Computerized tomography of cranial sutures

Part 2: Abnormalities of sutures and skull deformity in craniosynostosis


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Preoperative computerized tomographic (CT) scans of 24 children who had surgery for either single or multiple craniosynostoses were compared with skull radiographs and operative and pathological findings. In addition to providing accurate imaging of calvarial and skull base deformities secondary to premature suture closure, high-resolution CT with bone definition algorithms supplied valuable detail of anatomical changes at the abnormally developed suture. The CT findings varied with the location of the suture. Thickened bony ridges predominated at the sagittal suture, focal bone thickening and erosions were more likely to be found at the metopic suture, and parasutural sclerosis was the prevalent finding on one side of the lambdoid suture. No evidence of the suture could be detected in the majority of patients with complete coronal craniosynostosis. Radiographs of the skull were shown to be a relatively insensitive means of imaging the zone of limited fusion, especially the lambdoid suture. An excellent correlation was found between the CT scan and the operative and pathological findings. There was histological evidence of progressive suture fusion in virtually all patients. An asymmetrically narrowed lucent zone with parasutural sclerosis or bony ridges seen on CT scans correlated well with fibrous union of the suture found on histological examination. The authors conclude that high-definition CT used in conjunction with bone windows and thin and coronal slices for the evaluation of sagittal sutures is a useful imaging method for the evaluation of craniosynostosis.

KEY WORDS • craniosynostosis • craniostenosis • computerized tomography • cranial suture

The use of computerized tomography (CT) for the evaluation of craniosynostosis and craniofacial dysostosis has been discussed.\textsuperscript{1,3,10,13} Even though CT is an excellent method for achieving detailed evaluation of deformities of the skull base, calvaria, and parenchymal brain structure,\textsuperscript{1,3,13} compared with the efficacy of skull radiographs and radionuclide bone scans, the usefulness of CT in the care and management of patients with craniosynostosis is still controversial.\textsuperscript{10} In a companion paper,\textsuperscript{9} we reported evidence that CT could accurately define normal sutural anatomy, including the presence or absence and the extent of sutural synostosis. On the basis of this evidence, we investigated the ability of CT to differentiate fibrous from bony fusion, to identify the sutures involved, to define the extent of sutural synostosis, and to demonstrate the deformity of the shape of the skull secondary to craniosynostosis. Our findings are presented below.

Clinical Material and Methods

Computerized Tomography Scans

The preoperative CT scans of 24 children, aged 7 days to 19 months, who had surgery for either single or multiple craniosynostoses were studied. Scans were obtained with a GE 8800 scanner and processed with both soft-tissue and bone algorithms. Axial slice thickness was either 10 mm (12 patients), 5 mm (10 patients), or 1.5 mm (2 patients); 5-mm coronal sections were obtained in one child. Skull radiographs were compared with CT scans for all patients.
Pathology

Generally, craniectomy and synostectomy specimens were removed intact at surgery, and often the resected bone included the area of synostosis and adjacent patent suture. Gross specimens were photographed, and areas of synostosis were identified and marked with ink. After decalcification with ethylenediaminetetraacetic acid (EDTA), multiple histological sections, taken at full thickness through the bone plate perpendicular to the axis of the suture, were obtained from the areas of identified synostosis, from suture regions immediately adjacent to the synostosed suture, and from a distant, apparently patent, suture if present. In some patients, abnormal sutures were not removed intact, and histological evaluation was limited to representative sections taken through the bone fragments. Specimens were assessed for evidence of bony or fibrous union, bony sclerosis, vascularity, and other pathological abnormalities without knowledge of the radiographic appearance; only then were the histological and radiological findings compared.

Results

Sutures affected by craniosynostosis are listed in Table 1. A single suture was involved in 11 patients and multiple sutures in 13 patients. Changes in the parenchyma and the incidence of combined syndromes are listed in Table 2. Of the patients with combined syndromes, five had Pfeiffer's syndrome, two had Crouzon's disease, and there was one case each of chromosome 7-P syndrome, Saethre-Chotzen syndrome, and Goldenhar's syndrome. The skull radiographs and detailed CT findings of the calvaria and sutures in patients with isolated and multiple suture synostosis are described in Tables 3 and 4.

Sagittal Synostosis

In the six children with sagittal synostosis, one or more of the following signs were identified on axial CT scans: parasutural sclerosis on both sides of the non-

TABLE 1

Involved sutures in 24 children with craniosynostosis

<table>
<thead>
<tr>
<th>Affected Sutures*</th>
<th>No. of Sutures Involved</th>
<th>Cases of Isolated Synostosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>sagittal</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>metopic</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>lambdoid unilateral</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>lambdoid bilateral</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>coronal unilateral</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>coronal bilateral</td>
<td>9</td>
<td>5</td>
</tr>
</tbody>
</table>

* One patient had a cloverleaf skull, with complete fusion of several sutures; see text.

TABLE 2

Clinical and computerized tomography findings in 24 cases of craniosynostosis

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Surgery (mos), Sex</th>
<th>Involved Suture(s)</th>
<th>Parenchymal Changes*</th>
<th>Associated Syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3, M</td>
<td>sagittal</td>
<td>mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>2, M</td>
<td>sagittal</td>
<td>midline posterior CSF collection</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>3, M</td>
<td>sagittal, bilateral coronal</td>
<td>midline posterior CSF collection</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>8, M</td>
<td>metopic</td>
<td>mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>6, M</td>
<td>metopic</td>
<td>normal</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>6, F</td>
<td>right lambdoid</td>
<td>ipsilateral ambient cistern effacement</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>5, M</td>
<td>right lambdoid</td>
<td>prominent frontal &amp; temporal subarachnoid space</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>4, F</td>
<td>right lambdoid</td>
<td>occipital &amp; parietal sulcal compression</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>9, M</td>
<td>right lambdoid</td>
<td>enlarged subarachnoid space, mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>5, F</td>
<td>right lambdoid</td>
<td>prominent frontal &amp; temporal subarachnoid space</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>7, F</td>
<td>bilateral lambdoid, sagittal</td>
<td>effacement of occipital horns</td>
<td>Goldenhar</td>
</tr>
<tr>
<td>12</td>
<td>2, M</td>
<td>bilateral lambdoid</td>
<td>lipoma, partial agenesis of corpus callosum</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>1, M</td>
<td>bilateral lambdoid, sagittal</td>
<td>enlarged subarachnoid space, occipital horn &amp; sulcal effacement, mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>1, M</td>
<td>left coronal</td>
<td>ipsilateral frontal horn and sulcal compression</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>2, F</td>
<td>right coronal</td>
<td>frontal horn compression</td>
<td>Pfeiffer</td>
</tr>
<tr>
<td>16</td>
<td>4, F</td>
<td>bilateral coronal</td>
<td>minimally compressed frontal horns</td>
<td>chromosome 7-P</td>
</tr>
<tr>
<td>17</td>
<td>2, F</td>
<td>bilateral coronal</td>
<td>mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>18, M</td>
<td>bilateral coronal</td>
<td>moderate hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>1, F</td>
<td>bilateral coronal</td>
<td>partial agenesis of corpus callosum</td>
<td>Pfeiffer</td>
</tr>
<tr>
<td>20</td>
<td>8, F</td>
<td>bilateral coronal</td>
<td>cavum velum interpostum</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>3, F</td>
<td>bilateral coronal, metopic, sagittal</td>
<td>frontal horn compression, complete absence of sulci, mild hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>8, F</td>
<td>bilateral coronal, bilateral lambdoid</td>
<td>moderate to marked hydrocephalus</td>
<td>Pfeiffer</td>
</tr>
<tr>
<td>23</td>
<td>19, M</td>
<td>bilateral coronal, metopic</td>
<td>frontal horn compression</td>
<td>Crouzon</td>
</tr>
<tr>
<td>24</td>
<td>4, M</td>
<td>cloverleaf</td>
<td>marked hydrocephalus</td>
<td>Crouzon</td>
</tr>
</tbody>
</table>

* CSF = cerebrospinal fluid.
ossified suture (three cases), and an external or internal bony ridge along the suture with a prominent deep groove or bony ridge immediately over the sagittal sinus (six cases) (Tables 2 and 3). A completely fused portion of a suture was identified as a bony ridge without parasutural sclerosis in five patients. The bony ridge of the anterior portion of the affected sagittal suture could not always be detected on axial CT scans. The two children with isolated sagittal synostosis and the one child who had sagittal and bilateral coronal synostoses had a characteristic dolichocephalic head shape (Case 2, Fig. 1), a flattened frontal area, and a bicornal diameter that was greater than the biparietal diameter. This characteristic head shape was not present in three of four children with combined synostoses (two had bilateral lambdoid and one a metopic and bilateral coronal synostoses in addition to sagittal synostosis). These patients are described below in the sections on

![Fig. 1. Case 2. Computerized tomography scan showing sagittal synostosis. Note the bony ridge (arrow) without any evidence of the sagittal suture.](image)

**TABLE 3**

*Computerized tomography (CT) findings in isolated suture synostosis*

<table>
<thead>
<tr>
<th>Suture</th>
<th>Drawing</th>
<th>CT Scan</th>
<th>Focal Changes</th>
<th>Skull Shape Deformity</th>
<th>Skull Radiographic Picture</th>
</tr>
</thead>
<tbody>
<tr>
<td>sagittal</td>
<td><img src="image" alt="Sagittal Drawing" /></td>
<td><img src="image" alt="Sagittal CT Scan" /></td>
<td>parasutural sclerosis, external or internal bony ridge along suture, deep groove over sagittal sinus, bony ridge without sclerosis when completely fused</td>
<td>dolichocephaly, flattened frontal area, bicornal diameter greater than biparietal diameter</td>
<td>dolichocephaly, bone fusion sometimes seen on angled anteroposterior or lateral view</td>
</tr>
<tr>
<td>metopic</td>
<td><img src="image" alt="Metopic Drawing" /></td>
<td><img src="image" alt="Metopic CT Scan" /></td>
<td>thickening &amp; sclerosis, enostosis</td>
<td>trigonocephaly, hypotelorism, small anterior cranial fossa</td>
<td>trigonocephaly on basal view, hypotelorism</td>
</tr>
<tr>
<td>lambdoid</td>
<td><img src="image" alt="Lambdoid Drawing" /></td>
<td><img src="image" alt="Lambdoid CT Scan" /></td>
<td>increased bone deposition on inner table adjacent to suture, suture narrower on affected side than on patent side</td>
<td>plagiocephaly (flattened unilateral occipital area), bulging of anterior ipsilateral portion of calvaria, a relatively anterior petrous bone</td>
<td>sticky suture often missing, bulging of anterior ipsilateral portion of calvaria, a relatively anterior position of ipsilateral petrous bone</td>
</tr>
<tr>
<td>coronal</td>
<td><img src="image" alt="Coronal Drawing" /></td>
<td><img src="image" alt="Coronal CT Scan" /></td>
<td>parasutural sclerosis, no evidence of suture, no thickening when completely ossified</td>
<td>plagiocephaly (flattened unilateral frontal area), foreshortened anterior cranial fossa, sphenoid wing displacement, thickening of lowest portion of coronal suture, double sphenoid ridges, shallow orbit</td>
<td>parasutural sclerosis, plagiocephaly on basal view, sphenoid wing displacement</td>
</tr>
</tbody>
</table>
### TABLE 4

*Computerized tomography (CT) findings in multiple suture synostosis*

<table>
<thead>
<tr>
<th>Sutures</th>
<th>Drawing</th>
<th>CT Scan</th>
<th>Changes in Skull</th>
</tr>
</thead>
<tbody>
<tr>
<td>bilateral coronal</td>
<td><img src="image1" alt="Drawing" /></td>
<td><img src="image2" alt="CT Scan" /></td>
<td>foreshortened anteroposterior distance of cranial fossa, shallow orbit (narrow angle between lateral orbital wall &amp; lateral side of middle cranial fossa), thickening of bone around high-positioned frontozygomatic future, double sphenoid ridges, anterior &amp; superior displacement of petrous bones</td>
</tr>
<tr>
<td>bilateral lambdoid</td>
<td><img src="image3" alt="Drawing" /></td>
<td><img src="image4" alt="CT Scan" /></td>
<td>symmetrical occipital flattening, anterior displacement of petrous bones bilaterally</td>
</tr>
<tr>
<td>bilateral coronal and metopic</td>
<td><img src="image5" alt="Drawing" /></td>
<td><img src="image6" alt="CT Scan" /></td>
<td>very small anterior cranial fossa (foreshortened both in anteroposterior &amp; transverse distance), rounded shape of frontal bone</td>
</tr>
<tr>
<td>bilateral lambdoid and sagittal</td>
<td><img src="image7" alt="Drawing" /></td>
<td><img src="image8" alt="CT Scan" /></td>
<td>occipital flattening, marked concavity on midline below sagittal suture, relatively anterior placement of petrous bones bilaterally</td>
</tr>
<tr>
<td>cloverleaf</td>
<td><img src="image9" alt="Drawing" /></td>
<td><img src="image10" alt="CT Scan" /></td>
<td>brain bulging through anterolateral fontanels bilaterally &amp; through anterior fontanel (producing the bizarre appearance of a cloverleaf!), marked digital markings over inner table, shallow orbit &amp; exophthalmos, foreshortened anterior cranial fossa, double sphenoid ridges</td>
</tr>
</tbody>
</table>

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**Fig. 2.** Case 5. Computerized tomography scan showing metopic synostosis. Note the anteriorly pointed cranial vault and bone thickening at the site of the metopic suture (arrow).

**Fig. 3.** Case 21. Computerized tomography scan showing bilateral coronal, metopic, and sagittal synostosis. The site of the metopic suture (short arrow) and the bilateral coronal sutures (long arrows) are both closed. Note the prominent enostosis at the site of the metopic suture.
bilateral lambdoid and bilateral coronal synostoses, respectively.

**Metopic Synostosis**

Two children had an isolated metopic synostosis, and two had the combined type, with bilateral coronal synostosis in both and anterior sagittal synostosis in one (Tables 2 and 3). Both patients with isolated metopic synostosis had an anteriorly pointed cranial vault typical of trigonocephaly. Metopic sutures were not visible on axial CT slices in any patient. One of the children with isolated metopic synostosis had marked sclerosis and enostosis at the metopic suture site. The other had marked localized bone thickening at the suture site within which was a low-density area that at surgery was found to be highly vascular fibrous tissue (Case 5, Fig. 2). This patient had an abnormally foreshortened anterior cranial fossa, despite the lack of involvement of anterior fontanel, sagittal, and coronal sutures. In both patients with the combined type, a large enostosis extended as a cleft between the two frontal lobes. The anterior cranial fossa was foreshortened and small, as expected with bilateral coronal sutural involvement, and the frontal bone was rounded rather than pointed (Case 21, Fig. 3 and Table 4).

**Unilateral Lambdoid Synostosis**

The CT scans of all five children with unilateral lambdoid synostosis showed increased deposits of bone on the inner table of the vault that were limited to one side of the suture (Tables 2 and 3). In two patients the parietal side was involved (see, for example, Case 6, Fig. 4 left), and in another the occipital side. In the remaining two patients, abnormal deposits of bone were limited to a portion of the suture (Case 8, Fig. 4 right). The suture on the affected side was narrower than on the normal side, but in no patient had bone fusion occurred on the normal side. At surgery the affected sutures were immobile with bony ridges, but did not appear to be completely mineralized.

In one patient the junctional region between the lambdoid and occipitomastoid sutures was sclerotic, but bone fusion did not appear to have occurred. At surgery, the most inferior aspect of the lambdoid suture appeared to be closed by a fibrous union, but there was no evidence of bone fusion. In general, the skull vault was characterized by local thinning and flattening of the occipital bone on the affected side, compensatory bulging of the anterior ipsilateral portion of the calvaria, and a relatively anterior position of the ipsilateral petrous bone (Case 8, Fig. 5).

In four of five patients, skull radiographs did not show well the increased bone deposits along the suture or local vault thinning. In only one of the five patients did the skull radiograph show abnormal sutural sclerosis (Cases 6 and 8, Fig. 6).

**Bilateral Lambdoid Synostosis**

Of the four patients with bilateral lambdoid synostosis, only one had no other sutures involved. On CT scans, only occipital flattening and a poorly defined suture line with some evidence of adjacent sclerosis...
could be seen (Case 12, Fig. 7 right and Table 4). At surgery, a number of focal zones of hyperostosis with no sutural movement were found, between which sutures appeared to be patent. This was considered to represent bilateral segmental closure of the lower portion of the lambdoid sutures (Case 12, Fig. 7 left).

In the two patients in whom sagittal synostosis occurred with bilateral lambdoid involvement, the posterior portion of the sagittal suture was marked by internal and external ridging, with bilateral ridging of the inner table at the lambdoid sutures. Posterior aspects of the calvaria of both patients were flattened and had a midline concavity in the occipital bone between the lambdoid sutures (Table 4). The occipitomastoid sutures were open. At surgery, there was marked ridging of the inner table at the lambdoid sutures and a deep groove and significant bony ridge overlying the sagittal sinus that indented the underlying dura (Case 13, Fig. 8 left).

One patient with bilateral lambdoid and bilateral coronal synostosis was diagnosed as having Pfeiffer's syndrome. The lambdoid sutures were completely obliterated on both sides, resulting in a smaller posterior half of the cranium with distinct convolutional markings upon the inner table. The anterior half of the cranium was brachycephalic (Case 22, Fig. 8 right).

Unilateral Coronal Synostosis

The skull radiograph of one of the children had the classic harlequin appearance, in which the lesser wing of the sphenoid was elevated (Case 15, Fig. 9 upper left and Table 3). However, the sphenoid ridge was lower on the affected side in the other patient, which is the opposite of the more commonly described harlequin appearance (Case 14, Fig. 9 upper right). A CT scan showed that the anterior aspect of the calvaria on the affected side was flattened and the ipsilateral portion of the anterior cranial fossa was foreshortened (Fig. 9 lower left). The most inferior segment of the coronal suture was clearly obliterated and thickened, while its more superior segment appeared to be open. At surgery, it was found that the dura adhered to, and was intimately involved with, the most inferior portion of the coronal suture, where abnormal bone production had taken place (Fig. 9 lower right).

Bilateral Coronal Synostosis

Combined syndromes were seen in seven of nine children with bilateral coronal synostosis: four children had Pfeiffer's syndrome and there was one case each of chromosome 7-P syndrome, Saethre-Chotzen syndrome, and Crouzon's disease (Table 2). Closure of portions of coronal sutures that were evident on CT scans was consistent with the operative findings. Other major CT findings included: duplication of the sphenoid ridge, thickening of the bone around the frontozygomatic suture, foreshortened anteroposterior length of the anterior cranial fossa, shallow orbits, apparent anterior and superior displacement of the sphenoid bone and anterior displacement of the petrous bone, abnormally acute angle between the lateral orbital wall and the lateral wall of the middle cranial fossa,10 and superiorly situated frontozygomatic sutures, all of which are considered to be the result of early closure of the coronal sutures (Case 17, Fig. 10 and Table 4). In a patient in whom the coronal suture had definite bone fusion bilaterally, a non-ossified frontosphenoidal suture was clearly shown in a CT scan (Case 19, Fig. 11).

Cloverleaf Skull

In the one case of cloverleaf skull (Case 24), the patient was classified as having Crouzon's disease. The
FIG. 9. Upper: Plain radiographs in Case 15 (left) and Case 14 (right). In Case 15, the classic harlequin appearance is shown. Also, the sphenoid ridge is elevated (arrow). In Case 14, the reverse harlequin appearance can be seen. Note the depressed sphenoid wing (arrow). Lower: Corresponding computerized tomography scans of both patients clearly show the suture abnormalities and skull deformities. The anterior cranial portion is flattened and the anterior cranial fossa is foreshortened in both patients. In Case 15 (left), there was an undefined to absent coronal suture zone on the affected side (long arrow) and a patent suture (short arrow), and in Case 14 (right) the lowest portion of the coronal suture was thickened (arrow).

CT scan showed complete fusion of the metopic, coronal, lambdoid, and squamosal sutures that caused the bizarre cloverleaf skull deformity (Fig. 12). There were three leaflets: the upper leaflet was the brain bulging through the widely opened anterior aspect of the sagittal suture and anterior fontanel, and the lateral leaflets were the brain bulging below the squamosal sutures. The bilateral sphenosquamosal sutures were widely open. The CT scan also showed that the calvarial bone was markedly attenuated over the upwardly, bilaterally protruding brain. There were marked convolutional impressions thinning the inner table of the calvaria and

FIG. 10. Case 17. Computerized tomography scans showing bilateral coronal synostosis. Left: The arrows indicate the double sphenoid ridges. Right: Parasutural sclerosis can be seen along the non-ossified coronal sutures on both sides (arrow).

FIG. 11. Case 19. Computerized tomography scan showing bilateral coronal synostosis. The frontosphenoidal suture is apparent (arrows).
FIG. 12. Case 24. This patient had a cloverleaf skull. Left: Computerized tomography scan demonstrating lateral bulging of the brain (small arrows), the double sphenoid ridges (double arrows), and the completely fused metopic suture (large arrow). Right: Plain radiograph, anteroposterior view, showing the cloverleaf shape of the skull.

FIG. 13. Case 10. Computerized tomography scan showing microcephaly. The bilateral lambdoid sutures are the same width (arrows), and there is no evidence of a bony ridge or sclerosis even though the right occipital area is flattened.

Microcephaly with Unilateral Occipital Flattening

A 5-month-old girl (Case 10) who presented with developmental delay, hypotelorism, a fore-shortened anterior cranial fossa, and double sphenoid ridges were present in the frontal half of the skull (Table 4). The moderate hydrocephalus present preoperatively became severe enough after surgical correction that placement of a ventriculoperitoneal shunt was required.

growth and retarded development and/or deafness). A CT scan showed that the metopic suture was closed (a finding consistent with hypotelorism), and the anterior cranial fossa was slightly foreshortened. However, the other sutures were patent and there was no evidence of parasutural sclerosis or thickening. In fact, the parasutural bone was thinned on the side of the occipital flattening (Fig. 13).

Pathology

Sagittal Synostosis. Microscopic examination of a specimen from a typical patient with sagittal synostosis (Case 3) showed a broad ridge of thickened lamellar bone extending down into the cranial vault at the site of still identifiable suture (Fig. 14 lower). Histologically, a portion of the suture showed complete bone fusion characterized by a broad band of thick lamellar bone that obliterated the suture site. Immediately adjacent to this central area of fused bone was a focus of incomplete fusion, characterized by calvaria in close apposition with a single focus of fused bone (Fig. 14 lower right). Histological sections from areas more distant from the fused focus showed open sutures, but with prominent osteoblast activity and foci of horizontal spicules of new bone formation extending into the suture space from the calvarial bones. Every specimen of sagittal synostosis available for histological examination showed at least focal and usually complete bone fusion of the suture.

Lambdoid Synostosis. Pathological examination of a typical case of lambdoid synostosis (Case 8) failed to identify overt bone fusion despite multiple sections through the synostosis. There was abundant evidence of active bone growth that led to close apposition of the calvarial bones. This was characterized by individual spicules of horizontal growth of new bone extending out from each of the apposed calvarial bones; each
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FIG. 14. Upper: Gross specimen of the inner table of a fused sagittal suture with a prominent bony ridge. Portions of the suture are almost completely obliterated. Lower Left: Histological section of the sagittal synostosis pictured above showing homogeneous dense lamellar bone at the superior surface (top). The inferior portion corresponds to the bony ridge entering from the inner table, and consists of woven and lamellar bone with prominent osteoblastic activity, vascular-ity, and admixed normal marrow elements. This section represents complete bone fusion; no residual suture is evident. H & E, × 18. Lower Right: Adjacent section from suture shown at left. Only a single focus of bone fusion is present; the calvariae are closely apposed, but the preexisting suture site is apparent. Spicules of new bone growth with numerous osteoblasts lining their surfaces are present. This may be an earlier step leading to the process of complete bone fusion seen at left. H & E, × 15.

Spicule was capped by prominent layers of plump osteoblasts (Fig. 15). Spicules showed variable degrees of ossification that ranged from none at the tips to more extensive ossification where they joined the remaining calvarial surface. The fibrous region that comprises the suture space was focally and markedly narrowed at the site where spicules of new bone approached each other; the suture in this area was narrowed to less than one-fifth the width of the remaining suture and was less than 1 mm across. Actual fusion of the bone spicules could not be found on multiple histological sections, which supports the radiographic evidence of fibrous union in this instance.

This pattern of lambdoid synostosis was seen in four of five specimens examined. However, one specimen that showed a similar architectural pattern had a focal fusion between apposed spicules of bone outgrowth that was identified on a single section. The area of fusion consisted of unmineralized cartilage that measured no more than 1 to 2 mm in the greatest dimension. The lack of mineralization suggests that this is an early event leading to bone fusion. Because we cannot exclude the possibility that similar fusion existed focally but was not identified in the sections obtained from other patients, we believe that all pathological findings should be interpreted conservatively.

Metopic Synostosis. Only one example of metopic synostosis was available for pathological examination. This specimen showed diffusely thickened dense lamellar bone at the site where suture normally would be found. Complete bone fusion had occurred, and no residual suture could be identified.

Coronal Synostosis. Specimens of coronal synostosis had a variable appearance. Most specimens had focal bone fusion that typically involved only a portion of the suture thickness; residual suture could be readily identified, and the calvarial bone showed close apposition in the regions adjoining the suture. The adjacent bone showed either no or slight thickening; the prominent spicules of new bone formation found in the lambdoid synostosis were not present. One specimen had complete bone fusion of the lowest portion of the suture with marked thickening of surrounding bone (Case 14, with a reverse harlequin eye sign on plain radiograph, Fig. 9 upper right).

Discussion

Computerized tomography scanning has been shown to be a good diagnostic method for the detailed evaluation of deformities of the calvaria and skull base that are produced by craniosynostosis, and for the diagnosis of abnormalities of the brain parenchyma that accompany craniosynostosis.1,3 Because the scanners used in these earlier studies had limited capabilities, sutures were not evaluated. The influence of CT on the management of patients with craniosynostosis has been questioned. In one series of 13 patients diagnosed as having craniosynostosis by skull radiographs and radionuclide scans, CT was useful in the management of only one patient, who was evaluated with CT for possible shunt failure.10

However, no previous study has evaluated the use of high-resolution CT with bone definition algorithms to directly examine the abnormal anatomy of sutures of the calvaria and skull base, nor have these findings been compared with the operative and pathological findings. The results of this study indicate that CT is a very sensitive method with which to detect craniosynostosis, even in small sections of a suture.

Classifications of craniosynostosis based on anatomic considerations are numerous and problematic. There are no designations for some complex forms of cranio-
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FIG. 15. Photomicrographs showing lambdoid synostosis. **Left:** No overt bone fusion is present in this low-power view. The closely apposed bones have spicules of prominent new bone growth (arrows) that markedly narrow but do not obliterate the preexisting suture. The suture space (S) is composed of dense fibrous connective tissue. H & E, x 28. **Right:** Higher-power view of the specimen at left showing close apposition of bony spicules (arrows) with resultant narrowing of the suture (S) to a distance of less than 1 mm. H & E, x 46.

Synostosis. Furthermore, classifications usually do not take into account the various sutures and synchondroses of the skull base. Cohen has pointed out that what is important anatomically and therapeutically is the clear description of which suture or sutures are involved and the extent of involvement, irrespective of classification. High-resolution CT appears to be the most effective imaging technique for showing the presence and extent of these abnormalities.

In a recent study of imaging techniques for the diagnosis of craniosynostosis (Gellad FE, Haney PJ, Sun JCC, et al., in preparation), the accuracy of skull radiographs, CT scans, and radionuclide scans was 89%, 89%, and 58%, respectively. In our study we found that CT is more sensitive for the diagnosis of abnormalities of sutures than are plain skull films, especially when the area fused was limited or at the skull base. Thin-section scans are the most sensitive and can be used for three-dimensional image reformation. Anteroposterior and lateral projection radiographs obtained on the scanner may be used to augment axial and direct coronal CT scans to show overall head shape and to distinguish vascular structures that may simulate normal sutures. In our limited experience with one case of microcephalus, the CT scan was extremely valuable in showing that occipital flattening was not caused by craniosynostosis and that the hypotelorism could be explained on the basis of early closure of the metopic suture secondary to microcephalus.

Associated intracranial brain parenchymal abnormalities are easily detected on CT. Nine (38%) of our 24 patients had some degree of ventricular enlargement or hydrocephalus, an incidence that falls between the 31% reported by Goldstein and Kidd and the 67% reported by Marsh and Gado, who included only craniofacial dysostosis. In two of our nine patients with hydrocephalus (one with cloverleaf and one with bilateral coronal and lambdoid synostosis), the fourth ventricle was so small that aqueductal stenosis was suspected. It was not possible to distinguish between primary aqueductal stenosis and the constrictive effect of the cranial malformation. Other parenchymal abnormalities, such as sulcal and ventricular compression, abnormally prominent subarachnoid spaces, and defects in the corpus callosum, have been described. The CT scan shows that even isolated synostosis may affect growing brain in children to varying degrees. A statistical correlation between elevated intracranial pressure (ICP) in craniosynostosis and poor mental development has been reported.

In general, CT can show three varieties of changes in the calvaria and skull base in patients with craniosynostosis: local changes at the site of the involved suture, such as complete obliteration of the involved suture, parasutural sclerosis, bony ridge over the narrow suture, and enostosis; calvarial and skull base changes directly secondary to premature suture synostosis, such as thickening of the lowest portion of the coronal suture.
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and foreshortened anterior cranial fossa in the case of the coronal synostosis; and compensatory calvarial bulging to maintain brain volume, such as bulging of the anterior ipsilateral portion of the calvaria in the case of unilateral lambdoid synostosis. Multiple craniosynostoses should be carefully examined by CT so that areas of abnormal closure and secondary anatomical distortion can be identified and effective surgical management can be planned.

Specific CT findings varied with suture location. In complete bilateral coronal synostosis (six cases), no trace of the suture was detectable, nor was there a bony ridge to indicate where it had been. Parasutural sclerosis was seen in only two of 11 patients with coronal synostosis (Fig. 10 right). Sagittal and, to a lesser extent, lambdoid synostosis was characterized by thickened bony ridges on the inner and outer tables, with or without a deep groove on the inner table. Complete metopic synostosis was marked by local bony thickening at the suture site. Endostosis extending as a cleft between two frontal lobes was also characteristic of metopic synostosis. More complicated changes seen in coronal synostosis included thickening of the more inferior portion of the coronal suture, a superiorly positioned frontozygomatic suture, and a double sphenoid ridge. In one patient with unilateral coronal synostosis, there was a reverse harlequin eye sign (Case 14, Fig. 9 right pair). It was thought that growth of the sphenoid ridge on the affected side was limited by the very early closure and dural adhesion of the lowest portion of the coronal suture.

The frontosphenoidal suture may be viewed as the inferior portion of a continuous ring with the coronal suture, and therefore must be considered in any maldevelopment of the anterior cranial fossa. The lateral and inferior portions of the frontosphenoidal suture are well delineated by CT, while only the lateral portion is seen on skull radiographs. However, as shown in Fig. 11, the frontosphenoidal suture does not seem to be ossified despite total bone fusion of both coronal sutures in a 1-month-old patient. Distinguishing fibrous union from non-union remains difficult. Obviously, more studies will be needed to determine the contribution of the frontosphenoidal suture in coronal suture synostosis.

The reason why the appearance of sutures in complete craniosynostosis of coronal sutures should differ so greatly from that of complete sagittal craniosynostosis is not clear. A possible explanation is that the attachment of the falx and the dura of the superior sagittal sinus to the skull vault beneath the sagittal suture may protect it from the direct remodeling influence of elevated ICP, whereas the fused coronal suture directly overlies the dura and pulsating brain, which in the presence of elevated ICP may suppress the formation of a bony ridge around the fused suture.

In this series, operative and pathological findings correlated well with the CT findings. In addition to confirming the complete suture fusion seen on CT scans, careful histological examination of suture specimens shows that, when the involved suture had an asymmetrically narrowed lucent zone with parasutural sclerosis or a bony ridge, fibrous union with various amounts of new bone formation from both sides of the suture was invariably present. The pathological findings strongly support the CT evidence that complete bone fusion of a cranial suture is not necessary and fibrous union can cause the clinical sequelae of craniosynostosis.

Pathological examination also showed that in virtually all patients there was histological evidence of progressive suture fusion. In contrast, Albright and Byrd reported that, in a series of patients with craniosynostoses for which specimens were examined histologically, there were three identifiable regions: a region of complete bone fusion, an adjacent area of encroachment of calvarial bones, and an entirely normal distant suture. Fourteen of their 19 patients had sagittal synostosis, and in no patient were lambdoid sutures examined. They concluded that craniosynostosis begins in one area of a suture and that fusion progresses along the suture. While we reached the same conclusion, our histological evidence suggests that the process is a more gradual progression from fusion to near fusion with prominent outgrowths of new bone to normal distant suture. It is as if the fusion process begins as a localized process, perhaps initiated by local growth hormones or organizers, and over a period of time progressively involves the remaining suture, somewhat in the manner of a door slowly closing about a hinge.

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