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High-Resolution CT of the Temporal Bone in Dysplasia of the Auricle and External Auditory Canal

T. E. Mayer, H. Brueckmann, R. Siegert, A. Witt, and H. Weerda

PURPOSE: To determine CT findings in the external, middle, and inner ear of patients with microtia and external auditory canal dysplasia. **METHODS:** We used high-resolution CT, with multiplanar or axial 1-mm continuous sections, coronal or sagittal reformations, or low-dose spiral acquisitions, to examine 184 temporal bones of children with microtia. **RESULTS:** In cases of minor microtia, auditory canal stenosis was the most common associated abnormality; in those with major microtia, atresia was predominant. Middle ear malformations depended on the severity of the auricular anomalies. Inner ear changes could also be noted. Ossicle dysplasias occurred in 98% of patients (stapes, 72%), absence of the oval window in 36%, labyrinthine malformations in 13%, closed round window in 6%, facial canal displacement in up to 75%, and aberrations of the vascular canal in 38% of patients with third-grade auricular deformity. **CONCLUSION:** A variety of external, middle, and, less frequently, inner ear changes were detected in connection with microtia.

Index terms: Ear, abnormalities and anomalies; Temporal bone, computed tomography

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Congenital aural dysplasias occur in one of 3300 to 10 000 births (1, 2), except in the era of thalidomide embryopathy (1959–1962), when it was diagnosed in one of 900 neonates born in Germany (3). Microtia has been described even on Babylonian tablets (4) and has been seen in prehistoric skulls (5). The classification of auricular deformities was proposed by Marx (6) and described by Rogers (7). Microtia is often associated with abnormalities of other organs as the result of genetic disorders, chromosomal defects, intrauterine infections, or environmental teratogens.

Surgery for congenital aural atresia is one of the most difficult ear operations, as it requires reconstruction of the auricle, often done by using grafts of skin and rib cartilage (8, 9), and, if necessary, restoration of hearing by drilling a

new auditory canal and reconstructing the ossicular chain (10–12). Surgery is also performed for congenital cholesteatoma, labyrinthine fistula, infection, or facial nerve palsy from previous surgery (13). Even if there is labyrinthine involvement by the malformation, inner ear function is preserved in most cases. Endoscopic viewing is not possible in stenotic or atretic auditory canals. Staging by computed tomography (CT) is necessary to avoid the risk of facial nerve lesions, worsening of hearing, and bleeding, and to obtain a valid prognosis of success.

We present a review of the CT findings in 92 patients with microtia and external auditory canal dysplasia.

Subjects and Methods

All 92 patients had high-resolution CT, 89 of them with 1-mm-thick sections and three with 2-mm-thick sections. Fifty-five patients only had axial CT, sometimes with reformations of the coronal, oblique, or sagittal planes; 37 patients were scanned in axial and coronal planes, and five of these were additionally scanned in the sagittal orientation with low-dose (85 mAs) spiral CT. In these cases,

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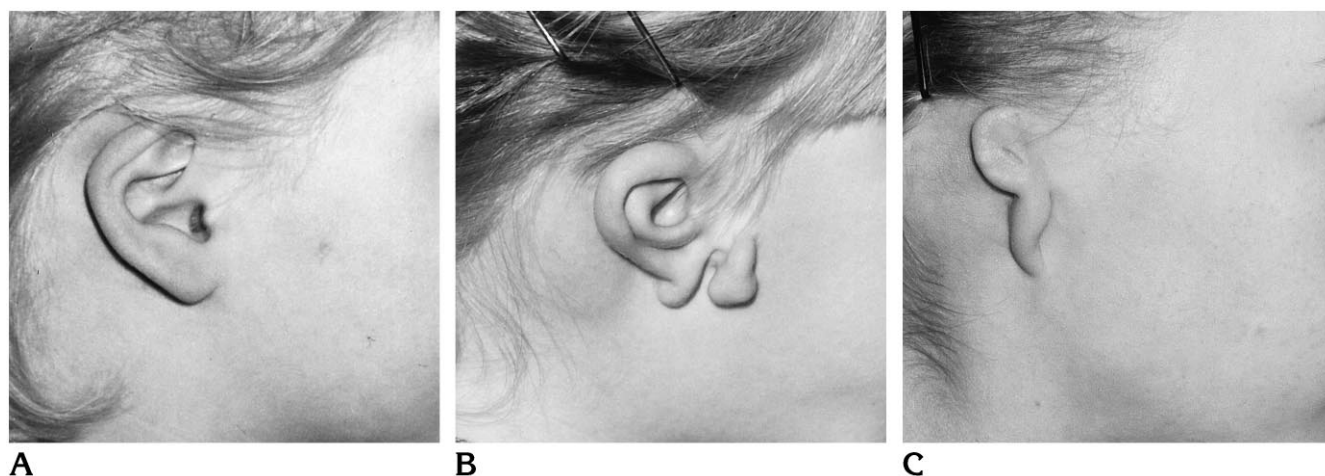


Fig 1. Pathologic anatomy of the auricle, classification after Weerda (14).

A, First-degree auricular dysplasia: small auricle with hypoplasia of the upper helix.

B, Second-degree auricular dysplasia: malformed ear, with preserved helical configuration (snail shell ear).

C, Third-degree auricular dysplasia: auricular remnant with no recognizable structures of a normal ear.

three-dimensional surface reconstructions and sagittal reformations were also made, if necessary. Spiral CT allowed a reduction of motion artifacts in examinations of children. Four patients had magnetic resonance (MR) imaging.

The following structures were evaluated in 184 temporal bones: 1) The external auditory canal: stenosis or atresia of the cartilaginous part of the auditory canal, stenosis or atresia of the bony part, thickness of the atresia plate (smallest dimension at the level of the hypotympanum). 2) Dysplasia or defects of the temporal bone, zygomatic bone, and mandibular condyle; if possible, the whole skull and the cervical spine (scout view) were analyzed for anomalies. 3) Vascular structures such as the carotid canal, sigmoid sinus, and jugular bulb were looked at for dehiscence, aberrant course, or remnant embryological vessels. 4) The auditory tube, including tensor tympani muscle and grade of pneumatization. 5) Congenital or secondary cholesteatoma or epidermoid. 6) The extent of the middle ear cavity, form of tegmen. 7) Diminution, dysplasia, rotation, fusion, ectopia, tympanic wall adherence, and absence of the ossicles. 8) Labyrinthine windows open or closed; fistula. 9) Cochlear turns, vestibule, semicircular canals, aqueducts. 10) The internal auditory canal and facial nerve canal: aberration, dehiscence, hypoplasia, thickening, splitting.

Classification criteria of auricular deformities used in this study were those described by Weerda (14), as follows. First-degree dysplasia: macrotia, prominent ear, pocket ear, absence of the upper helix (Fig 1A), absence of the tragus, clefts, lobular deformities, and cup ear deformities type 1 and 2. Second-degree dysplasia: cup ear deformity type 3 (Fig 1B) and mini-ear. Third-degree dysplasia: absence of normal auricular structure (Fig 1C) (unilateral or bilateral) and anotia; severely dysplastic ears might be displaced downward owing to incomplete ascension from the neck (15).

Results

Auricular Dysplasia

One hundred thirty-four of 184 ears examined were microtic. Consequently, 23% of the patients had bilateral microtia. Auricular deformities were often visible on CT scans. Some patients had undergone several reconstructive operations on the external ear.

CT findings were compared with the clinical appearance of the auricle. The results are presented as percentages of two groups: three quarters of the microtic ears were classified as third-degree dysplasia (including one case of anotia) and are referred to as major microtia (84 ears); one quarter of the dysplastic auricles had first-degree (16 ears) or second-degree (13 ears) dysplasia. The two groups are combined and referred to as minor microtia in the following sections (see Table).

External Auditory Canal

Microtia and external auditory canal dysplasia were highly correlated ($P < 10^{-30}$). Three quarters of our patients with major microtia had an atresia; none had a normal external auditory canal. In three quarters of the minor microtic patients, the bony or cartilaginous part of the external auditory canal was stenotic ($P < 10^{-7}$). The patients with a normal external ear (contralateral to a microtic ear) rarely had external auditory canal stenosis.

Petrus bone malformations correlated to the degree of auricular dysplasia (relative rate in each group of microtia)

Auricular Dysplasia, n = 184	Normal (Contralateral) Ear, n = 71	Minor Microtia (First and Second Grade), n = 16 + 13	Major Microtia (Third Grade), n = 84
External auditory canal			
Stenosis	5.6	72.4	21.4
Atresia	0	10.3	78.6
Malformations of:			
Temporal bone	9.9	55.2	89.3
Mandible*	1.4	20.7	64.3
Zygomatic arch†	0	13.8	44.0
Others	14.1	17.2	29.8
Carotid canal			
Slight aberrations	1.4	17.2	15.5
Hypoplasia	1.4	3.4	0
Sigmoid sinus, jugular bulb aberration, highriding	1.4	17.2	38.1
Eustachian tube, bony dysplasia	0	20.7	44.0
Pneumatization			
Reduced‡	7.0	10.3	19.0
Little or none‡	0	17.2	47.6
Middle ear space epitympanum			
Reduced§	1.4	24.1	20.2
Minimal§	0	3.4	41.7
Mesotympanum			
Reduced§	0	37.9	46.4
Minimal§	0	6.9	41.7
Hypotympanum			
Reduced§	1.4	24.1	17.9
Minimal§	0	13.8	73.8
Malleoincudal joint			
Dysplastic, adherent	7.0	51.7	63.1
Absent	0	3.4	34.5
Incudostapedial joint, dysplastic/absent	4.2	68.9	89.3
Stapes			
Dyplastic	1.4	17.2	15.5
Absent	0	34.5	56.0
Oval window, occluded	0	41.4	35.7
Fallopian canal abnormalities			
Hypoplasia of the labyrinthine segment	0	10.3	9.5
Aberration of the labyrinthine segment	0	3.4	4.8
Tympanic dehiscence	0	24.1	26.2
Severe anomalies of the tympanic segment	0	0	2.4
Mastoid segment anterior dislocation	0	55.2	61.9
Severe aberration of the mastoid segment	0	3.4	11.9
Internal auditory canal, inclined/widened/agenesis	5.6	13.8	16.7
Round window, occluded	0	3.4	6.0
Inner ear dysplasia			
Cochlea	0	0	3.6
Vestibulum	0	3.4	4.8
Lateral semicircular canal	0	6.9	13.1

Note.—Values given in percentages.

* n = 137.

† n = 122.

‡ Reduced indicates 75% less than contralateral side or only periantral cells bilaterally; little or none, complete loss of mastoid pneumatization; tympanum may or may not be air containing.

§ Reduced indicates 33% less; minimal, 67% less.

Fig 2. Axial CT scans of external auditory canal abnormalities.

A, Stenosis (arrow) is seen on the right.

B, Atresia and formation of an atretic plate (arrowhead) are on the right; a normal external auditory canal is on the left (arrow).

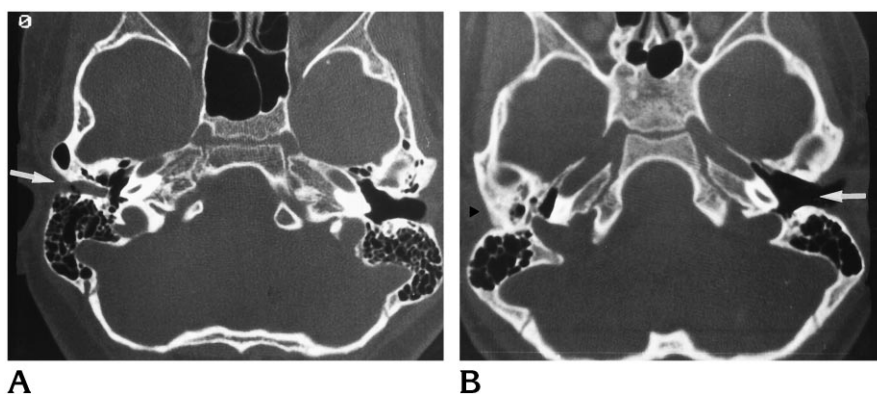


Fig 3. Cholesteatoma and dermoid.

A, Axial CT scan shows masses of solid tissue in the middle ear cavity and mastoid, with bony erosion (arrowhead) in external auditory canal stenosis.

B, Coronal T1-weighted (360/15/4 [repetition time/echo time/excitations]) spin-echo MR image shows normal external auditory canal on left (arrow) and external auditory canal missing and an epidermoid in the cerebellopontine angle on right (arrowheads).

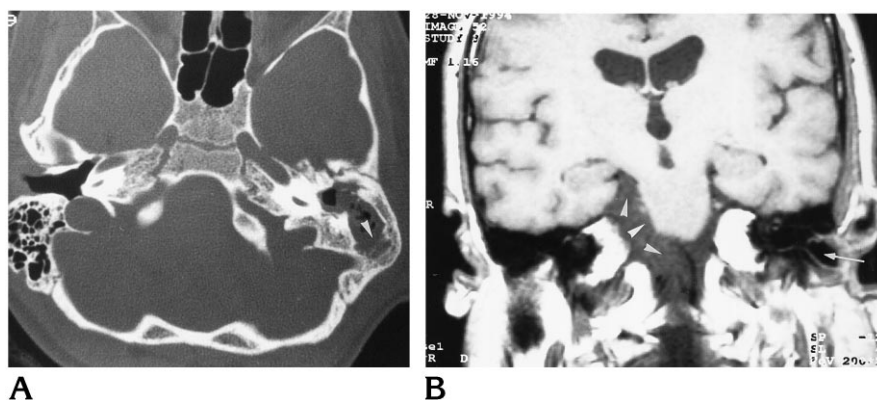
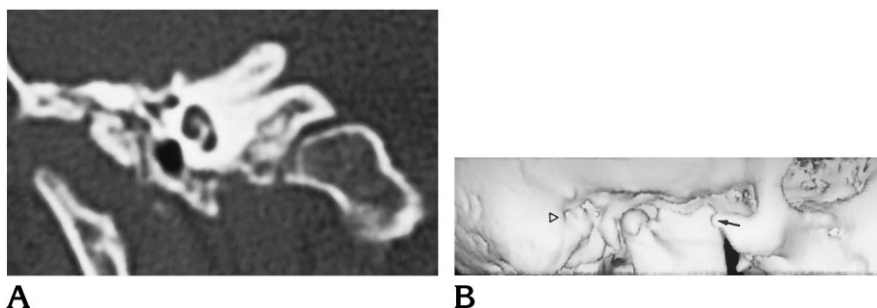


Fig 4. Associated anomalies.

A, Coronal CT scan shows dysplasia of the mandibular condyle and defect of the joint socket with a nearly obliterated middle ear in Franceschetti syndrome.

B, Three-dimensional CT surface reconstruction viewed from the right side shows interrupted zygomatic arch (arrow); no external auditory canal is present (arrowhead).



Stenotic auditory canals often showed a distinctly more angulated course than normal. The canals sloped from caudolateral to craniomedial. External auditory canal atresia varied from a fibrous closure to thick atretic plates (Fig 2) up to 31 mm in diameter (median, 14 mm).

Among patients with external auditory canal stenosis and atresia, 24% had a tympanic mass of solid tissue or fat density. Some patients had bone erosion, suggesting an epidermoid cyst or a congenital or secondary cholesteatoma (Fig 3A). One patient had fistulas to the skin of the side of the face combined with mandibular dysplasia. In another patient, an epidermoid of the cerebellopontine angle had to be removed surgically (Fig 3B).

Other Skull Abnormalities

Microtia and external auditory canal atresia were most often associated with aplasia or hypoplasia of the tympanic part or mastoid process of the temporal bone ($P < 10^{-15}$), which correlated with the degree of auricular dysplasia ($P < .001$). The second most frequent abnormality was dysplasia of the mandibular condyle ($P < 10^{-9}$), followed by defects of the zygomatic arch ($P < 10^{-4}$) (Fig 4). In genetic disorders, such as Franceschetti (Treacher Collins) syndrome or Crouzon disease, the whole skull was abnormal; and in Klippel-Feil syndrome, the cervical spine was dysplastic.

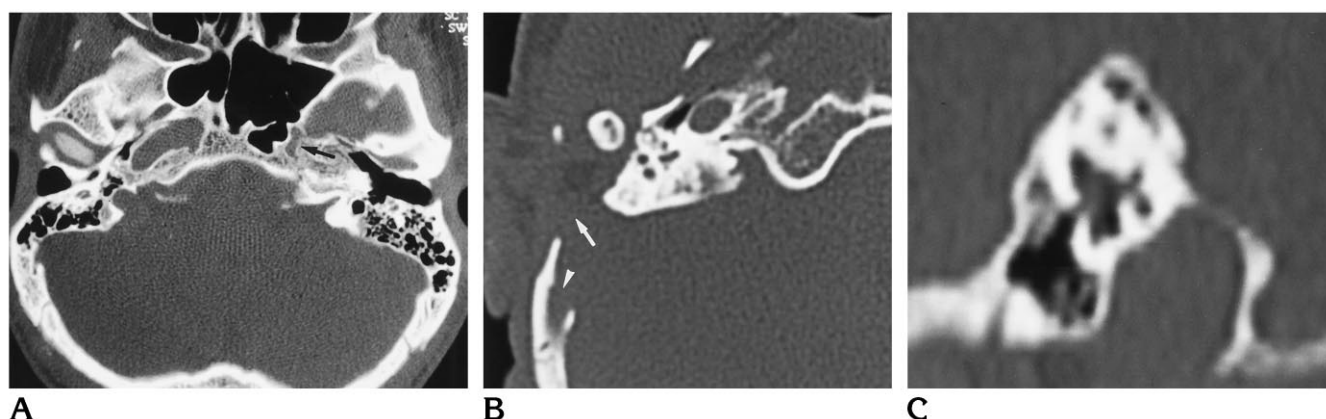


Fig 5. Vessel canal aberrations.

A, Axial CT scan at the level of the foramen lacerum and basal turn of the cochlea shows hypoplastic carotid canal (arrow) in first-degree auricular dysplasia.

B, Axial CT scan shows dehiscent sigmoid sinus (arrow) and an emissary (arrowhead) in a severely microtic ear with hypoplastic temporal bone.

C, Sagittal CT scan in the plane of the round window shows high-riding jugular bulb.

Temporal Bone Vessels

The carotid canal was asymmetric in one fifth of the patients with unilateral aural atresia, slightly rotated or elevated in relation to the hypoplastic base of the skull ($P < .05$). In two patients, the internal carotid artery was severely hypoplastic or aplastic, but these severe anomalies did not correspond with the degree of microtia (both had first-degree auricular dysplasia, and one was affected on the contralateral side). We did not find dehiscence into the middle ear or a remnant stapedia artery. The sigmoid sinus and jugular vein showed normal asymmetries and emissaries and often a high-riding jugular bulb. In cases of severe hypoplasia of the tympanic part of the temporal bone, the sigmoid was positioned somewhat ventrally ($P < 10^{-4}$). Only three patients had an erosion of the squamous part of the temporal bone by venous structures, but none showed dehiscence into the middle ear (Fig 5).

Pneumatization of the Temporal Bone

The air content of the middle ear and mastoid was partially or completely reduced in one quarter of the minor and in two thirds of the major microtic ears; only a few normal contralateral ears had reduced mastoid pneumatization ($P < 10^{-7}$). The severity and frequency of decreased pneumatization correlated with the degree of anomalies of the external ear ($P < .05$).

The bony eustachian tube had either a dys-



Fig 6. Axial CT scan shows eustachian tube (arrow) and complete lack of mastoid pneumatization with atresia of the external auditory canal (arrowhead).

plastic appearance (narrowing) or the tensor tympani muscle was hypoplastic ($P < 10^{-6}$) in about 20% of minor and 40% of major microtic ears. In two patients, the tube was excessively enlarged (Fig 6). The cartilaginous part of the tube was visible only in cases of enlargement.

The Tympanic Space

The width of the middle ear was frequently diminished ($P < 10^{-19}$), correlating with the degree of microtia ($P < 10^{-7}$). The epitympanum was affected least often, but could be totally obliterated. In the mesotympanum, at least a small residual cleft was usually left in other-



Fig 7. Axial CT scans of tympanic space reduction.

A, Obliterated epitympanum and loss of ossicles are on the right.

B, Rudimentary slitlike mesotympanic space is at the level of the round window on the left; note deep sinus tympani.

C, Remnant hypotympanum and bony eustachian tube (arrow).

wise nearly obliterated middle ears. The hypotympanum was reduced most often (Fig 7).

The Ossicular Chain

Changes of the ossicles were frequently present ($P < 10^{-18}$) and consisted of a dysplastic shape, diminution, thickening (rarely), axis rotation, arthrodesis, bony or ligamentous adherence to the attic wall, ectopia, or complete absence (Figs 8 and 9A).

We subdivided the complex dysplasias according to embryological origin:

Malleus-incus-complex.—Few of the normal contralateral ears showed dysplastic changes of the malleus or incus. In minor microtia, half the patients had dysplastic ossicles; rarely, the ossicles were absent. In severe microtia, only 2% of the ears were normal, almost two thirds were dysplastic, and one third were absent ($P < 10^{-7}$ for the dependency of the degree of microtia).

Incus-stapes joint.—The incus-stapes complex was often also dysplastic if the malleoincudal articulation was affected. In cases of an absent stapes, the remnant incus was always malformed and the joint counted as dysplastic or absent ($P < .05$ for the dependency of the degree of microtia).

Stapes.—The stapes were not involved as frequently as the rest of the ossicular chain. Even in severe microtia, about 30% of the stapes remained normal. In minor microtia, the results were similar, so the stapes anomalies had no significant correlation with the degree of auric-

ular anomalies. The most often observed abnormality was a missing stapes. Dysplasia was difficult to see and was identified less often.

Oval Window

The oval window was always open in normal ears. It was absent in more than one third of minor and major microtic ears ($P < 10^{-4}$), with no significant correlation with the degree of external ear deformity (Fig 9).

Facial Nerve Canal

The course of the fallopian canal was often abnormal in patients with microtia ($P < 10^{-13}$).

Anomalies of the labyrinthine segment occurred rarely—three slight aberrations were located caudally, two medially—four of them in patients with severe microtia. In 11 patients, the labyrinthine segment was hypoplastic, eight of these had major microtia.

The tympanic segment was affected more frequently. Typically, it was displaced caudally, extending as inferiorly as the round window and was often dehiscent (Fig 10A and B). Dehiscence of the tympanic part could be difficult to identify if the tympanum was filled with soft tissue. In one case, the course of the facial nerve was shown intraoperatively between the crura of the stapes (combined with closure of the oval and round window). In two patients, the facial nerve canal had an extremely anterior course, straight down from the geniculate ganglion.

The mastoid segment was ventrally displaced

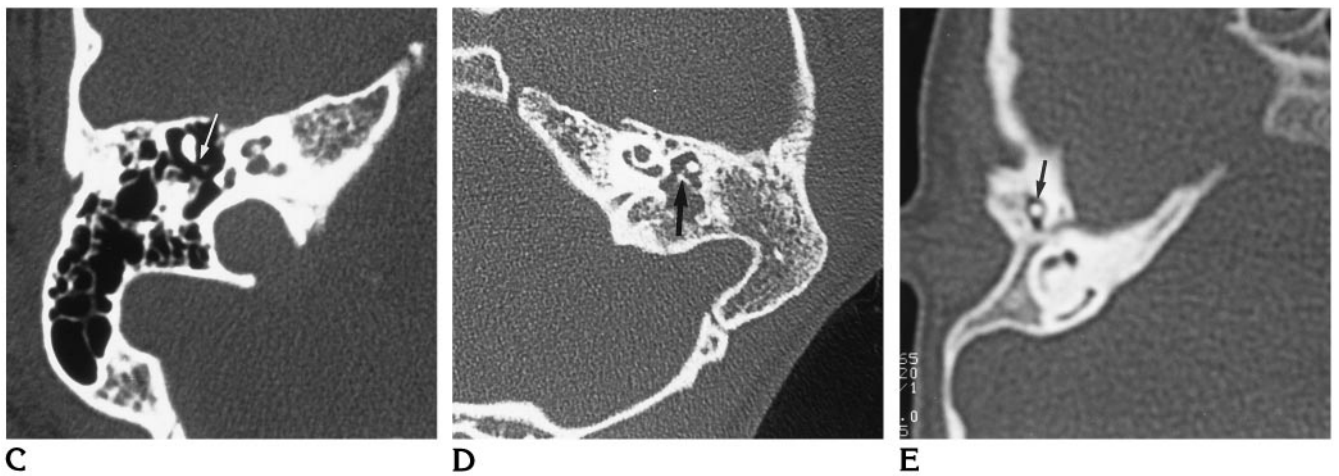
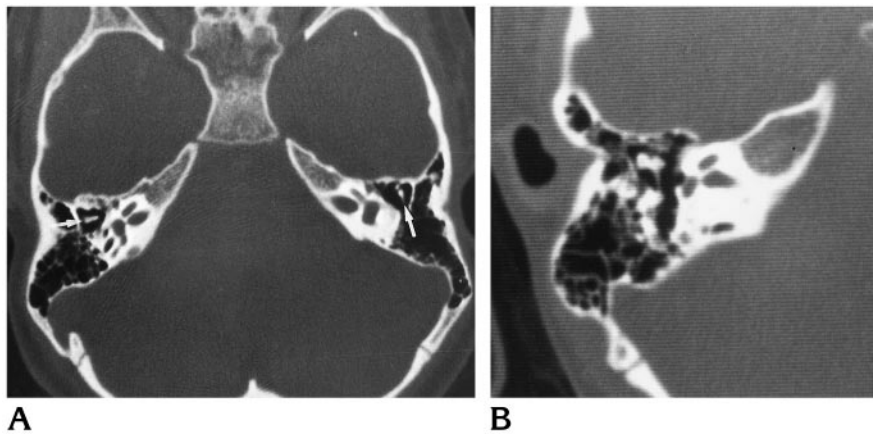


Fig 8. Axial CT scans of ossicle dysplasia.

A, Axis rotation and arthrodesis of the incudomalleal joint are on the right; the left side is normal (*arrow*).

B, Fusion and adherence of the incudomalleal joint.

C, Dysplasia of the incudostapedial joint (*arrow*).

D, Rudimentary ossicles (*arrow*) in an opacified middle ear.

E, Ectopic (anterior) ossicle (*arrow*) in an obliterated temporal bone.

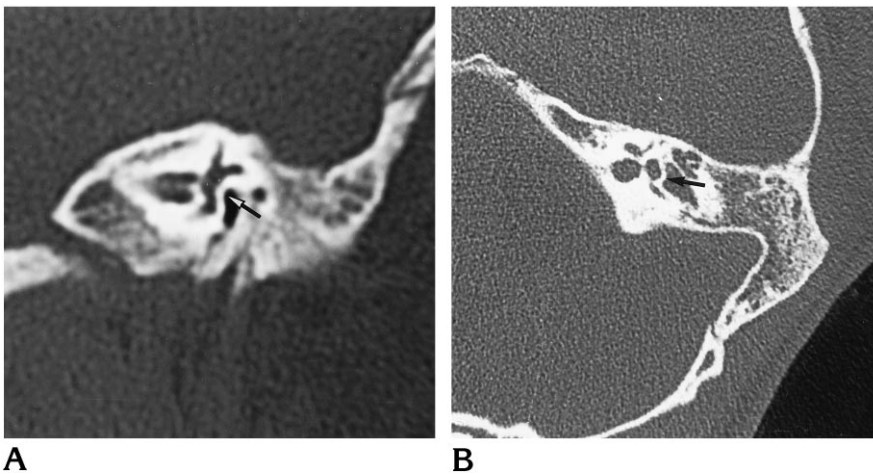


Fig 9. Oval window.

A, Coronal CT scan shows closed oval window (*arrow*) in a pneumatized middle ear, with loss of all ossicles and external auditory canal atresia.

B, Axial CT scan shows closed oval window (*arrow*) in an opacified temporal bone.

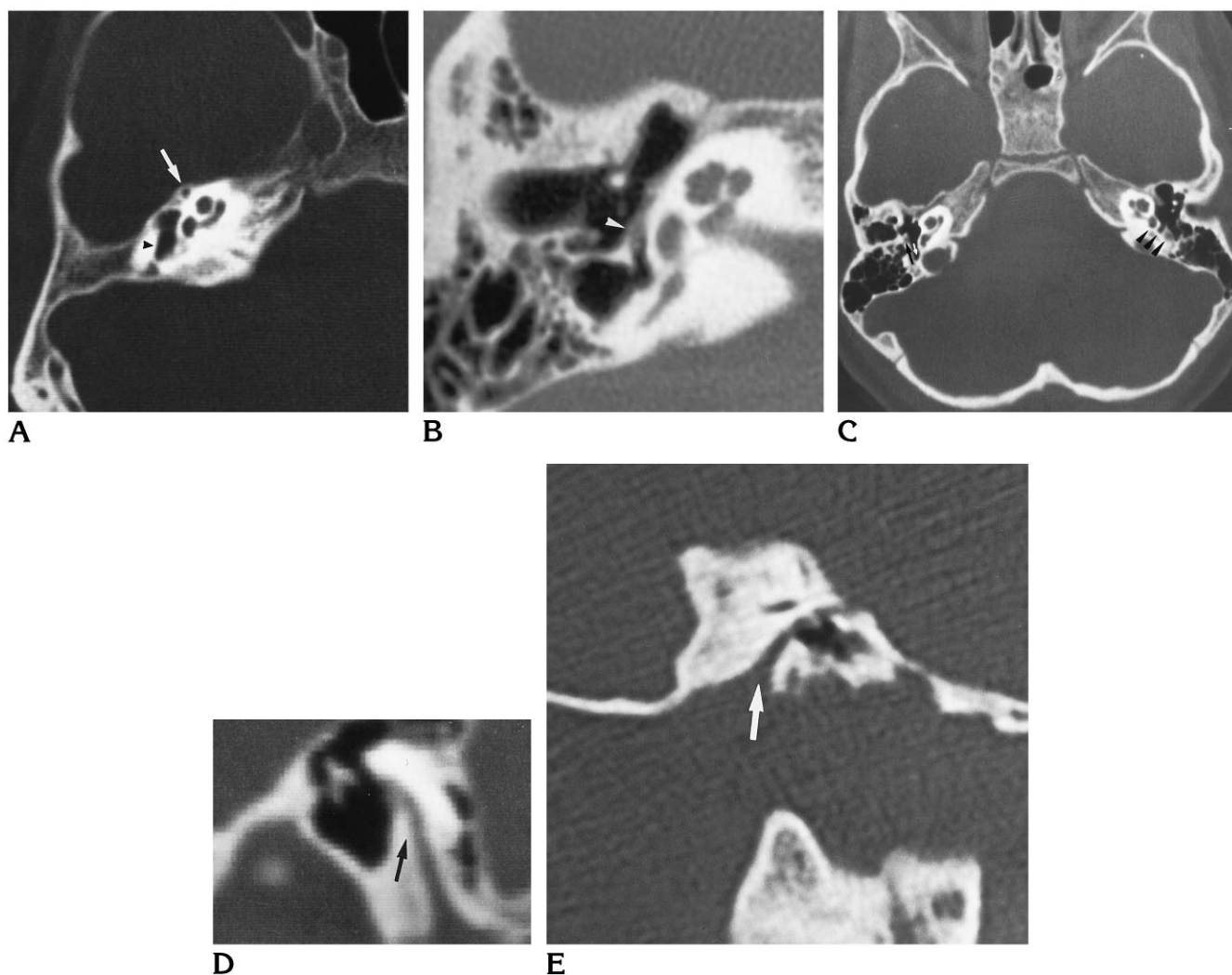


Fig 10. Axial CT scans of facial nerve canal aberration.

A, Caudal course of the second segment (*arrow*) and opacified narrowed tympanum (*arrowhead*).

B, Dehiscence in the caudally displaced tympanic segment (*arrowhead*).

C, External auditory canal atresia and ventral displacement of the mastoid segment (*black arrow*), deepened sinus tympany (*white arrow*), and closed round window (lacking "air bubble") on the right; normal round window, sinus tympani, and mastoid facial nerve (*arrowheads*) on the left, from ventral to dorsal.

D, Sagittal CT reformation of the normal mastoid facial nerve canal (*arrow*).

E, Sagittal CT scan of the flattened second genu and ventrally dislocated mastoid segment of the facial nerve canal (*arrow*).

in half the patients with minor microtia and in three quarters of those with major microtia. Mostly, the dorsal attic wall was displaced ventrally, reaching the level of the round window, leading to a deep tympanic sinus (Fig 10C and E). In eight of these cases, the facial nerve left the mastoid cranially to the stylomastoid foramen in a ventral direction to the temporomandibular joint; in three cases, the exit was in a lateral direction. All but one of these patients had major microtia. A posterior course of the seventh nerve was found in two microtic and two normal ears. In one case of first-degree

auricular dysplasia, the mastoid facial nerve coursed medial to the tympanic sinus. The CT appearance could suggest splitting of the facial nerve, but it was indistinguishable from vascular canals leading to the nerve. In one case, duplication was proved intraoperatively (combined with dehiscence, closure of the oval window, and loss of the stapes).

Internal Auditory Canal

The internal auditory canal was dysplastic in one sixth of all patients with microtia. Typical

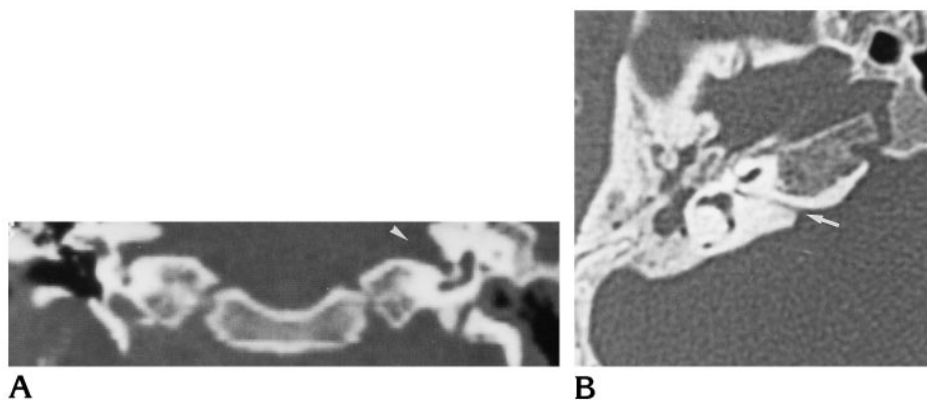


Fig 11. Abnormality of the internal auditory canal.

A, Coronal CT reformation of an abnormal craniomedial to caudolateral course and widening of the internal auditory canal (*arrowhead*) on the left; the right side shows a normally horizontal direction.

B, Axial CT scan of hypoplastic internal auditory canal (*arrow*).



Fig 12. Coronal CT scan shows fibrous closure of the round window (*arrow*) in a pneumatized middle ear.

was an inclined course from craniomedial to caudolateral and widening of the porus (Fig 11A). In one patient, instead of the internal auditory canal, only a thin canal of the seventh and eighth nerve was preserved (Fig 11B), combined with a hypoplastic labyrinthine segment of the facial nerve canal.

Round Window

The round window was absent in only 6% of the severely microtic ears (Fig 12). Closure of the round window was not significantly correlated with severe microtia or other dysplasias. It occurred also in two patients with well-pneumatized middle ear cavities (Fig 10C).

Labyrinth

The inner ear appeared slightly dysplastic in 13% of patients with microtia ($P < .02$); there was only one severe anomaly of this structure. In 13 cases, the lateral semicircular canal was hypoplastic (Fig 13A); in six cases, the vestibule was abnormal (Fig 13B). Minor labyrinthine anomalies were especially frequent in patients with Franceschetti syndrome. In one case of thalidomide embryopathy, only rudiments of the whole labyrinth, including the cochlea, were present bilaterally (Fig 13C). One case showed marked widening of the vestibular aqueduct (Fig 13D).

Discussion

To aid in understanding the complex anomalies of the external, middle, and inner ear, we present a short synopsis of its embryology.

Development of the inner ear begins in the third week of gestation with formation of the otic placode, an ectodermal thickening in the neighborhood of the myelencephalon. Invagination of the otic placode leads to an otic pit and to fusion of their external lips to the otic vesicle. In the eighth week, the otic capsule is formed, providing the stapes footplate and the ligament of the oval window. By the 12th week, the labyrinth is well differentiated. The facial nerve grows to reach its destinations between the fourth and fifth week.

The middle and external ear develops from the mesodermal first and second branchial arch and the endodermal first pharyngeal pouch between the fourth and 30th weeks. Developmental anomalies of the first pharyngeal pouch lead to disturbances of the eustachian tube and of the tympanic and mastoid pneumatization. Failure of differentiation of the first branchial arch leads to malformations of the incudomalleal joint, tensor tympani muscle, and mandible. Failure of differentiation of the second branchial arch affects the facial nerve canal, the stapedius muscle, the lower part of the ossicular chain, and the styloid process. Disorders of the first and second branchial arches also result in dysplasia of the auricular cartilage (leading to microtia in the seventh to eighth week, the earlier the more severe, or to anotia in the seventh week).

The external auditory canal arises from deepening of the first branchial groove in the ninth week. Opening of the bony part of the external auditory canal starts only in the 30th week, after complete differentiation of the inner, middle,

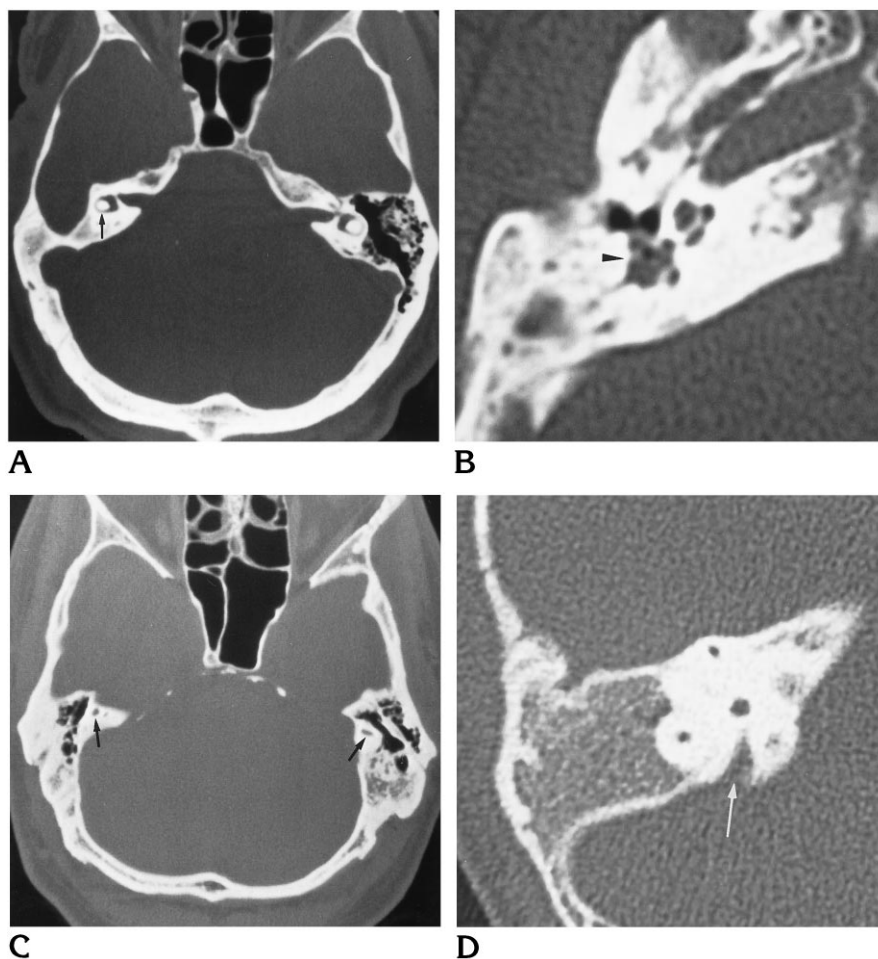
Fig 13. Axial CT scans of labyrinthine dysplasia.

A, Diminutive lateral semicircular canal (*arrow*) and hypoplasia of the tympanic part of the temporal bone on the microtic side (right); the left side is normal.

B, Dysplastic vestibulum (*arrowhead*) in Franceschetti syndrome.

C, Severe bilateral dysplasia of the labyrinth, including the cochlea (*arrows*) and the middle and external ear in thalidomide embryopathy.

D, Widened vestibular aqueduct (*arrow*), obliterated temporal bone, postoperative drainage in the auricle.



and outer ear. Failure of the epithelial cells of the first branchial groove to split causes stenosis or atresia of the external auditory canal, which might be isolated in an otherwise normal temporal bone (16–19). Anomalies of the internal carotid artery are thought to be caused by maldevelopment of the third branchial arch during the fourth week (20).

A variety of pathologic changes associated with microtia can be detected by high-resolution CT (17, 21–23). We found that the extent of the auricular anomalies corresponded to the severity of changes in the external auditory canal (ie, stenosis or atresia). Excessive inclination of the external and internal auditory canals may be explained by incomplete ascension of the auricle from the neck (15).

The degree of auricular anomaly was also highly correlated with dysplasias of the middle ear cavity, mastoid cells, malleus, and incus, presumably because of a common embryological origin. The connection between malforma-

tion of the auricle/external auditory canal and development of the middle ear as a whole was recently characterized (24) by means of Jahrsdoerfer's score (25). Just as frequent were aberrations of the facial nerve canal that developed from the second branchial arch. Ventral dislocation of the mastoid segment might be caused directly by hypoplasia of Reichert's cartilage and the ventrally migrated mastoid, as described by Curtin (23). Ventral placement of the dorsal tympanic wall leads to a deep tympanic sinus. The chorda tympani then courses lateral to the atresia plate (17). These changes corresponded to the severity of microtia and atresia. In some cases of anterior displacement of the mastoid segment and severe microtia, the facial nerve canal turned anteriorly or, less commonly, laterally, leaving the temporal bone to the temporomandibular articulation instead of continuing downward toward the stylomastoid foramen. This finding corresponds to the observations of Proctor (26). The more frequent

variation of the mastoid segment in humans with normal auricles is a posterior course (26, 27). We found this posterior "hump" less often, and not in connection with microtia. Caudal displacement and dehiscence of the tympanic segment were also frequent CT findings, corresponding to the cases reported by Swartz (28). In two of our patients, the seventh nerve on the microtic side went straight down from the geniculate ganglion ventral to the tympanum, in accordance with the description of a case of microtia by Fowler (27). Another interesting finding was a facial nerve medial to the tympanic sinus. A detailed classification of the facial nerve aberrations and splitting based on anatomic specimens and surgical findings has been provided by Proctor (26), Fowler (27), and Mündnich and Terrahe (29). CT findings sometimes suggest a separation of the facial nerve, but additional canals converging with the mastoid segment (or chorda tympani) are mostly vascular (30). A true nerve separation might be confirmed only by MR imaging (31–35). In opacified temporal bones, visualization of the fallopian canal by CT is difficult, and T1-weighted MR imaging with a 512×512 matrix and thin sections may have an advantage. Facial (and abducens) nerve palsy is more common in embryopathies. We did not find a correlation between facial nerve canal aberration and facial palsy; a bony facial nerve canal might be "empty" (26).

Abnormalities of structures representing the border between the branchial arches and the otic capsule, such as the stapes, oval window, and tympanic segment of the facial nerve canal, were less frequent and not significantly associated with the severity of auricular malformations. The derivatives of the otocyst, such as the round window and labyrinth, were rarely affected. Absence of the round window was exceptional (6%) and was occasionally found in otherwise minor malformations of temporal bone. The cochlea was involved (bilaterally) in only one of our cases, and that was due to thalidomide embryopathy. Embryopathies, whether toxic (eg, thalidomide) or viral (eg, rubella), generally carry a high risk of inner ear malformation and deafness (18). With regard to genetic disorders, a controversial discussion in the literature of the past concerned the issue of whether the inner ear is involved in malformations of the external and middle ear. Since the advent of polytomography, changes of labyrinthine

structures in microtia have most often been described at the lateral semicircular canal (2, 36, 37), which is in keeping with our findings. In our cases, lateral semicircular canal hypoplasias occurred relatively frequently (13%), particularly in patients with severe involvement of the middle ear. The relationship between these anomalies is unclear. The lateral semicircular canal has a spatial relationship to the tympanic cavity, but these structures have separate embryologic derivations. Although the otic capsule begins its development before the middle ear is formed, complete differentiation of the labyrinth might be influenced by branchial structures. In cases of inner ear malformation without microtia, lateral semicircular canal dysplasia appears in 40% (17). A detailed description of inner ear malformations was published by Mündnich and Terrahe (29) and radiologic findings by Curtin (23). Even changes of the vestibular organ seem to be a sign of inner ear dysfunction (38), but audiometry is decisive in this instance. We did not find a CSF fistula, common in labyrinthine dysplasias (but not microtia).

As in the report by Tanghe (17), we found few changes in the labyrinthine segment of the facial nerve canal. Severe dysplasias of the internal auditory canal are also rare. Only a few reports concern internal auditory canal abnormalities in the setting of microtia (29), and a few cases of aberrations of the canalicular segment of the facial nerve canal (without microtia) have also been reported (26, 29, 39).

Epidermoids or cholesteatomas, congenital or secondary, can involve the facial nerve, inner ear, and brain, and they require surgery (40). Congenital cholesteatomas account for 2% of all cholesteatomas and are located at varying places in the temporal bone. In some of our cases there was a typical pattern. Epidermoids also occur intracranially, most often in the cerebellopontine angle, as in our patients, or in the suprasellar cistern (41).

The frequent failure of pneumatization may be due to disturbances of the branchial arches and groove rather than of the first branchial pouch. In this study, severe anomalies of the eustachian tube were rare. However, the cartilaginous parts and the function of the auditory tube are not appreciable by CT. Anomalies of major vessels were also rare, but a ventrally dislocated sigmoid sinus in an anteroposteriorly reduced mastoid could be dangerous for surgery. The carotid canal may be elongated into

the middle ear (embryological inferior tympanic artery) and may result in enlargement of the tympanic segment of the facial nerve canal. This is caused by the persistent stapedia artery, which leads to the medial meningeal artery (42). Hypoplasia or agenesis of the carotid canal is uncommon and is more often unilateral (as in our two cases) than bilateral (43). It may be associated with intracranial aneurysms (44, 45) and neurovascular compressive symptoms due to compensatory vertebral artery ectasia (46).

In conclusion, we found a decrease in the rate of microtic anomalies from the external auditory canal to the middle ear, the boarder of the middle and inner ear, the labyrinth, the internal auditory canal, and the cerebellopontine angle. The current most important and validated preoperative criteria for hearing repair surgery (12), proposed by Jahrsdoerfer (25) and recently substantiated (47), include the clinical appearance of the external ear and the findings at CT examination. The ear surgeon is interested in multiplanar, especially sagittal, views when planning restoration of the external auditory canal. Therefore, direct scanning in these planes (at a low dose) or reconstruction of a three-dimensional data set, acquired by spiral CT (to prevent motion artifacts when imaging children), is necessary.

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