ORIGINAL ARTICLE

Sixteen years of cochlear implant surgery in cochleovestibular malformation and cochlear nerve deficiency: Insights from northern Malaysia

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ABSTRACT

Introduction: Congenital sensorineural hearing loss associated with cochleovestibular malformations (CVM) and/or cochlear nerve deficiency (CND) presents distinct surgical and audiological challenges. Patients with CVM face elevated risks of intraoperative cerebrospinal fluid (CSF) gushers and aberrant facial nerve courses, while those with CND have historically been viewed as poor candidates for cochlear implantation (CI) due to concerns regarding neural sufficiency. This study aims to bridge this gap by evaluating the surgical safety and audiological outcomes of CI in a focused cohort of patients with CVM and CND at a tertiary referral centre in northern Malaysia.

Materials and Methods: A retrospective cohort study was conducted on 20 cases with CVM and/or CND performed between January 2009 and December 2024 at Hospital Sultanah Bahiyah, Kedah, Malaysia. The study population included patients with radiologically confirmed CVM classified according to the Sennaroglu classification and CND. Surgical outcomes, including the incidence of CSF gushers, were analysed by Fisher's exact test following stratification into high-risk and low-risk gusher group. Primary audiological outcomes were assessed using the aided average pure tone audiometry (PTA), analysed longitudinally using a linear mixed-effects model. Secondary functional audiological outcomes were evaluated by comparing pre-implantation and 12-month post-implantation Categories of Auditory Performance-II (CAP-II) scores using the Wilcoxon signed-rank test.

Results: The cohort was predominantly prelingual (90.0%). Surgical analysis revealed a shift in technique over the 16-year period, moving from cochleostomy to round window insertion. Intraoperative CSF gushers were encountered in 12 of 20 ears (60%). We found no statistically significant difference in the incidence of gushers between the "high risk" group (enlarged vestibular aqueduct and incomplete partition type II) and to the "low risk" group (p=0.559). There were no incidences of facial nerve injury. In terms of audiology outcome, the linear mixed-effects model revealed a highly significant improvement in aided PTA over time for all ears (p<0.001). Crucially, comparing CND versus non-CND ears revealed no statistically significant difference in outcomes, with both groups following a parallel trajectory of auditory improvement. Functional analysis confirmed that these gains translated into real-world benefits, with CAP-II scores improving significantly from a median of 2.0 pre-operatively to 4.5 at 12 months (p=0.003).

Conclusion: Cochlear implantation is a safe and effective intervention for children with CVM and/or CND. Our findings indicate that the risk of intraoperative CSF gushers extends beyond specific high-risk groups, underscoring the need for broad surgical readiness across the spectrum of malformations. The audiological outcomes observed, irrespective of the presence of CND, support the expansion of CI candidacy to this challenging population, provided there is requisite surgical expertise and thorough family counselling.

KEYWORDS:
Cochlear implants; Inner ear; Cerebrospinal fluid; Cochlear nerve; Audiology

INTRODUCTION

Congenital sensorineural hearing loss (SNHL) affects 1.5 to 3 in every 1,000 newborns.[1] A proportion of these cases, approximately 20%, are associated with structural anomalies of the inner ear, known as cochleovestibular malformations (CVM).[2] For cases of severe-to-profound sensorineural hearing loss (SNHL), cochlear implantation (CI) is the gold standard of care to restore auditory function.[3,4] In Malaysia, the Ministry of Health (MOH) National CI Programme was established in 2008 to address this need.[5] According to the programme's 10-Year Report (2008-2018), a total of 413 surgeries were performed across ten satellite hospitals nationwide.[5] However, the national data indicates that most of these recipients (91.5%) presented with normal inner ear anatomy, with cases of cochlear malformation or nerve hypoplasia representing a small minority of the national cohort.[5]

However, CI in patients with CVM involves various surgical challenges. The anomalous development of the bony

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labyrinth can lead to an aberrant course of the facial nerve, difficulties in electrode placement, and, most notably, an increased risk of intraoperative cerebrospinal fluid (CSF) leakage, commonly termed a "gusher".[6,7] A CSF gusher, defined as a profuse, pulsatile egress of CSF upon opening the inner ear, occurs due to an abnormal communication between the subarachnoid space and the perilymph.[8] These events not only complicate the surgical procedure but also increase the risk of postoperative morbidity, including meningitis.[9] To navigate this complex surgical landscape, meticulous preoperative planning is essential. High-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) are vital for diagnosis and surgical road-mapping.[10,11] The imaging-based classification system proposed by Sennaroglu et al. is the globally accepted standard for categorizing CVM, providing a crucial link between specific developmental anomalies and the potential intraoperative challenges a surgeon is likely to encounter.[7,12] In addition to the anatomical challenges posed by CVM, a separate and equally critical consideration is the integrity of the neural pathway itself: cochlear nerve deficiency (CND). This condition has a reported prevalence ranging from 33% to 39% in cases of unilateral SNHL.[13,14] CND is radiologically classified according to the Birman classification, which categorizes the condition based on MRI visualization of the cochlear nerve and CT assessment of the bony cochlear aperture, ranging from normal morphology to hypoplasia or complete aplasia of the nerve.[15] Historically considered a contraindication for CI, recent literature presents a wide spectrum of results.[16,17]

Globally, the assessment of CI outcomes relies on various assessments designed to capture both the physiological restoration of hearing and the functional acquisition of auditory skills. While the aided pure tone audiometry (PTA) thresholds remain the fundamental test for determining sound detection thresholds and verifying device function, there has been increasing emphasis on functional outcomes that reflect real-world performance. Validated scales, most notably the Categories of Auditory Performance-II (CAP-II) and Speech Intelligibility Rating (SIR), are universally adopted to track the longitudinal trajectory of auditory development.[18,19] These standardized measures are essential not only to monitor individual progress, but also to facilitate comparison across different international centres and diverse patient populations.

CI outcomes in Malaysia have generally been positive, with major centres like Universiti Kebangsaan Malaysia (UKM) and University Malaya Medical Centre (UMMC) reporting excellent long-term functional results in the general population.[20,21] However, specific data regarding the management and outcomes of the sub-population with CVM and/or CND remains underreported in our region. This study aims to address these gaps by presenting a 16-year experience from a tertiary referral centre in northern Malaysia. Our analysis includes both CVM and CND. The primary objective was to retrospectively evaluate the surgical approaches, intraoperative findings, and postoperative complications. Secondarily, we aimed to correlate specific malformations with the incidence of CSF gushers and to evaluate audiological improvements.

MATERIALS AND METHODS

This study was conducted as a retrospective, single-centre, observational chart review. The reporting of this study was structured to adhere to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) guidelines for cohort studies.[22] The study protocol received approval from the National Medical Research Registry (NMRR) of Malaysia (NMRR ID-25-04235-JKH). Given the retrospective nature of the data analysis and the de-identification of patient information, the requirement for individual patient consent was waived by the committee.

The study was performed at Hospital Sultanah Bahiyah, a tertiary referral public hospital in northern Malaysia with an established cochlear implant programme. A comprehensive review was conducted of all CI surgeries performed between January 1, 2009, and December 31, 2024. The inclusion criteria for this study were all patients who underwent cochlear implantation during the specified period and who had radiologically confirmed evidence of CVM or CND on preoperative imaging. Diagnosis was established through a combination of HRCT of the temporal bones and MRI of the internal auditory meatus (IAM) and brain. Patients with normal inner ear anatomy, hearing loss secondary to labyrinthitis ossificans, or those with incomplete medical, surgical, or radiological records were excluded. For patients who underwent bilateral CI, each ear was treated as an independent case for the analysis of radiological findings and intraoperative surgical outcomes.

Relevant information was extracted from patient medical records, operative logs, audiological files, and the Picture Archiving and Communication System (PACS) for radiological images. All preoperative imaging studies (HRCT and MRI) were reviewed. Cochleovestibular malformation (CVM) was diagnosed based on radiological abnormalities of the cochlea and/or vestibular structures on HRCT and MRI. Diagnostic criteria included abnormal cochlear morphology (reduced number of turns, absent or deficient modiolus, cystic configuration), enlarged vestibule or vestibular aqueduct (midpoint diameter >1.5 mm), and dysplastic or aplastic semicircular canals (SCC).12 The identified anomalies were classified according to the Sennaroglu classification, which includes categories such as incomplete partition type I (IP-I), incomplete partition type II (IP-II), cochlear hypoplasia (CH), and enlarged vestibular aqueduct (EVA).[12] The status of the cochlear nerve (e.g., normal, hypoplastic, aplastic) was also documented. Anomalies were noted as being isolated or occurring in combination with other malformations.

Pre-operative variables collected were patient demographics of age at the time of implantation and radiological findings as mentioned above. Intraoperative surgical information collected included cochlear access (cochleostomy or round window membrane insertion), CSF gusher, other intraoperative difficulties and electrode insertion. A "gusher" was defined as a profuse, pulsatile, and rapid flow of CSF upon opening the inner ear, while "leak" was defined as a less vigorous, gentle flow of the fluid.[8] Medical records were then reviewed for any documented immediate or delayed postoperative complications, including but not limited to wound infection, seroma, device failure or migration,

Table I: Surgical details and intraoperative complications

Surgical details	n	Rate (%)
Technique		
Insertion route		
Cochleotomy	7	35.0
Round window	13	65.0
Electrode insertion		
Complete	18	90.0
Partial	2	10.0
Intraoperative complications		
CSF gusher		
Absent	8	40.0
Present	12	60.0
Facial nerve injury		
Absent	20	100
Present	0	0

Abbreviations: CSF, cerebrospinal fluid.

Table II: Distribution of cochleovestibular malformations in the study cohort and gusher rate (n=24)

Types of malformations Cochleovestibular malformations (Sennaroglu)	n (%)	Co-existing anomalies	CSF gusher n (%)
Cochlear hypoplasia	2 (13.30)	1 hypoplastic nerve, 1 EVA	1 (50)
IP I	4 (26.70)	3 hypoplastic nerve, 1 EVA	3 (75)
IP II	7 (46.70)	1 hypoplastic nerve, 6 EVA	6 (85.70)
EVA	2 (13.30)	Not applicable	2 (100)
Cochlear nerve deficiency (CND)		Co-existing anomalies	
Isolated hypoplastic nerve	3 (37.50)	Not applicable	0 (0)
Hypoplastic nerve related with CVM	5 (62.50)	3 IP-I, 1 IP-II, 1 CH	4 (57.14)

Abbreviations: CH, cochlear hypoplasia; CND, cochlear nerve deficiency; CSF, cerebrospinal fluid; CVM, cochleovestibular malformation; EVA, enlarged vestibular aqueduct; IP, incomplete partition.

Table III: Incidence of intraoperative CSF gusher stratified by anomaly risk group

Anomaly type	With CSF Gusher	No CSF Gusher	Total
High-Risk Group (EVA & IP-II)	8	1	9
Low-Risk Group (IP-I, CH)	4	2	6
Total	12	3	15

Abbreviations: CH, cochlear hypoplasia; CSF, cerebrospinal fluid; EVA, enlarged vestibular aqueduct; IP, incomplete partition.

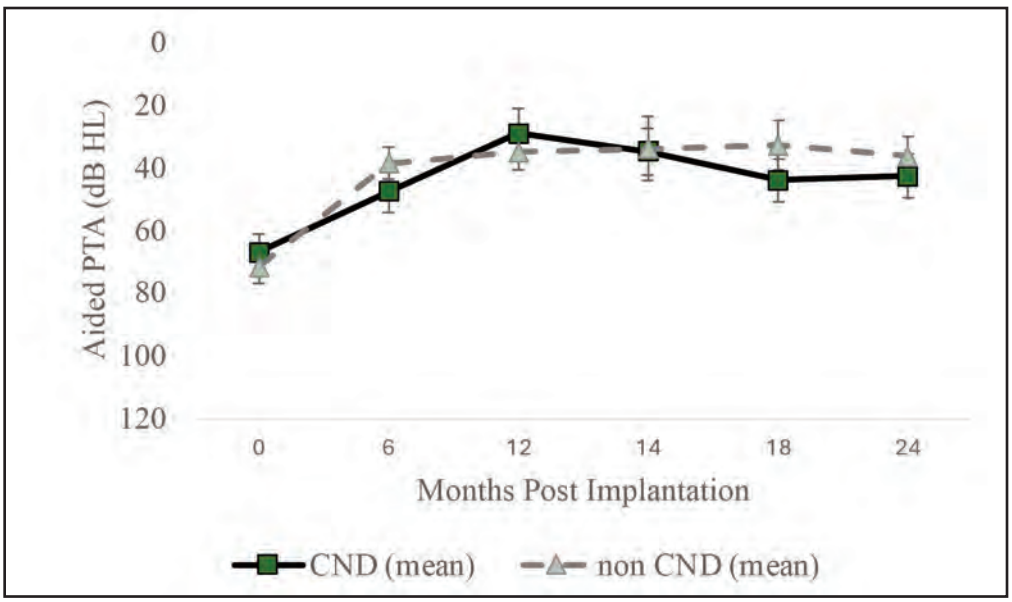

Fig. 1: Longitudinal comparison of aided pure tone average (PTA) between the cochlear nerve deficiency (CND) and non-CND groups. Data points represent the mean aided PTA at each follow-up interval. Error bars indicate the standard error of the mean. Both groups exhibit a significant and parallel improvement in hearing thresholds over time ($p<0.001$), with no significant difference between groups ($p > 0.05$).

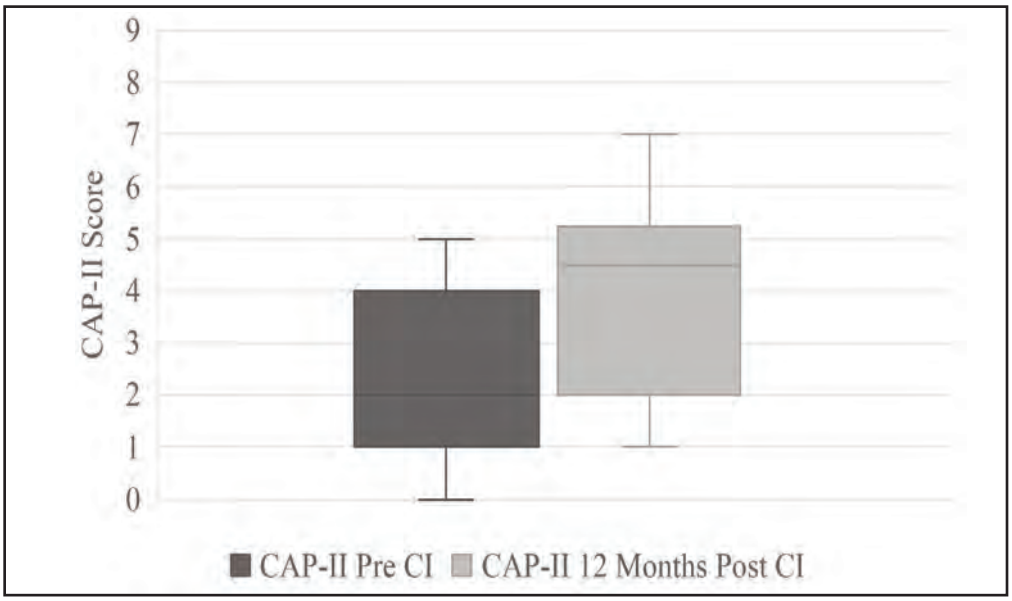

Fig. 2: Comparison of Categories of Auditory Performance-II (CAP-II) scores pre-implantation and 12 months post-implantation. The box plot illustrates the significant improvement in functional auditory performance following cochlear implantation (p=0.003, Wilcoxon signed-rank test). The horizontal line within the box represents the median score, while the box edges represent the interquartile range

persistent CSF leakage, and meningitis. Audiological outcomes were assessed using average aided pure tone audiometry (PTA) thresholds across four frequencies (500 Hz, 1 kHz, 2 kHz, and 4 kHz) in dB HL during follow-up visits up to 24 months. Functional auditory performance was evaluated using CAP-II scores obtained preimplantation and 12 months post implantation.

All data were analysed using SPSS Statistics for Windows, Version 29.0.2.0.[20] To assess the association between anatomical risk and CSF gushers, cases with only CND were first excluded. The remaining patients were stratified into a high-risk group (EVA & IP-II) and a low-risk group (IP-I and CH). A Fisher's exact test was used to determine the significance of this association. To analyse the longitudinal aided PTA data, the unit of analysis was the operated ear. A linear mixed-effects model was employed. The model included time (follow up months) and CND status (CND vs non-CND) and their interaction term as fixed effects, with a random intercept for each subject. For the functional CAP-II score analysis, the unit of analysis was the individual patient. To ensure independence of data, bilateral cases were considered as a single observational unit. A Wilcoxon signed-rank test was used to compare the paired pre- and post-implantation CAP-II scores. A p-value of < 0.05 was considered statistically significant.

RESULTS

Over the 16-year study period, a total of 20 CI cases met the inclusion criteria. This included 20 operated ears from 16 patients. There were 4 patients who underwent bilateral CI at different setting, with each ear considered as individual cases. Most patients were prelingual (n=18, 90.0%). There were only 2 syndromic cases, which included a single case of CHARGE syndrome and Down syndrome each. Age of implantation ranged between 8 months to 53 months old and 6 to 16 years old for prelingual and postlingual cases respectively.

A review of the surgical records revealed an evolution in the preferred technique for cochlear access over the study period. Prior to 2020, promontorial cochleostomy was the standard approach. From 2020 onwards, a deliberate shift was made towards round window insertion as the primary technique, reflecting a centre-wide adoption of principles aimed at less traumatic surgery and structural preservation.

Regarding electrode insertion, a successful full insertion of the electrode array was achieved in 18 of the 20 patients (90.0%) as shown in Table I. The two cases of partial electrode insertion both occurred in patients with an IP-I malformation. No other significant intraoperative difficulties, such as an aberrant facial nerve requiring a change in surgical approach, were documented in the operative reports.

Secondarily, the analysis of functional outcomes via CAP-II score was performed at the patient level. This analysis was performed in 14 patients, after the exclusion of 2 cases with incomplete CAP-II data. The Wilcoxon signed-rank test confirmed that the improvement in CAP-II scores from pre-implantation (median = 2) to 12 months post-implantation (median = 4.5) was statistically significant (p=0.003). This indicates that the objective hearing gains translated into meaningful, real-world improvements in auditory performance across the diverse range of complex cases.

Intraoperatively, no cases of facial nerve injury were recorded. CSF gusher was encountered in 12 of the 20 patients (60%). Major postoperative complications were confined to one patient with IP-II who had experienced an intraoperative gusher, who developed a CSF leak and subsequently bacterial meningitis but recovered fully with medical therapy.

The distribution of various anomalies with gusher rate is detailed in Table II. Among the 15 patients with CVM according to Sennaroglu classification, the most frequently

observed malformation was IP-II, accounting for 7 cases (46.7%). This was followed by IP-I in 4 cases (26.7%). The remaining CVM consisted of CH and isolated EVA. All cases of CND were radiologically classified as hypoplastic, and one-third of these were isolated findings.

The rate of CSF gusher varied across the different types of CVM. The highest rates were observed in patients with EVA, where 100% of cases experienced a gusher, and in patients with IP-II, with an 85.7% incidence.

To assess the relationship between anomaly type and CSF gushers, the cohort was stratified into a high-risk group (comprising patients with EVA and IP-II) and a low-risk group (comprising patients with IP-I and CH). The incidence of intraoperative CSF gusher was not statistically significant between the high-risk group and the low-risk group (p=0.559, Fisher's exact test) (Table III).

Of the 20 cases in the initial cohort, 1 were lost to follow-up and did not have post-implantation PTA data available. Therefore, statistical analyses of PTA outcomes were performed on the remaining 19 cases who had complete pre- and post-operative datasets.

To analyse the longitudinal trajectory of audiological outcome, a linear mixed-effects model was performed with aided PTA as the dependent variable. The model included fixed effects for time (in months), risk group (CND vs non-CND), and their interaction, with a random intercept for each patient. This statistical analysis performed revealed three key findings. First, there was a significant improvement in hearing thresholds over time for all patients in the cohort ($p<0.001$). Second, there was no statistically significant difference in the overall aided PTA between the CND and non-CND groups. Third, the interaction between group and time was not significant, indicating that both groups followed a similar, parallel trajectory of improvement over the 24-month follow-up period. This suggests that the rate of objective auditory improvement was comparable, regardless of the presence of CND.

DISCUSSION

This 16-year retrospective review provides valuable insights into the surgical management and outcomes of cochlear implantation (CI) in a diverse cohort of patients with CVM and/or CND from a regional centre in northern Malaysia. Our findings complement the broader National MOH CI Programme 10-Year Report (2008-2018).[5] While the national report identified normal inner ear anatomy in nearly 95% of prelingual recipients, our study focused exclusively on the remaining complex minority, specifically those with CVM and/or CND who presented distinct surgical and audiological challenges. Our findings underscore that meticulous preoperative planning is essential, and they help to refine our understanding of surgical risk and expand the boundaries of CI candidacy. Beyond simply reporting outcomes, this study offers a unique perspective from a regional centre in northern Malaysia, contributing to the global understanding of these complex cases.

CVMs are diagnosed radiologically according to the classification proposed by Sennaroglu.[7] The Sennaroglu classification categorizes CVMs according to the stage of embryologic arrest and associated surgical implications.[7] As encountered in our cohort, IP-I, IP-II, CH and EVA were identified. IP-I is characterised by a cystic cochlea lacking internal architecture, with complete absence of the modiolus and interscalar septa, typically associated with a dilated vestibule.[7] In contrast, IP-II demonstrates cystic fusion of the middle and apical cochlear turns with a preserved basal turn and is commonly associated with EVA.[7] CH refers to a cochlea that is smaller than normal with varying degrees of internal structural deformity.[7] It is typically characterised by reduced cochlear turns (often fewer than 2½ turns) and diminished overall dimensions on CT or MRI.[7] Lastly, EVA is diagnosed when the vestibular aqueduct diameter exceeds 1.5 mm at the midpoint between posterior labyrinth and operculum, in the background of normal cochlea, vestibule and SCC.[7] The status of the cochlear nerve was assessed on MRI, with hypoplasia defined as a smaller calibre nerve compared to the facial nerve, and aplasia defined as absence of a visible nerve within the internal auditory canal.7 Diagnostic criteria for other CVMs are not detailed here as they were not observed in our cohort.

The National MOH Report cited a low overall complication rate of 5.57%, with CSF gushers noted as an intraoperative finding rather than a major complication.[5] In stark contrast, our focused cohort of complex anomalies revealed a gusher incidence of 60%. While previous literature has often stratified risk based on specific anomalies, our analysis of this cohort revealed no statistically significant difference in the incidence of CSF gushers between the "high-risk" group (EVA & IP-II) and the "low-risk" group (IP-I & CH).[8,23,24] Nevertheless, cases of EVA and IP-II had the highest and second highest rate of CSF gushers in our cohort respectively. In IP-II, a deficient modiolus creates a wide, patent communication between the cochlea and the CSF-filled internal auditory canal.[23] Similarly, EVA is frequently associated with a patent and enlarged cochlear aqueduct, providing another direct conduit for CSF.[25] The lack of statistical significance in our risk stratification analysis must be considered along with two important factors. First, the small sample size (n=15) possibly limits the statistical power of the study, increasing the likelihood of a Type II error where a true difference fails to reach the threshold of significance. Second, and clinically more important, is the unexpectedly high incidence of gushers (66.7%) within the "low-risk" group. This was primarily driven by patients with IP-I. While typically considered lower risk than IP-II or EVA, IP-I is characterized by the absence of the cribriform plate, which creates a direct, low-resistance pathway for CSF to enter the cochlea.[7] Consequently, our data suggests that the risk of a gusher is not confined to specific "high-risk" categories but is a constant challenge across the spectrum of major CVM. Therefore, surgeons should maintain a high index of suspicion and prepare for a gusher in all cases of CVM. Literature describes various techniques of CSF gusher management, such as tight packing of the cochleostomy or round window niche with temporalis muscle, fascia, fat tissue or fibrin glue immediately following electrode insertion.[8,26] Interestingly, in our centre, we do not routinely perform any

packing. The fact that only one patient in our high-risk group developed a postoperative CSF leak suggests that a snug fit of the electrode array alone can effectively arrest the fluid egress, rendering the use of any packing unnecessary in most cases.

An important insight from our data is the clear link between anatomy, intraoperative events, and postoperative complications. The confinement of all major complications (persistent CSF leak and meningitis) exclusively to patients with IP-II malformations demonstrates a clear pathophysiological cascade. As mentioned by Sennaroglu et al., third window defect of EVA transmitting CSF pressure into the cochlea leads to a high probability of a high-pressure intraoperative gusher.[7] It is more challenging to seal such a CSF gush than managing a low-pressure ooze. Consequently, the imperfect seal then creates a persistent fistula between the non-sterile middle ear and the sterile subarachnoid space, serving as a direct pathway for bacteria and placing the patient at a significantly elevated risk for life-threatening meningitis.[9] This suggests that preoperative identification of IP-II is of importance not only for managing the gusher but, more critically, for mitigating the risk of severe postoperative morbidity. Interestingly, our study discovered a shift in surgical technique over the last decade, moving from promontorial cochleostomy to round window CI electrode insertion. This not only reflects global trends towards atraumatic and less invasive methods, but it also holds specific advantages in dysplastic cochlea.[27] In cases of IP-II or EVA where a gusher is anticipated, the round window approach avoids the drilling of cochlear bone, thereby reducing the risk of inadvertent injury to an aberrant facial nerve a common anomaly in this population.[9,28] Although our sample size is insufficient to statistically compare gusher rates between approaches, the absence of facial nerve injuries in our cohort supports the safety of this surgical strategy. Importantly, our study decouples this intraoperative surgical challenge from the audiological success. Despite a high incidence of CSF gushers, our data demonstrated significant improvements in both aided PTA and CAP-II scores. This supports the view that a CSF gusher is an intraoperative event to be managed, rather than a negative prognostic indicator for long-term hearing outcomes, provided a secure seal is achieved. The profound audiological benefits demonstrated in our cohort strongly support the argument that this predictable and manageable surgical risk is justified.

To assess the benefit of CI, we utilized the aided PTA as one of our audiological outcomes. While speech perception scores are often considered the gold standard, aided PTA was selected given that some of our patients are cases of bilateral CI in different period of life. While the National MOH Report utilized aided thresholds to categorize performance (≤40dBHL vs >40dBHL), we employed a linear mixed-effects model to track longitudinal continuous data at the ear level rather than the patient level.[5] Analysing at the ear level allowed us to isolate the specific impact of the anatomical anomaly on hearing thresholds in individual ear. The most compelling finding of our study and the one with the most significant clinical implication relates to the outcomes in patients with CND. Historically, CND has been viewed with caution or even considered a contraindication for CI due to concerns that the hypoplastic nerve would fail to transmit electrical stimulation effectively stimulation.17 In our study, when comparing the CND and non-CND groups, there was no statistically significant difference in aided PTA outcomes. Crucially, the interaction between the group (CND vs non-CND) and time was not significant. This indicates that both groups followed a parallel trajectory of improvement over the 24-month follow-up period. This suggests that the presence of a radiologically hypoplastic nerve does not inherently limit the rate of auditory learning or neural adaptation in the first two years post-implantation. These findings align with emerging evidence suggesting that children with CND can achieve comparable performance and should not be considered an absolute contraindication for CI.[29] Indeed, other studies have found that children with CNH can achieve performance comparable to their matched counterparts, especially with longer CI usage.[17] Therefore, CND should not be an absolute barrier to implantation. Instead, these excellent outcomes support the expansion of CI candidacy to this population, provided there is requisite surgical expertise and thorough family counselling regarding variable outcomes.

While aided PTA measures auditory detection, it does not necessarily reflect function as in how the patients uses hearing in daily life. To address this, we analysed the Categories of Auditory Performance-II (CAP-II) scores.18 Unlike the ear-level analysis used for aided PTA, CAP-II was analysed at the patient level, treating bilateral cases as a single unit to reflect the patient's overall functional ability. The significant increase in CAP-II scores confirms that the audiological detection measured by PTA translates into meaningful, real-world benefits. It demonstrates that even in distinctively complex cases such as those involving high-risk gushers, anatomical distortions, or nerve deficiency, patients are not just "hearing" sound but are developing the requisite listening behaviours to interact with their environment.

Our findings contribute to the growing body of evidence regarding cochlear implant safety and audiological efficacy within the Malaysian context. Landmark longitudinal studies by Goh et al. from UKM and Konting et al. from UMMC have previously established that the general paediatric CI population in Malaysia achieves high rates of oral communication and mainstream education integration.[20,21] While these studies primarily characterized the general population, recent local data indicates that this positive trend extends to the distinctively complex demographic of CVM and CND. Notably, Misron et al. at Hospital Sultan Ismail reported significant improvements in CAP-II and Meaningful Auditory Integration Scale (MAIS) scores within their CVM cohort, while Abdullah et al. at UKM observed similar significant gains in CAP-II and SIR scores among children with CVM and/or CND.[17,30] Supporting these findings, the statistically significant improvement in aided PTA and CAP-II scores observed in our cohort validates that with appropriate surgical planning, even patients with complex anatomy can achieve functional outcomes comparable to their peers.

Looking forward, there is a clear need for prospective, multi-centre research within the region to overcome the statistical

limitations inherent to single-centre retrospective reviews. The establishment of a collaborative national registry for complex CI cases encompassing both MOH and university hospitals would provide the necessary sample size to perform more detailed analyses of specific malformation subtypes. Most importantly, any future studies must incorporate standardized, long-term audiological and speech outcome measures. This will be essential for building a comprehensive understanding of the relationship between inner ear anatomy, surgical complexity, and the success of auditory rehabilitation in this challenging and deserving patient population.

CONCLUSION

This 16-year review from northern Malaysia demonstrates that cochlear implantation is a safe and effective intervention for patients with CVM and/or CND. Meticulous preoperative radiological evaluation is of vital importance. However, our findings highlight that the risk of intraoperative CSF gushers extends beyond classically defined high-risk groups, necessitating proactive surgical readiness for this complication across the spectrum of CVM. Most importantly, this study reinforces the evolving consensus that CI candidacy can and should be extended to anatomically complex cases, including those with CND, provided there is the requisite surgical expertise and a framework for thorough, realistic preoperative counselling for patients and their families.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no actual or potential conflict of interest in relation to this article.

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