Dehiscence or Thinning of Bone Overlying the Superior Semicircular Canal in a Temporal Bone Survey

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Objective: To determine the incidence and etiology of dehiscences of bone overlying the superior semicircular canal in a temporal bone archive.

Design: A microscopic study was performed of 1000 temporal bones from 596 adults in a university hospital registry. Specimens were sectioned vertically in the plane of the superior semicircular canal. Measurements of minimum bone thickness over the superior canal were made in a subset of 108 randomly chosen specimens. All bones were examined for thinning or dehiscence relative to these norms. Clinical histories, when available, were reviewed.

Results: Complete dehiscence of the superior canal was identified in 5 specimens (0.5%), at the middle fossa floor (n = 1) and where the superior petrosal sinus was in contact with the canal (n = 4). In 14 other specimens (1.4%), the bone at the middle fossa floor (n = 8) or superior petrosal sinus (n = 6) was no thicker than 0.1 mm, significantly less than values measured in the control specimens (P < .001). Abnormalities were typically bilateral. Specimens from infants demonstrated uniformly thin bone over the superior canal in the middle fossa at birth, with gradual thickening until 3 years of age.

Conclusions: Dehiscence of bone overlying the superior canal occurred in approximately 0.5% of temporal bone specimens (0.7% of individuals). In an additional 1.4% of specimens (1.3% of individuals), the bone was markedly thin (< 0.1 mm), such that it might appear dehiscent even on ultra–high-resolution computed tomography of the temporal bone. Sites affected were in the middle fossa floor or a deep groove for the superior petrosal sinus, often bilaterally. These abnormalities may arise from failure of postnatal bone development. Thin areas of bone over the superior canal may be predisposed to disruption by trauma.

Arch Otolaryngol Head Neck Surg. 2000;126:137-147

MINOR et al identified a syndrome related to dehiscence of bone overlying the superior semicircular canal. Vertigo, oscillopsia, and/or dysequilibrium resulted from sound, changes in middle ear pressure, and/or changes in intracranial pressure in these patients. Vertical-torsional eye movements were induced by these sound and pressure stimuli and documented by measurement of the eye movements in 3 dimensions. Ultra–high-resolution computed tomographic (CT) scans of the temporal bone demonstrated dehiscence of the bone overlying the affected superior semicircular canal. The dehiscence has been confirmed in surgical explorations of the middle cranial fossa, with plugging or resurfacing of the defect in 5 cases.

The eye movements evoked in clinical tests are those anticipated for excitation or inhibition of the affected superior semicircular canal. Excitation of this canal results from stimuli including the Valsalva maneuver against pinched nostrils and positive pressure in the external auditory canal. We hypothesize that these maneuvers create a positive pressure gradient from the middle ear to the labyrinth at the round and/or oval windows. The bone dehiscence represents a third mobile window, allowing dissipation of the pressure as the membranous labyrinth bulges into the adjacent dura and endolymph flows away from the ampulla. The resulting ampullofugal deflection of the superior semicircular canal cupula is excitatory. Conversely, stimuli causing opposite eye movements (Valsalva maneuver against a closed glottis, jugular venous compression, and negative pressure in the external canal) create a positive pressure gradient from the middle cranial fossa to the labyrinth at the dehiscence. The round or oval window dissipates this gradient, and the endolymph displacement is in-
MATERIALS AND METHODS

Temporal bones were extracted, fixed in formaldehyde, decalcified, dehydrated, and embedded in 35% nitrocellulose. Specimens were sectioned in a vertical plane that was perpendicular to the long axis of the petrous portion of the temporal bone. The sections were 24 µm thick, and every tenth section was mounted and stained with hematoxylin and eosin. Most of the specimens were obtained from autopsies performed at The Johns Hopkins Hospital from 1925 through 1975. Clinical histories and audiograms were available for many of the specimens in the collection.

Each temporal bone was screened by direct visual inspection and examination under a dissecting microscope at ×4 magnification. All bones with visibly less than 1 mm of bone over the superior canal were further examined under higher magnification. Measurements were made on digital images of the thickness of bone in 2 places. First, bone was measured at its thinnest point between the lumen of the bony superior canal and the middle cranial fossa floor. Second, the bone thickness was measured between the lumen of the bony superior canal and the depression for the superior petrosal sinus. In specimens in which the sinus did not form a discernible impression, the minimum distance between the lumen of the superior canal and the posterior fossa was recorded.

Images were captured with a charge-coupled device camera (CCD; Hamamatsu USA, Bridgewater, NJ) via a 50-mm, 1:2 macro lens (Olympus America, Melville, NY) mounted above a light table for ×15 total magnification. For measurements that were less than 0.1 mm, images were taken from an Axioskop microscope (Carl Zeiss, Inc, Thornwood, NY) with a CCD camera (Photometrics Ltd, Tucson, Ariz) in the magnification range ×15 to ×62.5. Images were digitally stored and analyzed on a Macintosh personal computer (Apple Computer, Inc, Cupertino, Calif) using a program available on the Internet (NIH Image, version 1.58; developed at the US National Institutes of Health, http://rsb.info.nih.gov/nih-image/). Reconstructions were performed on a personal computer (Gateway, North Sioux City, SD) using software available on the Internet (Scion Image Software; http://www.scioncorp.com/).

Results are presented as mean ± SD. Comparisons between groups are made with 2-sample t tests assuming unequal variances.

RESULTS

SURVEY OF ADULT TEMPORAL BONES

We screened 1000 temporal bones from 596 adults in The Johns Hopkins Temporal Bone Collection. There were right-left pairs from 404 individuals and single bones from 192 individuals. We first established normative values for the thickness of bone overlying the superior canal at sites where dehiscences were found. For this, we took measures at each site from temporal bones that did not display dehiscences or thinning (control specimens: 108 bones, including pairs from 45 individuals and single bones from 18 individuals).

The minimum thickness of bone overlying the superior canal was measured in adult specimens at the following 2 locations: the floor of the middle cranial fossa and the superior petrosal sinus. The latter was usually discernible as a distinct groove in the bone on its medial surface. In Figure 1, representative photomicrographs demonstrate the range of bone thickness overlying the superior semicircular canal encountered in our study. Pneumatization (Figure 1, A) accounts for the largest values of bone thickness overlying the canal. Pneumatized specimens typically measured thicker than 1.5 mm in the middle fossa and thicker than 4 mm in the posterior fossa. More commonly, we encountered nonpneumatized bone thicker than 0.10 mm at these locations (Figure 1, B and C). We considered bone no thicker than 0.10 mm over the canal to be abnormal (Figure 1, D).

The mean thickness in 100 control specimens for the middle fossa and superior petrosal sinus was 0.96 ± 0.61 mm (range, 0.13-3.28 mm) and 1.79 ± 1.20 mm (range, 0.13-6.24 mm), respectively. Histograms showing the distribution of bone thickness measured in the control specimens at both locations are shown in Figure 2. The broader distribution of thickness at the superior petrosal sinus results from the more frequent finding of pneumatization of bone between the sinus and the canal.

DEHISCENCE OF THE SUPERIOR CANAL IN ADULT TEMPORAL BONES

A dehiscence in the bone overlying the superior canal was identified in 5 (0.5%) of the specimens from 4 (0.4%) of the adults. One dehiscence was located between the super-
perior canal and the middle cranial fossa (Figure 3, A and B). The other 4 dehiscences in 3 individuals were found between the superior canal and the superior petrosal sinus (Figure 4 and Figure 5).

Figure 3 demonstrates the middle fossa dehiscence (parts A and B) and thinning of bone overlying the superior canal on the contralateral side (part C). Only dura and periosteum cover the membranous canal. This demonstrates the criterion we used when we encountered a fracture in thin bone overlying the canal. We relied on intact dura as a sign that bone had not been simply avulsed during processing. In approximately 3% of the specimens in the survey, however, a fracture occurred, and dura was disrupted. We did not consider such cases to be dehiscences, but, rather, damage resulting from processing. These specimens were excluded from further analysis.

Dehiscence at the superior petrosal sinus is demonstrated in Figures 4 and 5. The bony defect in the section containing the superior canal might appear to be a focal erosion (Figure 4, C). However, in each case we could trace the deep trough of the superior petrosal sinus along the length of the temporal bone, confirming that it was the cause of the dehiscence. This is demonstrated by computer-aided reconstruction (Figure 4, A). The temporal bone is opened like a book to the dehiscence at the superior canal. Sections anterior and posterior to this are shown to the right and left, respectively, indicating the course of the sinus.

Figure 5 demonstrates the only case of bilateral dehiscences. These specimens were taken from a 27-year-old man who had a large left vestibular schwannoma. He underwent an attempted resection in 1931, but died of brainstem complications postoperatively. On the left side (Figure 5, D-F), the deep trough of the superior petrosal sinus has been invaded by tumor. The tumor appears to be eroding bone, as evidenced by the irregular border and fingerlike projection of tumor into bone. In contrast, the right side (Figure 5, A-C) shows the more typical finding of mature, stable bone at the margin of the groove. Lamellae are seen parallel to the surface contour of the sinus, indicating that ossification conformed to the sinus and had remained stable for many years. This pattern was observed in other cases of thinning or de-
hiscence at the superior petrosal sinus. The tumor involvement of the left side was the only case of an erosive process that we found. Thus, with a single exception, we found lamellar bone at thin areas in the superior petrosal sinus, indicating that the process that led to the thin bone in this region was of long-standing duration.

THINNING OF THE SUPERIOR CANAL IN ADULT TEMPORAL BONES

In 14 specimens from 10 adults, the bone at one of these same locations was thin (≤0.1 mm) but not dehiscent. Eight of these occurred at the projection of the superior aspect of the canal into the middle fossa, where the bone was thinned to 0.04 ± 0.02 mm. Six occurred at the superior petrosal sinus groove where the bone was also thinned to 0.04 ± 0.02 mm. An example of bone thinner than 0.1 mm is shown in Figure 1, part D. Despite the fracture in the thin bone, it still forms a barrier between the dura and membranous canal, and thus we did not consider this case to be a dehiscence.

The Table summarizes measurements from the specimens and the associated clinical histories for the 12 individuals with thinning and/or dehiscence of the bone overlying the superior canal.

Bilateral involvement was a common feature. In addition to the individual with bilateral dehiscences at the superior petrosal sinus, 2 individuals had a dehiscence of a single superior canal and thinning of the contralateral one. In cases of thin but intact bone, there was also a strong correlation between the thickness of bone on both sides. For individuals with bone no thicker than 0.10 mm over the superior canal at the middle fossa in adults, the contralateral thickness at the middle fossa measured only 0.07 ± 0.05 mm. This was significantly less than the average thickness for adult controls (P<.001). At the superior petrosal sinus, bone no thicker than 0.10 mm on a given side was associated with contralateral thickness of 0.25 ± 0.30 mm, also significantly less than the average for adult controls (P<.001). Six of the 12 affected individuals had bilateral findings: bilateral dehiscences (1), unilateral dehiscence with contralateral thinning (2), or bilateral thinning (3).

Each specimen exhibiting thinning or dehiscence was examined for evidence of underlying bone disease that might account for the loss of bone over the canal. Aside from the vestibular schwannoma-related erosion noted in subject 2, we found none. Changes seen in osteoporosis, namely, thinning of trabeculae and relative increase in the fatty marrow,5 were absent in these specimens. Correspondingly, the thickness of bone overlying the superior canal was of normal thickness in 12 bones in the archive that exhibited osteoporosis. Otosclerosis was found around the oval window on both sides in subject 4; however, the semicircular canals were not involved. The incidence in this small group (8.3% of the individuals with thinning or dehiscence) is consistent with previous reports of an approximate 10% incidence of otosclerosis in large samples of temporal bones.5-7 Specimens from subjects (subjects 1, 3, 6, and 7) with a history of syphilis were examined for gummas as well as the nongummatous manifestations of otosyphilis, ie, periostitis, fibrosis, and deformation of the canals,8,9 but none was found. Pathologic changes of chronic otitis media were present in both ears from a subject with a history of middle ear disease (subject 7). However, the abnormality was confined to the middle ear and appeared unrelated to the thinning of the bone over the superior canal.

TEMPORAL BONES FROM INFANTS AND CHILDREN

The mean thickness of bone overlying the superior canal in the 36 specimens from 20 infants (≤1 year of age) measured 0.15 ± 0.15 mm, significantly thinner than that measured at the middle fossa floor in adults (P<.001). The superior petrosal sinus was usually not developed in these specimens, so only the measure of the middle fossa was taken. Figure 6 shows photomicrographs of specimens from 4 infants at 24 weeks estimated gestational age and 2, 4, and 10 months of age. In the premature infant (Figure 6, A), only a thin layer of inner peri-
osteal bone developed around the superior, horizontal, and posterior canals. This thin (0.02 mm) inner bone is the only covering of the superior canal at the middle fossa floor. Scattered intrachondral and endochondral ossification forms the middle layer, and outer periosteal bone completes the trilaminar structure. From 2 to 4 months after birth (Figure 6, B and C), bone thickens around each of the canals by fusion of the growing outer and middle layers with the thin inner layer. However, only a thin inner layer of bone covers the superior canal in the middle fossa, even up to 10 months of age (0.03 mm at 2 months; 0.10 mm at 4 months; and 0.11 mm at 10 months).

Figure 7 shows the relationship between mean bone thickness overlying the superior canal at the middle fossa and age from 27 pairs of temporal bones from infants and children no older than 4 years. In only 1 specimen was the bone overlying the canal extensively pneumatized (overall thickness, 3.51 mm). This atypical specimen was excluded from the data in this plot. The equation for the linear regression drawn was as follows:

\[
\text{Thickness} = (0.022 \text{ mm/mo}) \times \text{age} + 0.087 \text{ mm} \quad (R = 0.85)
\]

The line intersects the 99% confidence interval for adult middle fossa thickness (gray area) at 32.4 months of age. Thus, bone is added over the superior canal for almost 3 postnatal years, with the predicted thickness at birth being only 0.092 mm.

**CLINICAL CORRELATIONS**

Medical records were available for 3 of the 4 individuals with complete dehiscences. The 27-year-old man with bilateral dehiscences at the superior petrosal sinus had vertigo and ataxia, which were presumably related to the large vestibular schwannoma compressing his cerebellum and brainstem. Vertigo was not reported in the other 2 subjects. Caloric testing was only recorded for subject 4, who, interestingly, had no ice-water caloric responses. There was no history of head trauma noted in any of the subjects with complete dehiscence.

One of the 10 individuals with thin bone overlying the superior canal (subject 9) had a history of vertigo. Neither the nature of her vertigo nor the typical precipitating event was specified. However, nausea developed when audiometric testing was attempted. The examiner did not note whether vertigo or nystagmus accompanied the nausea. Caloric data were available in 3 subjects with thinning. Normal cold-water caloric responses were reported in subjects 8 and 12. Subject 11 had a prolonged warm-water caloric response on the right, the same side on which the superior canal was thin at the superior petrosal sinus.

High-frequency sensorineural hearing loss was noted in 3 subjects with thinning of the superior canal. Presbycusis may explain these findings, as these individuals were significantly older than the others with thinning (87 ± 10 vs 54 ± 17 years of age; \(P = .003, 2\text{-sample }t\text{ test with unequal variances}\)). Bilateral flat conductive hearing loss was noted in the patient with pathologic findings of bilateral otosclerosis.

Of the systemic diseases, syphilis occurred in 4 of the 12 subjects with superior canal thinning or dehis-
ence. As noted above, however, none of the temporal bone manifestations of syphilis was found in these cases.

COMMENT

The major finding of this temporal bone survey was that 19 (1.9%) of 1000 bones had dehiscence or thinning of the bone overlying the superior semicircular canal. Of 596 individuals, 12 (2.0%) had thinning or dehiscence, and in half of these individuals an abnormality was found on both sides. Limited clinical histories suggested that most of these individuals were not symptomatic. However, for reasons we shall discuss, such individuals may be predisposed to development of the syndrome if trauma disrupts thin bone or stable dura over the canal, particularly at the middle fossa.

INCIDENCE AND LOCATION OF DEHISCENCES

Defects in the bone at the tegmen tympani or tegmen mastoideum have been identified in previous temporal bone series. Åhren and Thulin10 found tegmen defects in 20 (21%) of 94 temporal bones examined macroscopically at autopsy. Lang11 examined 70 temporal bones and found tegmen defects in 14 (20%). Kapur and Bangash12 examined 50 intact temporal bones from cadavers and found tegmen or petromastoid defects in 17 specimens (34%). None of these studies, however, noted defects in the superior semicircular canal. Nager5 illustrated a single case of a defect of the superior canal in the middle cranial fossa. He attributed the bony defect to “senile osteoporosis of the petrous bone.” No clinical history accompanied the description. We could not find any temporal bone series that documented the prevalence of such defects in the bony superior canal.

Our study addresses the prevalence of defects of the bone over the superior semicircular canal. We examined a large series of temporal bones sectioned in the plane of the superior semicircular canal and determined overall prevalence for dehiscence of the bony canal to be 0.5% of specimens and 0.7% of individuals. We believe this estimate to be conservative, because we removed from consideration approximately 3% of the specimens that had defects in bone and dura overlying the canal. We did this to exclude cases in which bone was missing only because it was avulsed in processing, reasoning that avul-
sion of bone would necessarily result in a defect in the dura as well. However, the same appearance would have resulted if the technician had stripped dura from an already dehiscent specimen. We observed many intact bones from which the dura had been stripped. We suspect that some bones that were actually dehiscent were excluded as damaged and that our estimate of the prevalence of superior canal dehiscence in the middle fossa is low.

An additional 1.4% of specimens (1.3% of individuals) had bone no thicker than 0.1 mm over the lumen of the canal. We chose 0.1 mm as a criterion measure for abnormally thin bone for the following reasons. First, in reviewing the ultra–high-resolution CT scans from our patients with the superior canal dehiscence syndrome, 0.1 mm was the lower limit of thickness that we could measure reliably. Bone thinner than this might appear dehiscent on such scans, and we were interested in determining not only the true dehiscence rate, but also the expected rate of apparent dehiscences on such CTs. Second, 0.1 mm is well below the 99% confidence interval for the thickness of bone over the superior canal in our 100 control temporal bones. Finally, 0.1 mm is even at the lower end of the 90% confidence interval for thickness over the superior canal in our infant specimens. By any of these measures, then, 0.1 mm is abnormally thin.

Our estimate for the prevalence of bone no thicker than 0.1 mm over the superior canal is also probably conservative. Such thin bone over the canal at the middle fossa is rare. The vast majority of our specimens had bone that was abnormally thick for the lumen of the canal.
Bone Thickness, mm

<table>
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<th>Subject No./ Age</th>
<th>Sex</th>
<th>Middle Fossa</th>
<th>Superior Petrosal Sinus</th>
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<th>Systemic Diseases</th>
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*For subjects 1 through 4, bone overlying one or both superior canals was dehiscent; for subjects 5 through 12, it was thin (≤0.1 mm). Dehiscences are marked in bold and by boxes; thin values are in bold. Bone thickness was the minimum distance between the lumen of the bony superior canal and the upper surface of the temporal bone (middle fossa) or trough of the superior petrosal sinus. Ellipses indicate none identified; CRI, chronic renal insufficiency; CA, carcinoma; DM, diabetes mellitus; CVA, cerebrovascular accident; TB, tuberculosis; COM, chronic otitis media; and SNHL, sensorineural hearing loss.

The findings of this study also have important implications for diagnosis of the superior canal dehiscence syndrome. Current CT technology does not permit differentiation of bone thinner than 0.1 mm from dehiscence. Identification of this syndrome must, therefore, be based on the presence of the associated symptoms, signs, and findings on diagnostic studies in addition to findings on CT imaging. The symptoms include sound- and/or pressure-induced vertigo and oscillopsia, hyperacusis, and dysequilibrium. Abnormally low thresholds for sound-induced vestibular evoked myogenic potentials have also been reported in patients with superior canal dehiscence syndrome.

**ETIOLOGY AND PATHOLOGIC FEATURES**

We did not find any local bone abnormalities to explain erosion in these regions, with the exception of subject 2, in whom a left vestibular schwannoma may have contributed to loss of bone at the superior petrosal sinus (Figure 5, E and F). Even in this subject, however, the deep superior petrosal sinus appeared to be a preexisting feature, and many parts of the sinus groove were free of tumor. Moreover, the contralateral side, although free of tumor, also showed a dehiscence at the superior petrosal sinus (Figure 5, A). Lamellae in the bone near the dehiscence on the contralateral side conform to the sinus (Figure 5, C), suggesting that the bone had been stable for many years.

Elevated intracranial pressure may have been an etiologic factor in bilateral thinning of the skull base. However, we have no history of increased intracranial pressure in any of the other patients with thinning or dehiscence. Our clinical series corroborates this finding. Cranial imaging in patients with the superior canal dehiscence syndrome has not revealed ventricular enlargement or other sign of elevated intracranial pressure.

Syphilis can affect the otic capsule, in particular the semicircular canals. Lesions range from a proliferative peri-
ostitis and fibrosis of the canals to bony obliteration of the canals. Gummas may or may not be present. The medical records indicated syphilitic aortitis in 4 of the patients with superior canal thinning or dehiscence. We did not find any syphilitic lesions in the temporal bones of these or any of the other subjects in the Table. The high prevalence of syphilis in this group may simply reflect the high prevalence in the preantibiotic era, from which many of the subjects came.

Likewise, arachnoid granulations are known to cause tegmen erosion, encephaloceles, and cerebrospinal fluid leaks. However, we did not find any arachnoid granulations eroding bone over the superior canal. Meninges frequently were removed from the specimens before processing, so it is possible that we could have missed small arachnoid granulations confined to the upper surface of the temporal bone. In particular, arachnoid granulations have a predilection for the dural sinuses. The appearance of a deep erosion of bone at the superior petrosal sinus suggests that an arachnoid granulation may have been responsible. We argue against this for 3 reasons. First, in a number of specimens, the superior petrosal sinus was preserved intact in a deep trough, and no arachnoid granulations were seen in these troughs (Figure 1, D). Second, we were able to trace an elongated groove for the superior petrosal sinus along the length of the temporal bone in each case of thinning or dehiscence related to the sinus (Figure 4). An arachnoid granulation would not likely carve out such a smooth, elongated trough, but would instead create an irregular, focal erosion. We did not observe any such pattern over the superior canal. Third, erosions due to arachnoid granulations show histological evidence of recent bone erosion with disruption of the lamellae normally seen parallel to the surface of the bone.

Figure 6. Bone over the superior canal (sc) in infant temporal bones was consistently thin at the floor of the middle fossa (mf). A, Premature infant of 24 weeks’ estimated gestational age. B, Full-term infant, aged 2 months. C, Full-term infant, aged 4 months. D, Full-term infant, aged 10 months. A, At 24 weeks’ gestation, the labyrinth is formed and covered by the thin inner layer (endosteal) bone (i). Middle layer bone (m) is forming by the fusion of osseous globules. The outer layer bone (o) covers the temporal bone, except at the projection of the superior canal into the middle fossa. Here, only the thin inner layer covers the canal (arrowhead; 0.02 mm). B, At 2 months of age, the bony walls of the semicircular canals are thickening as middle layer bone is added (white arrowheads). However, the thin inner layer bone is all that covers the superior canal in the middle fossa (black arrowhead; 0.03 mm). C, At 4 months of age, the otic capsule continues to thicken (white arrowheads), but only minimally over the superior canal (black arrowhead; 0.10 mm). D, At 10 months of age, otic capsule bone is thick around all of the canals (white arrowheads), even over a portion of the superior canal in the middle fossa. Nevertheless, a relatively thin area remains over the canal near the posterior fossa (black arrowhead; 0.11 mm). The fibrous tract of the subarcuate fossa (saf) persists in each of these infant specimens. Each calibration bar indicates 1 mm. Hc indicates horizontal canal; me, middle ear; and pc, posterior canal.
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it can become pneumatized. The middle layer is unique
ference interval at 32.4 months. The line fit through the data from infants and
children is represented by the following equation: Thickness = (0.022
mm/month) \times \text{age} + 0.087 \text{ mm} (R = 0.85). It intersects the adult 99%
confidence interval at 22.4 months.

We did not see such disruption in the superior petrosal
sinus groove. Rather, we found lamellae of ossification par-
allel to the contour of the sinus (Figure 5, C). If erosion
occurred here, it must have stopped years previously, as
woven bone has been replaced by this mature lamellar bone.
Alternatively, we suggest that the sinus did not erode bone,
but that bone was deposited around the prominent sinus
during development.

The bilateral nature of thinning and the presence of
mature lamellar bone on the margins of thin areas point
toward a developmental cause. Our findings from the in-
fant temporal bones support this conclusion and ex-
pand on the description of development of the bony laby-
rinth given by Anson and Donaldson.17 They described
the development of the otic capsule bone from a carti-
lage model. Beginning in the 15th week of gestation, 14
ossification centers grow and fuse, so that, by the 21st
week, the membranous labyrinth is completely encased
in bone. Each center has a trilaminar structure, and, after
their fusion, does the entire otic capsule. The inner
periosteal (or endosteal) layer, which contacts the perilymphatic space, grows minimally after fusion. An
outer periosteal layer grows well into postnatal life, when
it can become pneumatized. The middle layer is unique
in the skeleton, because it forms by the invasion of os-
teoblasts into cartilage lacunae, forming osseous glob-
ules. Accretion of bone around these globules gradually
replaces marrow space, giving the bone its petrous char-
acter in approximately 5 postnatal months.

Figure 6 emphasizes these findings, but with an im-
portant difference. The thin, inner periosteal layer of bone
covering the superior canal at its projection into the middle
fossa is not fully covered by the growth of the other lay-
ers until much later. Even the specimen at 10 months
shows a persistent monolayer of bone at this site (Fig-
ure 6, D). The pattern of thinning or dehiscence at the
middle fossa seen in 9 adult specimens is strikingly simi-
lar to the appearance of these infant temporal bones (Fig-
ures 1, D, and 3). Findings in 27 pairs of temporal bones
from infants and children no older than 4 years confirm
that bone over the superior canal in the middle fossa re-
mains thin for the first 2 to 3 years of life (Figure 7).

We suggest that postnatal failure to develop outer
and/or middle layer bone over the superior canal may be
the cause of adult superior canal thinning and dehis-
cence in the middle fossa. The alternative explanation,
that bone developed normally, then was eroded sym-
metrically in subsequent years, seems less plausible. More-
over, the ossification pattern appeared to be stable in our
thin specimens, as evidenced by the lamellar structure
of the bone at the edges. Whatever process caused thin-
ning must have remained stable for many years.

PATHOPHYSIOLOGIC FEATURES

A dehiscence in the setting of thin superior canal bone
may result from a traumatic event such as closed head
injury or a sudden change in intracranial pressure. Such
a history was elicited in approximately half of the pa-
tients in whom superior canal dehiscence syndrome was
diagnosed at our institution.13 We hypothesize that such a
second event must occur to disrupt the bone and ex-
pose the membranous labyrinth to fluctuations in intra-
cranial pressure before symptoms ensue. Clinical histo-
ries from subjects in our temporal bone archive with thin
but intact bone also suggest that labyrinthine symptoms
or signs do not appear to result just from thinning of the
bone overlying the superior canal. No vestibular symp-
toms were noted in 7 of the 8 patients with thinning of the
canal at the middle fossa. Caloric nystagmus was still
elicited in each of 3 of these subjects who underwent ca-
loric testing.

Only 1 subject in the temporal bone archive had a
middle fossa dehiscence. Although she had no history of
episodic vertigo, she had no ice-water caloric responses,
indicating bilateral labyrinthine hypofunction. This does
not appear to be the usual natural history of dehiscence,
however, as none of the patients seen with the syndrome
at our institution have had abnormal results of caloric
tests.1 Bilateral labyrinthine hypofunction in the tempo-
ral bone archive subject may not have been related to the
status of the bone overlying the superior canal. However,
the hypofunction could explain why she had no episodic
vertigo.

The other 4 dehiscences occurred at the superior pe-
rosal sinus, a site for which we have not observed symp-
toms of the superior canal dehiscence syndrome. How-
ever, these bony dehiscences should be adequate conduits
for transmitting intracranial pressure to the superior ca-
nal. Perhaps, then, pressure alone is not the relevant stimu-
lus. Dura bulging into or out of the canal and displacing
endolymph may be the relevant stimulus in patients with
symptomatic dehiscences. Superior petrosal sinus dehis-
cences are of limited length because the sinus crosses the
canal at nearly a right angle (Figures 4 and 5). The asym-
omatic dehiscence in the middle fossa depicted in Fig-
ure 3 is about the same length. Perhaps dehiscences must

Figure 7. Scatterplot of bone thickness over the superior semicircular canal
for specimens from 26 infants and children no older than 4 years. Data are
measurements taken at the thinnest bone over the superior canal in the
middle fossa. For comparison, average adult bone thickness at this site (0.96
mm) is depicted by the dashed line, and the shaded area indicates 99%
confidence interval for adults. The line fit through the data from infants and
children is represented by the following equation: Thickness = (0.022
mm/month) \times \text{age} + 0.087 \text{ mm} (R = 0.85). It intersects the adult 99%
confidence interval at 22.4 months.
be of a critical length or area before enough dura can bulge into the canal to cause symptomatic endolymph displacement. We are investigating the possible relationship between the dimensions of superior canal dehiscences measured from high-resolution CT scans and specific signs and symptoms in our patients with the syndrome.

**CONCLUSIONS**

The findings of this temporal bone survey suggest that approximately 2% of individuals already have a dehiscence or extremely thin bone overlying the superior semicircular canal at the middle fossa or superior petrosal sinus. The bilateral nature of this abnormality and its similarity to the appearance of the infant temporal bone suggest that this is a developmental anomaly. Our clinical experience has been that symptoms and signs of superior canal dehiscence first develop in adulthood. We suggest, therefore, that a second event may occur, which fractures the thin bone or destabilizes dura over a preexistent dehiscence. This second event may be a closed head injury or sudden change in intracranial pressure.

Accepted for publication August 18, 1999.

This study was supported by grants R01 DC02390 and P60 DC00979 from the National Institute of Deafness and Other Communication Disorders, Bethesda, Md.

Presented as a poster at the 22nd Midwinter Meeting of the Association for Research in Otalaryngology, St Petersburg, Fla, February 13-18, 1999, and at the Ninth Annual Meeting of the Society for the Neural Control of Movement, satellite session on Vestibular Influences on Spatial Orientation, Princeville, Hawaii, April 16-19, 1999.

We thank Edward F. McCarthy, Jr, MD, for reviewing the histological material with us to rule out bone abnormalities, and the Johns Hopkins Pathology Photography and Graphics Department for photomicroscopy. We are indebted to the late Samuel J. Crowe, MD, and John E. Bordley, MD, for establishing, building, and maintaining the Johns Hopkins Temporal Bone Collection.