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Anomalous Facial Nerve Canal with Cochlear Malformations

Laura Vitale Romo and Hugh D. Curtin

BACKGROUND AND PURPOSE: Anteromedial “migration” of the first segment of the facial nerve canal has been previously identified in a patient with a non-Mondini-type cochlear malformation. In this study, several patients with the same facial nerve canal anomaly were reviewed to assess for the association and type of cochlear malformation.

METHODS: CT scans of the temporal bone of 15 patients with anteromedial migration of the first segment of the facial nerve canal were collected from routine departmental examinations. In seven patients, the anomalous course was bilateral, for a total of 22 cases. The migration was graded relative to normal as either mild/moderate or pronounced. The cochlea in each of these cases was examined for the presence and size of the basilar, second, and apical turns. The turns were either absent, small, normal, or enlarged. The CT scans of five patients with eight Mondini malformations were examined for comparison.

RESULTS: The degree of the facial nerve migration was pronounced in nine cases and mild/moderate in 13. All 22 of these cases had associated cochlear abnormalities of the non-Mondini variety. These included common cavity anomalies with lack of definition between the cochlea and vestibule (five cases), cochleae with enlarged basilar turns and absent second or third turns (five cases), and cochleae with small or normal basilar turns with small or absent second or third turns (12 cases). None of the patients with Mondini-type cochlear malformations had anteromedial migration of the facial nerve canal.

CONCLUSION: Anteromedial migration of the facial nerve canal occurs in association with some cochlear malformations. It did not occur in association with the Mondini malformations. A cochlea with a Mondini malformation, being similar in size to a normal cochlea, may physically prohibit such a deviation in course.

In a previous report, an anomalous course of the first segment of the facial nerve canal was described in a patient with a malformed cochlea (1). The location of the labyrinthine segment of the nerve was anteromedial to that of the normal position. The cochlea was represented by a bud-like diverticulum from the vestibule, and the upper two turns had not developed. Because of the intimate relationship between the development of the facial nerve with the development of the cochlea, the malformation of the cochlea in this case was postulated to be the cause of the anomalous course of the facial nerve.

In this study, we chose to examine other cases of anomalous first segments of the facial nerve to

learn whether the association with a particular cochlear malformation, as previously described, was an isolated phenomenon or a consistent and predictable finding.

Methods

The study included CT scans of the temporal bone of 15 patients identified as having anomalous first segments of the facial nerve canal. In seven of these 15 patients, the anomaly was bilateral, yielding a total of 22 cases of anomalous facial nerves. These cases were collected over a few years from two main institutions. They were collected in a random manner from teaching files and daily case material. The patients had undergone scanning for a history of sensorineural hearing loss. In all cases, CT was performed using bone algorithm and 1.0-mm contiguous section thickness and was angled to the superior or inferior orbitomeatal lines.

The first segment of the facial nerve canal, otherwise called the labyrinthine segment of the fallopian canal, as it originates from the internal auditory canal, was recorded in each case as normal or anteromedial to that of normal. There were two possible degrees of anteromedial displacement of the fallopian canal: mild/moderate and pronounced. The cases of mild and moderate displacement were placed together because it was difficult to visually distinguish between the two but it was

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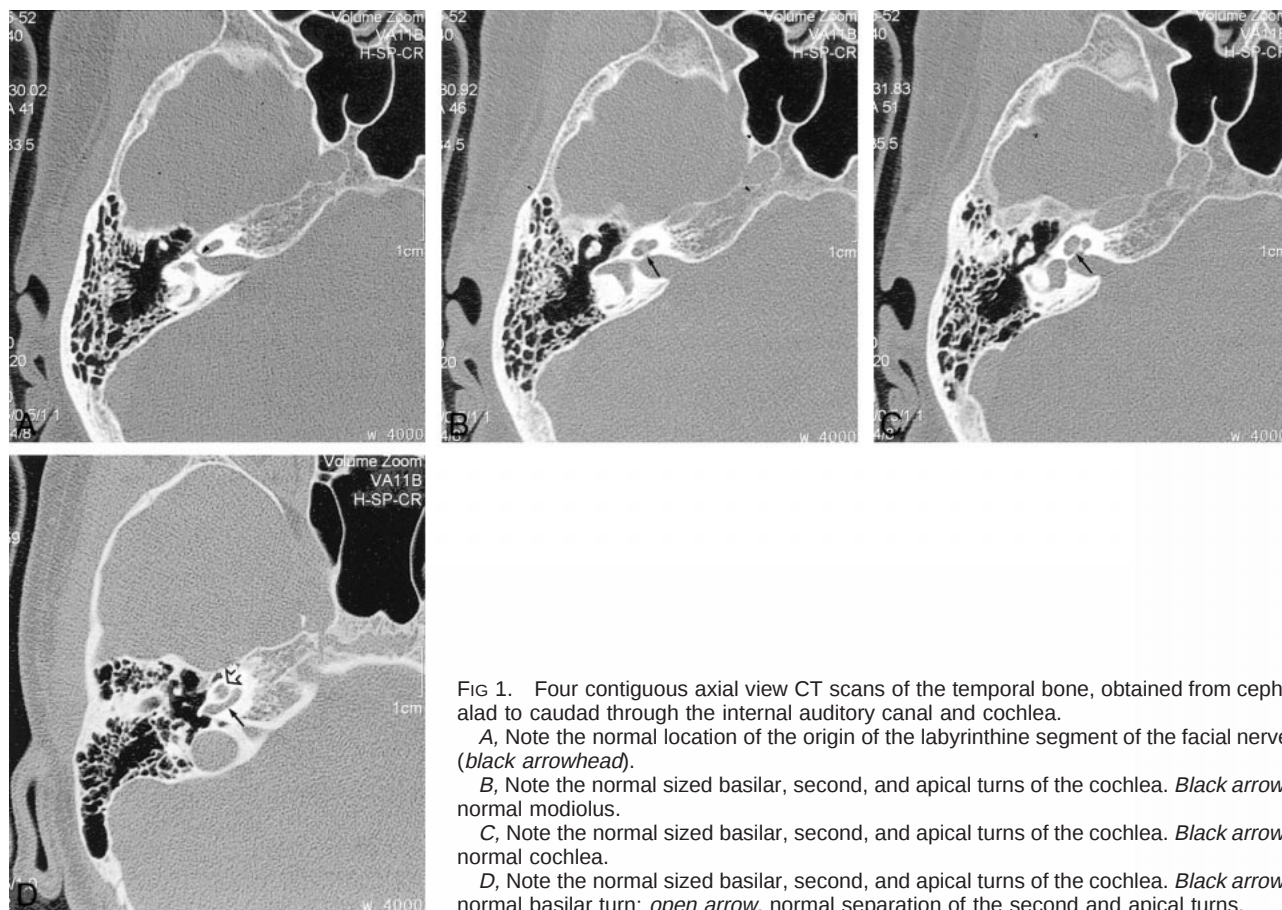


FIG 1. Four contiguous axial view CT scans of the temporal bone, obtained from cephalad to caudad through the internal auditory canal and cochlea.

A, Note the normal location of the origin of the labyrinthine segment of the facial nerve (black arrowhead).

B, Note the normal sized basilar, second, and apical turns of the cochlea. Black arrow, normal modiolus.

C, Note the normal sized basilar, second, and apical turns of the cochlea. Black arrow, normal cochlea.

D, Note the normal sized basilar, second, and apical turns of the cochlea. Black arrow, normal basilar turn; open arrow, normal separation of the second and apical turns.

feasible to separate them as a group from those cases with pronounced displacement. A pronounced degree of migration was a substantial displacement with the facial nerve canal exiting the internal auditory canal significantly more medially, with corresponding anteromedial positioning of the geniculate turn of the canal. Mild/moderate degrees of displacement were less than pronounced degrees of displacement, sometimes being only slight and sometimes approaching, but not quite reaching, the severity of pronounced displacement.

The tympanic segments of the facial nerve were measured in 14 of the 22 cases. The tympanic segments were not measurable in the remaining eight cases because the data were not available for review. The segments were measured relative to a scale depicted on the appropriate scans. The segments were measured to assess for any increase in length related to a more anterior and medial location of the geniculate ganglion. The measurements were compared with an established normal length of 12 mm (2).

Widening of Bill's bar (the thin bony crest separating the exit apertures of the facial and superior vestibular nerves) was also recorded, if present. The degree of widening was estimated using the same degrees as those for facial nerve canal displacement. All evaluations were made by consensus between the two radiologists.

Figure 1 shows CT scans of a normal temporal bone. The cochleae in each of the cases of facial nerve displacement were examined for the presence and size of the basilar, second, and apical turns. The turns were categorized as either absent, small, normal, or enlarged (Figs 2 and 3). The absence of all cochlear turns with the presence of only a rudimentary bud-like cavity connected to the vestibule was categorized as a common cavity (Fig 4).

The previously reported case of anteromedial migration of the fallopian canal occurred in conjunction with severe dys-

plasia of the cochlea. There was no normal basilar turn, and a sac-like diverticulum represented the cochlea. Five patients with eight typical Mondini-type cochlear malformations were also examined for comparison. These patients had either classic Mondini malformations with normal basilar turns and a common cloaca or cavity in place of the second and apical turns or slightly less severe modiolar deficiencies closely approximating true Mondini malformations. The course of the first segment of the facial nerve canal in each case was recorded in the same fashion as the others. The lengths of the tympanic segments of the facial nerves were also recorded and again compared with a normal length of 12 mm.

Results

Of the 22 anomalous facial nerves, the degree of anteromedial displacement was pronounced in nine (Figs 2 and 4) and mild/moderate in 13 (Fig 3) (see Fig 1 as a normal reference). The 14 measurable tympanic segments of the anomalous facial nerves were all longer than the established normal length of 12 mm. In two cases, the tympanic segment measured 22 mm; in two cases, it measured 16 mm; in nine cases, it measured 15 mm; and in one case, it measured 14 mm. No change in the position of the mastoid segments of the facial nerves was appreciated in these cases. Widening of Bill's bar was pronounced in six cases and was mild/moderate in 15 (Table). In one patient with minimal anteromedial facial nerve displacement, there was no appreciable widening of Bill's bar. All 22 cases of facial

FIG 2. Cochleae in cases of facial nerve displacement were examined for presence and size of basilar, second, and apical turns.

A and B, Axial view CT scans of the temporal bone show pronounced antero-medial facial nerve migration bilaterally (*black arrowheads*), with pronounced widening of Bill's bar (*black arrow*).

C, Axial view CT scan of the temporal bone shows the right-sided hypoplastic cochlea with small first turn (*black arrowhead*). The left-sided cochlea was also hypoplastic and is not shown. (Same magnification as that shown in Fig 1.)

D, Axial view CT scan of the temporal bone shows the right-sided hypoplastic cochlea with small second turn (*black arrowhead*). The left-sided cochlea was also hypoplastic and is not shown. (Same scale as that shown in Fig 1.)

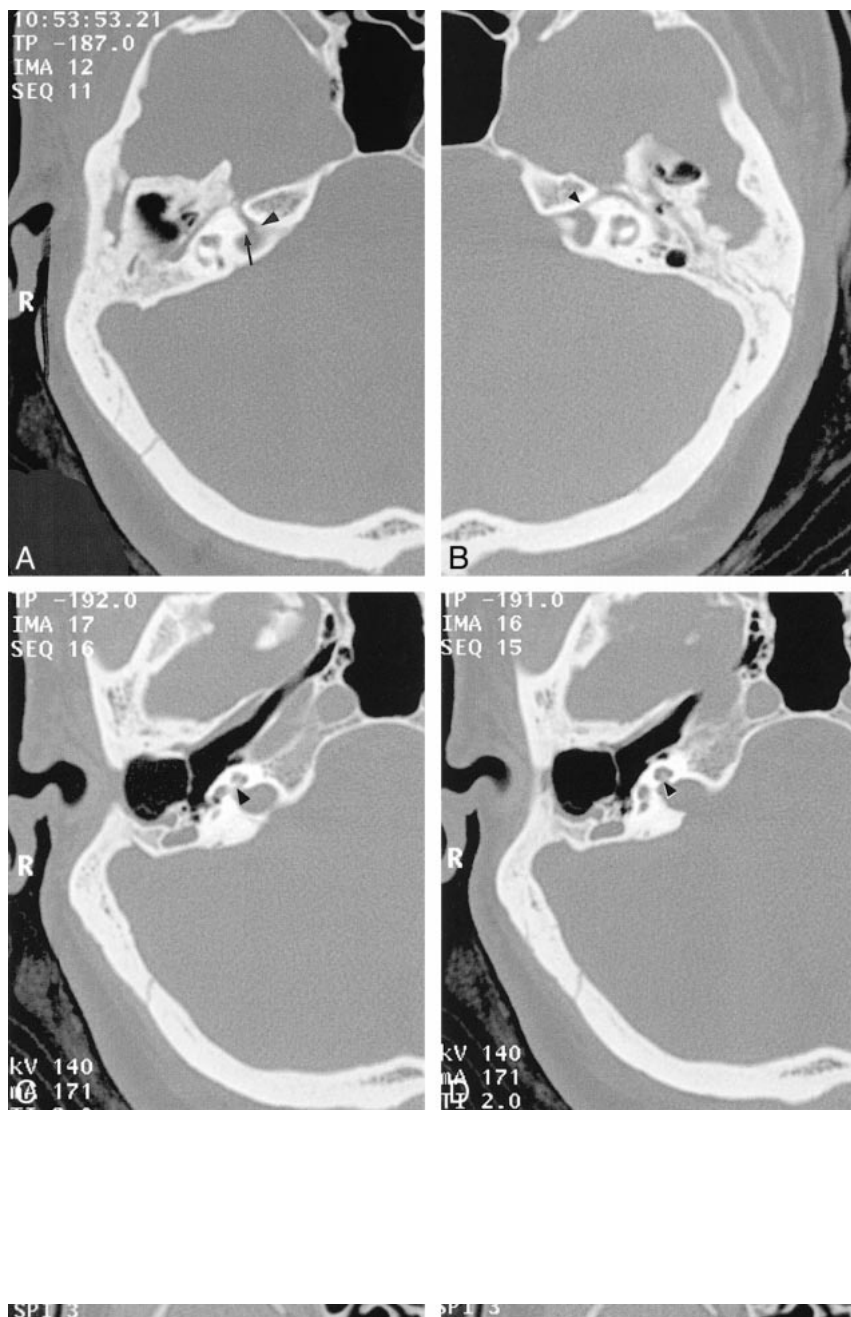


FIG 3. Cochleae in cases of facial nerve displacement were examined for presence and size of basilar, second, and apical turns.

A, Axial view CT scan of the temporal bone shows mild/moderate migration of the right facial nerve (*black arrowhead*) and mild/moderate widening of Bill's bar (*small black arrow*).

B, Basilar turn of the cochlea was normal, second turn was small, and third turn was absent (*large black arrow*). (Same scale as that shown in Fig 1.)

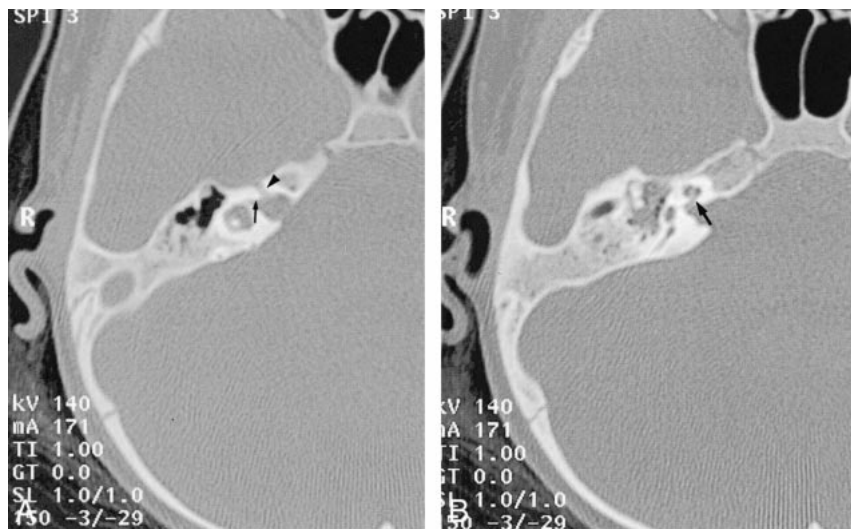




FIG 4. Absence of all cochlear turns with presence of only rudimentary bud-like cavity connected to vestibule was categorized as common cavity.

A and B, Axial view CT scans of the temporal bone show pronounced antero-medial facial nerve migration bilaterally (black arrowheads), with widening of Bill's bar (small black arrow) in a patient with a common cavity deformity bilaterally (large black arrows). (Same scale as that shown in Fig 1.)

Facial nerve migration and non-Mondini-type malformations

	Cochlear malformations*					
	CC	1E 2A 3A	1N 2S 3A	1S 2S 3A	1S 2A 3A	Total
Facial Nerve Migration:						
Pronounced	4	1		4		9
Mild/Moderate	1	4	3	4	1	13
Bill's Bar Widening:						
Pronounced	4			2		6
Mild/Moderate	1	5	2	6	1	15

* CC = common cavity; 1 = first cochlear turn, 2 = second cochlear turn, 3 = third cochlear turn; E = enlarged; A = absent; N = normal; and S = small.

nerve displacement had cochlear abnormalities and sensorineural hearing loss. None of the malformations were of the classic Mondini variety. In five cases, there were common cavities (Fig 4); in five, there were enlarged basilar turns with absent second and third turns; in three, there were normal basilar turns with small second and absent third turns (Fig 3); in eight, there were small basilar and second turns with absence of the third turns (Fig 2); and in one, there was a small basilar turn and absent second and third turns. Of the nine cases of pronounced facial nerve displacement, four had common cavities, four had small basilar and second turns with absent third turns, and one had an enlarged basilar turn with absent second and third turns. In the 13 cases of mild/moderate migration, four had small basilar and second turns with absent third turns, four had enlarged basilar turns with absent second and third turns, three had normal basilar turns with small second and absent third turns, one had a common cavity, and one had a small basilar turn with absent second and third turns. In eight of the 22 cases of migration, there were associated abnormalities of the semicircular canals, most commonly the lateral semicircular canals, either enlarged or abnormally small. Two of the eight

cases also had enlargement of the superior semicircular canals, and three had enlarged vestibules. In only one of the 22 cases was there an enlarged vestibular aqueduct, measuring > 1.5 mm.

None of the eight Mondini (Fig 5) or Mondini-type malformations had evidence of anteromedial facial nerve displacement, lengthening of the tympanic segments of the facial nerves, or widening of Bill's bar (Fig 3). In two cases, the tympanic segments of the facial nerve measured 12 mm; in four cases, it measured 11 mm; and in two cases, it measured 10 mm. All these malformations had associated enlargement of the vestibular aqueduct, measuring > 1.5 mm.

Discussion

It is well known that segments of the facial nerve canal will be anomalous in course in the setting of maldevelopment of various adjacent structures. Abnormal development of first and second branchial arch derivatives, including the bony wall of the facial nerve canal, the stapes, the styloid process and/or external auditory canal, have been associated with an anomalous course of the tympanic and mastoid segments of the facial nerve, the nerve of the second branchial arch (3–10). Normally, the facial nerve is located posterior to the structures formed from Reichert's cartilage, the cartilage anlage of the second branchial arch (10). The long, convoluted course of the facial nerve through the temporal bone is thought to be dependent on the normal development of the bony structures derived from Reichert's cartilage. Thus, with abnormal formation of, for example, the stapes crura and lack of fusion of the crura to the footplate, the tympanic and mastoid segments of the facial nerve may assume a more anterior and inferior course, possibly taking a more direct route or "migrating" anteriorly to the end organ, the muscles of facial expression (5).

This theory of anterior migration of the tympanic and mastoid segments of the facial nerve was first

FIG 5. CT scans of Mondini malformations.

A and B, Axial view CT scans obtained through the temporal bone show Mondini malformations of the cochlea bilaterally, with lack of separation of the second and third turns (*black arrowheads*).

C and D, Axial view CT scans of the same patient, obtained more cephalad, show normal origins of the facial nerves from the internal auditory canals bilaterally (*black arrowheads*). Note also prominent vestibules (*large black arrows*) and enlarged right vestibular aqueduct (*small black arrow*). (Same scale as that shown in Fig 1.)



proposed by Durcan et al (5) in 1967. To reiterate, they proposed that lack of normal development of the stapes and bony wall of the tympanic segment of the facial nerve from Reichert's cartilage allows the facial nerve to migrate anteriorly or "follow its natural tendency to move forward toward its target innervation," the muscles of facial expression. They used the difference in embryologic development of the right and left recurrent laryngeal nerves, branches of the vagus nerves that supply the sixth branchial arches, as supportive evidence. Normally, the left recurrent laryngeal nerve has a longer course than the right recurrent laryngeal nerve. The left is pulled inferiorly to the level of the ductus arteriosus, the persistent distal portion of the sixth aortic arch, by the development of the heart. However, on the right, the fifth and sixth aortic arches degenerate so that the nerve does not travel as inferiorly. The right recurrent laryngeal nerve hooks around the fourth aortic arch, taking a more direct route to the larynx.

From these two examples of variation in cranial nerve course related to variation in the development of surrounding structures, we have extrapolated a similar proposal with regard to the labyrinthine segment of the facial nerve. Anteromedial displacement of the labyrinthine segment of the facial nerve canal may be due to a malformation of the cochlea akin to anteroinferior migration of the tympanic and mastoid segments with malformation of a second arch cartilage derivative and superior displacement of the right recurrent laryngeal nerve with degeneration of the last two aortic arches. In our cases, the facial nerve would be allowed to take a more direct course toward the pterygopalatine fossa. The lengthening of the tympanic segment of the facial nerve might be taken as direct objective evidence of this phenomenon and the widening of Bill's bar as, perhaps, indirect perceptual evidence.

It is pertinent to this discussion to briefly review the embryologic development of the facial nerve and cochlea. The seventh and eighth nerves form

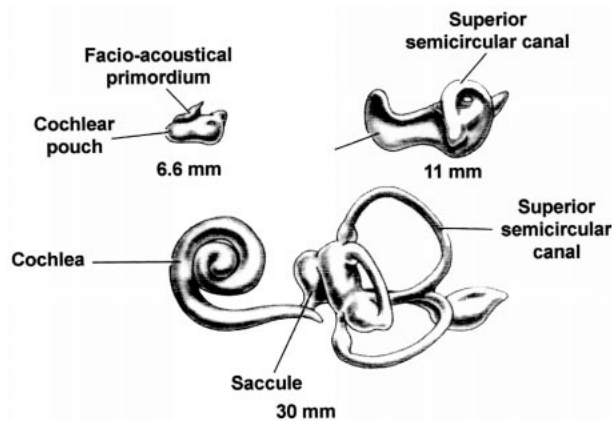


FIG 6. Drawing adapted from Streeter (11) and reprinted from Curtin et al (1) shows development of the labyrinth from the otocyst. The cochlear pouch develops with the upward curve as the cochlear duct curls into the adult form.

from the facioacoustic primordium immediately rostral to the otocyst from which the cochlear duct develops (Fig 6). The facioacoustic primordium begins to separate into distinct seventh and eighth nerves when the crown-rump length of the fetus is approximately 8 to 10 mm. The nerves are completely separate by a crown-rump length of 14 mm. At approximately the same time, the cochlear duct begins to develop, first as a small pouch from the otocyst ventrally and then coiling rostrally toward the facioacoustic primordium (11). The association of the geniculate to the vestibuloacoustic ganglion and the coiling of the cochlear duct explain the normal location of the geniculate ganglion, namely in the bony substance just above the basilar turn of the cochlea and between the superior semicircular canal and the cochlea proper (1).

In the previous radiologic case report of labyrinthine facial nerve migration, it was speculated that the cochlear malformation was causative (1). The cochlea, in this case, had no basilar turn and only a sac-like cochlear remnant connected to the vestibule. As such, the cochlear duct did not fully develop, possibly allowing the facial nerve to take a more direct, anteromedial route from the apex of the internal auditory canal to the pterygopalatine ganglion (1).

The question remains why some cochlear malformations, namely non-Mondini malformations, are associated with facial nerve migration and why Mondini malformations apparently are not. The Mondini deformity was described by Carlo Mondini in 1791 from an autopsy specimen in a deaf boy (12). The cochlea was described as not having the normal "two and one-half spirals" but rather one and one-half spirals with the "final spiral at the apex missing" and ending "in a large hollow corresponding to the cavity of the final spiral" (12, 13). Since Mondini's original description, several others have presented similar cases (13–17) with cochleae possessing only one and one-half turns. In 1987, Jackler et al (18) proposed a classification

system for congenital malformations of the inner ear into which the Mondini malformation was placed. They described cochlear malformations as a spectrum of partition defects. In descending order of severity, these included complete labyrinthine aplasia (Michel's deformity), cochlear aplasia, common cavity with lack of separation of the cochlea and vestibule without internal architecture, cochlear hypoplasia, and finally, an incomplete partition deformity with a small cochlea with incomplete or absent interscalar septum. This spectrum of abnormalities was postulated to result from arrest of cochlear development at different embryonic stages. In this scheme, the Mondini malformation would result from arrested development at the latter end of embryogenesis and represent an incomplete partition defect. However, as the authors attested, all cochlear malformations could not fit into this classification system. In a series of patients with hearing impairment reviewed by Valvassori et al (19), 50% showed only a reduction in the size of the cochlea and not in the number of coils. These cases were labeled "dwarf cochlea."

In our series, some of the non-Mondini type cochlear malformations approximated the classifications proposed by Jackler et al (18) of the common cavity and cochlear hypoplasias with absent second and third turns and others more closely resembled the description presented by Valvassori et al (19) with close to the normal number of turns but small overall size. In those cases with the common cavity or hypoplastic cochlea configuration, the rostral growth of the cochlea may have been either absent or stunted and, as such, allowed the anteromedial facial nerve migration. Even a delay in the rostral growth of the cochlea might permit the facial nerve to escape anteriorly. In those patients with the microcochlea, the rostral growth of the cochlea would have occurred but would have been less pronounced than usual or stunted. In this case, either the final structure was too small to block migration or perhaps the time of formation was also delayed, with the rostral growth occurring after the nerve had already separated and migrated. These speculations are all based on the hypothesis that a normal cochlea may behave in a manner similar to that of Reichert's cartilage derivatives, which prevent migration of the facial nerve and tympanic facial nerve canal anteriorly (1, 5).

In the Mondini malformation, although there is a decreased number of turns, the "large hollow" or sac-like remnant of the second and apical turns presumably forms at the normal time, as evidenced by the relatively normal basilar turn. This sac-like representation of the second and apical turns may actually represent an enlargement of the cochlear duct and may be large enough to act as a physical barrier, much like a normal cochlea, to facial nerve migration. We know of no literature describing an association between Mondini malformations and facial nerve canal anomalies. Similarly, facial nerve migration would not be expected in even

more subtle cochlear abnormalities, such as modular deficiencies (20). In these cases, the rostral growth of the cochlear duct would occur, with the cochlear abnormality occurring at a slightly later stage of development.

The degree of facial nerve migration in our cases ranged from mild/moderate to pronounced. Most of the pronounced cases (eight of nine cases) had common cavities or small first and second turns with an absent third turn. The cases of mild/moderate migration were more evenly distributed among the different types of cochlear abnormalities. Because all the cochlear malformations could be classified as more severe than the Mondini malformation and represent a spectrum of lesser development and hypoplasia relative to the Mondini malformation, it is not clear why the degree of migration varied. It may be secondary to the variability in time at which the seventh nerve completely separates from the eighth nerve. If this separation occurs closer to the 8- to 10-mm fetal length, more growth or migration might be expected than if it occurs closer to the 14-mm length, with cochlear development beginning in the middle of this interval at 11 mm.

Conclusion

Anteromedial displacement of the facial nerve may occur in association with some cochlear anomalies. This displacement did not occur in association with the Mondini malformations. The anteromedial growth or migration may be allowed by a lack of or an impairment of the normal rostral growth of the cochlea during the 9- to 13-mm stage of embryonic development.

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