Finally, incomplete partition type II (IP-II), which arises in the seventh week of embryonic development, is the mildest type of malformation in this classification system. In this malformation, 1.5 or more turns of the cochlea are formed and the septum between the basal and upper turns is clearly visible, but the part above the basal turn is cystic. The modiolus can be ascertained at the basal turn. On the other hand, in the vestibular system, malformation of the semicircular canals is insignificant and there is only slight enlargement of the vestibule, but it is accompanied by enlargement of the vestibular aqueduct. These findings are in accordance with findings from cases of inner ear malformation reported by Carlo Mondini in 1791 [6]. Mondini dysplasia is the most well-known inner ear malformation, but because this term has not necessarily been used according to its strict definition, here it tends to be understood as a blanket term for various different deformities. In Sennaroglu's research [5], he clearly defines the mildest category of malformation and accurately applies this definition to reported cases of Mondini dysplasia. In this sense as well, the Sennaroglu classification system is highly significant.

A note of caution, however: when viewing inner ear malformations in their entirety, there are many cases that are difficult to explain solely through the mechanism of arrested development during a specific period in the embryonic process. For example, there are cases in which the vestibule and semicircular canals are completely unformed, even though the cochlea has formed 1.5 turns, or in which only the vestibular aqueduct is enlarged, even though CT imaging indicates no anomalies whatsoever in the other components of the labyrinth. These exceptions imply the existence of some other internal factor involving, for example, a genetic mutation that selectively affects a specific part of the inner ear. In other words, there exist malformations in which multiple components of the inner ear are simultaneously damaged during the embryonic process (Sennaroglu calls these multi-branch abnormalities), those in which abnormalities occur independently in specific components (single-branch abnormalities), and also those in which the two coincide. Consequently, strictly speaking, one cannot obtain a complete evaluation of inner ear malformation unless one divides the inner ear into its various components and describes in further detail the degree of malformation in each, as is done in the Jackler classification system (table 3-II). However, such a strict approach is excessive for the majority of inner ear malformations. Since most cases conform to one of the categories shown in table 4-(1), the most practical approach in clinical terms is first to determine roughly which of the stages between Michel aplasia and IP-II applies, then if this is insufficient, append more detailed findings and, in cases that do not fall within the framework of systematic anomalies, individually record the anomalies for each part of the inner ear.

Complete formation of the inner ear follows a process of development, growth, and ossification. In malformations of the inner ear it is important first to determine whether or not development has taken place, then the extent of growth, as increase in size also affects final inner ear morphology. In the inner ear of a normal adult, the outer diameter of the basal turn of the cochlea is approx. 7 mm, the diameter of a cross-section of the basal turn just under 2 mm, and the height of the modiolus approx. 5 mm. The size of a normal vestibule is 4–5 mm anteroposteriorly and approximately 6 mm mediolaterally, and the outer diameter of the semicircular canals is 6–7 mm [7]. The diameter of the internal auditory canal is approx. 5 mm anteroposteriorly and approx. 4.5 mm vertically, with a length of approx. 11 mm [8]. In most cases involving malformations of the inner ear, categorization is determined by irregularities to the shape itself, but in determining, for example, the size of the cavity in common cavity deformity, the existence of vestibule enlargement or lateral semicircular canal hypoplasty for IP-II malformation, or the presence of stenosis for internal auditory canal anomalies, measurement values for each component are useful for attaining a more detailed understanding of characteristics with each category.

Role of CT and MRI in Diagnosis of Inner Ear Anomalies

Structurally the inner ear is composed of a membranous labyrinth inside a bony labyrinth, but the structures that actually sense sound and acceleration are the hair cells of the membranous labyrinth. Consequently the membranous labyrinth is important in functional terms, and it would be ideal if it were depicted in medical images. However, as stated previously, there is a limit to spacial resolution with current MRI performance, so it is impossible to differentiate the content of the bony labyrinth so long as there is no fibrosis. Also, in actual clinical practice, imaging of the temporal bone is often carried out to diagnose hearing loss, and occasionally also to check for malformations of the middle ear, and this requires CT imaging. For depicting the overall morphology of the inner ear both CT and MRI are roughly equal, but whereas bony tissues are not shown in MR images, CT also permits observation of malformations of the auditory ossicles, making CT the superior diagnostic tool. CT is the appropriate first choice in imaging for hearing impaired patients in which congenital malformation is suspected.

However recent years have seen a dramatic increase in cochlear implantations, making it important in cases of inner ear malformation not only simply to record the morphological classification, but also to evaluate from a functional perspective regarding the propriety and estimated prognosis of a cochlear implantation. For this reason, an MRI is necessary to evaluate the condition of structures other than the bones of the inner ear, including cranial nerve VIII, the cochlear nerve, and if possible, inside the modiolus. In this section we present cases of varying degrees of inner ear malformation and examine them using temporal bone CT images in combination with soft tissue findings obtained from MR images, to convey the procedure and focal points of comprehensive imaging diagnosis.

References

1 Jackler RK, Luxford WM, House WF: Congenital malformation of the inner ear. Laryngoscope 1987; 97 (suppl 40):2–14.

2 Swartz JD, Mukherji SK: Chapter 5 The inner ear and otodystrophy. 4th edition. Imaging of the temporal bone. Thieme, New York, 2009.

3 Schuknecht HF: Pathology of the ear. Second edition. Lea and Febiger, Philadelphia, 1993.

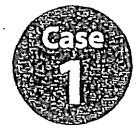
4 Ormerod FC: The pathology of congenital deafness. J Lryngol Otol 1960;74:919–950.

5 Sennaroglu L, Saatci I: A new classification for cochleovestibular malformations. Laryngoscope 2002;112:2230–2241.

6 Mondini C: Anatomical surdi nedi sectio. De Bononiensi Scientiarum et Artium Instituto Arque Acadamia Commentarii, Borogna 1791; 7:419 (quoted from reference 4).

7 Lang J: Anatomy of the brainstem and the lower cranial nerves, vessels, and surrounding structures. Am J Otol Supplement 1985;1–19.

8 McClay JE, Tandy RT, Grundfast K, et al: Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. Arch Otolaryngol Head Neck Surg 2002; 128:664–671.



Michel Aplasia (Inner Ear Aplasia)

Subject: male, 2 years old

History and Clinical Findings

The subject was born at 40 weeks gestation. No particular abnormalities were detected in the perinatal period. A pediatric exam one month after birth identified deficient reaction to sound and an ABR was carried out at three months, at which time it was confirmed that there was no response to 105 dB NHL stimulation in either ear. Pediatrically, aside from mild motor retardation, there were no other obvious problems. A hearing aid was fitted at a rehabilitation center, but with no effect, so the subject was referred to our department to determine whether a cochlear implant would be appropriate. There were no abnormalities of the auricle, external auditory canal, or tympanic membrane. There was nothing of note in the family history.

Patient CT Findings

The cochlea and vestibule are completely unobservable (fig. 22:1–4, fig. 24:1–3). The high-density bone mass of the labyrinth visible in the normal control images (fig. 22:n2–n4, fig. 24:n1–n3) are barely visible in this case in the vicinity of the internal auditory canal (fig. 22:2–4) and the superior part of the anterior semicircular canal (fig. 23, fig. 24:4). Normally, the structures of the inner ear and internal auditory canal are located between the middle ear and the intracranial space, but in this case as one moves medially in from the middle ear, one passes through a single bony wall before directly entering the intracranial space (fig. 24:3).

The internal auditory canal is located more inferiorly than normal (fig. 22:2&3, fig. 24:2) and, following its contents, one finds that it turns into the geniculate ganglion (fig. 22:2, fig. 24:1), then, after running slightly posteriorly (fig. 22:1), turns inferiorly and becomes the stylomastoid foramen (fig. 22:1, fig. 24:3). Also, it can be confirmed on the MR image that the space inside the internal auditory canal contains only the facial nerve (fig. 25:1, indicated by the numeral 7). The overall path of the facial nerve and position of the stylomastoid foramen is significantly anterior to normal (fig. 22:n1). The tympanic segment (fig. 24:2) is also more inferior than normal (fig. 24:n2). Cranial nerve VIII (cochlear and vestibular nerves) is completely unobservable.

Observing the structures of the middle ear, among the ossicles the malleus and incus show no defects, however the stapes is almost nonexistent, with merely a portion of the head of the stapes present.

A small portion of the arch of the upper extremity of the anterior semicircular canal is present (fig. 23:2&3, fig. 24:4), so it does not represent complete aplasia. Therefore, this case could be described as "cochlear aplasia, vestibular aplasia, lateral and posterior semicircular canal aplasia, anterior semicircular canal hypoplasia." However, aside from the vestigial presence of a portion of the anterior semicircular canal, the majority of the inner ear is aplastic and displays many of the characteristic image findings of inner ear aplasia, so it has been presented here

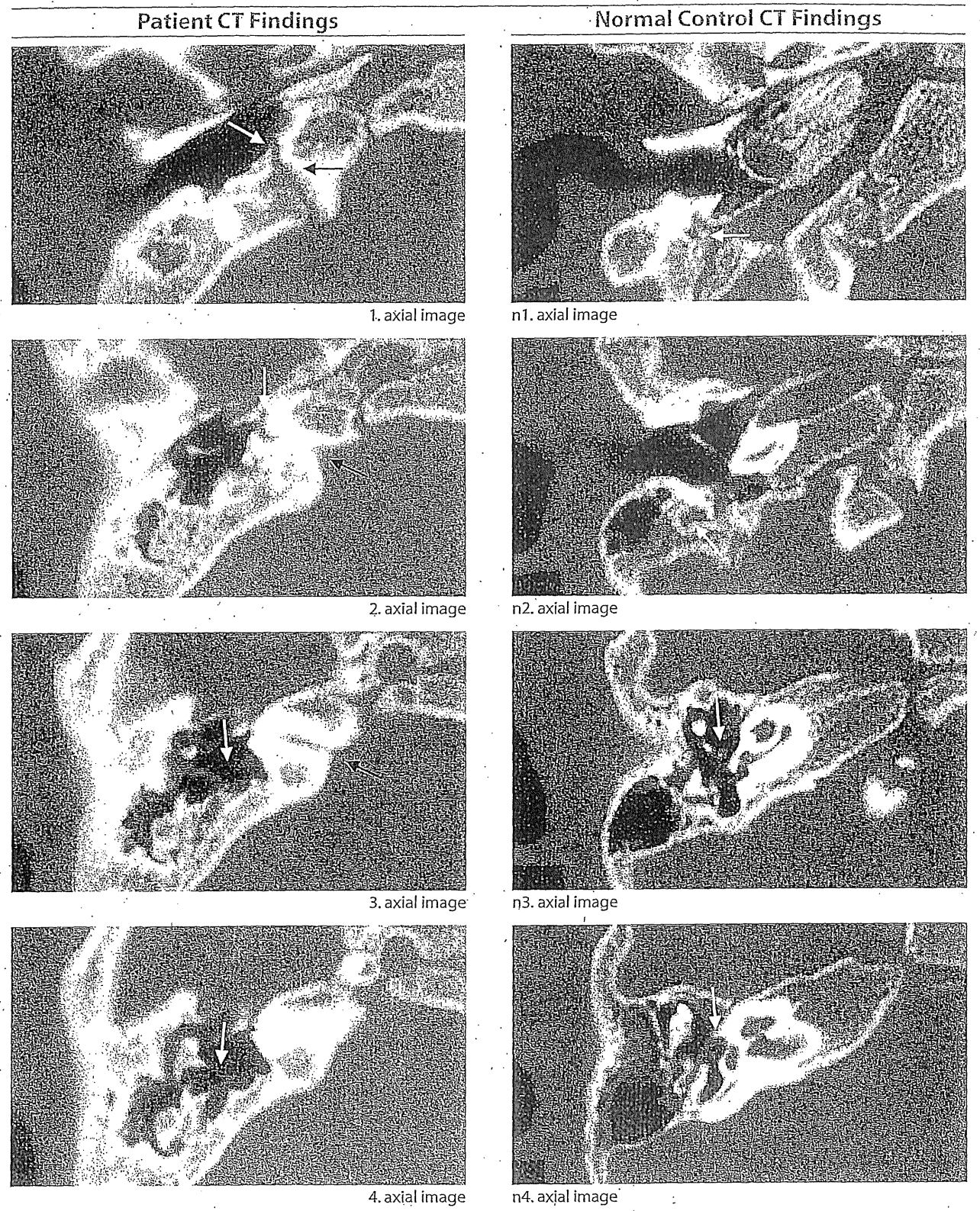


Fig. 22. (Case 1) CT

[Patient CT Findings]

The cochlea and vestibule are completely unobservable (1–4). The high-density bone mass of the labyrinth visible in the normal control images (n2–n4) is barely visible in this case in the vicinity of the internal auditory canal (2–4) and the superior part of the anterior semicircular canal (Fig. 23). The internal auditory canal is located more inferiorly than normal (2, 3: ↖) and, following its contents, one finds that it turns into the geniculate ganglion (2: ↓), then, after running slightly posteriorly (1: ↘), turns inferiorly and becomes the stylomastoid foramen (1: ←). The overall path of the facial nerve and position of the stylomastoid foramen are significantly anterior to normal (n1: ◇). Observing the structures of the middle ear, among the ossicles the malleus and incus show no defects, however the stapes is almost nonexistent (4: ↓), with merely a portion of the head of the stapes present (3: ·)

《Normal Control CT Findings》

n1: ← stylomastoid foramen. n2: ↖ mastoid segment of facial nerve. n3, n4: ↓ tympanic segment of the facial nerve and stapes.

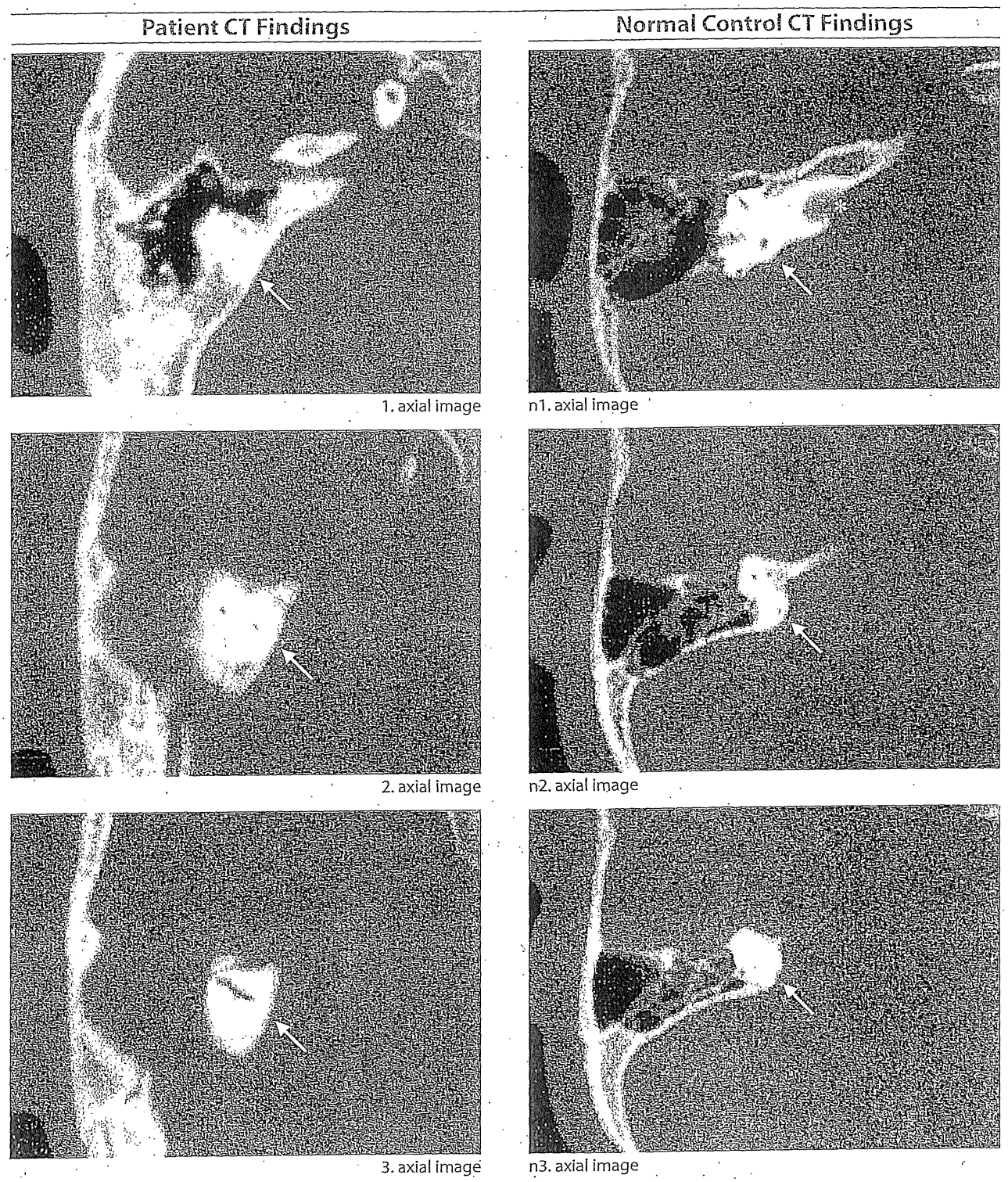


Fig. 23. (Case 1) CT

[Patient CT Findings]

In this case, the area below the anterior semicircular canal's base is not formed (1: ✎), but a small portion of the arch of its upper extremity is present (2, 3: ✎).

《Normal Control CT Findings》

n1–n3: ✎ anterior semicircular canal; ❁ internal auditory canal.

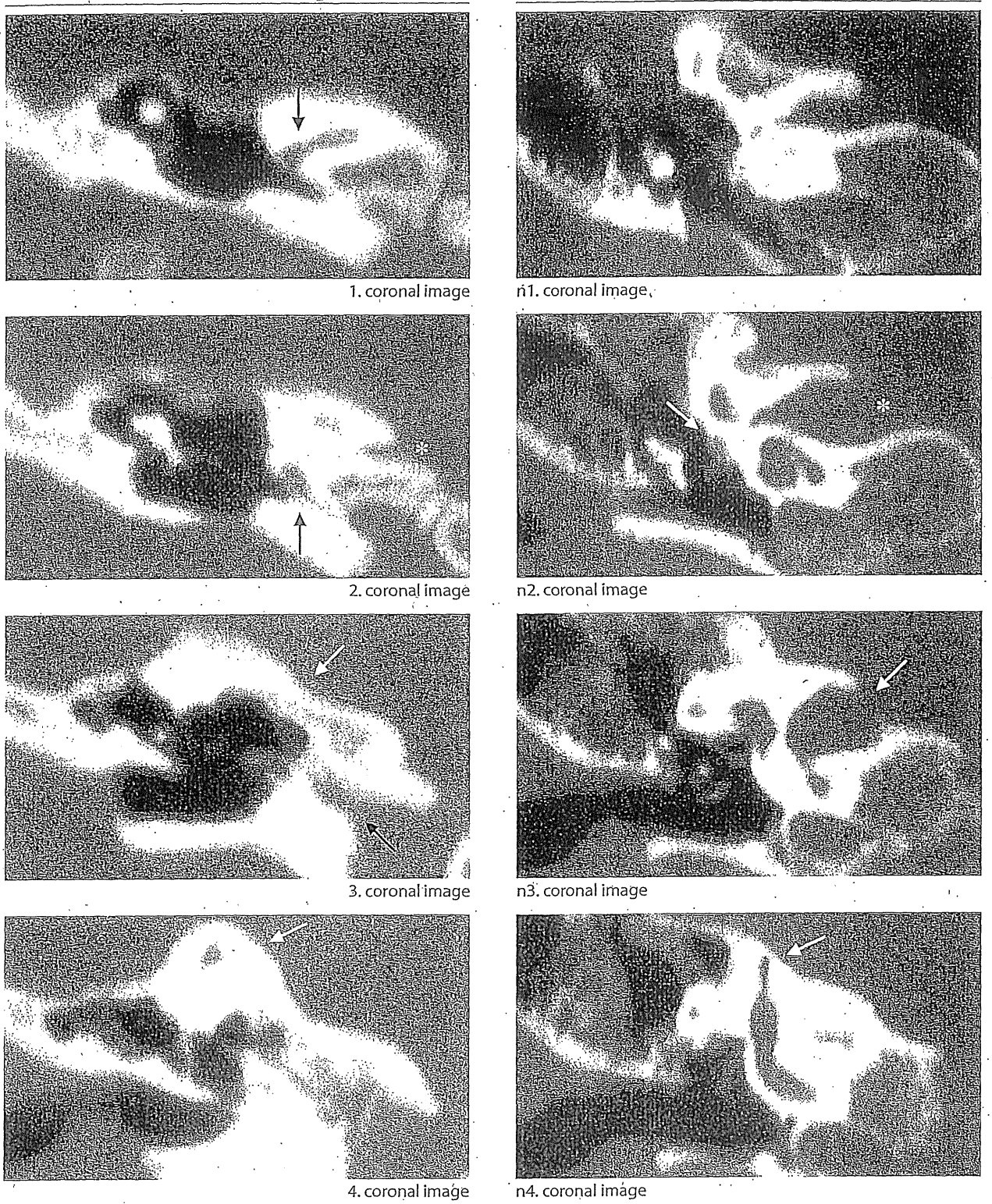


Fig. 24. (Case 1) CT

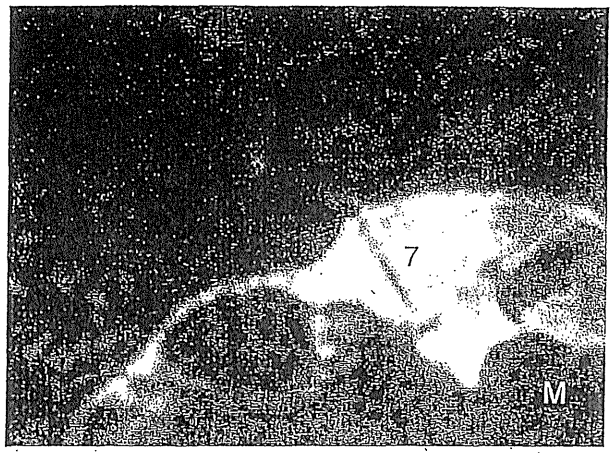
[Patient CT Findings]

The bone mass of the labyrinth is barely visible in the vicinity of the internal auditory canal and the superior part of the anterior semicircular canal (4: ↗). Normally, the structures of the inner ear and internal auditory canal are located between the middle ear and the intracranial space, but in this case as one moves medially in from the middle ear, one passes through a single bony wall before directly entering the intracranial space (3: ↗). The internal auditory canal is located more inferiorly than normal (2: ❁) and turns into the geniculate ganglion (1: ↓) then, after running slightly posteriorly, turns inferiorly and becomes the stylomastoid foramen (3: ↖). The tympanic segment (2: ↑) is also more inferior than normal (n2: ↘). In this case, a small portion of the arch of the upper extremity of the anterior semicircular canal is present (4: ↗), so it does not represent complete aplasia.

《Normal Control CT Findings》

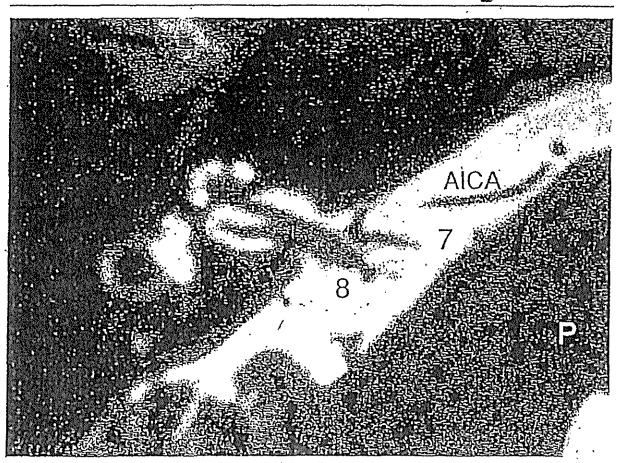
n1: foremost part of the bony capsule of the inner ear. n2: ↘ geniculate ganglion; ❁ internal auditory canal. n3: ↗ internal auditory canal. n4: ↗ anterior semicircular canal.

Patient MRI Findings



1. axial image

Normal Control MRI Findings



n1. axial image

Fig. 25. (Case 1) MRI

[Patient MRI Findings]
The space inside the internal auditory canal contains only the facial nerve (1: numeral 7). 1: M indicates the medulla oblongata.

《Normal Control MRI Findings》
In n1, the numeral 7 indicates the facial nerve, 8 indicates cranial nerve VIII, AICA indicates the anterior inferior cerebellar artery, and P indicates the pons.

as Michel aplasia. Because there is no area corresponding to the ampulla of the anterior semicircular canal it lacks the function of a semicircular canal, and so, as inner ear function can be considered to be completely absent, in functional terms this case belongs in the category of inner ear aplasia.

Michel Aplasia

Reported by Michel in 1863, Michel aplasia is the most severe inner ear malformation [1], caused by arrested inner ear development around the third week of embryonic development. In a normal temporal bone, arrangement of the parts of the ear is, from lateral to medial, the external auditory canal, the middle ear, the inner ear, and the internal auditory canal, but in Michel aplasia the inner ear is absent, with corresponding lateral thinning of the temporal bone. Cases have been reported both in which the inner ear is completely absent and in which, as in this case, it is hypoplastic containing only the facial nerve [2]. As a rule there are no abnormalities of the middle ear, but among the ossicles there is always abnormality of the stapes, including absence or deformation. The path of the facial nerve also deviates inferiorly and anteriorly to normal [3].

When diagnosing Michel aplasia, caution must be exercised to differentiate it from cases in which the inner ear was formed, but has ossified as a result of labyrinthitis due to meningitis or other causes, preventing confirmation of the inner ear in a CT image. As can be seen in this case, in Michel aplasia the bony capsule of the inner ear is essentially absent, with the internal auditory canal either hypoplastic or aplastic. Malformation of the stapes is also frequently observed [1]. Consequently, in cases where a bone mass corresponding to the inner ear is present, the internal auditory canal is normal, and there is no abnormality of the stapes, most likely it is not inner ear aplasia but postnatal inner ear ossification.

References

1 Michel P: Mémoire sur les anomalies congénitales de l'oreille interne. Gazette Méd de Strasbourg 1863;23:55–58.
2 Ozgen B, Oguz KK, Atas A, Sennaroglu L: Complete labyrinthine aplasia: clinical and radiologic findings with review of the literature. AJNR Am J Neuroradiol 2009;30:774–780.
3 Romo LV, Curtin HD: Anomalous facial nerve canal with cochlear malformations. Am J Neuroradiol 2001;223:838–844.

Point

1. Michel aplasia is accompanied by hypoplasia or aplasia of the bony capsule of the inner ear.
2. The internal auditory canal is either hypoplastic or aplastic, and cranial nerve VIII is also absent.
3. The path of the facial nerve is abnormal, and malformation of the stapes is frequent.
4. Caution is required to differentiate Michel aplasia from acquired inner ear ossification.

Cochlear Aplasia

Subject: female, 5 months old

History and Clinical Findings

Newborn hearing screening indicated a need for detailed bilateral examination and the subject received ABR testing at two and three months after birth, but all tests showed no response in either ear (105 dB NHL stimulation). Pediatric examination revealed microcephaly, developmental retardation, and corpus callosum hypoplasia. The subject was referred to our department to determine whether she was compatible for a cochlear implant and for general detailed examination relating to hearing loss. ASSR showed bilateral unresponsiveness for 110 dB at all frequencies, nor was there a response for Behavioral Observation Audiometry (BOA). No abnormalities were observed in the auricle or external auditory canal, but otitis media with effusion was present in both ears.

Patient CT Findings

The cochlea is completely absent, and in the location where it would normally be observed there is only the margin of the bony capsule of the vestibule (fig. 26:1, 2). On the other hand, the vestibule is present, but the lateral and anterior semicircular canals are fused and cystic, with the loops unformed. The structure corresponding to the posterior semicircular canal is not visible (fig. 26:3, 4). Particularly when viewing the coronal images (fig. 27), one can grasp that, even though the so-called pars superior of the inner ear is formed to some extent (fig. 27:3, 4), the pars inferior is almost completely absent (fig. 27:1). The internal auditory canal (fig. 26:3, 4; fig. 27:1–3) is located more anteriorly and inferiorly than normal and is short and narrow.

On examination of the middle ear, the ossicles in this case are almost normal and the malleus, incus, and stapes can all be observed (fig. 26:2–4; fig. 27: 1–4), but except for the epitympanum and mastoid antrum the pneumatic cavities are undeveloped and filled with soft tissue density, including the tympanic cavity. Clinically, because this subject has recurrent acute otitis media, the soft tissue density is assumed to be effusion. The labyrinthine segment of the facial nerve cannot be ascertained, and the tympanic segment cannot be identified due to effusion in the tympanic cavity. For the mastoid segment, a structure that one may assume to be the facial canal is visible lateroposterior to the tympanic cavity (fig. 26:1).

Cochlear Aplasia

Cochlear aplasia is the next most severe inner ear malformation after inner ear aplasia, with abnormality thought to arise around the fourth week of embryonic development. In both Michel aplasia and the cochlear aplasia shown here it is difficult to acquire hearing sensation, and because neither the space to implant an electrode nor the cochlear nerve are present, a cochlear implant is not compatible. However, in recent years, Auditory Brainstem Implant (ABI) surgery has been attempted in cases like this, and it has been reported that a definite hearing sensation can be obtained [1]. ABI was first used on cases of neurofibromatosis type 2 (NF2), but recently is increasingly being applied in cases other than NF2, including severe inner ear malformations such as those shown here. However, compared to acquired ossification of the cochlea, the results for ABI on severe inner ear malformation are relatively poor [2]. The reason for this is not clear but, in addition to the fact that severe inner ear malformation is often accompanied by multiple disabilities, it is possible that congenital deficit of the primary auditory nerve for the cochlea has some sort of negative effect on the development or function of the cochlear nucleus.

References

1 Sennaroglu L, Ziyal I, Atas A, et al: Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. Otol Neurotol 2009;30:708–715.

2 Colletti V, Shannon R, Carner M, et al: Outcomes in nontumor adults fitted with the auditory brainstem implant: 10 Years' Experience. Otology & Neurotology 2009;30:614–618.

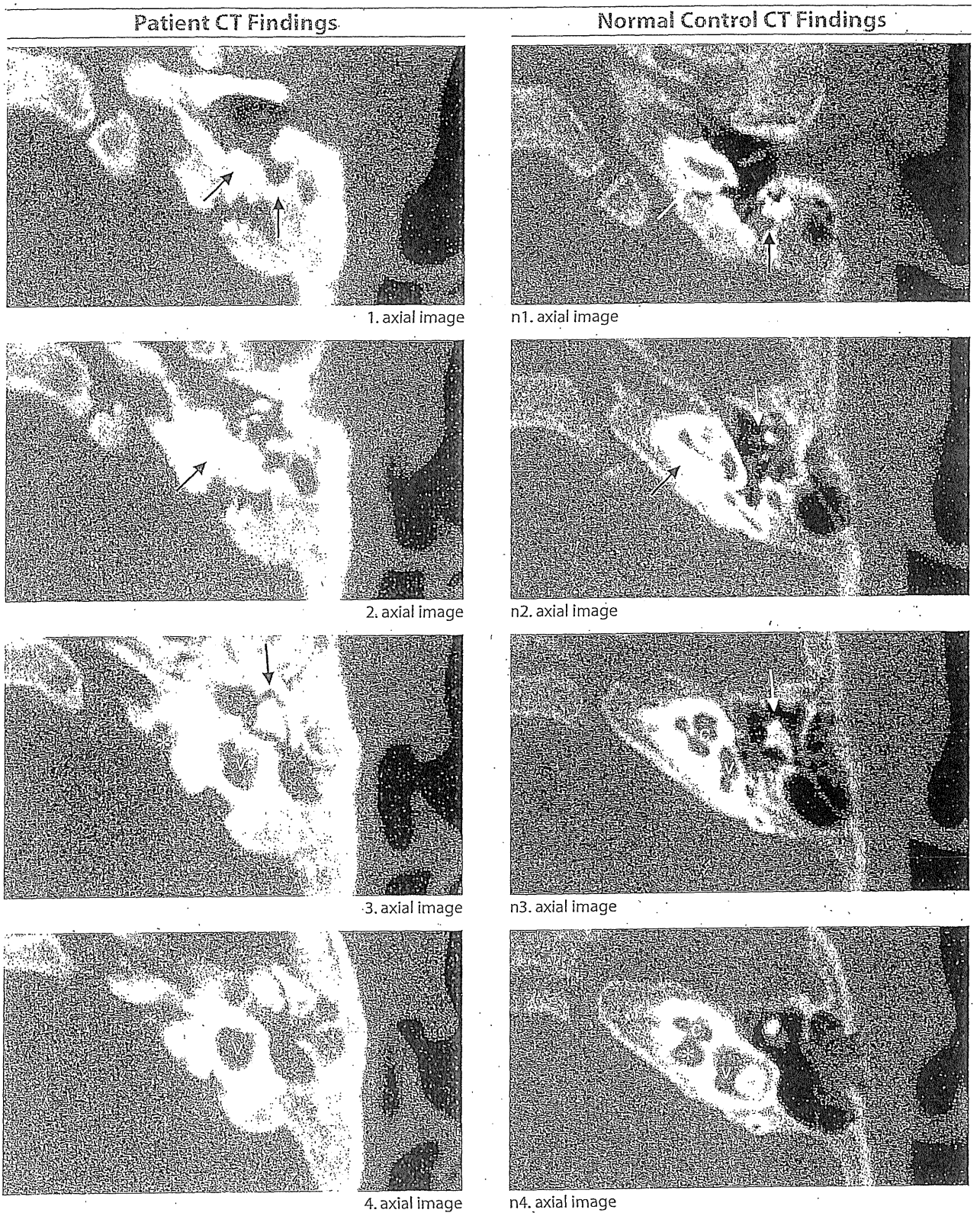


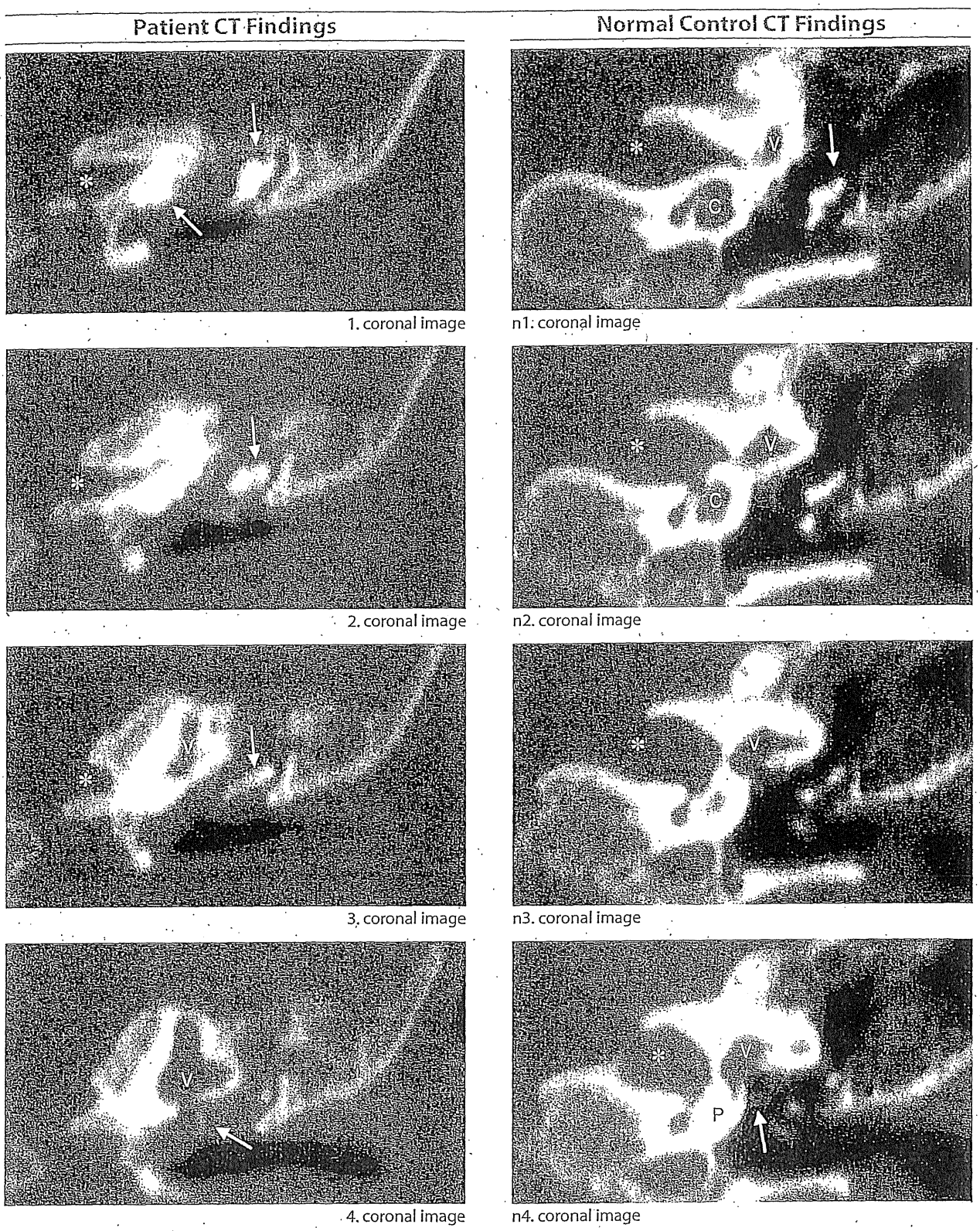
Fig. 26. (Case 2) CT

[Patient CT Findings]

The cochlea is completely absent, presenting only the margin of the bony capsule of the vestibule (1, 2: ↗). The vestibule is present, but the semicircular canals are undifferentiated (3, 4: V). The internal auditory canal (3, 4: ❁) is located more anteriorly and inferiorly than normal and is short and narrow. The ossicles in this case are almost normal and the malleus, incus, and stapes can all be observed (2: ⅄ ; 3: ↓), but except for the epitympanum and mastoid antrum the pneumatic cavities are undeveloped and filled with soft tissue density, including the tympanic cavity. In the mastoid segment, a structure that one may assume to be the facial canal is visible lateroposterior to the tympanic cavity (1: ↑).

《Normal Control CT Findings》

n1: ↗ cochlea; ↑ mastoid segment of facial nerve. n2: ↗ cochlea; ⅄ ossicles. n3: ⅄ malleus, incus; C cochlea; V vestibule. n4: C cochlea; V vestibule; ❁ internal auditory canal.



1. coronal image
n1: coronal image
2. coronal image
n2. coronal image
3. coronal image
n3. coronal image
4. coronal image
n4. coronal image

Fig. 27. (Case 2) CT

[Patient CT Findings]

In the inner ear, even though the so-called pars superior is formed to some extent (3, 4: V), the pars inferior is almost completely absent (1:). The lateral and anterior semicircular canals are fused and sacculated, with the loops unformed (4: V). The internal auditory canal (1–3: ❁) is located more anteriorly and inferiorly than normal and is short and narrow. On examination of the middle ear, the ossicles in this case are almost normal and the malleus, incus, and stapes can all be observed (1–3: ; 4:). but except for the epitympanum and mastoid antrum the pneumatic cavities are undeveloped and filled with soft tissue density, including the tympanic cavity.

《Normal Control CT Findings》

n1: V vestibule; C cochlea; ossicles; ❁ internal auditory canal. Repeated in n2, n3, and n4. n4: P promontory; stapes.

Common Cavity Deformity (1)

Subject: female, 1 year, 3 months

History and Clinical Findings

There were no particular perinatal abnormalities. The subject did not undergo newborn hearing screening. At around one year the subject was examined by a local otolaryngologist to address worries about delayed language development, then subject to auditory testing in the otolaryngology department of a general hospital. The subject was unresponsive to both ABR and ASSR, and was fitted with a hearing aid as an initial step. However, the hearing aid was ineffective, so the subject was brought to our department to determine candidacy for a cochlear implant. There were no abnormalities of the auricle or external auditory canal. The tympanic membrane itself displayed no abnormalities, but there was otitis media with effusion in the right ear along with a history of recurring otitis media. There were no developmental abnormalities except for delays in language and social interaction due to hearing loss.

Patient CT Findings

Findings are for the right ear. First, examining the axial images, the area corresponding to the cochlea is smaller than the normal control (fig. 28:1) and forms a common cavity with no separation from the vestibule. The vestibule and lateral semicircular canal form a single cavity (fig. 28:2–4), but the anterior and posterior semicircular canals are looped (fig. 28:2, 3, 4). Between this common cavity and the internal auditory canal, a thin bony partition can be found (fig. 28:2). The labyrinthine segment of the facial nerve is separated more than usual anteriorly from the internal auditory canal (fig. 28:1), while the tympanic segment is located immediately lateral to the common cavity and runs posterior to the inferior side of the prominence of the lateral semicircular canal. Overall air cell development of the middle ear is inhibited, but the ossicles display no clear abnormalities.

Next, examining the coronal images, it is readily apparent that the space corresponding to the cochlea is slightly protruding inferiorly from the cavity. Also, one can clearly see that, while the lateral semicircular canal is cystic, the anterior semicircular canal forms a separate loop (fig. 28:4).

Patient MRI Findings

Axial MR images for the left and right ears are shown in fig. 29:1R and 1L. Both sides form a common cavity with uniform T2 hyperintense signal inside the cavity and the modiolus, membranous labyrinth, and other structures indiscernible. The partition between the inner ear cavity and the internal auditory canal is clear on the right (fig. 29:1R), but unclear on the left (fig. 29:1L), with the possibility that the internal auditory canal and the common cavity are connected. In considering fenestration of the inner ear during cochlear implant surgery, because the left ear displays a high apparent risk for cerebrospinal fluid leakage (a "gusher") or meningitis due to retrograde infection from the middle ear, we settled on the right ear as more appropriate for surgery.

Examining the 3-dimensional reconstructed MR image (fig. 29:2R, 2L), the overall morphology and points of abnormality are easily discerned. The area corresponding to the cystic lateral semicircular canal displays almost no left-right difference, but the pars inferior, which corresponds to the cochlea, is more solidly formed on the right. Also, on examination of the semicircular canals, while in the right ear the posterior and anterior semicircular canals are separated and have a common crus (fig. 29:2R), in the left ear the two form a single, undifferentiated loop (fig. 29:2L). These minor disparities between the left and right ears are difficult to spot through observation of axial images alone.

Surgical Findings

Cochlear implantation was performed on the right ear, as it was determined from image findings that the probability of a gusher was lower on this side, overall differentiation of the inner ear was slightly more advanced than the left side, and the area corresponding to the cochlea was slightly larger. A mastoidectomy and posterior tympanotomy were performed, the incudostapedial joint severed and the incus extracted, and an approx. 2.5 mm diameter fenestration performed on the common cavity in the anterior end of the lateral semicircular canal prominence. No gusher occurred, and a straight electrode bent in the shape of a U was inserted through the fenestrated portion. During surgery we tried to find a way to implant the electrode as deeply as possible, but due to such factors as the electrode's tendency to spring straight we concluded the operation with it positioned slightly more shallowly than our original objective (fig. 30:1a, 1b, 1c). In postoperative mapping, the electrodes induced nystagmus in response to auditory stimulation, and it was established through verification of the CT images that the problem occurred on the portion contacting the area immediately lateral to the floor of the internal auditory canal. The effectiveness of the cochlear implant was examined with these electrodes disabled. The result was that, while response to sound was apparent, it was insufficient, and revision surgery was performed one year after initial surgery.

In revision surgery, we widely opened the area in the common cavity corresponding to the lateral semicircular canal and directly viewed the cavity's interior (fig. 30:2b). A thin bony partition was ascertained on the floor of the internal auditory canal (through which cerebrospinal fluid was faintly visible), and the electrode bent in the shape of a U was laid anteroinfeior to it. The wide fenestration allowed us to deliberately install the electrode on the anteroinferior segment of the cavity and stabilize it (fig. 30:2c). In the postoperative CT image as well, it was confirmed that the electrode was installed in the position assumed to correspond to the cochlea on the anteroinferior segment of the common cavity (fig. 30:2a, 2b). Postoperatively, a remarkable improvement in sound response compared to the results of the initial operation was obtained using the 12 electrodes in the central region of the array. However, leakage of stimulation current from the electrodes in the tip and base of the array near the fenestrated part of the cavity stimulated the facial nerve, so the device is being used with these electrodes disabled.

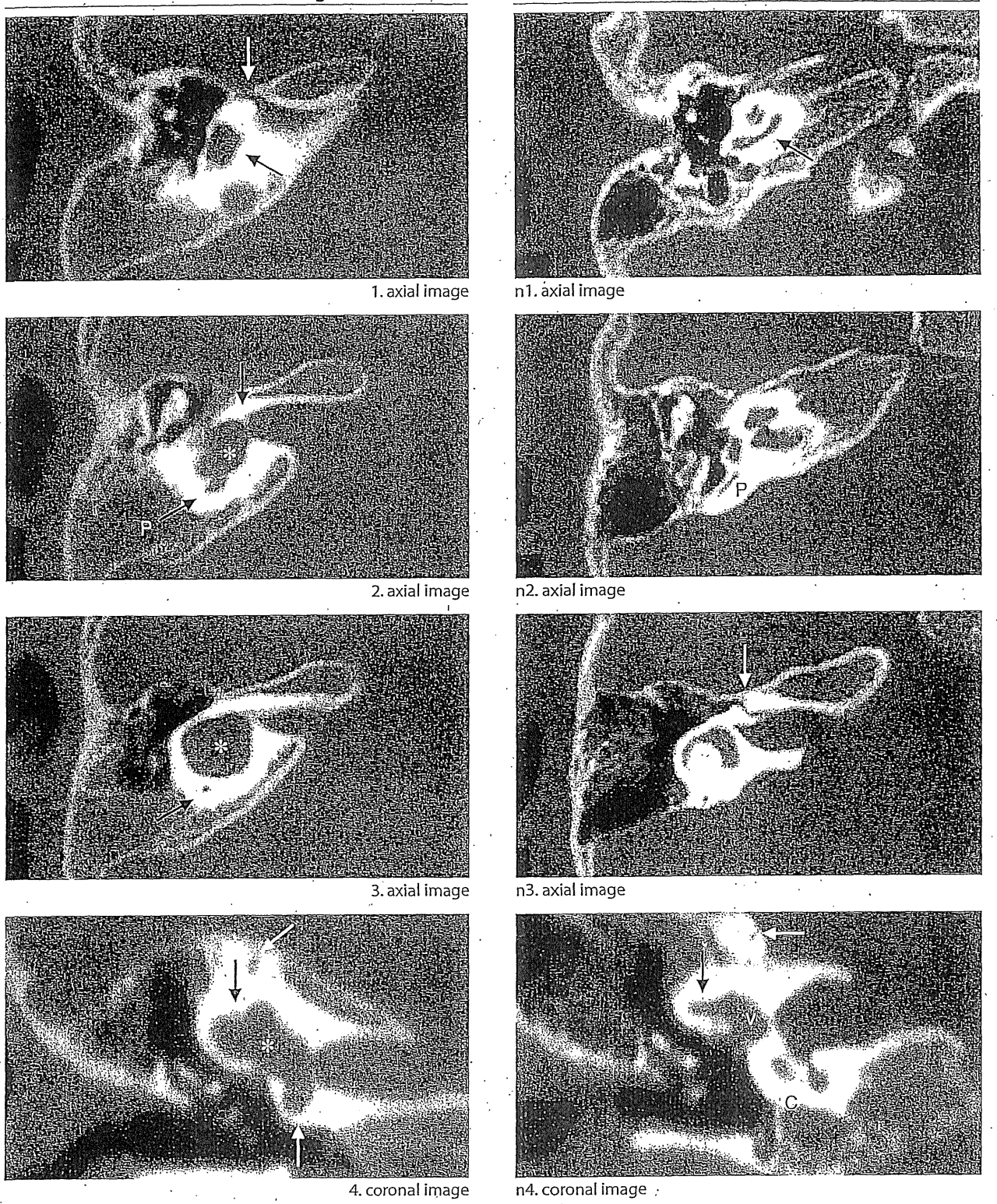


Fig. 28. (Case 3) CT

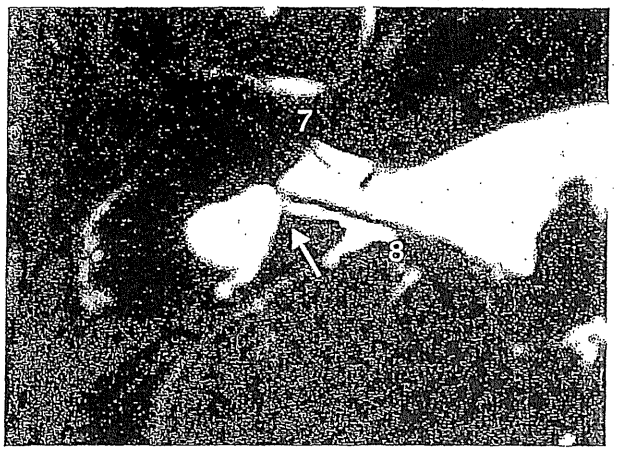
[Patient CT Findings]

The area corresponding to the cochlea is smaller than the normal control (1: ↖) and forms a common cavity with no separation from the vestibule. The vestibule and lateral semicircular canal form a single cavity (2–4: ❄), but the anterior and posterior semicircular canals are looped (2: P ↗, 3: ↗, 4: ♂). Between this common cavity and the internal auditory canal, a thin bony partition can be found (2: ↓). The labyrinthine segment of the facial nerve is separated more than usual anteriorly from the internal auditory canal (1: ⚷). Overall air cell development of the middle ear is inhibited, but the ossicles display no clear abnormalities. It is readily apparent that the space corresponding to the cochlea (4: ⚷) is slightly protruding inferiorly from the cavity. One can clearly see that, while the lateral semicircular canal is cystic (4: ↓), the anterior semicircular canal forms a separate loop (4: ♂).

《Normal Control CT Findings》

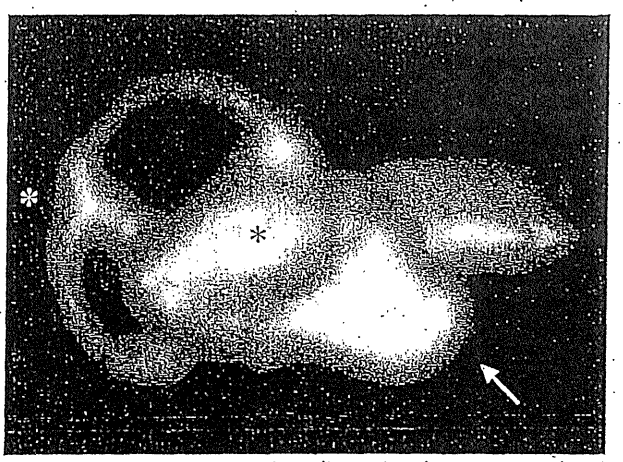
n1: ↖ cochlea. n2: P posterior semicircular canal. n3: ⚷ labyrinthine segment of facial nerve. n4: ↓ lateral semicircular canal; ⇦ anterior semicircular canal; V vestibule; C cochlea.

Patient MRI and 3-Dimensional Reconstructed MRI Findings

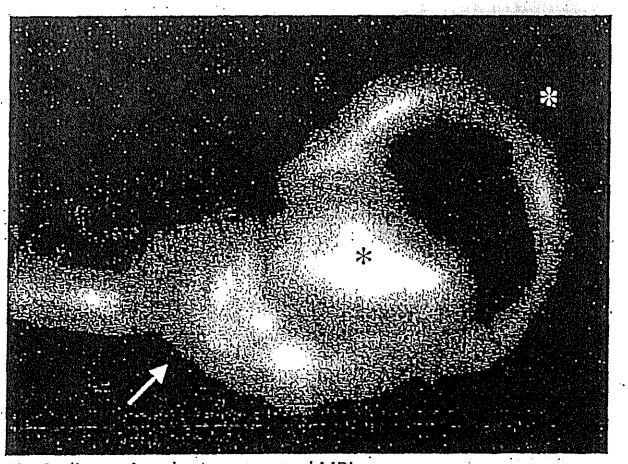


1R. axial image

1L. axial image



2R. 3-dimensional reconstructed MRI



2L. 3-dimensional reconstructed MRI

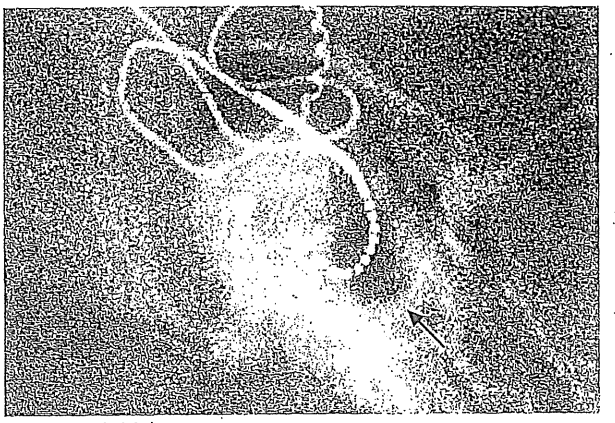
Fig. 29. (Case 3) MRI, 3-dimensional reconstructed MRI

[Patient MRI and 3-Dimensional Reconstructed MRI Findings]

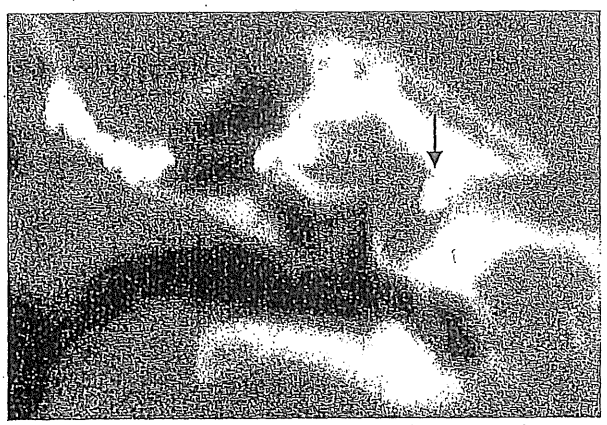
Both sides form a common cavity with uniform T2 hyperintense signal inside the cavity and the modiolus, membranous labyrinth, and other structures indiscernible. The partition between the inner ear cavity and the internal auditory canal is clear on the right (1R: ↖), but unclear on the left (1L: ↖), with the possibility that the internal auditory canal and the common cavity are connected. In considering fenestration of the inner ear during cochlear implant surgery, the left ear displays a high apparent risk of cerebrospinal fluid leakage (a "gusher") or meningitis due to retrograde infection from the middle ear. The area corresponding to the cystic lateral semicircular canal (2R, 2L: *) displays almost no left-right difference, but the pars inferior (2R: ↖, 2L: ↗), which corresponds to the cochlea, is more solidly formed on the right. On examination of the semicircular canals, while in the right ear the posterior and anterior semicircular canals are separated and have a common crus (2R: ❁), in the left ear the two form a single, undifferentiated loop (2L: ❁).

Patient CT and X-Ray Findings

1a. CT: Initial surgery



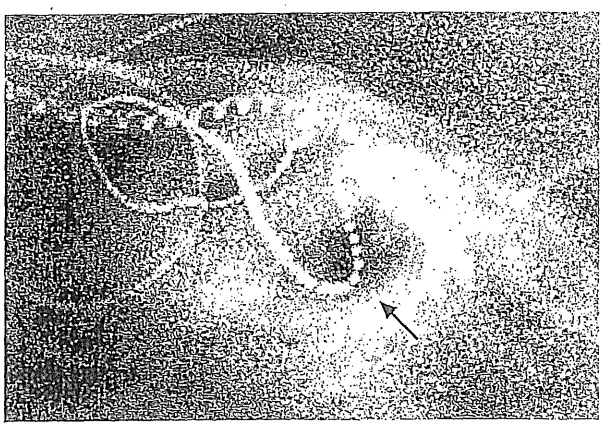
1c. X-ray: Initial surgery



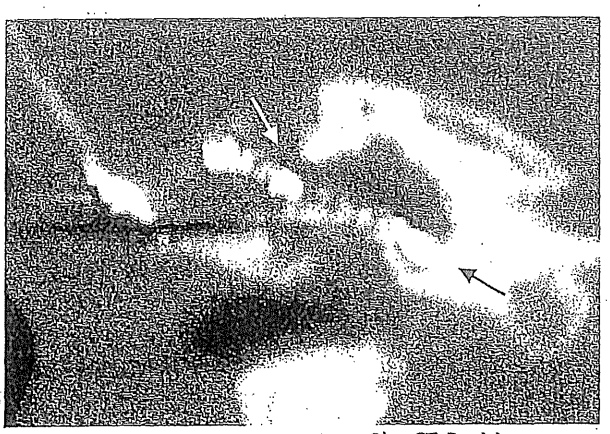
1b. CT: Initial surgery

[Patient CT and X-Ray Findings—1]
Initial Surgery: An approx. 2.5 mm diameter fenestration was performed on the common cavity in the anterior end of the lateral semicircular canal prominence. A straight electrode bent in the shape of a U was inserted through the fenestrated portion. During surgery we tried to find a way to implant the electrode as deeply as possible, but due to such factors as the electrode's tendency to spring straight we concluded the operation with it positioned slightly more shallowly than our original objective (1a: ↓, 1b: ↓, 1c: ↖).

2a. CT: Revision surgery



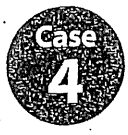
2c. X-ray: Revision surgery



2b. CT: Revision surgery

[Patient CT and X-Ray Findings—2]
Revision Surgery: In revision surgery, we widely opened the area in the common cavity corresponding to the lateral semicircular canal and directly viewed the cavity's interior (2b: ↖). A thin bony partition was ascertained on the floor of the internal auditory canal, and the electrode bent in the shape of a U was laid anteroinferior to it. The wide fenestration allowed us to deliberately install the electrode on the anteroinferior segment of the cavity and stabilize it (2c: ↖). In the postoperative CT image as well, it was confirmed that the electrode is installed in the position assumed to correspond to the cochlea on the anteroinferior segment of the common cavity (2a: ↖, 2b: ↖).

Fig. 30. (Case 3) CT, X-ray



Common Cavity Deformity (2)

Subject: female, 1 year, 10 months

History and Clinical Findings

At around one year five months, the subject's parents noticed that her language development was delayed. At the 18-month physical exam, hearing loss was identified and the subject underwent testing at a pediatric hospital, where she was diagnosed with severe hearing loss due to bilateral inner ear malformation. The infant patient was fitted with a hearing aid, but disliked it and resisted wearing it regularly. The subject was referred to our department at one year ten months to determine the indication for cochlear implant surgery. Both ears were unresponsive to ABR and ASSR, and even with COR no auditory response was observed at maximum sound stimulus level.

Patient CT Findings

The cochlea and vestibule are undifferentiated, forming a common cavity (fig. 31:2–4). The center area of the common cavity is slightly indented, and generally, the anteroinferior portion may be thought to correspond to the cochlea and the posterosuperior portion to the vestibule. The vestibular part is cystic overall, with only the posterior semicircular canal (fig. 31:2, 3) partially formed. As in Case 3, the path of the labyrinthine segment of the facial nerve is abnormal, running anteriorly starting from slightly anterior and medial position of the internal auditory canal (fig. 31:3). A bony partition can be observed between the common cavity and the internal auditory canal (fig. 31:3). In the coronal image, normally both the position and the shape of the vestibule and cochlea can be clearly determined (fig. 31:n4), but in this case, it is impossible to morphologically differentiate between the pars inferior of the cochlea and the pars superior of the vestibule (fig. 31:4).

Turning our attention to the middle ear, we find that mastoid air cell development is favorable and there are no anomalies of the malleus and incus. However, although normally the footplate of the stapes can be observed in the oval window (fig. 31:n2), in this case it cannot be ascertained. Also, a small soft tissue density mass is present lateral to the area corresponding to the oval window, continuous with the common cavity (fig. 31:1, 2, 4). This mass, which is further examined in the MRI findings, could be either a congenital cholesteatoma or a herniation of the inner ear structure.

Patient MRI Findings

The common cavity shows high T2 signal intensity due to labyrinthine fluid, but some low T2 signal structures are observed, which may be some sort of membranous labyrinth. The mass lateral to the oval window mentioned in the section on CT findings is T2 hyperintense and continuous with the common cavity (fig. 32:1), leading us to suspect herniation of inner ear structure. The structure marked P in figure 32:2 and 3 is a single vertical semicircular canal continuous to the posterior or anterior semicircular canal.

Cranial nerves VII and VIII can be observed inside the internal auditory canal (fig. 32:2, 3: 7 and 8), but division of cranial nerve VIII into the cochlear nerve and vestibular nerve cannot be confirmed. As suggested in the CT findings, the facial nerve runs slightly anteriorly starting from slightly central to the fundus (fig. 32:2: 7). The common cavity is independent of the internal auditory canal, separated by a thick septum (fig. 32:2).

Surgical Findings

At two years one month, cochlear implant surgery was performed on the right ear using a straight electrode. A mastoidectomy with posterior tympanotomy was carried out and the incudostapedial joint severed. The long process of the incus was deformed and curved, and the footplate of the stapes between the anterior and posterior limbs was absent. A thin, sacculated structure was herniated and protruding laterally; when incised, it exuded clear labyrinthine fluid. The incus was removed to secure a broad field of view of the posterior half of the epitympanum and an approx. 2.5 mm diameter fenestration performed in the vicinity of the anterior end of the lateral semicircular canal prominence to examine the cavity's interior (fig. 33:1).

A spiderweb-like membrane of soft tissue was present inside the common cavity. This was appropriately transected so as not to interfere with electrode insertion. The size of the common cavity had previously been measured on the images and the electrode bent into a loop to enable it to cover a broad area of the anterior portion of the cavity, then inserted through the fenestration (fig. 33:1). It was confirmed through the fenestration that the electrode was in contact along the cavity wall (fig. 33:2) and the cavity was filled with muscle pieces and fascia. The oval window in particular was filled with soft tissue from inside the cavity so as to occlude the defective area on the footplate of the stapes. Intraoperative X-ray images confirmed that the electrode had been positioned as planned and the bend was smooth (fig. 33:3). Neural Response Telemetry (NRT) testing confirmed a clear response from the cochlear nerve with electrical stimulation of all electrodes (fig. 33:4).

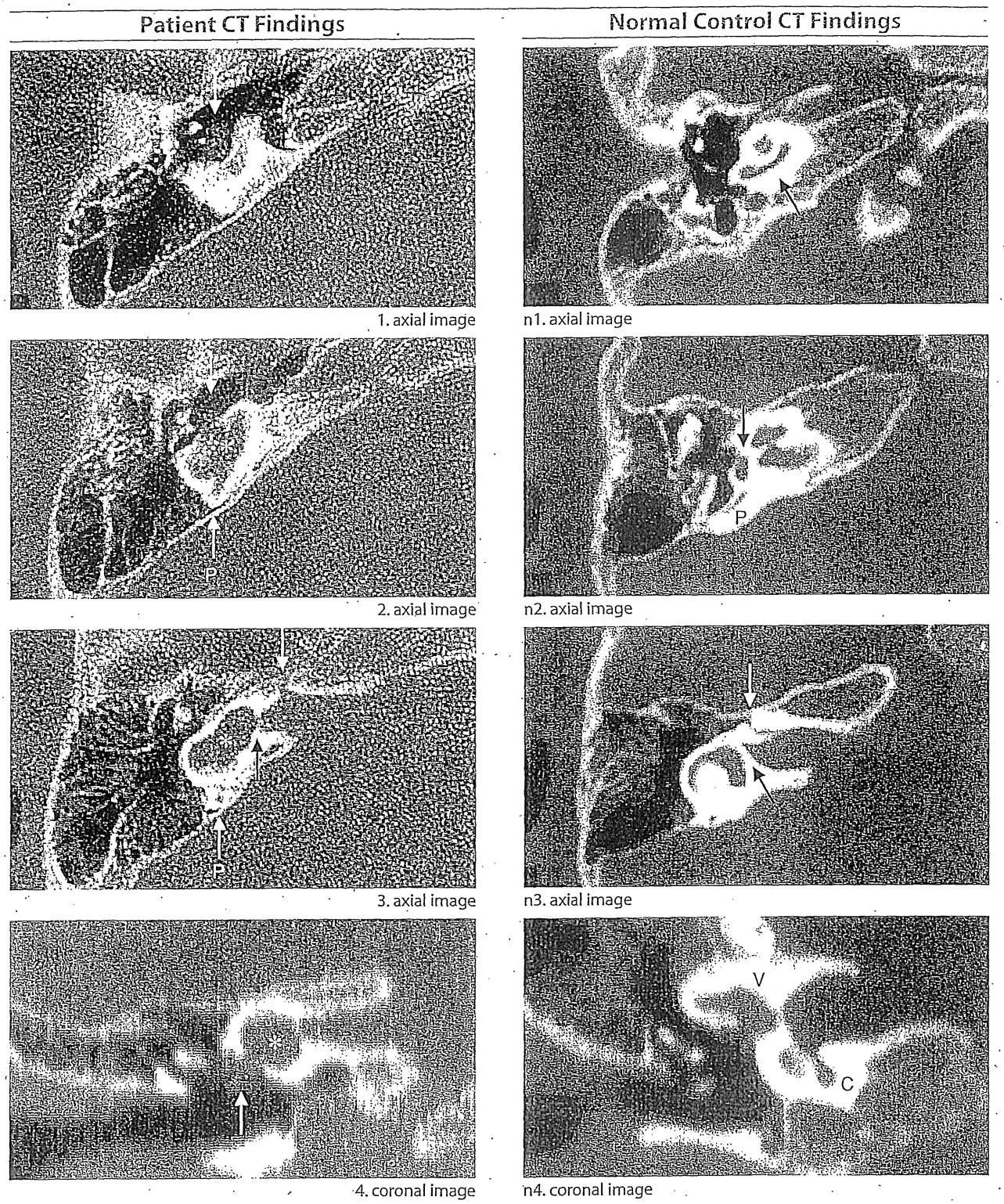


Fig. 31. (Case 4) CT

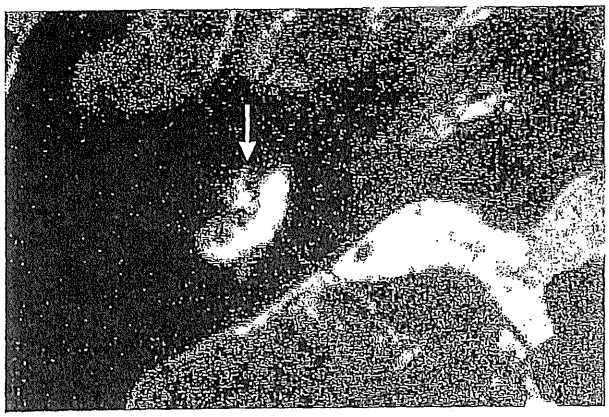
[Patient CT Findings]

The cochlea and vestibule are undifferentiated, forming a common cavity (2–4: ❁). The center area of the common cavity is slightly indented, and generally, the anteroinferior portion may be thought to correspond to the cochlea and the posterosuperior portion to the vestibule. The vestibular part is cystic overall, with only the posterior semicircular canal (2, 3: P ⇑) partially formed. The path of the labyrinthine segment of the facial nerve is abnormal, running anteriorly starting from slightly anterior and medial position of the internal auditory canal (3: ⇣). A bony partition can be observed between the common cavity and the internal auditory canal (3: ↑). In the coronal image, it is impossible to morphologically differentiate between the pars inferior of the cochlea and the pars superior of the vestibule (4: ❁). Mastoid air cell development is favorable and there are no anomalies of the malleus and incus, however the footplate of the stapes cannot be ascertained. Also, a small soft tissue density mass is present lateral to the area corresponding to the oval window, continuous with the common cavity (1, 2: ⇣; 4: ⇑).

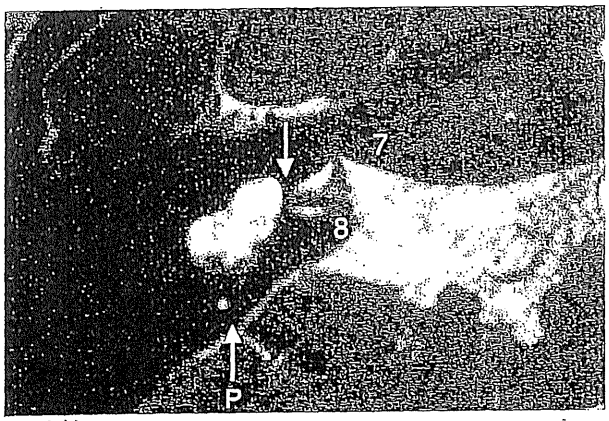
《Normal Control CT Findings》

n1: ↖ cochlea. n2: ↓ oval window, footplate of stapes; P posterior semicircular canal. n3: ⇣ labyrinthine segment of facial nerve; ↖ bony wall of internal auditory canal floor. n4: V vestibule; C cochlea.

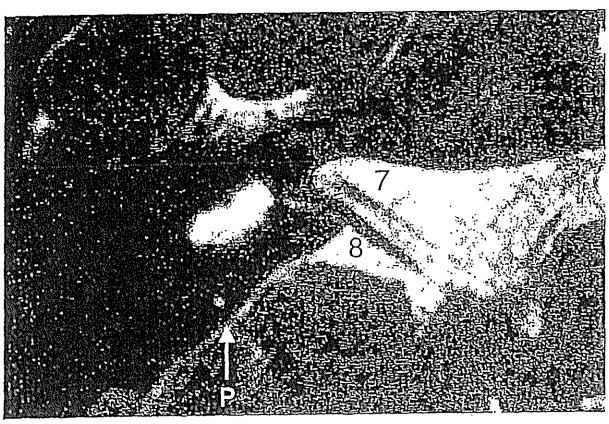
Patient MRI Findings



1. axial image



2. axial image

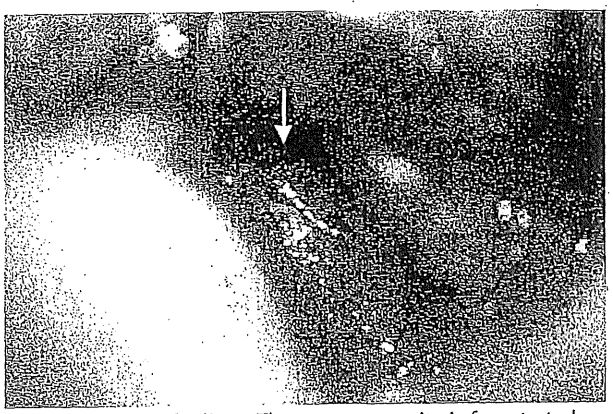


3. axial image

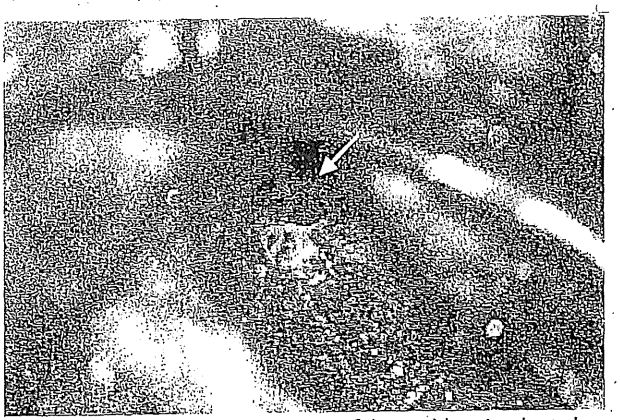
Fig. 32. (Case 4) MRI

[Patient MRI Findings]

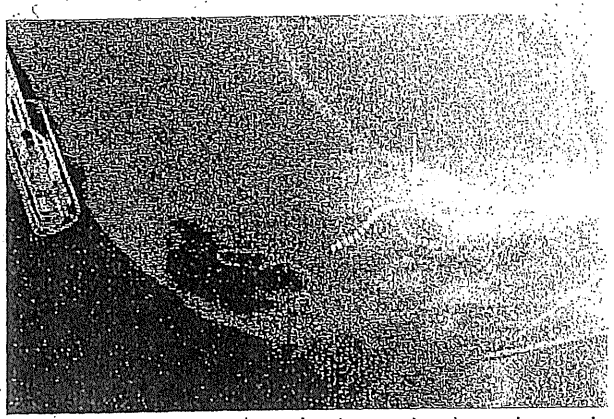
The common cavity shows high T2 singal intensity due to labyrinthine fluid, but some low T2 signal structures are observed, which may be some sort of membranous labyrinth formation. The mass lateral to the oval window is T2 hyperintense and continuous with the common cavity (1: ↓), and herniation of inner ear structure is suspected. The structure marked P in 2 and 3 is a single vertical semicircular canal continuous to the posterior or anterior semicircular canal. Cranial nerves VII and VIII can be observed inside the internal auditory canal (2, 3: 7 and 8), but division of cranial nerve VIII into the cochlear nerve and vestibular nerve cannot be confirmed. The facial nerve runs slightly anteriorly starting from slightly central to the fundus (2: 7). The common cavity is independent of the internal auditory canal, separated by a thick septum (2: ↓).



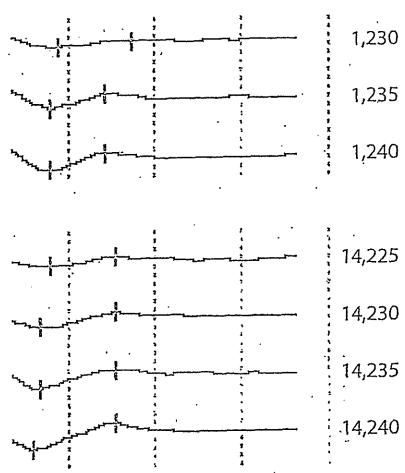
1. Intraoperative findings: The common cavity is fenestrated on the anterior end of the area corresponding to the vestibule and the electrode (↓) inserted.



2. Intraoperative findings: Insertion of the cochlear implant electrode (↙) has just been completed.



3. Intraoperative X-ray image: ↓ indicates the electrode array in the common cavity.



4 NRT testing

Fig. 33. (Case 4) Right ear cochlear implant surgery findings

Case 5 Aplasia of Cochlear Upper Turns and Semicircular Canals with Cochlear Nerve Canal Stenosis

Subject: female, 3 years old

History and Clinical Findings

This child did not undergo newborn hearing screening. Since just after one year old the parents had been worried by the child's unclear response to sound, and when even at two years she had not begun speaking they took her to an otolaryngology department of a general hospital. There she was diagnosed with severe hearing loss after ABR testing showed no response on either side with a stimulation of 105 dB NHL, whereupon the hospital referred the subject to our department for further testing. In COR testing no significant response was observed at 110 dB and, taking other findings into consideration, we recommended cochlear implantation. Since the family were concerned about the risks of surgical treatment, the subject was fitted with a hearing aid. It was confirmed that language development could not be obtained with a hearing aid, and the subject underwent cochlear implantation at the age of three.

Patient CT Findings

The temporal bone CT images taken during the initial medical examination by our department presented approximately the same findings for both ears. The results for the left ear are presented here. The cochlea's basal turn is thicker than normal (fig. 34:1C) and the upper turns can hardly be identified. The vestibule is present (fig. 34:2), but the semicircular canals are not identified (fig 34:3) except for a vestige of the posterior semicircular canal (fig 34:1).

Also, the structure of the area between the fundus of the internal auditory canal and the cochlea demands special attention in this case. Normally the cochlear base portion of the internal auditory canal, referred to as the cochlear area, is seen as a broad opening on CT images (fig. 34:n2) where the cochlear nerve fibers pass through a cribriform plate, but in this case only a narrow tube connects the fundus of the internal auditory canal with the modiolus (fig. 34:2). This type of tubular structure does not have a proper anatomical name as it does not exist in normal anatomy, but it is sometimes referred to as the cochlear nerve canal. When it is constricted, the cochlear implant efficacy is reduced, so it is important to check its condition when evaluating clinical images. This matter is discussed in further detail in the section titled "Anomalies of the internal auditory canal."

Patient MRI and 3-Dimensional Reconstructed MRI Findings

The modiolus can be ascertained (fig. 35:1). The individual nerves within the internal auditory canal cannot be identified as clearly as usual, but it is impossible to determine whether this is due to the nerves not being properly captured in the imaging slice or to a morphological abnormality in the nerves themselves. At the cerebellopontine angle, normally cranial nerve (CN) VIII is depicted thicker than CN VII, but here it is the reverse and CN VII is thicker, indicating the possibility that CN VIII (fig. 35:1) is atrophied or hypoplastic.

Observing the 3-dimensional reconstructed MR image of the inner ear, the overall abnormal condition is readily apparent. The cochlea in this case (fig. 35:2) ends after around 1.5 turns overall (①, ②). Compared to the normal control image, which has a second turn (fig. 35:n2, ②) and an apical turn (＊), it is clear that the upper turn is deficient. As seen in the CT images, hypoplasia of the vestibular system is conspicuous (fig. 35:2), with only a slight distention in the area assumed to be the base of the ampullae of the posterior and anterior semicircular canals.

Surgical Findings

Cochlear implantation was performed on the left ear. Normally in cases of inner ear malformation, use of a straight electrode array is recommended. This is because the design of a pre-curved electrode is based on the shape of a normal cochlea, so in cases of malformation it is uncertain whether or not the electrode can be properly inserted in accordance with the condition of the structure of the cochlea for each individual case. However, in this case, the modiolus was clearly present and the structure of the first turn, while slightly thicker than usual, appeared to be normal, so a pre-curved electrode was used. As a result, the electrode could be inserted smoothly without complication, and intraoperative X-ray confirmed that it had been suitably inserted in place.

Postoperative response to sound was fair, and the threshold level with cochlear implant stabilized at 45 dB.

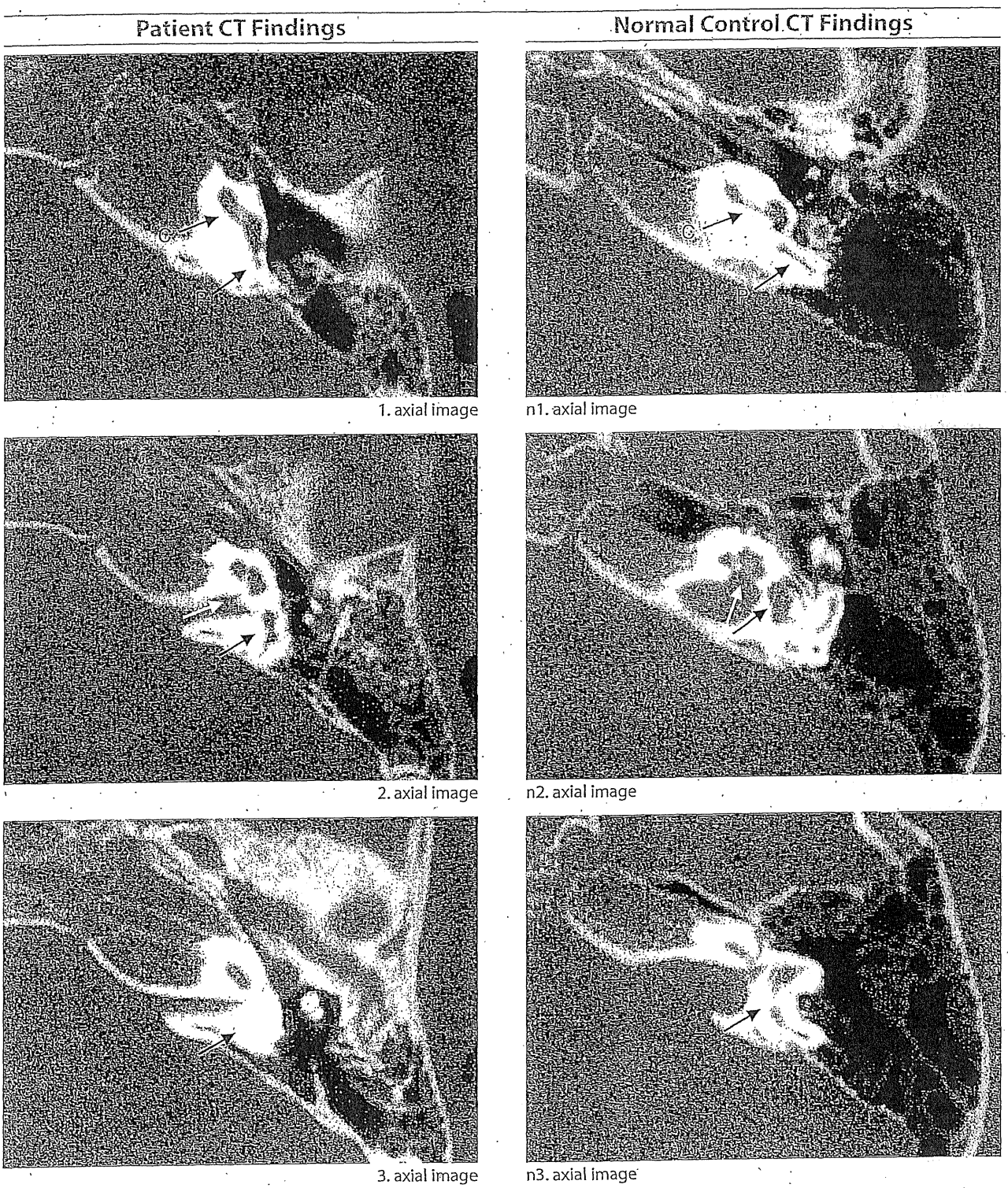


Fig. 34. (Case 5) CT

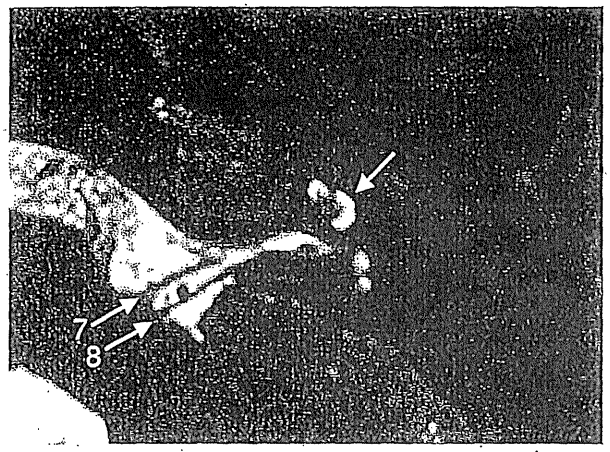
[Patient CT Findings]

The cochlea's basal turn is thicker than normal (1: C↗) and the upper turns can hardly be identified. Of the semicircular canals, only a vestige of the posterior semicircular canal can be seen (1: P↗). The vestibule is present (2:↗), but the semicircular canals are not identified (3: ↗). Normally the cochlear base portion of the internal auditory canal, referred to as the cochlear area, is seen as a broad opening on CT images (n2: ↗) where the cochlear nerve fibers pass through a cribriform plate, but in this case only a narrow tube connects the fundus of the internal auditory canal with the modiolus (2:↗).

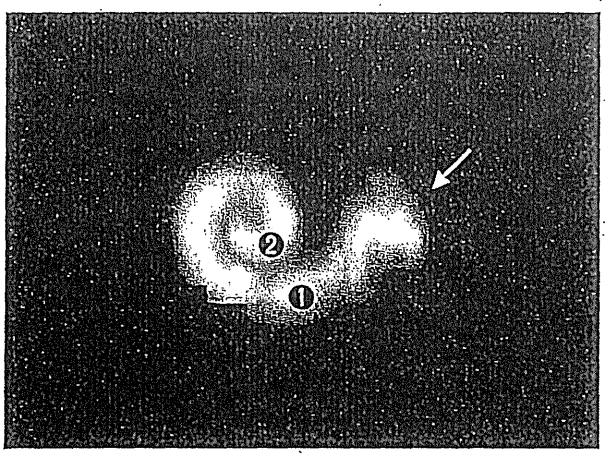
《Normal Control CT Findings》

n1: C ↗ cochlea; P ↗ posterior semicircular canal. n2: ↗ cochlear area on fundus of internal auditory canal; ↗ vestibule. n3: ↗ vestibule, semicircular canals.

Patient MRI and 3-Dimensional Reconstructed MRI Findings

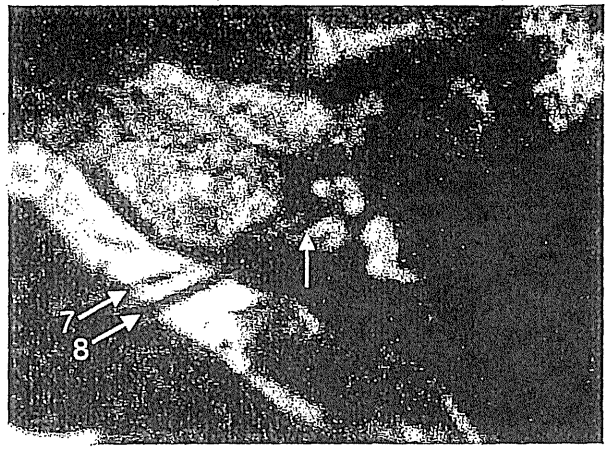


1. axial image

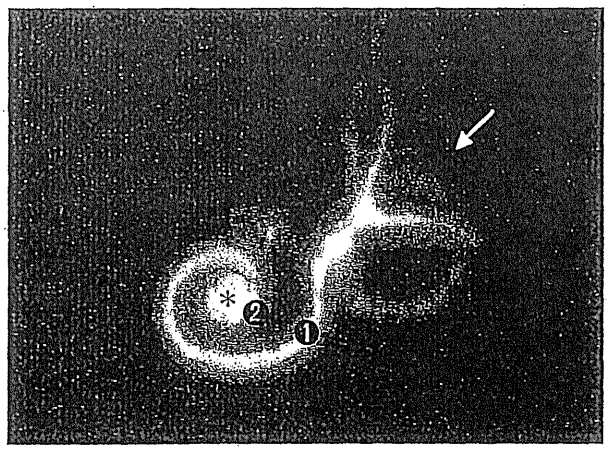


2. 3-dimensional reconstructed MRI

Normal Control MRI and 3-Dimensional Reconstructed MRI Findings



n1. axial image



n2. 3-dimensional reconstructed MRI

Fig. 35. (Case 5) MRI, 3-dimensional reconstructed MRI

[Patient MRI and 3-Dimensional Reconstructed MRI Findings]

The modiolus can be ascertained (1: ↙). The individual nerves within the internal auditory canal cannot be identified as clearly as usual. At the cerebellopontine angle, normally cranial nerve (CN) VIII is depicted thicker than CN VII, but here it is the reverse and CN VII is thicker (1: 7 ↗), indicating the possibility that CN VIII (1: 8 ↗) is atrophied or hypoplastic. The cochlea (2) ends after around 1.5 turns overall (①, ②). Compared to the normal control image, which has a second turn (n2: ②) and an apical turn (*), it is clear that the upper turn is deficient. Hypoplasia of the vestibular system is conspicuous (2: ↙), with only a slight distention in the area assumed to be the base of the ampullae of the posterior and anterior semicircular canals.

《Normal Control MRI and 3-Dimensional Reconstructed MRI Findings》

n1: 7 ↗ facial nerve; 8 ↗ cranial nerve VIII; ↑ cochlear nerve.
n2: ① start of turn at base of cochlea; ② start of second turn; * apical turn.

Incomplete Partition Type I (IP-I): Case of Cochlear Implantation Revision

Subject: male, 3 years old

History and Clinical Findings

Newborn hearing screening revealed the need for further testing and the subject received a detailed examination in the otolaryngology department of a local general hospital. He was then fitted with and began wearing a hearing aid. At the time, the threshold value while wearing a hearing aid was 40–50 dB and at first language development was observed, but gradually hearing deteriorated and the effects of the hearing aid diminished, so at three years old the subject was referred to our department. In our initial exam, hearing level was 120–130 dB in the right ear and 110–120 dB in the left ear. Aided thresholds were 50 dB at 500 Hz, 65 dB at 1,000 Hz, and off the scale at 2,000 Hz and above. The developmental quotients on the Kyoto Scale of Psychological Development were as follows: Posture-Movement: 100; Cognitive-Adaptive: 94; Language-Social: 64, with developmental delay shown only in language and socialization. It was a typical profile for a hearing-impaired child without accompanying general developmental retardation. It was decided that a cochlear implant would be indicated and surgery should be performed.

Patient CT and MRI Findings

In CT images of the temporal bones, inner ear malformations are observed on both sides. On the right side, the basal turn and upper turn of the cochlea are unseparated and cystic (fig. 36:1R). The lateral semicircular canal is also cystic (fig. 36:2R), with the vertical semicircular canal existing separately from this cystic vestibule. The reason we refer to this structure as the "vertical semicircular canal" here is because in this case there is no common crus and the posterior and anterior semicircular canals are unified as a single semicircular canal vertical to the horizontal plane. The right inner ear is an incomplete partition type I according to the Sennaroglu-Saatci classification. On the MRI of the right side, a partition is faintly visible between the inner ear and the internal auditory canal (fig. 36:3R). Meanwhile, on the left side, the basal turn of the cochlea is thicker than normal (fig. 36:1L), and the second turn and beyond is cystic. The vestibule and lateral semicircular canal form a single cyst (fig. 36:2L), but the vertical semicircular canal is separate from it. The left ear is also diagnosed as an incomplete partition type I. On the left side, no partition is visible between the internal auditory canal and the cochlea (fig. 36:3L), so it is possible that cerebrospinal fluid is freely exchanged with the inner ear and a cerebrospinal fluid gusher is anticipated if fenestration of the cochlea is performed during cochlear implantation.

In the MR images of the cochlea, a hypointense area corresponding to the modiolus is faintly visible (fig. 36:3L). Normally, this area is hypointense due to both the bony structure of the modiolus and the cochlear nerve and spiral ganglion cells inside. However, here we have already determined from the CT images that there is no bony modiolus, so the hypointense signal in the central part of the cochlea observable on the MRI is probably the cochlear nerve and spiral ganglion cells.

Observing the 3-dimensional reconstructed MR images for this case (fig. 37:R, L), the vestibular part is formed nearly the same on both sides. For the cochlea, however, whereas on the right side (R) the cochlea has approx. 1.5 turns overall and is cystic in the upper part, on the left side (L) the basal turn is almost completely formed and the second turn and beyond is cystic, leading us to believe that the left side is slightly more differentiated than the right.

Surgical Findings and Postoperative Course

Cochlear implantation was performed on the left ear. A mastoidectomy was performed and the opening of the posterior tympanotomy made wider than usual in anticipation of the need for steps to be taken later in the event of a gusher. Cerebrospinal fluid erupted on fenestration of the basal turn of the cochlea, but the eruption weakened after a short time and the cochlear implant electrode (straight type) was inserted. The area between the fenestration and the electrode was then filled with pieces of temporalis fascia and sealed and fixed using fibrin glue. After waiting to confirm that there was no eruption of cerebrospinal fluid, surgery was concluded. The postoperative course went well, favorable hearing was obtained, and language acquisition was promoted.

However, two years after the operation, facial spasms started to occur with sound stimulation, and speech perception scores began to deteriorate. On viewing simple X-ray images of the electrode, in the intraoperative image taken during the initial surgery full insertion could be confirmed (fig. 38:a), but in the image taken after the occurrence of facial spasms it was apparent that the electrode had slipped out slightly (fig. 38:b). The cochlear implant installed in the initial surgery was extracted, and this time a cochlear implant with a curved electrode was implanted (fig. 38:c). This approach was followed because, in this case, the left cochlea had a modiolus, so it was surmised that a curved electrode would provide more efficient nerve stimulation than a straight one. We also hypothesized that latent cerebrospinal pressure might have pushed the electrode array out of the cochlea, and hoped that a curved electrode around the modiolus would help maintain the electrode in a stable position. The postoperative course went well, with no stimulation of the facial nerve, and the electrode array remained stable.