Alternative techniques in the treatment of unilateral coronal synostosis

JOHN A. JANE, M.D., PH.D., F.R.C.S.(C), T. S. PARK, M.D., BARRY M. ZIDE, D.M.D., M.D., PHILLIP LAMBRUSCHI, M.D., JOHN A. PERSING, M.D., AND MILTON T. EDGERTON, M.D.

Department of Neurological Surgery and Department of Plastic and Reconstructive Surgery, University of Virginia School of Medicine, Charlottesville, Virginia, and Institute of Plastic and Reconstructive Surgery, University of New York Medical Center, New York, New York

Premature closure of one coronal suture results in bilateral abnormalities. There is always ipsilateral flatness of the orbital rim, and contralateral frontal bossing is often found. The authors have employed three operative techniques for correction of unilateral coronal synostosis: frontal bone overlay, lateral canthal advancement, and the tongue-in-groove procedure. The choice of operative technique depends upon the exact deformity to be corrected. The authors believe that altering the relations between the bone and dura by techniques such as radical remodeling and dural plication may improve the results of surgical correction of craniosynostosis.

KEY WORDS
- unilateral coronal synostosis
- bilateral correction
- lateral canthal advancement
- frontal bone overlay
- tongue-in-groove procedure
- craniofacial surgery

Premature closure of the cranial vault sutures, or craniosynostosis, has been a recognized clinical entity for over 100 years. Abnormal cranial vault formation results from this premature growth restriction, and with multiple suture closure there may occur, in addition, mental retardation, cranial nerve palsy, visual disturbances, or associated craniofacial deformity. The relationship between the sutural abnormality and the facial malformations is unclear. Whether one or the other is the leading event, whether both are produced by a common factor, or whether a causal relationship exists at all is unknown.

Patients with unilateral coronal synostosis demonstrate ipsilateral flattening of the frontal bone, including the lateral superior orbital ridge, since growth perpendicular to the suture has been restricted. As the brain attempts to grow to a predetermined volume, other cranial compartments whose growth has not been restricted expand (Fig. 1). Depending upon the severity of the condition, the opposite frontal bone will grow more or less excessively to accommodate the brain. This results in a characteristic, but poorly documented, contralateral frontal bossing (Fig. 1B and C). An associated compensatory deformity commonly occurs laterally at the ipsilateral temporal fossa (Fig. 1B and C). The clinical and experimental effects of single-suture closure are widely recognized and are of practical consequence for the surgeon, since adequate correction often requires a bilateral procedure. Cohen has recently analyzed the various theories concerning the etiology and pathogenesis of this condition. He states that “syndrome delineation and clinical evidence to date strongly suggest craniosynostosis is pathogenically heterogeneous.”

This report presents our understanding of the clinical and pathological entity of unilateral coronal synostosis. Operative correction should be technically simple, safe, and rapid, with prompt reversal of the cosmetic deformity.

Historical Background

Craniosynostosis Surgery

A brief history of craniosynostosis and the surgical attempts at its correction aid in understanding this problem. Although references to small and misshapen skulls date back to the time of Hippocrates, scientific attempts at understanding cranial growth were not doc-
Surgery of unilateral coronal synostosis

FIG. 1. Compensatory bossing in unilateral coronal synostosis. A: A mild case in which there is thickening of the bone in the lateral sphenoid wing and some flattening of the supraorbital ridge. Very little deformity has occurred at a distance. B: A more severe degree of deformity in which contralateral frontal bossing has occurred as well as a minor degree of ipsilateral bulging of the temporal fossa (arrows). The temporal fossa compensatory growth is, however, restricted by the synostosis itself. A surgical procedure to correct this condition probably should include a remodeling of the contralateral frontal bone as well as the ipsilateral orbital rim. C: Most extreme form of coronal synostosis in which the contralateral bulge is severe and the ipsilateral thickening is marked. In these cases the contralateral frontal bone should be corrected as well as the ipsilateral temporal deformity.

Documented until the mid 1800's. At that time, Virchow noted that patients with craniosynostosis demonstrated growth restriction in a direction perpendicular to the plane of the fused suture. He reasoned that the prematurely fused cranial vault suture acted as the primary cause of the observed abnormal vault form.

Nearly 50 years passed before Lane and Lanne-longue attempted surgical correction of abnormalities created by sutural fusion. Owing to the usually advanced age of the patient, the inability to maintain an open suture, and confusion with microcephaly, the results were mixed at best. However, in 1943, Faber and Towne established methods of surgical treatment for relieving various conditions, focusing on resection of the fused suture. Later, because of dissatisfaction with the long-term appearance of the skull and the necessity for repeat surgery due to sutural reclosure, renewed effort was directed at developing new surgical approaches to the problem. In 1947, Simmons and Peyton, using tantalum foil, and Ingraham, et al., using polyethylene film, attempted to prevent sutural re-fusion after linear craniectomy by interposing the material between the cut edges of bone at the suture line. In an effort to halt new bone growth, Anderson and Johnson, in 1956, advocated the application of Zenker's solution directly to the dura as a means of chemical cauterization of the bone-forming element of the meninges. Although the incidence of suture patency increased with the use of this agent, the toxic side effects, including seizures, caused many surgeons to cease or modify its use.

Recently, authors have recommended several new or modified approaches to the treatment of coronal synostosis. A significant conceptual advance was the emphasis on early surgical intervention for the skull deformity. Change in brain mass is positively correlated with changes in cranial form. The most rapid increase in brain mass occurs during the earliest stages of development. One might rationally infer that surgical treatment at an early stage of brain and skull development may likely produce more marked changes in skull form than treatment at a late stage. Experimental studies have confirmed this hypothesis.

Many neurosurgeons now recommend surgical treatment of the deformities of cranial synostosis before the 4th month of age.

Another conceptual advance involves the extension of craniectomies to release fused sutures previously not recognized as part of the disease process; notably, the frontosphenoidal, frontoethmoidal, and frontozygomatic sutures. Release and repositioning of abnormally shaped bone has been shown to result in a pleasing cosmetic result in coronal as well as in sagittal synostosis.
Theoretical Principles

Therapeutic decisions regarding various forms of craniosynostosis derive from the work of Moss and Young and their functional matrix theory. They postulated that the primary abnormality in the development of cranial deformity is malformation of the cranial base. In large part, the base consists of the sphenoid bone, which has extensive attachments to overlying dura. They also suggested that an abnormally developing cranial base transmits tensions through the dura to the overlying sutures, resulting in premature fusion of these sutures.

These observations, however, must be viewed in light of our findings in a rabbit model of isolated premature coronal suture fusion. In normal rabbits, premature coronal suture fixation resulted in morphological changes in the cranial base distant from areas of direct manipulation of the coronal suture. Therefore, manipulation of the cranial suture in a rapidly growing calvaria can result in an alteration of the cranial base.

Removal of a normal cranial suture in a normally developing skull does not affect the overall cranial form. Our interpretation is that whether the precipitant of the cranial deformity arises at the vault, suture, or base, it is the dura with its attachment to bone, particularly in the suture region and base, that may be central to the determination of cranial vault form. Because of our belief in this concept, we directly manipulate the form and presumably the stress of the dura at the time of surgery by extensively freeing the dura from the endocranial surface of the skull, then plicating it and/or inserting grafts of fascia to achieve a form that approximates the normal cranial outline. Besides removing the prematurely fused bone at the suture, we also change the cranial form by molding the bone fragments, previously removed by craniectomy, to achieve a normal appearance at the completion of surgery. We think this surgical intervention (namely, the manipulation of the dura, which alters its previously abnormal form and its attachments to overlying cranial vault bone, coupled with remodeling of the bone removed at craniectomy to correct the cranial deformity and change the relationship between bone and dura at the vault and the base) is responsible for the consistently pleasing appearance of our patients. Moreover, our long-term results have shown that early re-fusion necessitating reoperation does not occur, in spite of replacement of all bone fragments, and that no specific measures are necessary to prevent re-fusion. In order to prove the validity of this theory a controlled trial would be necessary. To date our theory is supported only by our impressions from our clinical material.

Operative Techniques

We present two techniques that we use, and describe in detail a third technique that has been alluded to before. The latter is basically a technique of Tessier for bilateral abnormalities which has been applied to the unilateral situation. In all cases of coronal synostosis, one must correct the position and often the shape of the deficient lateral orbital rim. The three ways of accomplishing this are: 1) by frontal bone overlay; 2) by lateral canthal advancement; and 3) by the “tongue-in-groove” procedure. In addition, in all cases the fused coronal, frontoethmoidal, and frontotemporal sutures are opened as part of an extensive removal of the orbital roof and lateral sphenoid wing.

Depending upon the severity of the condition, the contralateral bossing illustrated in Fig. 1 must be corrected, and we have found dural plication (Fig. 2A) to be an effective way of doing this. Reshaped bone can then be loosely applied to the now normally shaped dura. In very severe cases, plication of the ipsilateral temporal dura is an effective means of reducing the prominence of this area (Fig. 2B).
Surgery of unilateral coronal synostosis

A bicoronal incision is made and separate bilateral frontal craniotomies are performed (see Fig. 5). The bone thus freed includes the fused coronal suture. The most lateral aspect of the coronal suture is radically removed with rongeurs into the greater wing of the sphenoid bone up to the superior orbital fissure. Extensive undermining of the dura is performed from the cranial vault to the lateral aspect of the cranial base. The dura may be plicated in regions of abnormal bone prominence as in the frontal region contralateral to the prematurely fused suture, or augmented by pericranial grafts in regions of vault flattening as in the frontal region ipsilateral to the suture.

Inspection of the region of the orbit always demonstrates a lack of prominence of the superolateral orbital margin. As mentioned above, the options available to the surgeon include either an overlay of the free frontal bone fragment, a modified lateral canthal advancement, or a tongue-in-groove advancement.

Frontal Bone Overlay Technique

For the overlay procedure, extensive resection of the orbital bone walls is carried out first (Fig. 3). In this procedure, as well as the other two, it is extremely important to keep the pericranium in continuity with the periorbita. Disruption of the periorbita allows orbital fat to herniate through, complicating the procedure unnecessarily. The orbital rim is resected from the supraorbital notch laterally to the frontal zygomatic suture. The bone of the orbital roof is then removed. Attention is next directed to the area of fusion, which typically involves the coronal, sphenofrontal, and sphenozygomatic sutures. All of this bone is radically removed, and usually this removal is carried to the superior orbital fissure medially.

Modified Lateral Canthal Advancement

If the modified lateral canthal advancement approach is chosen, the pericranium and orbital fascia are bluntly dissected from the underlying bone to expose the superolateral orbital and temporal fossa. Gentle retraction of the frontal lobe exposes the roof of the orbit, following which an osteotomy to the lateral aspect of the orbital roof, including the suprallateral orbital margin, is carried out. The medial extent of the orbital osteotomy is the medial orbital margin. The lateral orbit is then angled forward with respect to the medial orbit. To increase the superolateral orbital prominence, a "green-stick" fracture is produced at the superomedial orbital rim. The bone position is then maintained in its new position by a buttress of bone obtained from the calvaria (Fig. 4A). This procedure can be performed rapidly in younger patients, but seems to us to be better for older patients in whom the final form can be achieved. The reason for this is that the fixation of the orbital fragment is posterior (Fig. 4C).

Tongue-in-Groove Technique

The tongue-in-groove technique involves orbital advancement, and therefore more extensive surgery than the other two methods. It gives excellent results for older patients in whom final fixation can be achieved. It is an example of LeRoux's posterior fixation (P LeRoux, personal communication, 1983). As seen in Fig. 5A, separate bifrontal craniotomies are performed. The "tongue" is outlined and cut with a sagittal saw (Fig. 5B and C), although the cut in the opposite orbit is best made with a circular saw from the outside in (Fig. 6A). The contralateral craniotomy is performed if the bulging on the opposite side is of sufficient magnitude to warrant correction. The temporalis muscle is...
J. A. Jane, et al.

FIG. 6. Tongue-in-groove technique. A: The bone is thinned out after the cut has been made. The oscillating saw cuts across the frontal bone as it articulates with the nasal bone and extends the cut into the contralateral orbit (arrowhead). B: The tongue of bone can be advanced 1 to 2 cm into the groove, and the proper bend made in the orbital rim with a Tessier rib bender.

FIG. 7. Tongue-in-groove technique. A: The tongue of bone has been wired in place in the groove. Rongeurs are used to thin the bone further. B: Pieces of morcellated bone are placed and wired. The temporalis muscle is then sewn back in place anterior to its previous position to correspond with the anterior movement of the lateral orbital rim.

FIG. 8. Illustrations demonstrating the corrections that must be made in unilateral coronal synostosis. A: The contralateral bossing, the sphenoid wing thickening, and the orbital roof must all be removed. B: Advancement of the orbital rim is carried across the midline to include the opposite side (arrowhead).

Dissected from the temporal bone (Fig. 5A) and retracted. At the end of the procedure, the temporalis muscle is advanced anteriorly (Fig. 7B). A back-cut in the fascia may be required. This is necessary since its orbital rim insertion has been moved anteriorly. If the back-cut is not made, a noticeable temporal "hollow" results behind the zygomatic process of the frontal bone. This is a critical part of all of the three corrections described. Since the lateral rim remains intact, the fused frontosphenoidal suture and lateral sphenoid bone must be removed by working from the inside out. This can be accomplished after the tongue has been cut, as illustrated in Fig. 6A. Here a burr is used to remove the fused suture and thin the bone so it can be bent by the rib bender. Rongeurs can also be used to effect the removal (Fig. 7A). The advancement of the orbital rim is carried across the midline to include the opposite side (Figs. 5A, 6A, and 8). The rim is usually advanced 1 to
Surgery of unilateral coronal synostosis

1.5 cm, more in males. After the bone has been cut and removed, Tessier rib benders can be used to achieve the proper shape (Fig. 6B). The frontal bone can be replaced utilizing a variety of techniques, the most effective being the creation of a mosaic of bone fragments. As has been our experience with the procedures for sagittal synostosis in which the relationships between dura and bone are radically altered, we find it possible to replace all bone fragments, and therefore the use of plastic on the edges is not required.

The only contraindication to the tongue-in-groove technique occurs in the situation in which the temporal fossa has bulged inordinately. In these cases, correction of the bulge takes precedence. Since excision of the bone by its very nature removes the "groove," this technique is precluded. We then use the frontal bone overlay or lateral canthal advancement methods.

We have utilized the same technique of tongue-in-groove advancement for cases of an asymmetrical skull in the absence of synostosis. Quite commonly only one brow is flattened and, in this instance, correction of the contralateral side is not necessary since secondary compensation has not taken place, probably because there are no fused sutures.

Comment

Unilateral coronal synostosis often results in a significant bilateral cranial deformity. In every instance, correction of the lateral orbital rim deficiency must be made. In our experience, this can be best achieved either by frontal bone overlay in young infants, a lateral canthal advancement in older infants, or a more radical tongue-in-groove advancement in children over 1 year of age. Depending on the severity of the case, it may be necessary to correct the contralateral frontal bossing. By a more radical manipulation of the bilateral defects resulting from this unilateral abnormality, it appears that a "cure" of the disease can be achieved.

Although it is now obviously feasible with present techniques to perform radical osteotomies and advance the orbital rim to an adequate position, the question still remains as to the position of the orbital contents. We have noted in some cases that a seemingly posterior orbit (that is, retropositioned relative to the normal side) may nonetheless appear in good position later, although no specific orbital advancement was performed. Our explanation for this is as follows. In addition to the osteotomies advancing the orbit, we also remove the orbital roof. Removal of the orbital roof results in a mild pulsating exophthalmos. It may well be that this exophthalmos is what achieves the ultimately pleasing position of the orbit on the side of the synostosis. The orbital roof eventually re-forms with a globe set forward.

One of us (B.Z.) has also seen a case of corrected unilateral coronal synostosis associated with an inferiorly placed orbit on the contralateral side 3 years following surgery. The orbit required elevation to over-come a severe cosmetic deformity. A patient in whom both contralateral bossing and the ipsilateral temporal fossa were corrected, and in whom the frontal dura was augmented, is seen in Fig. 9. A unilateral operation will not suffice to correct this type of deformity, and a bilateral procedure was indicated. The appearance of this child at 6 months of age (Fig. 9 center) is much

Fig. 9. Upper: Preoperative photographs of a 2-month-old infant with right coronal synostosis, showing left frontal bossing and a deficient right orbital rim. Center: The same patient at 6 months of age. Lower: The patient at 5 years of age. The left orbital rim is still slightly depressed. Hair covers the forehead, making evaluation of the contour difficult.
improved, and this has continued up to the most recent follow-up evaluation at 5 years of age (Fig. 9 lower).

Acknowledgment

We are grateful to Craig Luce of the Department of Ophthalmology for his excellent illustrations.

References

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

Manuscript received February 25, 1983. Accepted in final form March 21, 1984. This work was supported by NIH National Research Service Award 5 T32 DE07037-03-0031.

Address reprint requests to: John A. Jane, M.D., Department of Neurological Surgery, University of Virginia School of Medicine, Box 212, Charlottesville, Virginia 22908.