TRIGONOCEPHALY

IDENTITY AND SURGICAL TREATMENT

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TRIGONOCEPHALY is an uncommon variety of craniosynostosis caused by premature closure of the metopic suture, characterized by triangular appearance of the head when viewed from above, narrow pointed ("keel") forehead with mid-line ridging, and close approximation of the eyes. Roentgenograms of the skull demonstrate shortening of the frontal bones, hypotelorism, inward angulation of orbits, and, in the submentobregmatic projection, sharp anterior angulation of the forehead. On the basis of experiences with 18 patients, we wish in this report to help clarify recognition and surgical therapy of trigonocephaly by pointing out its salient anatomic and roentgenological characteristics, and to offer suggestions regarding operative indications and technique.

ANATOMICAL AND CLINICAL REVIEW

The metopic (Gr. metopon, "forehead") or frontal suture separates the frontal bones, extending from the base upward to the anterior fontanel or coronal suture. It is normally functional until at least 2 years of age, closure commencing late in the first year and proceeding to obliteration by the age of 8 years. Growth of frontal bones takes place by proliferation at right angles to the open coronal and metopic sutural margins, and normal rounding and broadening of the anterior cranium depend upon processes of maturation in and along the metopic suture. If part or all of this joint fuses prematurely frontal growth is stunted and the fetal type of angular forehead may persist. It is clear that the ethmoid bone must be narrow when the orbits are close, but whether this be a cause or result of the hypotelorism remains unknown. As in other types of craniosynostosis, the degree of deformation varies with time of onset and rate of premature fusion of the affected suture; the full-blown syndrome probably appears only if the metopic suture closes before or soon after birth and is rather rare, especially in females. Slight or equivocal narrowing of the forehead, mid-line ridging and reduced interorbital distance ascribable to familial characteristics (or possibly to partial synostosis) are common variants in physiognomy and are not to be confused with bona fide metopic synostosis.

Since 1940 approximately 185 patients with craniosynostosis have been observed at Children's Hospital of which 16 (Table 1) have shown premature metopic synostosis, an apparent incidence of about 9 per cent. (Our other 2 patients were treated elsewhere.) It is noteworthy, however, that no cases of trigonocephaly were identified here before 1954, yet 9 have been encountered in the past 18 months, a variation probably attributable to increased awareness and interest of the resident and visiting staffs. Ingraham and Matson (1954) noted 1 case in a series of 130 patients and Matson later commented on 6; Bertelsen reported 8 in a series of 219 individuals with craniosynostosis. Percentages from these clinics no doubt have been revised upward.

Premature metopic synostosis was first described by Welcker (1862) who observed 7 patients with this malady and coined the term trigonocephaly. He and later writers noted the occurrence of various other anomalies in such patients, including
### TABLE 1
**Summary of 18 cases of metopic synostosis**

<table>
<thead>
<tr>
<th>Age, Sex, Date</th>
<th>Appearance</th>
<th>Other Deformities</th>
<th>Mental Status</th>
<th>Roentgen-Ray Data</th>
<th>Operation</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. J.H. 2 mos. Male Oct. 1954</td>
<td>Classical &quot;keel&quot; forehead, hypertelorism</td>
<td>None</td>
<td>Normal at 9 yrs.</td>
<td>Characteristic. Some improvement 2 yrs. postop.</td>
<td>Frontal flaps with no regrowth-delay technique Oct. 1954 (Fig. 5)</td>
<td>Improved 2 yrs. postop., then lost. Forehead better; eyes same</td>
</tr>
<tr>
<td>3. V.G. 4 yrs. Female Nov. 1957</td>
<td>Typical</td>
<td>None</td>
<td>Prob. slow</td>
<td>Characteristic</td>
<td>None</td>
<td>Failed to return</td>
</tr>
<tr>
<td>5. R.A. 4 mos. Male Sept. 1958</td>
<td>Fair. Characteristic tri- gonocephaly (see 8 mos. postop.)</td>
<td>Cleft palate</td>
<td>Mild retardation by psychometric test</td>
<td>Characteristic</td>
<td>Performed 7/24/58 by Dr. J. Shillito. Frontal flaps</td>
<td>Prob. improved 2 mos. postop. Unable to trace later</td>
</tr>
<tr>
<td>9. J.G. 6 mos. Male May 1960</td>
<td>Moderate deformity. Hypotelorism but frontal eminences fair</td>
<td>None</td>
<td>Normal</td>
<td>Not advised</td>
<td>No real change in appearance 1 yr. later. Mental status good</td>
<td></td>
</tr>
<tr>
<td>10. R.K. 19 mos. Male July 1960</td>
<td>Typical, Doll attitude</td>
<td>Fair to poor</td>
<td>Characteristic</td>
<td>Not advised. Pt. of Dr. Hanson and F. L. Johnson</td>
<td>No change of note 1 yr. later. Recovered from 7 viral encephalitis</td>
<td></td>
</tr>
<tr>
<td>11. F.P. 15 mos. Male Aug. 1960</td>
<td>Typical but not extreme</td>
<td>None</td>
<td>Apparently normal</td>
<td>Characteristic; definite but orbits not angulated though close. Other changes well shown</td>
<td>Advised on cosmetic basis: declined by parents</td>
<td>Questionable improvement 1 yr. later</td>
</tr>
<tr>
<td>12. T.D. 44 mos. Male Oct. 1960</td>
<td>Typical</td>
<td>None</td>
<td>Apparently normal</td>
<td>Characteristic. Postop. changes slight in 5 mos.</td>
<td>Frontal flaps. No growth-retarding technique Oct. 1960</td>
<td>Improved appearance 2 mos. later (Fig. 1)</td>
</tr>
<tr>
<td>13. D.C. 5 mos. Male Nov. 1960</td>
<td>Typical</td>
<td>None</td>
<td>Apparently normal</td>
<td>Characteristic</td>
<td>Frontal flaps. No regrowth-retarding technique Nov. 1960</td>
<td>Much better 1 yr. later (Fig. 1)</td>
</tr>
<tr>
<td>15. T.B. 5 mos. Female June 1961</td>
<td>Well defined; frontal eminences not severely flat</td>
<td>None</td>
<td>Apparently normal</td>
<td>Characteristic, though tricho-suture visible</td>
<td>Frontal flaps. Zenker's solution placed on midline dura mater June 1961</td>
<td>Appearance enhanced 6 mos. later</td>
</tr>
<tr>
<td>18. J.M. 7 mos. Male Jan. 1962</td>
<td>Moderate</td>
<td>Epicantthus</td>
<td>Apparently normal</td>
<td>Characteristic</td>
<td>As in Fig. 5. Zenker's solution on midline dura mater</td>
<td>Short follow-up</td>
</tr>
</tbody>
</table>
cleft palate, coloboma, mental retardation, strabismus, polydactylysm, and arhinencephaly with hypoplasia of the forebrain.

In a comprehensive report, Currarino and Silverman reviewed many of the early writings on this subject, concluding that trigonocephaly occurs in two varieties: (1) simple, with no other defects, and (2) in association with malformation of the rhinencephalon and forebrain (arhinencephaly). We feel as they did that the cranial deformity in most instances is a primary lesion not caused by or specifically associated with cerebral defect, disagreeing with earlier opinions that impaired growth of the frontal lobe in these patients causes the changes in the skull. True, diffuse cerebral deformation may coexist, logically accounting for the mental impairment which is commoner here than in most other types of craniosynostosis; possibly 7 of our patients were retarded. It is well recognized, however, that the basic defect in craniosynostosis is cranial rather than cerebral, as opposed to the situation in microcephaly, wherein passive closure of the sutures follows undergrowth of the brain.

Elevated intracranial pressure does not occur if only the metopic suture fuses early, since other joints of the skull are functional, providing for the attainment of adequate cranial volume. However, restraint of the frontal lobes by the narrow anterior cranium may in some individuals lead to reduced functional capacity with consequent mental defect and, on the basis of this possibility, early diagnosis is desirable.

Diagnosis. (a) Clinical signs and symptoms: As indicated above, premature fusion of the metopic suture results in characteristic facial and cranial deformities (Fig. 1). Underdevelopment of frontal bones causes the forehead to be narrow and short with flat frontal eminences, and angulated rather sharply like a prow; the midline ridge is easily visible and palpable. These changes are clearer if the head is viewed from the top, when biparietal broadening contrasts with the small forehead, and the triangular form is defined strikingly. In classic examples close approximation of the eyes and ridging of forehead are quickly apparent when one sees the patient full face, and usually so marked that identity of the deformity virtually is established. Diagnosis is not easy in borderline cases and the very young; observation, reexamination and expert appraisal of roentgenograms of the skull often are essential to exclude facial hypoplasia and other dissimilar conditions. On general physical examination mental retardation and many other defects may be found, but there is no specific commonly associated anomaly, although 5 of our patients showed rather prominent epicanthic folds apparently caused by undergrowth of the root of the nose. The patient often is completely normal except for his peculiar cranial deformity.

History supplied by parents of these children usually is unremarkable. The condition is not inherited as a dominant, but family history may include a higher proportion of congenital defects than average. In our experience a few parents have noted the
baby's ridged forehead and close eyes at birth, and observed no change in these features with growth; others have considered their obviously deformed child normal. One of our trigonocephalic patients (Case 7) was considered by physicians to have only unusual moulding of the head at 6 months and surgery was discouraged; photographs and roentgenograms during the next 2 years showed no lessening of the characteristic facial and cranial deformities. Generally, the presence of mental defect, cleft palate, polydactyly or other gross variant will stimulate more extensive examination and consultations, increasing the chance of early diagnosis, but the child with no defect other than cranial has the greatest potential for salvage and therefore is most deserving of help.

(b) Roentgenography: The major roentgenographic signs of trigonocephaly mirror the clinical features and are diagnostic; in addition there are other characteristic radiographic changes not apparent clinically. In the mentobregmatic projection, the vertical profile of the forehead shows that the keel-shaped angulation is produced by a metopic ridge with sclerotic adjacent bone and flattened frontal eminences. There is well marked contrast between this and the normal rounded frontal contour, as shown in Fig. 2. It should be pointed out that the synostosed metopic suture may be visible in whole or part; this finding does not mean the joint is functional, nor does it conflict with the diagnosis.

A second principal clinical feature, ocular

![Fig. 2. Submentobregmatic views; (a) normal skull, and (b) one with trigonocephaly. Ridging and sclerotic borders of the metopic suture as well as the flattened frontal eminences are well seen.](image_url)
hypotelorism, is obvious in anteroposterior or postero-anterior view of the skull. In addition, these show differences from the normal round orbital contours, the orbits in trigonocephaly being oval or egg-shaped with the longer axis extending upward and medially from the inferolateral orbital margins. This suggests surprise and has given rise to the colloquial designation, the "surprised coon" sign. Coronal sutures are much more clearly defined than normal in the anteroposterior and postero-anterior projections by virtue of the fact that the frontal bone is short and reduced in breadth; lateral views reveal anterior curving of these sutures, whereas normally they are straight.

It is of interest that certain radiographic signs of trigonocephaly are opposite to those seen in coronal synostosis (brachycephaly): in the latter there is often hypotelorism, and the orbits cant outward producing a Mephistophelean appearance (Fig. 3). The forehead is flat and broad, thickened ridges of the fused coronal sutures are not well seen in anteroposterior views, and these curve posteriorly in the lateral projection (Fig. 4). The anterior fossa is short in both brachycephaly and trigonocephaly.

**Treatment.** It seems reasonable that the same fundamental principle of therapy should apply to patients with metopic synostosis as pertains to those with premature closure of other cranial sutures: namely, early provision of cranietomy channels to permit growth of the inhibited bones. The worth of appropriately chosen, properly executed operations has been demonstrated repeatedly in all types of craniosynostosis since the time of Faber and Towne (1927)\(^6\) and it is clear such surgery provides opportunity for ade-
quate enlargement and acceptable contouring of the head. Benefits from any craniectomy operation depend upon expansion of the growing youthful brain; surgery cannot be expected to help if it is too late, or if cerebral development is stunted, the brain hopelessly small and distorted. But, diagnosis of the cranial deformity must be accurate, and operation must be avoided on those who do not truly have premature fusion of sutures.

It is our opinion metopic synostosis should be treated surgically before patient is 3 months of age if he is not obviously retarded mentally and has no other serious contraindication such as disabling urological deformities, uncorrectable cardiac defects, or the like. We recognize it may well be impossible to assess the patient's over-all status fully at that time, and recommend delaying operation if there be serious question of the patient's salvageability. Pneumoencephalography may have value here, and is advised if deformity of the brain seems likely; admittedly a normal air study does not guarantee good mental growth, but severe defect (e.g., absence of corpus callosum) should convince one to avoid operation.

The technique suggested for surgery is as follows (Fig. 5):

Under general endotracheal anesthesia with patient supine, a coronal incision in the scalp is made from one temporal region to the other, well

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**Fig. 5.** (A) Illustrates some of cosmetic features of patient with metopic synostosis and indicates position of cuts in scalp and bone for surgical treatment. In most such cases the frontal bone is shorter, and incisions are farther forward. (B) and (C) show the exposed cranium and beginning removal of the thick metopic ridge. (D) Frontal bone has been trimmed about 1 cm. on all edges. Small bridge in each paramedian contouring cut helps hold the plates in place after they are bent gently to rounder configuration.
behind the hairline. The scalp is reflected downward to the supraorbital ridges providing full exposure of the frontal region, and the pericranium is left undisturbed. After strategic burr openings are made the frontal bone is carefully cut across low, immediately above the brows with a de Vilbiss rongeur, and a connecting channel is made from side to side across the calvarium parallel and just anterior to the coronal suture, avoiding injury to this. The free bone plate is separated from dura mater and lifted out, the mid-line ridge is resected completely and the margins are trimmed, making each half about 1 cm. smaller on all sides than the opening in the skull. An additional vertical cut through both flaps is useful to permit better contouring. In patients less than 6 months old, Zenker's acetic fixative may be applied for 2 minutes on the narrow strips of dura mater exposed around the repositioned plates, or polyethylene film is secured to the margins of the bone. In older children new-bone retarding techniques are used sparingly or omitted because this may delay solidification of the forehead longer than is desirable or necessary.

Results. The cosmetic effect of operation in our patients appears to have been reasonably good and there was no surgical mortality or morbidity. Removal of the mid-line ridge and provision for outward curving of frontal eminences seemed to provide a subtle, surprisingly effective change in appearance within a few days, and continuing growth of the forehead has in several patients enhanced facial contours considerably (Fig. 1). This perhaps adds to the impression that hypotelorism is gradually improved; eyes become set a little wider, and their closeness appears less disfiguring.

Serial postoperative roentgenograms show change in orbital contours with more nearly normal slanting of medial borders and mild increase in interorbital distance, better configuration of forehead in submentobregmatic views (Fig. 2c), straightening of the abnormally curved coronal sutures, and some increase in volume of the anterior fossa. These objective changes have not always been as pronounced as one would like, and are less marked in patients operated upon after the age of 6 months. However, they substantiate the favorable clinical impressions.

It is difficult to say whether there is benefit in freeing the immature frontal lobes from the variable growth-restraining effect of the fused frontal bones, though in more extreme cases this is a logical possibility. We have not detected specific evidence of increased intelligence postoperatively, but 3 patients reacted more normally and were far less hyperkinetic after surgery, changes which perhaps may be attributed to better mental function.

SUMMARY

A survey is presented of 18 patients with premature closure of the metopic suture (trigonocephaly, a type of craniosynostosis producing angled, keel-like forehead and hypotelorism. Identity, associated anomalies, and therapy are considered; it is noted the incidence of additional congenital defects is higher than in children with other varieties of craniosynostosis.

The well established radiological characteristics of trigonocephaly are discussed, including excessive closeness and inward canting of orbits; thickening along the metopic suture, and narrowing, angulation and foreshortening of frontal bones, which cause the abnormally curved coronal sutures to be unusually clear in anteroposterior and postero-anterior views.

Details of suggested corrective surgical procedure are given. This affords immediate enlargement of the anterior fossa, freeing the frontal poles of any constriction, and makes provision for broadening of the forehead and subsequent anterior cranial expansion. When diagnosis is established properly, it is recommended that operation be done before the age of 3 months, assuming that the patient is considered salvageable and shows no major uncorrectable defects.

General nature, diagnosis, therapy, and results are discussed; it is hoped this rather unusual ailment may be recognized more often.

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