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Imaging Diagnosis

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The temporal bone is a part of the cranial bone that constitutes the ear. Vital organs for hearing such as the external auditory canal (EAC), eardrum, auditory ossicles, cochlea, semicircular duct, otolithic organ, internal auditory canal, facial nerve, and internal carotid are crammed in this narrow space. Although simple X-ray of the middle and inner ear (Schüller method and Stenvers method) provide some findings and is still generally performed at present, it is difficult to observe the structure of the temporal bone clearly. Subsequent to the introduction of computed tomography (CT) in the 1970s, helical scanning was developed in the latter half of the 1980s, allowing the temporal bone to be shown more in detail [1]. Many patients with microtia/atresia of the EAC also have a middle ear anomaly, and a CT scan is essential for understanding the condition of the middle ear.

Age for CT Scanning

Babies are sometimes referred to our outpatient department with CT images at several months of age. The timing for performing CT differs among medical institutions. CT is performed within several months of birth for the following reasons: (1) it is the best method for revealing the condition of the external and middle ear; (2) it shows the presence or absence of an inner ear anomaly and enables identification of the cause of hearing loss; (3) it shows the presence or absence of congenital cholesteatomatous otitis media, and (4) it is used to understand the severity of the temporal bone anomaly at an early age in order to confirm the possibility of EAC reconstruction and to reassure the patient's family. Thanks to recent progress in CT equipment, the dose of radiation is decreasing, but the radioactivity exposure with CT is still dozens of times higher than that with simple radiography. Furthermore, because of their smaller body size, the level of exposure per organ is 2–5 times higher for children than adults, under the same conditions [2]. Also, image contrast is generally poorer in chil-

dren because they have less fat and smaller organs than adults. Thus, the dose of radioactivity must be increased when a higher contrast image is desired. CT in children should therefore be carefully planned considering the above.

It was reported that congenital cholesteatomatous otitis media was observed in 4–7% of patients with congenital stenosis or occlusion of the EAC [3, 4]. With progression of cholesteatomatous otitis media, surrounding bones are destroyed, and inner ear disorders and facial palsy are induced; serious symptoms associated with meningitis and brain abscess develop when the lesion reaches the cranium. It is therefore better to perform a CT scan as soon as possible to rule out congenital cholesteatomatous otitis media. However, diagnosis of cholesteatomatous otitis media based on CT images is sometimes difficult because fetal remnants and exudation are often retained in the middle ear cavity within 1 year of birth. Also, it may take several years before the disease becomes advanced enough to manifest obvious symptoms even if a diagnosis was made during infancy. MRI is useful diagnostically to help differentiate congenital cholesteatoma from other soft-density exudation of the middle ear. On MRI, cholesteatoma will appear with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

If a patient shows no auditory brainstem response or their auditory acuity is gradually deteriorating, CT should be performed as soon as possible to reveal the condition of the inner ear. However, residual auditory acuity is observed not only in patients with unilateral microtia whose normal ear has normal auditory acuity, but also in patients with bilateral microtia. Thus, little useful information is obtained from a CT scan performed at an early age in preparation for the use of hearing aids and commencing auditory training. Reconstruction of the EAC is often performed around the age of 10 years, and CT is performed immediately before to obtain information for the surgery. CT performed during infancy does not provide direct information for pinnaplasty and reconstruction of the external ear. Because the skull bone continues to grow until the age of 15 years, the mastoid process of the temporal bone to undergo mastoidectomy is not the same as that in infancy. CT findings during infancy are nevertheless useful for understanding the condition of the middle ear anomaly and the potential for improvement of auditory acuity after EAC reconstruction in the future. We perform the initial CT scan at around 3 years of age for patients without otitis media findings, such as obvious cholesteatomatous otitis media, or for patients who do not present with severe or progressive hearing loss.

Image Assessment

Patients with microtia/atresia of the EAC often have a complication of a middle ear anomaly due to developmental factors. The grading system of Jahrsdoerfer et al. [5] using CT findings is useful for judging whether or not auditory acuity will be improved by surgery (table 1). In this system, 1 point each is given to presence or absence of the oval window, round window, middle ear space, abnormal facial nerve pathway, malleus and incus complex, pneumatized mastoid, incus-stapes connec-

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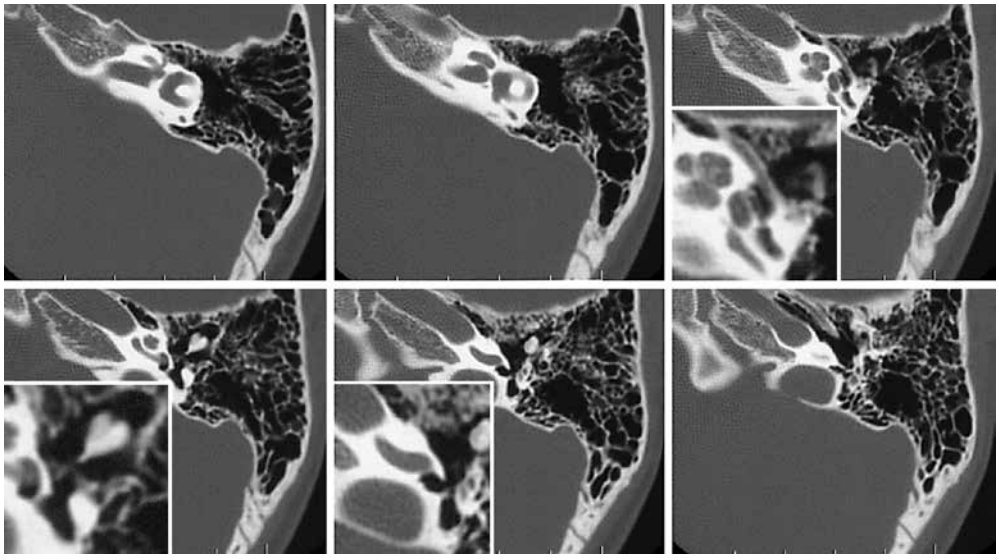

Fig. 1. Left temporal bone CT of a patient with unilateral microtia and atresia. The temporal bone is well developed.

Table 1. Jahrsdoerfer grading system

	Point
Stapes present	2
Oval window open	1
Middle ear space present	1
Anomalies of facial nerve	1
Malleus-incus completely present	1
Mastoid pneumatized	1
Incus-stapes connection present	1
Round window present	1
Appearance external ear	1
Total	10

tion, and EAC bone, and 2 points is given to stapes, for judgment by a total score (perfect score = 10 points). The lower the score, the more severe is the anomaly of the temporal bone.

Actual cases are shown below. Figure 1 is a patient with unilateral microtia. Stapes, malleus and incus complex, incus-stapes connection, oval window, round window, and middle ear space were observed, and the mastoid air cells grew favorably. The facial nerve follows a normal pathway, but the bony EAC is not observed. This patient scored 9 points. Figure 2 depicts a patient with bilateral microtia. The oval window

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