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Craniosynostosis: Diagnostic Imaging with Three-dimensional CT Presentation

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PURPOSE: To measure diagnostic performance and preference of two three-dimensional CT reconstruction modalities (voxel-gradient and surface-projection) displayed two ways (conventional and unwrapped) in craniosynostosis confirmed by surgical inspection and histologic analysis of resected sutures. **METHODS:** High-resolution 2-mm contiguous CT sections were obtained and three-dimensional reconstruction images generated for 25 infants and children with skull deformities before surgical treatment of craniosynostosis. Two pediatric radiologists and two neuroradiologists first ranked images by their own preferences for diagnostic use. Then they diagnosed craniosynostosis from images presented in random order and blinded. The standard of reference was inspection during surgery and histologic evaluation of excised sutures. Finally, reviewers repeated their subjective preference tests. **RESULTS:** The least experienced radiologist had 100% sensitivity for all imaging modalities and specificities ranging from 43% to 83%. The two most experienced radiologists performed nearly identically, with sensitivities of 96% and specificities of 100%. After performing diagnostic tasks using all image types, all radiologists preferred conventional surface projections. **CONCLUSION:** Experienced readers can achieve nearly perfect diagnostic performance using the latest three-dimensional CT reconstruction images, making it a contribution to the diagnostic process. Although performance is nearly identical for all modalities, readers strongly prefer conventionally presented surface-projection images.

Index terms: Skull, abnormalities and anomalies; Skull, computed tomography; Computed tomography, image display; Computed tomography, technique; Computed tomography, 3-D; Pediatric neuroradiology

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Radiography plays an important role in diagnosing craniosynostosis and planning its surgical treatment. It is generally agreed that three-dimensional computed tomography (CT) is the most useful imaging modality for planning surgical management (1–4). It has been demonstrated that CT allows more accurate diagnosis than plain radiography (5–7), and that 3-D CT reconstructions allow more accurate diagnosis than ordinary CT (6, 7). In the research reported here, we measure diagnostic performance of, and reader preference for, two greatly improved 3-D CT rendering methods (voxel-gradient and surface-projection) and a technique of image presentation (unwrapped). The diagnoses from these images are compared with a standard of reference comprising both inspection during surgery and histologic analysis of resected sutures.

Materials and Methods

The patient population consisted of 25 infants and children with skull deformities who underwent surgery for primary nonsyndromal craniosynostosis. Craniosynostosis

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was present in one or more calvarial sutures; the distribution according to the surgeon's report follows.

Type of Craniosynostosis	Number of Patients
Metopic	2
Unicoronal	7
Bicoronal	2
Sagittal	10
plus partial bilambdoidal	2
Unilambdoidal	2
Total	**25**

Patients entered the study prospectively between mid-1989 and mid-1991. The mean age of patients at surgery was 167 days, with a range from 62 to 484 days. Nine of the patients were girls and 16 were boys. With the exception of one black infant, all patients were white. Patients were entered into the study consecutively, with no attempt at randomization. Patients were excluded from the study if they did not undergo surgery or CT scans before surgery or if their craniosynostosis was syndromal (eg, Apert, Crouzon, or Carpenter) or not primary (eg, the result of shunting for hydrocephaly).

Surgery was performed at the St Louis Children's Hospital, Cleft Palate and Craniofacial Deformities Institute (23 patients) and at the Loma Linda University Medical Center (2 patients). Each infant and child in the study underwent surgical treatment with extended craniectomy and calvarial recontouring; some also received superior orbital reconstruction. In the course of surgery, both abnormal (synostosed) and portions of regionally adjacent clinically normal sutures were resected to establish calvarial symmetry. The resected sutures had a hole drilled in one end to indicate orientation, were preserved in formalin solution immediately after resection, and were labeled with information documenting the patient, suture, and orientation. A total of 66 excised sutures were included in the study (Table 1).

Histologic analysis took place in the Surgical Pathology Laboratory of St John's Mercy Medical Center. After decalcification using ethylenediaminetetraacetic acid solution for 1 to 2 weeks, suture specimens were serially cross-sectioned perpendicular to the sutural interface at 3-mm intervals and entirely submitted in embedding cassettes. Routine tissue processing and hematoxylin and eosin stains were used. Suture orientation was maintained by the application of tattoo dye, identifiable on routine microscopy, to the external surface. All sections from each case were reviewed and graded independently by two pathologists (B.B.K. and S.R.) on a five-point scale, which follows.

0	Completely patent suture
1	Minimal osseus encroachment on fibrous suture: no overlapping bony trabeculae
2	Fibrous suture predominates; overlapping bony projections
3	Minimal persistence of fibrous suture; bony trabeculae traverse suture
4	Fused suture

The standard of reference in this study was derived by comparing inspection during surgery and histologic inspection of resected sutures. Comparison of these independent sources of information showed that, if the division between pathologically normal and abnormal was set between categories 2 and 3, there were disagreements with surgical truth in only 6 of 66 sutures (Fig 1). Four of the disagreements concerned the two patients with partial bilateral lambdoidal synostosis. The synostosed portions of the lambdoidal sutures were excised with the synostosed sagittal suture, but the normal portions were excised separately and correctly judged to be histologically normal. The other two disagreements were metopic sutures, which had closed normally. The surgeon rated them normal, but the pathologists, who were blinded and did not consider the age of the patient, identified them as closed. In all 6 cases, therefore, the disagreements resulted from procedural difficulties, and the surgeon's findings were used as the standard of reference.

Patients were scanned using 1.5- to 2-mm contiguous sections. Scanning took place an average of 29 days before surgery, with a range of 0 to 105 days. Three-dimensional reconstruction images were created using Analyze software (Mayo Clinic, Rochester, Minn) (8, 9),

TABLE 1: Numbers of excised sutures analyzed in the study

Suture	Number Excised	Normal[a]	Abnormal[a]
Metopic	8	6	2
Left coronal	13	6	7
Right coronal	16	12	4
Sagittal	12	0	12
Left lambdoidal	8	6	2
Right lambdoidal	9	5	4
Total	66	35	31

[a] Diagnosis is according to surgeon's report.

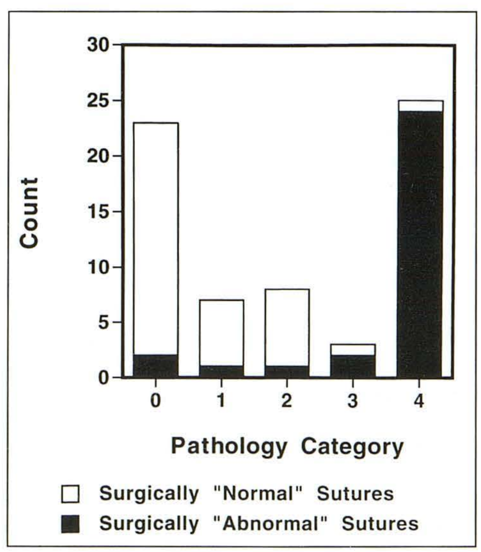


Fig 1. Comparison of two bases of diagnostic truth: surgical observations and pathologic categories.

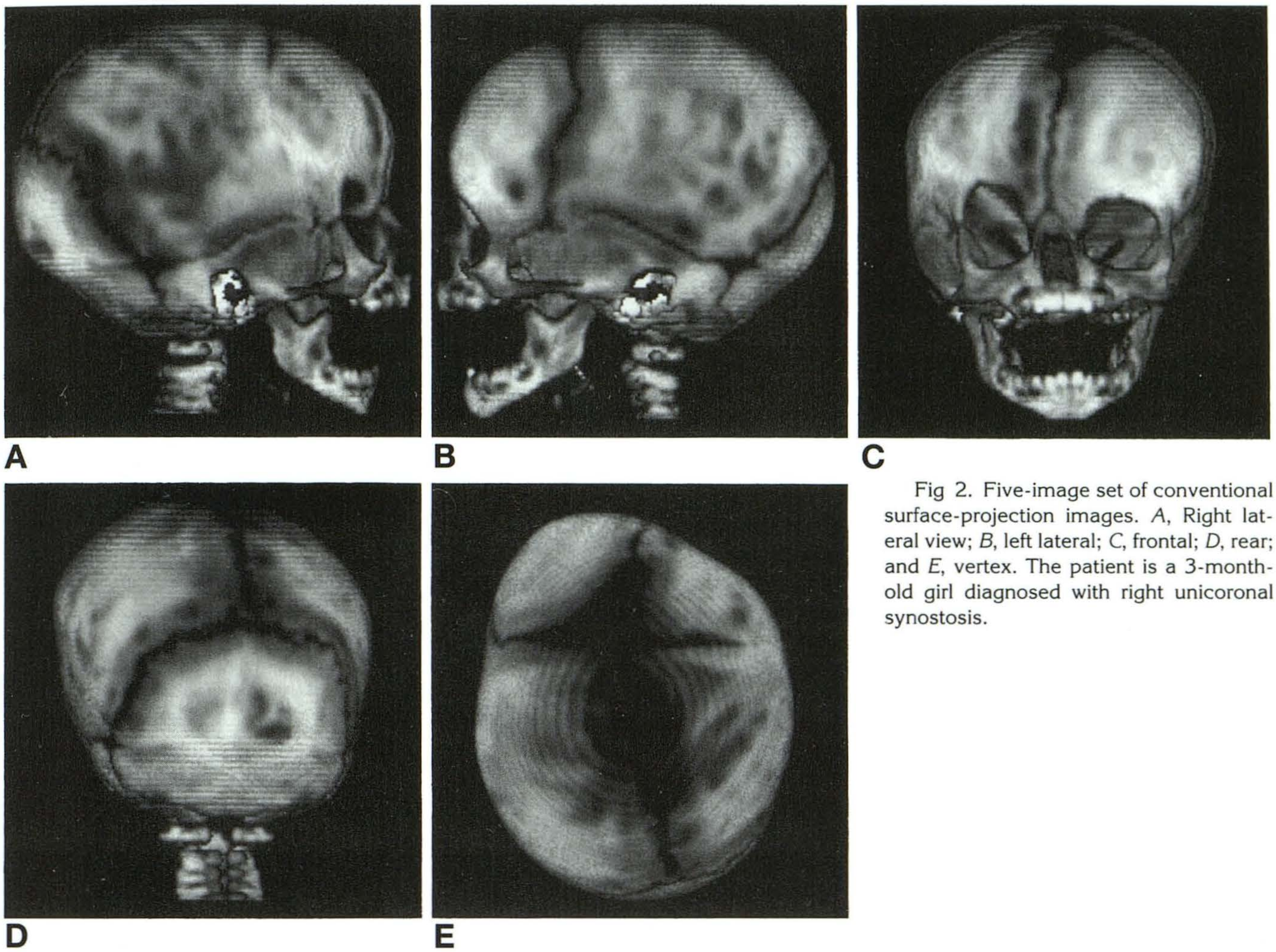


Fig 2. Five-image set of conventional surface-projection images. *A*, Right lateral view; *B*, left lateral; *C*, frontal; *D*, rear; and *E*, vertex. The patient is a 3-month-old girl diagnosed with right unicoronal synostosis.

version 5.0, on a Sun SPARCstation 2 GX (Sun, Mountain View, Calif). Hard copies were produced on a Seikosha thermal video printer with 64 gray levels, marketed as a Codonics VP 3500 (Codonics, Middleburg Heights, Ohio).

Two types of 3-D images were generated: voxel-gradient rendering and surface-projection rendering. Voxel-gradient rendering treats the computer reconstruction as a solid object and creates the image by calculating the angle at which a ray of light originating at the observer would strike the object. The closer the surface is to a right angle relative to the light ray, the brighter that portion of the image. Therefore, flat surfaces are highlighted, and angled surfaces are shaded (Fig 2). Although these principles are identical to those used to create the images in the most recent diagnostic test of 3-D imaging of craniosynostosis (6, 7), the images themselves are much improved because the algorithms that create the object surface preserve much more detail.

Projection rendering creates the image by calculating cumulative object density along a ray perpendicular to the display surface. In a situation directly analogous to x-ray films, the brightness of the image is determined by the density. If the rendering makes brightness inversely proportional to cumulative density, as ours did, the image will be very similar to an x-ray film. Our images had two important improvements over x-ray images or previously evaluated (6, 7) projection (then called volumetric) images. First, images were rendered using only the half the skull facing the viewer. This technique eliminates the possibility of confusing anatomic features from different sides of the skull. Second, we used surface projection, which calculates cumulative density only for those features a chosen distance below the object surface. Because calvarial shape and suture characteristics are both surface or near-surface features, we were able to select only the information relevant to them and eliminate potentially confusing internal structures (Fig 3).

Images were shown in frontal, posterior, right and left lateral, and vertex orientations (Figs 2 and 3). In addition to these conventional, or orthographic, presentations, images were also created in unwrapped (also known as cylindrical map or panoramic) form (Figs 4 and 5). An unwrapped image is created by placing a linear axis through the object. The location of points in the image is determined by distance along the axis and angular location about it, rather than by location in 3-D space. This is the

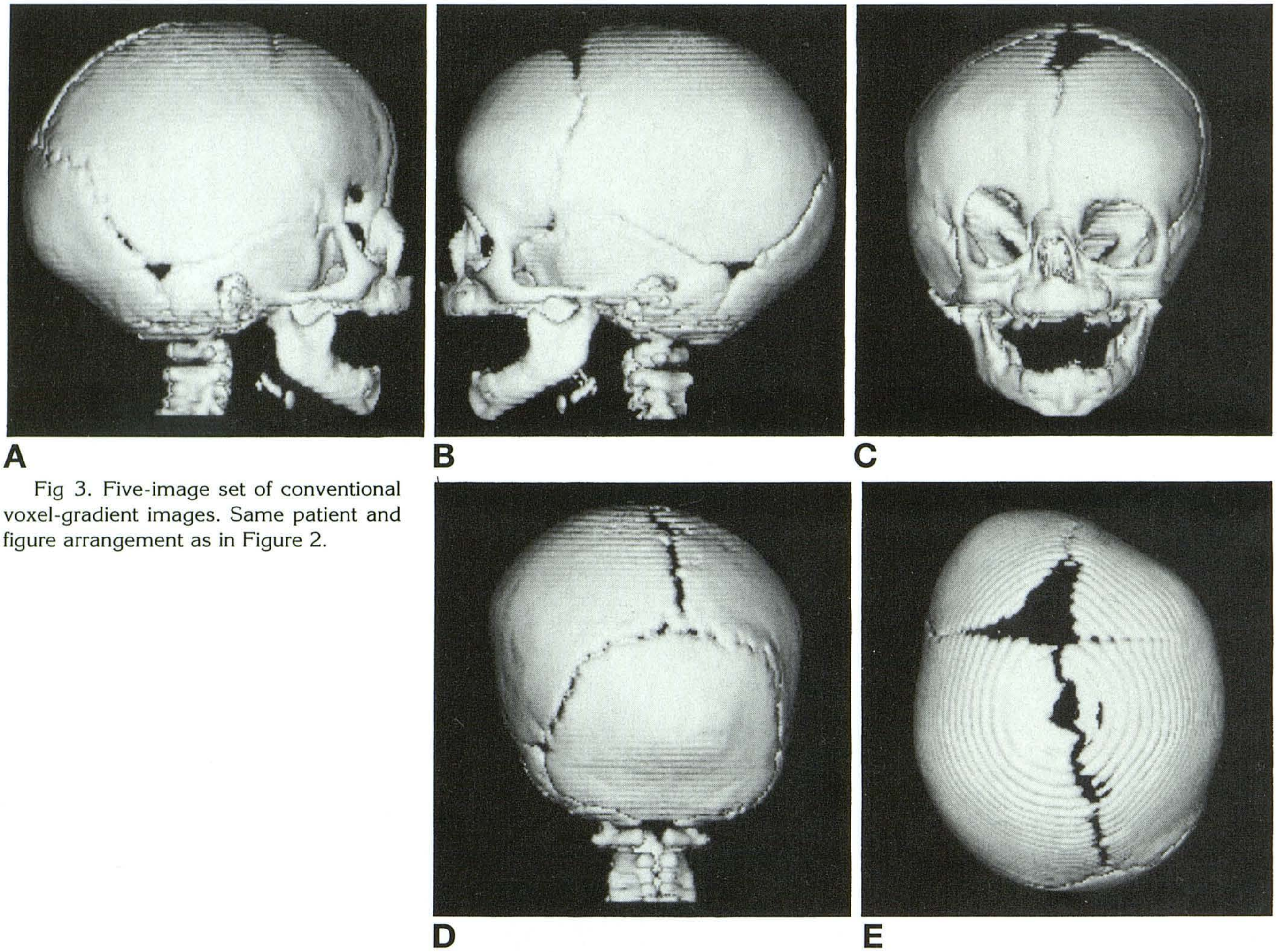


Fig 3. Five-image set of conventional voxel-gradient images. Same patient and figure arrangement as in Figure 2.

principle of the well-known Mercator projection used in mapping and results in distortion proportional to the object's deviation from a cylindrical shape.

Unwrapped images have two theoretical advantages over conventional images. First, the projection rays are always orthogonal to the surface of the skull, rather than to the plane of the image, as is the case with conventional views. This eliminates the "blotting out" of the suture at the edges of a projection image, where the curvature of the skull increases the quantity of bone through which the projected ray must pass. Second, the entire skull can be viewed at once, which makes it easier to examine bilateral sutures for symmetry.

Images were mounted in sets of three or five for each patient on gray matte board (Figs 2–5). Each board was identified with a random number. No information except the images and the random numbers was presented to the observers.

The four readers in this study (M.H.G., C.J.M., W.H.M., and G.D.S.) were all board-certified radiologists, with a minimum of 15 years experience. Two were neuroradiologists, who had had less exposure to craniosysnostosis and no experience making this diagnosis from 3-D CT reconstruction images. Two readers were pediatric radiologists, who were experienced in the evaluation and diagnosis of craniosynostosis and who had both participated in a previous study using 3-D CT reconstructions in the diagnosis of craniosynostosis.

We wished to measure two aspects of using these images: diagnostic performance and subjective reader preference. To measure subjective preference, readers were presented with all images for each patient, one patient at a time, and asked to rank the images in terms of their perceived ability to diagnose craniosynostosis using them. The random numbers assigned to the images determined the left-to-right presentation order, although readers were told they were free to change the order of the images if they thought this would aid them in making comparisons. The same images were presented in identical fashion before and after the diagnostic phase, and ranking was done the same way. Because these imaging modalities were unfamiliar to all reviewers at the beginning of the study, we were interested to see how well first impressions related to both diagnostic performance and subjective preference after diagnostic use.

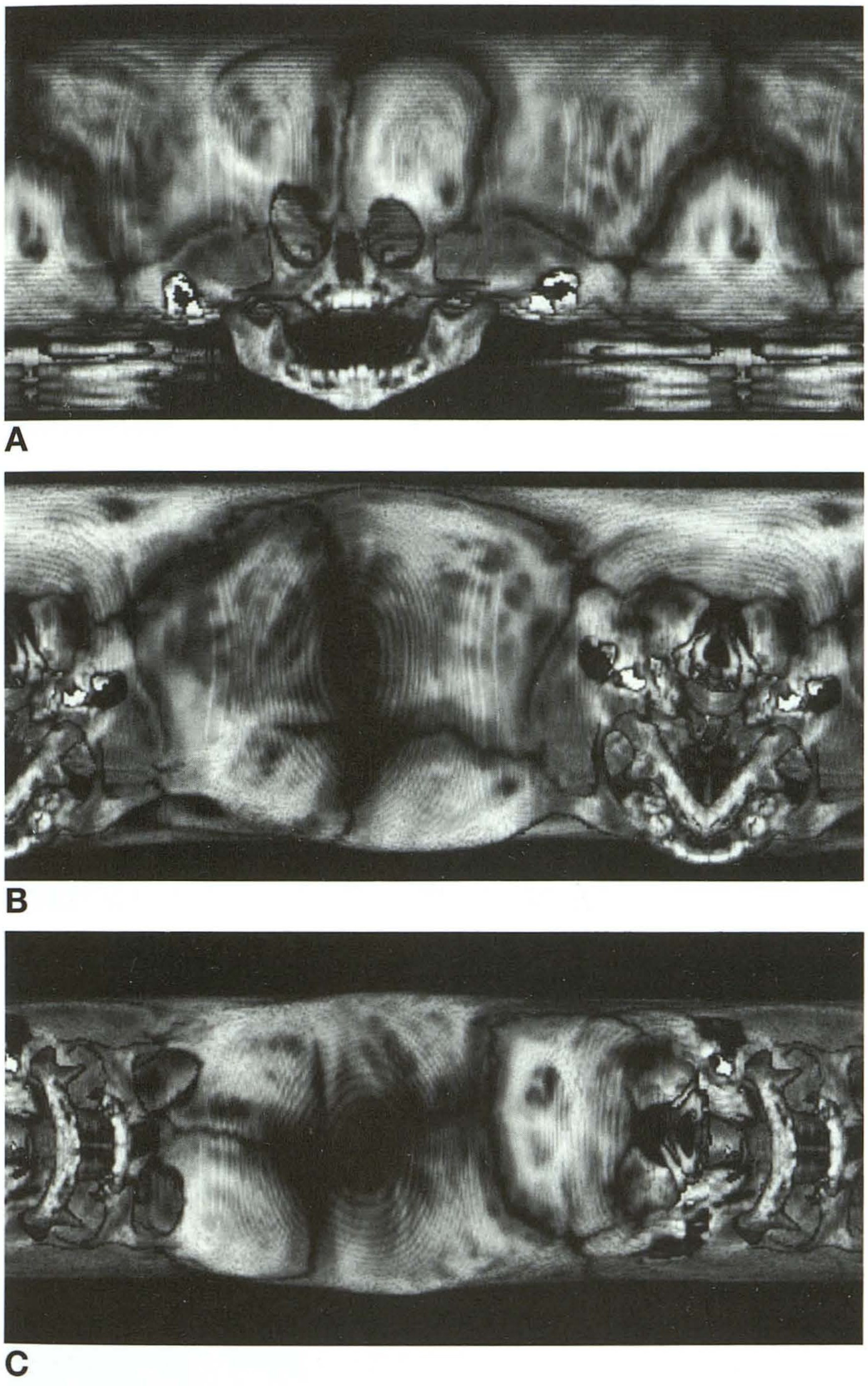
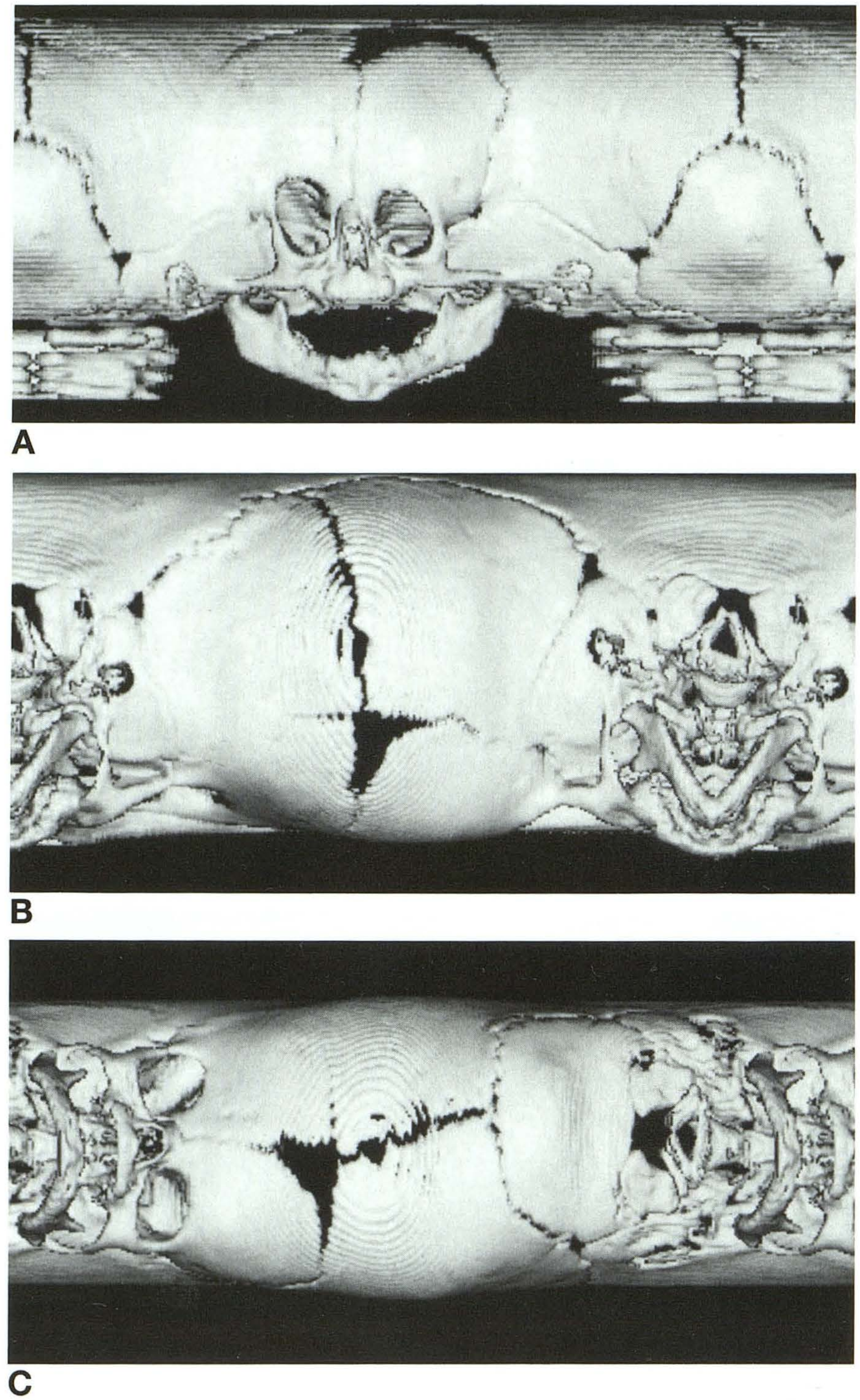


Fig 4. Three-image set of unwrapped surface-projection images. *A*, Unwrapped about a vertical axis; *B*, unwrapped about a horizontal (anteroposterior) axis; and *C*, unwrapped about a horizontal (lateral) axis. Same patient as in Figures 2 and 3.

For testing diagnostic performance, readers were presented with one board, containing one rendering and presentation combination, at a time. Presentation order of the images was determined by their randomly assigned numbers, so the images were randomly ordered with regard to both image type and patient. For each image set, readers were asked to diagnose craniosynostosis for six sutures: metopic, sagittal, right and left coronal, and right and left lambdoidal. Diagnosis was made in the form of a six-point scale (definitely or almost definitely abnormal, probably abnormal, possibly abnormal, possibly normal, probably normal, and definitely or almost definitely normal).

Results for the metopic sutures were not used in the analysis, because the diagnostic task was different from the rest. In most patients the metopic suture had closed normally at the time of the CT scan, so the readers were required to judge whether it had closed at the appropriate time rather than if it was patent or not, which was the task for the rest of the sutures.

Fig 5. Three-image set of unwrapped voxel-gradient images. Same patient and figure arrangment as in Figure 4.



In addition to making the diagnosis, readers assessed the quality of the image set for evaluating each suture on a three-point scale (good, satisfactory, or poor). Before beginning this phase of the study, each reader was presented with a set of standardized instructions.

Results

Before diagnostic use, the readers clearly preferred surface-projection images to voxel-gradient images. Three readers strongly pre-

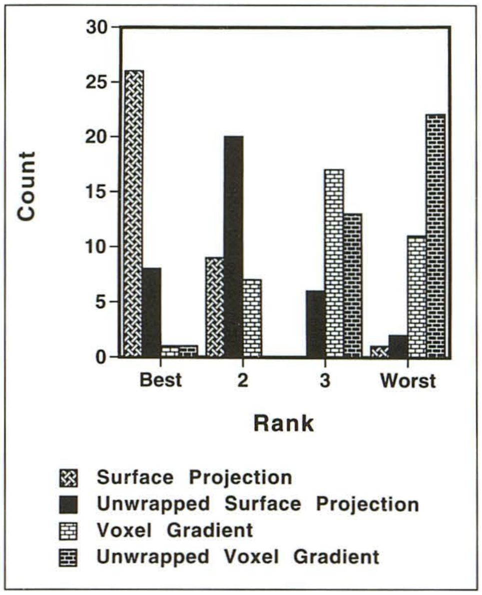


Fig 6. Subjective image preferences for all readers combined. These preferences were obtained before the observers used the images to perform diagnostic tasks.

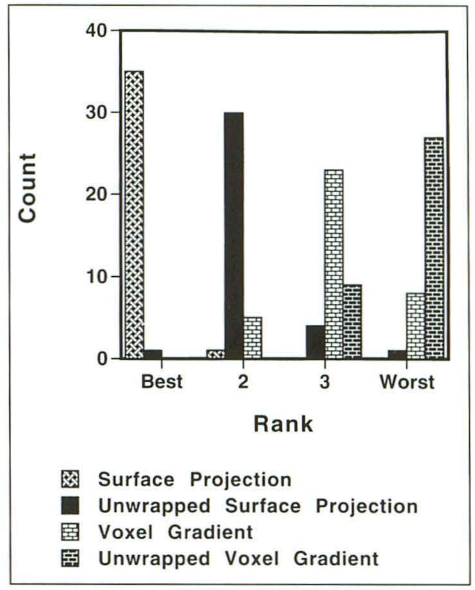


Fig 7. Subjective preferences for all readers combined. These preferences were obtained after the observers had used the images to perform diagnostic tasks.

ferred surface-projection images to be displayed conventionally, and one reader slightly preferred unwrapped images. When the preferences of all individuals were combined, there was a clear tendency to prefer surface-projection over voxel-gradient renderings and to prefer images in conventional rather than unwrapped form (Fig 6). After diagnostic use, readers consistently preferred conventionally presented surface-projection images to all others (Fig 7). Their second choice was consistently panoramic surface-projection images. Third and last choices were not so consistent, but generally conventional voxel-gradient images were the third choice, and panoramic voxel-gradient images were last.

Although this study was designed from the outset for receiver-operator characteristic (ROC) analysis, the level of performance was so high with these images that ROC analysis was impossible in almost all cases. The ROC programs we used, ROCFIT and CORROC2 (C.E. Metz, University of Chicago) (10), were unable to fit ROC curves to the data in all but 2 of the 16 cases. In these 2 cases, A_z, the area under the ROC curve, equal to the proportion of correct responses regardless of the decision threshold, was greater than 0.99. In place of ROC analysis, sensitivity and specificity were compared. Sensitivity and specificity were nearly perfect for most sutures, most imaging modalities, and most readers (Table 2). The deviations from 100% sensitivity for the pediatric radiologists were all attributable, with one exception, to the atypical presentation of one patient.

Although the radiologists who participated in the study achieved similar diagnostic perfor-

TABLE 2: Sensitivity and specificity, by imaging modality, for the four readers in the study

Reader	Imaging Modality	Sensitivity	Specificity
Neuroradiologist 1	Surface-projection	100.0	60.0
	Panoramic surface-projection	100.0	83.3
	Voxel-gradient	100.0	46.7
	Panoramic voxel-gradient	100.0	43.3
Neuroradiologist 2	Surface-projection	89.3	100.0
	Panoramic surface-projection	92.9	93.3
	Voxel-gradient	96.4	96.7
	Panoramic voxel-gradient	96.4	100.0
Pediatric radiologist 1	Surface-projection	96.4	100.0
	Panoramic surface-projection	96.4	100.0
	Voxel-gradient	92.9	100.0
	Panoramic voxel-gradient	96.4	100.0
Pediatric radiologist 2	Surface-projection	96.4	100.0
	Panoramic surface-projection	96.4	100.0
	Voxel-gradient	96.4	100.0
	Panoramic voxel-gradient	96.4	100.0

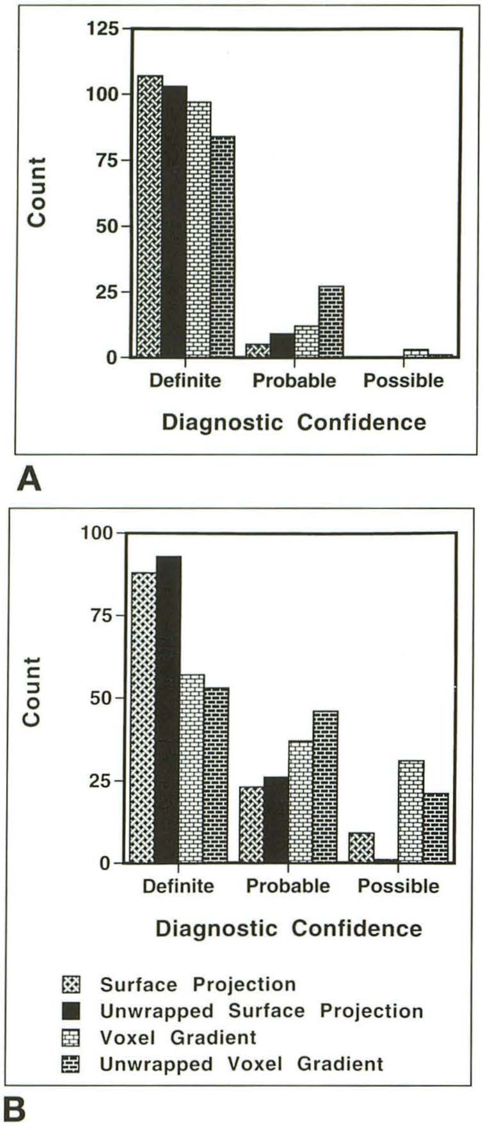


Fig 8. Diagnostic confidence for all readers combined, by image type and suture diagnosis. *A*, Abnormal sutures; *B*, normal sutures.

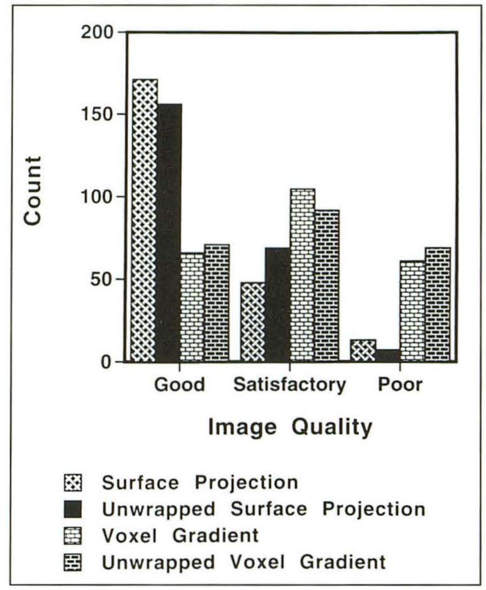


Fig 9. Evaluations of image quality for all readers combined, by image type.

mance with all imaging modalities, their levels of confidence in their diagnoses, as measured by relative strength of the diagnostic categories they chose, varied with the imaging modality (Fig 8). The differences in confidence were clearest with the normal sutures. Readers were most confident with the surface-projection images, particularly when they were conventionally displayed, and least comfortable with the voxel-gradient images, particularly the unwrapped versions. The differences in level of confidence were echoed in subjective evaluations of image quality (Fig 9). Surface-projection images were given the highest-quality ratings, with conventional images slightly favored over unwrapped ones. Voxel-gradient images received quality ratings markedly lower than surface-projection images, with no real difference between conventional and unwrapped images.

Discussion

The difficulty of diagnosing craniosynostosis varies by case. In some cases, especially of the simpler abnormalities, the diagnosis is clear. For more complex abnormalities the diagnosis can be more difficult. Improvements in radiologic imaging have improved its accuracy in diagnosing craniosynostosis consistently by making the more difficult cases clearer. CT allows more accurate diagnosis than plain radiography (5–7), and 3-D CT reconstructions allow more accurate diagnosis than ordinary CT (6, 7). This study, using the most recent 3-D CT algorithms, found nearly perfect performance with experienced readers. Gellad et al suggested that plain radiography is adequate for the diagnosis of most craniosynostosis, but that CT could be useful in cases not clearly positive or negative (5). Our results suggest that 3-D CT reconstructions may perform the same role in relation to CT.

Although all the 3-D reconstruction algorithms we tested achieved similar levels of diagnostic performance, and therefore have the same clinical potential, our readers strongly preferred conventionally displayed surface-projection images. The pediatric radiologists, who were most familiar with the diagnosis of craniosynostosis, described their pattern of viewing images as first looking at calvarial shape to see whether there was a reason to suspect craniosynostosis, and second, looking at the sutures to see which were open. The unwrapped images required more effort to interpret shape, and the voxel-gradient images required more effort to interpret suture patency. Although we did not record the viewing time necessary to reach a diagnosis, it was clear from observation that the readers required the least time to make diagnoses from the conventionally displayed surface-projection images.

Acknowledgments

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