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Abnormalities of the Facial Nerve in Temporal Bones With Inner Ear Malformations

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Keywords: cochlear implantation | facial nerve | inner ear malformations | temporal bone | temporal bone pathology

ABSTRACT

Background: Bony inner ear malformations (IEMFs) account for ~20% of congenital sensorineural hearing loss, often requiring cochlear or auditory brainstem implants. To ensure safe cochlear implantation in patients with IEMFs, understanding their anatomical features, particularly related to the facial nerve (FN), is crucial.

Methods: We examined 28 TBs obtained from donors with bony IEMFs. We classified cochlear and vestibular malformations and analyzed several anatomical features, including the diameter of the internal auditory canal (IAC), the angle of the first genu, the relationship of FN with the oval window (OW), the facial recess (FR), and the overall development of the FN.

Results: Among the TBs, 5 (17.8%) were cochlear hypoplasia-type II, 20 (71.4%) were cochlear hypoplasia-type III, 2 (7.1%) were incomplete partition-type II, and 1 (3.5%) had an isolated vestibular malformation. The IAC diameter was narrow in 2 of 26 TBs (7.7%). The first genu angle was obtuse or perpendicular in 14 of 20 TBs (70.0%). The FN was abnormally located in 8 of 27 TBs (29.6%). The FR was narrow (< 2.5 mm) in 16 of 27 TBs (59.3%). Additionally, the FN was hypoplastic in 18 of 28 TBs (64.3%).

Conclusion: Our study revealed a high prevalence of FN abnormalities among patients with IEMFs. Some of these abnormalities could pose significant challenges when using the traditional FR approach for cochlear implantation. Our findings underscore the importance of thoroughly evaluating the FN course preoperatively to mitigate the risks of surgical complications.

Evidence Level: N/A.

1 | Introduction

Congenital hearing loss is attributed to bony inner ear malformations (IEMF) in one out of every five cases [1, 2]. In children with severe or profound hearing loss, timely cochlear implantation (CI) is crucial to facilitate adequate language acquisition and learning [3–5]. Until the early 2000s, CI in children with IEMFs was discouraged due to concerns regarding surgical complexities and postoperative outcomes [6]. With recent advancements in imaging equipment and the refinement of surgical techniques, CI surgery is now widely accepted as the standard of care for most cases involving IEMFs [6–8].

CI in cases with IEMFs often involves a higher incidence of facial nerve (FN) anomalies [9–11]. Previous reports described that several middle ear anatomical landmarks, such as the promontory

and the round window (RW) membrane, are obscured in patients with IEMFs, therefore increasing the risks of FN injury [2, 12, 13]. Other abnormalities, including abnormal FN course, narrowing of the facial recess (FR), small and hypoplastic mastoids, among others, further increase the risks of surgical complications [9, 14–16]. FN injuries can occur in these cases regardless of the approach used (transmastoid, transmeatal, or fossa media) or the use of a FN monitor. While the frequency of intraoperative FN injury in cases of IEMF is yet to be determined, many studies have reported the risk of severe damage to the main trunk of the FN during surgery [2, 9, 11, 14–16]. Furthermore, postoperative FN stimulation symptoms are observed at a higher rate in CI surgeries for IEMF cases compared to cases with normal inner ear structures [17].

Despite remarkable advancements in imaging diagnostics and surgical techniques, there are a limited number of anatomical analyses and reports based on histological aspects of IEMFs cases using human temporal bone (TB) specimens. Otopathology research is crucial in advancing the understanding of ear surgeries by offering microscopic insights not attainable through radiological imaging alone. Qualitative and quantitative evaluations of middle and inner ear structures from tissue specimens of IEMFs cases are valuable for bridging the gap between otopathology and clinical aspects such as imaging diagnostics and surgical techniques.

In this study, we aimed to conduct a qualitative and quantitative evaluation of FN anomalies and the surrounding inner and middle ear structures by evaluating human TB specimens from cases with IEMFs. This study pioneers the examination of histopathology findings related to FN in patients with IEMFs, filling a critical gap in the existing literature.

2 | Materials and Methods

We selected specimens from donors with IEMFs from the collection of human TBs of Paparella Otopathology and Pathogenesis Laboratory at the University of Minnesota. This study was approved by the University of Minnesota Institutional Review Board (0206 M26181). TBs in the archived collection had previously been removed at autopsy, fixed in 10% buffered formalin, and serially sectioned in the horizontal plane at a thickness of 20 μm. Every 10th section was stained with hematoxylin–eosin, mounted on a glass slide for light-microscopic observation, and used in this study. Slides were evaluated with light microscopy.

We selected temporal bones with IEMFs by using the classification and criteria proposed by Sennaroğlu and Bajin [18]. Briefly, the classification system categorizes IEMFs into eight distinct types, including complete labyrinthine aplasia, common cavity deformities, cochlear hypoplasia types I–III, and incomplete partition types I–III. These classifications are based on radiological imaging features, particularly differences in the modiolus, interscalar septum, and cochlear structures. From the 2000+ temporal bones available in our collection, we selected 38 TBs that fulfilled the criteria. We excluded 10 TBs that had severe processing artifacts that prevented a full analysis of the FN. Consequently, the final study group included 28 TBs collected from 18 human donors (10 from male subjects and 15 from female subjects). The gender of the remaining 3 subjects could not be identified due to lack of data in the patient files. The donors' age range was from 2 days to 90 years, with a mean age of 14.99 ± 28.85 (SD) years.

From the 28 selected TBs, 5 (17.8%) were classified as cochlear hypoplasia-type II (CH-II), 20 (71.4%) as cochlear hypoplasia-type III (CH-III), 2 (7.1%) as incomplete partition-type II (IP-II, Mondini deformity), and 1 (3.5%) was classified as an isolated vestibular malformation (no cochlear malformation). Figure 1A–C shows the representative cochlear malformation findings for each IEMF case in this study.

To evaluate the most prominent anatomical features and potential factors associated with abnormalities of the FN, we conducted a series of histopathological analyses and measurements of the mastoid process, internal auditory canal (IAC), and associated structures.

2.1 | Measurement of Various Inner Ear Structures

All histopathologic evaluations and measurements of the specimens were performed using light microscopy (Nikon Eclipse

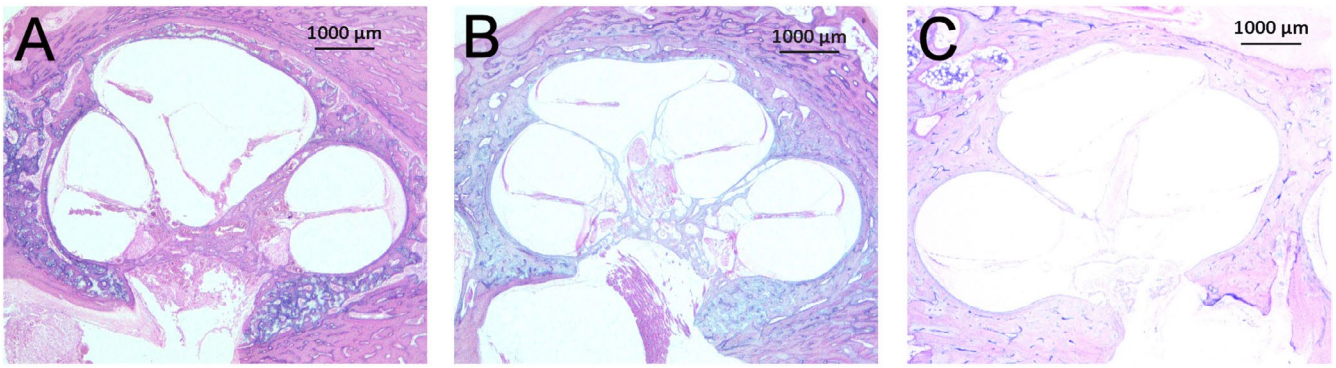

FIGURE 1 | Representative horizontal sections of the human temporal bones with inner ear malformations: (A) Cochlear hypoplasia type II (CH-II). In CH-II, while the bony contour is normal, the bony cochlear modiolus is absent, and there is a wide communication with the internal auditory canal. (B) Cochlear hypoplasia type III (CH-III). In CH-III, the cochlear contour resembles normal, but the bony cochlear modiolus is short, and it is generally smaller with fewer turns. (C) Incomplete partition type II (IP-II, Mondini's deformity). In IP-II, the bony septum is present only in the basal turn of the cochlea, and it is absent in the upper turns. The cochlear modiolus is also present only in the basal turn and is absent in the upper turns. The middle and apical turns of the cochlea appear to be fused into a sac-like structure.

E200—Nikon Co; Tokyo, Japan). The images were captured using the Ds-Fi2 camera (Nikon Co, Tokyo, Japan) (attached to the light microscope) at magnifications from ×10 to ×40 and calculated using the ImageJ software (U.S. National Institutes of Health, Bethesda, Maryland, USA).

2.2 | Development of Mastoid

Mastoid pneumatization was assessed through light microscopy of histological sections. Temporal bones with extensive air cells encompassing the antrum and adjacent mastoid areas were considered well-developed (Figure 2A), whereas those predominantly filled with bone tissue or bone marrow were classified as hypo-developed (Figure 2B). Cases with localized variations (e.g., poorly pneumatized mastoid tips) were categorized based on the overall extent of pneumatization within the mastoid.

2.3 | Internal Auditory Canal

The length of the IAC was primarily measured using Bill's bar [19], a bony prominence at the branching point of the FN and the superior vestibular nerve. To ensure consistent measurements, particularly given the potential deformities associated with IEMFs, we selected the section where the FN fibers, superior vestibular nerve fibers, and their branching were most clearly identifiable. The most prominent point of the bottom of the internal auditory meatus at that level was defined as the apex of Bill's bar or an equivalent vertical crest. The length was then measured as the straight-line distance from this apex to the center of the opening into the middle cranial fossa. The diameter was measured using a perpendicular line passing through the midpoint of the line used to measure the length. Following previous reports [1, 20], the IAC was considered "narrow" if the diameter was <2.0 mm, and "short" if the length was <5 mm. (Figure 3A–C).

2.4 | Facial Nerve

We studied the whole course of the FN within all intratemporal segments (IAC, labyrinthic, tympanic, and mastoid segments). The position of the nerve in relation to the anatomical landmarks was evaluated in the following portions:

a. The First Genu: To ensure thorough examination of the FN's path through its labyrinthine and horizontal segments, commonly referred to as the first genu, we conducted an evaluation to determine if the angle of the first

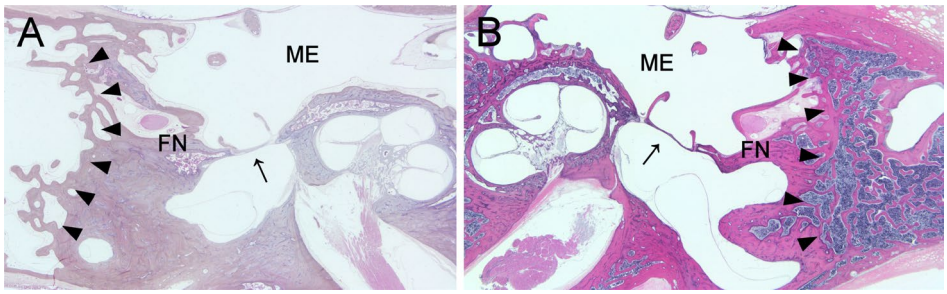

FIGURE 2 | Arrow: Stapes arrow head: Outline of the mastoidRepresentative examples of mastoid development. (A) Well-developed mastoid with robust pneumatization and clearly defined air cells. (B) Hypo-developed mastoid predominantly filled with bone and bone marrow, exhibiting significantly limited pneumatization and smaller, less distinct air cells.

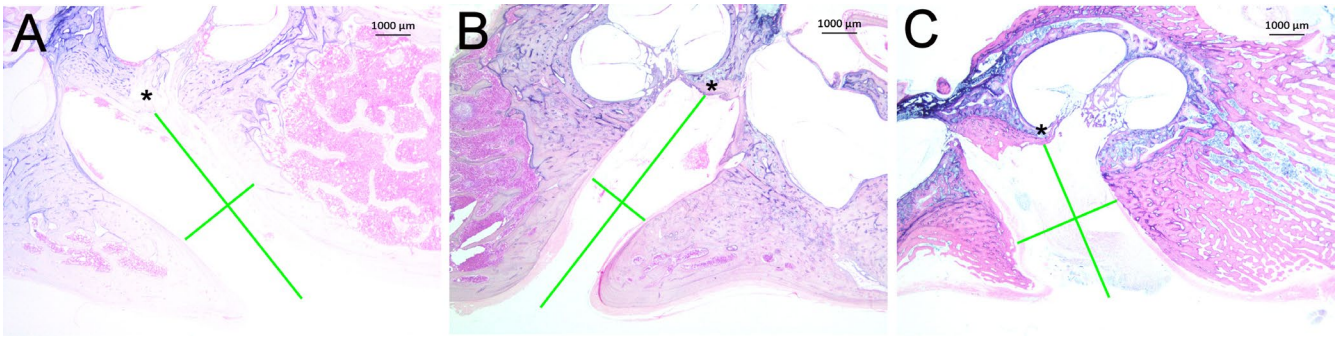

FIGURE 3 | Star: Apex of Bill's bar or the most prominent point of the bottom of the internal auditory meatus at that level where the branching of the FN and superior vestibular nerve can be identified most clearly evaluation of the diameter and length of the internal auditory canal. The longitudinal axis line from the star (*) to the center of the opening of the middle cranial fossa represents the length of the internal auditory canal. The other one indicates the diameter of the internal auditory canal (Interosseous distance) and is perpendicular to the longitudinal axis line and goes through the midpoint of it. (A) This specimen is from one donor with cochlear hypoplasia type II. In this case, the IAC has a normal diameter (≥ 2.0 mm) and length (≥ 5.0 mm). (B) This specimen is from one donor with cochlear hypoplasia type III. In this case, the IAC has a narrow diameter (1.9 mm) and length (≥ 5.0 mm). (C) This specimen is from one donor with cochlear hypoplasia type III. In this case, the IAC has a normal diameter (≥ 2.0 mm) and a short length (3.8 mm).

genu approximated the expected acute angle of ~75° [21, 22]. (Figure 4A,B)

b. Oval Window: Along its course in the tympanic segment, the FN is normally located superior and lateral to the oval window (OW) and stapes. We evaluated the directional relationship between the FN and OW to detect any abnormalities in this segment according to previously published criteria [23]. (Figure 5A–C)

c. Round Window/Facial Recess: The FR is the traditional route used in posterior tympanotomy for CI surgery [24]. Since it may influence the choice of surgery for CI, the maximum width of the facial recess was measured by calculating the perpendicular distance between the FN and the chorda tympani nerve at the RW membrane level. FR was defined as "narrow" when the maximum width was <2.5 mm [25]. (Figure 6A,B).

We also inspected the FN for the presence of nerve hypoplasia, defined as a marked reduction in the number (atrophy) of fibers and/or an increased ratio of the nerve diameter in relation to the facial nerve canal (Figure 4A,B).

3 | Results

The results of measurements of various inner/middle ear structures by type of inner ear malformation were presented in Tables 1 and 2.

3.1 | Development of Mastoid

Thirteen out of 28 TBs (46.4%) revealed hypo-developed mastoid air cells. In the cases classified as CH-II, 5 out of 5 (100%) exhibited hypo-developed mastoid, whereas in CH-III cases, 8 out of 20 (40%) showed that pathology. On the other hand, 2 TBs from one IP-II case and 1 TB from an isolated vestibular malformation case showed normal mastoid development.

3.2 | Internal Auditory Canal

Of the 28 TBs, aside from the 2 that were excluded due to removal artifacts, only 2 of 26 TBs (7.7%) exhibited a narrow diameter. Of those, one was identified with cochlear hypoplasia-III, while the

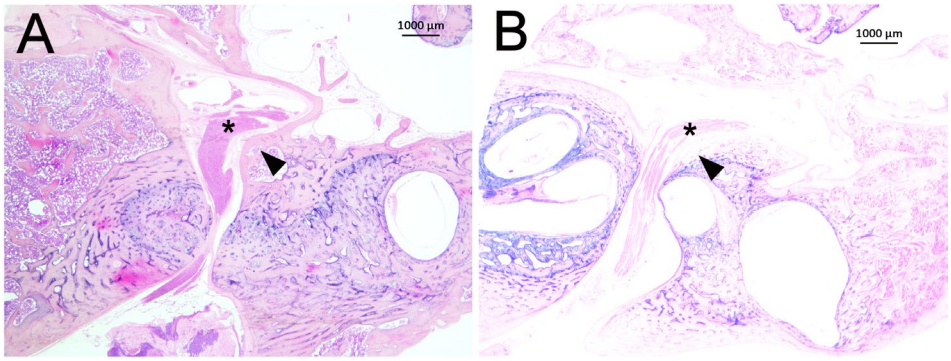

FIGURE 4 | Star: FN arrow head: First genu representative cases of the angle formed by the facial nerve at the first genu being normal (acute or $<75°$) and abnormal (obtuse). (A) This specimen is from one donor with cochlear hypoplasia type-III. In this case, the angle formed by the facial nerve at the first genu is normal (i.e., an acute angle or $<75°$). In this case, no apparent hypoplasia of the facial nerve is observed. (B) This specimen is from one donor with an isolated vestibular malformation. In this case, the angle formed by the facial nerve at the first genu is obtuse. In this case, the diameter of the facial nerve is significantly smaller as compared to that of a normal specimen, indicating facial nerve hypoplasia.

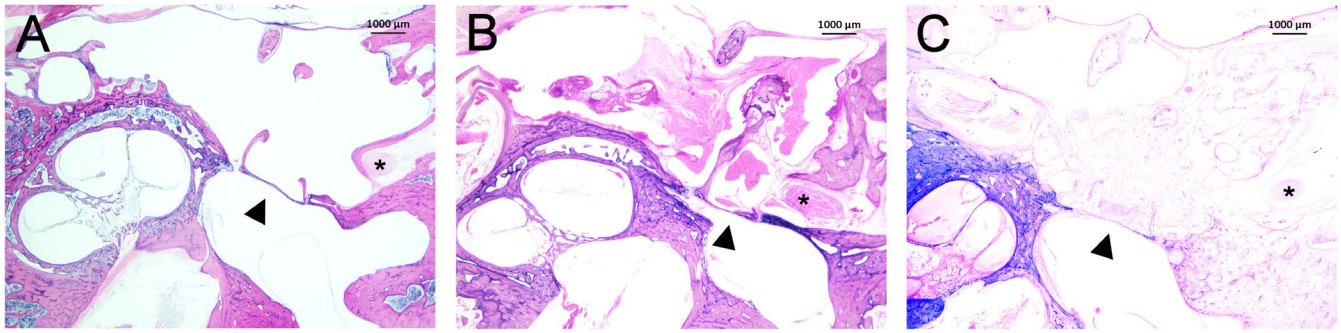

FIGURE 5 | Star: FN arrow head: OW. The positional relationship between the facial nerve and the oval window in the tympanic segment. (A) This specimen is from one donor with cochlear hypoplasia type III. In this case, the position of the facial nerve relative to the oval window is normal, but there is partial dehiscence of the facial nerve. (B) This specimen is from one donor with cochlear hypoplasia type III. In this case, the position of the facial nerve relative to the oval window is anterior and very close to normal. Furthermore, there is dysplasia of the oval window. Interestingly, this case exhibited cochlear hypoplasia type III on the right side and cochlear hypoplasia type II on the left side. (C) This specimen is from one donor with cochlear hypoplasia type III. In this case, the position of the facial nerve relative to the oval window is more posterior than normal.

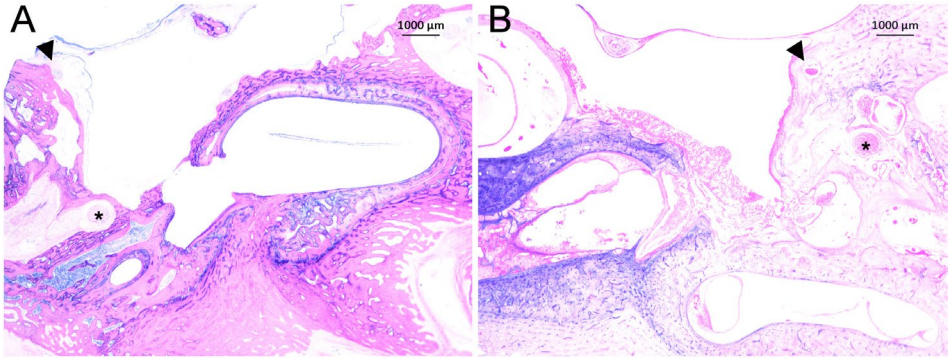

FIGURE 6 | Star: FN arrowhead: Chorda tympani representative cases of the facial recess (FR) width at the round window membrane level being normal (i.e., 2.5 mm or more) and narrowed (i.e., < 2.5 mm). (A) This specimen is from one donor with cochlear hypoplasia type III. In this case, the facial recess has a normal width (≥ 2.5 mm). (B) This specimen is from one donor with cochlear hypoplasia type III. In this case, the facial recess has a narrow width (1.5 mm).

TABLE 1 | Histopathological findings of the mastoid development and IAC in HTBs with IEMFs.

Type of IEMFs	Development of mastoid	Diameter of IAC	Length of IAC
Cochlear hypoplasia-type II ($N = 5$)	Hypo-developed; 100% (5)	Normal (≥ 2 mm); 80% (4) N/A(artifact); 20% (1)	Normal (≥ 5 mm); 40% (2) Short (< 5 mm); 40% (2) N/A(artifact); 20% (1)
Cochlear hypoplasia-type III ($N = 20$)	Normal; 60% (12) Hypo-developed; 40% (8)	Normal (≥ 2 mm); 90% (18) Narrow (< 2 mm); 5% (1) N/A(artifact); 5% (1)	Normal (≥ 5 mm); 50% (10) Short (< 5 mm); 45% (9) N/A(artifact); 5% (1)
Incomplete partition-type II (Mondini deformity) ($N = 2$)	Normal; 100% (2)	Normal (≥ 2 mm); 100% (2)	Normal (≥ 5 mm); 100% (2)
An isolated vestibular malformation ($N = 1$)	Normal; 100% (1)	Narrow (< 2 mm); 100% (1)	Short (< 5 mm); 100% (1)

Abbreviations: HTBs, Human Temporal Bones; IAC, Internal Auditory Canal; IEMFs, Inner Ear Malformations.

other had an isolated vestibular malformation, both presenting an IAC diameter of 1.9 mm. Overall, IAC diameter was measured as 3.48 ± 0.87 mm (mean ± SD), with a range of 1.9–5.4 mm.

In terms of IAC length, 12 of 26 TBs (46.2%) were detected with a short IAC. One temporal bone from the individual exhibiting isolated vestibular malformation also featured a narrow IAC. Two temporal bones were obtained from a patient classified as having CH-II, while the remaining samples exhibited CH-III. Overall, IAC length was 5.90 ± 2.27 mm (mean ± SD), with a range of 2.7–11.4 mm.

3.3 | The First Genu

It was not possible to assess the first genu in 8 TBs (7 TBs with artifacts, and 1 TB with aplastic FN) out of 28 TBs. The angle formed by the FN at the first genu was obtuse in 12 TBs and nearly perpendicular in 2 TBs. Among the TBs with an obtuse angle, one had an isolated vestibular malformation, the other 2 had CH-II, and the remaining 9 TBs had CH-III. The 2 perpendicular ones are also classified as CH-III. Overall, 14 out of 20 TBs (70%) showed abnormalities in the angle (aberrant course) of the first genu of the FN. In one of the TBs identified as CH-III, FN became aplastic before the first genu in the labyrinthine portion. (Figure 7).

3.4 | Facial Nerve and Oval Window

Of the 28 TBs, the positional relationship of the FN to the OW was not evaluated in 1 TB due to FN aplasia. In one of the TBs with CH-II, the FN was inferior to the OW as compared with normal. In cases of CH-III, the FN was positioned anterior to the OW in 6 TBs. Furthermore, in 4 out of these 6 TBs, the FN was present overlying the OW. In another one out of the CH-III cases, the FN was positioned superiorly and posteriorly to the OW. Out of 27 TBs, a total of 8 TBs (29.6%) exhibited abnormalities in the positional relationship of the FN to the OW.

Dysplasia or aplasia of the OW was observed in a total of 7 of 28 TBs (25.0%). In 11 out of 28 cases (40.7%) of TBs, the FN in the tympanic segment was completely or partially dehiscent (Figure 8A,B).

TABLE 2 | Characteristics of FN in HTBs with IEMFs.

Type of IEMFs	Angle of the first genu	Position of FN relative to the OW	Width of FR	Development of FN
Cochlear hypoplasia-type II	Normal (acute or < 75°); 40% (2)	Normal (superior & lateral); 80% (4)	Normal (≥ 2.5 mm); 40% (2)	Normal; 40% (2)
($N = 5$)	Obtuse; 40% (2)	Inferior; 20% (1)	Narrow (< 2.5 mm); 40% (2)	Hypoplastic; 60% (3)
	N/A(artifact); 20% (1)		N/A (Chorda tympani is missing); 20% (1)	
		FN Dehisence; 20% (1)		
		Aplastic OW; 20% (1)	Overhanging bone at RW niche; 20% (1)	
Cochlear hypoplasia-type III	Normal (acute or < 75°); 15% (3)	Normal (superior and lateral); 60% (12)	Normal (≥ 2.5 mm); 30% (6)	Normal; 30% (6)
($N = 20$)	Obtuse; 45% (9)	Anterior; 30% (6)	Narrow (< 2.5 mm); 65% (13)	Hypoplastic; 65% (13)
	Perpendicular; 10% (2)	Superior & posterior; 5% (1)	N/A (Aplastic FN); 20% (1)	Aplastic; 5% (1)
	N/A (aplastic FN); 5% (1)	N/A(aplastic FN); 5% (1)		
	N/A (artifact); 25% (5)		Overhanging bone at RW niche; 25% (5)	
		FN Dehisence; 45% (9)	Bony obliteration with aplastic/ hypoplastic RW; 15% (3)	
		Aplastic OW; 25% (5)		
		Dysplastic OW; 5% (1)		
Incomplete partition-type II (Mondini deformity)	Normal (acute or < 75°); 50% (1)	Normal (superior & lateral); 100% (2)	Normal (≥ 2.5 mm); 100% (2)	Normal; 100% (2)
($N = 2$)	N/A (artifact); 50% (1)			
			Overhanging bone at RW niche; 50% (1)	
An isolated vestibular malformation	Obtuse; 100% (1)	Normal (superior & lateral); 100% (1)	Narrow (< 2.5 mm); 100% (1)	Hypoplastic; 100% (1)
($N = 1$)				
		FN Dehisence; 100% (1)		

Abbreviations: FN, Facial Nerve; FR, Facial Recess; HTBs, Human Temporal Bones; IEMFs, Inner Ear Malformations; OW, Oval Window; RW, Round Window.

3.5 | Facial Recess and Round Window

Of the 28 TBs, FR width could not be measured in 1 TB due to FN aplasia. Sixteen (59.3%) of the remaining TBs were determined to have narrow FR. Two of the TBs had CH-II, and the remaining had CH-III. The mean ± SD of the FR diameter was 2.36 ± 0.56 mm, with a range of 1.2–3.4 mm. In addition, the RW niche showed stenosis or obliteration by bony tissues in 9 of 28 TBs (32.1%). In these 9 TBs, 6 TBs showed partial RW stenosis by overhanging bone, and the remaining 3 TBs showed the aplastic or hypoplastic RW with obliteration by bone tissue.

3.6 | Development of Facial Nerve

In the 18 of 28 TBs (64.3%), the FN fiber was hypoplastic or aplastic. One of them had an isolated vestibular malformation, 3 of them had CH-II, and all the remaining 14 TBs had CH-III. In one of the TBs identified as CH-III, the FN became aplastic in the labyrinthine portion. Clinical information related to the FN function was only available for one donor with CH-III, who presented with complete absence of FN function.

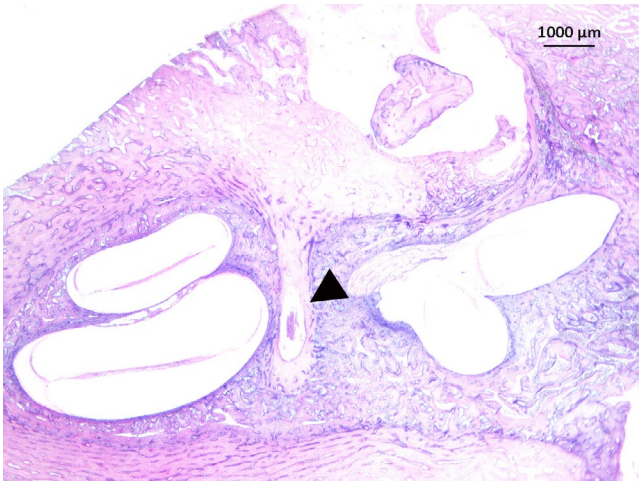

FIGURE 7 | Presentation of a case with prominent facial nerve hypoplasia and partial aplasia. This is a specimen from one donor with cochlear hypoplasia III and an example of a case that particularly demonstrates prominent facial nerve hypoplasia and partial aplasia in our current study. Interestingly, the facial nerve became aplastic before the first genu in the labyrinthine portion (arrow head).

4 | Discussion

Our findings demonstrate a constellation of abnormalities impacting the FN development, course, and position in bones with IEMF that can present significant challenges for proper CI placement and increase the risks of FN injuries. This detailed study of structural aspects is particularly valuable since current imaging techniques do not provide the same level of detail.

We found a high prevalence of FN hypoplasia in our sample (64.3%), being more commonly observed in cases of CH-II (60%) and CH-III (65%). Our findings may correlate with previous observations showing that patients with IEMFs can present with an associated facial weakness or palsy [26].

Our results revealed a high prevalence of aberrant FN course, particularly in CH cases. Although we faced challenges in studying the labyrinthine FN segment using horizontal sections, we used the first genu as an indicator of course abnormalities. Our TBs, particularly CH cases, presented with an anterior and inferior displacement of the first genu. Previous literature dedicated to evaluating the labyrinthine segment of the FN in IEMFs presents conflicting results: while some histopathological and imaging investigations showed similar findings [11, 21, 22, 27], Sennaroğlu et al. [23] reported that no major abnormalities were seen in high-resolution CT scans. Although our histopathological study only allows us to speculate on the causes for both labyrinthine and first genu malposition in CH cases, previous studies reported that the abnormal FN course can be the result of the stunted development of the otic capsule and cochlea as a whole [11].

We found a high prevalence of anatomic abnormality of the FN in the first genu region (70%). In this study, we found that the first

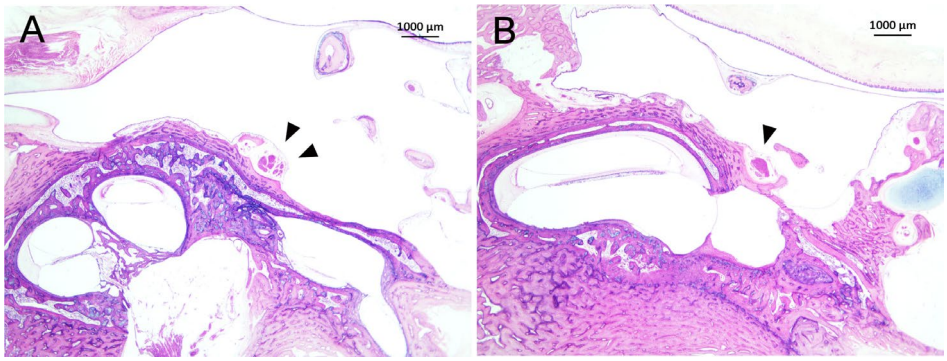

FIGURE 8 | Facial nerve which is significantly anteriorly displaced and dehiscent presentation of a case with prominent facial nerve dehiscence in the tympanic segment. (A) This is a specimen from one donor with cochlear hypoplasia type III and an example from our current study that particularly highlights dehiscence of the facial nerve in the tympanic segment. The course of the facial nerve is also significantly displaced anteriorly. (B) This is a section at the level of the round window in the same case of Figure 8A. In such cases, even if the surgeon chooses a trans-canal approach, special attention should be paid to the fenestration of the cochlear window.

genu was anteriorly and inferiorly displaced and formed obtuse or perpendicular angles. These abnormalities can impact CI surgeries via the middle cranial fossa (MCF) approach. The MCF approach, commonly used as an alternative or optional CI route when the posterior tympanotomy is not feasible [28–30], is extremely challenging in IEMF cases as the surgical landmarks can be blunted [28, 31, 32]. These anatomical issues, in association with an abnormal first genu, can yield additional risks of FN injury [29, 30].

In the tympanic segment, abnormal positioning of the FN was observed in 30% of our sample, most of which were CH-III cases. The most frequent and representative finding was displacement towards the OW. Additionally, OW dysplasia and/or aplasia was seen in 25% of the TBs. While the abnormal FN positioning confined to the tympanic segment alone is unlikely to impact CI (except in cases where an extended cochleostomy is required) [23], it could significantly impact other middle ear procedures such as chronic otitis media surgery or stapes procedures [33, 34].

We also found a high prevalence of FN dehiscence in the tympanic segment (40.7%). The location and frequency of these dehiscences were consistent with previous reports for non-IEMF cases, which ranged from 25% to 57% [35, 36]. As the course of the FN is frequently abnormal in cases of IEMF, the presence of dehiscences can yield additional risks for FN injury. Our findings corroborate the need for comprehensive preoperative high-resolution imaging study to assess the presence of these dehiscences, as these may be difficult to identify intra-operatively [37–42]. Additionally, the high rate of FN dehiscence as well as the abnormal FN course can result in higher risks of post-operative FN stimulation symptoms. Previous literature [17] showed that the patients with IEMFs have a higher incidence of post-operative FN stimulation symptoms following CI.

We identified significant abnormalities in the mastoid portion of the FN. The prevalence of a narrow FR was 59.3% in our cases, markedly higher than in previous studies using high-resolution CT of patients with IEMFs (estimated at 20%) [23]. Furthermore, the average width (2.36 ± 0.56 mm) was shorter than the typical range reported in normal cases (3–4 mm both for children and adults) [43]. A narrowed FR can impede access to the RW niche using the standard transmastoid/FR approach. In addition to FR narrowing, we frequently observed hypo-developed (46.4% of cases) and stenotic or obliterated RW niches (32.1% of cases), particularly in CH cases. These findings parallel those reported for IEMFs [11, 23]. Therefore, alongside the challenges posed by the narrow FR, other findings such as hypo-developed mastoids, RW niche abnormalities, abnormal promontory anatomy, and FN displacement within the tympanic segment significantly increase the risk of FN injury or entirely obstruct the transmastoid approach. Moreover, CI via the MCF approach, which demands high technical proficiency even in normal cases, becomes even more daunting in IEMFs due to abnormal cranial base anatomy, heightening the risk of severe complications [28, 31, 32]. Hence, thorough preoperative imaging assessments are crucial to carefully select the most appropriate surgical approach.

Our study has limitations. The small sample size for each type of IEMF limited our ability to conduct statistical analyses. Unfortunately, our collection does not have cases of IEMFs that had undergone CI surgery, and the available clinical records were limited. As a result, it was not possible to directly correlate the histopathological findings with clinical and surgical aspects (e.g., preoperative hearing including ABR, postoperative outcomes, subsequent language development, and clinical findings at the time of surgery, or the presence of facial nerve symptoms during the patient's lifetime).

5 | Conclusion

Our study shows a variety of abnormalities affecting the FN in IEMF cases, some of which can significantly impact CI surgery. Further research with larger sample sizes and multi-center collaborative studies that integrate clinical aspects, such as intraoperative findings and radiological diagnoses, is expected to provide deeper insights, ultimately improving patient outcomes and reducing surgical risks in IEMF cases.

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Conflicts of Interest

The authors declare no conflicts of interest.

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