ORIGINAL RESEARCH

Laryngoscope
Investigative Otolaryngology

Effect of inner ear malformations on intraoperative ECAP thresholds and postoperative auditory performance

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Abstract

Objectives: This study sought to characterize the influence of inner ear malformations (IEMs) on intraoperative electrically evoked compound action potential (ECAP) and auditory performance to better understand the underlying pathophysiology related to variabilities in cochlear implant (CI) outcomes that individuals with malformed cochlea may present.

Methods: The medical records of 222 ears implanted with Cochlear Nucleus CI were reviewed. Of the total, 64 ears had radiologic evidence of IEMs, and 158 ears were normal. Individuals with IEMs were grouped based on the severity of anomalies; 38 had mild IEMs (e.g., enlarged vestibular aqueduct, incomplete partition type II, etc.) and 26 had severe IEMs (e.g., cochlear nerve hypoplasia, common cavity, etc.). Intraoperative ECAP thresholds obtained via neural response telemetry (NRT) and the categories of auditory performance (CAP) scores measured at 12 months postoperative were compared and correlated.

Results: Absent ECAP responses were more apparent in the IEM group. ECAP thresholds were significantly elevated in the severe IEM group, while the mild IEM group had ECAP thresholds comparable to the normal group. The mild IEM group achieved CAP scores similar to the normal control. Patients in the severe IEM group showed significantly lower CAP scores at 12 months postoperative. Significant negative relationships existed between ECAP thresholds and CAP scores obtained from all subjects.

Conclusion: Measurable ECAP responses and NRT thresholds varied across groups. The inverse relationship between NRT thresholds and CAP scores may suggest that electrophysiological responses measured during surgery may potentially be indicative of postoperative performance in our CI population.

Level of Evidence: 2b.

KEYWORDS
categories of auditory performance, cochlear implant, electrically evoked compound action potential, inner ear malformation, neural response telemetry

1 | INTRODUCTION

Candidacy criteria for cochlear implantation (CI) have gradually expanded over the years to include those with inner ear malformations (IEMs). Individuals with IEMs were considered less desirable candidates in earlier phases of CI due to concerns surrounding increased surgical risk and poor prognosis, presumably associated with the lack of neural populations and abnormal cochleovestibular structures.[1] However, advances in surgical techniques and diagnostic radiology and accumulation of IEM cases have led to reported benefits after CI facilitated CI in individuals with the malformed cochlea.[1–6] While a good prognosis is generally anticipated for those with mild IEMs, individuals who have severe IEMs have achieved some functional benefits, including better detection of environmental sounds, increased ability to communicate with or without lip-reading over the years of implant use, and intensive aural rehabilitation. Currently, severe IEM is no longer considered a contraindication for CI.

Individuals with IEMs are likely to present with the aberrant distribution of the auditory nerve and a low survival rate of the spiral ganglion neurons along the modiolus.[4,5,7] The health of the auditory nerve may play an essential role in determining CI outcomes in individuals with IEMs compared to those with the normal cochlea. Thus, assessing the functional status of the auditory nerve may help us better understand the association between neural health and considerable variance in auditory performance in the CI population.

The status of the auditory nerve can be evaluated using electrically evoked compound action potential (ECAP). ECAP is a summed response to electrical stimulation across a large number of individual auditory nerve fibers. ECAP offers many clinical benefits. Most importantly, ECAP has been used as an objective tool to determine programming levels for individual CI electrodes, which can assist device programming for hard-to-test populations who cannot present consistent behavioral responses. ECAP has also been used as a routine procedure to verify implant function and ensure responses from the auditory nerve to electrical stimulation during surgery.[8] Accumulating evidence suggests that ECAP responses are sensitive to reflect the health status of auditory nerve fibers near recording electrodes that may play an important role in determining CI outcomes.[8–11] Based on these findings that ECAP reflects neural survival of auditory nerve fibers, there has been increased interest in exploring cochlear nerve responsiveness using different ECAP parameters in CI children with cochlear nerve deficiency or genetic mutations that may affect auditory nerve function.[12–15]

It is clinically challenging to manage individuals with IEMs due to the lack of understanding of their underlying neurophysiological mechanisms. Developing a clinical tool for these patients requires a better understanding of the functional status of the peripheral auditory system. ECAP may potentially be used as a guide to estimate CI progress based on neural status and customize individualized counseling that directs an optimal rehabilitation. This study evaluated the use of intraoperative ECAP measurements to characterize the influence of IEMs on the status of the auditory nerve. Understanding the association with postoperative auditory performance may also help us understand the underlying pathophysiology related to the large variability in outcomes that CI users with malformed cochlea may present.

2 | MATERIALS AND METHODS

2.1 | Study participants

The medical records of 222 ears from 155 subjects who underwent CI with Cochlear Nucleus CI (Cochlear Ltd.) at Samsung Medical Center between 2016 and 2021 were reviewed in this retrospective study. All subjects had no additional disabilities. Eighty-eight subjects were unilaterally implanted, while the other 67 subjects were bilaterally implanted. All participants had severe to profound hearing loss in the implanted ear and underwent CI after having minimal benefit from at least 3 months of hearing aid use and aural rehabilitation. Seventy-one ears were from adults (mean 54.54 years of age), and 151 ears were from children (mean 5.32 years of age).

All subjects were implanted with either Cochlear Nucleus lateral wall slim electrodes (e.g., CI422, CI522, CI622) or perimodiolar electrodes (e.g., CI512, CI532, CI632). The type of electrode was determined before surgery based on the clinical decision regarding the preoperative residual hearing in the implanted ear and anatomical structures. All participants underwent temporal bone computed tomography (TBCT) and magnetic resonance imaging (MRI) of the internal auditory canal (IAC) before surgery. Imaging study results were reviewed and the type of IEMs were classified by one neurotologist (author I.J.M.). The widely accepted categorizations based on embryogenesis of the inner ear by Jackler et al.[16] and Sennaroğlu et al.[17] were adopted for IEM classification. Based on these criteria, subjects in the IEM group were split into two groups according to the severity of malformations. Enlarged vestibular aqueduct (EVA), incomplete partition (IP) type II, and semicircular canal hypoplasia or aplasia were classified as the "mild" IEM group. Other anomalies such as cochlear hypoplasia or aplasia, IP type I and III, common cavity, cochlear nerve hypoplasia or aplasia, narrow IAC, or bony cochlear nerve canal stenosis were classified as the "severe" IEM group. When study participants showed more than one type of inner ear anomaly, they were assigned to the group based on the most severe malformation. For example, if a subject had IP-II, narrow IAC, and cochlear aplasia, this individual was categorized as having severe IEMs.

A total of 64 ears had radiologic evidence of malformations and were referred to as the "IEM" group; 38 ears had mild IEMs while the other 26 ears had severe IEMs. A total of 158 ears had normal cochlear structures and intact auditory nerves and were referred to as the "normal" group. Table 1 describes subject demographics and the types of intracochlear electrode arrays per group in detail.

This retrospective study was approved by the Institutional Review Board (IRB) at Samsung Medical Center (SMC 2021-05-011-001). All subjects signed an informed consent document before participating in this study.

TABLE 1 Demographics of study participants

| Groups | N (ears) | | Age (years) | | Intracochlear electrode array | | | | | | | Age at CI (years) | Postop CI use (months) |
	Adult	Children	Adult	Children	CI 512	CI 422	CI 522	CI 622	CI 532	CI 632		
Normal	66	92	54.53 (SD = 18.18)	5.45 (SD = 3.68)	2	38	34	49	22	13	24.55 (SD = 27.55)	26.75 (SD = 27.30)
Mild IEM	2	36	35.5 (SD = 2.12)	5.5 (SD = 3.76)	1	8	13	12	3	1	4.71 (SD = 7.46)	10.45 (SD = 3.05)
Severe IEM	0	26	N/A	4.65 (SD = 3.23)	0	7	10	5	4	0	1.92 (SD = 2.57)	10.61 (SD = 3.72)
Total	68	154	54.54 (SD = 18.20)	5.32 (SD = 3.61)	3	53	57	66	29	14	18.53 (SD = 25.32)	28.66 (SD = 17.48)

Abbreviation: IEM, inner ear malformation.

2.2 | Intraoperative recordings

CI was conducted using the standard facial recess approach. An intracochlear electrode was introduced via a round window membrane in an attempt to preserve residual hearing and cochlear structures. When the round window did not allow for full visualization or it was too anatomically challenging to advance an electrode through the round window, cochleostomy or the extended round window approach was implemented. In all cases, an electrode array was completely inserted.

Responses from the auditory nerve were recorded immediately after electrode insertion. Neural response telemetry (NRT) featured in the Custom Sound EP software was used to record ECAP responses. The following default NRT recording parameters were used; (1) a stimulation rate of 250 Hz, (2) 25-μs pulse widths, (3) 35 averages per measurement, (4) amplifier gain of 50 dB, and (5) a measurement delay of 125 μs. NRT thresholds were detected automatically by tracing measurable ECAP waveforms to given electrical stimulation across all 22 electrodes.

2.3 | Postoperative outcome measures

Categories of auditory performance (CAP) score obtained at 12 months postoperative were used as CI outcome measures for this study. CAP is an index consisting of eight performance categories designed to assess functional performance in everyday situations.[18] These categories cover a range of auditory performances arranged in order of increased difficulty from 0 (no awareness of environmental sounds) to 7 (use of a telephone with a familiar talker). CAP is a useful tool to provide clinicians the overall picture of auditory performance in CI users, including the "difficult-to-test" population who may not be able to develop open-set speech perception abilities.

2.4 | Data analysis

Intraoperative NRT thresholds measured from the 22 electrodes were analyzed using the linear mixed-effects (LME) model to compare differences between the normal and two IEM groups. The model included fixed effects described in the results and a random intercept for subjects. All reported *p* values for pairwise comparisons were adjusted using a Tukey adjustment for multiple comparisons. CAP scores measured at preimplant and 12 months postoperative were compared across groups using one-way ANOVA with post hoc Tukey's HSD test. The relationship between intraoperative NRT thresholds and postoperative CAP scores was analyzed using Pearson's correlation and univariate linear regression. Statistical analyses were performed using SPSS version 22.0 (IBM) and SAS version 9.4 (SAS Institute).

TABLE 2 Types of inner ear malformations (IEM) of study participants

IEM types by severity	Number of cases (ears)	Number of cerebrospinal fluid (CSF) gusher
Mild IEM		
Incomplete partition (IP) type II (IP-II)	1	0
Enlarged vestibular aqueduct (EVA)	7	4
Semicircular canal (SCC) hypoplasia	3	0
IP-II, EVA	26	16
SCC aplasia	0	0
EVA, SCC hypoplasia	1	0
Subtotal	38	20
Severe IEM		
Incomplete partition (IP) type I (IP-I)	0	0
Incomplete partition (IP) type III (IP-III)	0	0
Common cavity	0	0
Cochlear hypoplasia	0	0
Cochlear aplasia	0	0
Cochlear nerve hypoplasia	5	0
Cochlear nerve aplasia	0	0
Narrow internal auditory canal (IAC)	0	0
Bony cochlear nerve canal (BCNC) stenosis	1	0
BCNC stenosis, narrow IAC, cochlear nerve hypoplasia	2	0
Cochlear hypoplasia, narrow IAC	1	0
Cochlear hypoplasia, SCC hypoplasia, cochlear nerve hypoplasia	2	0
Cochlear nerve aplasia, SCC hypoplasia	1	0
Cochlear nerve hypoplasia, narrow IAC	3	0
Cochlear nerve hypoplasia, narrow IAC, BCNC stenosis	4	0
IP-II, narrow IAC, cochlear aplasia	2	0
Narrow IAC, cochlear nerve hypoplasia	2	0
Narrow IAC, cochlear nerve hypoplasia, SCC hypoplasia	1	0
SCC hypoplasia, cochlear nerve hypoplasia	1	0
SCC hypoplasia, narrow IAC, cochlear nerve hypoplasia	1	0
Subtotal	26	0
Total	64	20

3 | RESULTS

3.1 | Preoperative imaging findings

All subjects underwent high-resolution TBCT scanning and IAC MRI before CI. Imaging results were reviewed and subjects who had evidence of IEMs were placed into two groups according to the severity of malformations.[16,17] Table 2 shows the type of IEMs in our CI subjects. A total of 64 ears had radiologic evidence of IEMs (64/222 = 28%); 38 ears had mild form IEMs (38/64 = 59%) while the other 26 ears had severe form IEMs (26/64 = 40%). The most common malformation type was IP-II

concomitant with EVA in 26 ears. It was followed by EVA only in seven ears and cochlear nerve hypoplasia occurred in five ears. A cerebrospinal fluid (CSF) gusher was encountered in 20 ears having either IP-II with EVA (16/20) or EVA only (4/20). A CSF leak was not observed in any cases with severe IEMs in our CI population.

3.2 | Intraoperative NRT recordings

In general, intraoperative NRT thresholds were measurable from all 22 electrodes in more than 70% of ears (160/222 = 72.07%), while these

were completely absent in 12 ears (12/222 = 5.4%). Other subjects showed intermittent responses. NRT thresholds were all present except for one or two basal electrodes in 10% of ears (23/222 = 10.36%). The other 12% of ears (27/222 = 12.16%) had measurable NRT thresholds between 3 and 21 electrodes with varying locations.

The prevalence of measurable NRT thresholds varied between the normal and IEM groups. Individuals with normal cochlea tended to have a higher number of measurable ECAP thresholds (156/222 = 70.27%). The IEM group had 24% measurable NRT responses from at least one of the tested electrodes (54/222 = 24.32%); the mild IEM group had 16% measurable NRT responses (37/222 = 16.66%) and the severe IEM group had 7% measurable NRT responses (17/222 = 7.65%). Normal and IEM groups were also different in the number of electrodes with measurable NRT responses (see Figure 1). All present ECAP responses from 22 electrodes were more apparent in the normal group (126/222 = 56.75%) followed by the mild IEM group (30/222 = 13.51%). Subjects in the severe IEM group rarely had "all present" NRT responses from 22 electrodes (4/222 = 1.8%). Absent NRT responses were more noticeable in the IEM groups. Completely absent ECAP responses from all tested electrodes were frequently seen in subjects in the severe IEM group (9/222 = 4.05%), compared to the mild IEM group (1/222 = 0.45%) and normal group (2/222 = 0.9%).

Intraoperative NRT thresholds obtained from 22 electrodes were compared across groups using the LME analysis. The LME analysis included a fixed effect for the group varied by the severity of malformations and a random intercept for subjects. We focused on testing the main effect of the different groups on ECAP thresholds. Results showed a significant main effect of groups on NRT thresholds ($F_{(2,408)} = 34.287$, $p < .001$). Post hoc analysis revealed that average NRT thresholds of the severe IEM group were significantly elevated compared to those with the normal group ($p < .001$) and the mild IEM group ($p < .001$), and this trend was consistent across all electrodes. Interestingly, the mild IEM group showed average NRT thresholds comparable to those of the normal group ($p = .874$). This trend was consistent across all tested electrodes (see Figure 2).

3.3 | Postoperative auditory performance

The CAP measurements at 12 months postoperative were compared between groups using one-way ANOVA (see Figure 3). CAP scores measured from the normal and two IEM groups were significantly different ($F_{(2,157)} = 41.199$, $p < .001$). Post hoc analysis revealed that the severe IEM group remained at a considerably lower level of CAP scores ($p < .001$) compared to the normal and mild IEM groups. The mild IEM group achieved CAP scores similar to those of the normal group ($p = .475$).

CAP scores measured at 12 months postoperative (postop) were also compared with those measured before implantation (preop) to investigate the degree of improvement in auditory performance between groups (see Figure 4). The mild IEM group showed considerable improvement in CAP scores at postop 12 months and the level of improvement was comparable to the normal group. The severe IEM

group also showed improved CAP scores; however, they did not reach the level of the other groups at 12 months after CI. Improvement on CAP scores preop and postop was significantly different across groups ($F_{(2,157)} = 4.813$, $p = .009$). Post hoc analysis showed that the mild IEM group had similar improvement in CAP scores compared to the normal group ($p = .841$). Two IEM groups were significantly different in CAP score changes between preop and postop ($p = .006$).

3.4 | Relationship between intraoperative NRT threshold and auditory performance

Intraoperative NRT thresholds obtained from all 22 electrodes correlated with postoperative CAP scores. Significant inverse correlations
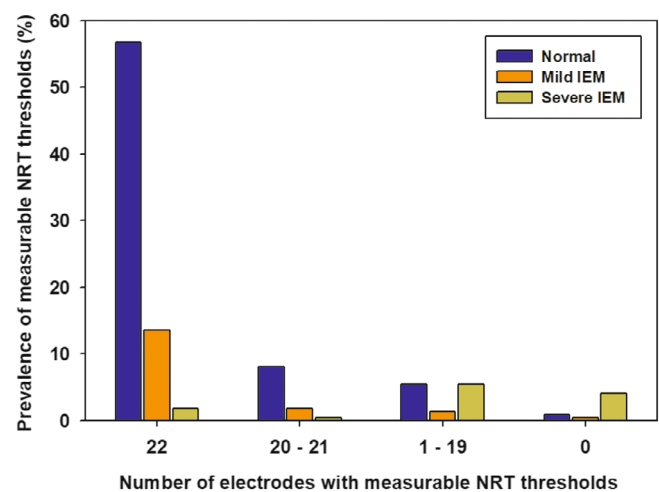

FIGURE 1 The number of electrodes with measurable NRT thresholds between normal and IEM groups. IEM, inner ear malformation; NRT, neural response telemetry
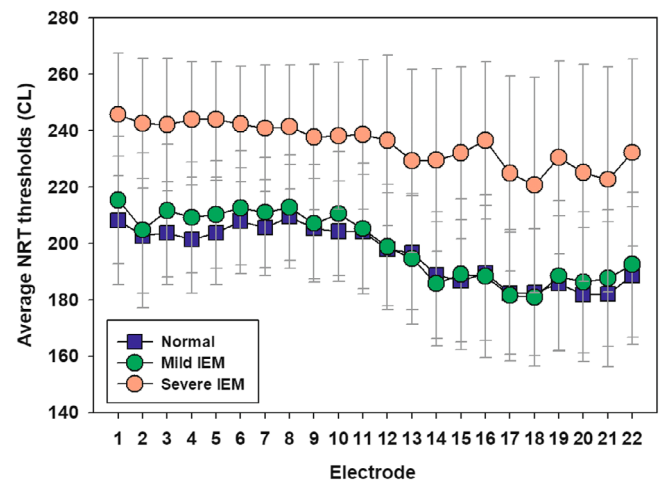
FIGURE 2 Average NRT thresholds measured across 22 electrodes between normal and two IEM groups. Gray error bars indicate +1 standard deviation (SD). IEM, inner ear malformation; NRT, neural response telemetry
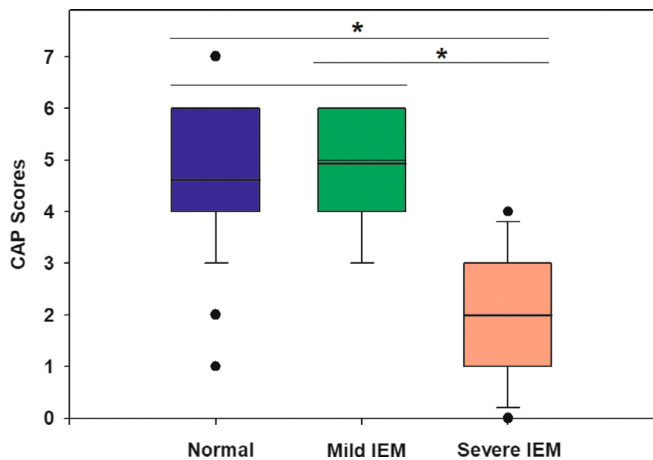
FIGURE 3 Box and whisker plots of CAP scores measured at postoperative 12 months across normal and two IEM groups. Asterisks (*) indicate statistical significance at $\alpha = .05$. CAP, categories of auditory performance; IEM, inner ear malformation
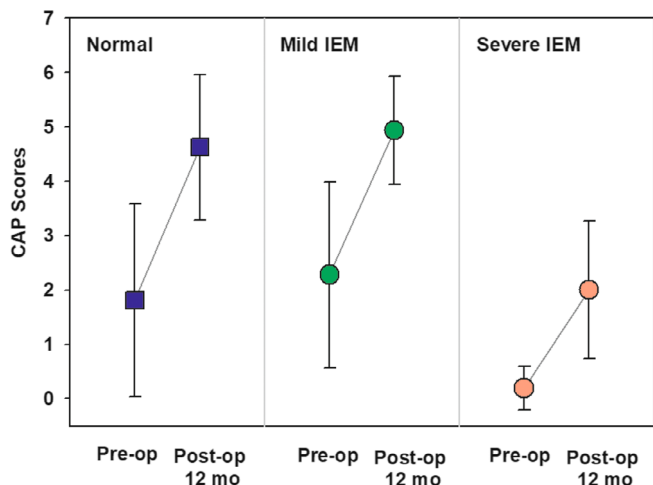
FIGURE 4 Comparison of CAP scores before cochlear implantation (preop) and 12 months postoperative (postop) across normal and IEM groups. Error bars indicate +1SD. CAP, categories of auditory performance; IEM, inner ear malformation

TABLE 3 Correlation statistics between intraoperative NRT thresholds and postoperative CAP scores combined across groups

| | Correlation coefficient | | |
Electrode number	r	r²	p Value
E1	−.383	.147	<.001***
E2	−.486	.236	<.001***
E3	−.467	.218	<.001***
E4	−.482	.232	<.001***
E5	−.478	.228	<.001***
E6	−.438	.192	<.001***
E7	−.452	.204	<.001***
E8	−.491	.241	<.001***
E9	−.471	.222	<.001***
E10	−.454	.206	<.001***
E11	−.400	.160	<.001***
E12	−.388	.150	<.001***
E13	−.423	.179	<.001***
E14	−.419	.176	<.001***
E15	−.417	.174	<.001***
E16	−.399	.159	<.001***
E17	−.470	.221	<.001***
E18	−.449	.202	<.001***
E19	−.448	.201	<.001***
E20	−.410	.168	<.001***
E21	−.359	.129	<.001***
E22	−.396	.157	<.001***

Note: *p < .05, **p < .01, ***p < .001.
Abbreviations: CAP, categories of auditory performance; NRT, neural response telemetry.

were found between NRT thresholds and CAP scores when all participants were combined (Table 3). These negative linear relationships indicate that subjects who have lower NRT thresholds tended to achieve higher CAP scores. Univariate linear regression was also conducted to reveal an association between ECAP thresholds and auditory performance. Overall, regression coefficients (B) obtained from all 22 electrodes showed significant negative relationships between intraoperative NRT thresholds and CAP scores (Table 4, see overall). These results suggest that one current level (CL) increase in the NRT threshold is associated with an approximately 0.02 or 0.03 decrease in CAP score at 12 months postoperative. CAP scores obtained from the normal group were associated significantly with the NRT thresholds at more basally located electrodes (e.g., E2–E10) and more

apically located electrodes (e.g., E17–E20). This negative-direction relationship between intraoperative NRT thresholds and postoperative CAP scores was not significant in both IEM groups.

4 | DISCUSSION

The current study explored the influence of ear anomalies on electrophysiological information that may reflect the health of the cochlea and the status of auditory nerve integrity. We hypothesized that the behavior of ECAP response and its association with postoperative auditory performance might be different between normal and IEM groups. The percentage of measurable ECAP thresholds obtained during surgery was far lower in individuals with IEMs compared to those with normal cochlear structures. The lower occurrence rate of measurable ECAPs in the IEM group was consistent with previous reports comparing intraoperative ECAPs between CI users with the malformed and normal cochlea. ECAP thresholds were present in 25% of the malformed cochlea group, compared to 74% in the normal control.[19] The percentage of measurable ECAPs in the group with normal

TABLE 4 Regression statistics between intraoperative NRT thresholds and postoperative CAP scores obtained from all subjects (overall) and by groups (normal, mild IEM, and severe IEM groups)

Electrode number	Overall		Normal		Mild IEM		Severe IEM	
	Regression coefficient B (S.E.)	p Value	Regression coefficient B (S.E.)	p Value	Regression coefficient B (S.E.)	p Value	Regression coefficient B (S.E.)	p Value
E1	−0.02 (0.005)	**<.001**	−0.005 (0.005)	.405	−0.01 (0.008)	.109	−0.02 (0.014)	.269
E2	−0.03 (0.004)	**<.001**	−0.01 (0.006)	**.044**	−0.02 (0.006)	**.014**	−0.02 (0.012)	.219
E3	−0.03 (0.005)	**<.001**	−0.02 (0.007)	**.008**	−0.01 (0.008)	.292	−0.01 (0.012)	.292
E4	−0.03 (0.005)	**<.001**	−0.02 (0.007)	**.033**	−0.01 (0.010)	.306	−0.02 (0.012)	.181
E5	−0.03 (0.005)	**<.001**	−0.02 (0.007)	**.015**	−0.01 (0.011)	.578	−0.02 (0.012)	.190
E6	−0.03 (0.005)	**<.001**	−0.02 (0.007)	**.021**	−0.003 (0.009)	.784	−0.02 (0.013)	.191
E7	−0.03 (0.005)	**<.001**	−0.02 (0.008)	**.010**	−0.004 (0.009)	.694	−0.01 (0.012)	.246
E8	−0.04 (0.005)	**<.001**	−0.02 (0.007)	**.001**	−0.01 (0.010)	.419	−0.01 (0.014)	.357
E9	−0.03 (0.005)	**<.001**	−0.02 (0.008)	**.015**	−0.02 (0.008)	.058	−0.02 (0.010)	.055
E10	−0.03 (0.005)	**<.001**	−0.02 (0.008)	**.002**	−0.01 (0.008)	.371	−0.01 (0.011)	.363
E11	−0.03 (0.005)	**<.001**	−0.01 (0.007)	.150	−0.01 (0.008)	.097	−0.01 (0.010)	.318
E12	−0.03 (0.005)	**<.001**	−0.01 (0.007)	.202	−0.01 (0.008)	.168	−0.01 (0.009)	.381
E13	−0.03 (0.005)	**<.001**	−0.01 (0.007)	.077	−0.01 (0.008)	.203	−0.01 (0.009)	.253
E14	−0.02 (0.004)	**<.001**	−0.01 (0.006)	.376	−0.03 (0.007)	**<.001**	−0.01 (0.009)	.406
E15	−0.02 (0.004)	**<.001**	−0.01 (0.007)	.151	−0.01 (0.007)	.374	−0.01 (0.009)	.450
E16	−0.02 (0.004)	**<.001**	−0.01 (0.006)	.148	−0.01 (0.006)	.415	−0.004 (0.010)	.690
E17	−0.03 (0.004)	**<.001**	−0.02 (0.006)	**.011**	−0.01 (0.008)	.144	−0.01 (0.008)	.397
E18	−0.03 (0.004)	**<.001**	−0.01 (0.006)	**.011**	−0.01 (0.008)	.113	−0.01 (0.007)	.194
E19	−0.02 (0.004)	**<.001**	−0.01 (0.006)	**.015**	−0.004 (0.007)	.579	−0.01 (0.008)	.437
E20	−0.02 (0.004)	**<.001**	−0.01 (0.006)	**.021**	−0.005 (0.007)	.475	−0.005 (0.007)	.529
E21	−0.02 (0.004)	**<.001**	−0.01 (0.005)	.126	−0.005 (0.007)	.486	−0.01 (0.007)	.348
E22	−0.02 (0.004)	**<.001**	−0.01 (0.005)	.072	−0.01 (0.007)	.341	−0.01 (0.008)	.418

Abbreviations: CAP, categories of auditory performance; IEM, inner ear malformation; NRT, neural response telemetry; S.E., standard error.
Note: Bold p values indicate statistical significance ($\alpha = .05$).

imaging findings was approximately 90% compared to 58%–81% of measurability across electrodes in the pathological group.[20] In addition, patients with IEMs tended to have a higher presence of "variable" ECAP responses referred to as reproducible responses only in a limited number of electrodes, and "no" response referred to no identifiable response waveforms in any of the electrodes.[21] Reduced incidence of measurable ECAP responses in individuals with ear anomalies is presumably due to pathological changes in the peripheral auditory system that may affect ECAP responses representing a synchronized response generated by a group of electrically activated auditory nerve fibers. The presence of IEMs is known to be associated with decreased neural survival, various degrees of modiolar defects, the likelihood of fewer spiral ganglion cells, and hypoplastic or aplastic vestibulocochlear nerve.[3,7,22,23]

There has been increased interest in using ECAP metrics to estimate neural survival of auditory nerve fibers to identify this association in human and animal models. ECAP thresholds and maximum ECAP amplitudes are believed to reflect the size of the electrically activated neural population and the density of the surviving neural population.[9,11,12,24] The presence of identifiable ECAP responses could also be dependent upon structural changes of the cochlea.

Electrical stimulation could cause atypical waveform morphology and prevent the appearance of measurable ECAP waveforms in the presence of cochlear malformations, otosclerotic bone growth, and ossification of the cochlea.[25,26] There were more missing cases of identifiable ECAP responses in individuals with inner ear anomalies, presumably related to the irregular formation and changed the anatomy of the cochlea.[19,27]

In the current study, a significant difference in intraoperative ECAP thresholds was identified between IEM groups and the normal control. Significantly higher CLs were required to evoke identifiable ECAP responses for individuals in the severe IEM group. The mild IEM group needed far lower CLs to obtain measurable ECAP responses. This was consistent with previous findings that children with cochlear nerve deficiency had higher ECAP thresholds with smaller ECAP amplitudes and smaller slopes of ECAP amplitude growth function compared to children with normal-sized cochlear nerves.[12-14] This suggests that intraoperative ECAP thresholds might reflect the physiological status and the severity of inner ear anomalies. ECAP is a method to investigate neural responsiveness in the peripheral auditory system, and measured responses largely depend on the health of spiral ganglion cells. In cases with cochlear nerve deficiency and

modiolus defective types of the malformed cochlea, the firing of spiral ganglion neurons requires an increased level of electrical current to evoke action potentials.[19] In that sense, highly elevated ECAP thresholds observed in the severe IEM group may indicate the presence of structural differences and suboptimal neural function. Manipulating parameters for electrical stimulation such as widening pulse widths and adjusting the stimulation rate may be helpful for individuals with severe IEMs to ensure enough dynamic range below out of compliance and maximize the sound quality of cochlear implant in clinical programming.

Individuals with mild IEMs, such as EVA and IP-II, achieved excellent CAP scores comparable to the normal group. Otherwise, subjects with severe malformations such as cochlear hypoplasia, narrow IAC, or cochlear nerve hypoplasia had considerably lower CAP scores, even 12 months after implantation. Previous studies reported similar results. The Mondini/IP-II group showed significantly better open- and closed-set word scores at postop 12 and 24 months compared to groups with common cavity and cochlear hypoplasia.[4] Groups with vestibular anomaly and mild cochlear malformation achieved better or comparably similar word and sentence recognition scores compared to the normal inner ear group.[1,6,7] Even though the auditory performance of individuals with severe IEMs remained suboptimal, they also benefit from CI compared to preimplant conditions (see Figure 4). When using their implant, they were able to recognize environmental noises and respond to speech sounds that they were not able to enjoy before implantation. While the functional benefit of CI was observed across groups either with or without malformation, the rate of increase was less steep in the severe IEM group compared to other groups. This might be related to aberrations in the normal anatomy course associated with more frequent nerve abnormalities such as hypoplastic or aplastic cochlear nerve and deformed bony structures such as narrow IAC and stenotic bony cochlear nerve canal.[17,22,23]

Various ECAP metrics (e.g., thresholds, amplitudes, the slope of amplitude growth function, etc.) have been explored to evaluate the relationship between ECAP responses and auditory performance.[9,10,28,29] Due to the retrospective nature of this study, ECAP thresholds via intraoperative NRT testing were allowed to access. The predictive ability of ECAP thresholds on speech perception was evaluated in a few reports, and the results were inconclusive. No significant association of speech perception with NRT thresholds has been reported in both adults and children.[29–32] In contrast, significant differences in ECAP thresholds were observed between good and poor CI performers.[33] In this study, intraoperative NRT thresholds measured from 22 electrodes had significant inverse correlations with CAP scores measured from all subjects. When compared by group, IEM subgroups showed similar inverse relationships with no statistical significance. This lack of significance in the IEM subgroups might be due to the wide variance in auditory performance that is often seen in individuals with ear anomalies and a relatively smaller number of participants in the IEM group.[1] A previous empirical report showed that some poor performers have more robust ECAP responses than good performers, which may add innate intersubject variability that is uncommon in electrophysiological measures obtained from CI users.[29] These results demonstrate the need for further investigation with a large dataset using extended ECAP metrics to understand these inconsistencies in the association between ECAP measures and auditory performance. This may also yield an alternative clinical application of intraoperative ECAP measurements to provide helpful information to estimate the future course of aural development based on neural responsiveness acquired during the surgery and determine directions to maximize CI benefits in this challenging CI population.

5 | CONCLUSION

Measurable ECAP responses and NRT thresholds varied across the normal and IEM groups. The significant inverse relationship between intraoperative NRT thresholds and postoperative CAP scores existed in general. This may suggest that electrophysiological responses routinely measured during surgery may potentially be indicative of postoperative auditory performance in our CI population.

CONFLICT OF INTEREST
The authors declare no conflict of interest.

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