Use of Osteogenesis Distractors in Cloverleaf Skull Reconstruction

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Abstract: Cloverleaf skull deformity (Kleeblattschädel-Syndromen, trilobular skulls) results from synostosis of multiple cranial sutures. The number of sutures involved, the pathogenesis of the synostosis, and the associated anomalies and syndromes are variable. All forms of cloverleaf skull are associated with a high morbidity and mortality. Management of surviving infants requires multiple decompressive and reconstructive operative procedures. Maximal advancement of the forehead/brow at the initial surgery is usually not enough to correct the associated proptosis and a second brow advancement must be done. We present a patient with Apert syndrome and cloverleaf skull deformity that required early (1 month old) cranial vault decompression due to severe proptosis and papilledema. Our management included the placement of osteogenesis distracters on the forehead/brow to gain additional advancement and expand the soft tissue.

Key Words: cloverleaf, Kleeblattschädel, craniosynostosis, osteogenesis distraction, Apert syndrome, fronto-orbital advancement

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Cloverleaf skull deformity results from the synostosis of multiple cranial sutures. It is a rare deformity with several case reports and only a few case series. The bony constriction on the brain can create life-threatening and permanent neurological sequelae if left untreated. Early intervention with ventricular shunting followed by staged craniotomies with forehead/brow reconstruction and advancement is indicated to expand the cranial vault, decrease intracranial pressure, and correct proptosis.[1] We present a case of newborn with cloverleaf skull deformity and severe proptosis.

CASE STUDY

A newborn girl with Apert syndrome and cloverleaf skull deformity was diagnosed by ultrasound during pregnancy. The girl was stabilized in the neonatal intensive care unit and a ventricular shunt was placed when evidence of hydrocephalus was obtained. Because of the severity of the synostosis, persistent intracranial hypertension, and papilledema, a semiurgent craniotomy was planned at 1 month of age (Fig. 1). A zigzag bicoronal incision was made to expose the anterior two thirds of the skull and supraorbital rims. Severe constriction was noted in the area of the coronal sutures along with multiple bony defects with brain herniation through these openings. The entire anterior two thirds of the skull and supraorbital rims were removed. This allowed extensive decompression of the brain and release of superior and lateral orbits down to the inferior orbital floor. Bone grafts were harvested from the parietal area to reconstruct the defects in the supraorbital bone. Resorbable plates were used to secure bone grafts to the supraorbital bone as well as to help maintain the new contour. Two large pieces of bone were recontoured and used to reconstruct the forehead plate (Fig. 2). The supraorbital bone and forehead were then secured together and positioned with 2 cm of advancement. Bilateral distractors were used to secure the lateral supraorbital bone to the temporal area of bone to provide additional advancement of the forehead/brow with distraction osteogenesis. Flexible arms were attached to each distractor and exited in the temporal scalp. On postoperative day 5, distraction was initiated twice a day (1 mm/d). Bilateral distraction was successfully performed from 25 days for an additional 2.5 cm of forehead/brow advancement. The distractors were removed 5 months later (Fig. 3). Postoperative photographs show the forehead and brow still in good position 1 year later (Fig. 4).

DISCUSSION

Cloverleaf skull (also known as Kleeblattschädel, trilobar skull, triphyllocephaly) was first described by Holtermüller and Wiedemann in 1958. It is a rare deformity resulting from synostosis of more than 3 major cranial sutures. The synostosis creates a trilobar skull with varying levels of temporal bulging and bossing of the forehead. Most infants will also have a flattened posterior skull. Although cloverleaf skull deformity encompasses any combination of synostosis producing the trilobar skull, the coronal and lambdoid sutures are almost always involved.[2] The etiology of cloverleaf skull is unknown; however, the deformity is usually associated with syndromes (achondroplasia, Crouzon, Apert, and Pfeiffer syndromes).[3] Although there are reports in nonsyndromic children, a high morbidity and mortality rate accompanies the diagnosis.[4–6] Besides the cosmetic facial deformity, severe

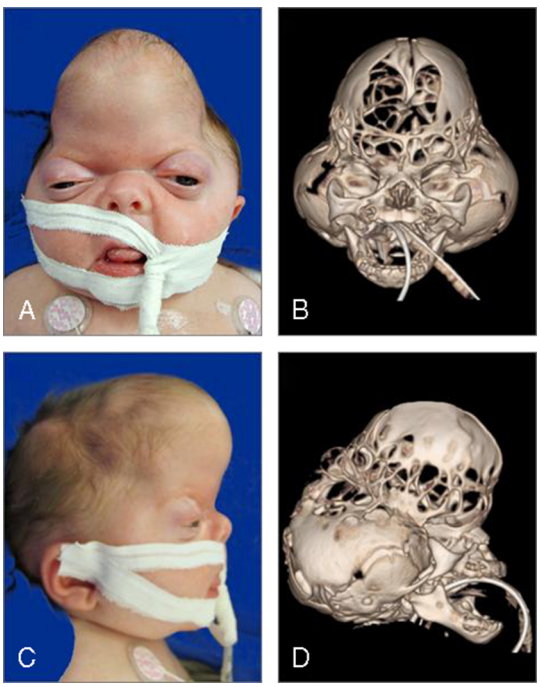

FIGURE 1. Preoperative photographs (A, C) and 3-dimensional computed tomographic scans (B, D) of a 1-month-old girl with cloverleaf skull deformity.

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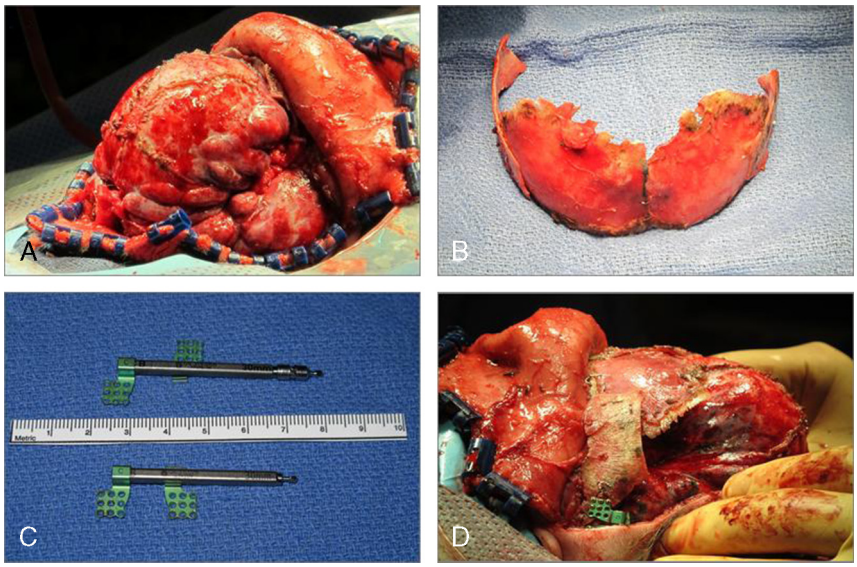

FIGURE 2. Intraoperative photographs showing decompression with removal of anterior two thirds of skull (A), forehead reconstruction with bone grafts (B), and bilateral internal distractors (C) attached to reconstructed forehead and brow for additional advancement (D).

problems can arise with the associated micromyelia, high intracranial pressures, proptosis, papilledema, hydrocephalus, hindbrain herniation, and skull base dysplasias.[7] Those who survive early infancy have a high incidence of neurological impairment that can continue throughout life. The neurological sequelae arise from hydrocephalus, hindbrain herniation, and venous hypertension.[8]

Surgical correction of the cloverleaf skull has evolved since the first reported treatment in 1972 involving craniotomies along fused suture lines.[9] Subtotal craniectomy (the suggested procedure in *Plastic Surgery* by Mathes[10]) has been the treatment of choice even with the expectant poor outcomes. Several authors have described staged release of the involved sutures with far superior statistics in morbidity and mortality.[11–13] Current recommendations from centers who have dealt with higher numbers of these recommend early control of intracranial hypertension with ventricular shunting and a staged cranial decompression/remodeling. Because of the limited amounts of quality calvarial bone and the limitations of the soft tissue, fronto-orbital advancement can sometimes fall short of the distance needed to correct the associated problems of turricephaly and proptosis.[12,14] For these children, this means a second and sometimes third procedure to advance the forehead/brow. Distraction osteogenesis offers the advantage of obtaining additional advancement with the gradual stretching and expansion of the soft tissue that can limit the initial expansion. This technique can potentially limit the number of advancements needed.

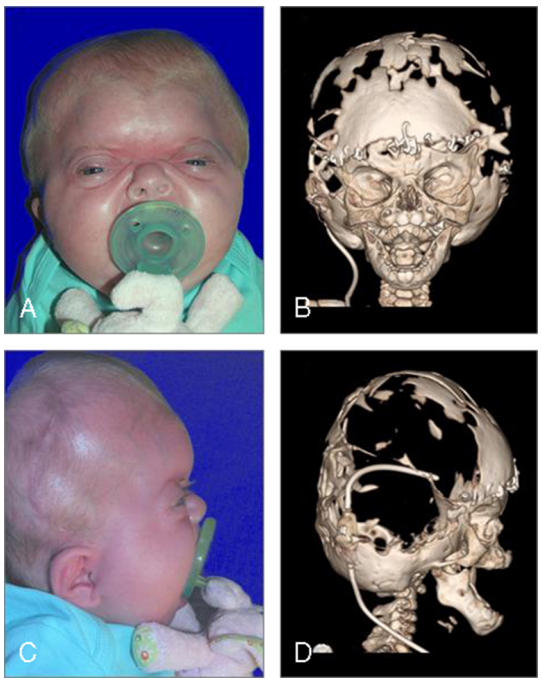

FIGURE 3. Postoperative photographs (A, C) and 3-dimensional computed tomographic scans (B, D) 5 months after decompression, reconstruction, and distraction.

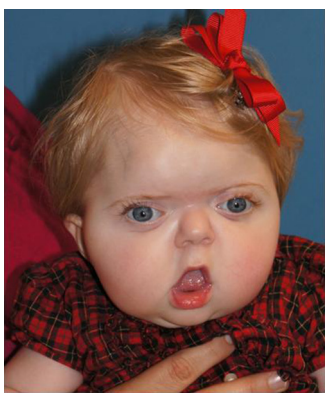

FIGURE 4. Postoperative photograph at 1 year of age shows forehead and brow in good position.

CONCLUSIONS

In infants born with craniosynostosis requiring early decompression, forehead/brow advancement with the addition of distraction osteogenesis may decrease the number of procedures required to correct the deformity.

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