices for MCDO are easier to remove than are the internal distractors of FODO. Removal is achieved under sedation and without any incision, typically within 5 min.

The major limitation of this investigation is the small number of patients involved. Although satisfactory results were obtained in all five subjects, a greater number of patients and long-term monitoring are essential to validate our approach.

## 4. Conclusions

Our results suggested that MCDO enables both cranial expansion and correction of a flat forehead, constituting a valid treatment alternative in instances of bicoronal synostosis.

## Conflicts of interest statement

The authors declare no conflicts of interest.

## Financial sources

No financial sources were sought or provided for this project.

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