Aplasia and Hypoplasia of the Vestibulocochlear Nerve: Diagnosis with MR Imaging

PURPOSE: To introduce aplasia or hypoplasia of the vestibulocochlear nerve (VCN) as a possible cause of hearing loss and to identify the magnetic resonance (MR) imaging characteristics of this entity.

MATERIALS AND METHODS: In seven patients with congenital deafness or unexplained sensorineural hearing loss, MR imaging enabled diagnosis of aplasia or hypoplasia of the VCN. Axial (0.7-mm) three-dimensional Fourier transformation-constructive interference in steady state (3DFT-CISS) images and parasagittal reconstruction images perpendicular on the course of the VCN were obtained. Twenty normal inner ears were also studied; their findings were compared with those of the patients.

RESULTS: The facial nerve and inferior and superior vestibular and cochlear branches of the VCN were identified on the MR images in the 20 normal inner ears. Aplasia of the VCN was detected in two patients with normal labyrinths but with a severe stenosis of the internal auditory canal. A common VCN was absent in the cochlear branch was found bilaterally in two patients with a congenital malformation of the labyrinth. A common VCN with absence of the cochlear branch was found in three patients with normal internal auditory canals and labyrinths.

CONCLUSION: Submillimetric gradient-echo images (eg, 3DFT-CISS) should always be used to exclude aplasia or hypoplasia of the cochlear branch of the VCN in all cochlear implant candidates and patients with congenital deafness. This entity, which can occur with or without associated labyrinthine malformation, should be confirmed in two planes.

MAGNETIC resonance (MR) imaging is accepted as the method of choice to look for abnormalities in patients with sensorineural hearing loss and/or vertigo (1-5). Moreover, gradient-echo MR imaging proved its value in patients with congenital inner ear malformations (6) and especially in candidates for cochlear implantation (7-9). The MR demonstration of a normal cochlea, with a fluid-filled scala tympani and vestibuli, indicates that these patients are ideal candidates for implantation. Nevertheless, in some of these patients the cochlear implant did not result in auditory perception, although the implant itself worked and was not causing the problem (O’Donoghue GM, personal communication, 1996). A possible explanation could be an absent or abnormal cochlear branch of the vestibulocochlear nerve (VCN). With the advent of submillimetric T2-weighted gradient-echo images, these nerves and nerve branches can be visualized in a reliable way inside the internal auditory canal and cerebellotopine angle (10). In the internal auditory canal, the facial nerve and the cochlear branch, inferior vestibular branch, and superior vestibular branch of the VCN are best recognized on axial three-dimensional Fourier transformation-constructive interference in steady state (3DFT-CISS) images and can be confirmed on parasagittal reconstruction images (5).

The purpose of our study was to introduce aplasia or hypoplasia of the VCN as a possible cause of sensorineural hearing loss or congenital deafness and to identify the MR imaging characteristics of this entity with use of the 3DFT-CISS sequence.

MATERIALS AND METHODS

Subjects

During a period of 24 months (from February 1994 to January 1996), the diagnosis of aplasia or hypoplasia of the cochlear branch of the VCN was made at MR imaging in seven patients (six male patients, one female patient; age range, 2-42 years; average age, 11 years). The common clinical finding that led to computerized tomography (CT) and MR imaging was sensorineural hearing loss. The sensorineural hearing loss was congenital and bilateral in three patients (who were part of a cochlear implantation selection program) and congenital and unilateral in three patients, and sudden hearing loss occurred in one patient. More details of the clinical presentation of these patients are listed in the Table.

Imaging Protocol

All patients underwent MR imaging performed with a 1-T, active-shielded su-
Clinical and Imaging Data of the Affected Inner Ears of the Seven Patients with Aplasia or Hypoplasia of the VCN

<table>
<thead>
<tr>
<th>Patient No./Sex/Age (y)</th>
<th>Clinical Presentation</th>
<th>Side</th>
<th>CT Findings</th>
<th>MR Findings</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Labyrinth</td>
<td>IAC</td>
</tr>
<tr>
<td>1/F/2</td>
<td>Until age 9 months, parents thought the child had normal hearing; at 14 months, bilateral congenital deafness was confirmed with audiogram (vibration deafness) and ABR; normal facial nerve function; potential cochlear implant candidate</td>
<td>Right</td>
<td>IAC stenosis</td>
<td>Normal</td>
</tr>
<tr>
<td>2/M/42</td>
<td>Bilateral aplasia of external ear and the IAC, normal facial nerve function, plastic and otologic surgery (approximately 30 interventions), BAHA on the right side and bilateral epiphyses, sudden SNHL on the right side (at age 42 years) with completely deaf ear on the left (per audiogram)</td>
<td>Left</td>
<td>Stenosis, separate canal for facial nerve</td>
<td>Normal</td>
</tr>
<tr>
<td>3/M/6</td>
<td>Congenital deafness, noted by parents at age 6 months; at 18 months, deafness confirmed with ABR and audiogram (corner audiogram); presumably autosomal or X-linked recessive; electric promontory stimulation: response on left ear; potential cochlear implant candidate</td>
<td>Right</td>
<td>Common cavity, abnormal semicircular canals</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Left</td>
<td>Common cavity, abnormal semicircular canals</td>
<td>Normal</td>
</tr>
<tr>
<td>4/M/10</td>
<td>Bilateral congenital deafness confirmed with audiogram; electric promontory stimulation is planned; potential cochlear implant candidate</td>
<td>Right</td>
<td>Common cavity, IAC stenosis, partial development of SSC</td>
<td>Stenosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Left</td>
<td>Common cavity, IAC stenosis, partial development of SSC</td>
<td>Stenosis</td>
</tr>
<tr>
<td>5/M/5</td>
<td>Congenital hearing loss on the left side since birth; at 6 years, total congenital deafness on the left side confirmed with audiogram (phantom curve), normal hearing on the right side</td>
<td>Right</td>
<td>Not applicable</td>
<td>Normal</td>
</tr>
<tr>
<td>6/M/3</td>
<td>Congenital deafness on the right side since birth; only mild hearing loss on the left; completely deaf ear on the right confirmed with audiogram</td>
<td>Right</td>
<td>Not applicable</td>
<td>Normal</td>
</tr>
<tr>
<td>7/M/12</td>
<td>Child turns head to the left to hear with normal right ear; congenital high frequency hearing loss on the left side confirmed with audiogram; normal hearing on the right</td>
<td>Left</td>
<td>Normal</td>
<td>Normal</td>
</tr>
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</table>

Note.—ABR = auditory brain-stem responses, BAHA = bone-anchored hearing aid, EAC = external auditory canal, IAC = internal auditory canal, SNHL = sensorineural hearing loss, SSC = superior semicircular canal.

periconductive system (Magnetom SP 42; Siemens, Erlangen, Germany). A standard circular polarized head coil was used to allow simultaneous imaging of both inner ears. All patients underwent a routine inner ear study with an axial, 7-mm-thick, T2-weighted spin-echo sequence (2,000/16, 90 [repetition time msec/echo time msec], one signal acquired) of the brain and a selective, axial, 3-mm-thick, contiguous, T1-weighted spin-echo sequence (500/14, four signals acquired) with and without administration of an intravenous gadolinium chelate. Their temporal bones were also examined with a coronal gadolinium-enhanced T1-weighted sequence. A dose of 0.1 mmol/kg gadoterate meglumine (Dotarem; Guerbet Laboratories, Aulnay-sous-Bois, France) or gadopentetate dimeglumine (Magnevist; Schering, Berlin, Germany) was used. The patients were also examined with a 3DFT-CISS gradient-echo sequence. The details of this gradient-echo sequence have been published previously (4,10). The most important parameters of this sequence are the following: 15/21, 65° flip angle, one axial slab of 22.4-mm thickness, 32 partitions, 256 × 256 matrix, 170-mm field of view, two signals acquired. This results in 0.7-mm axial sections with an in-plane resolution of 0.66 × 0.66 mm and a total acquisition time of 8 minutes 6 seconds. The absence of nerves or nerve branches was always checked on parasagittal 3DFT-CISS reconstruction images. These reconstruction images were made through the cerebellopontine angle and middle and lateral thirds of the internal auditory canal and were angled perpendicular on the course of the nerves (Figs 1, 2a, 2b).

In six of the patients, 15-mm-thick axial CT sections also were obtained every 1 mm, and in two patients (patients 2 and 4) additional coronal CT scans were acquired.

Control Group

Twenty normal inner ears were also studied with 0.7-mm-thick 3DFT-CISS images. The selected 3DFT-CISS examinations were free of movement or flow artifacts and therefore allowed adequate evaluation of the nerves in the cerebellopontine angle and internal auditory canal. The reliability to distinguish the facial nerve and the inferior vestibular, superior vestibular, and cochlear branches of the VCN in the cerebellopontine angle and internal auditory canal was checked on these axial images and on the parasagittal 3DFT-CISS reconstruction images, made
perpendicular on the course of the nerves in the cerebellopontine angle and in the middle and lateral thirds of the internal auditory canal. The thickness of the nerves in relation to one another was evaluated on the parasagittal reconstruction images. Especially the thicknesses of the VCN (in the cerebellopontine angle) and its cochlear branch (in the internal auditory canal) were compared with the thickness of the facial nerve.

RESULTS

Control Group

The facial nerve and the cochlear branch, inferior vestibular branch, and superior vestibular branch of the VCN (Fig 1) could be visualized on the axial 3DFT-CISS images in all 20 normal inner ears (Fig 2a, 2b). On the parasagittal reconstruction images made at the level of the cerebellopontine angle, both the facial nerve and VCN were always visible. At this location, the VCN was 1½–2 times larger in diameter than the facial nerve in 19 of the 20 normal inner ears (Fig 2c). In one inner ear, they were the same size. In 19 inner ears, the facial nerve and the cochlear branch of the VCN and the common vestibular nerve (the vestibular branch of the VCN, not yet bifurcating into a superior and inferior branch) could be distinguished on the parasagittal reconstruction images made through the middle third of the internal auditory canal. The three branches of the VCN and the facial nerve could be separated from one another on the parasagittal reconstruction images made near the fundus of the internal auditory canal. In only one patient was it difficult to visualize the facial nerve because this nerve probably was small and adjacent to the wall of the internal auditory canal; the three branches of the VCN were clearly visible. On these images made through the internal auditory canal, the cochlear branch of the VCN was larger than the facial nerve in 12 inner ears, was as large as the facial nerve in five inner ears, and was smaller than the facial nerve in only three inner ears (Fig 2d, 2e).

Patients

In all patients, the T2-weighted spin-echo images of the brain were normal, and the unenhanced and gadolinium-enhanced T1-weighted images of the inner ear helped confirm only the stenosis of the internal auditory canal (five inner ears [three patients]) and the malformations of the osseous and membranous labyrinths (four inner ears [two patients]). However, these inner ear malformations were far easier to visualize on the 3DFT-CISS images. The absence of gadolinium enhancement in the cerebellopontine angle, internal auditory canal, and membranous labyrinth ruled out an acoustic neuroma or acute labyrinthitis as the cause of the deafness or hearing loss. Chronic labyrinthitis was ruled out on CT scans (no calcifications) and on the 3DFT-CISS images (no fibrous obliteration of the intralabyrinthine fluid spaces). Both inner ears were involved in three of the seven patients.

Two patients (three inner ears) had normal osseous and membranous labyrinths but had very narrow internal auditory canals. One of them (patient 2) even had a separate bony canal for the facial nerve, parallel to the internal auditory canal. In these two patients, the VCN was completely absent, and only a facial nerve was visualized on the gradient-echo images (Fig 3).

Associated malformations of the osseous and membranous labyrinths were seen in only two patients (four inner ears [patients 3 and 4]), and in one of them, the internal auditory canal was also stenotic on both sides. These patients had a common VCN, without trifurcation into cochlear, inferior vestibular, and superior vestibular branches (Fig 4).

Three other patients (three inner ears) had normal internal auditory canals and labyrinthine structures. Two of them (patients 5 and 6), however, had a common VCN running toward the vestibular labyrinth, and in both patients the cochlear branch of the VCN was absent (Fig 5). In the other patient (patient 7), hypoplasia of the cochlear branch of the VCN was found (Fig 6). All MR and CT findings are listed in the Table.

DISCUSSION

For years, CT was the best technique to look for congenital malformations of the inner ear in patients with congenital deafness. With the advent of MR imaging, it became possible to exclude abnormality along the acoustic pathway and in the auditory cortex on T2-weighted spin-echo images. Later on, gadolinium-enhanced T1-weighted spin-echo images were used in these patients to exclude acoustic neuromas, labyrinthitis, and other possible pathologic conditions in the cerebellopontine angle, internal auditory canal, and membranous labyrinth (1-5). In recent years, T2-weighted gradient-echo techniques have been used more and more to study the inner ear. These gradient-echo images are needed to evaluate the very small structures of the inner ear and to detect some of the abnormalities in the cerebellopontine angle, internal auditory canal, and membranous laby-
Good gradient-echo images must provide high contrast between the cerebrospinal fluid—intralabyrinthine fluid, nerves, and bone, and the sections must be very thin. In this study, axial 0.7-mm-thick 3DFT-CISS images were used because they provide excellent contrast between the different inner ear structures, and even the cerebrospinal fluid around the brain stem remains completely white on these images (10). Similar results were reported with other gradient-echo techniques, including 3D gradient-recalled acquisition in the steady state (11), contrast material—enhanced fast acquisition in the steady state (12), and 3D fast imaging with steady-state precession (13), and even a fast spin-echo (14) sequence proved to be beneficial in the study of the inner ear. The congenital malformations, detectable with CT, could now also be recognized on these gradient-echo images.

The big advantage, however, was that it now became possible to detect unexpected inner ear malformations in patients with sensorineural hearing loss and/or vertigo by using MR alone, obviating an additional CT study (6).

CT and MR imaging could now be used to confirm the presence of a normal cochlea in candidates for cochlear implantation and to exclude calcifications in the cochlea (at CT) and/or fibrous obliteration in the membranous labyrinth (at MR imaging). In the absence of calcifications and fibrous obliterations, it is possible to introduce a cochlear implant into the cochlea. It is, however, obvious that the next important structure to be checked in these patients is the cochlear branch of the VCN. However, the MR technique must be fine-tuned to be useful for detection of nerve aplasias or hypoplasias. These malformations can be detected only on thin-section T2-weighted gradient-echo images (eg, 3DFT-CISS, etc) or comparable fast spin-echo images.

Visualization of the Nerves in the Internal Auditory Canal

It is crucial to use images with a thickness of 1 mm or less to visualize the facial nerve and the three branches of the VCN separately inside the internal auditory canal. Nerve visualization becomes more accurate when, for instance, 0.7-mm-thick sections are used instead of 1.0-mm sections (10). The four nerves could be visualized in the internal auditory canal on the axial 0.7-mm-thick 3DFT-CISS images in all 20 normal inner ears and on the parasagittal reconstruction images in 19 of them. This proves that in the absence of movement artifacts, in cooperative patients, this technique is reliable enough to demonstrate the four nerves (Fig 2). Verification of the nerves in a second plane makes the technique even more reliable. It can also happen that a nerve is lying against the anterior or inferior wall of the medial part of the internal auditory canal, and in these patients the parasagittal reconstruction images can help in the demonstration of a nerve that is difficult to see on the axial images.

The thickness of the nerves inside the internal auditory canal can be measured, but these nerves are too small to be measured in a reliable way on images with an in-plane resolution of 0.66 x 0.66 mm and a thickness of 0.7 mm. The voxels are nearly isotropic or cubically shaped when these in-plane and section-thickness dimensions are used, and the use of such voxels will result in good quality multiplanar reconstruction images. The diameter of the common vestibular
nerve and its superior and inferior vestibular branches is difficult to evaluate. Sometimes the bifurcation is seen only near the fundus of the internal auditory canal, and in the region just medial to the bifurcation, the nerve then becomes shaped like a figure eight and therefore has a large cranio-caudal diameter. The diameters of the cochlear branch and the facial nerve were easier to depict inside the internal auditory canal on the parasagittal images and were more constant in size. Most frequently, the cochlear branch of the VCN was larger than the facial nerve, although the latter can be as large or even larger. The findings were more constant in the cerebellopontine angle, where the facial nerve and the VCN are found, and the latter was nearly always 1½–2 times larger than the facial nerve and was never smaller. These findings can be used as a reference when the nerves in the cerebellopontine angle and internal auditory canal have to be checked.

Embryologic Development

Aplasia of the complete VCN and aplasia or hypoplasia of its cochlear branch were demonstrated on MR images in seven patients (10 inner ears). These abnormalities occurred isolated or in association with a stenosis of the internal auditory canal and/or congenital malformation of the labyrinth. A possible explanation for these isolated and associated nerve abnormalities can be sought in the embryologic development of the labyrinth and VCN.

The development of the human cochlea starts with the appearance of the otic placode at the third embryonic week. This placode transforms into the otic vesicle that gives rise to the endolymphatic duct, the utricular, saccular, semicircular canals, and cochlea. At 9 weeks, the cochlear windings are fully developed and the appearance of the neural epithelium has started. Neuroblasts of the cochlear ganglion separate from the otic epithelium. Fibers from these ganglion cell bodies grow peripherally back into the otic epithelium and centrally into the brain stem (15). The first afferent fibers entering the undifferentiated otic epithelium are seen at 9–10 weeks (16). In the avian embryo, an abundant neuronal population invades the epithelium (17,18). This is followed by the disappearance of many redundant fibers, which may lead to a 25% reduction in the number of fibers. This reorganization is called “neural stabilization” (17). Although it has been thought that the cochlear development or differentiation depends on the innervation, this has been proved not to be the case. Explants of chicken otic vesicles that lack neural fibers give rise to inner ear sensory structures with normal morphologic features in vitro. This implies that the inner ear development is not dependent on any neuronal trigger or stimulus or trophic effect (19). Also, the differentiation of the hair cells is controlled by location-specific cues that originate in the ear itself. Conversely, the developing inner ear appears to have an important trophic effect on the survival and the cytokidifferentiation of the afferent neurons. The otic vesicle releases a nerve growth factor-like substance that is essential for the survival of the neurones and for “neuronal stabilization” (18).

These data can explain why the VCN or its cochlear branch can be absent in patients with an abnormal or absent cochlea. It is equally conceivable that a disturbance in the trophic effect that the cochlea exerts on the cochlear neurones may result in a well-developed cochlea without a surviving cochlear nerve. The association of an absent VCN and a stenosis of the internal auditory canal is not explained by these findings, but a possible hypothesis is that a normal internal auditory canal is formed only in the presence of a normal nerve.
Therefore, a disturbance in the trophic effect of the cochlea can result in loss of too many or all neuronal fibers, and in these patients, the internal auditory canal formed around the initial labyrinthine structures that can be visualized.

The upper part of the posterior semicircular canal was missing on the adjacent images (not shown) made higher through the inner ear. (e) Axial 0.7-mm-thick 3DFT-CISS image (15/21, 65° flip angle) obtained through the middle part of the left internal auditory canal. Both the facial nerve (black arrows) and the vestibular nerve (large white arrows) can be followed toward the porus of the internal auditory canal where they come close together and touch the posterior lip of the porus. No fluid is found in the cochlear region (thin white arrow). A fluid-filled common cavity (small white arrows) and malformed posterior semicircular canal (arrowhead) are the only labyrinthine structures that can be visualized. The absence of fluid in the normal region of the cochlea (thin white arrow), and a fluid-filled common cavity (small white arrows) posteriorly and a facial nerve (black arrow) can be recognized. (f) Parasagittal 3DFT-CISS reconstruction image obtained at the level of the lateral third of the left internal auditory canal. Near the fundus of the internal auditory canal, the common VCN (white arrow) is still seen instead of the three branches of the VCN that should be seen at this level. The facial nerve (black arrow), seen in the anterior part of the internal auditory canal, has an abnormal low position inside the internal auditory canal. A = anterior, P = posterior.

Classification Proposal

The MR findings in the seven patients with aplasia or hypoplasia of the cochlear branch of the VCN were in accordance with the clinical findings (Table) but as yet cannot be confirmed with surgery or at pathologic examination. The authors believe that the presented cases represent different stages or grades of developmental anomalies of the VCN. According to the CT, MR imaging, and clinical findings and with respect to embryologic knowledge, we propose the following classification.
but these fibers can again disappear when there is a disturbance in the VCN. The absence of a normal VCN might predict that a cochlear branch was initially overgrown with cells from other structures. Therefore, it is advisable to look for evidence of aplasia of the VCN. Partial volume effects cannot explain the disappearance of a nerve in this plane, and in normal inner ears, both the facial nerve and VCN were always visible anterior to the flocculus (Fig 2). Therefore, it is advisable to look for aplasia of the VCN at this level in patients with congenital hearing loss and associated stenosis of one or both internal auditory canals.

In these patients, the findings on the parasagittal 3DFT-CISS reconstruction images made through the cerebellum are very reliable in the exclusion or confirmation of aplasia of the VCN. Partial volume effects cannot explain the disappearance of a nerve in this plane, and in normal inner ears, both the facial nerve and VCN were always visible anterior to the flocculus (Fig 2). Therefore, it is advisable to look for aplasia of the VCN at this level in patients with congenital hearing loss and associated stenosis of one or both internal auditory canals.

The MR demonstration of the absence of a VCN might predict that a cochlear implant will not work. Nevertheless, it could be interesting to perform an electric promontory stimulation test and functional MR imaging of the auditory cortex to exclude the presence of very thin cochlear fibers, too thin to be detected on MR images. However, it seems unlikely that a completely invisible VCN on MR images would possess enough fibers to result in worthwhile hearing.

Type 2: Common VCN with Aplasia or Hypoplasia of Its Cochlear Branch

Type 2A: type 2 in the presence of a labyrinthine malformation.—In two patients (patients 3 and 4), the cochlea and vestibule formed a single, large, fluid-filled cavity, known as a common cavity (20). They also had associated malformations of the semicircular canals (Table). In both patients, the axial gradient-echo images and the parasagittal reconstruction images showed that the cochlear branch of the VCN was absent (Fig 4). As mentioned above, the absence of a normal developing cochlea can impede the survival or cytodifferentiation of the afferent neurons (cochlear branch) and can explain this combination of malformations. The absence of the cochlear branch was initially over-
looked on the axial MR images of one of these patients (patient 3), and at that time, parasagittal reconstruction images were not made. The diagnosis was made only several months later when the images were reviewed for a study on congenital malformations. In the meantime, electric stimulation of the left ear had elicited clear subjective hearing sensation, and as a consequence, the ear surgeon (F.E.O.) placed a cochlear implant (LAURA; Antwerp Bionic Systems, Antwerp, Belgium) into the inferior part of the common cavity, which looked like a basal turn of the cochlea (Fig 4c). The postoperative electrodacoustic test was within the normal range and stable throughout the 6-month postoperative period, and the child showed clear signs of auditory progress (voice control, auditory behavior). The most probable explanation or hypothesis is that the single common VCN probably carries some fibers projecting into the auditory cortex. These results make it, of course, difficult to make a decision in the second patient (patient 4), with similar malformations on both sides and bilateral stenoses of the internal auditory canal. The results of the electric promontory stimulation can guide the surgeon, and in the future, functional MR imaging of the auditory cortex might become the technique of choice. Only this imaging modality has the potential to demonstrate whether "cochlear fibers" connect an abnormal labyrinth with the brain stem and auditory cortex, even in the absence of a cochlear branch of the VCN.

Type 2B: type 2 in patients with a normal labyrinth—Three patients presented with completely normal labyrinths and internal auditory canals. The axial and parasagittal gradient-echo images showed an absent cochlear branch of the VCN in two patients (patients 5 and 6), and in these patients, the common vestibular nerve could be followed to the fundus of the internal auditory canal without a clear bifurcation or a very late (not visible on MR images due to partial volume effect with the fundus) bifurcation in a superior and inferior vestibular nerve (Fig 5). In a third patient (patient 7), a very thin cochlear branch could be seen on the axial and parasagittal gradient-echo images (Fig 6). In these three patients, a disturbance in the trophic effect (nerve growth factor) that the cochlea exerts on the cochlear neurones could once again explain the hypoplasia or complete absence of the cochlear branch of the VCN. Of course, these findings had no clinical consequences as these three patients had a normal functioning contralateral ear; nevertheless, the MR findings correlated well with the unilateral hearing loss. However, in patients with normal labyrinths and bilateral aplasias of the cochlear branch of the VCN, electric promontory stimulation and/or functional MR imaging should be used as a selection tool for cochlear implant candidacy.
The absence of the cochlear branch can be recognized on thin-section gradient-echo images, although once again parasagittal reconstruction images are mandatory to confirm the findings seen on the axial images. The danger exists that the cochlear branch lies adjacent to the floor of the internal auditory canal and is therefore not seen on the axial gradient-echo images due to partial volume effects. The parasagittal plane is better suited to study this region and can be used to exclude partial volume effects. The diagnosis of hypoplasia of the cochlear branch is more difficult. In three of the 20 normal inner ears, the cochlear branch of the VCN inside the internal auditory canal was smaller than the facial nerve. But even in these cases, the diameters of both nerves were nearly the same. In patients 7, the diameter of the cochlear branch was much smaller than the diameter of the facial nerve, whereas on the opposite normal side the cochlear branch was much larger than the facial nerve.

Type 3?: Presence of a Common VCN with Aplasia or Hypoplasia of the Vestibular Branch(es)

Theoretically, a third type of VCN aplasia or hypoplasia with only absence of the vestibular branches but with a normal cochlear branch could be conceived. To our knowledge, such a type of VCN malformation has not yet been detected on MR images. Since the vestibule develops at an earlier stage than the cochlea, one may expect that a vestibular malformation will always lead to an associated cochlear malformation. In such a case, a type 1 aplasia would be the result. Therefore, the authors speculate that an isolated aplasia of the vestibular branch or type 3 aplasia of the VCN may not exist.

References