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Congenital Malformations of the External and Middle Ear: High-Resolution CT Findings of Surgical Import

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The external auditory canal, middle ear, and bulk of the ossicular chain develop from the first branchial groove, first and second branchial arches, and first pharyngeal pouch. Embryologic development of these structures is complex and only rarely are two anomalies identical. Development of the inner ear structures occurs independently of external ear structures, and concomitant involvement is unusual. This study includes 11 cases of unilateral external auditory canal atresia and two cases of bilateral atresia. Eight cases (four bilateral) of isolated congenital ossicular anomalies are also included. Emphasis is placed on findings of surgical import. All patients were studied with computed tomography only, because it was believed that the bony and soft-tissue detail achieved is superior to that with conventional multidirectional tomography.

High-resolution computed tomography (CT) has emerged as the method of choice for evaluation of the temporal bone. The purpose of this study was to provide a detailed analysis of congenital aural malformations to emphasize findings deemed critical by surgeons.

Materials and Methods

A General Electric 8800 CT/T scanner was used in all cases. Overlapping CT sections of 1.5 mm thickness were obtained in axial and coronal projections [1]. Infants and very young children required heavy sedation or general anesthesia. If an endotracheal tube was used, a coronal view in the supine position was necessary to facilitate patient monitoring. Excellent images were obtained in all patients, usually within 40 min. All images were targeted using high-bone-detail algorithms.

Results

Twenty-one patients with congenital deformities of the external and/or middle ear were evaluated. These included 11 patients with unilateral external auditory canal atresia, two with bilateral external auditory canal atresia, and eight with isolated congenital ossicular deformity. Of the 11 patients with unilateral atresia, five were male and six were female. In six there was moderate to marked ossicular deformity and in one of these there was inner ear deformity as well. Five patients had minimal to no ossicular deformity. Six had anteriorly located descending facial nerve canals (mastoid segment), five had very poor mastoid pneumatization, five had very small tympanic cavities with high and posteriorly oriented mandibular condyles, and two had very thick irregular atresia plates. Three had incomplete atresia plates and two had membranous occlusion. Of interest, none of these patients had any noticeable structural deformity of the opposite ear.

Of the two patients with bilateral atresia, the male patient had marked ossicular deformity, anteriorly located descending facial nerve canal (mastoid segment), small

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tympenic cavities, high mandibular condyles, and very thick atresia plates. The female patient had minimal to no ossicular deformity and none of the other findings.

Of the eight patients with isolated congenital ossicular deformity, five had involvement of the incus and stapes (two bilateral) and two of the entire ossicular chain (one bilateral). Of these latter two patients one had severe cochlear hypoplasia and the other was diagnosed with Goldenhar syndrome (oculoauriculovertebral dysplasia). A single patient had unilateral stapes maldevelopment with an apparent ossified stapedius tendon. Two patients had dehiscence of the tympanic segment of the facial nerve canal. None had anteriorly located descending (mastoid) segments.

Embryology

Development of the external ear, middle ear, and mastoid is due to a complex interaction of all three germ layers.

The first branchial groove (ectoderm) develops between the first and second branchial arches and becomes the external auditory canal [2–5]. Invagination occurs about 6 weeks, and at this time the lateral one-third of the external auditory canal is formed (cartilaginous part). A solid core of epithelial cells then begins to develop at the medial end of the groove. This persists until about the 26th fetal week, at which time it splits, first in its medialmost part to form the outer surface of the tympanic membrane and then laterally to join the cartilaginous part of the canal. Thus, the medial two-thirds (bony external auditory canal) is subsequently developed. Failure of canalization of this core results in atresia of the external auditory canal.

The connective tissue around the tympanic membrane ossifies and forms the tympanic bone at about 12 weeks. The tympanic bone is a U-shaped membranous bone that surrounds this ectodermal core and forms the sides and floor of the bony external auditory canal [6]. Failure of canalization results in tympanic bone deformity with resultant bony overgrowth. This is the atresia plate [4]. This plate may also in part be from downward extension of the squamous temporal bone [7].

The first and second branchial arches (mesoderm) differentiate into the bulk of the ossicular chain between the 16th and 30th week. The head of the malleus, tensor tympani muscle and tendon, and the body and short process of the incus develop from the first arch (Meckel cartilage). The stapedius muscle and tendon and the rest of the ossicular chain with the exception of the stapes footplate (otic capsule) develop from the second arch (Reichert cartilage) [2–5]. The mandibular condyle, styloid process, and facial nerve canal also develop wholly or partly from the second arch and are thus interrelated. Concomitant anomalies of these structures are therefore not uncommon. Development of the middle and external ears is, therefore, closely intertwined. The inner ear structures develop from the auditory placode at an earlier stage. Since this occurs independently from middle and external ear development, simultaneous anomalies of the inner ear should not occur. In reality, such anomalies do occur with frequencies of 11%–30% [8, 9].

The first pharyngeal pouch (endoderm) develops into the eustachian tube and the tympanic cavity with all its extensions (attic, antrum, mastoid air cells). The inner surface of the tympanic membrane also forms from pharyngeal pouch endoderm. Pneumatization of the tympanic cavity begins at 4 weeks and continues to week 30; however, mastoid pneumatization may continue even into adult life.

A disturbance in its differentiation affects the architecture of the middle ear and mastoid, which may be of considerable surgical significance [5]. It must be emphasized that due to complex embryologic development, the possible deformities of the external and middle ear are endless in variety, and it is unusual for any two branchial arch deformities to be identical.

External Auditory Canal Atresia

Clinically, patients born with external auditory canal atresia are seen with deformity of the auricle and no visible external auditory canal. An obvious jaw deformity may also be present. There is a marked direct correlation between the severity of deformity of the jaw and that of the middle ear and external canal but not between the degree of auricular deformity and the severity of the middle ear and external canal disorder [4]. Abnormalities in other organ systems may exist as well [5, 8].

External auditory canal atresia may be either bony (figs. 1 and 2) or membranous (fig. 3) and may occur with or without associated middle ear malformations. The process is said to be bilateral in 29% of cases, and 61% of those affected are male. When unilateral, it is most often right-sided (58%), and there is a positive family history in 14% [5, 8]. There are numerous possible ossicular anomalies associated with this disorder. The most common is fusion of the malleus and incus in the attic. Bony ankylosis of the neck of the malleus to the atresia plate is also common (fig. 2). Hypoplasia of the manubrium of the malleus is a constant finding in the presence of an atresia plate. Various anomalies of the incus and stapes also occur [5, 7].

There are many classifications of external auditory canal atresia. The most often quoted, that of Altmann [10], separates deformities into slight, moderate, and severe, referring to the degree of difficulty involved in surgical reconstruction (table 1).

Bilateral external auditory canal atresia always requires surgical repair, provided high-resolution CT analysis reveals normal inner ears and normal oval and round windows. The ideal time for reconstruction is at age 4, to prepare the child for entering grade school [11]. The operation for unilateral external auditory canal atresia however may be delayed indefinitely with a normal-hearing opposite ear. Many of these patients wait until their teenage years when correction of the cosmetic deformity becomes most important. Surveillance of these processes with high-resolution CT is important to rule out the presence of concomitant primary middle-ear cholesteatoma [11]. In the presence of such a lesion, surgery is recommended as soon as possible to avoid further damage to the middle or inner ear [11, 12].

The major objectives of high-resolution CT analysis include

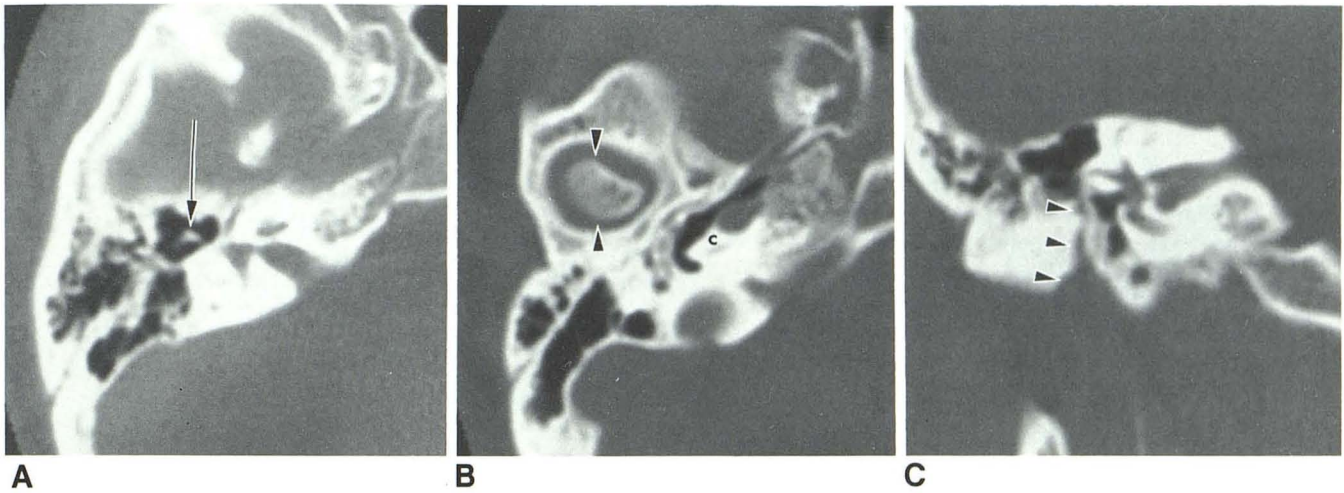


Fig. 1.—Bony external auditory canal atresia in 17-year-old girl contemplating cosmetic repair. **A**, Thick, irregular atresia plate with markedly deformed ossicular mass (*arrow*). Small middle ear cavity. **B**, Mandibular condyle (*arrow-*

heads) is too superior, being at level of cochlea (*c*) **C**, Descending facial nerve canal (mastoid segment) (*arrowheads*) at coronal level of vestibule.

Fig. 2.—Bony external auditory canal atresia in 3-year-old girl. **A**, Fusion of neck of malleus to atresia plate (*arrow*). **B**, Anteriorly located descending facial nerve canal (mastoid segment) (*arrows*). Middle ear is opaque, likely due either to concomitant cholesteatoma or to fluid.

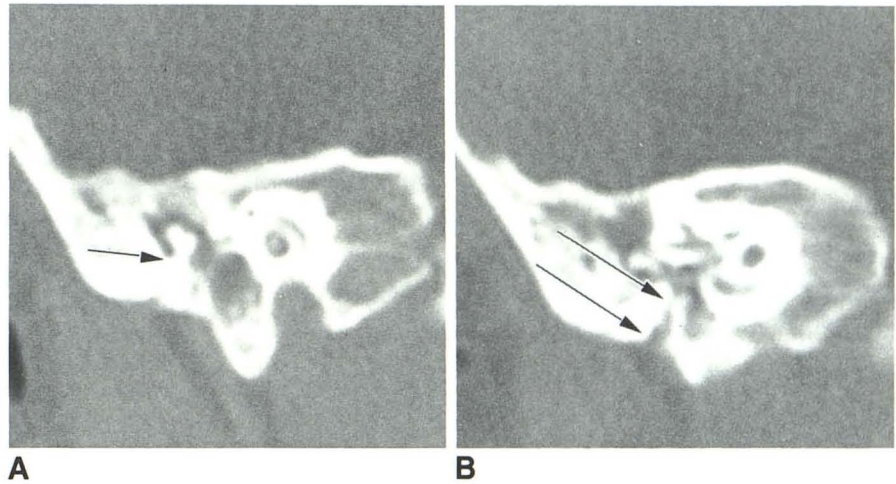
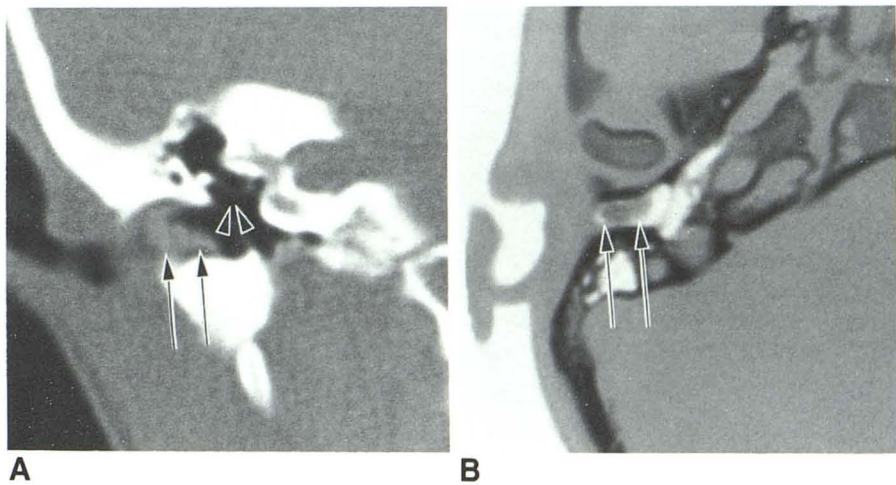


Fig. 3.—Membranous external auditory canal atresia in 10-year-old boy with deformed auricle and no visible canal. External auditory canal is occluded by soft-tissue density (*arrows*). In **A**, ossicular chain is unremarkable (*arrowheads*). At surgery a fibrous plug was found within an otherwise normal bony external auditory canal.



identification of the type of anomaly and determination of surgical correctability [13, 14]. One must evaluate the atresia plate, the mandibular condyle, the size of the tympanic cavity, the ossicular chain, the inner ear, and the fenestrae. Evalua-

tion of the facial nerve canal, sigmoid sinus, and mastoid pneumatization is also critical (table 2).

The atresia plate may be of varying thickness and is either complete or incomplete [15] (fig. 4). The surgical implications are obvious. The position of the mandibular condyles is highly variable. In the absence of a normal tympanic bone the condyle must articulate with the mastoid directly and often must assume a more posterior and superior location (fig. 1B). The size of the tympanic cavity and the degree of pneumatization reflect the size of the surgical field and are of paramount importance. The status of the ossicular chain is variable and is of obvious significance, as the surgeon may need to plan tympanoplasty and ossicular reconstructive procedures. Any inner-ear abnormality of course precludes operation. The middle ear must also be studied to rule out associated cholesteatoma or other complications of tubotympanic disease such as fluid. CT has a clear advantage over conventional multidirectional tomography in this respect (fig. 2).

Many authors have discussed the associated frequency of the anomalous course of the facial nerve canal [5, 7, 12-14]. The most common variation both in the literature and in our experience is an anteriorly located descending (mastoid) segment. This often is seen at the level of the vestibule on coronal images (fig. 1C).

TABLE 1: External Auditory Canal Deformity

Classification of External Auditory Canal Atresia	
Slight (small external auditory canal):	
Hypoplastic tympanic membrane	
Hypoplastic tympanic bone	
Small middle ear cavity	
Varying ossicular deformity	
Well aerated mastoid	
Moderate (absent external auditory canal):	
Small middle ear cavity	
Atresia plate	
Deformed malleus and incus	
Well aerated mastoid	
Severe (absent external auditory canal):	
Hypoplastic middle ear cavity	
Severe ossicular deformity	
Poor mastoid aeration	

Note.—Modified from [10].

TABLE 2: External Auditory Canal Atresia: Associated Anomalies

Anomaly	No. (n = 15)
Bony atresia	13
Membranous atresia	2
Acellular pneumatization	5
Abnormal round window	0
Abnormal oval window	1
Anteriorly located sigmoid sinus	1
Small tympanic cavity	7
Superiorly and posteriorly placed mandibular condyle	7
Thick atresia plate	5
Incomplete atresia plate	3
Anteriorly located descending facial nerve canal (mastoid segment)	8

Isolated Congenital Ossicular Deformity [5, 13, 16]

Isolated congenital ossicular deformity was seen in eight of our patients and is not rare (figs. 5 and 6). In four of these cases the anomaly was bilateral. Most of these patients were young adults with a conductive hearing loss, and the diagnosis of otosclerosis was suspected. A 40-60 dB conductive hearing loss in the absence of a history of trauma or infection indicates congenital ossicular deformity in a child; in an adult it implies either fenestral otosclerosis or congenital ossicular deformity. Numerous anomalies are possible and bilaterality is common [16-18].

The stapes is the most common ossicle involved and was abnormal in all our cases of isolated deformity. Reported anomalies include both congenital absence and crural deform-

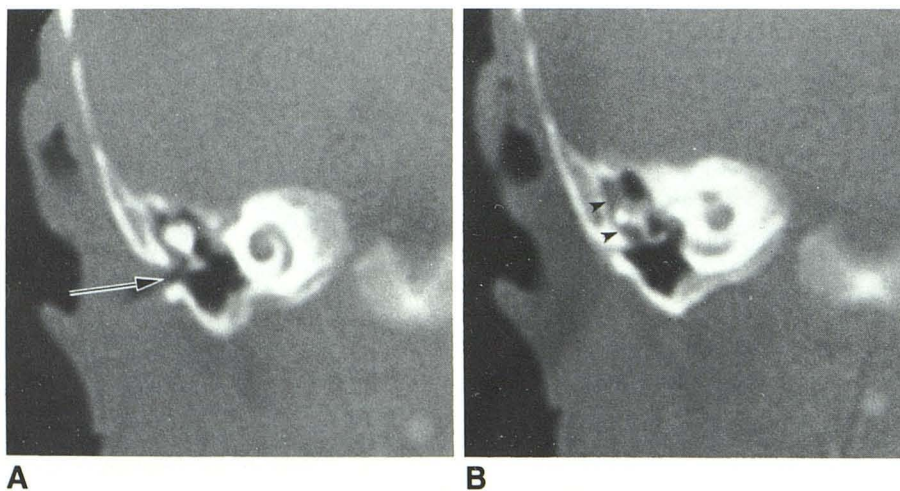


Fig. 4.—Bony external auditory canal atresia in 4-year-old girl with deformed auricle. **A**, Very thin, incomplete atresia plate. Small, patent lumen (arrow). Intact ossicular chain. Normal malleus head. **B**, Debris in Prussak space (arrowheads).

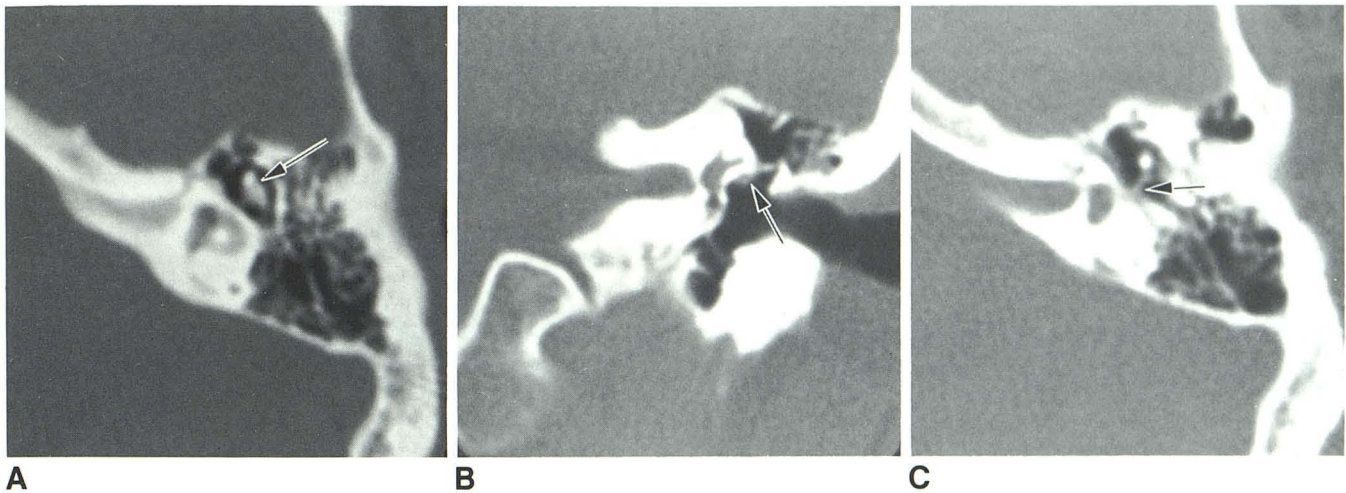


Fig. 5.—Isolated congenital ossicular deformity (Reichert) in 23-year-old man with conductive hearing loss, no history of chronic otitis, and normal tympanic membrane. **A**, Normal articulation between malleus head and incus body on

more superior axial view (*arrow*). **B** and **C**, Long process of incus is hypoplastic with much more horizontal orientation; it apposes horizontal part of facial nerve canal (tympanic segment) (*arrows*). Normal stapes was not imaged.

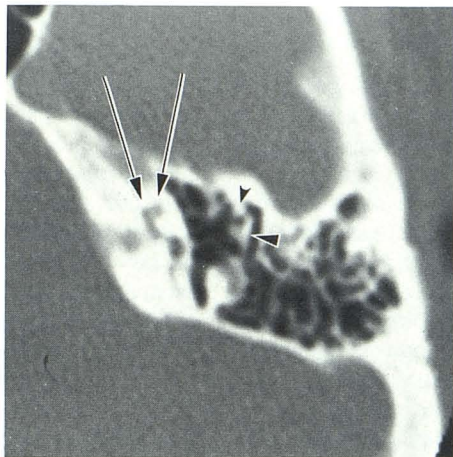


Fig. 6.—Isolated congenital ossicular deformity (Meckel and Reichert) in 40-year-old woman with profound mixed hearing loss bilaterally. Small malleus head (*small arrowhead*) has poor articulation with peculiarly shaped incus (*large arrowhead*). Significant cochlear hypoplasia (*arrows*). Identical abnormality was present on opposite side.

ity. We have not seen a surgically verified case of congenital oval window nondevelopment; however, this entity does not appear to be rare as it has been well documented. Congenital incus abnormalities include absence or hypoplasia of the long process (fig. 5), total absence, incudostapedial joint separation, and attic fixation. The least involved ossicle is the malleus. Attic fusion is probably the most common anomaly identified. This is often associated with fenestral otosclerosis [17]; however, we had only one such case.

The long process of the incus, lenticular process of the incus, and stapes superstructure are all derived from the second branchial arch (Reichert cartilage). Concurrent anom-

TABLE 3: Isolated Congenital Ossicular Deformity: Summary

Affected Structures	No.
Incus and stapes only	5 (2 bilateral)
Entire chain	2 (both bilateral)
Stapes only	1 (unilateral)

alies of these structures would therefore be expected to occur on a consistent basis. In fact, this combination type of isolated ossicular anomaly was the most common in our experience (table 3).

The major thrust of analysis involves the determination of the exact type of ossicular anomaly; however, one must also evaluate the size and orientation of the tympanic cavity, condition of the oval window, and the position of the facial nerve canal. Careful evaluation of the opposite ear is also critical.

Virtually all of these anomalies can be surgically corrected. The type of procedure used varies extensively among individual surgeons [19]. Total ossicular replacement prostheses (TORPs) and partial ossicular replacement prostheses (PORPs) are commercially available; however, many highly skilled and experienced otologic surgeons do not use them [20]. A simple wire prosthesis may be preferred. The surgeon may also prefer to personally resculpture the existing ossicular chain or employ allograft ossicular implants [17]. It should be emphasized that the end result in these cases is of paramount importance, and the means by which the surgeon attains a safe hearing ear is relatively inconsequential.

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