Occipital flattening in the infant skull

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Occipital plagiocephaly caused by lambdoid synostosis is rare. Positional flattening is more common and will most often respond to conservative measures. Surgical correction of a flat occiput is warranted if the deformity is profound. Skull molding devices may be effective for treating mild abnormalities but are ineffective in the more severe cases. An operative procedure is described that uses a microplate-reinforced median bar to provide a rigid scaffold to maintain the occipital correction. Seventy-three consecutive patients were evaluated over a 3-year period for occipital plagiocephaly. Of these individuals, only one had true lambdoid synostosis and six required surgery. There were no operative complications and cranial length was increased from 84 to 94% of age-matched controls after surgery. The need for operative intervention is rare; however, it should be based on the severity of the posterior deformity, especially when accompanied by compensatory frontal bossing, and not on the etiology of the flattening.

Key Words * plagiocephaly * skull deformity * positional flattening * children

Unilateral or bilateral occipital flattening in infants is becoming more commonly recognized. In the majority of cases, flattening is thought to be the result of prone positioning, which has had an important role in decreasing the risk of sudden infant death syndrome. True lambdoid synostosis with bone fusion of the suture remains extremely rare.[1] In our experience skull molding therapy has been only modestly effective. Therefore, we believe that surgical correction of occipital flattening without true synostosis is warranted in a severe and aesthetically unacceptable skull deformity that does not improve over time.

Fig. 1. Left: Photograph of a patient with true lambdoid synostosis. Center: True lambdoid synostosis is demonstrated on a three-dimensional CT scan obtained in a 6-month-old patient. Right: Artist's illustration showing typical changes in true lambdoid synostosis, including downward displacement of the pinna as seen in panels left and right.

CLINICAL MATERIAL AND METHODS

From 1992 to 1995, 73 patients were evaluated for unilateral or bilateral occipital flattening. There was only one child noted to have true lambdoid synostosis. In this case correction was not undertaken because of the mildness of the presenting deformity. Physical examination of this child demonstrated an ipsilateral ear that was displaced downward and posteriorly. In patients with unilateral positional occipital plagiocephaly, the ear is typically displaced forward (Fig. 1). Differences in skull characteristics between positional flattening and true lambdoid synostosis have recently been described by Huang, et al.[2]

Fig. 2. Artist's illustration of the operative technique for repair of posterior skull deformities with a bilateral correction. The patient is place in the modified prone position (A) and bilateral paramedian osteotomies are created (B). Bilateral parietooccipital craniotomies are fashioned (C) and the sagittal sinus is freed and cut posteriorly (D). A piece of bone taken from the parietooccipital craniotomies is used to lengthen the posterior strip (E). The construct is secured with microplates after posterior contouring (F). The craniotomies are reversed and rotated (G) and loosely sutured to the midline strut and the surrounding bone (H). For unilateral deformity, the midline arc is less exaggerated and the flap rotation and reversal creates a symmetrical result. Preoperative three-dimensional CT scanning enables visualization of the transverse sinuses and torcula; the foramen magnum is also revealed, which aides in a safer dural exposure (I).

Of the 73 patients, six underwent surgical correction. The children's ages at the time of correction ranged from 3 to 9 months (mean of 6 months). Two patients had bilateral positional flattening and four showed unilateral asymmetry. No child responded adequately to conservative treatment measures such as positional changes or skull molding therapy. The technique we used to repair the skull deformity is illustrated in Fig. 2. The common feature in the reconstruction is the judicious use of microplates for internal fixation. In the past, we noted a disturbing incidence of breakdown of correction when the child returned to the supine position; the rigidity provided by the

microplates maintains the correction achieved at surgery and allows the child to lay supine as quickly as 48 hours after surgery.

RESULTS

There was no operative morbidity or mortality in our series. Specifically, there was no need for delayed blood transfusion, no cerebrospinal fluid leak, no brain injury, and no transosseous migration of plates or screws at follow-up evaluation 1 year postoperatively. Cranial length was increased from 84% of age-matched controls to 94% of age-matched controls, as determined by comparing the preoperative with the 1-week postoperative computerized tomography scans. Pre- and postoperative lateral radiographs are provided as Fig. 3.

DISCUSSION

Conventional understanding of the nature of posterior skull deformities has recently undergone revision. In two major series of craniofacial operations performed between 1968

and 1984, lambdoid synostosis accounted for 15 to 20% of all surgical procedures for craniosynostosis.[1,5,9] The skull deformity that is described in these reports, however, is now thought to be more consistent with positional deformation rather than true synostosis. In fact, histological evaluation of the sutures showed bone fusion in only 7% of the patients in their series.[5] Because this deformity is now believed to be the result of positional molding, which may be amenable to nonsurgical means of treatment, the decision to not operate is now more often made.

Fig. 3. Preoperative (left) and postoperative (right) lateral radiographs obtained in a 6-month-old patient who underwent correction of a bilateral positional deformity.

Our indications for surgery are based on the severity and course of the deformity rather than its etiology. Surgery is recommended if there is a progressively more severe deformity, characterized by marked uni-

or bilateral occipital skull flattening and significant compensatory frontal bossing with elevation of the vertex that does not improve spontaneously or after use of a skull molding cap over a trial period of no less than 3 months. In our experience, the use of a skull molding cap alone has led to disappointing results either because of patient noncompliance or intractability of the bone deformity.[7,8]

Techniques for operative correction described previously include isolated strip craniectomies, limited cranioplasties with reversal and rotation of bone segments, and extensive total cranial vault cranioplasties with barrel stavelike osteotomies to reshape the effected bones. Additionally, the use of prolonged prone positioning in the postoperative period is required.[1,3,4-6,10] A more extensive cranioplasty is usually needed to resolve all primary and compensatory problems involved with significant positional molding.

Our modification to existing techniques is the creation of a median strut of bone posteriorly to define a new arc for the posterior skull, which is rigidly fixed. Because of the rigid fixation, it also acts as a scaffolding to which the remaining parietooccipital bones can be attached (Fig. 2). Rigid fixation is only used on the median strut to incorporate the interpositional bone graft and to provide enough stability to prevent relapse in the early postoperative period. The remaining bones are only loosely attached with absorbable suture to allow for enough flexibility to accommodate further brain growth and expansion. The procedure is applicable to either unilateral or bilateral positional flattening. For bilateral deformities the arc of the strut is more pronounced. In unilateral flattening the arc of the midline strut is less exaggerated, and rotation of the parietooccipital flaps is used to produce symmetry. In the unilateral cases, the frontal deformity is addressed if needed. The patient is kept prone for the first 48 hours; after that time period, there are no restrictions on the patient's positioning.

CONCLUSIONS

The indications for operative management of both true lambdoid synostosis and positional deformations remain controversial. In this series of 73 patients, six children underwent surgical correction. All children had severe unilateral or bilateral positional deformities and a trial of conservative management consisting of behavior modification and helmet therapy had failed. The procedure described has been safe and has provided excellent cosmetic results; nonetheless, because most infants respond so well to conservative measures, only a small number of severe cases will require surgery. The ultimate discriminator is not the etiology but the severity of the deformity along with any consequential compensatory changes.

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