including temporal bone CT images, air and bone conduction ASSR, ABR, stapedial reflex, and so on.

In cases of (C) and (D) involving complete atresia, aberrant courses of the facial nerve are commonly observed. In middle ear surgery that does not involve malformations, the prominence of the lateral semicircular canal and the short process of the incus form the referrence point for surgical site orientation. However, in cases involving EAC atresia and middle ear malformations, as was evident in the two cases presented here, this relative spatial relationship changes, and in order to avoid damaging the facial nerve it is necessary to use CT images to confirm the facial nerve path and spatial relationships of the ossicles and lateral semicircular canal for each case. It is also essential to use facial nerve monitoring during surgery.

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Points

1. Congenital EAC atresia occurs with a frequency of approx. one in 10,000; 15–25% of cases are bilateral.
2. Microtia often accompanies EAC atresia and stenosis.
3. The embryological origins of the external/middle ear and the inner ear are different, but combined malformations sometimes occur.
4. Auditory, language development, and other functional testing is required along with visual, imaging, and other morphological examination.
5. A diagnosis of conductive hearing loss is facilitated by temporal bone CT imaging, bone conduction ABR, bone conduction ASSR, stapedial reflex, and other testing.
6. In cases of cartilaginous part stenosis, temporal bone CT examination is essential as external auditory canal cholesteatoma often occurs.
7. Cases of EAC atresia and stenosis are often accompanied by fixation of the ossicles to the bony wall of the attic and the atretic plate.
8. In cases of EAC atresia, facial nerve anomalies also occur, with the nerve's path running inferior (tympanic segment) and anterior (mastoid segment) to normal.
9. In determining the surgical indication, overall consideration must be given to such factors as location of the malformation, complications, whether the malformation is bilateral or unilateral, and other factors.
10. The Jahrsdoerfer score, which provides a numerical evaluation of the condition of EAC atresia, is useful in estimating the effectiveness of surgery.

2 Auditory Ossicles and Middle Ear

Congenital Ossicular Malformation

Congenital ossicular malformation may accompany EAC atresia or stenosis, or it may occur independently. Here we will examine cases in which there are no anomalies of the external auditory canal, as cases accompanying EAC atresia and stenosis were already covered in the previous section. Approx. 30% of cases of ossicular malformation are bilateral [1], with some reports finding no sex difference [2] and others finding it slightly more common in females [1]. Ossicular anomalies are diverse, with possible malformation and partial defects in each of the three auditory ossicles, along with resulting joint disruption (primarily the incudostapedial joint) and fusion/fixation either among the auditory ossicles or with the surrounding bone.

Classifications of ossicular malformation have been attempted from the standpoints of morphology, embryology, and treatment, but here as one example we will present the classification system advocated by Cremers et al [3], which is based on the type of surgery required (table 2). According to this classification, Class 1 is when there is fixation of the stapes but no other anomalies; Class 2 is when, in addition to fixation of the stapes, there are anomalies in other auditory ossicles; Class 3 is when the stapes is mobile and there are anomalies in other auditory ossicles; and Class 4 is when there is atresia of the labyrinthine window. In terms of surgical intervention, Class 1 indicates that standard stapes surgery is sufficient; Class 2 indicates that, in addition to stapes surgery, surgical procedures on other auditory ossicles will be required; Class 3 indicates the anomaly can be dealt with through ossicular chain reconstruction involving procedures on auditory ossicles other than the stapes; and Class 4 indicates the need for new fenestration to the inner ear. Following this classification system, Cases 1 and 2 presented here are Class 2, Case 3 is Class 3, and Case 4 is Class 4 (accompanied by drooping of the facial nerve). Of these cases, only Case 4 failed to obtain hearing improvement. A survey of the literature indicates that the record of postoperative hearing improvement for Class 4 cases is less favorable than in other classes [2, 3], indicating that prudence is required when considering surgical indication.

Fixation of the footplate of the stapes occurs in 69% of overall cases according to a report by Teunissen and Cremers [3], in 59% according to Park et al [2], and 26% according to Kojima et al [1]. Despite the variation, a high incidence rate is indicated in all samples. Kojima's report also indicates a frequency of over 46% for incudostapedial joint discontinuity. Fixation of the footplate of the stapes cannot be diagnosed through imaging, and even supposing there is no stapedial reflex this may also be due to discontinuity or fixation of the malleus or incus, making it impossible to determine in advance whether or not the stapes is fixed in cases of ossicular malformation. Consequently, diagnosis of ossicular malformation frequently requires an exploratory tympanotomy in addition to physiological and imaging tests. If anomalies of the ossicular chain are confirmed during the exploratory tympanotomy, the surgeon can then proceed to perform conduction reconstructive surgery in accordance with the findings obtained. However it is necessary to formulate a surgical plan in advance that includes the possibility of stapes surgery, and also to obtain the informed consent of the patient.

As mentioned in the section on otosclerosis, even in children there are cases of stapes fixation due to otosclerosis [4] or bone metabolism disorders that are difficult to differentiate from stapes fixation as a result of congenital malformation. Findings that indicate otosclerosis include decalcification in the anterior end of the oval window and gradual progression of hearing loss, but decalcification is not observed in all CT images of otosclerosis and it takes time to confirm longterm hearing deterioration, so it would seem that in practical terms differential diagnosis is not always possible.

Stapes Surgery in Children

There has been much debate over whether or not to conduct stapes surgery for congenital stapes fixation and otosclerosis in children. The main points to consider are whether or not longterm, stable hearing improvements can be obtained, what risks are involved in surgery, whether or not otitis media affects the inner ear, and so on. House et al [5] started their article stating "Stapedectomy on a child? Never! But we believe there are indications.", and showed that their results of stapedectomy in children appear to be as satisfactory as results in adults. Ordinarily,

Table 2. Classification of congenital anomalies of middle ear

Class	Main Anomaly	Subclassification		No. of Ears
1	Congenital stapes ankylosis			44
2	Stapes ankylosis associated with another congenital ossicular chain anomaly			55
3	Congenital anomaly of ossicular chain but mobile stapes footplate	Discontinuity in ossicular chain		11
		Epitympanic fixation		20
4	Congenital aplasia or severe dysplasia of oval window or round window	Aplasia		10
		Dysplasia	Crossing facial nerve	3
			Persistent stapedial artery	1

From Teunissen and Cremers [3]

we can perform stapes surgeries safely in children and can obtain favorable results with no greater incidence of complication than in adults [4–6]. However, the youngest recipients of stapes surgery in these reports are around five years old. Taking into consideration the general age at which postoperative rest becomes substantially possible and the occurrence of otitis media is reduced, it would seem safer to wait until children reach school age or later to perform stapes surgery. Prior to that, during infancy, it is important to provide hearing aid appropriate to the level of hearing loss and take action to ensure that hearing and language development are not impaired. These cases essentially involve conductive hearing loss, so the effect to be gained from a hearing aid is substantial.

CT Diagnosis of Ossicular Malformation

In CT diagnosis of congenital ossicular malformation one observes the morphology of each auditory ossicle and whether or not there is space between it and the surrounding bone. In particular, fixation of the the head and neck of the malleus or the lateral process of the incus to the surrounding bone and discontinuity of the incudostapedial joint due to defects in the lenticular process of the long process of the incus or the head of the stapes are frequently observed. There are also many slight variations from normal in the shape and position of each of the auditory ossicles, necessitating a close comparison with the normal control images. The thickness of the anterior limb of the stapes is 0.5 mm or less and only occupies a small part in the CT image slab, so it is depicted in the CT image as grey rather than white and, if not properly captured in the cross-section, cannot be viewed in its entire form. Therefore when the stapes is poorly depicted in the CT image it is difficult to determine whether a finding is due to an anomaly in the stapes itself or to the partial volume effect in the image, and one must also consider functional test results such as hearing and the stapedial reflex. As mentioned above, in the end an exploratory tympanotomy is often required.

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Ossicular Disruption with Stapes Fixation

Subject: male, 3 years, 6 months

History and Clinical Findings

The subject was born with auricular deformity and was placed under the observation of a plastic surgeon since soon after birth. At around one year old, his parents noticed that he spoke few words, but at his three-year health exam he was able to understand two-word sentences so observation continued as before. However, recently he frequently failed to react to whispers or answer even when called in a loud voice and so was referred to our department. The auricles on both sides were bent over—so-called lop ear—but there was no stenosis of the external auditory canal and no clear abnormal findings for the tympanic membrane. However, no stapedial reflex was observed despite a type A tympanogram for both ears (fig. 6). Static compliance (SC) for this case was 1.0 cc, slightly higher than the normal range of 0.2 to 0.9 cc, suggesting some form of excess mobility in the tympanic membrane or ossicular chain. ASSR (air conduction) test thresholds were elevated, with 100–110 dB in the low frequency range and 50–60 dB for the mid-to-high ranges. A bone conduction ASSR test revealed thresholds in the neighborhood of 30 dB for both ears, for a finding of conductive hearing loss (fig. 7).

Patient CT Findings

Here we discuss the left side, but the findings are essentially the same for both sides. Mastoid air cell development is normal and there is no soft tissue density to indicate inflammation. In observing the vicinity of the ossicular chain, first we note that the anatomical structures around the tip of the incus long process are not clearly depicted, so that the lenticular process is unobservable (fig. 8:1), and that the stapedius tendon appears to be inside the bony tube (fig. 8:1). Normally the stapes can be observed as an arch formed by the anterior and posterior crus, but in this case such an arch structure is not apparent (fig. 8:2). In the cross-section in which the base of the crus can be observed, in the normal control sample both the anterior and posterior crus are identified, but in this case there is only a thickening in one location at the center of the footplate (fig. 8:3), leading one to suspect a malformed stapes with just a single crus. Also, the tympanic segment of the facial nerve runs inferior to normal, so that the facial canal is thickly depicted immediately lateral to the footplate of the stapes in the same cross-section as the footplate (fig. 8:3).

Surgical Findings

Because conductive hearing loss is suspected in this case, we decided to first perform a left exploratory tympanotomy, with ossicular chain reconstruction if possible. Observing inside the tympanic cavity, the tip of the long process of the incus was gradually tapered, and the lenticular process was deficient and fixed to the head of the stapes by funicular soft tissue. Also the stapedius tendon was inside the bony tube and this tube was fused directly to the head of the stapes. The footplate of the stapes was a

bluish color due to slight thinness and a single crus stood up from the center of the footplate. The footplate of the stapes was fixed to the surrounding bone, with no mobility whatsoever. The tympanic segment of the facial nerve was more inferior than normal and slightly overhung the oval window.

In this case, in order to carry out ossicular chain reconstruction, it was necessary to either extract the deformed upper structure of the stapes and perform a small fenestration slightly inferior to the oval window, or perform stapedectomy. Also, because the long process of the incus was hypoplastic, if a teflon wire piston was used the wire would need to be attached to the malleus. Since the child was still only three years old and there were various risks, it was decided not to perform stapes surgery but rather to close the incision as is and use a hearing aid. The parents were consulted and plans have been made to perform stapes surgery once the child reaches school age.

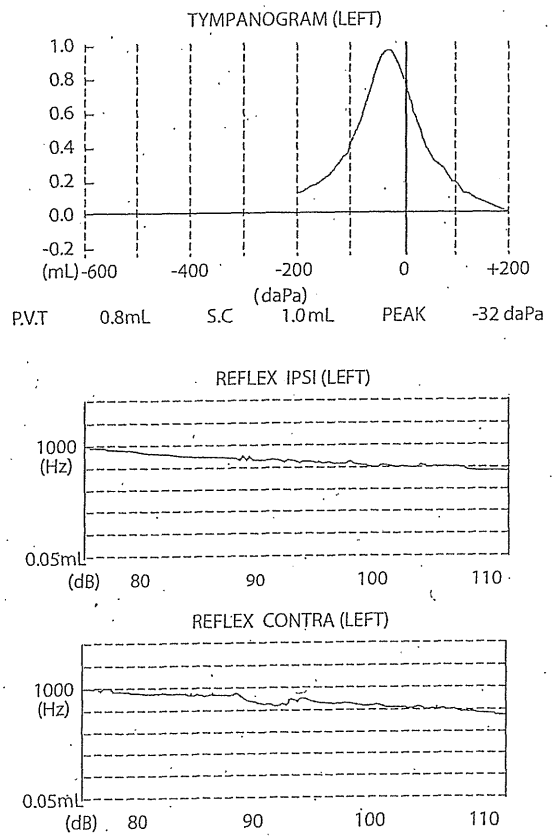


Fig. 6. (Case 1) Tympanogram and stapedial reflex test findings: preoperative.

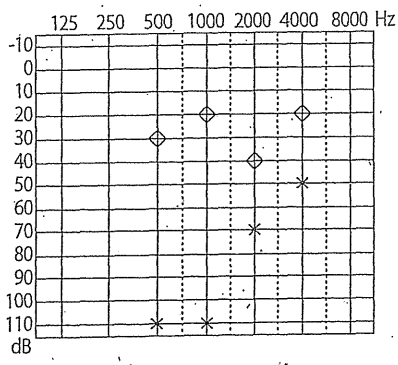


Fig. 7. (Case 1) ASSR findings; preoperative
(×: air conduction threshold; ◇: bone conduction threshold)

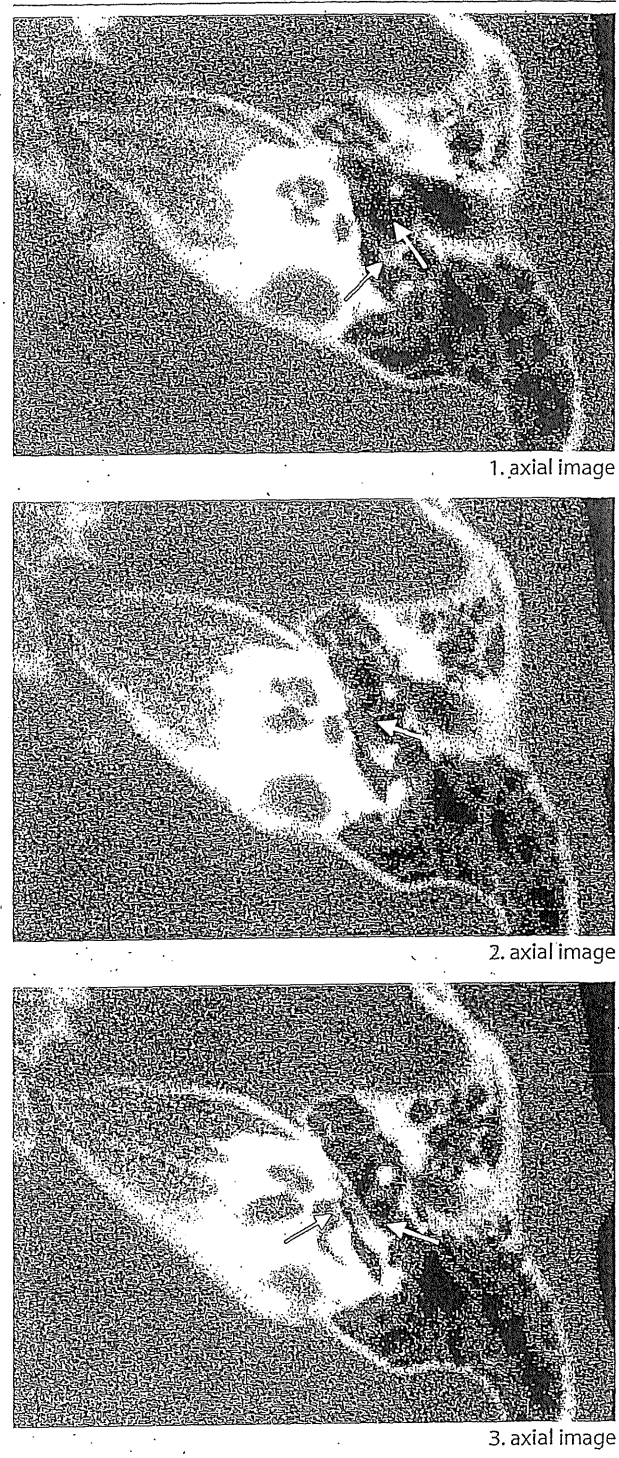
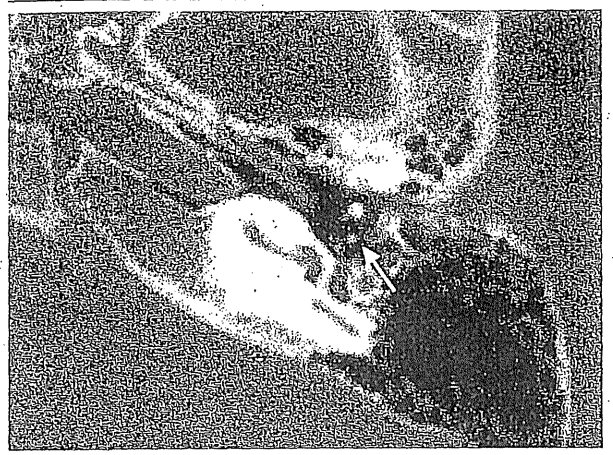


Fig. 8. (Case 1) Left ear CT: preoperative

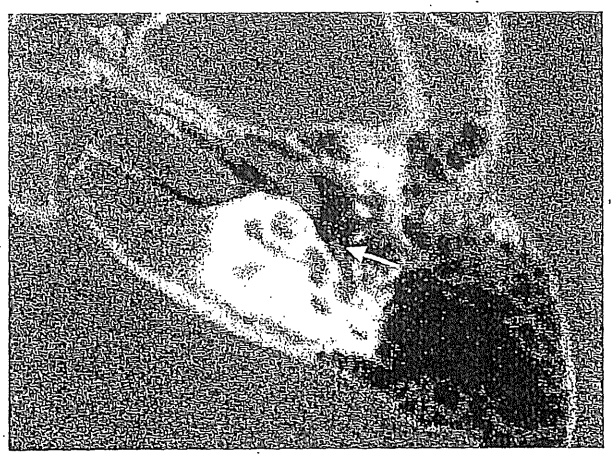
[Patient CT Findings]

The anatomical structures around the tip of the incus long process are not clearly depicted, so the lenticular process is unobservable (1:), and the stapedius tendon appears to be inside the bony tube (1:). Normally the stapes can be observed as an arch formed by the anterior and posterior crus, but in this case such an arch structure is not apparent (2:). In the cross-section in which the base of the crus can be observed, there is only a thickening in one location at the center of the footplate (3:), leading one to suspect a malformed stapes with just a single crus. Also, the tympanic segment of the facial nerve runs inferior to normal, so that the facial canal is thickly depicted immediately lateral to the footplate of the stapes in the same cross-section as the footplate (3:).

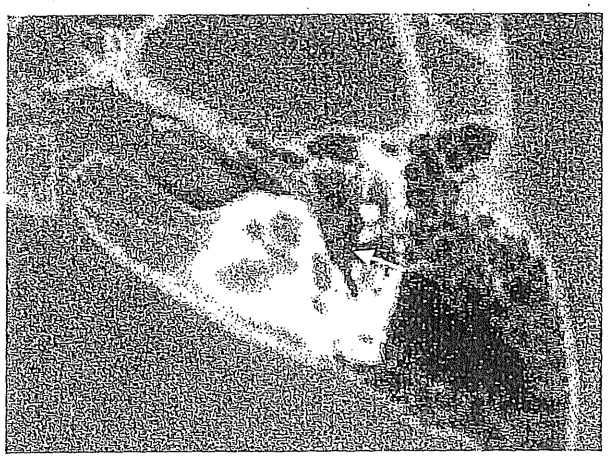
Normal Control CT Findings



n1. axial image



n2. axial image



n3. axial image

《Normal Control CT Findings》
n1: ↖ lenticular process of incus; n2: ⌒ arch of stapes;
n3: ⌒ anterior and posterior crus of stapes

Ossicular Deformities with Stapes Fixation

Subject: female, 12 years old

History and Clinical Findings

The subject underwent plastic surgery at nine months old for a cleft palate. A family member noticed her hearing loss at around age five. Testing done at her regular pediatric hospital found bilateral mixed hearing loss between moderate and severe, and she was referred to our department to assess whether or not surgical intervention would be appropriate.

Pure tone audiometry conducted by us indicated mixed hearing loss with average hearing level of 50.0 dB right and 66.7 dB left (fig. 9). On initial inspection the tympanic membrane appeared almost normal, but further observation revealed that its diameter was slightly smaller and the handle of the malleus slightly shorter than normal. Surgery was performed on both sides, however here we will discuss only the left ear, in which hearing loss was worse.

Preoperative Patient CT Findings

Mastoid air cell development is deficient, but no effusion or other soft tissue density is apparent. The vicinity around the tip of the long process of the incus is unclear and the lenticular process is unrecognizable (fig. 10:1). This case does not include a cross-section in which the incudostapedial joint itself is displayed, and disruption is suspected. The superstructure of the stapes is slightly asymmetrical in form, but the head, anterior crus, and posterior crus of the stapes are all visible (fig. 10:1). The cross-section of the long process of the incus is flatter than normal (fig. 10:2) and the neck of the malleus contacts the anterolateral wall of the external auditory canal (fig. 10:2), with suspected fixation. Also, the space lateral to the body of the incus, which normally can be observed as quite wide, is narrow in this case (fig. 10:3). Fixation is probable here as well.

Surgical Findings

The posterosuperior wall of the bony segment of the external auditory canal and the lateral attic wall were carefully removed. With the ossicular chain clearly visible, it was apparent that the neck of the malleus contacted and was fixed to the anterior tympanic spine, and furthermore the short process of the incus was fixed to the surrounding bone, with no mobility in either. The long process of the incus was bent medially toward the tip, and the

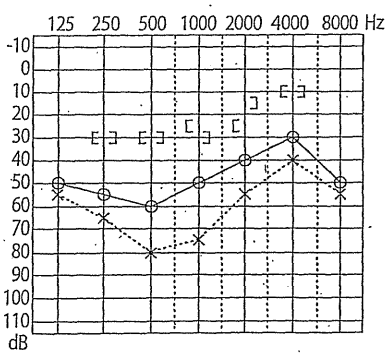


Fig. 9. (Case 2) Audiogram: preoperative

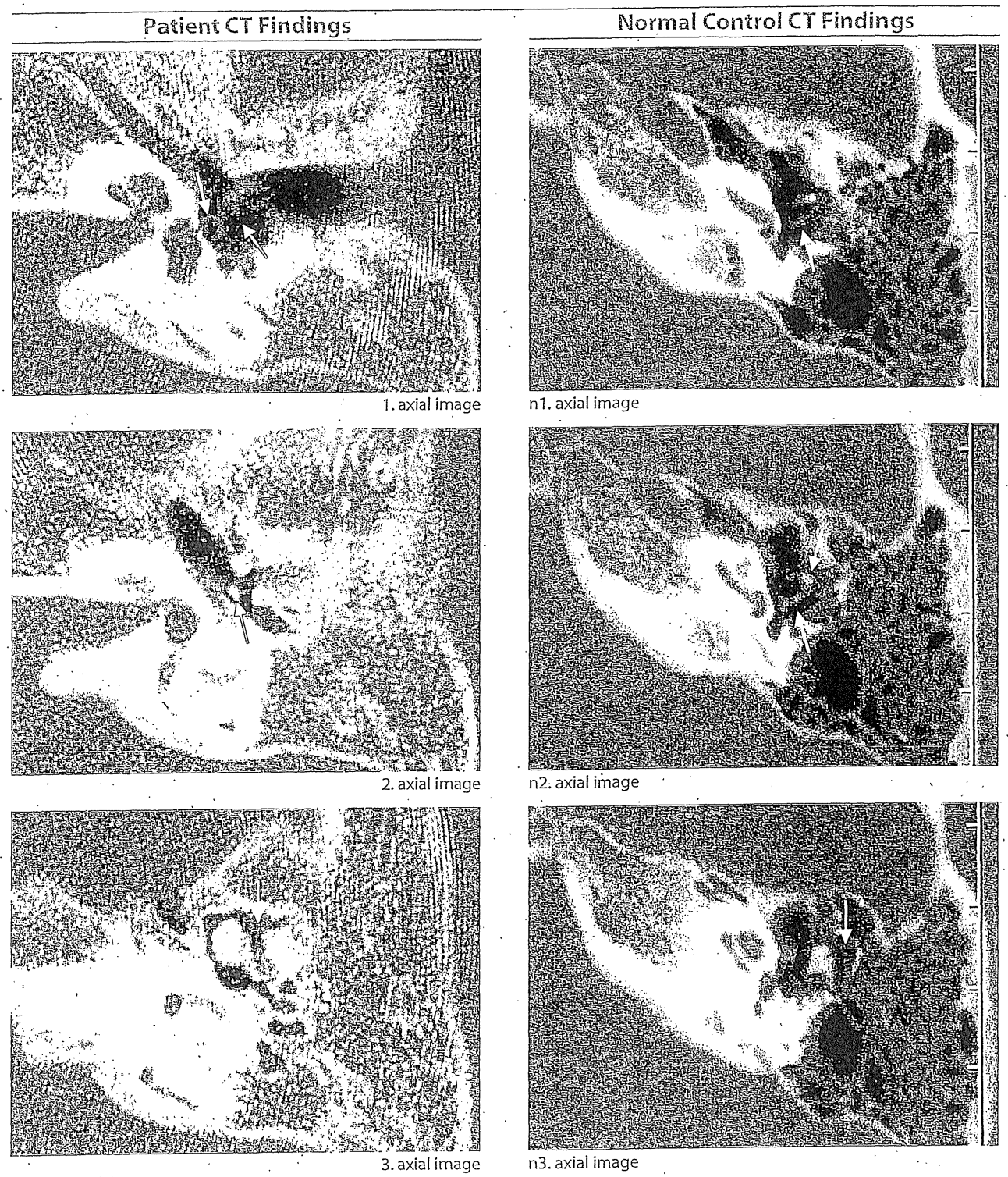


Fig. 10. (Case 2) Left ear CT: preoperative

[Patient CT Findings]

The vicinity around the tip of the long process of the incus is unclear and the lenticular process is unrecognizable (1:). This case does not include a cross-section in which the incudostapedial joint itself is displayed, and disruption is suspected. The superstructure of the stapes is slightly asymmetrical in form, but the head, anterior crus, and posterior crus of the stapes are all visible (1:). The cross-section of the long process of the incus is flatter than normal (2:), and the neck of the malleus contacts the anterolateral wall of the external auditory canal (2:), with suspected fixation. Also, the space lateral to the malleus head and the body of the incus, which normally can be observed as quite wide, is narrow in this case (3:). Fixation is probable here as well.

《Normal Control CT Findings》

n1: incudostapedial joint. n2: neck of malleus; long process of incus and head of stapes. n3: Wide space ( ) between the head of malleus/body of incus and the lateral attic wall.

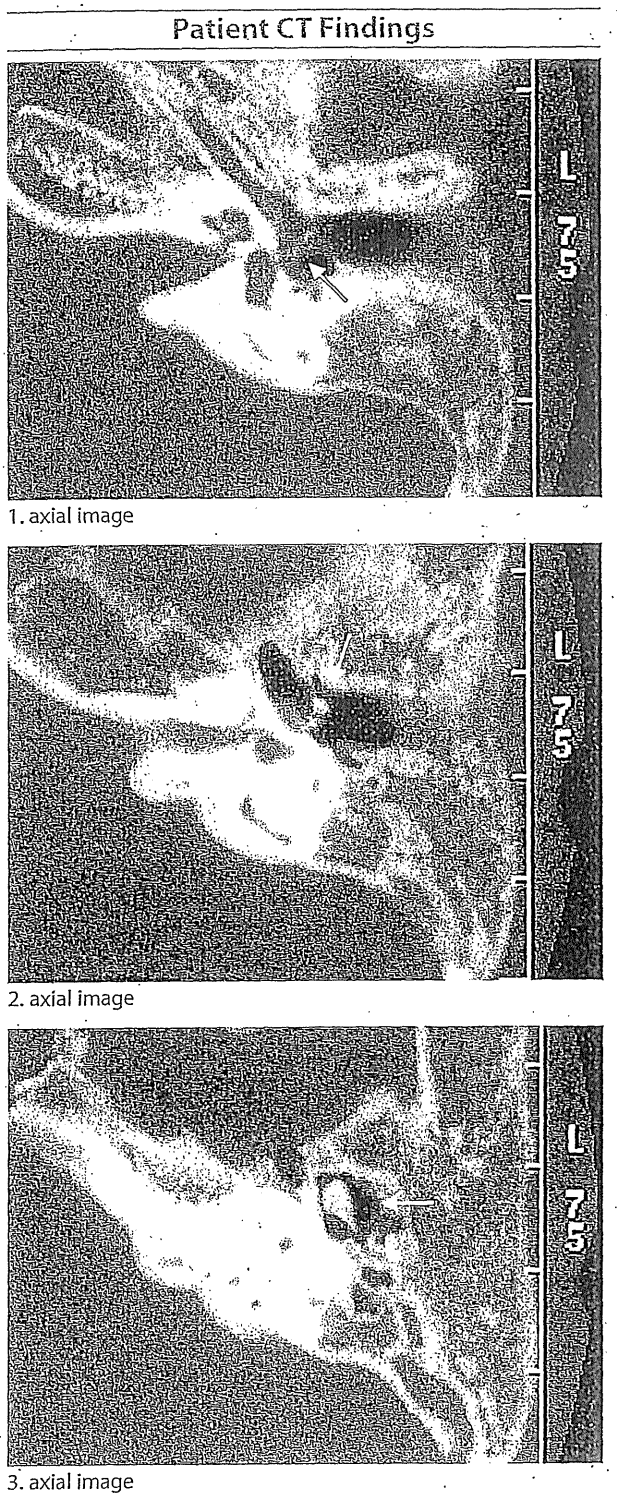


Fig. 11. (Case 2) Left ear CT: postoperative

[Patient CT Findings]

The incudostapedial joint is slightly enlarged and is connected via a tissue shadow with moderate density, which we may assume is the interpolating segment made from the sliver of auricular cartilage (1: ↖). A new space has been created anterolateral to the neck of the malleus (2: ↙) and lateral to the body of the incus (3: ←), due to the surgical removal of the superior wall of the bony segment of the external auditory canal and the lateral attic wall.

lenticular process was absent and contact with the head of the stapes was only via funicular soft tissue. The head, anterior crus, and posterior crus of the stapes were present, but the footplate was fixed, with no apparent mobility.

The funicular soft tissue connecting the long process of the incus to the head of the stapes was severed and the bone in the vicinity of the malleus and incus adhesions carefully removed, freeing up the malleus and incus to move normally. It was decided that, as the long process of the incus was malformed, it would be difficult to perform standard stapes surgery by fastening the hook of the teflon wire piston to the long process, so a Rosen probe was used to carefully mobilize the stapes so that it slanted in the facial canal/promontory direction. The fixation between the stapes footplate and surrounding bone was released, resulting in a substantial gain in stapes mobility. Finally, type III tympanoplasty was performed by inserting a sliver of auricular cartilage into the space between the head of the stapes and the stump of the long process of the incus.

Postoperative Patient CT Findings and Postoperative Course

On examination of the postoperative CT image (fig. 11), first we notice that the incudostapedial joint is slightly enlarged and is connected via a tissue shadow with moderate density, which we may assume is the interpolating segment made from the sliver of auricular cartilage (fig. 11:1). Also note that, compared to the preoperative image, a new space has been created anterolateral to the neck of the malleus (fig. 11:2) and lateral to the body of the incus (fig. 11:3), due to the surgical removal of the superior wall of the bony segment of the external auditory canal and the lateral attic wall.

Postoperatively, hearing level in the left ear averaged 25.0 dB and the air-bone gap had improved to 3.3 dB (fig. 12). Currently, four years after the surgery, there is no change. Only mobilization of the stapes footplate was performed and there was concern that it would re-fixate, but currently it is progressing favorably. If the stapes footplate should re-fixate, we plan to perform stapes surgery. Also, a canal wall down tympanoplasty with soft wall reconstruction was performed in this case, with no bony reconstruction of the deep part of the bony segment of the external auditory canal that had been removed. This prevents fixation of the ossicular chain to the surrounding bone and vibrates from the tympanic membrane to the continuous external auditory canal wall, which is thought to contribute to acquisition and maintenance of favorable postoperative hearing.

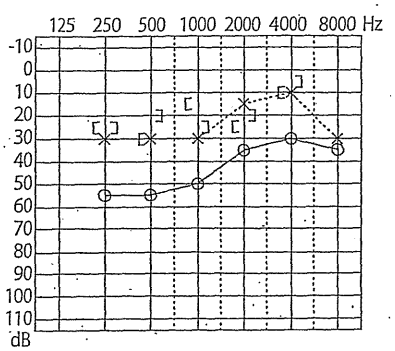


Fig. 12. (Case 2) Audiogram: postoperative

Case 3

Ossicular Deformities

Subject: female, 7 years old

History and Clinical Findings

Originally the subject was not aware of any abnormalities, nor had any been identified, but upon visiting ENT clinic due to a cold, an abnormality in her left tympanic membrane was identified and she was referred to our department. There were no abnormalities in the right ear. The left tympanic membrane showed no redness or retraction, but the handle of the malleus appeared slightly shorter and more vertical than normal. In pure tone audiometry the left ear had average air conduction hearing of 65.7 dB and bone conduction of 25.0 dB, with an air-bone gap of 40.7 dB (fig. 13a). The tympanogram was type As (fig. 13b), and no thickening or other qualitative abnormalities were observed in the tympanic membrane itself, so it was inferred that there must be some sort of mobility restriction in the ossicular chain. No stapedial reflex was present (fig. 13c). Basically it was conductive hearing loss, so an imaging examination was conducted to study the possibility of improving the subject's hearing through surgery.

Preoperative Patient CT Findings

Overall mastoid air cell development is satisfactory. There is no soft tissue density in the middle ear, negating the possibility of inflammatory disease. There is also no indication of anomalies in the inner ear or internal auditory canal. Observing the left ossicular chain, the malleus is slightly deformed and somewhat inferior to normal. There is no space where the anterior mallear ligament should normally be, and the neck of the malleus and the anterior tympanic spine are touching (fig. 14:1). In the normal control image, the handle of the malleus is pointing in the direction of the promontory and there is sufficient space between it and the anterior wall of the external auditory canal and other surrounding bone (fig. 14:n1, n2). There are no abnormalities in the stapes, but the tip of the long process of the incus cannot be clearly ascertained (fig. 14:2), nor can the joint between it and the stapes head (the incudostapedial joint) be confirmed. In the corresponding normal control image, the lenticular process of the incus is clearly depicted (fig. 14:n2). Viewing the cross-section of the central part of the incus, the head of the malleus (fig. 14:3) is depicted slightly smaller in this case than that of the normal control (fig. 14:n3). Also, as is often seen in cases of middle ear malformation, the space between the body of the incus and the lateral attic wall is narrow (fig. 14:3), in clear contrast to the normal control (fig. 14:n3).

In the coronal section as well, normally one can observe the corner at the tip of the long process of the incus where it bends at right angles from the lenticular process to reach the head of the stapes (fig. 15:n1), but in this case it cannot be identified (fig. 15:1). Also, the incus is adjacent to the lateral attic wall (fig. 15:1), and clearly lacks the usual space (fig. 15:n1).

Based on the above CT findings, we suspected fixation of the neck of the malleus and body of the incus to the surrounding bone, along with congenital disruption of the incudostapedial joint due to absence of the tip of the incus (lenticular process).

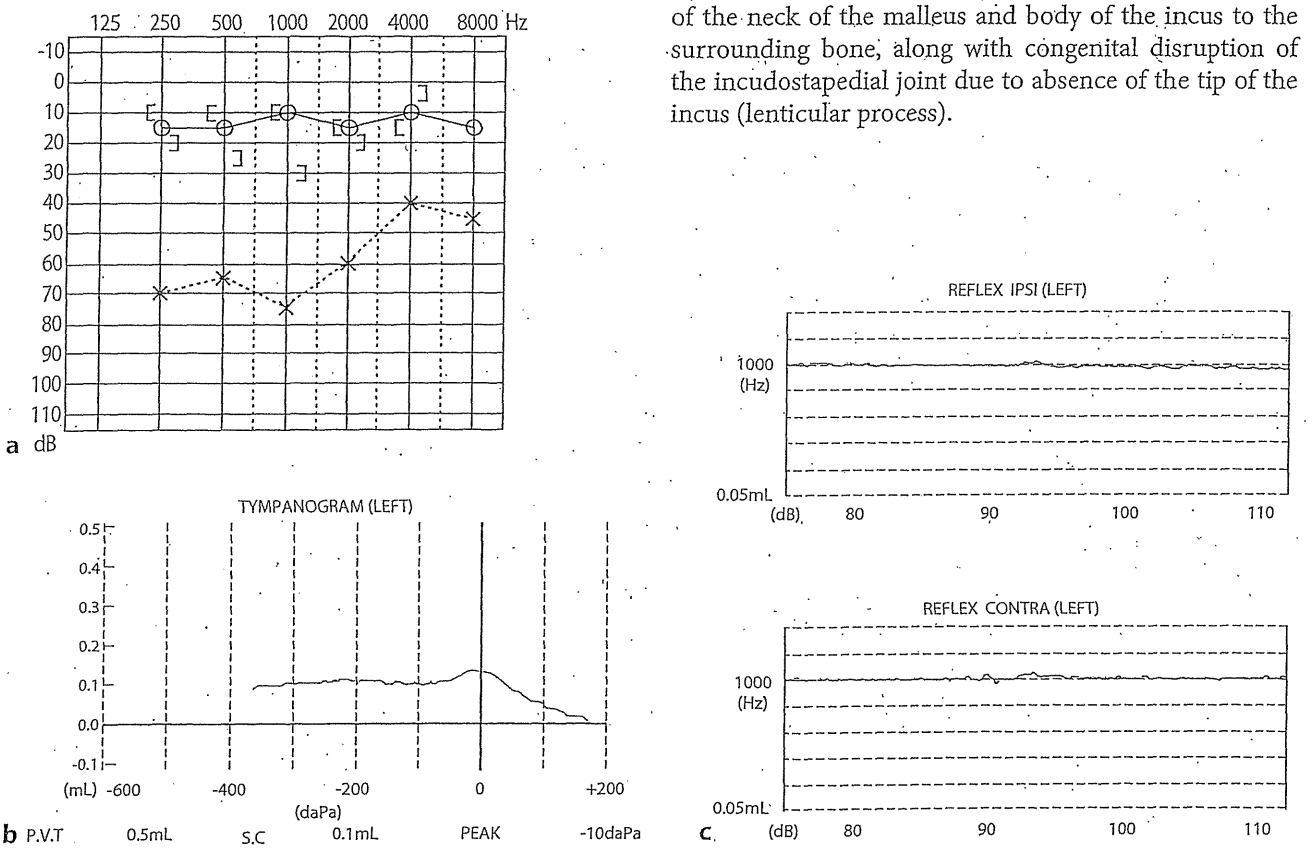


Fig. 13. (Case 3) Hearing test findings: preoperative. a= audiogram; b= tympanogram; c= left ear stapedial reflex

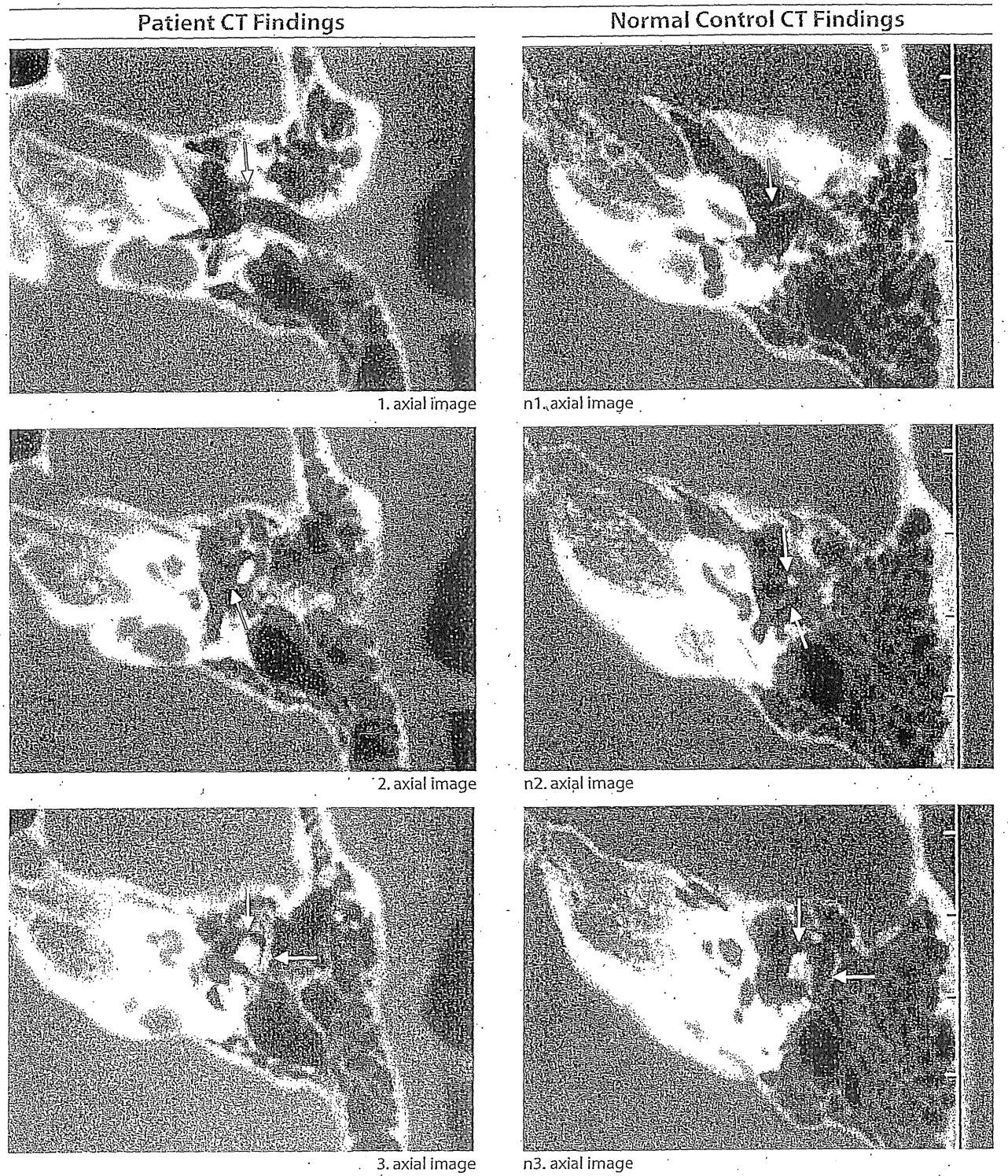


Fig. 14. (Case 3) Left ear CT: preoperative

[Patient CT Findings]

The malleus is slightly deformed and somewhat inferior to normal and the neck of the malleus and the anterior tympanic spine are touching (1: ↓). There are no abnormalities in the stapes, but the tip of the long process of the incus cannot be clearly ascertained (2: ↖); nor can the joint between it and the stapes head (the incudostapedial joint) be confirmed. Viewing the cross-section of the central part of the incus, the head of the malleus in this case (3: ↓) is depicted slightly smaller than that of the normal control (n3: ↓). Also, as is often seen in cases of middle ear malformation, the space between the body of the incus and the lateral wall of the attic is narrow (⇦).

《Normal Control CT Findings》

n1: ↓ handle of malleus. n2: ↓ neck of malleus; ↖ long process of incus and lenticular process. n3: ↓ head of malleus; ⇦ lateral attic wall.

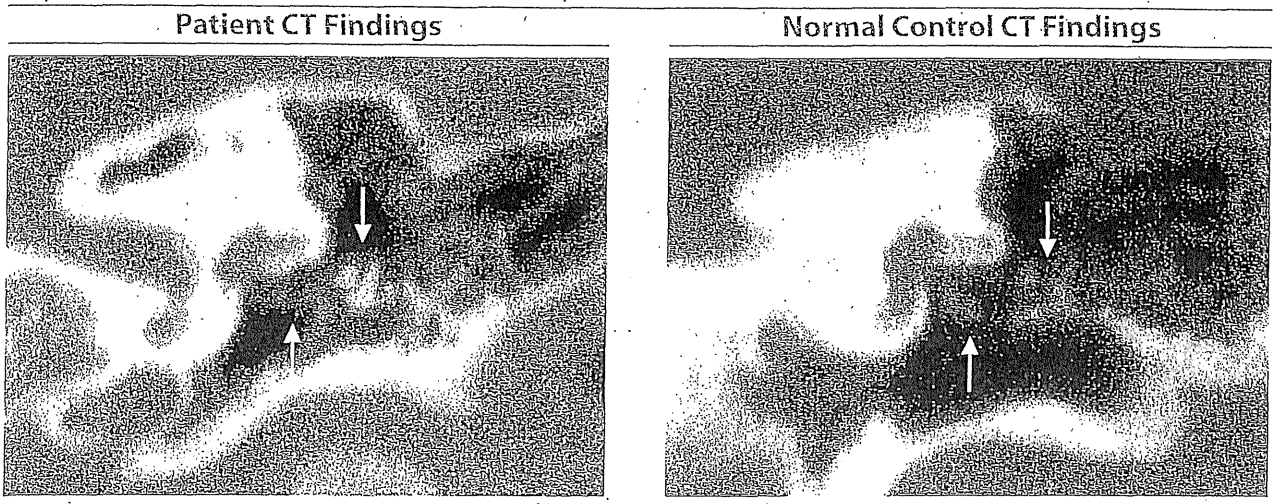


Fig. 15. (Case 3) Left ear CT: preoperative

[Patient CT Findings]
The corner at the tip of the long process of the incus where it bends at right angles from the lenticular process to reach the head of the stapes cannot be identified (1: ⇑). Also, the body of the incus is adjacent to the lateral attic wall (1: ⇓)

《Normal Control CT Findings》
n1: ⇑ area where the tip of the long process of the incus bends from the lenticular process to reach the head of the stapes; ⇓ space between the body of the incus and the lateral attic wall.

Surgical Findings

A left exploratory tympanotomy was performed. The neck and head of the malleus were malformed and fixed to the anterior tympanic spine and the lateral attic wall. The posterosuperior part of the bony segment of the external auditory canal was carefully removed to expose the incudostapedial joint, revealing that approx. ⅓ of the tip of the long process of the incus was extremely thin and curved gently to contact the head of the stapes. What appeared as a disruption in the CT image may have been due to the thinness of the tip of the long process of the incus and the lack of a bone mass corresponding to the lenticular process. Upon severing the incudostapedial joint, it was observed that the stapes displayed no abnormalities and its mobility was favorable. A portion of the lateral attic wall was removed and the incus that had been fixed to it extracted. It was also established that the head and handle of the malleus were separate in the vicinity of the neck, a finding that could not be confirmed in the preoperative CT image. The head of the malleus was severed and a type III ossiculoplasty was performed using the sculpted incus as a short columella to connect the stapes head and malleus handle.

Postoperative course was good, with left ear average hearing level of 23.3 dB eight months after surgery—a greater than 40 dB improvement over preoperative values—and the air-bone gap had virtually disappeared (fig. 16).

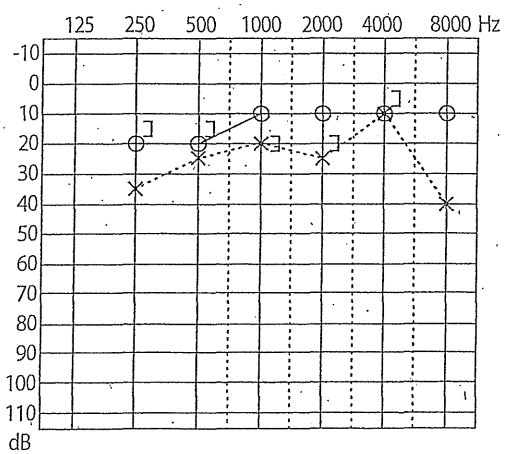


Fig. 16. (Case 3) Audiogram: 8 months postoperative

Case 4

Oval Window Atresia

Subject: male, 29 years old

History and Clinical Findings

The subject had suffered from bilateral hearing loss since infancy and came to us with the hope of improving hearing in his right ear. He had mixed hearing loss, with average hearing level of 78 dB in the right ear and 59 dB in the left ear.

Patient CT Findings

The CT image is of the right temporal bone. Both mastoid air cell development and pneumatization are satisfactory. Observing the oval window in the axial section, it is apparent that it is obstructed by bone roughly 1 mm thick, though the central portion is slightly thinner (fig. 17:1). The thickness of the oval window atresia in this case is readily apparent when compared to the image of a normal stapes footplate (fig. 17: n1). The same finding can be made in the coronal section. In all of the cross-sections, including the coronal image, the oval window is obstructed by bone (fig. 17:2). The head of the malleus and body of the incus are normal (fig. 17:1), but the long process of the incus is unclear and the superstructure of the stapes cannot be identified. There are no clear abnormalities in the shape of the bony labyrinth.

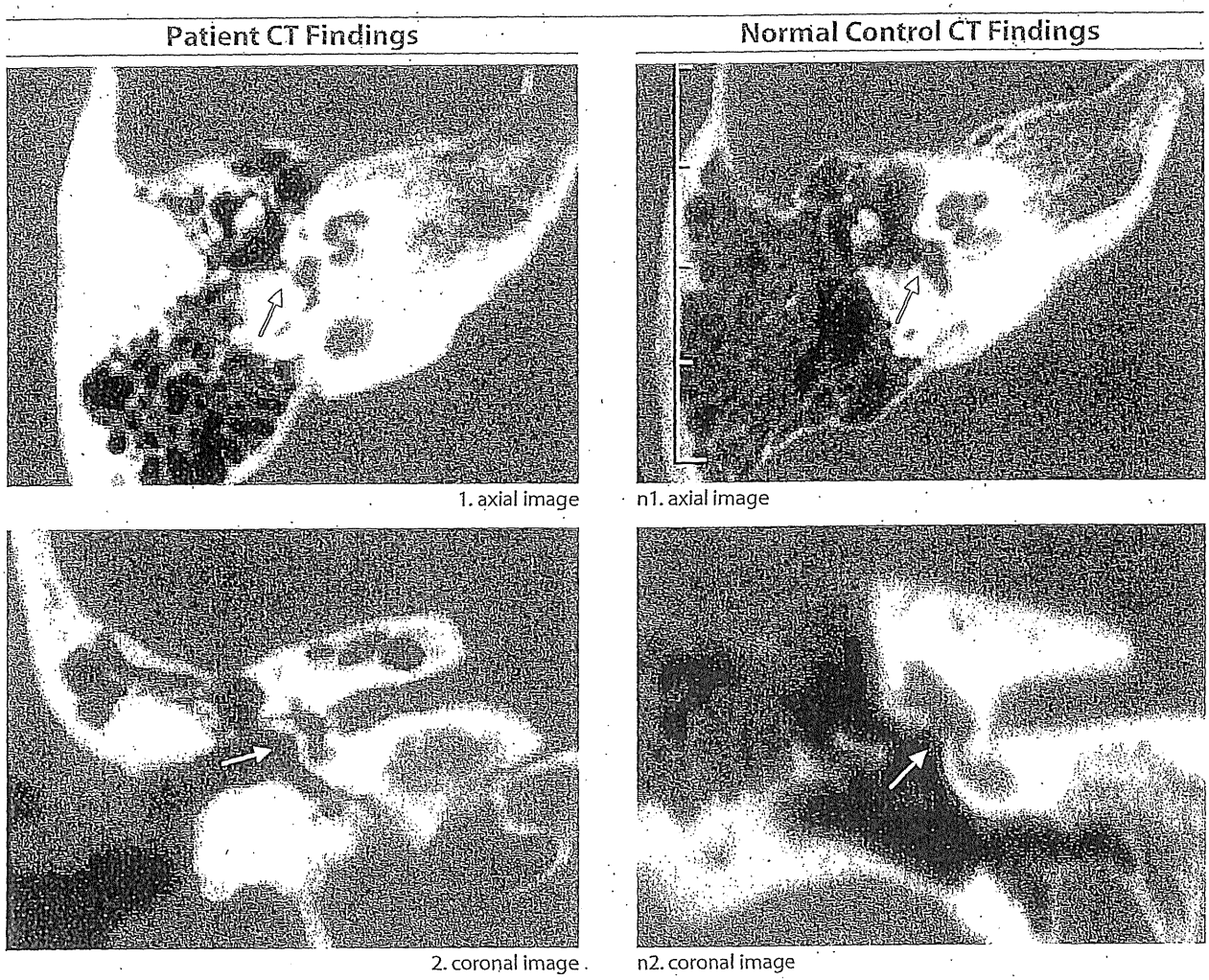


Fig. 17. (Case 4) Right ear CT: preoperative

[Patient CT Findings]
Both mastoid air cell development and pneumatization are satisfactory. The oval window is obstructed by bone roughly 1 mm thick, though the central portion is slightly thinner (1:). The same finding can be observed in the coronal section (2:). The head of the malleus and body of the incus are normal (1), but the long process of the incus is unclear and the superstructure of the stapes cannot be identified.

《Normal Control CT Findings》
n1: footplate of stapes. n2: oval window and footplate of stapes.

Surgical Findings

This was a difficult case with oval window atresia, but the possibility was pursued of improving hearing through an exploratory tympanotomy. A right exploratory tympanotomy was performed with prior explanation that stapes surgery may be carried out if circumstances warranted. During surgery it was observed that the stapes was missing and the oval window could not be identified. The tympanic segment of the facial nerve ran inferiorly to the area corresponding to the oval window. (In this case the resolution of the CT image was low, making it impossible to accurately confirm the path of the facial nerve.) The long process of the incus was malformed but present, so fenestration of the inner ear was carried out in the area corresponding to the oval window, a teflon wire piston inserted, and the wire attached to the long process of the incus. Hearing improved temporarily postoperatively, but later dropped to preoperative levels.

Points

1. Diagnosis of ossicular malformation requires pure tone audiometry, a stapedial reflex test, and a temporal bone CT exam.
2. In cases of hearing loss where mastoid air cell development and pneumatization are satisfactory, one should be mindful of the possibility of congenital malformation.
3. In cases of unilateral hearing loss in children when the subject is only partially aware of the hearing loss, there is a high probability that it is congenital.
4. The frequency of stapes footplate fixation is high, but confirmation through CT imaging is difficult.
5. In cases where either the neck of the malleus, head of the malleus, body of the incus, or short process of the incus are in extremely close proximity to the lateral attic wall, there is a possibility of fixation.
6. Ossicular chain disruption occurs frequently at the incudostapedial joint, making it important when examining the CT image to focus attention on whether the lenticular process of the incus and the head of the stapes are making contact.

Case 5 Skull Base Vascular Anomalies (CHARGE Syndrome)

Subject: male, 14 years old

History and Clinical Findings

The subject had responded poorly to sound since birth. He had other physical impairments, including partial choroid membrane deficiency (coloboma) of the eye, right nasal cavity stenosis (without choanal atresia), hypoplasia of the semicircular canals in the inner ear, right facial nerve paresis with dysphagia, underweight (growth hormone deficiency), middle ear malformation, patent ductus arteriosus, mild developmental retardation, and square face, which resulted in a diagnosis of CHARGE syndrome. The use of hearing aids was not effective. Due to the coexistence of multiple handicaps, the expected level of effectiveness from cochlear implants needed to be set lower than for hearing loss alone, but after consultation with the child's parents he underwent surgery at five years old, with the objective of achieving environmental sound recognition and partial recognition of spoken language.

In the child's preoperative CT exam, bilateral mastoid air cell development was found to be deficient and the middle ear space more or less limited to the tympanic cavity. Moreover, on the left side, the mastoid was occupied by a broadly meandering blood vessel. Existence of cranial nerve VIII was confirmed bilaterally via MRI. Because the left mastoid was occupied over a broad area by an aberrant blood vessel and there was a risk of macrovascular damage when preparing both the electrode insertion path and the well for embedding the receiver-stimulator unit, it was decided to perform the operation on the right side. Since, except for the sealed external auditory canal and the cochlear implantation area, the preoperative image is essentially the same as the postoperative image shown below, it is not shown here in order to avoid duplication.

Surgical Findings

The right mastoid air cells were completely undeveloped, so in order to insert the electrode via the external auditory canal a postauricular incision was made and all of the external auditory canal skin and tympanic membrane detached and extracted through the external auditory canal. Then a groove was created in the bone running posteriorly from the external auditory canal and a depression formed posterosuperiorly in the temporal bone to accommodate the receiver-stimulator unit. Transcanal confirmation of the round window niche was made, and fenestration of the cochlea's scala tympani performed anteriorly. The tip of the electrode array was fully inserted without problem, but the conducting wire at the base would not fit, so it was looped around once inside the external auditory canal. In order to prevent the cochlear implant's electrodes from being exposed to the external auditory canal, the cartilaginous segment of the external auditory canal was severed and sutured, and the inlet to the bony segment of the external auditory canal sealed with a bony plate.

Postoperatively a very gradual response to sound stimulation was obtained, and currently eight years later the average threshold with cochlear implant is 40 dB. The subject is unable to discern words through hearing alone,

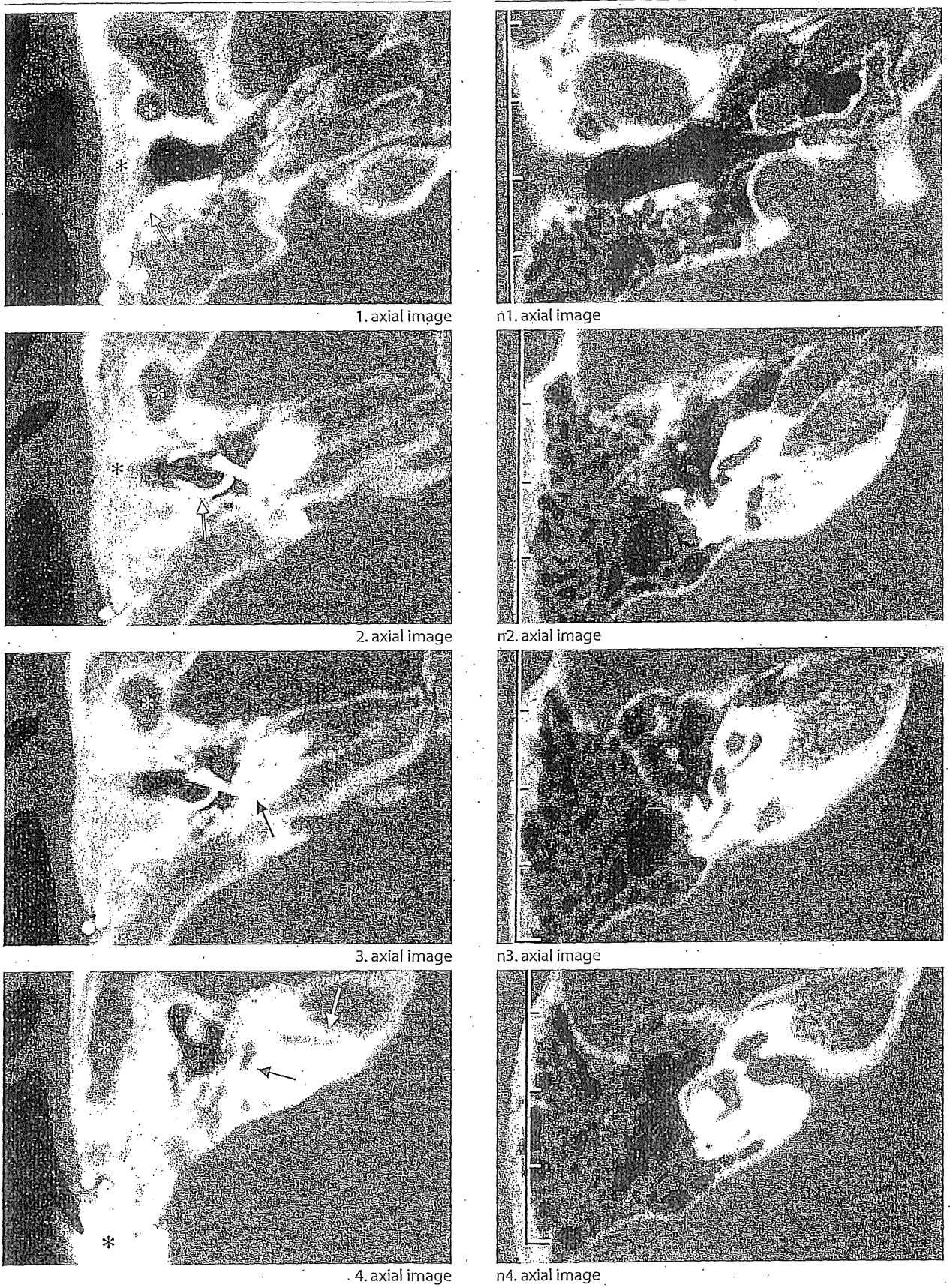


Fig. 18. (Case 5) Right ear CT: 9 years postoperative (age 14)

[Patient CT Findings]

The external auditory canal shows thick bony occlusion (1: *, 2: *). The conducting wire of the cochlear implant passes through the temporal bone (1: ↖) and forms a loop in the lumen of the external auditory canal (2: ↑) before entering the cochlea (3: ↖). In the vestibular system, the lateral semicircular canal is missing, forming only the vestibule (4: ←), and the internal auditory canal is also narrow (4: ↓). The receiver-stimulator unit (4: *) is appropriately embedded in the lateral part of the skull. The structures marked "❀" are the PGF in figure 18:1 and the PSS in figures 18:2–4 (refer to text for explanation)

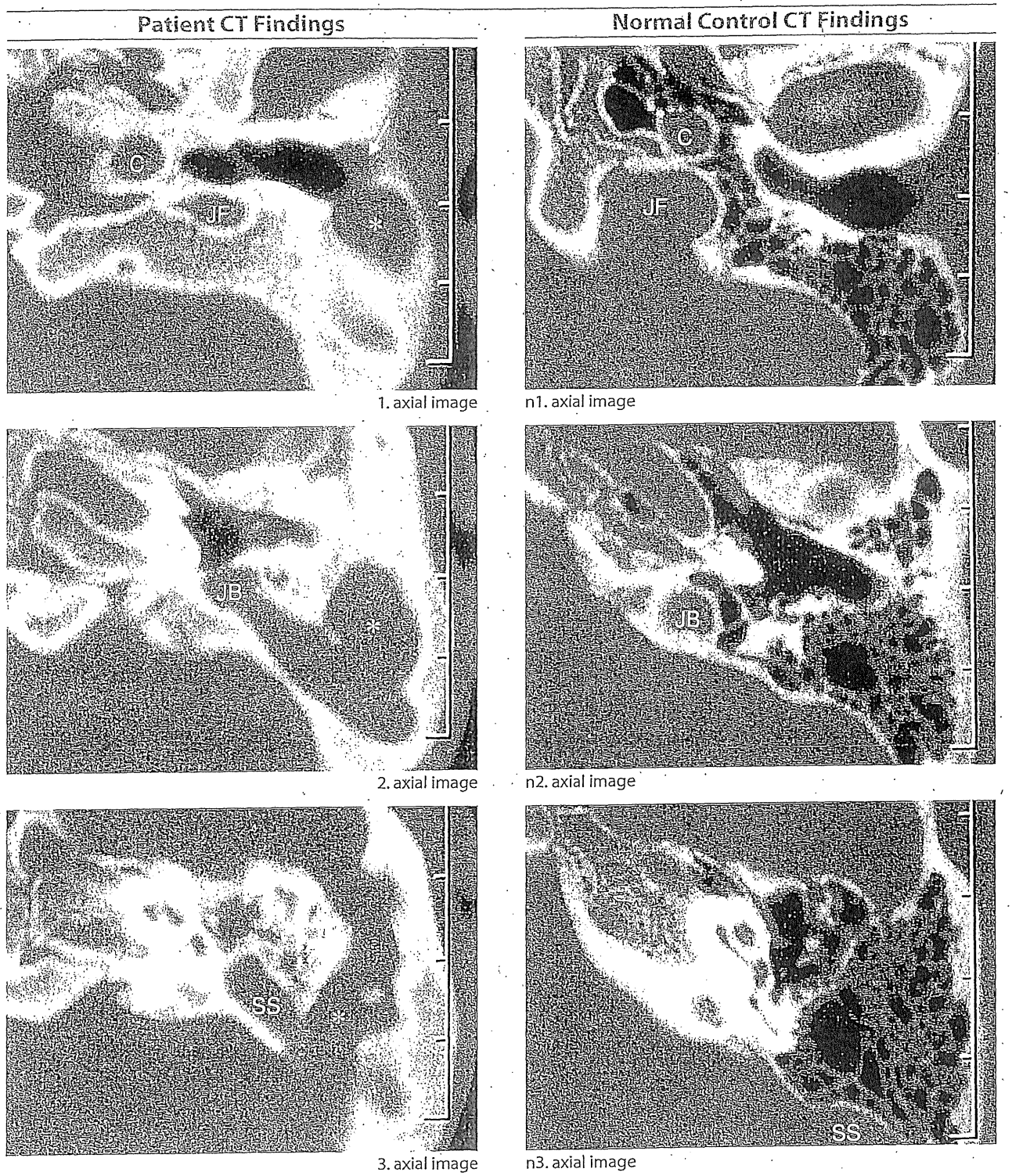


Fig. 19. (Case 5) Left ear CT: 9 years postoperative (age 14)

[Patient CT Findings]

On the left side, the PSS (❁) is enlarged, bifurcating from the sigmoid sinus (3: SS and ❁) and running anteriorly (2: ❁), exiting the temporal bone at the upper external auditory canal (1: ✍). The internal carotid artery is of normal size (1: C), but the jugular foramen (1: JF) is clearly smaller than normal. Also, the position of the jugular bulb (2: JB) is located more laterally than normal.

《Normal Control CT Findings》

n1: C=internal carotid artery; JF=jugular foramen.
n2: JB=jugular bulb. n3: SS=sigmoid sinus.

but in combination with lip reading has 100% vowel recognition and can perceive simple words. Hearing, while auxiliary, plays a firm role in the subject's everyday life.

Postoperative CT Findings

The CT image shows the child at 14 years old, nine years after surgery. Mastoid air cell development is deficient, but there is no soft tissue density in the tympanic cavity, nor is there effusion retention or cholesteatoma due to remnant skin. The external auditory canal shows thick, bony occlusion (fig. 18:1, 2). The conducting wire of the cochlear implant, which is external to the cochlea, passes through the temporal bone and along the external auditory canal, forming a loop in the lumen of the external auditory canal (fig. 18:2) before entering the cochlea (fig. 18:3). In the vestibular system, the lateral semicircular canal is missing, forming only the vestibule (fig. 18:4), and the internal auditory canal is also narrow (fig. 18:4). The receiver-stimulator unit (fig. 18:4) is appropriately embedded in the lateral part of the skull, with no excessive protrusion or intracranial invagination observable. In the area of the border between the lateral margin of the pyramid and the pars squamosa there is a band of soft tissue density about 5 mm wide (fig. 18:4) which, if followed anteriorly, exits the temporal bone posterior to the temporomandibular joint (fig. 18:1–3). This structure, marked "❄", which I will discuss later, is the petrosquamosal sinus (PSS) and its emissary the postglenoid foramen (PGF).

On the left side (fig. 19), the PSS is enlarged, bifurcating from the sigmoid sinus (fig. 19:3) and running anteriorly (fig. 19:2), exiting the temporal bone at the upper external auditory canal (fig. 19:1). The internal carotid artery is of normal size (fig. 19:1), but the jugular foramen (fig. 19:1) is clearly smaller than normal. Also, the position of the jugular bulb (fig. 19:2) is located more laterally than normal.

CHARGE Syndrome

Since CHARGE syndrome was first reported by Hall [1] in 1979, it has appeared in a comparatively large number of reports. It is thought to result from abnormal differentiation, setting, interaction, and migration of neural crest cells. A number of criteria for diagnosis of this disease have been proposed, but an updated diagnostic criteria recently proposed by Verloes [2] is as follows. He lists three major signs and five minor signs. The major signs are: 1) coloboma (iris or choroid, with or without microphthalmia); 2) atresia of choanae; 3) hypoplastic semicircular canals. The minor signs are: 1) rhombencephalic dysfunction (brainstem dysfunctions, cranial nerve VII to XII palsies and neurosensory deafness); 2) hypothalamo-hypophyseal dysfunction (including GH and gonadotrophin deficiencies); 3) abnormal middle or external ear; 4) malformation of mediastinal organs (heart, esophagus); 5) mental retardation. A patient with three major signs or two major signs and two or more minor signs is categorized as typical CHARGE syndrome; two major signs and one minor sign as partial/incomplete CHARGE syndrome; and fewer than that as atypical CHARGE syndrome. The case currently under consideration displays two major signs and all five minor signs, and so is classified as typical CHARGE syndrome.

Vascular Malformations of the Temporal Bone

Cerebral blood flow drains mainly through the internal jugular vein, but a portion drains through the external jugular vein. There are two routes that connect the two: one passes from the superficial and deep middle cerebral veins and through the cavernous sinus to the pterygoid plexus, and the other is via the emissary veins. Of the emissary veins, the one running from the sigmoid sinus to the mastoid foramen is observed frequently. However, the emissary route referred to as the petrosquamosal sinus (PSS), which runs from the anterior portion of the transverse sinus anteriorly along the petrosquamous suture and exits from the temporal bone through the postglenoid foramen (PGF) located anterior to the external auditory canal and posterior to the temporomandibular joint to arrive at the temporal fossa, ordinarily undergoes involution in the fetal or early postnatal stages, so it is rare to encounter it in a clinical setting [3]. The PGF can be observed in around 3.5% of humans, but in 40–50% of monkeys [4]. However, with advances in imaging diagnosis, the number of cases of skull base malformation in which the PSS can be confirmed is rising [5], and a case of CHARGE syndrome involving enlarged PSS has been reported [6]. The PSS normally either does not exist or is vestigial with no functional significance, however in cases of malformation such as the present one it has a large diameter and is believed to play an important role along with the internal jugular vein system in draining blood from the brain. This vascular malformation is difficult to diagnose due to its rarity, but because in otologic surgical procedures there is a danger it could cause hemorrhaging or other complications or, if obstructed, result in insufficient venous blood drainage from the brain, one should be mindful of this anomaly especially when treating hearing impaired children with syndromes.

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Points

1. When skull base malformation is involved, it sometimes is accompanied by vascular malformations such as sigmoid sinus anomalies or persistence or enlargement of the PSS.
2. PSS and PGF can be the cause of complications in otologic surgical procedures, so it is important to use imaging to make a full prior evaluation.

Facial Nerve Anomaly

Subject: male, 3 months old

History and Clinical Findings

There were no particular problems at birth, but the mother noticed that the child had trouble closing his right eye and that when he smiled the right corner of his mouth did not rise, so she had him examined by a local doctor who referred him to our department. Because the subject was an infant and could not yet intentionally make facial expressions, a detailed evaluation was difficult, but based on his expressions made while laughing or crying, he was diagnosed as grade 5 on the House-Brackmann scale.

Patient CT and MRI Findings

Mastoid air cell development is judged to be normal for this age on both sides. Observing the right facial nerve along its path from the internal auditory canal, the labyrinthine segment (fig. 20:4) displays no abnormalities, but from the geniculate ganglion (fig. 20:4) and tympanic segment to the second genu (fig. 20:3) and in the proximal portion of the mastoid segment (fig. 20:2), an enlargement of the neural canal is apparent compared to the normal control. The diameter of the facial nerve changes slightly in each segment, but the normal value for the diameter of the facial nerve distal to the tympanic segment is reported as 1.42 mm in neonates and an average of 1.87 mm in infants [1]. In this case, the diameter of the same segment of the facial nerve as measured on the image is 3.1 mm—roughly twice normal. On the other hand, no anomalies were found in the ossicles, including the incudostapedial joint and the superstructure of the stapes. Also, in the distal portion of the mastoid segment, the bony tube is bifurcated (fig. 20:1).

For this case, an MRI was also taken under general anesthesia using an intravenous anesthetic. In a contrast-enhanced MRI, the swollen facial nerve with contrast enhancement can be observed from the geniculate ganglion and tympanic segment to the mastoid segment (fig. 21:1). At the same time, a clear contrast enhancement can be seen in the middle ear cavity (fig. 21:1), showing strong otitis media.

Because symptoms of otitis media did not improve even with treatment using antibiotics and intravenous administration of steroids, a mastoidectomy was performed to directly observe and decompress the facial nerve.

Surgical Findings and Postoperative Course

A mastoidectomy and posterior tympanotomy were performed, at which point no ossicular abnormalities were discernible. When the incus was temporarily extracted to observe the tympanic segment of the facial nerve, it was discovered that the bony tube was absent from the geniculate ganglion to the second genu, exposing the tympanic segment of the facial nerve in the tympanic cavity. A decompression was performed from the mastoid segment to the stylomastoid foramen to expose the facial nerve, whereupon it was confirmed that the nerve was divided in two in the distal portion of the mastoid segment. Response of the facial muscles to intraoperative electrical stimulation was clear on stimulation of both branches of the nerve, in both the tympanic and the mastoid segments. When the nerve's epineurium and perineurium were incised from the tympanic segment to the mastoid segment (the area of pronounced neuronal swelling) to further decompress the nerve, swelling in the edematous nerve became more pronounced, but no tumor was visible. When electrical stimulation was applied directly to the nerve, it produced stronger facial muscle contraction than when applied through the perineurium. The incus was returned to its original position and a steroid-soaked gelatin sponge interned over the nerve to complete the operation.

Postoperative MRI and Clinical Course

Six months after facial nerve decompression surgery another contrast-enhanced MRI exam was performed. The otitis media observed preoperatively had healed and the contrast enhancement for the tympanic and mastoid segments of the facial nerve had abated (fig. 21:2).

Postoperatively, facial nerve function gradually improved to a grade 2 on the House-Brackman scale, as of two years after surgery.

Congenital Facial Nerve Anomalies

As mentioned in the section on EAC atresia and stenosis in this chapter, abnormal location of the facial nerve path often accompany external auditory canal anomalies. There are also reports of facial nerve bifurcation in cases of severe systemic deformity such as multiple anomalies [2, 3] and chromosomal abnormality [4]. A case such as this, however, in which there is a morphological defect to the facial nerve with no other defects beyond the ear, is extremely rare [5]. The facial nerve differentiates from the neural crest and otic vesicle in the fourth week of embryonic development and is complete after eight weeks, but the nerve's bony tube originates from the second branchial arch, with formation starting around the end of the fifth month. The distal portion of the mastoid segment in particular continues to extend peripherally after birth along with temporal bone development. The tip of the long process of the incus and the stapes also originate from the second branchial arch, and combined malformations of these structures and the facial nerve have been reported [6]. However, the present case is unique in that there were no anomalies of the ossicles. Initially, I concluded that the bifurcation of the distal region of the facial nerve's mastoid segment was a deformity. Certainly one does not see this kind of bifurcation of the facial nerve between the mastoid segment and the stylomastoid foramen in adults. But on examination of CT images in multiple cases of other young infants around one year old without facial paralysis or middle or external ear anomalies, it became clear that images showing bifurcation were not uncommon (fig. 20:n1), so perhaps this finding may be considered as within the normal range of the developmental process for the mastoid segment of the temporal bone.

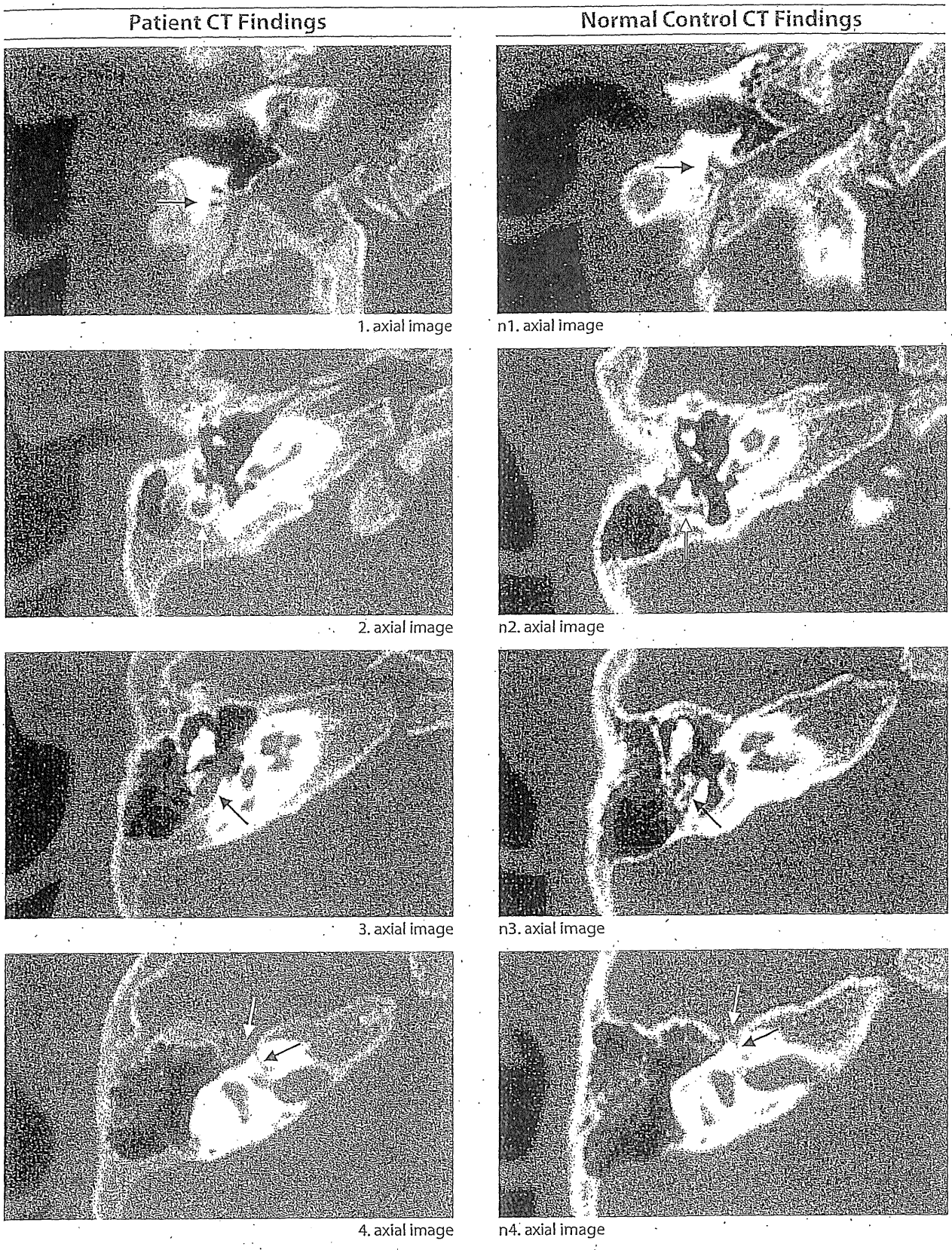

Fig. 20. (Case 6) Right ear CT: preoperative

[Patient CT Findings]

The labyrinthine segment of the facial nerve (4: ↙) displays no abnormalities, but from the geniculate ganglion (4: ↓) and tympanic segment to the second genu (3: ↖) and in the proximal portion of the mastoid segment (2: ↑) an enlargement of the neural canal is apparent. In the distal portion of the mastoid segment the bony tube bifurcates (1: →).

《Normal Control CT Findings》

n1: → The distal part of the facial nerve's mastoid segment is sometimes bifurcated in young infants, even in normal control samples. n2: ↑ proximal portion of mastoid segment. n3: ↖ second genu. n4: ↓ geniculate ganglion, ↙ labyrinthine segment.

Patient MRI Findings

1 2

Fig. 21. (Case 6) Right ear contrast-enhanced MRI: 1 = preoperative, 2 = six months after facial nerve decompression surgery

[Patient MRI Findings]

In a contrast-enhanced MRI, the swollen facial nerve with contrast enhancement can be observed from the geniculate ganglion and tympanic segment to the vicinity of the mastoid segment (1: ⇩). At the same time, a clear contrast enhancement can be seen in the middle ear cavity (1: ⇧), indicating otitis media. In another contrast-enhanced MRI exam performed six months after facial nerve decompression surgery, the otitis media observed preoperatively had healed (2: ⇧) and the contrast enhancement for the tympanic and mastoid segments of the facial nerve had abated (2: ⇩).

Facial paralysis accompanies facial nerve anomalies involving aplasia or extreme hypoplasia [3], but in cases reported by Takahashi et al [6], the only symptom was hearing loss due to attendant ossicular deformities, with no accompanying facial paralysis despite the existence of facial nerve path anomaly and bifurcation. In a report by Glastombury et al [5], all three cases were accompanied by congenital hearing loss, with two cases displaying facial paralysis since birth. However, whether or not the paralysis in these cases was congenital cannot be confirmed, as their histories included such factors as forceps delivery and physical trauma. In the present case, snapshots taken of the infant since birth showed no asymmetry, indicating that the paralysis was probably acquired. The fact that complications due to otitis media were present at the time the MRI was taken, the facial nerve's bony tube was absent in the tympanic segment and the nerve exposed in the middle ear, and the contrast enhancement reduced and nerve function recovered after decompression, it is possible that this case of facial paralysis was caused by neuritis that had spread to the nerve from otitis media, though because the previous CT image indicated no finding of otitis media we cannot be certain. In cases such as this, in which not just morphology but also the possibility of inflammation or tumor is being considered, both simple and contrast-enhanced MRI are required.

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Points

1. In infant facial paralysis there is a possibility of facial nerve anomalies.
2. In this case the facial nerve anomaly was isolated, but care must be taken to determine if there are combined malformations of the tip of the long process of the incus or the superstructure of the stapes, which originate from the second branchial arch.
3. A contrast-enhanced MRI is useful in diagnosing neuritis, but requires general anesthesia in young infants.

3 Inner Ear

Congenital Malformation of the Inner Ear

Congenital malformations of the inner ear can cause a variety of deficits in hearing and vestibular function. Recently congenital malformations are often discovered in newborn hearing screening tests, but in cases of mild hearing loss where the tests were not performed, deficiency symptoms may be unclear and remain unnoticed until the child is older. Also, one must be cautious as hearing loss that progresses after birth may not be picked up in neonatal screening. In cases of severe hearing loss, a doctor will eventually be consulted when the parents notice unresponsiveness to sound or delayed language development, or if abnormalities are identified during a regular physical exam. On the other hand, with congenital peripheral vestibular dysfunction, it is often difficult to determine whether symptoms such as delayed walking deviate from the normal range of individual developmental variations, making it more difficult to arrive at a definitive diagnosis than with hearing loss.

Malformations of the inner ear account for around 20% of congenital hearing loss [1], making it a major cause of disease. As with other causes, in cases of hearing loss due to inner ear malformation early diagnosis and intervention is important. Particularly in cases of severe to profound hearing loss with a high probability that a cochlear implant will be required, an accurate evaluation of the malformation is required not only for diagnosis and classification but also for surgical planning. Points to keep in mind when performing cochlear implantation in cases of inner ear malformation will be covered in detail in each of the cases in this chapter.

Genesis of the Inner Ear

Genesis of a normal inner ear begins around the third week of gestation with the formation of the otic placode, which forms a depression that becomes the otic pit, which in turn becomes the otic vesicle and separates around the fourth week. The otic vesicle divides into the masses that will form the basis for each component of the inner ear, which then enlarge and differentiate to form discrete shapes until the precise structure is complete. The bony labyrinth is basically formed first, with the formation of the membranous labyrinth following later.

Looking first at the cochlea, at around the fifth week of embryonic development the cochlear duct begins to extend from the saccule, reaching completion between the ninth and tenth weeks with approx. 2.5 turns. In the membranous labyrinth, the sensory cells arise in the organ of Corti starting at seven weeks and are complete at around 24 weeks, at which point the fetus is capable of hearing sounds. With the vestibular system, as the endolymphatic duct extends from the dorsal surface of the otic vesicle around the end of the fourth week, the eminences which will form the basis for the semicircular canals appear. In the seventh week, the vestibule specializes into the utricle and the saccule, and around the end of the eighth week the mesenchymal tissue of first the anterior and posterior semicircular canals, then the lateral semicircular canal, is absorbed and they become tubular [2].

After nine weeks, the mesenchymal tissue around the otic vesicle becomes cartilage and after ten weeks the cartilage around the membranous labyrinth is absorbed and replaced with fluid and the perilymphatic space is formed. From around 15 weeks the cartilage around the labyrinth begins to ossify, and this process is completed in the 23rd week, with overall completion of the inner ear at around 26 weeks.

Histopathological Classification of Inner Ear Malformation

Histopathologically, inner ear malformation may be classified [3] as 1) Michel aplasia: the inner ears is completely failure of development of the inner ear; 2) Mondini dysplasia: the cochlea is flat, the cochlear duct is short, the modiolus is hypoplastic, the auditory and vestibular sense organs and nerves are immature, the vestibule is large, the semicircular canals are wide, small, or missing, and the endolymphatic sac is bulbous; 3) Bing-Siebenmann dysplasia: normal bony labyrinth with underdeveloped membranous labyrinth; 4) Scheibe dysplasia: disorder of the membranous labyrinth of the saccule and cochlea, with normal development of the utricle and semicircular canals and 5) Alexander dysplasia: aplasia of the cochlear membranous labyrinth, especially when accompanied by high-frequency hearing loss due to abnormalities in the basal turn [4]. This classification system is based on findings of microscopic observations on histopathological temporal bone sections, however diagnosis of inner ear malformation in everyday clinical settings is performed using temporal bone CT and MRI examination so it is not possible to use the above classification as is. Current CT and MRI spatial resolution and contrast resolution permits observation of the modiolus and the septum between the cochlear turns, but because the membranous labyrinth and sensory cells are not depicted, in clinical imaging examination it is not possible to distinguish between 3) to 5) above, namely Bing-Siebenmann, Scheibe, and Alexander dysplasia. This classification system also makes no reference to inner ear malformations in which the cochlea and vestibule are fused in an undifferentiated cyst (explained later as common cavity deformity). Moreover, 1) Michel aplasia is rare, so as a result the majority of inner ear malformations encountered everyday are 2) Mondini dysplasia, rendering this classification system meaningless. Given this state of affairs it is apparent that, in addition to histopathological classification, a classification system of inner ear malformations based on clinical imaging findings is also required.

Classification Based on Clinical Imaging

The Jackler-Luxford-House classification system (table 3) [1] indicates the basic thinking behind classification of inner ear malformations based on clinical imaging findings, and up to now has served as the standard in

this field. Category II, relating to the bony labyrinth and membranous labyrinth, is epoch-making in that it systematically classifies inner ear malformations based on findings for the cochlea, vestibule, vestibular aqueduct, and cochlear canaliculus that can be observed by imaging, allowing the inner ear malformation classification system previously centered on histopathological classification to be applied clinically. Furthermore, based on the Jackler classification system, Sennaroglu and Saatci have introduced changes to create a more practical system overall (table 4) [5], particularly by clearly distinguishing between Mondini dysplasia (incomplete partition type II) and a similar but embryologically more immature malformation (incomplete partition type I). Henceforth we will follow the Sennaroglu and Saatci classification when describing inner ear malformations from a clinical imaging perspective.

Classification of inner ear malformation, particularly the severity of cochlea malformation, using temporal bone CT imaging is shown in table 4-(1). The earlier the arrested development that causes the malformation occurs, the more advanced the malformation.

The most serious malformation is Michel aplasia, in which the inner ear is unformed, but this is due to an anomaly around the third week of embryonic development and as previously mentioned is rare. The next most serious malformation is cochlear aplasia, which is thought to result from problems occurring slightly after the third week.

Next, due to an anomaly occurring around the fourth week of embryonic development, the primodium of both the cochlea and vestibule is formed, but thereafter does not differentiate so the inner ear becomes a single, cystic structure. This is referred to as common cavity deformity. With common cavity deformity, there is a wide variation in the overall size of the cavity and the relative sizes of the areas corresponding to the cochlea and the vestibule.

In the next category, incomplete partition type I (IP-I), the vestibule, semicircular canals, and cochlea are clearly separated and formed to some extent but, even though the contours of the basal and upper turns of the cochlea are present, the septa separating cochlear turns and the modiolus are not discernible on the CT image. IP-I is thought to occur due to an anomaly in the fifth week of embryonic development. If inner ear development is impeded in the sixth week, the inner ear is divided into cochlea and vestibule, but their development is arrested before reaching maturity and the turns of the cochlea are slightly undersized and the vestibule and semicircular canals are either aplastic or hypoplastic, resulting in what is referred to as cochleovestibular hypoplasia.

Table 3. Jackler-Luxford-House classification of congenital malformations of the inner ear

I. Malformations limited to the membranous labyrinth
A. Complete membranous labyrinthine dysplasia
B. Limited membranous labyrinthine dysplasia
i. Cochleosaccular dysplasia (Scheibe)
ii. Cochlear basal turn dysplasia (Alexander)
II. Malformations of the osseous and membranous labyrinths
A. Complete labyrinthine aplasia (Michel)
B. Cochlear anomalies
i. Cochlear aplasia
ii. Cochlear hypoplasia
iii. Incomplete partition (Mondini)
iv. Common cavity
C. Labyrinthine anomalies
i. Semicircular canal dysplasia
ii. Semicircular canal aplasia
D. Aqueductal anomalies
i. Enlargement of the vestibular aqueduct
ii. Enlargement of the cochlear aqueduct
E. Abnormalities of the internal auditory canal
i. Narrow internal auditory canal
ii. Wide internal auditory canal

From Jackler, Luxford and House [1]

Table 4. Sennaroglu-Saatci classification of congenital malformation of the inner ear

Cochlear malformations
Cochlear malformations include the following: 1) Michel deformity. There is complete absence of all cochlear and vestibular structures. 2) Cochlear aplasia. The cochlea is completely absent. 3) Common cavity deformity. There is a cystic cavity representing the cochlea and vestibule, but without showing any differentiation into cochlea and vestibule. 4) Cochlear hypoplasia. Malformation is further differentiated so that the cochlea and vestibule are separate from each other but their dimensions are smaller than normal. Hypoplastic cochlea resembles a small bud off the internal auditory canal (IAC). 5) Incomplete partition type I (IP-I). The cochlea is lacking the entire modiolus and cribriform area, resulting in a cystic appearance. This is accompanied by a large cystic vestibule. 6) Incomplete partition type II (IP-II) (Mondini deformity). The cochlea consists of 1.5 turns, in which the middle and apical turns coalesce to form a cystic apex, accompanied by a dilated vestibule and enlarged VA.
Vestibular malformations
Vestibular malformations include Michel deformity, common cavity, absent vestibule, hypoplastic vestibule, and dilated vestibule.
Semicircular canal malformations
Semicircular canal malformations are described as absent, hypoplastic, or enlarged.
Internal auditory canal malformations
Internal auditory canal malformations are described as absent, narrow, or enlarged.
Vestibular and cochlear aqueduct findings
Vestibular and cochlear aqueduct abnormalities are described as enlarged or normal.

From Sennaroglu and Saatci [5]