ISSN: 2083-389X
eISSN: 2084-3127

J Hear Sci, 2025; 15(4): 61–68
DOI: 10.17430/jhs/215470

Received:
August 21, 2025
Revision accepted:
December 11, 2025
Published online:
February 2, 2026

LOW-FREQUENCY COCHLEAR IMPLANT (CI) STIMULATION AND PRESERVED HIGH FREQUENCY HEARING IN A CASE OF PARTIAL DEAFNESS: POSSIBLE EXPANSION OF CI CANDIDACY

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: Thanks to ongoing advancements in cochlear implant (CI) technology and surgical techniques, the eligibility criteria for CIs have expanded to include patients with various levels of low-frequency hearing. However, there is another group of patients with non-functional (or borderline functional) hearing at low frequencies but preserved residual hearing in the high-frequency range, thus making them off-label for a CI even though they get limited benefit from a hearing aid.

Case report: This study presents a 47-year-old patient with residual hearing at the functional border (75 dB HL or better) for low and mid frequencies (125–1500 Hz) and functional residual hearing (70 dB HL or better) for high frequencies (2000–8000 Hz). CI surgery was performed using the Med-El Flex26 electrode via round window insertion. Hearing preservation (HP) was complete up to at least 24 months and partial up to 36 months. For the high-frequencies only (2000–8000 Hz) there was complete HP for the entire 36 months. At 12 months post-surgery, the patient's word recognition scores (WRS) had improved by 75 percentage points in quiet and 70 points in noise.

Conclusions: The patient's results demonstrate that preserving functional residual hearing in the basal cochlea is possible after CI surgery, even though this region is very susceptible to insertion trauma. The presence of functional high-frequency hearing should not be the only reason for withholding CI surgery, especially if a hearing aid is ineffective.

Keywords: cochlear implant • partial deafness treatment • round window insertion • hearing preservation • electrode insertion trauma

STYMULACJA ZA POŚREDNICTWEM IMPLANTU ŚLIMAKOWEGO W ZAKRESIE NISKICH CZĘSTOTLIWOŚCI I ZACHOWANEGO SŁUCHU WYSOKOCZĘSTOTLIWOŚCIOWEGO W PRZYPADKU CZĘŚCIOWEJ GŁUCHOTY: MOŻLIWE ROZSZERZENIE WSKAZAŃ DO IMPLANTU ŚLIMAKOWEGO

Streszczenie

Wprowadzenie: Dzięki stałemu postępowi w technologii implantów ślimakowych oraz technik chirurgicznych kryteria kwalifikacji do wszczepienia implantu ślimakowego uległy rozszerzeniu i obecnie obejmują pacjentów z różnym stopniem zachowanego słuchu w zakresie niskich częstotliwości. Istnieje jednak grupa pacjentów z niefunkcjonalnymi resztkami słuchu (lub resztkami słuchu na granicy funkcjonalności) w zakresie niskich częstotliwości, przy jednoczesnym zachowaniu funkcjonalnych resztek słuchu w zakresie wysokich częstotliwości. Pacjenci ci pozostają poza aktualnymi wskazaniami audiologicznymi do wszczepienia implantu ślimakowego, pomimo ograniczonych korzyści słuchowych ze stosowania aparatów słuchowych.

Opis przypadku: W pracy przedstawiono przypadek 47-letniego pacjenta z resztkowym słuchem na granicy funkcjonalności (≤75 dB HL) dla niskich i średnich częstotliwości (125–1500 Hz) oraz funkcjonalnymi resztkami słuchu (≤70 dB HL) dla wysokich częstotliwości (2000–8000 Hz). Procedurę wszczepienia implantu ślimakowego przeprowadzono z użyciem elektrody Med-El Flex26, stosując dostęp przez okienko okrągłe. Uzyskano całkowite zachowanie słuchu (*hearing preservation*, HP) do 24 miesięcy po operacji oraz częściowe zachowanie słuchu do 36 miesięcy obserwacji. W zakresie wyłącznie wysokich częstotliwości (2000–8000 Hz) odnotowano całkowite zachowanie słuchu przez cały 36-miesięczny okres obserwacji. Po 12 miesiącach od zabiegu wyniki rozpoznawania słów (word recognition scores, WRS) poprawiły się o 75 punktów procentowych w ciszy oraz o 70 punktów procentowych w szumie.

Wnioski: Uzyskane wyniki wskazują, że zachowanie funkcjonalnych resztek słuchu w podstawnej części ślimaka jest możliwe po implantacji ślimakowej, chociaż obszar ten jest najbardziej podatny na uraz związany z insercją elektrody. Obecność funkcjonalnych resztek słuchu w zakresie wysokich częstotliwości nie powinna stanowić jedynego powodu do wykluczenia pacjenta z kwalifikacji do wszczepienia implantu ślimakowego, szczególnie w przypadku, gdy aparaty słuchowe są nieskuteczne.

Słowa kluczowe: implant ślimakowy • leczenie częściowej głuchoty • insercja przez okienko okrągłe • zachowanie słuchu • uraz ślimaka związany z insercją elektrody

Key to abbreviations	
AC	air-conduction
BC	bone-conduction
BIAP	International Bureau of Audiophonology
CI	cochlear implant
CT	computed tomography
ENS	electro-natural stimulation
ESPCI	European Symposium on Paediatric Cochlear Implantation
HP	hearing preservation
PDT-EAS	Partial Deafness Treatment–Electro-Acoustic Stimulation
PDT-EC	Partial Deafness Treatment–Electric Complementation
PDT-ES	Partial Deafness Treatment–Electric Stimulation
PTA	pure-tone average
SNHL	sensorineural hearing loss
SRA	Straight Research Array
WRS	word recognition scores

Introduction

The term “cochlear implant” (CI) was first introduced into the medical literature in 1973 during an international conference in San Francisco on the use of electrical stimulation of the cochlear nerve to treat deafness in humans [1,2]. (Incidentally, that conference was attended by Blair Simmons, Robert White, William House, Jack Urban, and Claude Henri Chouard.) Not long afterwards, cochlear implantation became a standard procedure for treating deafness [3,4].

In Poland, the first cochlear implantation surgery was performed by Henryk Skarzynski in 1992 [5] and the technique has since given hope to thousands of deaf children and adults in the country. The positive results achieved in cases of complete deafness encouraged scientists and clinicians to gradually expand the indications for CI surgery in individuals with some residual hearing (referred to at the time as “non-functional” hearing).

The initial group included patients with preserved but non-functional residual hearing (Partial Deafness Treatment–Electric Stimulation, PDT-ES). Another group comprised patients with residual low-frequency hearing needing combined electro-acoustic stimulation, referred to as Partial Deafness Treatment–Electro-Acoustic Stimulation (PDT-EAS). In 2002, indications for cochlear implantation were expanded to adults with partial deafness [6], and in 2004, to children [7]. This included patients with Partial Deafness Treatment–Electric Complementation (PDT-EC), who had normal or nearly normal low-frequency hearing (up to 500 Hz) but required electrical stimulation for mid and high frequencies. Finally, indications were extended to patients with fully functional hearing in the 125–1500 Hz range but deafness at other frequencies, classified as electro-natural stimulation (ENS) [8,9].

All CI procedures in PDT treatment are based on a minimally invasive surgical approach, referred to earlier as the 6-step Skarzynski procedure, which involves inserting the electrode through the round window into the scala tympani [10]. Further development of PDT using CIs was also made possible by advances in technology, including the creation of delicate, flexible electrodes of varying lengths [11]. Since 1997, we have mostly used the Med-El system with flexible electrodes, including the Flex20, Flex24, Medium, and Flex28. In 2009, during the 9th European Symposium on Paediatric Cochlear Implantation (ESPCI) in Warsaw, Skarzynski performed the first demonstration operation using the Cochlear Nucleus Straight Research Array (SRA) electrode, developed according to his concept [12,13]. Subsequently, the next-generation Cochlear electrodes (422, 522, 622) have been used. Since 2017, the Advanced Bionics HiFocus SlimJ electrode has also been used in PDT [14]. Additionally, the Neuro Zti Oticon electrode has been used exclusively for patients diagnosed with minimally functional residual hearing prior to surgery [15].

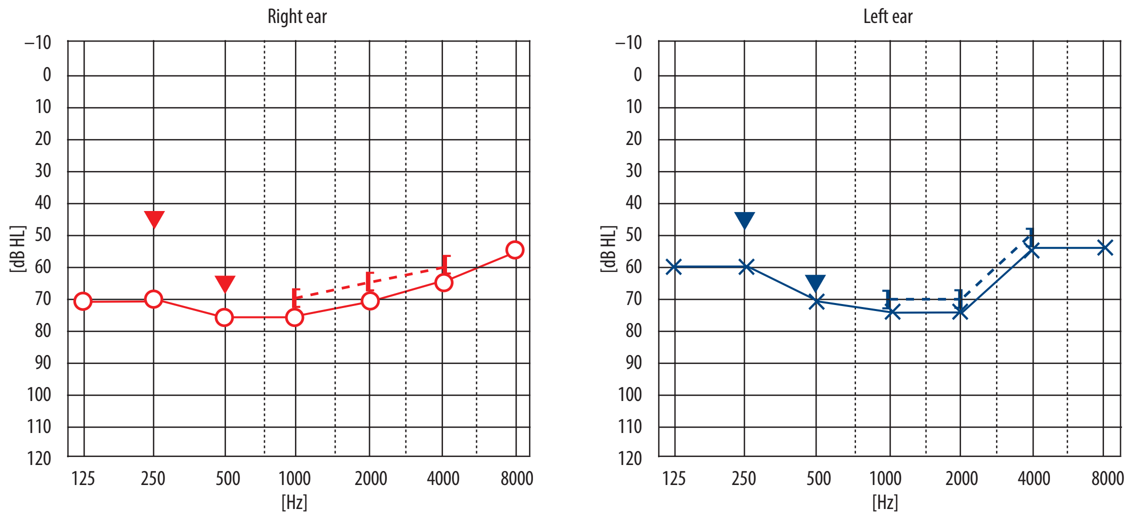

Figure 1. Patient's preoperative pure-tone audiometry for the right and left ears

Worldwide, similar procedures aimed at preserving residual hearing have also been carried out at other centers. However, the level of functional residual hearing varies among clinicians [16–23].

New electrodes of novel designs and varying lengths have been particularly successful in patients with high-frequency hearing loss (a "ski-slope" audiogram). At the same time, they have led to fresh challenges in everyday clinical practice, since they expand the current surgical indications and options. One notable new scenario, and the focus of this report, is called Partial Deafness Treatment–Low Frequency Stimulation. This type of hearing loss can, in a sense, be considered the 'reverse' of the typical partial deafness, since such patients have non-functional (or borderline functional) hearing in the low-frequency range but have preserved residual hearing in the high-frequency range. Traditionally, to preserve hearing at high frequencies, the management of this kind of partial deafness has generally involved the use of multichannel hearing aids; however, they often result in limited effectiveness in terms of speech understanding.

It now appears that a CI can effectively work in this scenario. This paper reports the effectiveness of a CI, including a measure of HP outcome, in a patient with preoperative functional residual hearing in the high-frequency range.

Case report

A 47-year-old patient with bilateral, sensorineural hearing loss (SNHL) first presented to the Oto-Rhino-Laryngology Surgery Clinic at our center in 2018. During the medical interview, it was noted that at age 40, the patient experienced his first episode of sudden SNHL in the right ear, accompanied by severe tinnitus and vertigo. Despite hospitalization and intravenous dexamethasone treatment, the hearing loss in the right ear progressed slowly and gradually At age 46, the patient experienced another episode of bilateral sudden SNHL with vertigo. Despite hospitalization and treatment, which included oral steroids, intratympanic steroid injections via a drain, and hyperbaric oxygen therapy, no hearing improvement was achieved. The patient had been using hearing aids bilaterally, although irregularly, due to limited effectiveness. The patient has not undergone genetic testing.

Preoperative audiometric evaluation

Pure-tone audiometry was performed to assess air-conduction (AC) thresholds at 125–8000 Hz, and bone-conduction (BC) thresholds at 250–4000 Hz, in accordance with clinical standards (ISO 8253-1: 2010) [24]. Results of preoperative pure-tone audiometry are presented in **Figure 1**. The pure-tone average (PTA) for AC at frequencies of 500, 1000, 2000, and 4000 Hz was 71.2 dB HL for the right ear and 68.8 dB HL for the left. According to the classification of hearing loss proposed by the International Bureau of Audiophonology (BIAP) [25], the patient's hearing loss was classified as severe in the right ear and moderate in the left. For this reason, the right ear was classified as suitable for CI surgery. Preoperative hearing thresholds in the right ear were 75 dB or better for low and medium frequencies (125–1500 Hz), while for high frequencies (2000–8000 Hz) they were 70 dB HL or better.

Surgical procedure

In March 2021, the patient underwent CI surgery. To protect the delicate structures of the inner ear and, specifically, to preserve residual hearing at low frequencies, which were on the border of functional hearing, the decision was made to use a flexible electrode array from Med-El (GmbH, Innsbruck, Austria). Based on preoperative computed tomography (CT) images of the temporal bone and Med-El's Otoplan tool (**Figure 2**), the Flex26 electrode was selected. Virtual 3D scala tympani insertion analysis revealed that the 26 mm electrode array had a high likelihood of providing complete cochlear coverage across the full range of cochlear duct lengths, with a cochlear angle of 475° corresponding to a frequency of 521 Hz. The surgical procedure was performed by the senior surgeon

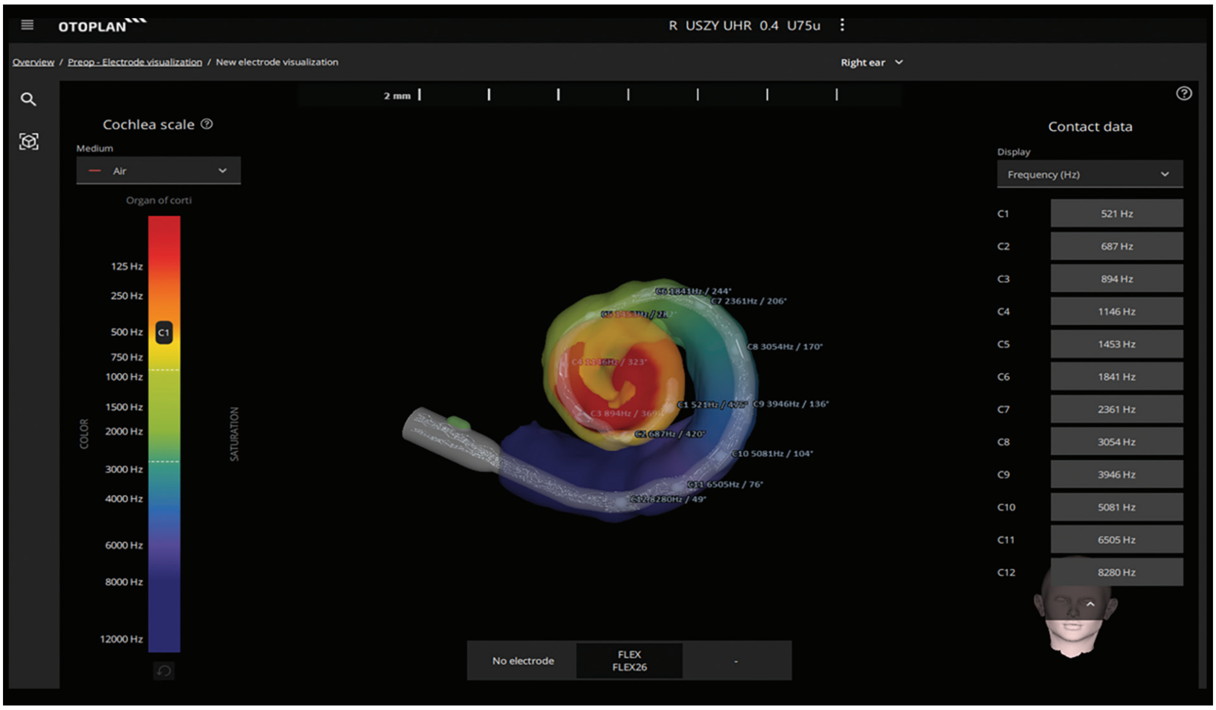

Figure 2. Preoperative visualization of the patient's cochlea and insertion of the Med-El Flex26 electrode, based on Med-El's Otoplan

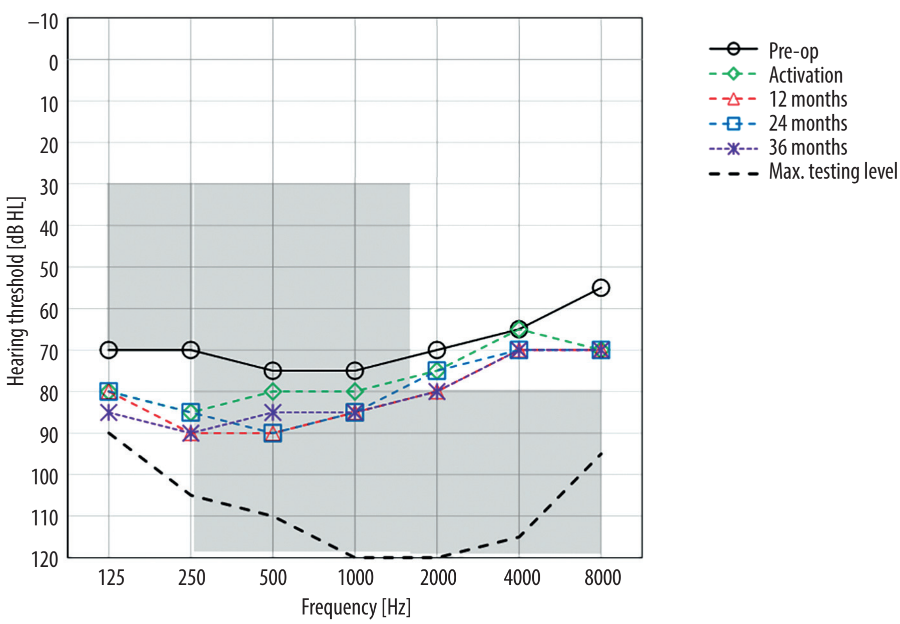

Figure 3. Pre- and postoperative air conduction hearing thresholds for the implanted ear. The shaded area represents the range of indications for electro-acoustic stimulation according to Skarzynski's concept

(the first author of this paper). A minimally invasive round-window approach, following the Skarzynski six-step procedure [10] was used, with full insertion of the CI electrode. Steroid (dexamethasone) was administered to the patient according to the procedure previously described by Skarzynska et al. [26].

Postoperative audiometric evaluation

Postoperative pure-tone audiometry was performed 1 month after surgery (at activation), and subsequently at 12, 24, and 36 months. The pre- and postoperative AC thresholds for the operated ear are shown in **Figure 3**.

Before surgery, the mean AC threshold at all tested frequencies was 68.5 dB HL. At activation, the mean AC thresholds increased by 8.1 dB HL. In the longer postoperative follow-up periods (i.e., 12, 24, and 36 months), the mean AC thresholds increased, with the maximum shift being 12 dB HL compared to preoperative values. When analyzing only the high-frequency range (at 2, 3, 4, 6, and 8 kHz), the mean preoperative AC thresholds were

Table 1. Results of hearing preservation according to Skarzynski's HP classification

Postoperative follow-up	Hearing preservation [%]	Evaluation
1 month	85.4	Complete HP
12 months	75.3	Complete HP
24 months	78.7	Complete HP
36 months	74.2	Partial HP
Key		
S	**Evaluation**	
> 75%	Complete HP	
26–75%	Partial HP	
1–25%	Minimal HP	
No detectable hearing	Loss of hearing	

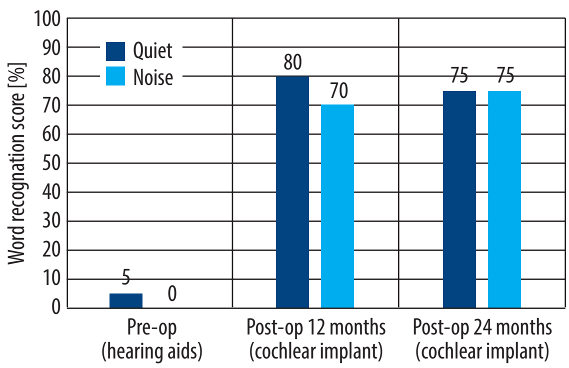

Figure 4. Word recognition scores in quiet and noise, before and after cochlear implantation

63.5 dB HL. During activation, the mean high-frequency AC thresholds increased by 6.5 dB HL. In the longer postoperative follow-up periods (i.e., 12, 24, and 36 months), the mean high-frequency AC thresholds increased, with the maximum shift being 9.5 dB HL compared to preoperative values.

Hearing preservation evaluation

The HP classification system proposed by Skarzynski et al. [27], based on preoperative and postoperative pure-tone audiometry thresholds (including 11 audiometric frequencies), was used to calculate HP. Hearing thresholds were measured five times: preoperatively, 1 month after surgery (at activation), 12 months post-surgery, 24 months post-surgery, and 36 months post-surgery. The exact HP values (S values) obtained for the patient are presented in **Table 1**. Calculating the percentage change in hearing thresholds postoperatively according to this HP classification showed that hearing was completely preserved up to 24 months post-surgery and partially preserved up to 36 months post-surgery.

Additionally, HP was assessed separately for high frequencies (at 2, 3, 4, 6, and 8 kHz). After surgery, the calculated S-value at the 1-month follow-up was 86%, at 12 months it was 81.7%, at 24 months 83.9%, and at 36 months 79.6%. These results indicate complete HP throughout the postoperative follow-up period.

Speech discrimination evaluation

The Demenko & Pruszewicz Polish Monosyllabic Word test [28] was used to assess the word recognition score (WRS) in free field at 65 dB SPL. Prior to surgery, the test was conducted with a fitted hearing aid in both quiet and noisy conditions (at a signal-to-noise ratio of +10 dB). To assess speech recognition gain, the WRS with the CI processor was performed 12 and 24 months post-surgery. The WRS results (**Figure 4**) show that over a period of 12 months, speech recognition improved from 5% to 80% in quiet and from 0% to 70% in noise. These improvements remained stable throughout the 24-month postoperative follow-up.

Discussion

The insertion of an electrode array during CI surgery often causes trauma to the cochlea, leading to ganglion cell death, necrosis, and apoptosis [29,30]. For patients with residual acoustic hearing, preserving both the residual hearing and the structural integrity of the cochlea during and after CI surgery is crucial to improving hearing outcomes [31]. CI users with residual low-frequency hearing exhibit better speech understanding, particularly in challenging listening conditions [32]. The design of the electrode array, including its length, is crucial in minimizing trauma during surgery [33]. While shallow electrode insertions may help preserve apical organ of Corti structures, the extent of trauma to the basal cochlea during insertion remains unclear.

This paper reports what is thought to be the first results of an adult patient with partial deafness and functional residual high-frequency hearing who underwent cochlear implantation. The etiology of his sudden hearing loss could not be determined; however, a genetic contribution cannot be excluded. Phenotypic analysis using AudioGene software [34] indicated that the patient's audiogram most closely matches the hearing loss pattern associated with pathogenic variants in the *WFS1* gene, which is typically linked to autosomal dominant low-frequency sensorineural hearing loss. The characteristic audiometric profile – marked by the largest threshold deterioration at low and mid frequencies, with relatively preserved hearing above 4 kHz – is consistent with the described phenotype of this gene. Additional loci suggested by the analysis, CCDC50 and MYO7A, may also contribute to autosomal dominant forms of hearing loss; however, their associated phenotypes generally involve higher frequencies or demonstrate greater progression. The resulting gene ranking supports the prioritization of *WFS1* in molecular diagnostic testing, which may carry both prognostic value and implications for clinical management and genetic counseling.

Although hearing aids are typically recommended for residual high-frequency hearing, their benefits can be limited, which supports the decision to proceed with cochlear implantation [35]. Audiograms alone may not reveal cochlear dysfunction, as dead regions in outer and inner hair cells can limit hearing aid effectiveness [36]. Vinay et al. [37] found dead regions in 59% of ears with SNHL, especially at frequencies above 70 dB HL. This may explain the low WRS outcomes observed in our patient, which were not consistent with their hearing thresholds. Cochlear implantation was the only viable option to restore effective hearing. After 12 months, the patient showed significant improvement in speech recognition, with a 75 percentage point increase in quiet and 70 points in noise. The decision to use a 26 mm electrode array was made to minimize intracochlear trauma during implantation, especially at low frequencies (up to 500 Hz), where the patient's preoperative hearing was on the borderline of functional residual hearing.

The Flex26 is the newest lateral-wall electrode array developed by Med-El, having a length between their medium and long arrays [38]. It offers the same active stimulation range (20.9 mm) as the shorter Flex24 but provides greater angular insertion depth [39]. Ketterer et al. [40] reported that the insertion angle of the Flex26 ranged from 377° to 601°, with a mean of 517° (*SD* 60°), showing no displacement from scala tympani. In contrast, displacement was observed with longer electrodes, such as the FlexSoft and Flex28. Other studies have shown that the insertion angle depends not only on electrode array type but also cochlear length [41,42]. Timm et al. [43] recommend incorporating preoperative cochlear length assessment into routine care due to cochlear size variations. Additionally, interventions like steroids or intraoperative electrocochleography [30] can further improve HP. In our patient, preoperative CT imaging and the Otoplan tool predicted that the Flex26 electrode would provide cochlear coverage and full electrode placement in the scala tympani up to 521 Hz. Low-frequency HP was crucial, as we expected potential loss of residual high-frequency hearing due to insertion trauma. However, at the 24-month postoperative follow-up, HP was complete, and at 36 months there was only a slight deterioration, mainly in low-frequency thresholds. High-frequency HP remained intact throughout the 36-month observation period, indicating that full electrode insertion had not affected HP.

In our center, for many years we have preferred the round window approach surgical technique. According to Dhanasingh and Jolly [44], an atraumatic insertion depends on surgical technique, electrode array design, cochlear duct length, anatomical variations, insertion angle, and cochlear height. In a histological analysis of 33 temporal bones using Med-El electrodes, Adunka et al. [45] identified two mechanisms of basal trauma: array buckling and trauma from drilling. They found that the caudal approach (cochleostomy) led to 48% destructive basal trauma, compared to less than 15% with round window insertions. Recent studies have shown growing interest in biomarkers such as impedance telemetry, which measures inner ear events such as impedance spikes [31]. Gu et al. [46] found that electrodes implanted via the round window approach showed significantly lower impedance than those implanted with the traditional cochleostomy method, particularly at the cochlear base. The authors emphasise that the use of an atraumatic surgical approach probably helps in preserving cochlear architecture and preventing cochlear fibrosis.

To our knowledge, this is the first reported case in which pre-operative high-frequency hearing was assessed following cochlear implantation. The study's limitations should be acknowledged, arising from its retrospective design and the analysis being limited to a single patient. Further systematic investigations involving a larger group of patients with this type of partial deafness are needed to better understand the potential for preserving residual hearing following surgery.

Conclusions

Our patient's results demonstrate that preserving functional residual hearing in the basal cochlea is possible after CI surgery, even though this region is most susceptible to insertion trauma. The presence of functional high-frequency hearing should not be a major reason for withholding cochlear implantation, especially if a hearing aid is ineffective. Larger studies are needed to assess the potential for expanding CI candidacy in patients with this type of partial deafness.

Funding

This work was carried out as part of the research project No. 2024/ABM/03/KPO/KPOD.07.07-IW.07-0221/24-00, funded by the Medical Research Agency, Poland.

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