

# Gradual Orbital Contraction after Facial Bipartition: Correction of Wide No. 0 to 14 Craniofacial Cleft

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**M**edian craniofacial clefts or Tessier no. 0 to 14 clefts are a spectrum of midline anomalies with tissue agenesis and holoprosencephaly at one end (hypoplasia), frontonasal hyperplasia and excessive tissue at the other end, and median anomalies with normal tissue volume (dysrhapsia) in the middle of the spectrum.<sup>1-3</sup> Anterior encephaloceles are congenital cystic malformations of the central nervous system structures herniating through a defect in the cranium in communication with cerebrospinal fluid pathways.<sup>4,5</sup> Embryologically, they occur between normally developed zones, where a weakness permits the brain to escape. A large mass may further push fields apart to create different degrees of hypertelorbitism.<sup>6,7</sup>

Tessier laid the foundation for surgical correction using intracranial-extracranial approaches and facial bipartition.<sup>8,9</sup> Appropriate age for correction of hypertelorbitism remains controversial.<sup>10,11</sup> Corrective surgery for these anomalies at a young age (<8 years), particularly in severe cases (wide intraorbital distances), is thought to result in higher relapse requiring subsequent reoperations. The widest interdacryon distance previously reported in the literature was 59 mm.<sup>10</sup> We describe an anterior encephalocele case with severe hypertelorbitism (interdacryon distance, 81 mm) and report the outcome of a novel technique for gradual medial translocation of facial bipartition segments for operative correction.

## PATIENTS AND METHODS

A 29-year-old G6P4 woman gave birth, by means of cesarean delivery, to a male infant noted to have a large midline anterior encephalocele, associated with a wide Tessier no. 0 to 14 cleft (Figs. 1 and 2). On examination, the patient had separation of the orbits, nasal, and maxillary regions and a large central mass herniating centrally down through a cleft palate. There was no binocular vision; rather, each side functioned independently, including ocular movement. Lack of oral competence created feeding and long-term drooling problems. Computed tomography revealed displacement of frontal lobes, anterior to the orbits, with brain parenchyma herniating down through the palate. Orbits were angled outward at 90 degrees from each other.

Shortly after birth, a ventriculoperitoneal shunt was placed, the dural defect was repaired, and initial frontal bone reconstruction was performed using split bone and recombinant bone morphogenetic protein-2 on collagen sponges fixed to a resorbable plate. At 5 years of age, the patient had developmental delays and no recognizable speech. There were also episodes of meningitis with intermittent cystic swelling and fevers. Because of these problems, we elected to perform corrective surgery at this age. Radiographic studies identified an interdacryon distance of 81 mm and the central canines to be separated by 48 mm (Fig. 3).

Through a zigzag coronal incision, the encephalocele was dissected from the skin flap and regenerated frontal bone from the previous op-

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*Received for publication April 5, 2010; accepted June 17, 2010.*

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DOI: 10.1097/PRS.0b013e3181f44802

**Disclosure:** *The authors have no financial interest to declare in relation to the content of this article.*



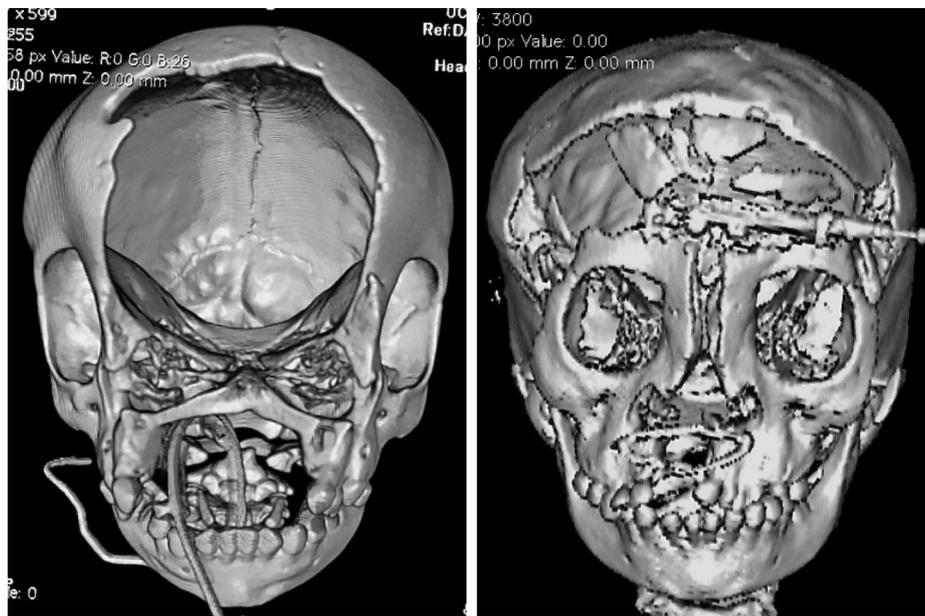
**Fig. 1.** Frontal views of a patient with Tessier no. 0 to 14 craniofacial cleft. (Left) Preoperative image demonstrating large midline frontonasal encephalocele. (Right) Postoperative image after facial bipartition, gradual soft-tissue contraction, and subsequent median cleft lip and nose repair.



**Fig. 2.** Lateral views of a patient with Tessier no. 0 to 14 craniofacial cleft. (Left) Preoperative image demonstrating anterior displacement of the encephalocele with functional problems of independent ocular movement and drooling. (Right) Postoperative image after corrective procedures. Functional improvements in ocular movement, oral competence, and speech were noted.

eration. There was only a small amount of abnormal brain parenchyma present that could be safely resected. The rest of the anteriorly displaced frontal lobe had to be mobilized superiorly. A facial

bipartition was then performed. After downfracture of each segment, accessory central bone and septal cartilage was resected. It was necessary to resect medial orbital bone posteriorly, close to the



**Fig. 3.** Three-dimensional computed tomographic scans. (Left) Preoperative image with a large central osseous defect and an 81-mm interdacyron distance. (Right) Postoperative image after orbital soft-tissue contraction with the midline device in place.

optic foramen, to allow for mobilization of the bipartition segments.

Intraoperative medialization of the facial halves was accomplished to an interdacyron distance of 41 mm. A further attempt at medialization was limited by compression of the brain and soft-tissue traction. We therefore anchored a 30-mm internal KLS Martin (Jacksonville, Fla.) distractor to the supraorbital bone of the bipartition halves. This was performed to facilitate sequential soft-tissue contraction of the bipartition halves. The distractor device was brought through a separate stab incision in the frontal scalp. Maxillomandibular fixation screws were placed on both maxillary segments, and 24-gauge wire was passed between them to provide inferior stabilization and for subsequent tightening during device contraction.

The patient underwent 17 days of sequential contraction, closing the distractor daily to reach an interdacyron distance of 18 mm. Immediately after this, the patient was taken back to the operating room for removal of the distractor along with a small segment of central bone. The medialized bipartition segments were fixed using two 1.0-mm box titanium plates. Soft-tissue transnasal wires with Xeroform (Covidien, Mansfield, Mass.) bolsters were also placed for 7 days. The midline abnormal skin and soft tissues were resected at this time.

Four weeks later, the patient was taken back to the operating room for repair of his true median cleft lip and nose. Midline excess tissue and duplicate structures were marked for excision. After bilateral subperiosteal mobilization, the upper lip was repaired in layers: mucosal, interdigitating orbicularis muscle slips, and skin. For nasal reconstruction, a cantilever cranial bone graft was used. Four weeks later, the wide cleft palate and alveolar cleft were repaired. A superiorly based pharyngeal flap was used for supplemental nasal lining. Iliac crest bone was used for the alveolar defect.

At 1-year follow-up, the patient had developed recognizable speech, had good vision bilaterally with signs of coordinated ocular movement, and showed improved social interaction. Years of follow-up for signs of relapse will be needed.

## DISCUSSION

Because of the variations of median craniofacial dysplasias, a variety of surgical approaches have been described for the correction of Tessier no. 0 to 14 clefts.<sup>12-16</sup> These strategies have been aimed at repositioning the orbits, reconstructing the nose, and repairing the cleft lip and palate. For hypertelorbitism, early reports described resection of the central bony portion of the nose and medial movement of the inner eyebrow for camouflage.<sup>15,16</sup> Contemporary techniques (facial bipartition/orbital box osteotomy) take root in the work per-

formed by Tessier and colleagues.<sup>8,16</sup> Converse demonstrated this strategy to be safe, successfully performing encephalocele reduction, dural repair, orbital osteotomy, and median bone resection.<sup>16</sup>

In this report, we present a novel surgical strategy of “orbital soft-tissue contraction” after facial bipartition mobilization for one of the widest interdacryon distances reported in the literature. After narrowing the orbital distance to 41 mm intraoperatively, a distraction device (used in reverse) was placed for postoperative activation. This particular strategy was undertaken for two reasons: (1) to avoid undue pressure on the brain and (2) to avoid excess stretching/trauma to the optic nerves. This successfully allowed for gradual reduction to an acceptable interdacryon distance. Distractor removal and fixation with titanium plates was performed, as bony transport has been shown to produce a fibrocartilaginous cap which, if not removed, would prevent bone healing.<sup>17–19</sup> We offer this report as a viable approach for treatment of patients with excessively wide hypertelorbitism.

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### PATIENT CONSENT

*Parents or guardians provided written consent for the use of the patient image.*

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