Rotating Distraction Osteogenesis in a Child with Secondary Craniosynostosis

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Abstract: Distraction osteogenesis has been acknowledged as a most effective surgical method to maximize intracranial space in patients with craniosynostosis by progressive displacement of the skull flap. However, it is difficult to control the growth direction of the skull flap even with application of four distraction bars. The authors of this study performed a cranioplasty procedure in a four-year-old girl with brachicephalic microcephaly, who showed a high lumbar puncture pressure of 21 cm H$_2$O. After bilateral frontal craniectomy, the frontal bone and the midline of the superior orbital bone were fixed with a single wire, thus converting the linear displacement of distraction to a rotation flap and raising the skull flap superiorly. This rotational distraction osteogenesis resulted in a longer distance of distraction and upward rotation of the frontal skull flap with only two distraction bars. The authors suggest that rotating distraction osteogenesis has several merits: lesser depression of the bone flap, longer distance of distraction, and it is simpler and more economic than linear distraction. We also advocate that more aggressive surgical procedures should be considered for even older children with high intracranial pressure.

Key Words: Distraction osteogenesis, craniosynostosis, intracranial pressure

Although various procedures regarding cranial vault remodeling have been indicated in infants or children with craniosynostosis; in order to increase intracranial space and to produce a better aesthetic effect, it is difficult to increase the cranial size more than 2 cm due to the limits of the scalp pressure. Distraction osteogenesis starting from the mandible has been introduced in patients with craniosynostosis, and is at present being widely employed because of the relative simplicity of the procedure and larger increase of the intracranial space compared to previous methods. Instability during the distraction process from discrepancy between the axis of the distraction bar and the scalp pressure, resulting in depression of the bone flap and shorter distance of distraction, have been observed in many patients. The authors attempted to overcome these problems by replacing the linear distraction with rotational distraction and observed good results after application of this procedure to a patient with craniosynostosis.

CLINICAL REPORT

A four-year-old girl presented to our surgical center with microcephaly and developmental delay. Physical examination showed that her head circumference was 45 cm (less than third percentile), and the lumbar puncture showed a cerebrospinal pressure of 21 cm H$_2$O. She had a previous history of general seizures and had been diagnosed with congenital injury of the right hemisphere and craniosynostosis by computed tomography (CT) when she was one month old. She received rehabilitation therapy without any surgery and had showed some movement limitation, especially fine movement of the left upper extremity, and a low intelligence quotient of 70 with passive characteristics.

The operation findings at our hospital demonstrated that the frontal lobe forcefully protruded forward just after frontal craniectomy, suggesting high intracranial pressure from the craniosynostosis. A frontal free bone flap was fixed to the midline of the frontal bone for rotation with a 0.015 inch wire. Two distraction bars were fixed to a point 3 cm lateral from the midline of the frontal bone flap and parietal skull bilaterally (Fig 1A). Postoperatively, a distance of 1 cm appeared between the skull flap and the parietal skull (Fig 1B) resulting in increased intracranial volume, which was almost completely filled by expansion of the brain. We distracted the frontal bone flap for 0.6 mm everyday for 2 months beginning 1 month after the operation; after a further consolidation phase of 3 months, the resulting displacement was 32 mm on the midline (Fig 1C).

Follow-up CT at the second and fourth postoperative month showed increased intracranial space and sufficient expansion of the underlying brain and only small free epidural space. We removed the two distraction bars 6 months after initial establishment of distraction system. The CT also showed proper ossification in the distracted area (Fig 1 D). The patient showed good cranial contour aesthetically with upward elevation and forward displacement of the preoperatively depressed frontal skull (Fig 2A,B) and her parents were also satisfied with this result.
At 12 months postoperatively, she showed a slightly elevated intellectual quotient of 80, and her character changed from passive to active resulting in improved learning and rehabilitation exercise without any unexpected postoperative complications.

**DISCUSSION**

There are two major management modalities of craniosynostosis: tunnel craniectomy and cranial vault reshaping by various methods. Tunnel craniectomy is not as effective when the bivalved skull growth does not follow progressively with brain growth, and results in retardation of brain growth or bulging out of the brain through the craniectomy site. However, tunnel craniectomy is simpler, safer, and more economic compared to cranial vault reshaping, especially with the introduction of the endoscopic craniectomy method. On the other hand, cranial vault reshaping has the advantage of immediate expansion of the intracranial volume irrespective of brain growth. Cranial vault reshaping may be a more suitable method for patients with low-pressure brain growth such as syndromic craniosynostosis, craniosynostosis with ventriculo-peritoneal shunt, and secondary craniosynostosis after shunting. Although cranial vault reshaping results in immediate expansion of the intracranial volume, the overexpanded skull may then exert increased tension.
Fig 2 (A) Initial preoperative photography of patient shows depressed frontal skull and small head. (B) Postoperative photography shows elevated frontal skull and larger head 8 months after initial operation.

on the scalp, resulting in breakeage of the cranial fixation and increased chances of infection in the epidural dead space during the prolonged period it takes for the brain expansion and growth to fill the cranium.\textsuperscript{4} Infection rates are increased especially in syndromic patients or older children who show slow expansion or growth rate of the brain.\textsuperscript{5} This limits the length of skull expansion to 1–2 cm per single operative cranial vault reshaping that does not relieve the elevated intracranial pressure sufficiently.\textsuperscript{1,6} Cranial vault reshaping can also result in either poor growth of the skull and brain as a consequence of the fixated skull, or good brain growth recovery after brain expansion or filling of the skull dead space.

Distraction osteogenesis incorporates several advantages from tunnel craniectomy and cranial vault reshaping: skull expansion is slow and progressive permitting the scalp to lengthen sufficiently, resulting in absence of skull flap breakage or scalp necrosis due to decreased tension on the scalp.\textsuperscript{7,8} In addition, slow and progressive expansion of the skull flap results in only a small extradural dead space and therefore low infection rates.\textsuperscript{5} Also, it is not necessary to dissect the underlying dura from the skull for distraction, which simplifies the operation and lessens bone flap resorption.\textsuperscript{5,9} Interestingly, usual cranial vault reshaping results in remnant epidural dead space while distraction osteogenesis without dissection of the underlying dura may produce a subdural dead space. These differences could be important for the management of the slowly growing brain with low expansile force, such as in older children or a child with shunted or atrophied brain. Distraction osteogenesis may permit the brain to grow and expand under more favorable circumstances such as a large space, but also a lower resultant intracranial pressure compared to the epidural decompression after cranial vault reshaping. Therefore, operative management by distraction osteogenesis may be more appropriate for older children with craniosynostosis, or for children with shunted or atrophied brain.

However distraction osteogenesis has some disadvantages such as limited initial reshaping, and the necessity for additional operation for removal of the device.\textsuperscript{7} Cohen et al.\textsuperscript{10} advocated narrower range of indications for distraction osteogenesis in 1998. They proposed this procedure in patients with syndromic craniosynostosis who have showed slow brain expansion or growth, rather than for patients with non-syndromic craniosynostosis. Another disadvantage could be an infection problem through the scalp opening for the rotational rod that is situated outside the scalp. Postprocedural infection rates has been reported to vary widely from no infection\textsuperscript{11,12} to 22% in 19 patients.\textsuperscript{6}

Distraction osteogenesis also has some mechanical limitations; there is instability and limitation of the upward distraction from summation of the two major forces that develops during distraction in various angles. The first force is from the distraction rod that usually exerts forward, and the second force develops passively from the scalp that pushes the
distraction rod downward. The summation of these two forces causes the distraction direction to be less forward and downward, resulting in frontal depression and short expansion during frontal skull distraction. Although many operators have employed four distraction rods to increase stability and elevate the frontal flap, the operation time to apply four rods is longer, and the adjustment of each direction of the four rods can be troublesome and less economic.

The authors developed a modified form of rotating distraction osteogenesis by the addition of wirings to the distracted frontal flap and the base of the frontal skull, and using only a two-rod distraction system that resulted in successful rotation of the frontal bone flap. This rotation of the frontal skull flap produced $10^\circ$ rotation and 13 mm upward movement of the frontal skull flap compared with the imaginary conventional distraction osteogenesis method (Fig 3). There were three advantages of the authors' rotating distraction osteogenesis with two rods: 1) the adjustment of the rod direction was simple; and 2) strong stability from fixation of the distal tip of the free skull flap to the base of the frontal skull, third, a larger intracranial volume could be achieved from rotation of the frontal skull flap, fourth, more natural curves were produced without malposition between the frontal skull base and the frontal skull flap after the initial treatment, and last, the procedure was more economic and shorter in terms of operation time. This maneuver could be especially effective for patients with oxycephalic depressed frontal bone conditions.

In conclusion, the authors of this study applied the aforementioned rotating distraction osteogenesis to a four-year-old girl with secondary craniosynostosis and relatively high intracranial pressure. We distracted and rotated the frontal bone flap progressively, resulting in good brain expansion and growing, which successfully filled the whole expanded intracranial space. She showed good cranial contour, to the satisfaction of her parents. Her intellectual quotient also increased, and her character changed from passive to active. These changes facilitated her learning ability and we therefore envisage additional Intellectual development with continued rehabilitation process.

REFERENCES