Journal of Neuroradiology (2013) 40, 192—197

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ORIGINAL ARTICLE

# Does microtia predict severity of temporal bone CT abnormalities in children with persistent conductive hearing loss?

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**KEYWORDS**
Conductive hearing loss;
Children;
Microtia;
Temporal bone CT

**Summary**
*Purpose:* This study aimed to determine the spectrum of temporal bone computed tomography (CT) abnormalities in children with conductive hearing loss (CHL) with and without microtia.
*Patients and methods:* From 1993 to 2008, a total of 3396 pediatric records including CHL were reviewed at our institution and revealed 180 cases of persistent CHL, 46 of whom had diagnostic temporal bone CT examinations. All of these examinations were systematically reviewed by two pediatric neuroradiologists, working in consensus, who had 5 and 18 years, respectively, of dedicated pediatric neuroradiology experience.
*Results:* Of the 46 children, 16 were boys and 30 were girls (age: 0.2—16 years; mean: 5 years). Also, 21 (46%) children had microtia and 25 (54%) children did not, as determined by clinical evaluation. External auditory canal atresia/stenosis (EAC-A/S) was the most common anomaly in both microtia and non-microtia groups. Two or more anomalies were observed in 18/21 children with microtia. The frequency of EAC-A/S was greater in children with microtia versus those without it (86% versus 32%, respectively; $P = 0.0003$). Syndromic diagnoses were also significantly more frequently made in children with microtia versus those without microtia (76% versus 20%, respectively; $P = 0.0001$). Temporal bone CT scans were normal in 10 children (22%) with persistent CHL.
*Conclusion:* Microtia is an important finding in children with CHL. EAC and middle ear/ossicle anomalies were significantly more frequently seen in children with microtia, and multiple

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0150-9861/$ - see front matter 
http://dx.doi.org/10.1016/j.neurad.2012.10.002

anomalies and bilateral microtia were more common in children with syndromic associations. These findings highlight the importance of understanding the embryological development of the temporal bone. The presence of one anomaly should raise suspicion of the possibility of other anomalies, especially in the setting of microtia. Bilateral microtia and multiple anomalies should also raise suspicion of genetic syndromes.


## Introduction

Conductive hearing loss (CHL) is defined as hearing loss resulting from interference of the mechanical reception or amplification of sound by the cochlea. A wide range of pathologies involving the external, the middle and, rarely, the inner ear may result in CHL.

The etiology of CHL can be congenital, post-infectious or post-traumatic in origin. In addition, CHL may be seen in complex syndromes such as the rare autosomal-dominant inherited Treacher Collins—Franceschetti syndrome or as part of the oculo-auriculo-vertebral spectrum (OAVS), which includes the Goldenhar syndrome. Typically, microtia—anotia refers to a congenital malformation of the pinna, ranging from partial deformity to complete aplasia of the pinna, commonly associated with anomalies of the external auditory canal. In addition, middle-ear anomalies with abnormalities of the ossicular chain are frequently noted along with variable degrees of deafness [1—3]. The exact etiology of these complex malformations is unknown, but they are believed to result from a malfunction or disruption of the early embryonic development of the first/second branchial arches and first pharyngeal pouch. This would explain the combined facial malformation/asymmetry and ear malformation [4,5]. More important, in most children with CHL, appropriate treatment can improve hearing function and consequently enhance speech development. For this reason, a detailed and correct diagnosis of CHL is a *sine qua non*.

History-taking, physical examination, and thorough air-conduction and bone-conduction audiometry are essential for identifying CHL and assessing the severity of deafness. Cross-sectional imaging such as computed tomography (CT) and magnetic resonance imaging (MRI) are recommended for evaluating the precise anatomy of the temporal bone and retrocochlear structures. CT and MRI of the temporal bone give partially overlapping and complementary information. CT is especially helpful in the delineation of the external auditory canal (EAC), the middle-ear cavity including the ossicles, the mastoid and petrous air-cell system, the cochlea and vestibular system, the inner auditory canal (IAC), and cochlear and vestibular aqueducts, and osseous canal of the facial nerve. MRI is more sensitive for the internal architecture of the cochlea and for retrocochlear structures, including the cochlear, vestibular and facial nerves as well as the brain stem. CT is typically performed in the axial plane, and coronal multiplanar reconstructions (MPR) are acquired. MRI also has the advantage of T1-weighted and heavily T2-weighted three-dimensional sequences that enhance the contrast between bone/soft tissue and cerebrospinal fluid (CSF)/endolymph [6].

The goal of the present retrospective study was to evaluate temporal bone CT examinations in a cohort of children with persistent CHL to determine the spectrum of temporal bone CT abnormalities in CHL children with and without microtia. This evaluation may further optimize the diagnostic approach and, eventually, the treatment of CHL children.

## Patients and methods

### Patient population and selection criteria

Institutional review board (IRB) approval was obtained for this retrospective study. The clinical database of our hospital was electronically searched for children that had been referred/evaluated for suspected or confirmed CHL. The clinical records were subsequently studied by an experienced pediatric otorhinolaryngologist (S.I.) for a confirmed diagnosis of persistent CHL. Children with a history of cholesteatoma, tympanic membrane perforation, chronic otitis media and/or middle-ear effusion or sensorineural hearing loss were excluded. Presence or absence of microtia was based on clinical evaluation.

### Imaging method

The available CT studies were systematically examined by two pediatric neuroradiologists working in consensus, with 5 and 18 years, respectively, of experience dedicated to pediatric neuroradiology. The data were kept anonymous and randomly allocated to the reviewing neuroradiologists.

The CT studies were first evaluated for completeness of data and image quality. The minimum requirements for study inclusion were that the CT study had to include axial and coronal images of thin sections (0.5—0.75 mm), and data reconstruction using a high-resolution bone algorithm. Multiplanar reconstructions from the original dataset were performed at the scanner and made available for review in the picture archiving and communications system (PACS). The availability of contrast-enhanced soft-tissue images was not considered necessary. The studies were graded as non-diagnostic, diagnostic or highly diagnostic. Temporal bone CT scans with incomplete or poor/non-diagnostic image quality were excluded from the data evaluation.

### Image analysis

All CT data were systematically reviewed according to a predefined evaluation sheet. The following anatomical structures were evaluated: pinna; EAC; tympanic membrane; size and morphology of the middle-ear cavity; presence and integrity of the middle-ear ossicles/ossicular chain (malleus, incus, stapes); oval window; cochlea; round window; vestibulum; facial nerve; and mastoid cells. All listed structures were evaluated for both ears. In addition,