Treatment of bilateral coronal synostosis in infancy: a holistic approach

JOHN A. PERSING, M.D., JOHN A. JANE, M.D., AND JOHNNY B. DELASHAW, M.D.

Departments of Neurological Surgery and Plastic and Reconstructive Surgery, University of Virginia School of Medicine, Charlottesville, Virginia

Bilateral coronal synostosis often results in a turribrachycephalic skull shape. Reduction of skull height and elongation of the anteroposterior axis of the skull while preserving normal cerebral function are the major therapeutic goals. A surgical technique is described which can successfully accomplish these goals in a single operative procedure.

Key Words • coronal synostosis • oxycephaly • skull reconstruction • cranioplasty

Children born with bilateral coronal synostosis have a distinctly abnormal skull which is difficult to treat. In these infants, the overall skull shape is shortened anteroposteriorly, widened mediolaterally, and elongated vertically compared to normal. The anterior cranial base is foreshortened, the orbital rims are hypoplastic (particularly superiorly and laterally), the superior frontal and squamous temporal bones are protuberant, and the occiput is flattened.

The treatment of these deformities has undergone major advances in the last 90 years. Emphasis on release of the stenotic suture, on early surgical intervention, and more recently on major reshaping of the disfigured bone have all led to significant improvements in skull shape. In spite of this, a truly normal skull shape is relatively infrequently achieved, even following the most extensive cranioplasty procedure. In patients with bilateral coronal synostosis, the problem is often related to an inability to obtain satisfactory reduction of the abnormal height of the skull and to elongate its shortened anteroposterior axis. A technique is presented which can accomplish both goals in one operative procedure.

Operative Procedure

Preoperative Evaluation

Preoperatively, the patient is assessed by clinical and radiographic examination. If the child is less than 1 year of age and has the typical clinical stigmata of bilateral coronal synostosis, as described above, the following approach is undertaken. Computerized tomography (CT) scans of the skull base and plain radiographs of the skull are obtained. In addition, cervical spine radiographs, including lateral flexion and extension films, are taken to exclude craniovertebral abnormalities or instability which might jeopardize safe placement of the patient in the modified prone position. This position is essential to allow for simultaneous access to the anterior and posterior aspects of the skull. Simultaneous anteroposterior access is necessary for full reshaping because the capacitance of the skull must be expanded posteriorly in order to sufficiently reduce the skull height while at the same time avoiding significant compression of the brain.

Operative Technique

The patient is intubated while supine and is then placed prone on a supple beanbag. Foam padding is placed to cushion the child, particularly beneath the chin and adjacent to the cheeks up to the level of the zygoma. The beanbag is brought up around the lower level of the face to the same level, leaving the lateral orbit, postauricular region, and occiput freely visible. The beanbag is vacuum-stiffened to support the skull in this position (Fig. 1).

The operative procedure is begun with a bicoronal skin incision. Supraperiosteal subgaleal dissection is performed in children less than 1 year of age to elevate the scalp flaps so as to lessen the amount of blood loss. The scalp flaps are reflected anteriorly to the supraor-
bital rims, and posteriorly superficial to the semispinalis capitus musculature. At approximately 1 cm above the supraorbital rim, a transversely oriented incision is made in the peristeme and a subperiosteal and subperiobital dissection is performed in continuity. The subperiobital dissection is carried intraoorbitally for a distance of 1 to 2 cm on the superior, lateral, and inferolateral walls of the orbit. Additionally, because the squamous portion of the temporal bone is deformed by a lateral bulge, the temporalis muscle is elevated subperiosteally and laterally out of the infratemporal fossa to the level of the zygomatic arch. Posteriorly, the occipital musculature is elevated subperiosteally to the level of the rim of the foramen magnum medially, and slightly cephalad to that laterally.

The cranial vault shape is again assessed. Note is made of areas of skull irregularity, taking into account the height of the skull and the position of its vertex on the anteroposterior axis. The goals in design of the osteotomy are multiple. One goal is to leave a platform of bone at the vertex of the skull adjacent to the sagittal sinus (to avoid injury to the sinus), with lateral parietal bone struts extending from the vertex bone plate to the skull base, which will later serve to reduce the height of the skull. A second aim is to remove the frontal and parieto-occipital bone plates as bilateral bone grafts. The latter maneuver is attempted so as to reduce the likelihood of asymmetry developing between the right and left hemicrania, which would be more likely to occur with multiple independently mobile bone segments in the frontal or parieto-occipital regions. A third goal is to expand the skull posteriorly in order to allow neurocranial capsule displacement posteriorly as the skull height is reduced.

After the proposed craniotomy lines are drawn, burr holes are placed, avoiding the forehead region. One burr hole is placed posterior to the hairline and the anterior fontanel (if fused), and one lateral burr hole is placed in the region of the pterion bilaterally. A bifrontal craniotomy is performed fracturing the frontal bone plate forward at the glabella (Fig. 2). If the temporal squami are excessively protuberant, a craniotomy is performed to remove the irregularly convex-shaped bone in one piece, if possible.

In the parieto-occipital region, the torcular and transverse sinuses are dissected free from overlying bone and a craniotomy is performed, including parietal and occipital bone, at the level of the lambdoid suture just above the transverse sinus. The sagittal sinus and its overlying parietal bone are left undisturbed. The parieto-occipital craniotomy may still be performed in one piece at this stage, but with a shorter bridge attachment across the midline (that is, only occipital bone is bridging the midline).

After completion of the parieto-occipital craniotomy, epidural dissection is carried out over the cerebellum to just above the level of the foramen magnum. Parallel, vertically oriented "barrel stave"-like osteotomies are performed in the remaining basal, parietal, and occipital bones (Fig. 3). The caudal extent of the osteotomies reaches to a point approximately 2 to 3 cm above the level of the foramen magnum medially and 3 to 5 cm above this level laterally. The osteotomies have weakened the occipital bone, which may then be fractured posteriorly. This green-stick fracture accomplishes two things: it increases the bone capacitance of the skull overall, particularly in the occipital region, and it acts to elongate the basal vault skeleton in the anteroposterior axis.

Attention is redirected to the periorbital region. The burr hole at the pterion level is enlarged to expose a larger portion of the greater wing of the sphenoid bone bilaterally. Much of the superolateral aspect of the greater wing of the sphenoid bone is removed, as it is abnormally displaced cephalad, and may represent the basal extension of the coronal suture synostosis. In addition, an osteotomy of the orbital roof is carried out
Bilateral coronal synostosis in infancy

approximately 5 mm posterior to the superior orbital rim. The sectioning of the orbital roof and walls is carried laterally to encompass the frontozygomatic suture, the frontal process of the zygoma, and the body of the zygoma as it forms the infraorbital rim. Thus, a C-shaped ring of bone is formed which may be advanced forward as a single unit (Fig. 4). The rim is supported forward in this advanced position by stacked segments of bone taken from the parieto-occipital regions and wedged into the oblique osteotomy in the zygoma. The advanced orbital rim is secured inferolaterally to the stacked bone grafts by wire; however, no wire bridges the orbital rim to the zygoma or to the cranial base in the growing child. In essence, the lateral rim is allowed to "float free." The rims are advanced into a slightly overcorrected position. The squamous portions of the temporal bone are cut radially and reshaped, then secured inferolaterally to the remaining temporal bone.

Prior to reduction of skull height, an intracranial pressure (ICP) monitoring device is inserted in the paramedian right parietal bone through to the subarachnoid space (Fig. 5). The parietal bone struts extending caudally from the vertex of the skull are severed low and lateral in the parietal region. The vertex of the skull is slowly shifted caudally and posteriorly, avoiding displacement of the ICP monitor, to reduce the abnormal height of the skull. Simultaneously, the anteriorly displaced bregma is shifted posteriorly by the posterior positioning of the parietal bone struts. The skull height is usually reduced 1 to 1.5 cm but, regardless of the height reduction, it should be done slowly (over the course of 30 to 60 minutes). This is accomplished by slowly cinching down the wire segments placed through both the caudal portion of the parietal bone struts and the remaining cranial base temporoparietal bone. Intracranial pressure monitoring is useful during this maneuver to assess the patient's tolerance for this height-reduction procedure.

Empirically, the ICP has been allowed to reach as high as 40 mm Hg for short periods (< 2 minutes) as height reduction occurs. Within this time frame, the ICP must reduce to less than 20 mm Hg, as venous and cerebrospinal fluid (CSF) compensation occurs. Further incremental height reduction may then be achieved if

FIG. 3. "Barrel stave"-like osteotomies are placed in the basal parieto-occipital skeleton. When the osteotomy segments are fractured outward, they elongate the anteroposterior axis of the skull and increase the capacitance of the posterior cranial vault. Radial osteotomies are placed in the frontal and parieto-occipital bone grafts to weaken and remodel the bone. A vaulted arch is created based on bilateral parietal struts.

FIG. 4. A: A C-shaped orbital osteotomy is made to advance the superior (1), lateral (2), and inferior (3) orbital rim. B and C: The lateral portion of the greater wing of the sphenoid bone (as the basal extension of the coronal suture) is removed. Bone grafts are inserted into the oblique osteotomy in the zygoma, as the orbital rims are green-stick fractured forward.

FIG. 5. A: The vertex of the skull is shifted posteriorly approximately 1 or 2 cm. The desired amount of bone is removed from the basal parietal bone struts. Skull height is reduced by slowly cinching down the wires placed through the parietal bone segments. Intracranial pressure is recorded during this maneuver. Dotted line denotes the preoperative profile. B: The frontal and parieto-occipital bone grafts are reshaped. The frontal bone graft is securely fixed to the superior orbital rim, whereas the parieto-occipital bone graft is attached loosely to the dura.
ICP reduction to 20 mm Hg or less has occurred. During ICP recording, conditions of normocapnia and normotension must be met to simulate the postoperative state. If, after 2 minutes, the ICP is not 20 mm Hg or less, or cerebral perfusion pressure is not 60 mm Hg or greater, the restriction is released or the height reduction is achieved more slowly.

The amount of posterior shift of the skull vertex is usually 0.5 to 1.0 cm. This serves not only to place the vertex in a more normal position, but it also results in a less rounded contour to the superior frontal region. This is further supported by plicating the dura frontally in the regions of increased prominence if necessary. The ICP monitor may be capped at this point to allow easier access to the skull, unhindered by ICP recording tubing.

Following this, the frontal and parieto-occipital bone plates are cut radially and contoured. The frontal bone plate is fixed firmly to surrounding bone anteriorly at the supraorbital rim. Conversely, the parieto-occipital bone plate is secured only loosely to the underlying dura, and a gap of bone of approximately 1 cm is left at the anterior margin of the "barrel staves" in the occipital region to encourage further neurocranial capsule migration and expansion in this region (Fig. 5). The scalp incision is closed (Fig. 6). A negative image mold of Elastomere is made of the skull while the patient is still on the operating table. Subsequently, this is used to make a skull molding cap to be used on the child within 1 week after surgery.

Discussion

The turribrachycephalic skull appearance of children with bilateral coronal synostosis is a significant and progressive deformity that has proved difficult to treat adequately. In this report, a technique is described that addresses treatment of the major skull deformities in one operative procedure. Although the approach described is used for most children with bilateral coronal synostosis deformities, the surgeon must individualize the procedure to each child's deformity and condition. Modifications may be necessary to include the variation in patency of the metopic suture and mobility of the frontal bone plate(s). It may be impossible to remove the frontal bone as a single rigid bone plate; in such cases it is best to preserve the ectocranial peristeum in the frontal midline and accept the midline mobility and therefore the greater potential for right-to-left hemicranial asymmetry. The use of a postoperative skull molding cap enhances the symmetry of the frontal bone region.

In the orbital region, the frontozygomatic suture may be widely patent and not allow anterior migration of the lateral orbital rim in the form of a rigid G. If this is the case, we recommend splitting the frontal bone from the zygoma at the frontozygomatic suture, in order to advance the orbital rim forward on the frontal process of the zygoma. As before, individual segments of parieto-occipital bone are stacked on the zygoma to keep the rim forward in the desired position. We avoid attachment of the rim to the shortened anterior cranial base because of the belief that it may further restrict migration of the rim with advancing age. The validity of this belief, however, requires further documentation.

Additionally, the most significant modifier is the extent to which it is possible to safely reduce the height of the skull. As the height of the skull is reduced, it unavoidably compresses the brain-CSF-dura capsule. If compression is too great or too fast it may induce neurological damage.

Theoretically, one would not want to induce a change in ICP, which would critically reduce the perfusion pressure to the brain or cause downward displacement or sheering of the brain stem. Monitoring of ICP is a practical guide in cranial height reduction. Increments in height reduction should be staged such that the ICP does not exceed 40 mm Hg — a level that has been used as an empirical guide by others (MS Muhlbauer, RA Sanford, WC Clark: unpublished data). Ordinarily, the ICP will then drift down to a value of less than 20 mm Hg in under 2 minutes, with no change in mean arterial blood pressure or CO2 tension. Further height reduction can then be accomplished. If the ICP does not reach a value of less than 20 mm Hg during that time interval, the compression should be released or achieved in smaller increments over a longer period of time. One may have to accept a lesser amount of cranial-height reduction in order to assure safety for the patient. Elevation of ICP at the time of surgery is usually transient because the CSF and vascular spaces compensate initially; in addition, the cranial bone compartment has been released or expanded posteriorly, so that the dura may readily stretch into this space in the young patient. Of 11 patients undergoing cranial-height reduction for varied suture and syndrome pathology, the only patient who manifested signs of increased ICP postoperatively requiring release of the cranial height reduction was a 4 1/2-year-old child with...
Bilateral coronal synostosis in infancy

Crouzon's syndrome. This patient had previously undergone two cranioplasty procedures (therefore, the dura was thickened and scarred), had external hydrocephalus, and had the cranial height reduced by 2.5 cm before we began monitoring ICP intraoperatively. Despite this, to date we have not observed any negative neurological sequelae in any of our patients.

Another precaution with this procedure relates to the use of the modified prone position. Basal CT scans, cervical spine films, or (in selected cases) magnetic resonance imaging should be obtained preoperatively to check for the presence of craniovertebral anomalies or instability. The presence of such abnormalities may preclude positioning the patient with the neck hyperextended. In such cases, the operative procedure may be performed in two stages with the patient alternately positioned supine and prone.

As with any craniotomy procedure in virtually any position, the possibility of developing an air embolus exists. We therefore recommend volume loading prior to making the skin incision so as to lessen the possibility, and appropriate testing such as Doppler ultrasound and end-tidal CO₂ monitoring be used during these operative cases.

As more craniotomies are performed with this technique than in previously described procedures, there is a greater potential for blood loss. A useful aid in minimizing blood loss is related to controlled hypotension during the operative procedure. Further safety is afforded by the avoidance of hypothermia, a confounding feature which may mimic hypovolemia. To avoid this complication and to optimize fluid replacement, the trunk and extremities are wrapped circumferentially with Webril following intravenous and arterial line placement. All irrigation fluid is warmed, and the skull is repeatedly bathed in this fluid during the operative procedure.

With these precautions, this form of cranial remodeling has been performed effectively and safely, with the results demonstrating a degree of improvement significantly greater than we have achieved using previously described treatment regimens. This surgical approach appears to have good results because it addresses both hypoplastic and prominent compensatory bone abnormalities present throughout the skull. Virtually all forms of craniosynostosis result in abnormalities throughout the entire skull and surgical correction should reflect the generalized nature of the bone pathology.

Acknowledgments

The authors thank Craig Luce for his illustrations and Lucille Staiger for editorial assistance.

References

9. Lane LC: Pioneer craniectomy for relief of mental imbecility due to premature suture closure and microcephalus. JAMA 18:49-50, 1892
10. Lannelongue OM: De la craniectomie dans la microcephalie. Compt Rend Acad Sci 110:1382-1385, 1890

Manuscript received January 2, 1989.
Accepted in final form August 1, 1989.

Address reprint requests to: John A. Persing, M.D., Department of Plastic and Reconstructive Surgery, University of Virginia School of Medicine, Box 376, Medical Center, Charlottesville, Virginia 22908.